The ability to operate successfully in the posterior fossa requires a thorough understanding of its neuroanatomy and physiology, accurate localization of lesions, and optimal surgical technique. Principles of Posterior Fossa Surgery provides an in-depth review of this complex surgical region with detailed coverage of anatomy, pathology, imaging, disease-based management, and surgical approaches. Written by a team of highly respected specialists, it will be a valued reference and refresher for clinicians who perform posterior fossa surgery and for trainees.

Features:
• Begins with a useful framework in neuroimaging, neuropathology, and microsurgical anatomy of the posterior cranial fossa
• Covers a wide range of approaches and pathologies in the region, including congenital Chiari malformations, infections, trauma, aneurysms, and tumors
• Highlights the anatomy of common surgical approaches, with numerous radiographic and endoscopic images that aid in visualizing concepts
• Provides full coverage of surgical techniques, starting with basic concepts and progressing to operations on more challenging entities like petroclival meningiomas, jugular bulb tumors, acoustic neuromas, complex basilar aneurysms, and posterior circulation aneurysms
• Includes comprehensive sections on surgical management of pediatric posterior fossa tumors and shunt surgery for lesions
• Shares the insights of prominent neurosurgeons from top centers around the world, who discuss their preferred strategies for tackling this challenging area of the brain

Focusing solely on the posterior fossa, this book fills an important gap for neurosurgeons, skull base specialists, and residents and fellows who are training in this anatomically challenging region. It will enrich their understanding and knowledge of the field, expand their surgical armamentarium, and help achieve the most successful clinical outcomes.

Anil Nanda, MD, FACS, is Professor and Chairman, Department of Neurosurgery, Louisiana State University Health Sciences Center, Shreveport, Louisiana.

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Principles of Posterior Fossa Surgery

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Thieme
New York • Stuttgart
Dedication

Matru devo bhava
Pitru devo bhava
Aachaarya devo bhava

Revere your Mother as God
Revere your Father as God
Revere your Teacher as God

Taittiriya Upanishad, 3000 B.C.

To my parents, Dr. and Mrs. Krishan and Uma Nanda
My wife, Laura, Ubi Amor Ibi Fides
My children, Alexander, Christopher, and Mary Catherine
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Foreword

When I first met Anil Nanda, I was impressed by his poise and natural brilliance. I immediately accepted him as a postgraduate fellow in microneurosurgery. Since he began the fellowship, I have never once been disappointed. I am reminded of the anticipation we all have before a trip, educational meeting, or social event. With Dr. Nanda, it is safe to anticipate, for he always produces in a way that exceeds all possible expectations.

And, so it is with Principles of Posterior Fossa Surgery. Nanda, with a select group of colleagues, each an established and knowledgeable leader in a specific area, exposes the “gloomy corner” of neurosurgery. They bring to it the Klieg light of forthright inspection, brilliant evaluation, lucid exposition, and faultless description of the nuances of operative treatment.

Principles of Posterior Fossa Surgery is organized into four logically divided sections. General Considerations contains the background one needs just to think clearly about the region, that is, historical perspectives, anatomy, imaging, pathology, and neurological entities.

Next is a disease-based series of discussions, spanning congenital, neoplastic to vascular, and infection problems. This is followed by details of operative techniques for a diverse group of problems that all neurologists and neurosurgeons see and treat, and must treat well. Finally, the last section contains a brief two-chapter discussion of miscellany important to the surgery of the posterior fossa.

A book in science or medicine is a good book when it is well written, well organized, and informative. It is a great book when it is organized by a master, is written by proficient and articulate leaders in a field, is fully informative, and meets a need. This volume is a great contribution which every neurosurgeon, neurologist, and otolaryngologist should read and have on the bookshelf.

Peter J. Jannetta, MD
I am delighted to welcome you to a journey in the posterior fossa. Historically, Harvey Cushing labeled this as the “gloomy corner” of neurological surgery because the morbidity and mortality in the posterior fossa and cerebellopontine angle was dismal. So much so that Cushing, who was a quintessential historian, called the cerebellar pontine angle “the fence corner of the Gettysburg Battlefield—the bloody angle.” Much has changed over the past few decades. This text is an attempt to focus on a difficult terrain of neurological surgery with a wide exposure on multiple facets. Currently, most textbooks in neurological surgery focus on supratentorial surgery with some attention to the posterior fossa, but we have tried to elevate this entire area and look at its multiple aspects from different angles. Much has changed since Harvey Cushing’s day, but his enviable drop in mortality has not been matched; although developments in microsurgery and technology have helped considerably.

As a resident, I was always intimidated by the posterior fossa. With apologies to Churchill, to me it was a riddle wrapped in a mystery inside an enigma. After I watched Professor Peter Jannetta waltz through the posterior fossa and saw how effortless he made it, my fear lessened. Technical maestro that he is, he always made it alarmingly simple. Minor technical tips like opening the cisterna magna, planning your bone flap, and meticulous closure have helped the field considerably, as well as advances in skull base surgeries. It was with this in mind that I decided to focus purely on the posterior fossa and fill a niche for an area that is sometimes neglected.

This book begins with a historical perspective, and then a detailed anatomical description, followed by radiological and neuropathological chapters. After a chapter on the neurological manifestation of disease in the posterior fossa, there are five chapters on disease-based management that include congenital malformations such as Chiari, trauma, and infections, and vascular entities, like hemangiomas and dural fistulas.

Attention then shifts to pure surgical technique, and basic concepts in posterior fossa surgery are followed by more difficult entities, such as petroclival meningiomas, jugular bulb tumors, and acoustic neuromas, as well as complex basilar aneurysms and posterior circulation aneurysms. The book concludes with a chapter on pediatric tumors and shunt management. There has been some duplication to provide novel approaches from different surgeons, such as cavernous angiomas by both Dr. Roberto Heros and Dr. Robert Spetzler’s group. Hopefully, these chapters will serve as entries to mastery in this anatomically challenging region. I felt that covering more eclectic topics on this terrain would enhance our understanding, enrich our techniques, and result in better clinical outcomes.

I owe an incredible debt of gratitude to Professor Peter Jannetta, who was able to shine a light into this gloomy corner of neurosurgery and make it easy for so many of us. I am also indebted to Dr. Perry Black, my Program Chairman, and many teachers along the way. I would like to acknowledge the help of several individuals in getting this book off the ground. Numerous authors have contributed and enriched the fabric of this offering, and I owe a debt to all the fellows who have worked in my department over the years, including Dr. Amitabh Chanda, Dr. Vijay Javalkar, Dr. Lissa Baird, Dr. Anirban Banerjee, and Dr. Haim Ezer, as well as all of our neurosurgical residents. I am also grateful to my editorial assistants, Latisia Mills and Lauren Sowell, as well as Lauren Henry and Kay Conerly from Thieme Publications, who were a pleasure to work with.

Lastly, I am deeply indebted to my wife, for supporting me during this project and my children who were always kidding me as to when the book would finally be done. As America’s poet, Mary Oliver, said “Every day I walk out into the world to be dazzled, then to be reflective.” I hope this modest effort is surgically challenging, but in the end contemplative enough to result in a deeper understanding of a difficult terrain.
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General Considerations
1

Surgery of the Posterior Cranial Fossa: Historical Aspects

Lissa C. Baird and Anil Nanda

The earliest known surgeries performed on the cranium date as far back as prehistoric times. The vast majority of these, however, were performed on the cranial convexity. The history of posterior fossa operations, as compared with the entire history of neurosurgical procedures, is relatively brief. This is not surprising, considering the vulnerability of the vital neural structures found in the posterior fossa. Surgical manipulation of the cerebellum, brainstem, and cranial nerves resulted in forbiddingly high mortality prior to the sophisticated techniques and operative environment available in the modern neurosurgical era. Even with the diminished risk associated with contemporary technological advancements, surgical procedures involving the posterior fossa continue to carry higher morbidity than elsewhere in the central nervous system.

The first successful surgeries in the posterior fossa occurred at the end of the 19th century and involved drainage of cerebellar abscesses through trephine openings behind the mastoid process.1,2 In 1893 Charles McBurney, an American surgeon best known for the eponymic landmark used in diagnosing appendicitis, reported the first successful removal of a cerebellar tumor.1,3 Surgical treatment of lesions in the posterior fossa has expanded since that time to include a wide spectrum of pathologies.

The ability to successfully operate in the posterior fossa resulted from a detailed understanding of the neuroanatomy and physiology, accurate localization of lesions, improved surgical techniques, and the development of antisepsis and anesthesia. Today’s neurosurgeon is indebted to the numerous individuals who contributed to these elements.

◆ Galen

The first recorded technique for a posterior fossa approach can be found in the Anatomic Procedures written by Claudius Galen in AD 177.4 Galen (AD 129–200) was born in the cultural and intellectual Hellenistic center of Pergamon (Fig. 1.1). His induction into medicine has mythological origins that tell of Aesculapius, the Greek god of medicine and son of Apollo, inspiring Galen’s future career in a dream his father had. Galen went on to become the most prominent physician of his time, holding positions that included personal physician to Marcus Aurelius and his son Commodus, and surgeon to the gladiators. He made extensive contributions to the understanding of neuroanatomy, including the first recorded attempt at identifying and numbering the cranial nerves.5

Human cadaveric dissections were forbidden in the Roman Empire during the 2nd century AD, leading Galen to develop his approach to the fourth ventricle through live animal dissections.4 He chose monkeys as his primary subjects, not only for their assumed anatomic similarity to humans, but to avoid the inconvenience of having the animal “shout at the top of their lungs” during the dissection, as tended to occur with pigs and goats.4,6 Galen’s efforts were primarily for educational purposes, and there is no conclusive evidence that he ever attempted this approach in a human.4 Regardless, the methodology he developed for approaching the cerebellum and fourth ventricle is remarkably similar to the standard approach used 1700 years later by contemporary neurosurgeons.

Galen recommended making a straight incision through skin and muscle in the midline of the occipital region. Blood loss was controlled by distracting and elevating the edges of the wound, and hooks were placed to keep the tissue spread apart.4,6 Following removal of the periosteum, he described a suboccipital craniectomy using special chisels to protect the dura mater. He specifically suggested to “take care not to tear the thick membrane” when removing bone, and to extend the craniectomy “until the suture shaped as the Greek letter lambda.”4,6 When explaining the dural opening, Galen emphasized elevation of the dural edges to avoid injury to the underlying cortex. The vermis was then divided to expose
I General Considerations

the fourth ventricle. Galen found in his investigations that in order for the animal to survive and remain conscious, only a small incision through the vermis could be made, as an extended division would result in irreversible impairment of consciousness.4

Much of the intracranial work done by Galen was described only in Arabic and thus was not available to the anatomists of the Renaissance.4 The precocious nature of his work is demonstrated by the superior accuracy of his descriptions of fourth ventricular anatomy to those made during the 16th century, as well as through a comparison of his surgical approach with the technique described in Charles Frazier’s 1926 publication entitled The Midline Bloodless Approach to the Posterior Fossa.7

Galen’s approach to his own education as a physician remains a relevant example for modern-day students of neurosurgery. He emphasized the importance of learning anatomy through dissections of dead animals prior to attempting surgery on living animals, and his anatomic studies were undertaken with the motivation to improve his abilities as a surgeon and physician.4

◆ Advances in Neuroanatomy and Neurophysiology

The 16th century encompassed pivotal events and key individuals whose contributions laid the foundation for modern neurosurgery. The passion for new knowledge that infused this period of time and the subsequent explosion of new discoveries marked this era as the beginning of the modern anatomic period.5 Physicians of this era began to connect disease to anatomic structure for the first time, and the desire to better understand the human body led to the reintroduction of cadaveric dissections.8 Exploration of the intricate anatomy of the posterior fossa was no exception.

Leonardo da Vinci (1452–1519) is recognized as the founder of iconographic and physiologic anatomy.8,9 He conducted numerous anatomic studies on the brain and ventricular system and provided the first diagrams of the cranial nerves (Fig. 1.2). His endeavors laid the groundwork for subsequent anatomists to expand the knowledge of exact neuroanatomic relationships.10,11

The first illustration of the posterior fossa can be found among the drawings of Johannes Dryander (1500–1560), a professor of surgery in Marburg (Fig. 1.3). In 1536 Dryander published what can be considered the first textbook of neuroanatomy, containing 16 plates of his dissections of the brain.12 He was also one of the first anatomists to perform public dissections of the skull, brain, and dura mater.8,13

The most well known anatomist of the period was the surgeon Andreas Vesalius (1514–1564). Vesalius made fundamental contributions to the field of medicine through his efforts to provide the most accurate descriptions possible of the human body.14 He emphasized the importance of learning anatomy alongside a cadaver and directly applied the discoveries of his own dissections to his surgical practice.8 For his studies of the brain he noted that “heads of beheaded men are the most suitable since they can be obtained immediately

Fig. 1.1 Claudius Galen (AD 129–200). (From Wellcome Institute Library, London.)
Fig. 1.2  Leonardo da Vinci’s drawings of the skull, brain, ventricular system, and cranial nerves. His drawings provided the first diagrams of the cranial nerves.
after execution with the friendly help of judges and pre-
fects.8,15,16 Vesalius drew exceptional illustrations of the
brain, which included detailed dissections of the brainstem
and cranial nerves (Fig. 1.4). Two centuries later, Samuel
Thomas von Soemmerring (1755–1830), a German physician
and anatomist, would be the first to accurately describe all
12 cranial nerves. His drawings of the posterior fossa struc-
tures were the first to incorporate almost the same degree of
detail as seen in modern-day illustrations (Fig. 1.5).8

The work of these and other key Renaissance figures created
a basic foundation for the intense neuroanatomic investiga-
tions that followed during the subsequent period of scientific
growth. Contemporary physicians began to appreciate
the value that accurate understanding of anatomy held for the
success of their surgical attempts. The expanded knowledge
of brain anatomy, combined with more sophisticated surgical
tools and generally improved medical conditions, enabled
surgeons to consider attempting operations on the brain.8

Inquiry into the physiologic function of nervous structures
followed their anatomic descriptions. Some of the first ob-
servations on the cerebellum were made by Thomas Willis
(1621–1675). Willis, along with his study of what became
known as the circle of Willis, improved the classification of
the cranial nerves (he recognized nine), and he published in
1664 the most accurate text of neuroanatomy to date, Cerebri
Anatomie (Fig. 1.6).5,8,17 In his text, Willis suggested that the
cerebellum regulated involuntary movements.1,5 This theory
was later confirmed by the Italian physician Luigi Rolando
(1773–1831) in 1809 with his experiments demonstrating
cerebellar control over movement. Marie Jean Pierre Flourens
(1794–1867) in 1824 restricted the functions of the cerebel-
lum to the coordination of movement with his studies in-
volving cerebellar ablations in animals.1 In the first American
textbook of neurology published in 1881, William Hammond
(1828–1900) described symptoms associated with cerebellar
tumors including headache, vomiting, staggering gait, and
convulsions.1,18 By the beginning of the 20th century, the
classic findings of cerebellar tumors (ataxia, headaches, vom-
itating, papilledema, and hydrocephalus) were common knowl-
gedge among contemporary neurologists, leading to more ac-
The ability to correlate a functional role with a neuroanatomic structure enabled surgeons to localize lesions based on the patient’s symptomatology. Walter Dandy’s development of ventriculography in 1918 went on to supplement the physical examination for localization of posterior fossa tumors. Elucidation of the intricate functions of the brainstem began with the work of Julien Le Gallois (1770–1814), who in 1812 discovered the vital respiratory centers in the medulla. Subsequent investigators, including Thomas Lumsden and Sir Charles Sherrington, went on to describe these brainstem centers in more detail. Knowledge of the neurophysiologic functions of the structures of the posterior fossa was vital for the surgeon attempting a surgical approach to the area. Although the cerebellum held an important role in the fine regulation of motor activity, its function was not pertinent to life. Conversely, the vital centers contained in the lower brainstem structures had to be handled extremely delicately.

Louis Sebastian Saucerotte (1741–1814) was one of the first surgeons to realize that certain areas of the brain were far more susceptible to injury than others. As a surgeon in the French Army, he was exposed to a myriad of head injuries, and eventually published a review that gave a detailed description of the symptoms associated with certain brain injuries. He was the first to describe loss of coordination with opisthotonos and eye rolling from a cerebellar lesion. He divided the brain into areas of susceptibility, and noted that the structures of the posterior fossa—the brainstem and cerebellum—were the regions associated with the most severe injuries. Insight into neurologic function rapidly expanded after investigators realized that the brain did not function as a single unit and that function could be localized. G.T. Fritsch (1838–1927), E. Hitzig (1838–1907), and Paul Broca (1824–1880) each contributed to the concept that each part of the brain correlated with a particular function. Robert Bartholow (1831–1904), an American physician working in Ohio, reported in 1868 his observations in three patients with brain tumors who had clinical symptoms that correlated with his anatomic findings.

Over the course of the 18th and 19th centuries, the concept of operating in the posterior fossa gained acceptance. Percival Pott (1714–1788), described as the first modern neurosurgeon, was a strong proponent of intervention in brain injury. He felt that infections creating compression of the brain should be removed, including in the region of the posterior fossa. Surgeons became increasingly comfortable with their knowledge of brain anatomy and function. Symptoms of central nervous system pathology were recognized as such, leading to more aggressive clinical management. The addition of antisepsis, anesthesia, and improved techniques for cerebral localization then set the stage for the more complicated neurosurgical approaches that would be developed during the 19th century.

Cerebellar Surgery

Walter Dandy (1886–1946) made the observation that “surgery of the brain is the outgrowth of three discoveries of the nineteenth century, namely, anesthesia, asepsis and cerebral localization.” The progress in these areas of neurosurgery led to more routine attempts to treat pathology of the posterior fossa; however, the initial associated surgical mortality was shockingly high. Herman Oppenheim (1858–1919) reported a mortality rate of 71% in patients treated for cerebellar tumors. Henri Duret (1849–1921) reported a slightly lower rate of 60% in 1903, and Charles Frazier (1870–1936) reported 42% in 1905. The lack of understanding of cerebrospinal fluid (CSF) dynamics along with inadequate intraoperative supportive measures such as intravenous fluids and blood transfusion likely contributed to these initial poor results. It is obvious that these limitations made the duration of the surgery a critical factor in patient survival, and it is not surprising that tumors were only partially resected in these early cases.

Neurologic surgery did not become its own specialty until the end of the 19th century. Prior to that time the operative procedures on the brain were undertaken by general surgeons. There were many talented surgeons during this early modern era who played pivotal roles in the progress that would be made in the surgical treatment of neurologic pathology. They focused their creative efforts on developing new techniques that would push the field forward.

Positioning of the patient in a manner that would facilitate suitable access to the posterior fossa presented one challenge. Victor Horsley (1857–1916) and Fedor Krause (1857–1937)
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recommended a lateral position on the operating table, whereas Harvey Cushing (1869–1939) and Frazier each developed their own specialized head rests to allow the patient to be placed in the prone position. Although Cushing also endorsed some elevation of the head to reduce venous pressure, French surgeon Thierry de Martel (1875–1940) introduced the sitting position and designed a special chair for use during his cerebellar surgeries (Fig. 1.7). The incisions and muscle flaps created to approach the cerebellum varied significantly and changed a great deal before evolving into the straight linear incision that is most commonly used today. Krause, considered to be the father of German neurosurgery, was the first to publish detailed techniques for approaches to the posterior fossa. He used a unilateral or, when necessary, bilateral horseshoe flap that he reflected inferiorly (Fig. 1.8). Horsley used a curved incision from mastoid to mastoid with the apex above the inion. Similarly, Charles Ballance (1856–1936) used an upward-curving incision from mastoid to midline for a unilateral exposure. Cushing devised his “cross-bow” incision in 1905 in an effort to improve exposure. Starting with a curved transverse incision between the mastoids, he added a midline extension down to the lower cervical spine. Few other surgeons adopted this approach because of the additional time required to close the wound. In 1926 Frazier described his “midline bloodless approach to the posterior fossa.” The incisions used prior to that time caused inconvenient bleeding when cutting through the occipital arteries and veins as they crossed the midline. Frazier’s straight vertical incision avoided this problem with a smaller extension from 2 cm above the inion to the upper cervical region. He made a small (2 cm) transverse extension of the incision at the superior apex to facilitate retraction and provide a site for trephine opening and ventricular puncture should it be necessary. Howard Naffziger described an approach to expose the anterior cerebellum in 1928 involving opening the tentorium to the incisura tentorii while retracting the occipital lobe. Naffziger also used Frazier’s midline incision to approach midline cerebellar lesions (although he eliminated the small transverse extensions at the inion) and “hockey-stick” incisions extending from the tip of the mastoid to the inion and then vertically down the midline when approaching one cerebellar hemisphere (Fig. 1.9). Alfred Adson (1887–1951) described in 1941 the use of a straight vertical incision over the lateral suboccipital regions for unilateral cerebellar exposure, but the upward-curving mastoid-to-mastoid lateral incision seems to be the most generally popular incision used by surgeons of this era. Dandy favored this incision in a large series of cerebellar surgeries he described in 1932, and Paul Bucy (1904–1992), William Cone (1897–1959), and Wilder Penfield (1891–1976) also used this incision routinely.

Given the thickness of muscle in the occipital area, most surgeons were not hesitant to perform suboccipital craniectomies, as there would be no deforming defect. Although many replaced the bone just as they would at the cerebral convexity, they realized early on that discarding the bone
Fig. 1.8  Krause’s approach to the cerebellopontine angle using a hinged horseshoe osteoplastic flap.

Fig. 1.9  Illustrations demonstrating the midline (A) and hemispheric (B) approaches to the posterior fossa used by Naffziger.
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Cerebellopontine Angle Tumors

The Scottish surgeon Sir Charles Bell (1774–1842) described one of the first cases of a cerebellopontine angle (CPA) tumor in 1833 after diagnosing the lesion at autopsy. The first attempts to resect tumors in this area began at the end of the 19th century when surgeons attempted to remove vestibular schwannomas and meningiomas. The first report of a successful operation on a tumor of the CPA is credited to Ballance in 1894 after he removed an acoustic neuroma in a 49-year-old woman who had presented with headache, vertigo, and tinnitus. Unsurprisingly, the initial attempts to approach the CPA met more commonly with failure than success. Moritz Borchardt in 1906 reported 18 cases with 13 deaths. Several years later in 1913 Anton von Eiselsberg reported 16 cases with 12 deaths, and Krause reported 31 cases with 26 deaths. In 1917, Cushing reported a much lower operative mortality of 15% with CPA tumors, likely due to his meticulous surgical methods. Many of the approaches used to access the CPA were quickly found to be unsuccessful. One example is the translabyrinthine approach suggested by R. Panse in 1904. Despite the successful use of this approach today, difficulty in removing larger tumors without traumatizing the surrounding structures along with the frequency of postoperative CSF leakage quickly made this approach unpopular. Charles Elsberg, Borchardt, and von Eiselsberg all attempted a combined suboccipital and petrosal approach without success. Frazier’s approach of choice involved a unilateral suboccipital craniectomy combined with an occipital bone flap that extended to the lateral sinus.

Cushing not only was a talented surgeon with meticulous technique but also was responsible for many of the most important contributions to his field. Cushing realized that none of the approaches attempted by his predecessors had provided sufficient working space. This combined with the common technique of finger enucleation led to mechanical trauma of the posterior fossa structures. His approach utilized the crossbow incision, bilateral craniectomies, and drainage of CSF via tapping of the lateral ventricle. He opened the dura in a stellate fashion and retracted the cerebellum medially to expose the CPA tumor. His attempts to lower the mortality rate led him to forgo a complete resection, instead debulking the tumor through a capsular incision and leaving the capsule behind. For the larger acoustic tumors he resected a portion of the cerebellar hemisphere to obtain adequate exposure, a technique he referred to as “cerebellar uncapping.” Dandy went on to improve on Cushing’s technique, eventually favoring a smaller unilateral flap and craniectomy in the lateral position. Dandy trained under Cushing and made some of the most innovative and influential contributions seen in neurosurgery. The methods he developed in his approach included enucleation of the tumor from within the capsule followed by complete removal of the capsule by carefully dissecting it away from its attachments. He reported five such operations in 1934, with all five patients surviving. (Fig. 1.10). Cushing and Dandy notoriously clashed over their differing approaches to treating acoustic neuromas. When Dandy neglected to mention Cushing’s work in an article describing his own technique for the Johns Hopkins Hospital Bulletin, Cushing was incensed, and wrote to Dandy, “You must not forget your manners, and this last note of yours is in extremely bad taste.”

The different approaches to these tumors created some controversy in the field as surgeons attempted to weigh the consequences of facial paralysis, tumor recurrence, and surgical mortality to determine the best method for operative treatment. In general, facial paralysis was felt to be preferable to tumor recurrence, as reoperations carried even greater mortality risk. Surgeons did not begin attempting to spare the facial nerve during complete resections until Hugh Cairns reported such a success in 1931. With the refinement of microsurgical techniques and addition of facial nerve monitoring, the preservation of facial nerve function today has dramatically improved.

Conclusion

Considerable progress has been made in the surgical management of posterior fossa lesions. The advances in anesthesia,
Antisepsis, and critical care have had a major impact on operative mortality. Introducing the operating microscope to the field of neurosurgery in the 1950s enabled revolutionary advances in technique to be made, especially when operating on the small and sensitive structures found in the posterior fossa. Stereotactic radiosurgery has demonstrated excellent results for many pathologies, and has become an important adjunct for the treatment of posterior fossa lesions, especially those that carry a high risk of morbidity with surgical treatment. The modern-day neurosurgeon has an excellent understanding of the complex anatomy of the posterior fossa, diagnostic modalities that facilitate the exact localization of lesions, and technological tools to supplement surgical skills. All of these advances have resulted in a dramatic reduction in morbidity and mortality. The ongoing evolution of treatment modalities and surgical techniques will continue to improve the care of patients with posterior fossa pathology.

References

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Microsurgical Anatomy of the Posterior Cranial Fossa

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The posterior cranial fossa is unique in the cranial cavity, as it is the largest of the three cranial fossae and has the most complex intracranial anatomy. It harbors the parts of the brain that control respiration, cardiac cycle, consciousness, and balance. It is the conduit of fibers of practically all important motor and sensory pathways. Here lies the conduit in which the cerebrospinal fluid (CSF) emerges from the ventricular system into the subarachnoid space. Moreover, the complex arterial relationships of the vertebrobasilar system make this region even more complex. In addition to discussing surgical anatomy, this chapter highlights the anatomy of common surgical approaches to this region, and provides illustrations of radiologic anatomy and some endoscopic anatomy (Figs. 2.1, 2.2, 2.3, and 2.4), which is essential for a neurosurgeon in practice or in training.

◆ Panoramic View of the Posterior Fossa

Superiorly, the posterior fossa starts from the tentorial hiatus and extends down to the foramen magnum. It is encased by occipital, temporal, and sphenoid bones, with a small contribution from parietal bone. In front of it is the dorsum sellae and the clival part of the occipital bone; behind it is the squamous part of the occipital bone, and on both sides are the petrous and mastoid part of the temporal bone along with the lateral part of the squamous part of the occipital bone. The upper peripheral part is bounded by the sulcus of the lateral sinus starting from the internal occipital protuberance and extending anterolaterally to the posterior part of the petrous ridge. The boundary continues along the petrous ridge to end at the petroclinoid junction (Fig. 2.5). We will discuss the bony architecture as well as the neural structures and vessels.

◆ Bony Anatomy

Foramen Magnum and Its Adjacent Region

When the posterior cranial fossa is viewed from above or below in a dry skull specimen, the foramen magnum is the most prominent landmark that comes into view. This oval-shaped foramen is wider posteriorly, and this posterior part transmits the medulla. The narrower anterior part sits over the odontoid peg.

The posterior part of the foramen magnum is contributed to by the squamous part of the occipital bone, which is an internally concave plate. This part not only forms the posterior boundary but also continues above the foramen magnum. The convex external surface has several prominences on which muscles of the neck attach; these muscles include the sternocleidomastoid, trapezius, splenius capitis, semispinalis capitis, rectus capitis posterior major, and superior oblique. The largest prominence, the external occipital protuberance or inion, is located about a centimeter below the apex of the internal occipital protuberance and is slightly below the torcular Herophili. This is an important landmark for performing a craniotomy in the posterior fossa exposure. The inner surface has the internal occipital protuberance near its center. The internal surface is divided into four somewhat unequal parts by bony ridges. The lower two parts are bounded medially by the internal occipital crest (a prominent ridge descending from the internal occipital protuberance, which gives attachment to falx cerebelli) and above by the paired sulci for lateral sinuses that extend laterally from the internal occipital protuberance. The sulcus for the right lateral sinus is usually larger (Fig. 2.5). The internal occipital crest bifurcates above the foramen magnum to form paired lower limbs, between which there is a depression called the vermis fossa, which is occupied by the lower end of the vermis of the cerebellum.
Fig. 2.1 Bone window of computed tomography (CT) scan showing the crucial anatomy of the posterior fossa. (A) Axial section of the posterior fossa showing the relationship of the carotid canal and the jugular foramina. (B) Section through the temporal bone showing the external ear, middle ear, and eustachian tube. Note the relationship of the eustachian tube and the carotid canal. (C) Another section through the temporal bone showing the bony labyrinth and the internal auditory canal. (D) A section through the posterior semicircular canal. Note its proximity to the posterior surface of the petrous bone. (E) A section through the superior semicircular canal.
Fig. 2.2  Magnetic resonance imaging (MRI) scan showing important anatomy of the posterior fossa. (A) Sagittal T1 MRI scan showing the brainstem, fourth ventricle, and cerebellar tonsil. (B) Axial section through lower medulla showing the cisterna magna and the tonsil of the cerebellum. (C) Axial section a little above the previous section showing parts of the medulla, uvula, and tonsil of cerebellum. (D) Axial section through the midpons.
The anterior part of the foramen magnum is formed by the basilar part of the occipital bone or the clivus, which is a thick quadrangular plate of bone that extends forward and upward at an angle of 45 degrees from the foramen magnum. If traced upward, the sphenoid-occipital synchondrosis (where the occipital bone joins the sphenoid bone) is found just below the dorsum sellae. The dorsal part of the clivus is concave from side to side, and laterally it is bounded by the petroclival fissure (which separates the clivus from the petrous temporal bone) on both sides (Fig. 2.6). The petroclival fissure harbors the inferior petrosal sinus, and if traced back, it ends in the jugular foramen. On the ventral surface of the clivus just in front of the foramen magnum there is a small elevation called the pharyngeal tubercle (Fig. 2.6), which gives attachment to the pharyngeal raphe.

Laterally, the foramen magnum is bounded by the condylar parts of the occipital bone. The most prominent part of the condylar part is the paired occipital condyles, which are oval in shape and convex downward; they face downward and laterally, with the long axes in the anteromedial direction, and they are located lateral to the anterior half of the foramen magnum and protrude from the condylar part of the occipital bone. They articulate with the lateral masses of the atlas. On the external surface behind the condyle there is the condylar fossa (Fig. 2.7), a depression that is often perforated to form the condylar canal transmitting the emissary veins. During the transcondylar approach, these veins can cause troublesome bleeding. From the posterior half of the condyle, the jugular process, a quadrilateral plate of bone extends laterally to form the posterior border of the jugular foramen. On the medial side of each condyle there is a tubercle, which gives attachment to the alar ligament of the odontoid process. The jugular tubercle, an oval prominence on the intracranial surface of the condylar part, sits just above the hypoglossal canal and medial to the lower extent of the petroclival fissure (Fig. 2.6).

**Jugular Foramen Region**

The jugular foramen, sitting at the posterior end of the petroclival fissure, is situated lateral and slightly superior to the anterior half of the occipital condyle (Fig. 2.8). It is bordered posteriorly by the jugular process of the occipital bone, and
anterosuperiorly by the jugular fossa of the petrous temporal bone. It can be considered a hiatus between the temporal and occipital bones. The right foramen is larger than the left. The long axis of the foramen is directed anteromedially, so that its anterolateral margin is formed by the temporal bone, and the posteromedial margin is formed by the occipital bone. It is directed anteromedially and downward from the intracranial aspect. The foramen cannot be seen when the base of the skull is viewed directly from above or below. It has a large oval component laterally, where it receives the drainage of the sigmoid sinus, and a smaller medial part or petrosal part, which receives the drainage of the inferior petrosal sinus (Fig. 2.8A). If the foramen is viewed from below, the part of the jugular foramen that is seen is the part on which sits the jugular bulb. The intrajugular processes on the opposing surfaces of the occipital and temporal bones are connected by a fibrous strand, called the intrajugular septum. This septum, which can sometimes be bony, separates the petrosal and sigmoid part of the foramen. The intrajugular ridge extends forward from the intrajugular process of the temporal bone along the medial edge of the jugular bulb (Fig. 2.9B). The glossopharyngeal nerve courses along the medial edge of this

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**Fig. 2.3** Demonstration of endoscopic anatomy in a cadaver after a third ventriculostomy. The endoscope was navigated through the preoptic cistern and below. (A) Oculomotor nerve on the left side is seen. (B) Acousticofacial bundle (AFB) is seen. (C) Abducens is seen entering Dorello’s canal. CN, cranial nerve. (D) Lower cranial nerves.
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ridge. Sometimes a canal is formed through which the glosso-pharyngeal nerve courses.

The sigmoid sinus courses forward into the sigmoid portion of the jugular foramen. In this foramen there is a high domed recess, called the jugular fossa, which forms the roof over the jugular bulb and is usually larger on the right side due to the larger size of the jugular bulb on the right side. On the medial side of the intrajugular process of the temporal bone, a small triangular recess, called the pyramidal fossa, extends forward along the anterior wall of the petrosal part.

Fig. 2.4 The endoscope was navigated back from the third ventricle to guide through the aqueduct into the fourth ventricle. (A) The roof of third ventricle is clearly seen, along with the aqueduct and the suprapineal recess. (B) Entry into the fourth ventricle.

Fig. 2.5 A panoramic view of the bony posterior cranial fossa from above.

Fig. 2.6 Foramen magnum and the adjacent region from above.
of the foramen. The external aperture of the *cochlear canaliculus*, housing the perilymphatic duct and a tubular prolongation of the dura mater, opens into the anterior apex of the pyramidal fossa (Fig. 2.9A). The glossopharyngeal nerve enters this fossa below the point at which the cochlear aqueduct joins its apex.

The *jugular process of the occipital bone*, forming the posteromedial wall of the foramen, extends laterally from the area above the posterior half of the occipital condyle. It is penetrated by the hypoglossal canal. An oval prominence, the *jugular tubercle*, is found on the superior surface of the intrajugular process and is exactly above the hypoglossal canal (Fig. 2.9A). Occasionally, the jugular tubercle is marked by a shallow furrow indicating the passage of the glossopharyngeal, vagus, and accessory nerves.

The *hypoglossal canal* passes through the condylar part of the occipital bone and is located medial to the jugular foramen just below the jugular tubercle and approximately 5 mm
inferomedial to the petrosal part of the jugular foramen (Figs. 2.8A and 2.9A).

Just inside the lateral edge of the jugular foramen there is the mastoid canaliculus, through which the auricular branch of the vagus (Arnold’s nerve) enters the mastoid. The nerve exits the bone in the inferolateral part of the tympanomastoid suture. A small canal, the tympanic canaliculus, is located at the site where the intrajugular ridge of the temporal bone meets the carotid ridge, which separates the carotid canal and jugular foramen, and is directed upward. The tympanic branch of the inferior glossopharyngeal ganglion (Jacobson’s nerve) courses through this area to the tympanic cavity (Fig. 2.9B).

**Petrosal Temporal Bone and the Adjoining Region**

The petrosal temporal bone is probably the most important bone for neurosurgeons. Its intricate anatomy must be learned in detail before venturing into surgery of this region. As many as seven cranial nerves are related to this bone. The internal carotid artery has a sizable length in this bone. The petrous part of the temporal bone is a pyramid-shaped with a base, an apex, three surfaces, and margins, and is wedged between the sphenoid and occipital bones. The anterior or superior surface faces the middle cranial fossa and is not discussed here.

The posterior surface of the petrous bone is continuous with the mastoid surface and faces the cerebellopontine angle region. The internal auditory meatus (IAM) is located midway between the base and apex on the posterior surface (Figs. 2.8A and 2.9A). The lateral end of the internal auditory ends in the meatal fundus, which is divided into superior and inferior parts by the transverse crest. The superior part is occupied by the facial nerve anteriorly and the superior vestibular nerve posteriorly. The inferior part is occupied by the cochlear nerve anteriorly and inferiorly. The superior part is incompletely divided by another bony bar, called the vertical crest or Bill’s bar (Fig. 2.10A), which separates the anteriorly located facial canal from the posteriorly located superior vestibular nerve. Posterior to the entry of the inferior vestibular nerve into the semicircular canal, in the posteroinferior corner of the meatal fundus, there is the singular foramen for the singular branch of the inferior vestibular nerve, innervating the posterior ampullae. Just lateral to the porus acusticus on the posterior surface of the petrous bone is the site of a small bony opening, called the subarcuate fossa, which gives passage to the subarcuate artery, a branch of the anterior inferior cerebellar artery (AICA), which usually ends blindly in the region of the superior semicircular canal. The opening for the vestibular aqueduct transmitting the endolymphatic duct, which opens below into the endolymphatic sac in between the dural layers, is infelateral to the porus (Fig. 2.9A).

Inferior to the porus, at the anteromedial edge of the jugular foramen is the opening of the cochlear aqueduct, occupied by the perilymphatic duct, just superolateral to the entry of the glossopharyngeal nerve into the intrajugular part of the jugular foramen. The superior border of the petrous bone or the petrous ridge, grooved by the superior petrosal sinus (connecting the cavernous sinus and the transverse-sigmoid junction), serves as the attachment to the tentorium cerebelli.

The inferior surface of the petrous temporal bone is rough. The apex is connected to the clivus medially by a fibrocartilage and gives attachment to the levator veli palatini and the cartilaginous portion of the eustachian tube. Just posterior to this there is an opening of the carotid canal, which is bounded posteriorly by the carotid ridge (vide supra). The carotid ridge separates the carotid canal from the jugular foramen (Fig. 2.9B).

The petrous temporal bone contains the carotid canal, bony labyrinth, and tympanic cavity (Fig. 2.10B, C). The mastoid air cells are contained in the mastoid part of the temporal bone. The bony labyrinth has three parts: the vestibule, the semicircular canals, and the cochlea. The vestibule, a small cavity in the center of the bony labyrinth, is located at the confluence of the ampullae and the nonampullated ends of the semicircular canals. Medial to it is the meatal fundus, lateral to it is the tympanic cavity, and anterior to it is the cochlea, and it sits on the bony plate harboring the apex of the jugular bulb. The bony plate separating the vestibule and the apex of the jugular bulb is thicker on the left side (average thickness: 8 mm) than on the right side (average thickness: 6 mm). This thickness is important during the translabyrinthine approach as the height of the jugular bulb is a major factor determining the degree of exposure. Posterosuperior to the vestibule are the semicircular canals (SCCs). The horizontal or lateral semicircular canal is situated above the tympanic segment of the facial nerve and serves as an excellent guide to locating the tympanic segment of the facial nerve. Henle’s spine, located along the posterosuperior edge of the bony external auditory meatus and anteroinferior to the suprameatal triangle, serves as a good landmark for the lateral semicircular canal, which is approximately 1.5 cm deep to this landmark. The canal is deep to the suprameatal triangle. The posterior semicircular canal lies parallel and close to the posterior surface of the petrous bone just behind and lateral to the lateral end of the internal auditory meatus. This anatomy is important in the retrosigmoid transmeatal approach to vestibular schwannoma. While drilling bone of the posterior edge of the internal auditory meatus to expose the internal auditory canal, the surgeon must be careful not to damage the posterior semicircular canal if the patient’s hearing is to be preserved. The superior semicircular canal projects toward the floor of the middle cranial fossa just below the arcuate eminence. Each of these canals has an ampullated and nonampullated end opening into the vestibule. The ampullae are located at the anterior end of the lateral and superior canals and the inferior end of the posterior canal. The posterior ends of the posterior and superior canals join to form the common crus, which opens into the vestibule. The ampullae of the superior and lateral canals are innervated by the superior vestibular nerve. The posterior ampulla of the posterior canal is innervated by a singular branch of the inferior vestibular nerve. The internal auditory canal (IAC) is medial to the arcuate eminence at an angle of approximately 60 degrees medial from the long axis of the superior semicircular canal.
The internal carotid artery (ICA) enters the carotid canal and runs vertically upward. The petrous carotid artery starts at the point of entry of the ICA through the periosteal-lined carotid canal in the petrous bone. The external orifice of the carotid canal is directly anterior to the jugular foramen, and its internal orifice is located at the petrous apex. Except at the entrance of the artery to the vertical canal, where it is anchored to the bone by dense bands, the artery can be easily separated from the connective tissue adhesion. The petrous ICA has two segments, the vertical and the horizontal, and they join at the genu. The vertical segment, as the name implies, passes vertically upward in the carotid canal. It is surrounded by the jugular fossa posteriorly, the eustachian tube (ET) anteriorly, and the tympanic bone anterolaterally. The vertical segment turns anteromedially at the genu to form the horizontal segment, which continues anterolaterally. It runs anterior to the cochlea, from which it is separated by a thin plate of bone. The anteromedial part of the roof of the horizontal part is formed by the dura or a thin plate of bone, which separates the ICA from the trigeminal/gasserian ganglion. There are one to two branches of the ICA in the carotid canal, and there are a few variations. (Interested readers can find further information in the literature.) A periarterial venous plexus, which is an extension of the cavernous sinus, extends around the petrous ICA for a variable distance. In the majority of the cases, it runs along the anterior and inferior side of the artery, and its extension is limited to the horizontal segment in the majority of the cases. This venous plexus lies within the periosteal covering of the canal, and if the periosteum is intact, bleeding can be avoided. Moreover, the ICA is also accompanied by the sympathetic fibers. The ET and the tensor tympani (TT) lie anterior and parallel to the horizontal segment of the petrous ICA, below the floor of the middle cranial fossa (MCF). Usually the TT lies superior to the ET, and, during dissection, exposure of the TT heralds the imminent exposure of the ET. In most patients, a thin plate of bone separates them. But in many patients they may be separated by fibrous tissue only. In turn, the TT muscle is separated from the carotid canal by a plate of bone, which varies in thickness. Sometimes a part, the superior surface of the muscle, is exposed through a bony dehiscence between the carotid canal and the foramen spinosum (FS). The ET usually
is separated from the ICA by a thin layer of bone. The ET crosses the anterolateral aspect of the genu as it exits the middle ear cavity.

The facial nerve in the temporal bone (Fig. 2.10) is divided into four segments: (1) the canalicular segment, located in the IAC, extends from its entry through the IAM up to the meatal fundus; (2) the labyrinthine segment, extending from the meatal fundus to the geniculate ganglion, from where the greater superficial petrosal nerve arises, is situated between the cochlea anteromedially and the semicircular canal posterolaterally; (3) the tympanic segment, running posterolaterally along the medial surface of the tympanic cavity, is situated between the lateral semicircular canal above and the oval window below; and (4) the mastoid segment, which runs vertically from below the midpoint of the lateral semicircular canal, ends by exiting through the stylomastoid foramen. The facial nerve has a very complex course in the temporal bone. The parts of the facial nerve in the temporal bone are part of the IAC, the labyrinthine part, the tympanic part, and the mastoid part. We focus on the labyrinthine part. The canal of Fallopius or the facial canal through the base of the skull is quite long; its course is Z-shaped, threading its way between the labyrinth and the tympanic cavity. This is further complicated by the fact that the segments do not exist in a single plane. The labyrinthine portion begins at the fundus of the IAC, bending forward by approximately 50 degrees the general direction of the IAC. Thus the direction of the canal here is at a right angle to the long axis of the petrous pyramid. It curves forward and inward, skirting the superolateral flank of the basal turn of the cochlea and moving in the direction of the greater superficial petrosal nerve (GSPN). It has a slightly ascending course. Posterolaterally, it comes in close relationship to the superior SCC, where it is sandwiched between the superior SCC and the cochlea, and it is liable to be damaged during drilling of the bone here. The facial nerve now makes a sharp bend posterolaterally, creating the first genu. This is where the geniculate ganglion lies and from here the GSPN arises.

The mastoid part is situated posteriorly in the temporal bone. It projects downward to form the process that is the site of attachment of the sternocleidomastoid, splenius capitis, and longissimus capitis muscles. The lower part medial to the mastoid process is grooved by the digastric notch, to which the posterior belly of the digastric attaches. The posterior border of the mastoid process is perforated by one or more foramina through which an emissary vein to the sigmoid sinus passes. The posterior to the mastoid part is the asterion, which is the confluence of the lambdoid, occipitomastoid, and parietomastoid suture. Deep to this is the lower margin of the junction of the transverse and sigmoid sinuses. The upper edge of the junction is just anterior to the junction of the supramastoid crest and squamosal suture. The medial surface of the mastoid part is grooved by the sigmoid sinus, which represents the posterior boundary of the mastoid cavity. At the level of the petrous ridge the sinus meets the roof of the mastoid cavity. The sinus dural angle, formed by the angle between the superior petrosal and sigmoid sinus and the middle fossa dura, is an important landmark when performing a mastoidectomy (Fig. 2.10C). The sigmoid sinus courses downward, medially and forward, crossing the occipital bone to enter the jugular foramen.

The medial boundary of the mastoid cavity is formed by the otic capsule, which is a block of bone containing the bony labyrinth. The area of the posterior fossa dura that is exposed through the mastoid cavity between the sigmoid sinus, petrosal sinus, otic capsule, and middle fossa dura is called Trautmann's triangle. The size of this triangle determines the amount of exposure in the presigmoid retrolabyrinthine approach. The average distance between the anterior margin of the sigmoid sinus and the otic capsule at the level of the posterior semicircular canal in a cadaveric study was 8 mm on the right side and 7 mm on the left side. Apart from this distance, the distance between the apex of the jugular bulb and the superior petrosal sinus is also an important determinant in this surgical exposure. The exposure is reduced if the jugular bulb is high. The apex of the jugular bulb is usually inferior to the ampulla of the posterior semicircular canal. The average distance from the apex of the jugular bulb to the superior petrosal sinus in a cadaveric study is 14 mm on the right side and 16 mm on the left side. The inside of the mastoid bone is full of air cells, which coalesce to form a cavity called the mastoid antrum, which communicates to the epitympanic part of the tympanic cavity through the aditus. The air cells of the mastoid may extend back behind the sigmoid sinus, up into the squamous part of the temporal bone, anteriorly to the root of the zygomatic process and the floor of the tympanic cavity. The mastoid antrum lies approximately 1.5 cm deep to the suprameatal triangle. The lateral semicircular canal is medial to the epitympanic recess. The posterior semicircular canal faces the medial wall of the antrum. The roof of the antrum is formed by the tegmen in the floor of the middle fossa. The mastoid segment of the facial nerve courses adjacent to the anteroinferior margin of the antrum. The facial nerve has three important sources of blood supply: the AICA provides branches to the proximal segment; the petrosal branch of the middle meningeal artery travels along with the GSPN and supplies the genu, labyrinthine segment, and tympanic segment; and the stylo mastoid branch of the posterior auricular artery supplies the mastoid segment. There is good anastomosis between these supplies, and preservation of any two will result in very minimal dysfunction.

The middle ear cavity or the tympanic cavity (Fig. 2.10B) is a narrow air-filled space between the tympanic membrane and the inner ear. Anteriorly it communicates with the nasopharynx by the ET, and posteriorly it communicates with the mastoid antrum by the aditus. The roof is formed by the tegmen, which separates the tympanic cavity from the middle fossa. The floor separates the cavity from the jugular bulb. The lateral wall is formed by the tympanic membrane and the bony ring to which it attaches. The ring is deficient above, near the openings of the anterior and posterior canaluli of the chorda tympani nerve. The cavity contains the malleus, incus, and stapes. The tympanic cavity opens up into the epitympanic recess containing the heads of the malleus and incus. The medial aspect of the neck of the malleus is crossed by the chorda tympani nerve. The medial wall has the promontory, the oval window, the round window, and the promi-
nence of the facial nerve. The promontory, which is grooved by the tympanic plexus, represents the basal turn of the cochlea. The apex of the cochlea is medial to the tympanic cavity anterior to the promontory. Posterosuperior to the promontory is the oval window, which connects the tympanic cavity with the vestibule and is occupied by the footplate of the stapes. Above the oval window is the facial nerve prominence. Posteroinferior to the oval window and just below the overhanging edge of promontory is the round window.

◆ Neural Structures

The parts of the brain parenchyma occupying the posterior cranial fossa are the midbrain, pons, and medulla, which constitute the brainstem and the cerebellum (Fig. 2.11).

Midbrain

The midbrain is the uppermost part of the brainstem and it traverses the tentorial hiatus. It is divided in the midline into a right half and left half. Anteriorly and posteriorly deep fissures can be appreciated dividing the midbrain into two halves. Anteriorly the fissure is quite deep to form the interpeduncular fossa. Each right and left half is called the cerebral peduncle. From anterior to posterior each peduncle is subdivided into the crus cerebri or basis pedunculi, the tegmentum, and the tectum. The division between the crus cerebri and the tegmentum is demarcated by the substantia nigra in the parenchyma and the lateral mesencephalic sulcus on the surface. The lateral mesencephalic sulcus is an important intraoperative landmark and is recognized by a longitudinal vein, which runs on the sulcus and connects the basal vein of Rosenthal superiorly and the brachial vein inferiorly. The tectum is behind the aqueduct of Sylvius. Superior and inferior colliculi (quadrigeminal plate) are parts of the tectum and are located on the dorsal aspect of the midbrain. The part superior to the superior colliculus can be a route for entry into the cisterna velum interpositum. The oculomotor nerve exits the midbrain medial to the crus cerebri into the interpeduncular fossa. The plane between the crus cerebri and the oculomotor nerve is a potential place of surgical entry into the midbrain.2 The pontomesencephalic sulcus, separating the midbrain from the pons, starts in the depth of the interpeduncular fossa and runs around the inferior margin of the crus cerebri to join the lateral mesencephalic sulcus. The midbrain continues superiorly with the diencephalic structures. Just above the quadrigeminal plate, at the midline, is the pineal gland. Internally at the third ventricular level, structures inside the posterior third ventricle (posterior commissure, pineal gland, and suprapineal recess) are located above the superior colliculi. The velum interpositum, the space located between the superior and inferior membranes of the tela choroidea, is located just above the pineal gland. The internal cerebral vein and vein of Galen are located just above the pineal gland, and the terminal portion of the vein of Rosenthal is superolateral to the pineal gland.

Pons

The pons is below the midbrain and has a prominent anterior surface, which is convex from side to side. Laterally it is continued in the middle cerebellar peduncle on both sides, which consists of transverse crossed fibers from the pons and higher up. In the midline anteriorly there is a sulcus called the basilar sulcus, which lodges the basilar artery trunk. The lateral pontine sulcus is a shallow groove separating the pontine belly and middle cerebellar peduncle. Just lateral to the lateral pontine sulcus emerges the trigeminal nerve. The trigeminal nerve has two roots. The smaller motor root is in the superomedial position at the emergence, and the larger sensory root is inferolateral in position. For practical microneurosurgical purposes, the emergence of the trigeminal nerve is considered a demarcation point between the pons and the middle cerebellar peduncle. In its posterior part the pons forms the upper part of the floor of the fourth ventricle.

Medulla

The medulla is located inferior to the pons and is separated from the pons by the pontomedullary sulcus. On its anterior part are one median and two paramedian fissures. The median fissure is also called the anterior median fissure, which continues inferiorly as the anterior median fissure of the spinal cord. Just lateral to the anterior median fissure there is a pyramid, which is condensation of the corticospinal tract, on each side. The paramedian fissure, also called the anterolateral sulcus or the preolivary sulcus, is continued below as the anterolateral sulcus of the spinal cord. The rootlets of the hypoglossal nerve come out from the preolivary sulcus. At the upper end of the preolivary sulcus, at its junction with the pontomedullary sulcus, the abducens nerve emerges. Lateral to the preolivary sulcus are the olives on each side, behind which the rootlets of the accessory, vagus, and glossopharyngeal
I General Considerations

The cerebellum wraps the brainstem from the posterior and posterolateral aspects (Fig. 2.9). The fourth ventricle, described as a tent-shaped, midline structure, comes in close contact with the vermian components of the cerebellum. The anatomy of the cerebellum and fourth ventricle are discussed together.

The cerebellum has three surfaces: (1) the petrosal surface, related to the petrous part of the temporal bone anterolaterally; (2) the tentorial surface, related to the tentorium cerebelli superiorly and the upper part of the roof of the fourth ventricle inferiorly; and (3) the suboccipital surface, related to the squamous part of the occipital bone inferiorly and the inferior part of the roof of the fourth ventricle anteriorly.

The suboccipital surface of the cerebellum is located below the lateral sinus and faces inferiorly. In the midline there is the posterior cerebellar incisura and just lateral to that, on two sides, are the velohemispheric or paravermian fissures separating the inferior vermis and cerebellar hemispheres. The parts of the inferior vermis and the corresponding parts of the cerebellar vermis are listed in Table 2.1. The great horizontal fissure starts in the midline between the folium and tuber, and runs laterally between the superior and inferior semilunar lobule. Traced laterally it continues onto the petrosal surface as the petrosal fissure. The secondary fissure is located between the tonsil and biventral lobule. The tonsils are two kidney-shaped structures and are hemispheric components of the uvula (Table 2.1). They are attached to the cerebellum through the superolaterally located peduncles of the tonsil (Fig. 2.12). The tonsil is a relatively free structure, and the superior, anterior, medial, posterior, and most of its lateral surfaces can be separated easily from the surrounding structures. Between its superior pole and the inferior medulary velum is the supratonsillar space; in between the two tonsils is the vallecula; between the anterior surface of the tonsil and the medullar surface is the cerebellomedullary fissure. The cerebellomedullary fissure is one of the routes into the fourth ventricle, instead of splitting the vermis.

The floor of the fourth ventricle (Fig. 2.13) is rhomboid-shaped and presents a strip between the lower margin of the cerebellar peduncles and the site of attachment of the tela choroidea. This strip, called the junctional part, is characterized by the medullary striae that extend into the lateral recesses. The junctional part divides the floor of the fourth ventricle into two unequal triangular parts. The larger superior part, with the apex directed toward the aqueduct, is the pontine part, and the smaller inferior part, with the apex directed toward the obex, is the medullary part. The floor is divided longitudinally into two equal halves by the median sulcus. Each half is again divided into two parts by the sulcus limitans and the sulcus limitans, which is discontinuous at the junctional area. The medial part is the median eminence and the lateral part is the vestibular area. The sulcus limitans demarcates the motor and sensory areas. The motor nuclei of the cranial nerves are located medial to the sulcus limitans, and the sensory nuclei are located lateral to the sulcus limitans. In the pontine part on the median eminence on each

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Table 2.1 Parts of the Cerebellar Vermis and Corresponding Parts of the Cerebellar Hemisphere (Moving from Top to Bottom)

<table>
<thead>
<tr>
<th>Part of Vermis</th>
<th>Corresponding Part of Cerebellar Hemisphere</th>
</tr>
</thead>
<tbody>
<tr>
<td>Superior vermis</td>
<td>Lingula</td>
</tr>
<tr>
<td></td>
<td>No hemispheric component</td>
</tr>
<tr>
<td>Central lobule</td>
<td>Wings of central lobule</td>
</tr>
<tr>
<td>Culmen</td>
<td>Quadrangular lobule</td>
</tr>
<tr>
<td>Decive</td>
<td>Simple lobule</td>
</tr>
<tr>
<td>Inferior vermis</td>
<td>Folium</td>
</tr>
<tr>
<td></td>
<td>Superior semilunar lobule</td>
</tr>
<tr>
<td>Tuber</td>
<td>Inferior semilunar lobule</td>
</tr>
<tr>
<td>Pyramid</td>
<td>Biventral lobule</td>
</tr>
<tr>
<td>Uvula</td>
<td>Tonsil</td>
</tr>
<tr>
<td>Nodule</td>
<td>Flocculus</td>
</tr>
</tbody>
</table>

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Fig. 2.12 Exposure of the suboccipital surface of the cerebellum during surgery. PICA, posterior inferior cerebellar artery.
side, just lateral to the median sulcus, there are facial colliculi, which are rounded prominences. Each facial colliculus is bounded laterally by the superior fovea, a dimple formed by the sulcus limitans. The inferior part is characterized by three triangular areas overlying the hypoglossal and vagus nuclei and the area postrema. The areas overlying the hypoglossal and vagal nuclei are the hypoglossal and vagal trigones. Just lateral to the hypoglossal trigone, there is another dimple formed by the sulcus limitans, called the inferior fovea.

The tentorial surface faces the tentorium. It has two incisura. The brainstem fits into the anterior cerebellar incisura and the falx cerebelli fits into the posterior cerebellar incisura. The anterosuperior margin, extending from the top of the culmen downward, forward, and laterally to a point above and behind the middle cerebellar peduncle, forms the posterior wall of the cerebellomesencephalic fissure. The anterolateral and posterolateral margins separate this surface from the petrosal and suboccipital surfaces, respectively. The lateral angle, which is seen in the lateral projection of angiography at the junction of the transverse and sigmoid sinuses, is the confluence of the anterolateral and posterolateral margins. The anterior angle is the confluence of the anterosuperior and anterolateral margins. The folia of the tentorial surface are represented by the superior vermis and its hemispheric counterpart (Table 2.1). Also part of the folium and superior semilunar lobule comes into the tentorial surface. The semilunar lobule occupies all three surfaces of the cerebellum. Most of the cerebellar lobules (except the lingual, tonsil, and nodule) occupy more than one surface. The cerebellomesencephalic or precentral cerebellar fissure is located between the cerebellum and midbrain. It is bounded posteriorly by the culmen above and the central lobule below. The lateral boundary is formed by the anterior surface of the quadrangular lobule above and the wing of the central lobule below. The anterior boundary in the midline is lingual and formed laterally by the superior and middle cerebellar peduncles. Of the cerebellar nuclei (fastigial, globose, emboliform, and dentate), the dentate nucleus is the most laterally located and the largest one. Most of the fibers of the superior cerebellar peduncle originate from the dentate nucleus, which is located at the posterior projection of the superior cerebellar peduncle. The lateral boundary of the dentate nucleus extends 0.5 to 2.0 cm from the midline. The lateral boundary of the dentate nucleus roughly correlates well with the posterior continuation of the interpeduncular sulcus (the sulcus between the superior and middle peduncles). This information is important when part of the cerebellum needs to be resected. The deep nuclei of the cerebellum must be preserved at all costs.

The petrosal surface faces the posterior surface of the petrous temporal bone and has two halves because of the intersection of brainstem. The two halves are separated by the petrosal fissure (Fig. 2.11), which runs lateral to medial. At the level of the flocculus, the petrosal fissure bifurcates into a superior limb and an inferior limb. The infralhfreeolar portion or the inferior limb separates the flocculonodule lobule from the rest of the cerebellum. It communicates with the cerebellomedullary fissure. The folia constituting the upper half of the petrosal surface are those of the tentorial surface that have folded over the middle cerebellar peduncle and the core of the cerebellum; similarly, the folia of the inferior petrosal surface of the cerebellum are formed by folia of the suboccipital surface of the cerebellum, which have folded over the inferior cerebellar peduncle. The choroids plexus and the rhomboid lip of the foramen of Luschka are located anteriorly and inferiorly to the flocculus. Just in front of the choroid plexus is the single superior rootlet of the glossopharyngeal nerve. The flocculus is located immediately below the lateral extension of the pontomedullary sulcus.

The fourth ventricle is a rhomboid-shaped structure, whose overall shape resembles that of a tent that has been turned over, with its base facing anteriorly with its two open lateral walls. The posterior surface of the pons and medulla forms the floor. The superior part of the roof is formed by the superior cerebellar peduncles, the superior medullary velum, and the adjacent lingula. The inferior part of the roof is formed by the inferior medullary velum, tela choroidea, choroids plexus, uvula, and nodule. The two open lateral walls of the fourth
ventricle are represented by the lateral recesses through which the fourth ventricle communicates with the cerebellopontine cistern. The conduit is also known as the foramen of Luschka through which the choroid plexus protrudes. The upper half of the roof of the fourth ventricle is composed of neural elements such as the superior cerebellar peduncles, superior medullary velum, and lingula. The lingula is easily visualized through the transparency of the superior medullary velum. The lower half of the roof is composed of non-neural elements and has a horizontal component and a vertical component. The horizontal part is the inferior medullary velum covering the nodule and superior pole of the tonsils. The vertical portion consists of the tela choroidea and the choroid plexus, covering the anterior aspect of the nodule, uvula, and part of the tonsils. These two parts unite at the telovelar junction and continue laterally as the floor of the lateral recess. The telovelar junction is important in gaining entry into the fourth ventricle. Entry through this route precludes splitting of the vermis, which results in severe truncal ataxia. Through the lateral recess the fourth ventricular cavity communicates with the cerebellopontine cistern. The direction of the lateral recess anteroinferiorly and laterally makes an angle of 45 degrees with the sagittal plane. The lateral recess has an anterior wall, superior wall (both formed by the inferior cerebellar peduncle), posterior wall, and floor (formed by the tela choroidea, choroid plexus, and inferior medullary velum from anterior to posterior). The inferior medullary velum becomes thick at the foramen of Luschka where it forms the peduncle of flocculus, which forms the posterior wall of the foramen of Luschka. The choroid plexus of the fourth ventricle is T-shaped with two vertical bars. The horizontal part starts from the fourth ventricle and protrudes into the cerebellopontine cistern. The vertical and proximal parts of the horizontal limb are supplied by the posterior inferior cerebellar artery (PICA), and the rest of the horizontal limb is supplied by the AICA.

◆ Vascular Anatomy

Arteries of the Posterior Fossa

The arteries of the posterior cranial fossa mainly come from the vertebrobasilar system. Arising from the first part of subclavian artery, the vertebral artery enters the foramen transversarium of the C6 vertebra. It then courses up through the transverse foramina of the cervical vertebrae to C2. After exiting from the foramen transversarium of C2, it curves laterally to enter the laterally placed foramen transversarium of C1. The C2 nerve root crosses this segment of the vertebral artery from behind. After exiting from the C1 transverse foramen, the vertebral artery sweeps behind the lateral mass of the atlas, just above the posterior arch in the suboccipital triangle to course superomedially to pierce the dura at the foramen magnum. The posterior spinal artery and posterior meningeal artery emerge from the vertebral artery at this level. At this level, the vertebral artery is accompanied by the suboccipital nerve or C1 nerve.

The vertebral artery can be divided into four parts: the first part extends from the origin to the entry into the foramen transversarium of the C6 vertebra. The second part is inside the transverse foramen of the cervical vertebrae. The third part is in the suboccipital triangle ([Fig. 2.14](#)). The fourth part is intradural and can be divided into lateral and anterior medullary segments, before it joins the mate from the opposite side to form the basilar artery. Usually one of the ver-
Cervical arteries is the dominant one; usually it is the left one, as it is wider than the right one. From the entrance into the intradural compartment the vertebral artery courses antero-medially and superiorly through the lower cranial nerve rootlets and lateral to the medulla to reach the preolivary sulcus. This segment is the lateral medullary segment of the vertebral artery. This segment begins at the dural foramen just inferior to the lateral edge of the foramen magnum. The dura in this region forms a funnel-shaped foramen around the first 4- to 6-mm length of the proximal part. The first cervical nerve exits and the posterior spinal artery enters the spinal canal through this dual foramen. These structures are bound by fibrous bands. The initial intradural segment of the vertebral artery passes just superior to the first cervical nerve and just anterior to the posterior spinal artery, the dentate ligament, and the spinal accessory nerve. After the preolivary sulcus, the anterior medullary segment begins, which runs in between the hypoglossal rootlets or anterior to them and crosses the pyramid to reach the pontomedullary sulcus, where it unites with the opposite vertebral artery to form the basilar artery. The branches of vertebral artery are the posterior spinal artery, anterior spinal artery, PICA, and anterior and posterior meningeal arteries. Several perforators arise from the vertebral artery, which penetrate the anterior and lateral surface of the medulla (Fig. 2.15).

The PICA arises from the vertebral artery at the anterolateral aspect of the brainstem (Figs. 2.15 and 2.16) near the inferior olive, and it supplies the medulla, inferior vermis, inferior part of the fourth ventricle, tonsils, and inferior aspect of the cerebellum. It is the largest branch of the vertebral artery. It has the most complex relationship with the cranial nerves of any artery, and it is frequently exposed in approaches directed to the fourth ventricle. After its origin it passes posteriorly around the medulla either caudally or rostrally to the hypoglossal rootlets. On reaching the posterolateral margin of the medulla, it passes rostral to the fila of the glosopharyngeal, vagus, or accessory nerves or in between them. After passing these nerves, it winds around the cerebellar tonsil and courses posterior to the roof of the fourth ventricle. After turning away from the roof of the fourth ventricle, it enters a series of deep fissures among the tonsil, vermis, and hemisphere. On exiting the fissures, the branches are distributed to the tonsil and the inferior surface of the vermis and hemisphere. In one study, 12.5% of PICAs passed between the glosopharyngeal and vagus nerves, 20% between the vagus and accessory nerves, and 65% through the rootlets of the accessory nerve.

The PICA is divided into five segments.7 The anterior medullary segment is short, located anterior to the medulla, and extends from the origin to the level of inferior olive. The lateral medullary segment extends from the inferior olive to the origin of cranial nerves IX, X, and XI and is located lateral to the brainstem. The lateral medullary segment of the PICA often has a lateral loop, which in 20% of cases is pressed against the inferior surfaces of the facial and vestibulocochlear nerves. The lateral medullary segment of the PICA passed superior to the hypoglossal nerve in approximately 20% of cases, through the rootlets of the hypoglossal nerve in 47.5%, and inferior to the hypoglossal nerve in 30%.8 The posterior medullary segment (also known as the tonsillomedullary segment) begins at the level of the origin of the cranial nerves IX, X, and XI and loops below the inferior pole of the cerebellar tonsil and upward along the medial surface of the tonsil toward the inferior medullary velum. This is also known as the caudal loop. The next segment, the supratonsillar segment (also known as the telovelotonsillar segment), runs in the cleft between the tela choroidea and the inferior medullary velum superiorly and the tonsil inferiorly. This begins below the fastigium, where the PICA turns posteriorly over the medial side of the superior pole of the tonsil. This part is called the cranial loop. The junction of the posterior medullary segment and the supratonsillar segment is called the choroidal point. The last segment is the cortical segment. After a short distance distal to the apex of the cranial loop, the PICA continues posteriorly in the retrotonsillar fissure.

Fig. 2.15 Relationship of the right vertebral artery with the lower cranial nerves. AFB, acousticofacial bundle; AICA, anterior inferior cerebellar artery; CN IX, glosopharyngeal nerve; CN X, vagus nerve; CN XI, accessory nerve; CN XII, hypoglossal nerve; JT, jugular tubercle; PICA, posterior inferior cerebellar artery; VA, vertebral artery.

Fig. 2.16 Course of the PICA below the cerebellum.
Here it bifurcates into two terminal branches, the tonsillo-hemispheric and inferior vermian branches. The tonsillo-hemispheric branch courses inferiorly near the prepyramidal sulcus and gives off anterior or tonsillar branches and posterior or hemispheric branches, which curve posteroinferiorly around the biventral lobule to the underside of the cerebellar hemisphere. The inferior vermian branch lies on the lower aspect of the inferior vermis and forms a convex loop (the pyramidal loop) around the copula pyramidis (connects the pyramid to the biventral lobule and forms the posterior wall of the retrotonsillar space). The anteriormost point of the curve of the pyramidal loop is called the copular point. Apart from terminal branches, the PICA gives rise to perforating, choroidal, and cortical branches; 5 to 20% of PICAs have an extradural origin.

The AICA usually arises from lower third of the basilar artery (Fig. 2.17) and less frequently from the middle third. (The AICA and PICA are defined according to their origin rather than by the part of the cerebellum that they supply.) After emerging from the basilar artery, the AICA courses postero-laterally and downward on the belly of the pons in contact with the abducens nerve. It may cross superior or inferior to the abducens nerve. On reaching the acousticofacial bundle, just before or after crossing the rootlets, the AICA bifurcates into its two major branches, the caudomedial and rostro-lateral (the main trunk). The caudomedial artery runs inferomedially toward the anterior and medial border of the cerebellum and supplies the middle cerebellar peduncle and biventral lobule. After its origin on the lateral aspect of the pons, the artery runs toward the pontomedullary sulcus to form its caudal loop on the lateral aspect of the pons and medulla. This loop can lie on the antero-inferolateral aspect of the flocculus, on the petrosal aspect of the biventral lobule, or the petrosal aspect of the undersurface of the biventral lobule. Distal to this loop, the next segment (the biventral segment) turns posteroinferiorly to reach the posterior surface of the cerebellum. It may also anastomose with the PICA or give off ascending hemispheric branches, which supplement or may entirely replace the hemispheric branches of the PICA. The rostrolateral trunk runs laterally and curls around the flocculus, and then it courses within the petrosal fissure from where the hemispheric branches to the superior and inferior semilunar lobule are given off.

The rostrolateral trunk is divided into three segments according to their relationship to the acousticofacial bundle. The premeatal segment begins at the origin of the rostrolateral trunk and courses around the brainstem to reach the acousticofacial bundle and the region of the meatus. The rostrolateral trunk, with the main trunk of the AICA, forms a caudal loop on the lateral aspect of the pons. The most posteroinferior point is called the caudal point. The rostrolateral trunk then continues superolaterally to reach acousticofacial bundle anterior to the flocculus. On the acousticofacial bundle and the middle cerebellar peduncle it describes either a single arterial or a double arterial loop. The single arterial loop is the single meatal loop because it is related to the internal acoustic meatus. The rostrolateral trunk ascends on the anterior aspect of cranial nerve VII, forming the apex of the meatal loop on the facial nerve near or within the internal auditory canal. From this part occasionally the internal auditory artery is given off. It then reverses the course and descends medially and posteroinferiorly to course between cranial nerves VII and VIII (Fig. 2.17B) or between cranial nerve VIII and the flocculus onto the middle cerebellar peduncle. Here it gives off branches to the middle cerebellar peduncle and the pons. The double arterial loop is called the M segment. The M segment has a proximal meatal loop and a brachial loop closely related to the middle cerebellar peduncle. When the M segment is present, after the meatal loop, the rostrolateral trunk reverses course to ascend as the brachial loop either on the superficial aspect of cranial nerve VIII or on the middle cerebellar peduncle. The apex of the brachial loop is also called the trigeminal point and it can pass anterosuperiorly on the middle cerebellar peduncle to reach the sensory root of the...
trigeminal nerve. At the trigeminal point it reverses its direction and runs posteroinferiorly on the middle cerebellar peduncle to form the loop. Distal to the single meatal loop or M segment is the postmeatal segment of the rostrolateral trunk. The rostrolateral trunk runs posteriorly and usually inferiorly to enter the suprafloccular portion of the petrosal fissure, where it divides into an ascending branch to the petrosal fissure, which supplies the petrosal surface of the cerebellum, and a descending branch to the posterolateral fissure, which supplies the retrofissural region.

The superior cerebellar artery (SCA) arises near the apex of the basilar artery (Fig. 2.18) just proximal to the origin of the posterior cerebral artery (PCA) and is the rostral-most artery of the infratentorial compartment. The SCA encircles the lower midbrain and pons and supplies the tentorial surface of the cerebellum, the deep cerebellar nuclei, the upper brainstem, and the inferior colliculi. The SCA can be divided into four segments. From its origin to the anterolateral part of the brainstem is the anterior pontomesencephalic segment. It runs laterally on the anterior aspect of the upper pons, often with a convex arc inferiorly. At the anterolateral margin of the brainstem it lies inferior to the third cranial nerve. The initial course of this segment depends on the site of the basilar bifurcation. With a low bifurcation, this segment passes upward, whereas in a high bifurcation, it initially passes anteroinferiorly to reach the anterior surface of the pons. The lateral pontomesencephalic segment starts from the anterolateral margin of the brainstem and runs caudally on the lateral side of the upper pons in the infratentorial portion of the ambient cistern to end at the anterior margin of the cerebellomesencephalic fissure. From this segment arises the first major cortical branch of the SCA, which runs anterolaterally to reach the anterolateral margin of the cerebellum. It then runs posterolaterally in the region of the petrosal fissure to supply the adjacent areas. Its area of supply is inversely related to the area supplied by the AICA. This vessel is an important angiographic landmark to locate the anterolateral margin and anterior angle of the cerebellum. Medial to this segment is the brainstem; lateral to this segment is the wing of the central lobule; inferior to this segment is middle cerebellar peduncle. Its caudal loop projects toward and often reaches the root entry zone of the trigeminal nerve. The bifurcation of the SCA into rostral and caudal branches occurs in this segment. The rostral branch supplies the superior vermis and the inner part of the tentorial surface of the cerebellum. The caudal branch supplies the rest of the tentorial surface not supplied by the rostral branch. The third segment, the cerebellomesencephalic segment, runs in the cerebellomesencephalic fissure and reaches the anterosuperior margin of the cerebellum through a series of hairpin bends. From this segment small cortical branches from the rostral and caudal branches arise, and these are precentral branches (twigs from the rostral branch supply the inferior colliculi and superior medullary velum, and twigs from the caudal branch supply the deep cerebellar nuclei). The last segment is the cortical segment. The hemispheric and vermic branches represent this segment.

### Veins of the Posterior Fossa

The veins of the posterior fossa can be divided into three groups: (1) the petrosal group or the anterior group draining into the superior and inferior petrosal sinuses, (2) the superior or the galenic group draining into the vein of Galen, and (3) the posterior or tentorial group draining into the venous sinuses in and around the torcular sinus. Usually the veins tend to drain into the venous sinus closest to them. The exceptions are the veins on the surface of the midbrain, which drains into the galenic system. The veins of the posterior fossa are summarized in Table 2.2.

#### Cisternal Anatomy

There are several subarachnoid cisterns in the posterior fossa or infratentorial compartment. Understanding the anatomy and disposition of these cisterns is extremely important in performing microsurgical dissection (Figs. 2.15, 2.16, 2.17, 2.18, and 2.19).

The **interpeduncular cistern** is a cone-shaped cul-de-sac occupying the interpeduncular fossa and is bounded above by the inferior surface of the mesencephalon, the lower diencephalons, the posterior perforated substance, and the mammillary bodies. Anteroinferiorly it is bounded by the clivus and superolaterally it is bounded by the carotid and crural cisterns and the medial temporal lobes. Inferolaterally it is continuous with the cisterna ambiens. The anterosuperior wall of this cistern is thick. Like a curtain, it stretches from one medial temporal surface to another and is fused with the chiasmatic cistern around the infundibulum and pituitary stalk. Above, it is attached to the mammillary bodies, and inferiorly it is attached to dorsum sellae. This is Liliequist membrane. The inferior part of the cistern extends in a triangular shape down to the middle portion of the basilar artery. The origins of the PCA and SCA are in this cistern. Within the interpeduncular cistern, the PCA and SCA do not have much arachnoid membrane around them, but at the level of the...
The **oculomotor nerve**, the **PCA and SCA** acquire their own arachnoid sleeve and enter the cisterna ambiens. The contents of the interpeduncular cistern are listed in **Table 2.3**.

The **prepontine cistern** is located between the anterior surface of the pons and the clivus and surrounds the basilar artery. The AICA originates in this segment, and moderately dense arachnoid fibers encircling the AICA denote its exit from this cistern into the cerebellopontine cistern. Laterally, the cistern is bounded by an arachnoid membrane, which forms the medial boundary of the cerebellopontine cistern. Inferiorly, the arachnoid wall of the cistern is thickened at the confluence of the vertebral arteries forming the basilar artery. Superiorly, a plane separates the prepontine cistern from the interpeduncular cistern. The contents are listed in **Table 2.3**.

The **premedullary cistern** extends superiorly from the pontomedullary sulcus to the upper cervical area inferiorly. The

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### Table 2.2  Veins of Posterior Fossa

<table>
<thead>
<tr>
<th>Major Group</th>
<th>Divisions</th>
<th>Veins</th>
</tr>
</thead>
<tbody>
<tr>
<td><em><em>Petrosal</em> group</em>*</td>
<td>Veins related to the anterior aspect of the brainstem</td>
<td>• Anterior pontomesencephalic vein</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Transverse pontine vein</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Lateral pontine vein</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Anterior medullary vein</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Parenchymal perforating vein</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Brachial veins</td>
</tr>
<tr>
<td></td>
<td>Veins in the wing of precentral cerebellar fissure</td>
<td>• Superior hemispheric veins, lateral group (draining lateral portion of the wing of central lobule, quadrangular lobule, simple lobule, tentorial part of superior semilunar lobule)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Inferior hemispheric veins</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Includes veins of great horizontal fissure</td>
</tr>
<tr>
<td></td>
<td>Veins on the superior and inferior surfaces of the cerebellar hemisphere</td>
<td>• Superior hemispheric veins, lateral group (draining lateral portion of the wing of central lobule, quadrangular lobule, simple lobule, tentorial part of superior semilunar lobule)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Inferior hemispheric veins</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Includes veins of great horizontal fissure</td>
</tr>
<tr>
<td></td>
<td>Veins on the cerebellar side and medullary side</td>
<td>• Medial tonsillar vein</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Retro-olivary vein</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Vein of inferior cerebellar peduncle</td>
</tr>
<tr>
<td><strong>Superior or galenic group</strong></td>
<td>Veins in the wing of precentral cerebellar fissure</td>
<td>• Median anterior pontomesencephalic vein</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Lateral anterior pontomesencephalic vein</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Lateral pontomesencephalic vein</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Lateral mesencephalic vein</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Peduncular vein</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Posterior mesencephalic veins</td>
</tr>
<tr>
<td></td>
<td>Cerebellar tributaries</td>
<td>• Tectal veins</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Precentral cerebellar vein</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Superior vermian vein (draining central lobule and culmen)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Superior emispheric veins, anterior group (draining intermediate portion of the wing of the central lobule and quadrangular lobule)</td>
</tr>
<tr>
<td><strong>Tentorial or posterior group</strong></td>
<td>Veins on the cerebellar side and medullary side</td>
<td>• Superior retrotonsillar vein</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Inferior retrotonsillar vein</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Drains tuber, pyramid, uvula</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Superior hemispheric veins, posterior group (draining declive, folium, intermediate portion of simple and superior semilunar lobules)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Inferior hemispheric veins</td>
</tr>
</tbody>
</table>

*Superior petrosal vein is formed by the junction of the transverse pontine and brachial veins, and the vein of great horizontal fissure.
The cistern's anterior boundary is the clivus, and the posterior boundary is the anterior surface of the medulla.

The ambient cistern (cisterna ambiens) skirts the lateral aspect of the mesencephalon and has both supratentorial and infratentorial components. It is a paired structure. Medially it is bounded by the cerebral peduncle and interpeduncular cistern. Laterally it is bounded by the medial temporal lobe (supratentorial) and the quadrangular lobule of the cerebellum (infratentorial). Inferiorly it shares the wall with the cerebellopontine cistern. Anteriorly it is related to the crural cistern. The superior extension of the ambient cistern, although not in the infratentorial compartment, warrants special mention. It is called the wing of the ambient cistern, which includes the portion of the cistern extending from the uncus of the temporal lobe, over the pulvinar of thalamus, and anteromedially to the area of velum interpositum. The contents are listed in Table 2.3.

### Table 2.3  Arachnoid Cisterns of the Posterior Fossa and Their Contents

<table>
<thead>
<tr>
<th>Cistern</th>
<th>Nerve</th>
<th>Artery</th>
<th>Vein</th>
</tr>
</thead>
<tbody>
<tr>
<td>Interpeduncular cistern</td>
<td>Oculomotor nerve</td>
<td>• Upper part of basilar artery</td>
<td>• Pontomesencephalic veins</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• PCA origin</td>
<td>• Basal vein of Rosenthal</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• SCA origin</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Thalamoperforating arteries</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Medial posterior choroidal arteries (origin)</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Quadrigeminal arteries</td>
<td></td>
</tr>
<tr>
<td>Preptone cistern</td>
<td>Abducens nerve</td>
<td>• Basilar artery</td>
<td>• Pontine veins</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Origin of AICA</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Brainstem perforators</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Anterior spinal artery</td>
<td></td>
</tr>
<tr>
<td>Premedullary cistern</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Cisterna ambiens/ambient cistern</td>
<td>Trochlear nerve with its arachnoid sheath</td>
<td>• PCA (P2, P3)</td>
<td>• Median medullary vein</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• SCA with its arachnoid sheath</td>
<td>• Lateral pontomesencephalic vein</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Lateral posterior choroidal artery (origin)</td>
<td>• Basal vein of Rosenthal</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Quadrigeminal artery</td>
<td></td>
</tr>
<tr>
<td>Cerebellopontine cistern</td>
<td>Facial nerve</td>
<td>• AICA and its branches</td>
<td>• Superior petrosal vein</td>
</tr>
<tr>
<td></td>
<td>Vestibulocochlear nerve</td>
<td></td>
<td>• Lateral recessus vein</td>
</tr>
<tr>
<td></td>
<td>Trigeminal nerve</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Trochlear nerve</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Lateral cerebellomedullary cistern</td>
<td>Glosopharyngeal nerve</td>
<td>• Vertebral artery</td>
<td>• Inferior petrosal vein</td>
</tr>
<tr>
<td></td>
<td>Vagus nerve</td>
<td>• PICA (origin)</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Accessory nerve</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Hypoglossal nerve</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Cisterna magna</td>
<td>C1, C2 nerves</td>
<td>• PICA (distal)</td>
<td>• Inferior vermian vein</td>
</tr>
<tr>
<td>Superior cerebellar cistern</td>
<td></td>
<td>Medial and terminal branches of SCA</td>
<td>Tributaries to the tentorial dura, straight sinus and precentral cerebellar veins.</td>
</tr>
</tbody>
</table>

Abbreviations: AICA, anterior inferior cerebellar artery; PCA, posterior cerebral artery; PICA, posterior inferior cerebellar artery; SCA, superior cerebellar artery.
The cerebellopontine cistern is also a paired structure. The lateral surface of the pons forms the medial boundary of the cistern. Superiorly the cistern shares an arachnoid wall with the ambient cistern, and inferiorly the arachnoid membrane separates it from the lateral cerebellomedullary cistern. Posteriorly the cistern is covered by the posterior quadrangular and superior semilunar lobules of the petrosal surface of the cerebellum. Medially the flocculus is immediately posterior to the cerebellopontine cistern. Laterally the cistern extends along the posterior petrous portion of the temporal bone entering the internal auditory canal and extending outward into Meckel’s cave. The trigeminal nerve, like the oculomotor nerve in the interpeduncular cistern, has its own arachnoid or cisternal sleeve that is separate from it, but that forms a recess into the cerebellopontine cistern.

The lateral cerebellomedullary cistern is also a paired structure and is located anterolateral to the medulla. The anterosuperior border is the sulcus between the pons and medulla. Arachnoid over the glossopharyngeal, vagus, and accessory nerves separates this cistern from the cisterna magna dorsally and from the cerebellopontine cistern superiorly. A less clear arachnoid layer separates it from the premedullary cistern. The cistern extends from the pontomedullary sulcus to the foramen magnum. Sleeves of arachnoid run along with the posterior medullary segment of the PICA. The tonsil is then retracted laterally, which exposes the roof of the fourth ventricle. The tela choroidea or the inferior medullary velum is dissected, and the tonsil is freed from the opposite tonsil uvula, inferior medullary velum, tela choroidea, and posterior medullary segment of the PICA. The tonsil is then retracted laterally, which exposes the roof of the fourth ventricle. The tela choroidea or the inferior medullary velum can be resected to gain better access. Occasionally, removal of the tonsil may be necessary.

Supracerebellar Infratentorial Approach to the Posterior Third Ventricle

The space between the tentorium and the tentorial surface of the cerebellum is a wide and excellent natural pathway to access the posterior third ventricular region. The operation is usually performed with the patient in a sitting position to take the advantage of gravity. A midline incision is made from the lambda to the tip of C2. The suboccipital muscles are resected to gain better access. Occasionally, removal of the tonsil may be necessary.
vermian vein or the precentral cerebellar vein or both in the cerebellomesencephalic fissure have to be sacrificed. The arachnoid in the quadrigeminal cistern is very thick and must be opened carefully, because there is a chance of injury to the branches of the SCA and PCA and even the great veins. Usually after opening the arachnoid adequately the lesion in the posterior third ventricle comes into view.

Preauricular Subtemporal-Infratemporal Approach to the Petroclival Region$^{11,12}$

This approach is used mainly to access the extradural petroclival junction, and access can be made from the nasopharynx to the foramen magnum. After the skin and the subcutaneous tissue are mobilized in the temporal region, parotidomasseteric fascia is dissected and separated from the parotid gland. This maneuver is extremely important. Later, when the mandibular condyle is retracted downward, the facial nerve is liable to become stretched. To minimize the traction on the facial nerve, the parotidomasseteric fascia is dissected free. The zygomatic arch and the frontozygomatic process are completely denuded of periosteum and exposed. A temporal or a frontotemporal craniotomy is made. Under the microscope the temporal dura is separated gently from the lateralmost part of the MCF. While separating, the arcuate eminence (AE) is identified. This is usually the posterior boundary of the dissection. The dural separation is then continued anteriorly to reach the foramen spinosum where the middle meningeal artery (MMA) is seen entering the cranial cavity. Further forward dissection exposes the lateral rim of the foramen ovale transmitting the mandibular division of the trigeminal nerve (V3). At this stage it is not possible to see the V3 clearly as it is covered by dural reflections. Medial dissection in between the AE and the MMA exposes the lesser superficial petrosal nerve (LSPN), which can be seen coursing toward the foramen ovale. The LSPN is cut and medial separation of the dura is continued. The MMA is cut at this stage. Further medial dissection exposes the GSPN emerging from the facial hiatus to supply part of the facial nerve. It is important to remember that if the GSPN is found, which is continuous with the periosteum covering the V3 nerve, the V3, and the V2 is drilled. This exposes the lateral part of the root of the pterygoid process. Drilling the bone of the MCF between V2 and V3 may expose the sphenoid sinus.

Attention is then turned toward exposure of the petrous ICA. Initially a search is made posteroinferomedial to V3, whether or not any part of the ICA is visible. If it is visible, it serves as a good guide. If the ICA is completely covered by bone, one must take extra caution while drilling. Drilling lateral to the horizontal part of the petrous ICA is relatively safe. The ICA is covered by a venous plexus variably. During drilling, bleeding from this venous plexus may occur. But if the periosteal sheath covering the ICA is kept intact, bleeding does not occur. While drilling the bone laterally, one first sees the muscle and glistening tendon of the tensor tympani muscle (TTM). The muscle is cut and reflected away. Just inferior to the TTM, the cartilaginous part of the ET is found. The ET may be separated from the ICA by a very thin plate of bone. Dissection is made around the ET, to free it from the ICA, and then it is cut to expose the lateral wall of the horizontal part of the petrous ICA completely. Bone drilling is continued to expose the superior, lateral, and inferior walls of the horizontal part of the petrous ICA. During exposure of the genu, care must be taken to avoid damage to the cochlea. However, if the drilling is done lateral to the ICA, the chance of injuring the cochlea is minimal. In this approach, it may not be necessary to drill the bone superior to the genu completely. The cochlea is posteromedial to the genu.

The petrous bone and the tympanic plate lateral and anterior to the vertical limb of the petrous ICA are drilled completely. During this part of the procedure, the condyle of the mandible has to be retracted inferiorly, and the downward tilt of the vertex is reduced. It may be necessary to tilt the vertex a little upward. This improves the view of the lower part of the petrous ICA. Here, a thick fibrocartilaginous band is found, which is continuous with the periosteum covering the ICA. This fibrocartilaginous ring is cut and is gently dissected away from the wall of the ICA. This frees the ICA to be retracted anteriorly and laterally. However, a thin bone plate, between the V3 and the horizontal part of the petrous ICA, will hinder complete anterior mobilization. This bone forms the medial rim of the foramen ovale. This bone is removed. Then the ICA is completely free to be retracted anteriorly. One
or two tapes are passed around the ICA, and the ICA is retracted anteriorly and laterally. Part of the petrous bone medial to the carotid canal is left now. This can be drilled freely as hardly any important structure is there. One may expect some bleeding here during drilling. However, while drilling posteriorly, one must be careful not to go too far back and encounter the cochlea. Further medially and backward the anterior wall of the IAC is seen. The petrous pyramid is drilled completely to expose the posterior fossa dura bounded above and below by the superior petrosal sinus (SPS) and below by the inferior petrosal sinus (IPS). Just below the IPS, the clivus is visible. The dura can be opened along the region of Meckel's cave or according to the demands of the patient's condition. Usually the dura is opened in the posterior fossa first. Then we can see a triangular space exposed in the posterior fossa dura bounded inferiorly by the IPS and the clivus bone, superiority by the trigeminal root and the SPS, and posteriorly by the superior SCC, IAC, and the jugular bulb. The dura is opened in a triangular fashion, with the base posteriorly placed. If necessary, the triangle can be split in two by incising the dura along the anterior aspect of the IAC. This exposes cranial nerve VI first exiting the pontomedullary junction on the ventral surface. The contralateral VI nerve can also be visualized, but the origin may not be. The midbasilar artery, the origin of the AICA, is ipsilateral cranial nerve VII–VIII and IX–X–XI complexes and cranial nerve XII can be seen. In fact it is possible to see the cranial nerve XII of both sides. Intradural pathology ventral to brainstem can be removed by this approach. To gain exposure above, the tentorium is cut. First the dura is cut along the inferior surface of the temporal lobe in a line along the anteroposterior axis. Another incision is made at a right angle to the first one, forming a T. The incision parallel to the coronal plane meets the SPS. The SPS is ligated and cut. Now the temporal lobe is gently elevated to expose the tentorial notch. Cranial nerve IV is seen diving below the tentorium. The tentorium is incised posterior to the point at which nerve IV is diving below the tentorium. The tentorial incision is extended to the point where the SPS was ligated. Sutures are placed in the tentorium and the tentorium is retracted. Now all the cranial nerves from III to XII are exposed. Cranial nerve V is seen exiting the brainstem and entering Meckel's cave, which has already been drilled.

Retrosigmoid Approach to the Cerebellopontine Angle

This approach, one of the major workhorse approaches to the posterior fossa, is directed through an opening located just behind the sigmoid sinus and is guided down the plane between the posterior face of the petrous temporal bone and the petrosal surface of the cerebellum.13 As mentioned before, during a craniotomy the asterion is an important landmark in localizing the genu of the lateral sinus. While operating on vestibular schwannoma through this approach, it is necessary to remove the posterior meatal wall. In that case it is often necessary to sacrifice the subarcuate artery because it passes through the dura on the posterior meatal wall to reach the subarcuate fossa. This artery has enough length so that it can be coagulated safely without jeopardizing AICA, but sometimes the stem is too short or the loop of the AICA along with the subarcuate artery is incorporated into the dura. In that case, a cuff of dura should be cut to separate the dura and the artery from the posterior meatal lip to prepare the opening of the meatus. The posterior semicircular canal (PSCC) and its common crus with the superior semicircular canal are situated just lateral to the posterior meatal lip, and should be preserved while exposing the IAC by drilling the posterior meatal lip, if hearing preservation is an issue. Similarly, injury to the endolymphatic sac, which expands under the dura on the posterior face of the petrus temporal bone inferolateral to the posterior meatal lip, should be avoided; it may be entered while removing the dura from the posterior meatal lip. Injury to the vestibular aqueduct, situated inferolateral to the posterior meatal lip, should also be avoided. An unusually high jugular bulb may block access to the posterior meatal lip. Encountering mastoid air cells is common while drilling this region and they must be obliterated meticulously to prevent CSF leak.

After the removal of the posterior wall of the internal auditory canal, the dura lining the canal is slit open to expose the contents. The nerves in the lateral part of the IAC are the facial, the cochlear, and the superior and inferior vestibular nerves. The position of the nerves is most constant in the lateral portion of the canal, which is divided into a superior and an inferior portion by a horizontal ridge, called the transverse crest or the falciform crest. The facial and the superior vestibular nerve are superior to the crest. The facial nerve is anterior to the superior vestibular nerve here and is separated from the superior vestibular nerve by Bill’s bar (vide supra). The cochlear and the inferior vestibular nerve run inferior to the transverse crest with the cochlear nerve anterior to the inferior vestibular nerve. Knowing this relation is of utmost importance in surgery of vestibular schwannoma. The tumor usually displaces the facial and cochlear nerves anteriorly. Variable growth of the tumor may displace the facial nerve anteriorly, anterosuperiorly, or anteroinferiorly. It is easier to locate the facial nerve at the lateral end of the IAC, as the relation between the four nerves is most constant here, rather than in the medial location. The landmarks that are helpful in guiding the surgeon to the junction of the facial nerve with the brainstem are the pontomedullary sulcus, the junction of cranial nerves IX, X, and XI with the medulla, the foramen of Luschka with its choroid plexus, and the flocculus. The facial nerve arises from the brainstem near the lateral end of the pontomedullary sulcus approximately 1 to 2 mm anterior to the point where the vestibulocochlear nerve joins the brainstem, which is at the lateral end of the pontomedullary sulcus (where it joins the foramen of Luschka). At this point the interval between the facial nerve and the vestibulocochlear nerve is the greatest.13 Another way of locating the medial end of the facial nerve is to project an imaginary line along the brainstem attachment of cranial nerves IX, X, and XI to the brainstem, and the junction of facial nerve is at the junction of the pontomedullary sulcus and this imaginary line, which is approximately 2 to 3 mm rostral to the rostral-most rootlet of the glossopharyngeal nerve. Distal to the junction with the brainstem, the facial nerve and the ves-
tibulocochlear nerves come closer as they approach the IAM. Apart from the facial nerve, the vestibulocochlear nervus intermedius is also a component of the acoustofacial bundle. There are three segments of nervus intermedius: the proximal segment adheres closely to the vestibulocochlear nerve, the intermediate segment lies free between the motor root of the facial and vestibulocochlear nerves, and distal segment joins the motor root to form the facial nerve. In most instances, it arises as a single root at the brainstem anterior to the superior vestibular nerve. Even when it arises as multiple rootlets, it quickly converges to a single trunk before joining the motor root of the facial nerve.

Retrolabyrinthine Petrosectomy Approach to the Cerebellopontine Angle or the Petroclival Region

This presigmoid approach starts with a standard mastoidectomy. The bone removal extends from the supramastoid crest superiorly, the mastoid tip anteriorly, and the posterior wall of the external auditory canal anteriorly. A straight cut made along the temporal line posteriorly into the sinodural angle delineates the upper portion of the dissection. A second cut is made perpendicularly to the first and toward the mastoid tip. This cut is made immediately posterior to the canal wall. The mastoid cortex is then removed in a systematic fashion, with the deepest penetration occurring at the junction of the two perpendicular lines. The mastoid cortex should be unroofed from the posterior canal wall back to and slightly beyond the sigmoid sinus and at an adequate distance into the mastoid tip. This results in a bean-shaped cavity, with the inferior portion formed by the mastoid tip below the sigmoid sinus and the external auditory canal, and the upper portion above the sigmoid sinus extending posteriorly into the sinodural angle and anteriorly into the zygomatic root. The sigmoid sinus, the sinodural angle, and the middle fossa dura should be exposed. Exposure of the middle fossa dura is essential to achieve the best possible access into the antrum and the epitympanic areas.

The next step exposes the mastoid antrum, which is invariably present, although its size may vary. It lies immediately below the deepest point of penetration into the temporal bone, posterior to the spine of Henle and the zygomatic root. Occasionally, before the exposure of the antrum, Köhler’s septum, which is actually a segment of the petrosquamous suture line, representing the fusion of the squamous and petrous bone (extending from posterior canal wall at the tympanomastoid suture line and blends with the air cells in the immediate proximity of the middle fossa dural plate), is usually observed. The antrum is identified as a large, air-containing space, at the bottom of which lies the basic landmark of the labyrinthine bone, the horizontal or lateral SCC. Just anterior to the lateral SCC is the aditus through which the middle ear can be seen. Here we can also find the facial buttress. Next, the fossa incudis, the epitympanum (anterior and superior to lateral SCC), and the external genu of the facial nerve (medial and inferior to lateral SCC) are exposed. Drilling the inferior mastoid cells will expose the digastric ridge, which is the cortical indentation of the attachment of the digastric muscle. Traced anteriorly, the digastric ridge leads to the facial nerve at the stylomastoid foramen. Thinning of the posterior wall of the external auditory canal exposes the bone in the facial recess area, bounded medially by the vertical mastoid segment of the facial nerve, laterally by the chorda tympani nerve, superiorly by the fossa incudis, and anteriorly by the tympanic membrane. Unless the facial nerve is transposed, it is prudent to preserve a thin shell of bone over the facial nerve, thereby preserving the periosteum. Then the vessels over the facial nerve can be seen. Preservation of these vessels is of the utmost importance if facial nerve function is to be preserved. Drilling the infralabyrinthine cells exposes the jugular bulb. To expose the jugular bulb completely, the facial nerve needs to be transposed anteriorly by mobilizing. The disorders that require the retrolabyrinthine petrosal approach do not require exposure of the jugular bulb.

Drilling the bone posterior to the lateral SCC exposes the posterior SCC, which is bisected by the lateral SCC, and runs parallel to the posterior fossa plate. The posterior SCC starts from just medial to the facial nerve below the external genu and ends superiorly at the common crus with the superior SCC. The endolymphatic sac, which is medial to the sigmoid sinus and inferior to the posterior SCC, is found by thinning the posterior fossa dural plate, immediately medial to the sigmoid sinus. It is identified by the presence of thickened white dura next to the darker, single-layered dura. The superior SCC is oriented perpendicularly to the lateral SCC and is approximately parallel to the floor, with its long axis orthogonal to the long axis of the petrous bone. It originates deep to the lateral SCC and arches posteriorly and superiorly to join the common crus. Next, a temporal craniotomy is added with its posterior boundary extending beyond the transverse sinus and the sigmoid sinus junction. The posterior fossa dura is opened first and the CSF in the cerebellopontine cistern is released to make the brain slack. A longitudinal incision is made in the inferior temporal dura. Here the utmost care is needed to preserve the vein of Labbé. The dural incision is then extended in a T-shaped manner to the superior petrosal sinus, which is ligated and the incision is extended medially to cut the tentorium up to its edge, with a special care to preserve the trochlear nerve. This exposes the cerebellopontine angle from the anterolateral direction, and it makes the approach to the clivus shallower. To increase or widen the exposure, the approach can be enlarged by converting it to a partial labyrinthectomy petrous apicectomy approach, a translabyrinthine approach, or a total petrosectomy approach.

Partial Labyrinthectomy Petrous Apicectomy Approach to the Cerebellopontine Angle or the Petroclival Region

The bone removal initially is exactly like that in the retrolabyrinthine approach. After completing the temporal craniotomy, the middle fossa floor is dissected to locate the GSPN, which acts as a guide to the position of the petrous ICA while drilling the medial petrous bone. The superior SCC and part
of the posterior SCC are removed by drilling. Medial petrous bone is drilled up to the apex of petrous bone. The cavernous sinus is reached. The dura is opened as in the retro labyrinthine approach. If necessary, the trigeminal nerve may be mobilized by opening Meckel’s cave so that retraction of this nerve can be facilitated. The trigeminal nerve arises from the anterolateral surface of the pons. The sensory root is larger and the motor root is smaller. The motor root is medial to the sensory root. The nerve traverses the upper part of the cerebellomedullary cistern and comes in close contact with the superior petrosal vein. The nerve enters Meckel’s cave below the superior petrosal sinus by pushing a layer of dura along with it. The dura blends with the epineurium at the level of the gasserian ganglion. From the ganglion the three branches arise. The motor root joins with the mandibular division.

Translabyrinthine Approach

After the mastoidectomy the labyrinthine procedure is performed. It involves removal of the semicircular canals and vestibule to expose the dura lining the internal auditory canal. After drilling and removing the SCCs, the vestibule is reached and removed. Care is needed to avoid injury to the facial nerve as it courses below the lateral SCC and the ampulla of the posterior SCC, and around the superolateral margin of the vestibule. Bone is further removed at the lateral end of the canal to expose the transverse and the vertical crest along with the four nerves at the lateral end. While removing the bone behind the posterior wall of the IAC, it is important to remember that the jugular bulb may bulge upward behind the posterior SCC or IAC. The vestibular aqueduct and endolymphatic sac are opened and removed as bone is removed between the meatus and the jugular bulb. The cochlear canalculus is seen deep to the vestibular aqueduct as the bone is removed in between the jugular bulb and the IAM. The lower end of the cochlear canalculus is situated just above the area where the glossopharyngeal nerve enters the medial side of the jugular foramen. Sometimes the subarcuate artery or even the AICA may be encountered in Trautmann’s triangle.

Total Petrosectomy Approach

This approach is reserved for giant tumors or aneurysms in the clival-petroclival region. It is also suitable for patients who were previously operated and radiated. After making a wide C-shaped skin incision extending forward to the anterior aspect of the superior temporal line, the external auditory canal is transected and over-sewn. The temporomandibular joint capsule is dissected free from the glenoid fossa. A complete mastoidectomy and a labyrinthectomy are done. The facial nerve is exposed from the stylomastoid foramen to the IAM. A temporal craniotomy is done with an anterior extent up to the sphenoid wing. A zygomatic osteotomy including the condylar fossa is done to gain access to the vertical segment of the petrous carotid artery. Occasionally, for greater exposure, the mandibular condyle needs to be resected. Next, the GSPN and the MMA are cut. The cartilaginous eustachian tube is exposed, cut, and sewn. The petrous carotid artery is completely mobilized from the proximal cavernous sinus portion to the upper cervical segment, as mentioned previously. The fibrocartilaginous ring surrounding the cervical carotid at its entrance into the skull base is divided, and the ICA is mobilized anteriorly. The cochlea and the bone medial to the mastoid part of the facial nerve are removed. The jugular foramen and its associated structures are skeletonized, and the facial nerve is mobilized posteriorly. Next, the medial petrous apex and clivus can be resected.

Far Lateral Approach to the Foramen Magnum and the Craniovertebral Junction

This approach is used to access anterior or anterolateral lesions in the lower clivus, C1, and C2. A lazy S-shaped, straight, or an inverted U-shaped incision is used. The advantage of an inverted U-shaped incision is the ease of doing a fixation if necessary. Some surgeons cut through the muscle in layers; others prefer an anatomic dissection, detaching the muscles from their attachments in layers. The advantage of the anatomic dissection is the ease of identifying the vertebral artery, which is located below the deepest layer of the superior and inferior oblique muscles. These muscles are detached, and the vertebral artery covered by the venous plexus is identified. This step must be done under the microscope. The foramen transversarium of the atlas and often the axis are unroofed to expose the vertebral artery. The venous sheath is coagulated and slit open longitudinally along the course of the artery, and the artery is completely taken out of the sheath. Considerable venous bleeding is encountered during this step, but once the artery is removed from the sheath, the venous bleeding is easily controlled. The vertebral artery is then followed up to the intradural entry. The venous sheath is adherent to the capsule of the joint formed by the occipital condyle and the C1 lateral mass, and the vertebral artery must be carefully detached to mobilize it completely. During this procedure, it must be remembered that the PICA may have extradural origin. The mobilization and proximal control depends on the pathology and the available intradural length of the vertebral artery. The suboccipital bone, the posterior part of the mastoid, and the ipsilateral posterior arch of the atlas are exposed. A suboccipital craniotomy is performed to include the foramen magnum. The posterior border of the sigmoid sinus is exposed. Below the jugular bulb, the bone removal is taken up to the occipital condyle. The ipsilateral posterior arch of C1 is also removed up to the lateral mass. The removal of the atlas depends on the lower extent of exposure needed. Up to this point, the exposure is adequate for the lesions in the anterolateral aspect of the foramen magnum. For the lesions placed more anteriorly, further bone removal is necessary. The posterior one third of the occipital condyle and superiorly the jugular tubercle are drilled away. For tumors involving the occipital condyle, drilling of the entire condyle is necessary. In this case, stabilization is necessary. After dural opening, the vertebral artery, dentate ligament, and cranial nerves, apart from the cerebellar tonsil and brainstem, come into view. The dentate ligament is a white fibrous
The spinal component arises as a series of rootlets situated midway between the ventral and dorsal roots. Its lateral border is attached at intervals by fibrous triangular processes to the dura. The rostral-most process is attached to the dura at the foramen magnum level, and the caudal process is attached posterior and inferior to the initial intradural segment of the vertebral artery. The lateral border of the dural ligament between the two rostral-most triangular processes is attached to the vertebral and posterior spinal arteries and to the C1 nerve roots, making separation of these structures difficult.

The rootlets of the hypoglossal nerve arise form the medulla along a line that is continuous with the line along which the ventral spinal roots arise. This line in the medulla is between the pyramid and the olive. These rootlets pass anterior to the vertebral artery and exit the intracranial compartment through the hypoglossal canal along with the venae comitantes. The glossohypharyngeal, vagus, and accessory nerves arise in a series of rootlets along a line posterior to the olive. They exit the intracranial compartment through the jugular foramen. At this point, where the glossohypharyngeal nerve penetrates the dura, it is separated from the vagus nerve by a dural septum.² The accessory nerve has two components. The cranial part is composed of rootlets arising from the medulla and upper rootlets joining the vagus nerve. The spinal part is formed by the union of a series of rootlets, which arise from the lower medulla and upper spinal cord. The spinal component passes through the foramen magnum and is joined by the lower-most rootlets of the cranial component. The spinal component arises as a series of rootlets situated midway between the ventral and dorsal roots of spinal nerves. The rootlets contributing to the formation of the spinal accessory may arise as low as the C7 root level.

Approach to the Jugular Foramen

The jugular foramen area is one of the most complex areas to reach. The choice of surgical approach depends on the exact location of the lesion and the presence of vascular encasement.

Transjugular/Mastoid and Neck Approach

This approach is used for lesions in the jugular bulb without extension into the neck or posterior fossa. A curved postauricular incision is made. The skin incision is carried down to the neck along the anterior border of the sternocleidomastoid muscle, and the sternocleidomastoid muscle is detached from the mastoid and reflected posteriorly. The digastric muscle is detached from the digastic groove and is reflected anteriorly. A complete mastoidectomy is performed, and the mastoid tip is transected lateral to the digastic groove. This exposes the major neurovascular bundle. The spinal accessory nerve is identified crossing the internal jugular vein (IJV) lateral to it. The glossohypharyngeal and vagus nerves are also identified and preserved. For paragangliomas, the IJV is doubly ligated in the neck. Next, the bone over the sigmoid sinus and the jugular bulb are completely drilled with a diamond bur. This exposes the entire jugular foramen region.

Mastoid and Neck Approach with Limited Facial Nerve Rerouting

This approach is needed for approaching more superiorly located lesions. Here, in addition to the approach described above, a total mastoidectomy is performed. The infralabyrinthine and retrofacial air cells are drilled to expose the facial nerve from the second genu to the stylomastoid foramen. The periosteum of the facial nerve canal is preserved, which in turn preserves the vascular supply of the facial nerve. The fibrous attachment at the stylomastoid foramen is sharply cut to mobilize the facial nerve anteriorly and laterally. This facilitates access to the jugular bulb. This approach is suitable for jugular foramen schwannomas and paragangliomas limited to the jugular bulb.

Infratemporal Approach

This approach is suitable for paragangliomas intimately involving the carotid artery. A C-shaped incision is made about 2 inches behind the postauricular crease. It extends upward and forward along the superior temporal line and extends below along the anterior margin of the sternocleidomastoid to the level of the thyroid cartilage. The skin and the galea flap are raised. The posterior wall of the external auditory canal is identified. Dissection of cervical flap is performed deep to the platysma muscle. The external auditory canal is transected at the level of the bony-cartilaginous junction. The cartilage is removed from the ear canal and the skin is fashioned as a cuff ready to be everting sutured. A mastoidectomy is performed. The chorda tympani is sacrificed to widen the facial recess. The posterior wall of the external auditory canal is drilled and removed. The ossicles of the middle ear, tympanic membrane, and the remaining skin of the external auditory canal are removed. The facial nerve is exposed from the geniculate ganglion to the stylomastoid foramen level. Next, the neck is exposed to take proximal control. The tympanic ring is removed to expose the jugular bulb completely. The tympanic bone is then drilled along with the bone in the region of the temporomandibular joint to expose the petrous carotid artery. The facial nerve is transposed anteriorly and laterally. The rest of the bone covering the petrous carotid artery and jugular bulb is drilled to skeletonize all the neurovascular structures in the jugular foramen area.

Extended Subfrontal Approach to the Clivus and Foramen Magnum

A bicoronal skin flap is raised and the skin is reflected downward, exposing the orbital rim completely on both sides and beyond the frontonasal suture in the midline. The temporalis muscles are detached from the anterior temporal fossa and are retracted posteriorly. The periiorbita is dissected from the roof and the upper medial wall of the orbit. A bifrontal craniotomy is performed. A bilateral orbital osteotomy is performed.
including the ethmoid. The orbito-fronto-ethmoid segment is loosened with an osteotome and removed after the attachments of the nasal mucosa and septum have been divided. The nasal cavity, ethmoid sinus, and orbits come into view. The ethmoid and nasal mucosa are vascular and should be cauterized. The posterior ethmoidal artery must be identified and cauterized. This is an important landmark, as the optic canal is just behind it. The optic canals may have to be decompressed depending on the pathology. The ethmoid cells are removed completely, which exposes the sphenoid sinus. The planum sphenoidale is resected. Then the sellar floor and lateral wall are drilled, completely exposing the cavernous sinus. Although it is difficult to drill the dorsum sellae, especially if it is prominent, it can be done with the help of a 30-degree endoscope. The medial cavernous sinus is protected by a thin periosteal layer, which may become torn or may have to be opened intentionally. Bleeding from the cavernous sinus is controlled with packing Surgicel or Gelfoam. The intracavernous ICA is followed along its anterior bend and medial surface of the horizontal and vertical segments to the petrocavernous junction. The medial part of the petrous apex as well as the body of the sphenoid bone may be removed this way. The clivus is drilled next, which exposes the clival dura. This may cause brisk bleeding from the basilar venous plexus. In young adults the dura is thick, but in the elderly it may be very thin and may be breached during drilling. The lateral boundaries of this approach are the cavernous sinuses, petrous apices, abducens nerves, and the hypoglossal canals. The lower boundary is the foramen magnum.

References


Neuroimaging of the posterior fossa is a diagnostic challenge for neuroradiology, radiology, neurology, and neurosurgery.\textsuperscript{1–5} Due to the dramatic technical progress that has been made in imaging modalities, clinicians must be knowledgeable about the imaging techniques and the indications for determining when these examinations are necessary. This chapter provides an overview of the present use and interpretation of imaging modalities for the posterior fossa. A discussion of the physical and technical basic principles of the imaging modalities is beyond the scope of this chapter. But a brief summary of the different imaging techniques, their indications, and the major advantage of focused techniques are presented here, along with the anatomic details of the posterior fossa and some historical background information.\textsuperscript{6–13} Radiologic cases are presented that demonstrate subtle pathologic findings, and interlinking the different modalities of neuroimaging can confirm the diagnosis.

**Topographic Anatomic Details of the Posterior Fossa**

The posterior cranial fossa is the deepest and most capacious of the three cranial fossae. It contains the cerebellum, pons, and medulla oblongata. The great foramen is centrally located in the posterior fossa. The posterior fossa is surrounded by deep grooves containing the transverse and sigmoid sinuses (Fig. 3.1). Neuroimaging can portray this detailed neuroanatomy to assist in evaluating the posterior fossa.

The brainstem is formed by the midbrain, pons, and medulla oblongata, and is partially obscured by the cerebral hemispheres and cerebellum. Small motor or sensory nuclei of the cranial nerves are scattered in the gray matter in the brainstem. The midbrain is located between the pons and the cerebral hemispheres. The dorsal segment of the midbrain is called the tectum, and the more central and ventral part is called the tegmentum. The pons lies anterior to the cerebellum and superior to the medulla, from which it is separated by a groove through which the abducens and the facial and acoustic nerves emerge. The medulla oblongata is the pyramid-shaped segment of the brainstem between the spinal cord and the pons. The lower half contains the remnants of the central canal. The posterior portion of the superior half forms the floor of the body of the fourth ventricle. The cerebellum is located in the posterior fossa of the skull, behind the pons and the medulla. It is separated from the overlying cerebrum by an extension of dura mater and the tentorium of the cerebellum. The tentorium of the cerebellum is oval in form, with its widest diameter along the transverse axis. It is composed of a small, unpaired central portion, called the vermis, and two large lateral masses—the cerebellar hemispheres (Fig. 3.2).

**Imaging Modalities for the Diagnosis of the Posterior Fossa**

The imaging modalities can be divided into noninvasive and invasive, as discussed in the following subsections.

**Ultrasound**

Since its introduction in the late 1950s, ultrasonography has become a very useful prenatal diagnostic tool in obstetrics and gynecology. The ultrasound scan is currently considered to be a safe, noninvasive, accurate, and cost-effective examination of the fetus. Many structural abnormalities in the fetus can be reliably diagnosed by an ultrasound scan, and these scans can usually be performed before the 20th gestational week. Common examples of neurologic malformations include hydrocephalus, anencephaly, and myelomeningocele. Ultrasound scan will be supplemented with Doppler ultrasound, and the three-dimensional (3D) and four-dimensional (4D) ultrasound techniques.
Plain X-Ray of the Skull

The oldest modality of imaging is a plain skull x-ray. This technique can show abnormalities and fracture of the skull, signs of chronic intracranial hypertension, and calcification. In cases of a dermoid cyst, a bone defect with sclerotic margins may be detected. But this technique is limited to direct or indirect bone lesions (Fig. 3.3).

Computed Tomography Scanning

Since 1976 examinations of the head have been performed with computed tomography (CT). As CT scan times have gotten faster, more anatomy can be scanned in less time. With an older CT scanner, the examination of the posterior fossa was limited because of the artifact produced from the surrounding thick bone. However, with the new technique of
Neuroimaging of the Posterior Fossa

Multidetector (4-row, 16-row) CT scanners, high-quality images can be reconstructed in multiple planes from a single volume data set (Fig. 3.4). Multidetector CT (MDCT) is rapidly becoming the new standard in radiologic imaging (Table 3.1).

The most important primary indications for CT imaging, including CT angiography and CT venography, in neuroradiology are acute head trauma, suspicion of acute intracranial hemorrhage, immediate postoperative evaluation for surgical treatment, shunted hydrocephalus, brain herniation, suspected mass or tumor, and acute cerebral infarction (Fig. 3.5).

Usually, CT is the imaging method of choice in patients with posterior fossa masses who often present with nausea, vomiting, ataxia, and other signs of increased intracranial pressure. CT is a quick, available, and relatively inexpensive method to assess neurologic emergencies including hydrocephalus, hemorrhage, and herniation syndromes.

Magnetic Resonance Imaging Scanning

In the early 1980s, magnetic resonance imaging (MRI) caught the attention of clinicians because of its ability to visualize abnormalities in the posterior fossa of the brain and in the upper cervical spine. Since its clinical development, the application of MRI sequences has rapidly evolved. New techniques and sequences are constantly being developed. To select the right technique, an understanding of the relationships

Fig. 3.2 Sagittal view of a T2-weighted magnetic resonance imaging (MRI) scan of the head of an 8-year-old boy. The indication for MRI examination was a continuous headache after the boy fell from his bicycle. The diagnosis was Chiari I malformation with herniation of the cerebellar tonsils through the great foramen into the cervical spinal canal.

Fig. 3.3 This figure demonstrates a fracture of the occipital bone on the right side in a 25-year-old man who arrived at the hospital unconscious and inebriated. (A) The injury is demonstrated in a conventional frontal plain film of the head. On the plain skull X-ray this fracture can be seen in projection at the right frontal sinus. (B) Three-dimensional VR scan images of the skull base, shows the fracture is in the occipital bone.
between the various imaging parameters is necessary. With all these techniques, it is evident that MRI is not a modality in which pathology will show itself. The imaging parameters need to be modified to yield the optimum contrast for the particular pathology under investigation (Table 3.2).

Furthermore, intrauterine MRI facilitates subtle prenatal neurologic diagnosis. The most important primary indications for MRI in neuroradiology are congenital disorders, posterior fossa lesions, early infarction, demyelinating and other white matter diseases, sensorineural deafness, and inflammatory lesions. The technical parameters of MRI examination vary with the presenting indication (Table 3.3).

Diagnostic Cerebral Angiography

In searching for an intracerebral aneurysm, the second diagnostic procedure to be performed immediately after emergency CT is selective cerebral angiography. Both the carotid and the vertebral arteries must be injected. External carotid arteries should also be visualized, particularly if intracranial angiography is negative, as subarachnoid hemorrhage may sometimes be due to a rupture not of an aneurysm but of a dural arteriovenous malformation. Angiography should aim at recognizing the aneurysm, its precise location, its size, the size of the neck, the relationship with the parent vessel, and its multiplicity. To achieve this goal, the ideal procedure is rotational angiography with 3D reconstruction.

Furthermore, cerebral angiography is useful in assessing the vascular supply of the tumor. With the wide availability of MRI, cerebral angiography is no longer the first option in brain tumor assessment.

The risks of cerebral angiography are bleeding at the site of the catheter insertion, allergic reaction to x-ray contrast (approximately 1/50,000 to 1/150,000 people), arterial embolism, and stroke.

Examples of pathologic findings in the posterior fossa are presented in Figs. 3.6, 3.7, 3.8, 3.9, 3.10, 3.11, 3.12, and 3.13.

Table 3.1 Sixteen-Row Multidetector Computed Tomography Scan Parameters

<table>
<thead>
<tr>
<th>Spiral Mode</th>
<th>mA</th>
<th>kV</th>
<th>Section Collimation (mm)</th>
<th>Rotation Time (s)</th>
<th>Scan Time (s)</th>
<th>Slice (mm)</th>
<th>Volume Contrast (mL)</th>
<th>Flow Rate</th>
<th>Delay (s)</th>
</tr>
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<tbody>
<tr>
<td>Head</td>
<td>330</td>
<td>120</td>
<td>16 × 0.75</td>
<td>1</td>
<td>3.76</td>
<td>3</td>
<td>100</td>
<td>2</td>
<td>50/60</td>
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<tr>
<td>Head angiography</td>
<td>200</td>
<td>120</td>
<td>16 × 0.75</td>
<td>0.5</td>
<td>10.45</td>
<td>4</td>
<td>100/120</td>
<td>3</td>
<td>4</td>
</tr>
</tbody>
</table>

kV, kilovolts; mA, milliamperes.
Table 3.2  Sequence and Parameters for Magnetic Resonance Imaging of the Head and Head Angiography

<table>
<thead>
<tr>
<th>Sequence</th>
<th>TR (ms)</th>
<th>TE (ms)</th>
<th>FA (°)</th>
<th>SN (n)</th>
<th>Orientation</th>
<th>ST (mm)</th>
<th>FOV (mm)</th>
<th>DF (%)</th>
<th>BR</th>
<th>Bandwidth (Hz/Px)</th>
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<tbody>
<tr>
<td>T2 se</td>
<td>3590</td>
<td>13</td>
<td>150</td>
<td>25</td>
<td>Tran</td>
<td>5</td>
<td>240</td>
<td>10</td>
<td>256</td>
<td>65</td>
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<tr>
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<td>5900</td>
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<td>150</td>
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<td>5</td>
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<tr>
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<td>14</td>
<td>90</td>
<td>25</td>
<td>Tran</td>
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<td>89</td>
</tr>
<tr>
<td>T1 + KM</td>
<td>522</td>
<td>14</td>
<td>90</td>
<td>25</td>
<td>Tran + cor</td>
<td>5</td>
<td>240</td>
<td>10</td>
<td>256</td>
<td>89</td>
</tr>
</tbody>
</table>

Head angiography

| FISP 3D        | 4.97    | 25      | 40     | Tran    | 0.83       | 81.3    | –37.5    | 512    | 65  |
| FLASH 3D       | 3.3     | 1.07    | 25      | Cor     | 1.3        | 360     | 20       | 512    | 390 |

Abbreviations: BR, base resolution; cor, coronary; DF, distance factor; FA, flip angle; FISP, fast imaging with steady precession; FLASH, fast low angle shot; FOV, field of view; KM, contrast medium; se, spin echo; SN, slice number; ST, slice thickness; TE, time of echo; TR, time of repetition; Tran, transversal; tse, turbo spin echo.

Parameters are for the Sonata MRI unit, with 1.5 Tesla, manufactured by Siemens, Washington, DC.

Table 3.3  Common Lesions of the Posterior Fossa

<table>
<thead>
<tr>
<th>Child</th>
<th>Adult</th>
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<tbody>
<tr>
<td>Extraaxial</td>
<td></td>
</tr>
<tr>
<td>Dandy-Walker malformations</td>
<td>Schwannoma</td>
</tr>
<tr>
<td>Epidermoid cyst</td>
<td>Meningioma</td>
</tr>
<tr>
<td>Arachnoid cyst</td>
<td>Epidermoid cyst</td>
</tr>
<tr>
<td>Rhabdoid cyst</td>
<td>Aneurysm</td>
</tr>
<tr>
<td>Intraaxial</td>
<td>Metastasis</td>
</tr>
<tr>
<td>PNET (medulloblastoma)</td>
<td>Arachnoid cyst</td>
</tr>
<tr>
<td>Pilocytic astrocytoma</td>
<td>Petrous lesions</td>
</tr>
<tr>
<td>Ependymoma</td>
<td>Infarction</td>
</tr>
<tr>
<td>Pontine astrocytoma</td>
<td>Metastasis</td>
</tr>
</tbody>
</table>

Abbreviation: PNET, permeative neuroectodermal tumor.


Fig. 3.6  Postcontrast axial T1-weighted MRI. At the right cerebello-pontine angle a contrast-enhanced acoustic neurinoma is located.
Fig. 3.7  Multiple sclerosis with lesions in the posterior fossa at the right cerebellopontine angle.

Fig. 3.8  Noncontrast CT shows bilateral symmetric calcification of the cerebellar hemispheres that could be idiopathic, familial cerebrovascular ferrocalcinosis, postinflammatory, congenital, or postanoxic/toxic.

Fig. 3.9  Edema of the cerebrum and cerebellum with compression of the mesencephalon by the tentorium of the cerebellum.

Fig. 3.10  Dandy-Walker malformation with these key features: large posterior fossa cyst, elevated confluence of the sinus (torcular Herophili), hypoplastic vermis, and hypoplastic cerebellar hemispheres.
Fig. 3.11  Thrombosis of the basilar artery (A) with an infarction area developing in 12 hours (B).

Fig. 3.12  Ruptured aneurysm of the basilar artery.

Fig. 3.13  CT scan (A) show a subtle hypodense area in the pons at the left side, whereas diffusion MRI scan (B) detects ischemia in this region with high signal intensity. (continued)
References

Neurosurgical Pathology of the Posterior Fossa

Marjorie Fowler

Diverse types of pathology occur in the posterior fossa. The specific pathologic lesion is diagnosed based on the location of the lesion; the age of the patient; the associated clinical history, including signs, symptoms, and duration; and the imaging studies. The most common types of lesions, besides trauma, that the neurosurgeon will encounter in the posterior fossa include neoplasms, both metastatic and primary; vascular lesions; and infections that present as masses. For the purposes of this discussion, this chapter divides the pathology into that associated with (1) parenchyma and leptomeninges, (2) cranial nerves, and (3) bone and dura mater. The discussion is limited to the most common lesions, and it focuses on gross pathology. The chapter also discusses some of the newer techniques that are emerging to aid neurosurgeons in the management of their patients.

**Parenchymal and Leptomeningeal Lesions Neoplasms**

One major type of lesion that neurosurgeons encounter in the posterior fossa is the neoplasm. The type of neoplasm is highly affected by the age of the patient. Most childhood nervous system neoplasms arise in the posterior fossa. In adults, although neoplasms certainly do occur in the posterior fossa, they more commonly present in the cerebral hemispheres.

The three most common posterior fossa neoplasms encountered in children are pilocytic astrocytomas, ependymomas, and medulloblastomas. All of these neoplasms tend to be well circumscribed and well demarcated from the parenchyma. The least likely of these to be well demarcated is the medulloblastoma, which may extend into and seed the subarachnoid space. Cerebellar pilocytic astrocytomas classically grow as cystic masses with mural nodules. In the brainstem, they are usually dorsal, exophytic, and not usually within the parenchyma. Both medulloblastomas and ependymomas tend to grow into and fill the fourth ventricle, presenting with symptoms of hydrocephalus. A gross clue to the cell type in these intraventricular neoplasms is the more common origin of medulloblastomas from the roof of the fourth ventricle and the origin of ependymomas from the floor. Growth within the subarachnoid space would lead one to suspect medulloblastoma, which has a strong tendency to grow and spread via the cerebrospinal fluid. It is more unusual for astrocytic neoplasms and ependymomas to spread in this manner.

Ependymomas, pilocytic astrocytomas, and medulloblastomas are generally soft, granular, and gray-pink. Primary nervous system tumors, in general, have a consistency that varies with their cellularity; the more cellular the neoplasm, the softer the consistency. Thus, medulloblastomas are generally the softest of these three, followed by ependymomas and then the mural nodule of the pilocytic astrocytomas. The less common childhood diffuse astrocytoma is usually firm and ill-defined, as it is in adults. As the grade of astrocytomas increases, they are generally softer, grayer, and may develop frank areas of hemorrhage and necrosis. Although high-grade astrocytomas may appear grossly to be more demarcated than their low-grade counterparts, when examined microscopically the tumor is generally found well beyond the apparent margins of the lesion. Glioblastomas, although not as common in the posterior fossa as in the cerebral hemispheres, do occur.

With older patients, the expected type of neoplasm changes, with an increase in the incidence of hemangioblastomas and diffuse astrocytomas and a decrease in the incidence of medulloblastomas and ependymomas. Also, as the patient ages, epidermoid cysts, choroid plexus papillomas,
lymphomas, and metastatic neoplasms are more commonly encountered. Hemangioblastomas are typically very red, spongy, and resilient, due to the large numbers of small blood vessels. They are characteristically well defined and frequently present as a mural nodule within a cyst. It is important when taking a biopsy from one of these slowly growing cystic lesions to be sure to send the mural nodules, rather than the cyst wall, to the pathology laboratory for frozen section. The often-florid gliotic tissue in the cyst walls frequently contains numerous Rosenthal fibers, which may lead to a mistaken diagnosis of pilocytic astrocytoma, if the pathologist is unaware that the biopsy is from the cyst wall rather than the mural nodule.

Epidermoid cysts characteristically have a thin cyst wall, appear somewhat translucent, and contain large amounts of pearly keratinous material. Hair is not usually seen within the cyst, but if it is seen, it would indicate that the diagnosis is more likely to be a teratoma. Teratomas and other germ cell tumors must always be considered when the tumor presents in the midline.

Choroid plexus papillomas of the fourth ventricle grossly appear to be papillary lesions that arise from the native choroid plexus of the fourth ventricle. Although they mostly grow as intraventricular neoplasms, they can rarely present as a mass at the cerebellopontine angle.

Primary lymphoma that is most commonly seen in the setting of immune suppression may present as multiple or single lesions. When associated with systemic lymphoma, it usually involves the leptomeninges rather than the parenchyma. Primary lymphoma more frequently involves the white matter than the gray matter. It generally is slightly gray, opaque, and duller than the surrounding white matter.

Metastatic neoplasms are one of the most common types of neoplasms encountered, especially in adults. The appearance of metastatic neoplasms is highly variable. They are generally sharply circumscribed and multiple. However, they may be soft or firm. If they have significant necrosis, they may appear cystic and mimic an abscess. They may be hemorrhagic, mucinous, or granular appearing.

**Vascular and Miscellaneous Lesions**

Vascular malformations are mostly cavernous angiomas and arteriovenous malformations, and they compose another group of lesions that, when seen in the posterior fossa, are most likely to be present in the cerebellum. Rupture of the vessels within these lesions leads to a hematoma of the parenchyma that may rupture into the subarachnoid space or the ventricle. Larger bleeds may destroy the lesion in the process. These vascular malformations are usually well circumscribed and may be surrounded by firm brown, yellow, or green discolored parenchyma, representing areas of old hemorrhage with gliotic repair. Other vascular lesions such as aneurysms, although less common than those in the anterior circle of Willis, may occur in the posterior fossa.

Arachnoid cysts occur in the leptomeninges of the posterior fossa. They are frequently asymptomatic, but may result in a mass effect if they are large. These cysts are generally very thin walled, contain clear colorless fluid, and rupture easily during surgery. They push the adjacent parenchyma to the side. In infants and small children, they may deform and enlarge the adjacent skull if they continue to grow.

**Inflammatory Lesions**

Inflammatory lesions may present as mass lesions of the parenchyma or the leptomeninges. Within the parenchyma, abscesses are the most common type of inflammatory lesion encountered. The earliest lesions present as areas containing soft loculated purulent material surrounded by red, wet, congested, edematous parenchyma. As the abscess matures, it forms a firm fibrous wall surrounded by parenchyma that is at first edematous and later gliotic and firm. The center remains soft, at first containing purulent material, and then later it becomes cystic, containing thinner fluid as it matures.

Granulomatous disease can affect either the parenchyma or the meninges and may be related to infectious organisms (e.g., fungi or mycobacteria) or sarcoidosis. Individual small granulomas are generally very firm, white, and discrete. As the granulomatous process evolves, the granulomas may become confluent, and, depending on the organism, may become centrally necrotic and soft. It is important to culture all lesions that have a possibility of being infections. If the lesion appears granulomatous, fungal and mycobacterial cultures are in order. Cultures done in the surgical suite are much preferred to those done in the frozen section rooms where contamination is much more possible. When the patient is immune suppressed, the neurosurgeon should also consider culturing for unusual viral agents.

Small early infarcts may also masquerade as tumors and require diagnostic biopsies. The earliest lesions are hemorrhagic because of dilated blood vessels. As the lesion evolves, it becomes edematous and then softens as it becomes necrotic. The eventual cystic infarct is generally not a problem to diagnose clinically, and thus is unlikely to result in neurosurgical biopsy.

**Cranial Nerve Lesions**

The main cranial nerve lesion that is encountered is the schwannoma, which most commonly arises from sensory nerves. It may be difficult to determine whether a lesion is arising from a nerve or from the dura, especially at the cerebellopontine angle. Schwannomas are well-circumscribed lesions that generally grow to the side of the nerve of origin. They can vary from white to yellow to red-brown, and frequently contain cystic areas and areas of hemorrhage. Although the original lesion is generally firm, the lesion may become softer as it develops areas of hemorrhage or cysts. The occurrence of schwannomas at a young age or in a bilateral location should prompt the suspicion of a genetic abnormality such as von Recklinghausen’s neurofibromatosis.
Bone and Dural Lesions of the Posterior Fossa

Neoplasms

Meningioma is the most common lesion arising from the dura mater. These lesions are usually firm nodular masses that indent and do not invade the brain. Because they are usually slow-growing tumors, they may grow to substantial size before presenting clinically. As in the case of schwannomas, they may also vary in color and consistency, although they are less likely to become cystic or to contain hemorrhage. The softer tumors are generally composed mostly of epithelioid-appearing meningothelial cells, and the firmer are tumors composed of spindle-shaped fibroblastic-appearing cells. They are usually shades of off-white but can have yellow or reddish areas. Meningiomas may penetrate into or through the bone and frequently cause an osteoblastic response with thickening of the bone. Locally aggressive hemangiopericytomas in the nervous system generally arise from the dura. Like meningiomas, these lesions are nodular and tend to push the adjacent brain to the side. They tend to be redder than the usual meningioma because of the dense vascularity, and thus will bleed more profusely.

Bony lesions may also involve the posterior fossa. Chordoma classically is seen in adults. The posterior fossa lesions most frequently involve the clivus, although they may grow predominantly on one side or the other or anteriorly into the sphenoid sinus. They are usually soft, lobulated, sometimes mucinous appearing, gray lesions that expand the bone and if growing into a space are covered by a fibrous capsule. Occasionally chordomas contain fragments of firm cartilaginous tissue or in some cases are composed almost entirely of cartilaginous tissue.

Chondrosarcoma occurs most frequently in patients in the fifth to seventh decades of life, and those arising in the posterior fossa are more common in females. In the skull base, the most common location is the temporo-occipital junction followed by the clivus and the sphenoethmoid complex. These tumors usually present with nervous system complaints, including headache, gait disturbances, and cranial nerve problems. Grossly, chondrosarcomas are infiltrative and nodular lesions have borderline histology.

Inflammatory Lesions

Lesions resulting from infections uncommonly involve the pachymeninges. When they do, they are almost always the result of infections of adjacent structures such as the middle ear, mastoid, or sinuses. Subdural empyemas are easily recognized by the purulent material within the space. Mucus-containing mucoceles usually arise and extend from the sinuses, and cholesterol granulomas penetrate through the petrous bone and are composed of rubbery brown or tan fibrous tissue.

Frozen Section Cautions (or How to Get the Best Pathologic Diagnoses)

Precise neuropathologic diagnosis in most cases requires a combination of the gross and microscopic pathology, the imaging studies, and the clinical history. Without all three components, the neurosurgeon risks making mistakes in diagnosis. When sending tissue for frozen section, it is essential that the neurosurgeon include the precise location of the lesion and the apparent tissue of origin, representative imaging studies, and an adequate history. The imaging characteristics of the lesion and adequate knowledge of the clinical symptoms, the progression of these symptoms, and other related history aid the pathologist in the diagnosis in the same way that they aid the neurosurgeon. Neither the pathologist nor the neurosurgeon can do his or her best job without this information.

New Advances in Neuropathology

In addition to the usual diagnostic immunohistochemical stains, there are two more recently developed types of testing that are beginning to play a very important role in neuropathology. The first of these are quantitative methods for estimating the rates of growth of tumors. The most widely used are those directed against the Ki-67 antigen. Semi-quantitative or quantitative estimation of the proliferative index using a computerized automated image analysis system is a useful adjunct to histologic grade of the tumor when determining the best treatment for patients, especially if the lesions have borderline histology.

The second group of tests that are beginning to influence treatment and occasionally diagnosis of neoplasms are molecular methods such as fluorescent in-situ hybridization, loss of heterozygosity, and microarray technologies used to map gene expression in tumors. Findings such as the loss or deletions of chromosomal regions on 1p and 19q in oligodendrogliomas that indicate a tumor that is particularly responsive to chemotherapy are likely to influence the treatment decisions made by the neurosurgeon. Newly emerging microarray technologies that make it possible to test for the expression of thousands of genes with one test promise new advances in tumor diagnosis and treatment in the near future.

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The posterior fossa contains the brainstem and cerebellum. The brainstem houses all the cranial nerve nuclei and many efferent and afferent fiber tracts that connect the brain with the rest of the body. The cerebellum is the major organ of coordination for all motor functions. Various neurologic entities can involve posterior fossa structures. These entities can affect the cranial nerves, cerebellum, and brainstem, alone or in combination. Some of the more common neurologic entities of the posterior fossa include multiple sclerosis (MS), acute disseminated encephalomyelitis, neurosarcoidosis, progressive multifocal leukoencephalopathy, Behçet disease, neurocysticercosis, Lyme disease, Whipple’s disease, and central nervous system (CNS) vasculitis. This chapter discusses these most significant neurologic entities.

**Multiple Sclerosis**

Multiple sclerosis is an immune-mediated disorder of the CNS that is usually characterized by a relapsing-remitting course, the presence of inflammatory lesions involving both the gray and white matter, and loss of the myelin/oligodendrocyte complex. MS is a CD4+ T-lymphocyte–mediated disorder that develops in genetically susceptible individuals following some as yet unidentified environmental exposure. Increased permeability of the blood–brain barrier (BBB) followed by transendothelial migration of monocytes and myelin basic protein–specific CD4+ T lymphocytes are among the significant features of the pathogenesis of MS. Indeed, disruption of the BBB has been demonstrated in vivo in acute and chronic active MS lesions using contrast-enhanced magnetic resonance imaging (MRI) (gadolinium–diethylenetriamine penta-acetic acid [Gd-DTPA] MRI). The presence of enhancing lesions on MRI is a strong indicator of the formation of new lesions and the expansion of former inflammatory lesions in relapsing-remitting multiple sclerosis (RRMS). Correlative MRI-neuropathologic studies of acute MS lesions have shown that these focially enhanced areas consist of fresh lesions, characterized by severe inflammation, edema, perivascular cuffing, and, in many cases, infiltration of mononuclear cells. Activated monocytes and myelin basic protein–specific CD4+ T lymphocytes are the major cell types in the perivenular infiltrates characteristic of MS. MS lesions show a unique predilection for the cerebral hemisphere, periventricular white matter, brainstem, cerebellum, optic nerves, and spinal cord.

Posterior fossa involvement in MS occurs frequently with development of demyelinating lesions in the cerebellum and brainstem. Cerebellar lesions of MS can cause disturbances in motor control, impairment of muscle tone regulation, and loss of coordination of skilled movements. Clinically, cerebellar involvement manifests with hypotonia, ataxia, dysarthria, tremor, and ocular motor dysfunction. Hypotonia is more common with acute cerebellar hemispheric lesions and is ipsilateral to the side of the cerebellar lesion. More often, hypotonia is noticeable in the upper limbs, particularly in the proximal musculature. Hypotonic limbs show decreased resistance to passive stretching of the muscles, and frequently exhibit pendular and diminished reflexes. Ataxia in MS results from loss or impairment of timing of the sequential contractions of agonist and antagonist muscles. Cerebellar ataxia refers to failure in the smooth performance of voluntary motor acts. This failure of performance affects speed, timing, force, and range of movements. Cerebellar dysarthria is characterized by abnormalities of articulation and prosody, either independently or in combination. Dysarthria may result from hypotonia and may affect intonation rather than articulation. Tremor in MS patients usually is disabling, and it may occur in patients who have normal strength. It is not unusual to observe tremor in MS patients with moderate cognitive impairment, for which tremor is termed cerebellar-cerebral. Tremor of MS patients is usually kinetic in nature. Cerebellar lesions in MS can cause various ocular motor disorders, particularly nystagmus. Gaze-evoked nystagmus, rebound nystagmus, abnormal optokinetic nystagmus, and periodic alternating nystagmus are among the most frequent ocular abnormalities that are observed in MS. Downbeat
General Considerations

Acute disseminated encephalomyelitis (ADEM) is another immune-mediated acute inflammatory demyelinating syndrome that occurs in association with immunization, vaccination, or systemic viral infections. ADEM is characterized by multifocal white matter involvement of the brain and spinal cord and diffuse neurologic signs. ADEM differs from MS in that it is generally a monophasic disorder with no relapses and with favorable long-term prognosis. Currently, there are no definitive diagnostic criteria to differentiate ADEM from MS. In MS demyelinating plaques, INO or medial longitudinal fasciculus (MLF) syndrome manifests with adduction weakness on the side of the MLF involvement and mono-ocular nystagmus of the abducting eye. In a young woman with INO, the first consideration in the differential diagnosis is MS. In patients with INO, unless the lesion is high enough to involve the midbrain, convergence is intact. Patients with INO may not have any complaints or may complain of diplopia or oscillopsia. INO is also commonly accompanied by skew deviation, with the higher eye on the side of the lesion. Acute vertigo is common in MS patients, and its recurrence may herald a new relapse. In severe cases, vertigo is associated with severe nausea and vomiting.

Acute Disseminated Encephalomyelitis

Acute disseminated encephalomyelitis (ADEM) is another immune-mediated acute inflammatory demyelinating syndrome affecting vestibulocerebellum.

Other clinical manifestations of MS lesions in the posterior fossa include trigeminal neuralgia, internuclear ophthalmoplegia (INO), and vertigo. Trigeminal neuralgia (TN) (also known as tic douloureux) is characterized by a sudden, stabbing, excruciating pain of paroxysmal nature. TN is more commonly unilateral and can manifest in the distribution of one or more of the divisions of the trigeminal nerve. Paroxysms of facial pain in TN are brief and usually last less than a minute. This painful syndrome is more common with advancing age, affects more females than males, and more commonly involves the right hemiface than the left hemiface. The presence of bilateral TN at the same period of time strongly raises the possibility of MS and necessitates further investigation. In MS patients, TN is possibly due to compression or irritation of the entry zone of the trigeminal nerve root by MS demyelinating plaques. INO or medial longitudinal fasciculus (MLF) syndrome manifests with adduction weakness on the side of the MLF involvement and mono-ocular nystagmus of the abducting eye. In a young woman with INO, the first consideration in the differential diagnosis is MS. In patients with INO, unless the lesion is high enough to involve the midbrain, convergence is intact. Patients with INO may not have any complaints or may complain of diplopia or oscillopsia. INO is also commonly accompanied by skew deviation, with the higher eye on the side of the lesion. Acute vertigo is common in MS patients, and its recurrence may herald a new relapse. In severe cases, vertigo is associated with severe nausea and vomiting.

Neurosarcoiidxosis

Sarcoidosis is a multisystemic granulomatous disease of unknown etiology. It involves the nervous system in 5% of patients. Although cutaneous and pulmonary systems are most commonly seen, neurologic manifestations of sarcoidosis can be potentially as disabling as other nonneurologic impairments. Neurosarcoidosis has a predilection for the base of the brain. The main clinical manifestations of neurosarcoidosis include cranial neuropathy, meningeal involvement, brain parenchymal disease, encephalopathy and seizures, myelopathy, peripheral neuropathy, and myopathy. Cranial neuropathy and meningeal involvement are the most prominent complications of neurosarcoidosis in the posterior fossa.

Single or multiple cranial neuropathies are common and the majority of patients have more than one cranial neuropathy. The facial nerve is most commonly affected. Facial nerve involvement occurs in 25 to 50% of patients with cranial nerve involvement, and this involvement is bilateral in one third of cases. The site of facial nerve involvement in neurosarcoidosis remains unknown; facial neuropathy frequently is associated with dysgeusia, which indicates that the lesion is probably proximal to the stylomastoid foramen. The next most commonly affected cranial nerve is the optic nerve. The next most common cranial neuropathy in neurosarcoidosis is eighth cranial nerve involvement, which occurs in 10 to 20% of patients and may be asymptomatic with only prolonged brainstem auditory evoked potentials, or can present with vestibular or hearing impairment. Optic neuropathy is less common in neurosarcoidosis and is observed in roughly 15% of patients. However, uveitis is a common ocular presentation of sarcoidosis. Optic neuropathy in neurosarcoidosis is usually due to local granulomatous invasion of the optic nerve or extraneural compression by a granulomatous mass. Papilledema is present in 14% of patients with ocular involvement. Olfactory nerve impairment in patients with neurosarcoidosis can manifest with anosmia or hyposmia and occurs in 2 to 17% of cases. Meningeal involvement presents with aseptic meningitis. It can manifest in 64 to 100% of patients with neurosarcoidosis and occasionally occurs as a meningeal mass lesion. Cerebrospinal fluid (CSF) examination in these patients reveals a mononuclear pleocytosis with elevated white blood cells and elevated protein levels.
Progressive Multifocal Neurocysticercosis

Meningitis can present as an acute monophasic illness or can be recurrent; it usually has a favorable outcome. Another complication of neurosarcoidosis is hydrocephalus, which can be either communicating or obstructive. Mechanisms of hydrocephalus formation include chronic basilar meningitis with obliteration of cerebrospinal flow, development of granulomatous lesions in the ventricular system causing obstruction, and compression of the cerebral aqueduct.

**Progressive Multifocal Leukoencephalopathy**

Progressive multifocal leukoencephalopathy (PML) is caused by a primary infection or is secondary to viral reactivation and infection of oligodendrocytes with JC virus. It almost always occurs in individuals who have underlying diseases that alter their cell-mediated immunity or who are iatrogenically immunosuppressed; currently, AIDS is the most common clinical syndrome that predisposes patients to PML. PML, in a diffuse and asymmetric distribution, typically involves, in descending order of occurrence, the cerebral hemispheres, cerebellum, brainstem, and spinal cord. PML is mainly restricted to the subcortical white matter and presents itself in a variety of neurologic syndromes. Patients are generally afebrile and without any manifestations of a systemic infection. Neurologic compromise slowly occurs over weeks to months. The well-recognized clinical triad of PML consists of visual impairment, motor weakness, and altered sensorium; prominent homonymous hemianopsia, monoparesis, or hemiparesis are among the typical initial manifestations of PML. Progressive loss of myelin in the cerebral visual centers may eventually cause cortical blindness. Subtle intellectual decline is a prominent early presentation, manifesting as personality change and progressing to blunted intellect, frank dementia, and eventually coma. Although patients usually present with cerebral symptoms, some patients present with brainstem or cerebellar involvement or seizures. The presence of ataxia in a patient with PML usually indicates the presence of lesions in the cerebellum and brainstem. Symptoms referable to both gray- and white-matter involvement can occur. JC virus, a 45-nm icosahedron, is one of the simplest of circular supercoiled double-stranded DNA viruses, which contains 5243 base pairs (bp) of DNA, and is mainly composed of VP1.

Most individuals are infected with JC virus at some point in their lives, and by age 5 at least 10% of individuals are JC-antibody–positive; by age 6, this number rises to 50% seropositive, and by age 14, 65% are seropositive. Eventually, 76% of individuals are seropositive for JC virus. Specific immunoglobulin G (IgG) antibodies to JC virus VP1 have been detected in 84.5% of adult control individuals. Initial infection with JC virus is asymptomatic, and the virus rapidly enters a latent phase in the kidney, other extraneural organs, and possibly the brain. In most individuals, the virus does not reactivate or cause problems. However, in individuals who develop diseases that suppress their cellular immunity or in individuals who are iatrogenically immunosuppressed, primary viral infection or reactivation of latent virus can occur. Immunosuppressed individuals commonly shed JC virus into their urine, as do 12.9% of leukemia and renal transplant patients.

**Behçet Disease**

Behçet disease is a chronic recurring multisystemic disorder that presents mainly with mucocutaneous and ocular manifestations. According to the work of the International Study Group, the major diagnostic criteria for Behçet disease include (1) oral ulcers recurring at least three times per year; (2) genital ulcers or scars; (3) eye involvement; (4) skin lesions (erythema nodosum, folliculitis, acneiform lesions); and (5) pathergy skin test observed by a physician. Minor diagnostic criteria include arthritis or arthralgia, deep venous thromboses, subcutaneous thrombophlebitis, epididymitis, family history, and gastrointestinal, CNS, or vascular involvement. Oral ulcers plus two other major criteria are required for the diagnosis.

In 4 to 49% of cases, Behçet disease involves the CNS, usually during active disease; in 5%, neurologic syndromes are the presenting manifestations. CNS involvement is usually polysymptomatic, with a relapsing-remitting or chronic progressive course with dementia, ataxia, or dysarthria, and a tendency to recur when immunosuppressive therapy is reduced. Brainstem or corticospinal tract signs (neuro-Behçet syndrome), acute confusion, increased intracranial pressure due to dural sinus thrombosis, meningoencephalitis, and isolated behavioral symptoms (psycho-Behçet syndrome) are among the most frequent neurologic manifestations. Rare cases with demyelinating lesions have been reported. Pure cerebellar or parkinsonian syndrome, peripheral neuropathy, and myelopathy are possible but uncommon. Neuropathologically, cerebral lesions of Behçet disease consist of multifocal necrotizing lesions with marked invasion of the inflammatory cells. In addition, necrosis may involve both the gray and white matter, probably secondary to vasculitis.

**Neurocysticercosis**

Neurocysticercosis is the most common parasitic infestation of the CNS, which is caused by the larval stage of *Taenia solium*, the pork tapeworm. Four forms of neurocysticercosis have been reported: meningal, parenchymal, ventricular, and mixed. Parenchymal lesions consist of small cysts, large cysts, and calcified lesions. Clinical manifestations of neurocysticercosis depend on the number of cysts, their location within the CNS, and the cyst’s state of health. Generally, cysticerci do not produce clinical symptomatology until the cysts begin to degenerate. Cyst degeneration begins from 2 to more than 10 years after the original infestation. As the cyst degenerates, cysticercal antigens leak into the adjacent brain or meninges and provoke a massive inflammatory response. The inflammatory reaction cause clinical manifestations such as
seizures, headaches, altered mental status, and focal neurologic signs like hemiparesis, visual loss, and paraparesis.

◆ Lyme Disease

Lyme disease is a multisystem infectious disease caused by the closely related group of spirochetes, known originally as *Borrelia burgdorferi* and now referred to as *Borrelia burgdorferi sensu lato*. The members of the *B. burgdorferi sensu lato* complex are transmitted by the bite of *Ixodes* ticks. Three members of this genus (*B. burgdorferi sensu stricto*, *B. garinii*, and *B. afzelii*) are pathogenic for humans. Skin, nervous system, joints, and heart are the most commonly affected organs. Up to 90% of infected patients develop the characteristic erythematous, macular, and painless rash, which evolves over days to become many centimeters in diameter. Following this acute and usually localized cutaneous infection, patients may develop subacute problems secondary to bacterial dissemination. Often, this dissemination is accompanied by a flu-like syndrome with fever, malaise, and diffuse aches and pains. In some, bacterial dissemination results in a multicentric erythema migrans. About 5% of patients develop cardiac conduction abnormalities. Others may develop a mild hepatitis or myositis, whereas some develop arthralgias or frank arthritis.

In 15% of patients, Lyme disease affects the nervous system, and these patients present with lymphocytic menigitis, cranial neuritis, and painful radiculitis. Every cranial nerve may be affected; however, the facial nerve is the most commonly involved cranial nerve. The painful radiculitis may resemble a mechanical monoradiculopathy or may be more disseminated, causing plexitis or even a diffuse disorder that may clinically resemble Guillain-Barré syndrome. In rare cases, patients (probably approximately 0.1% of infected and untreated individuals) may present with encephalomyelitis with prominent white matter involvement, frequently manifesting with a myelopathic picture. Studies of experimental infection in immunosuppressed primates have shown meningeal, but not CNS, parenchymal infection.

◆ Whipple’s Disease

Whipple’s disease is an infectious multisystemic disorder that mainly involves the small bowel and causes a malabsorption syndrome. Osteoarticular, cardiac, and CNS involvements are frequently observed in the context of Whipple’s disease. Whipple’s disease is caused by a gram-positive bacillus, *Tropheryma whippelii*. The pathogenesis and epidemiology, however, remain obscure. The most common neurologic manifestations of Whipple’s disease consist of cognitive decline, supranuclear gaze palsy, altered level of consciousness, psychiatric signs, and upper motor neurons signs. Postmortem studies have shown that these neurologic symptoms are associated with a granulomatous infiltration of the different levels of the CNS, particularly the brain, chiasm, pituitary gland, pons, and spinal cord. Cerebral lesions are essentially confined to the gray matter, preferentially the basal part of the telencephalon, the hypothalamus, and the thalamus.

◆ Central Nervous System Vasculitis

Vasculitis is defined as inflammation involving blood vessels with associated structural damage and usually tissue necrosis. Vasculitis primarily affecting the CNS, also known as isolated CNS angiitis, is confined to the brain, spinal cord, and their covering membranes. Isolated CNS vasculitis manifests with headache, signs of meningeal irritation, seizures, encephalopathic episodes, and hemispheric strokes. CSF examination is abnormal in more than 90% of cases, and neuroimaging of the brain and spinal cord reveals evidence of multifocal ischemic change of varying ages. Neuropathologic examination shows vasculitis of small and medium vessels of leptomeninges and underlying cortex with variable degrees of granulomatous changes. Occasionally giant cells may be present.

References

II

Disease-Based Management
Chiari malformations include a group of complex anomalies of the hindbrain with different etiology, pathophysiology, and clinical feature. These malformations range from the simpler to the more complex varieties of presentation, signifying their stages of appearance during embryologic differentiation and development. Various management modalities have been put forth to treat this entity. Over the years there has been a significant advance in understanding the pathophysiology, diagnostic modalities, and management strategies.

It was John Cleland (1835–1925), a Scottish anatomist, who first described a case of hindbrain malformations in 1883. In his article, “Contribution to the Study of Spina Bifida, Encephalocele, and Anencephalus,” Cleland described the pathophysiologic findings in nine infants at autopsy and two chick embryos. Later Hans Chiari (1851–1916), an Austrian pathologist who practiced medicine in Vienna, Prague, and Strasbourg, published his work in 1891. Chiari described the type I malformation as “peg-like elongation of tonsils and medial divisions of the inferior lobes of the cerebellum into cone-shaped projections which accompany the medulla oblongata into the spinal canal,” while sparing the medulla. In 1894, Julius Arnold (1835–1915), who studied under Virchow and Friedreich, described a case of an infant with spina bifida and elongation and descent of the inferior part of the cerebellum into the spinal canal. In 1907, Schwalbe and Gredig, while working in Arnold’s laboratory at Heidelberg, described four cases with meningomyelocele and alterations in the brainstem and cerebellum, and used the term “Arnold-Chiari.”

With the advent of magnetic resonance imaging (MRI), the intracranial anomalies associated with various Chiari malformations have become much clearer, and MRI became the imaging modality of choice in the diagnosis of Chiari malformations and the associated syrinx. MRI had been used to quantify the extent of tonsils below the foramen magnum. Aboulezz et al quantified the extension of tonsils below the foramen magnum based on MRI findings. The extension of tonsil below the foramen magnum is considered normal if it is less than 3 mm, borderline if it is between 3 and 5 mm, and clearly pathologic when it exceeds 5 mm. According to Barkovich et al, an MRI demonstration of tonsillar ectopia of <2 mm is probably of no clinical significance in the absence of syringomyelia. Further studies have shown that a small posterior fossa predisposes to hindbrain overcrowding, especially in patients with Chiari type I malformations. Further advances in MRI techniques, such as cine-MRI, greatly helped to study the alterations in the cerebrospinal fluid (CSF) flow at the foramen magnum and became a useful tool to evaluate the results of posterior fossa decompression radiologically.

**Pathogenesis of Chiari Malformations**

**Developmental Arrest**

The embryologic timing of the occurrence events responsible for the development of the Chiari malformation is not very clear. Daniel and Strich suggested that the abnormalities might develop early in the embryonic life. Based on their observation, they postulated that failure to form pontine flexure at the 6th week of fetal life may be one of the factors accounting for the marked elongation of the brainstem. They observed that in the initial part of fetal life (about 37 days), the length of hindbrain relative to forebrain is very great, and only after the formation of the pontine flexure does the hindbrain shorten. They also postulated the mechanism for the development of the kink. The cervical flexure is produced in the fourth week of fetal life and undone during the second month. If there is an elongated hindbrain during this process of straightening out the cervical flexure, it may result in a possible kink. The caudal displacement of the cerebellum and brainstem would result in an enlarged foramen magnum and a smaller posterior fossa that permits the tentorium to have a low insertion. However, this theory fails to explain the other associated spinal and cranial abnormalities.
The study by Marin-Padilla and Marin-Padilla showed that administration of vitamin A to pregnant hamsters induces types I and II Arnold-Chiari malformations. According to this theory, the primary defect is mesodermal, involving the cranial base. This has resulted in a small and short posterior fossa that is inadequate to contain the developing neural structures of the region. According to Sarnat, the crowding theory postulated by Marin-Padilla and Marin-Padilla would only impart a significant influence in late gestation. Nishikawa et al quantitatively measured the volume of the posterior cranial fossa and volume of the neural structures in the posterior cranial fossa. Their results strongly suggest the overcrowding hypothesis. Overcrowding of the neural structures (normal-sized hindbrain) in the underdeveloped posterior cranial fossa induces a downward herniation of the hindbrain. Hydrodynamic Mechanisms

According to Gardner, a balance exists between the pulsatile flow in the lateral ventricle choroid plexus and the fourth ventricle choroid plexus. The final position of tentorium is determined by the hydrodynamic effects of the anterior and posterior choroid plexus. According to Gardner, if the caudal end of the neural tube ruptures, the competing effect of the posterior choroid plexus is largely lost. The relatively increased effectiveness of the anterior choroid plexus causes the lateral ventricle to overdistend.

**Table 6.1 Description of Chiari Malformations**

<table>
<thead>
<tr>
<th>Type</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Chiari type I</td>
<td>• Tonsillar herniation greater than 5 mm below the foramen magnum&lt;br&gt;• Most commonly seen clinical entity&lt;br&gt;• Also referred to as adult-type Chiari malformation&lt;br&gt;• Syringomyelia common</td>
</tr>
<tr>
<td>Chiari type II</td>
<td>• Caudal descent of the cerebellar vermis along with brainstem and fourth ventricle&lt;br&gt;• Associated with myelomeningocele&lt;br&gt;• Hydrocephalus is commonly seen</td>
</tr>
<tr>
<td>Chiari type III</td>
<td>• Rarest and most severe form&lt;br&gt;• Occipital or high cervical encephalocele containing herniated cerebellar or brainstem tissue</td>
</tr>
<tr>
<td>Chiari type IV</td>
<td>• Marked hypoplasia or aplasia of the cerebellum&lt;br&gt;• Associated tentorial hypoplasia</td>
</tr>
</tbody>
</table>

Chiari malformations result from imbalances between the pulsating choroid plexus in the fourth ventricle and in the lateral ventricle. Overactive supratentorial pulsations might cause tentorial migration, resulting in the development of a Chiari malformation. This theory also fails to explain the occurrence of the other associated anomalies. The hydrodynamic theory does not explain the associated supratentorial anomalies.

**Oligo–Cerebrospinal Fluid or the Unified Theory**

This theory was proposed by McLone and Knepper. According to this theory, distention of the embryonic ventricular system is critical to normal development of the brain. If during the early part of the fetal development the presence of an open neural tube defect leads to escape of the CSF and fails to distend the embryonic ventricular system, the small posterior fossa is a consequence.

**Neuroschisis**

According to Padget, splitting open the neural tubes results in escape of the fluid from the neural tube and the formation of “neuroschistic blebs.” These blebs might heal totally, lead to formation of a spina bifida occulta, or might rupture with the eversion of neural cleft margins, leading to an open spinal lesion. Padget explained that a Chiari malformation is a sequel to a neural cleft, which allows fluid to escape from the neural tube. Embryonic microcephaly results, and the cerebellar primordia prematurely approximate and fuse in the posterior fossa, which is already small. Further development of the cerebellum in a small posterior fossa results in herniation out of the foramen magnum with subsequent obstruction of the fourth ventricle, producing hydrocephalus, whereas the microcephaly leads to folding and fusion at the level of the midbrain, resulting in aqueduct stenosis.

**Pulsion Theory**

According to this theory, fetal hydrocephalus pushes the contents of the posterior fossa downward from above. This theory was originally proposed by Chiari. This theory cannot explain the occurrence of the Chiari malformations without hydrocephalus and other associated anomalies.

**Neuroectodermal-Mesodermal Spatial Dyssynchrony**

Jennings et al hypothesized that the etiologic event responsible for the Arnold-Chiari malformation is the caudal displacement of the site of the initial fusion of the neural folds. They postulated that the normal zone of fusion at the third and fourth somites is displaced caudally below the third to fifth somite pairs, thus displacing the area of formation of the cervicomедullary junction. Their observations were based on the study of a 130-day-old human fetus with associated Arnold-Chiari malformation and thoracolumbar myeloschi-
sis, which revealed evidence of neuroectodermal mesodermal spatial dysynchrony.

**Traction Theory**

This theory, proposed by Penfield and Coburn in 1938, states that traction due to tethering of the spinal cord at a lower level prevents upward migration during early development, while the cerebellum and brainstem are pulled down as the vertebral column grows. Goldstein and Kepes tested the effect of spinal cord tethering on the development of Arnold-Chiari malformation in experimental animals and could not find any evidence of the malformation in the experimental animals that survived and reached adulthood. It is very unlikely that traction forces affect the cervical spinal cord and medulla. Traction theory fails to explain the lack of other observations, such as the medullary kink, and the spinal cord is not always tethered in patients with Chiari malformations.

**Molecular Genetic Hypothesis**

Sarnat postulated that Chiari malformation II is a disturbance of rhombomeric segmentation and ectopic expression in the embryonic hindbrain due to genetic mutation. Hox family genes in particular are implicated because they not only program hindbrain segmentation but also are important in the development of basioccipital, exoccipital, and supraoccipital bones of paraxial mesoderm origin. The same group studied several markers like vimentin in the ependyma of patients with Chiari II malformations. The results of the study showed that vimentin is focally upregulated only in areas of dysgenesis. Based on their observation, they speculated that the focal upregulation of vimentin could be a secondary event as a result of defective expression of another gene. This observation may also support a molecular genetic hypothesis.

◆ **Classification of Chiari Malformations**

**Chiari 0**

Iskandar et al in 1998 reported resolution of syringohydromyelia without hindbrain herniation in five patients after posterior fossa decompression. They referred to such cases of syringomyelia without hindbrain herniation as the “Chiari 0” malformation. Later the same researchers showed that the contents of the posterior fossa are compromised and distorted in these patients with syringohydromyelia without hindbrain herniation. They suggest that a smaller than normal posterior fossa is just enough to compromise CSF egress out of the cranium.

Mihorat feels that cerebellar tonsils are paramedian structures, and on midsagittal MRI scans minimal tonsillar herniation might be missed. Mihorat et al, in their study of 364 patients with Chiari I malformation, showed that in these patients the CSF volume in the posterior fossa is significantly smaller, and tonsillar herniation of less than 5 mm does not exclude the diagnosis of Chiari I malformation. Milhorat suggests that a better description of such cases with “Chiari 0” malformation would be “borderline Chiari malformations.” Rather than classification issues, one may have to investigate these patients without significant tonsillar herniations using cine-MRI scans to document disturbance in the CSF circulation in the posterior fossa before subjecting them to hindbrain decompression.

**Chiari I**

Chiari I malformation is characterized by caudal displacement of the cerebellar tonsils by more than 5 mm into the cervical spinal canal below the plane of the foramen magnum. According to Mihorat et al, tonsillar herniation of less than 5 mm does not exclude the diagnosis of Chiari I malformation. This is the most commonly seen clinical entity and is described as synonymous with tonsillar herniation. It is usually referred to as adult-type Chiari malformation due to its incidence in the second or third decade of life. The exact incidence of Chiari I malformations is not known. Meadows et al reviewed 22,591 patients who underwent MRI of the head and cervical spine. Of these only 175 patients (0.8%) had Chiari I malformation.

**Associated Anomalies**

Craniovertebral anomalies are frequently seen in patients with Chiari malformation. In the study by Mihorat et al, the most common bone anomalies were reduced height of the supraocciput (84%), increased slope of the tentorium (82%), reduced length of the clivus (49%), retroflexion of the odontoid (26%), and basilar invagination (12%). Tubbs et al graded the posterior angulation of the odontoid process in patients with Chiari I malformation and found that higher grades of posterior angulation are more often associated with syringomyelia. Other commonly seen osseous abnormalities are platybasia, midline occipital keel, remnants of proatlases, atlantoaxial dislocations, Klippel-Feil anomaly, empty sella, and clival concavity.

The incidence of syringomyelia associated with Chiari malformation varies between 50% and 75%. The common location of the syrinx is cervical followed by the thoracic spinal cord (Fig. 6.1), occasionally with holocord syrinx (Fig. 6.2). A thickened dural band posterior to the arch of C1 may be frequently encountered during surgery. Chiari I malformation is associated with hydrocephalus in 7 to 10% of cases. An occasional patient might show an elongated brainstem with medullary kinking (Fig. 6.3).

**Chiari 1.5**

This group comprises patients with tonsillar ectopia as seen in Chiari I malformation. In addition, they exhibit caudal descent of the brainstem (Fig. 6.4). This has been specifically referred to as “Chiari 1.5.” No other sign or symptom was peculiar for Chiari 1.5 malformations.
Chiari II malformation is characterized by caudal descent of the cerebellar vermis along with the brainstem and fourth ventricle.

**Associated Anomalies**

Myelomeningocele (MMC) is nearly always associated with Chiari II malformation. Luckenschadel skull is an ossification disorder in which the fetal skull appears fenestrated. It is almost always associated with Chiari II malformation and MMC. Other anomalies associated with Chiari II malformation are petrous scalloping, enlargement of the foramen magnum, fenestrations of the falk, falk hypoplasia, and hypoplastic tentorium with wide incisura. The inion is lower, and there may be an occipital keel with less frequency of basilar invagination. The upper cervical spine shows Klippel-Feil anomaly with hypoplastic posterior arch of C1 and scalloped dens.

Other abnormalities associated with Chiari II malformation, as detected in MRI scans, are hypothalamic adhesions (48.6%), low anterior commissure (38%), abnormalities of the corpus callosum and hippocampal commissure (57%), callosal ridge (60%), cortical posterior stenogryia (72%), gray matter heterotopias (19%), hippocampal abnormalities (85%), and atypical sulcation of the adjacent temporomesial cortex (93%). The midbrain is elongated with a shortened quadrigeminal plate. Peaking is seen with fusion of the colliculi (tectal beaking). Callen et al observed tectal beaking in 66% of patients with Chiari II malformation on prenatal sonography. The caudal displacement of the lower brainstem may create a medullary kink in the cervical canal.

Hydrocephalus is seen in 90% of patients with Chiari II malformation. The other ventricular abnormalities found in patients with Chiari malformation II are “shark-tooth deformity” of the third ventricle, colpocephaly, beaking of the
Congenital Chiari Malformations

The vast majority of type II patients exhibit syringohydromyelia, sometimes with exophytic components mimicking arachnoid cysts (Fig. 6.5). In approximately 6 to 15% of cases, there is an associated split-cord malformation.47,48

Chiari III

Type III is the rarest and most severe form of all the Chiari malformations. It is characterized by occipital or cervical encephalocele along with the above intracranial abnormalities seen with type II malformation.49,50 The sac of encephalocele contains dysmorphic and ischemic neural elements (Fig. 6.6).
Other cranial abnormalities, such as small posterior fossa, low tentorial attachment, scalloping of the clivus, tectal beaking, and dysgenesis of severely deformed corpus callosum, are seen.49

Chiari IV

This type is characterized by marked cerebellar hypoplasia or aplasia and tentorial hypoplasia. There is no hindbrain herniation.

◆ Clinical Presentation

Of all the Chiari malformations, type I is most commonly encountered. After the advent of MRI scans, several atypical, silent, or incidental entities have been reported. At Louisiana State University Health Sciences Center in Shreveport, Louisiana, the experience with these malformations amounts to 95 patients, of whom approximately 20% were children, although 9.5% presented after 55 years of age. There was female preponderance: 72% were females and 28% were males.

Type I Malformations

The signs and symptoms in patients with Chiari I malformation are generally related to the compression of the neural structures by the herniated tonsils51 and by the presence of an associated syrinx. Clinical manifestations can be grouped into cranial nerve dysfunction, long tract deficits, and cerebellar dysfunction.

Suboccipital headache is one of the common presentations of the Chiari I malformation. These headaches are usually exaggerated by physical exertion and the Valsalva maneuver.7 Ocular disturbances such as retro-orbital pain, floaters or flashing lights, blurred vision, photophobia, and diplopia are frequently reported by patients.7 Otoneurologic disturbances such as dizziness, disequilibrium, tinnitus, decreased hearing or hyperacusis, vertigo, and oscillopsia are commonly seen.7,52 Downbeat nystagmus is characterized by slow upward drift and fast downward phases.53 One of the common causes for downbeat nystagmus is Chiari malformation.53

Lower cranial nerve, brainstem, and cerebellar disturbances are seen in up to 52% of patients.7 The most common symptoms are dysphagia, sleep apnea, dysarthria, and incoordination.7 Objective signs are absence or impairment of the gag reflex, vocal cord paralysis, facial hypoesthesia, dysmetria, and truncal ataxia. Long tract signs include posterior column dysfunction and pyramidal weakness.

Patients might present with clinical features due to an associated syrinx. The signs and symptoms vary with the location and extent of the syrinx. The most common symptoms are muscular weakness, paresthesias, dysesthesia, nonradic-
ular segmental pain, analgesia, spasticity, trophic phenome-
on, and poor position sense.7 Dissociated sensory loss en-
tails loss of pain and temperature sensation with preservation
of light touch. An expanding syrinx disrupts the decussating
sensory fibers passing from the dorsal horn to the opposite
lateral spinothalamic tract. Disassociated sensory loss is not
always a necessary finding for the diagnosis of syringomy-
elia.54 These patients might have features of both upper
motor neuron and lower motor neuron dysfunction.

The presenting features in children sometimes differ from
the presenting features in adulthood. Children under 3 years
of age might present with oropharyngeal dysfunction, and
slightly older children (3 to 5 years of age) might present
with scoliosis, headache, or neck pain.55

**Type II Malformations**

The first sign in these patients would be an MMC, taking into
account that Chiari II malformation is universally associated
with MMC.45 Infants and children younger than 2 years of
age present most frequently with cranial nerve and brain-

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Fig. 6.6  (A–C) An occipital encephalocele in a young child. Intraoperative pictures show
the atrophic neural elements inside the meningeal sac and after excision.
stem dysfunction, and the most common and potentially fatal symptom in these age groups involves respiratory difficulties. Other symptoms in infants include hypotonia, opisthotonus, nystagmus, weak cry, and developmental delay. Symptoms in older children are not life threatening and tend to be more insidious, and features of cervical myelopathy are the hallmark finding in these age groups. Older children can also present with ataxia, occipital headache, neck pain, and features of syringomyelia.

**Type III Malformations**

These children usually present with breathing difficulty, swallowing problems, seizures, developmental delay, weakness and hyporeflexia in the upper extremities, and spasticity in the lower extremities.

**Diagnostic Imaging**

Magnetic resonance imaging is the diagnostic test of choice. It provides simultaneous visualization of the cervical spine and the entire neuraxis. Syringomyelia and syringobulbia are easily demonstrated. MRI facilitates the identification of the venous anomalies of the posterior fossa. The level of the torcular and the transverse sinus are important to plan the posterior fossa decompression. In cases of Chiari III malformation, MRI enables the surgeon to identify the amount of neural tissue in the encephalocele and the position of the brainstem. The anomalies associated with various Chiari malformations were described above.

Dynamic studies are useful to demonstrate CSF flow around the foramen magnum, and can show obstruction by hindbrain structures or evidence of syringomyelia. Cine-MRI is helpful in demonstrating a CSF flow disturbance at the foramen magnum. McGirt et al demonstrated that normal preoperative hindbrain CSF flow is an independent risk factor for treatment failure after decompression for Chiari I malformation regardless of the degree of tonsillar ectopia. In another study the presence of decreased CSF flow both ventral and dorsal to the cervicomedullary region was associated with improved response to posterior fossa decompression. Flow studies and cine-mode MRI are useful for postoperative evaluation following decompressive surgery. Flow obstruction, decreased velocities, and a shorter period of caudal CSF flow in the foramen Magendie and foramen magnum in patients with more than 5 mm of tonsillar herniation are reported.

**Brainstem Auditory Evoked Potentials**

Zamel et al studied the role of brainstem auditory evoked potential (BAEP) monitoring during posterior fossa decompression. They showed that a predominant improvement in central conduction in most of their patients occurred during the period of bony decompression. Similarly, Anderson et al found that improvement in conduction velocity occurs after bone decompression and division of the dural band. The predictive value of the improvement in BAEP needs to be assessed further in larger studies to establish its role in the surgical management of Chiari malformations.

**Surgical Treatment and Outcome**

**Chiari I Malformations**

With the advent of MRI an increasing number of patients are being diagnosed with asymptomatic Chiari I malformation. An important controversy in the management of asymptomatic Chiari I malformation concerns the role of prophylactic surgery in asymptomatic patients. Novegno et al studied the natural history of Chiari I malformation. Their findings suggest that a conservative approach is indicated in both asymptomatic and slightly symptomatic patients, with periodic follow-up. Haroun et al reported that in their survey of the members of the pediatric section of the American Association of Neurological Surgeons, the majority of the respondents rejected the routine use of prophylactic surgery for Chiari I malformation.

**Cerebrospinal Fluid Diversion Procedures**

In patients with hydrocephalus, the CSF diversion procedure is considered the best initial treatment option. Ventriculoperitoneal (VP) shunting is the most common CSF diversion procedure. Recently, endoscopic third ventriculostomy has been shown to be efficacious in patients with hydrocephalus associated with Chiari I malformation. Following the CSF diversion procedure, if the symptoms persist or if the syrinx does not show improvement or enlarges, then one needs to direct one’s attention to a Chiari decompression.

**Posterior Fossa Decompression**

**Surgical Technique**

In brief, this operative procedure is performed with the patient in the prone position. A midline skin incision is made from 2 cm above the inion up to C2. The incision is deepened down to the bone along the midline avascular plane. Self-retaining retractors are placed, and the musculature is gently retracted. At the level of the superior nuchal line, the suboccipital muscles are divided with a thin rim attached to the bone. The muscles are separated subperiosteally from the underlying occipital squamous bone. Inferiorly the muscular attachment to the retractor arch of the atlas is detached by sharp dissection. Weitlaner is placed superiorly and inferiorly. While separating the muscle and soft tissue, one needs to be extremely careful to avoid injury to the vertebral artery. The pulsations of the vertebral artery can be seen and felt. Any brisk bleeding from the venous plexuses can be controlled with Gelfoam, and it is a warning sign that the verte-
bral artery is nearby. We prefer a small craniotomy (5 cm). The foramen magnum rim is removed with either a high-speed drill or a Kerrison punch. The dural arch of C1 is excised. The dura is opened in a Y-shaped manner. At this stage, the microscope is brought into the field. The arachnoid is opened separately. The tonsils are usually tongue-like and smooth, very often pulling down the posterior inferior cerebellar artery along with them into the spinal canal. Care has to be taken to avoid injury to the artery while dissecting the tonsils from the arachnoid adhesions (Fig. 6.7). We routinely shrink the tonsils with bipolar coagulation. A fascia lata graft is used for duraplasty, and the dura is closed in a watertight manner.

◆ Outcome

In the vast majority of centers, the procedure has yielded good results. Postoperative MRI established the efficacy of this method in providing patent CSF pathways around the craniocervical junction.65,66 Park et al65 found improvement in all their patients over long-term follow-up. Klekamp et al66 reported an 87% decrease in the size of the syrinx in their series of 131 patients, whereas Garcia-Uria et al68 found 50% improvement and 75% stabilization over a follow-up period of 5 to 10 years, respectively. Fischer69 reported resolution of the syrinx in 93% of cases in a literature survey. Anderson et al62 reported postoperative improvement in BAEPs following craniocervical decompression. Not only did syringomyelia and symptoms of hindbrain compression improve, but also scoliosis, which is seen with long segment syringomyelia, responded to posterior cranial fossa (PCF) decompressive surgery.70,71

◆ Controversies

There are several controversies regarding intradural procedures such as dissection of the arachnoid overlying the tonsils, shrinkage of the tonsils by bipolar coagulation, and subpial resection of the tonsils. A recent study found that tonsillar management has no significant effect on improvement of syringomyelia.72

In some studies the results are good without any intradural manipulation.73-75 Genitori et al74 studied the role of posterior fossa bony decompression without dural opening in the management of symptomatic children affected with Chiari type I malformation. With this approach they achieved improvement in symptomatology in a high percentage of cases (97.2%). A similar strategy was adopted by James et al.76 Zamel et al75 described their experience with neurophysiologic intraoperative monitoring of BAEPs during posterior fossa decompression surgery for the management of Chiari I malformation. Their findings suggest that posterior fossa decompression with bone removal alone significantly improves conduction time in most pediatric patients with Chiari I malformation. The use of duraplasty facilitated a small improvement in conduction time in only 20% of the patients.61

In another study, the authors adopted a dura-splitting decompression for pediatric Chiari I malformation.73 The early clinical results were good, and this technique significantly reduced resource use.73

Others have described foramen magnum decompression with removal of the outer layer of the dura as a treatment option for syringomyelia occurring with Chiari I malformation.75,77

Recently, Sindou and Gimbert78 described decompression for Chiari type I malformation by extreme lateral foramen magnum opening and expansile duraplasty with arachnoid preservation. They claim that this type of craniocervical
decompression achieved the best results with minimal complications and side effects.

Chiari II Malformations

This is a difficult entity for decision making as well as for operative detail. There is no uniform consensus on the management of these lesions. The problem lies in choosing the appropriate patients for surgery and the type and extent of the surgical intervention. In patients presenting with symptomatic Chiari II malformation due to medullary dysfunction, early surgical intervention may be life sustaining.79

Ventriculoperitoneal Shunting

According to Tubbs et al.,79 a properly functioning ventricular shunt can obviate the need for hindbrain decompression. The difficulty lies with the evaluation of shunt function in these patients. In the presence of a shunt, progressive hydro-syringomyelia occurs due to shunt malfunction, unless proven otherwise.79 Periodic evaluations for brainstem malfunction need to be performed, especially by otolaryngologists and internists focusing on respiratory function, swallowing, and speech.

Posterior Fossa Decompression

In cases where progressive brainstem compression is clinically evident, posterior fossa decompression is indicated. One needs to keep in mind that the foramen magnum is wide in these patients and the torcular and the transverse sinus are low lying. Extensive bone removal may not be necessary and is dangerous if the sinuses are entered inadvertently.79 Patients may need postoperative cervical spine evaluation, because there is a possibility of delayed cervical instability.80 After midline durotomy, the arachnoid adhesions need to be lysed, and cerebellar vermis is released from compressing the cervicomedullary junction. Some patients require coagulation of excessive vermis that might be obstructing CSF flow. The foramen Magendie is carefully dissected free of the adhesions, and the CSF pathways are opened up.

Outcome

Posterior fossa decompression relieves the symptoms of syringomyelia in more than 75% of the patients and is preferred over syrinx shunting.81 If the syrinx persists and the patient is symptomatic, then a syringosubarachnoid, syringopleural, or syringoperitoneal shunt could be considered.79 Intrauterine repair of a myelomeningocele may reduce the extent of hindbrain abnormalities.82 The intrauterine repair of a myelomeningocele might reduce the fluid loss and thus the hindbrain herniation through the foramen magnum.

Chiari III Malformations

Much of the evidence comes from a relatively small number of series with very few cases. According to a recently published literature review, there are only 34 cases with Chiari III malformation.57 These patients may require VP shunting for hydrocephalus, repair of an encephalocele, and management of other congenital anomalies. If there is a hydrocephalus, VP shunting should be performed first.57 Snyder et al65 managed a patient with Chiari III malformation with an initial VP shunt. The shunt allowed the brainstem and the cerebellum to regress into the cervical spinal canal. A delayed closure of the cervical encephalocele was performed at 30 months of age. In another study, hydrocephalus was noted in seven of eight patients with Chiari III malformation, and five of them with initial hydrocephalus underwent shunting followed by closure of the encephalocele.57

The repair of the malformation follows the same principles that are applicable to open neural tube defects. The amount of neural tissue in the encephalocele varies. Great care is required to preserve neurologic function. MRI enables the surgeon to identify the amount of neural tissue in the encephalocele and the position of the brainstem and venous anomalies.50 Delayed treatment of hydrocephalus, initial severity of neurologic deficits, the amount of brain tissue within the excised encephalocele, the presence of intermittent apnea, and surgery in the newborn period were established as prognostic factors for a poor outcome.57

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Trauma to the Temporal Bone

Prasad Vannemreddy and Anil Nanda

Samuel Thomas von Soemmerring (1755–1830), a German medical student, established the present classification of cranial nerves in 12 numbered pairs two centuries ago based on the different foramina in the base of the skull through which the nerves exit the cranial cavity. The first description of traumatic cranial nerve injury appears in the Edwin Smith Papyrus, and describes a patient with a facial droop following head injury. This is probably the first account of traumatic facial nerve paralysis recorded.

◆ Applied Anatomy of the Facial Nerve

The two roots of the facial nerve arise from the pons lateral to the recess between the inferior olive and the inferior cerebellar peduncle. The sensory root is called the nervus intermedius and lies close to the eighth cranial nerve until it reaches the internal auditory meatus. Inside the internal auditory canal, the facial nerve is connected to the vestibular nerve by the fibers of Rasmussen. At the lateral fundus of the meatus it pierces the arachnoid and the dura to enter the facial canal superior to the transverse crest, separated from the superior vestibular nerve by Bill’s bar and accompanied by the labyrinthine branch of the anterior inferior cerebellar artery (AICA). This labyrinthine segment runs across the axis of the petrous pyramid to the geniculum and forms the geniculate ganglion and then turns 130 degrees as it becomes the tympanic or horizontal segment. The pyramidal part connects the horizontal and the mastoid segments and gives off the nerve to the stapedius muscle. The nerve now descends to the stylomastoid foramen and gives off the chorda tympani nerve (Fig. 7.1).

The temporal bone is developed from five developmentally and morphologically distinct parts. The cartilaginous petromastoid part develops to preserve the precise orientation of the membranous labyrinth. The mastoid serves as an anatomic door to the petrous pyramid, the base of which arbitrarily corresponds to the suture between the petrous and squamous elements. The apex of the petrous pyramid is blunt and irregularly angled between the greater wing of the sphenoid and the basiocciput bone. The anterior surface partly forms the floor of the middle cranial fossa and is continuous with the cerebral surface of the squamous part adapted to the inferior temporal gyri. Laterally the apex forms the roof of the vestibule and part of the facial canal. Between the arcuate eminence (lodging the superior semicircular canal) and the squamous temporal bone is the tegmen tympani, which forms the roof of the mastoid antrum. The posterior surface of the petrous pyramid is an anterior part of the posterior cranial fossa and is continuous with the internal mastoid surface. The superior border of the petrous bone is grooved by the superior petrosal sinus and gives attachment to the tentorium cerebelli along this groove. The posterior border has a gutter for the inferior petrosal sinus. At the junction of the petrous and squamous parts are two canals separated by a thin bone; the upper canal contains the tensor tympani and the lower, the auditory canal (Fig. 7.2).

◆ Etiology

The causes of facial nerve palsy vary. Blunt head trauma in motor vehicle accidents or from gunshot wounds is responsible for the majority of lesions. Other causes include penetrating stab wounds and operative trauma. Symmetric middle cranial neuropathies can result from crushing injuries of the skull. Blunt head trauma is accompanied by basilar fractures and involves the cranial nerves near the fractures. In gunshot wounds the bullet has a trajectory—horizontal in suicidal attempts and anteroposterior in homicidal injuries. Avulsion and stretching of the nerve roots can occur with acceleration and deceleration trauma and in blunt injuries. These forces damage the nerves at their points of fixation or angulation. Minor trauma may precipitate palsy in a nerve previously stretched by tumors.
Epidemiology

The incidence of cranial nerve injury varies by age group and demographics. Keane and Baloh provide a comparative tabulation of the incidence of cranial nerve injury after trauma. Facial and eighth nerve injuries have the greatest incidence, closely followed by temporal bone fractures. Road accidents are responsible for 50% of temporal bone fractures, and nearly 22% of all skull fractures are of the temporal bone; 7 to 8% of the latter have associated facial nerve injury. The association between types of temporal bone fracture and facial nerve injury is described in detail below.

Clinical Manifestations

In general, the olfactory, facial, and vestibulocochlear nerves are damaged most frequently following blunt head trauma, with the lower cranial nerves (IX, X, XI, and XII) being the least commonly injured. In children younger than 10 years of age, the abducent and facial nerves are commonly injured. Because of the close proximity of the facial nerve with the vestibulocochlear nerve, the combination of hearing loss with facial nerve paralysis might be seen in cases of middle cranial fossa fractures with cerebrospinal fluid fistula. Although a facial nerve lesion is easily detected, manifestation of vestibulocochlear nerve injury in states of obtunded sensorium may be delayed. However, maintaining a strong clinical suspicion helps in detecting such combinations early. The type of temporal bone fracture sometimes correlates very well with the nerve lesions. The various degrees of facial nerve paralysis are described by House and Brackmann in their classification system. By and large, traumatic injury of facial nerve in the temporal bone produces infranuclear palsy (also known as Bell's palsy).

Etiopathologic Correlation Between Facial Nerve Injury and Temporal Bone Fractures

The long, tortuous, intraosseous course of the facial nerve in the temporal bone makes this nerve highly susceptible to injury in temporal bone fractures. It is also injured in penetrating and blunt trauma to the head and face. Middle ear injury due to penetrating objects can easily damage this nerve. In approximately 50% of cases of transverse temporal bone fractures, the facial nerve within the internal auditory canal is damaged. With longitudinal fractures, the nerve is not directly involved, but a delayed paralysis may ensue secondary to edema, and facial nerve injury is seen in nearly 20% of cases. Temporal bone fractures are classified as transverse, longitudinal, and mixed, based on the relation of the fracture line to the axis of petrous pyramid.

Longitudinal fractures follow a blow to the temporoparietal bone and start from a weak point in the squama or the mastoid part of the temporal bone. They constitute approximately 80% of all temporal bone fractures and present with the classic Battle's sign of bloody otorrhea and a bony step-off in the external auditory canal. The fracture line traverses anteromedially through the middle ear, disrupts the ossicular chain, and is deflected anteriorly by the hard otic capsule. It may terminate in the middle cranial fossa or extend medially to the sphenoid bone, crossing the midline in 30% of cases. Transverse fractures account for only 20% of temporal bone injuries. These require a force great enough to break the occipital bone at the foramen magnum, reaching the petrous
Fig. 7.3  (A) The types of temporal bone fractures. The arrow points to the area of impact to create the tangent of force for a given fracture. The weak squama of the temporal bone requires relatively a small force to yield and produce a longitudinal fracture (L), which runs parallel to the length of the petrous pyramid. This courses along the weak areas, such as air cells, the middle ear cavity, and skull base foramina. In contrast, a greater force is required along the thick occipital bone traversing the thick structure of the posterior cranial fossa to produce a transverse fracture (T) of the petrous bone rupturing the hard otic capsule also.  (B) The longitudinal temporal bone fracture originates in the weak squamous temporal bone either the external auditory canal (A) anteriorly or the air cells of the mastoid sinus (M) posteriorly. With an anteriorly placed fracture, the tympanic membrane is ruptured, resulting in bloody otorrhea, whereas posterior fractures present with Battle’s sign and an intact tympanic membrane. The fractures course along the middle ear and reach the foramen lacerum deflected by the tough otic capsule, thus sparing the inner ear.  (C) The transverse fracture requiring much greater force originates in the thick occiput bone and reaches the foramen magnum from where the force reaches the floor of the middle cranial fossa, fracturing the petrous pyramid perpendicular to its long axis, disrupting the otic capsule, thus damaging the inner ear and facial nerve. The fracture sometimes can pass lateral or medial to the otic capsule. With the medial course of the fracture, the internal auditory canal and its contents are at risk.
pyramid. The intensity of such a force usually causes severe brain damage and disrupts the tough otic capsule, damaging both vestibular and cochlear components of the labyrinth. This fracture line generally spares the middle ear and has fewer external otologic findings. Facial nerve injury occurs in approximately 50% of cases, and the labyrinth is usually damaged by the fracture.

Mixed fractures have a combination of these findings and are thought to be less common. However, 65 to 80% of fractures have been reported to be neither longitudinal nor transverse, with the fracture lines traversing the petrous pyramid in an oblique way. Severe head injury can also avulse the nerve root from the brainstem; the patient usually has features of brainstem dysfunction. Facial paralysis is usually striking in cases of infranuclear paralysis. Injury to the vertical mastoid segment produces loss of taste on the ipsilateral anterior two thirds of the tongue. A horizontal segment (middle ear portion of the facial nerve) injury results in loss of the stapedius reflex (hypersensitivity to loud sound) and ipsilateral taste. Additionally, a lesion of the labyrinthine segment results in impaired ipsilateral lacrimation. Trauma involving the internal auditory canal injures both the facial and the vestibulocochlear nerves, and facial nerve symptoms, loss of hearing, and vertigo are present.

Grading of Facial Nerve Dysfunction

House and Brackmann proposed the clinical grading for facial nerve function [Table 7.1], originally intended for facial nerve paralysis and recovery in vestibular schwannoma treatment.

Differential Diagnosis

Traumatic cranial neuropathy is a difficult diagnosis and requires continuous and repeat examination in cases with high-risk factors, including basal skull fracture, bleeding from the nose or ear, and orbital injury. As an exception, facial nerve paralysis may be easy to diagnose in comatose patients compared with other cranial nerve deficits. Facial paralysis in coma may be incidental and preexisting. A stroke-like presentation of head injury with upper motor neuron facial paralysis needs to be considered in cases where correlation is difficult. Neuroelectrophysiologic studies are helpful in these instances.

Diagnostic Workup

Clinical suspicion is the best diagnostic tool in dealing with trauma. High-risk factors include basal skull fractures, which are best demonstrated by high-resolution computed tomography (CT) scan with thin slices. A good correlation exists between facial nerve injury or eighth nerve injury and type of temporal bone fracture. For both of these nerves, electrophysiologic monitoring is very useful in determining the diagnosis and prognosis of the injuries. Magnetic resonance imaging (MRI) is currently utilized to visualize the cranial nerves. Contrast-enhanced MRI can reveal facial nerve lesions including clinically silent traumatic lesions. Enhancement of the distal intrameatal and labyrinthine segments is described as specific for facial nerve palsy.

Biologic Basis

The common forms of direct injury to the cranial nerves are stretch injury, contusion, laceration, and disruption. Trauma to the vicinity of a cranial nerve leads to delayed injury, resulting in compression of the nerve or edema of the nerve within its coverings. Indirect injury can also be vascular, with resultant ischemia that is either transient or permanent with

Table 7.1 House and Brackmann Clinical Grading for Facial Nerve Function

<table>
<thead>
<tr>
<th>Grade</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Grade 1</td>
<td>Normal: normal facial function in all areas</td>
</tr>
<tr>
<td>Grade 2</td>
<td>Mild dysfunction</td>
</tr>
<tr>
<td>A. Gross: slight weakness noticeable on close inspection, may have very slight synkinesis</td>
<td></td>
</tr>
<tr>
<td>B. At rest: normal symmetry and tone</td>
<td></td>
</tr>
<tr>
<td>C. Motion:</td>
<td></td>
</tr>
<tr>
<td>Forehead: slight to moderate movement</td>
<td></td>
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<tr>
<td>Eye: complete closure with effort</td>
<td></td>
</tr>
<tr>
<td>Mouth: slight asymmetry</td>
<td></td>
</tr>
<tr>
<td>Grade 3</td>
<td>Moderate dysfunction</td>
</tr>
<tr>
<td>A. Gross: obvious but not disfiguring asymmetry, noticeable but not severe synkinesis</td>
<td></td>
</tr>
<tr>
<td>B. Motion:</td>
<td></td>
</tr>
<tr>
<td>Forehead: slight to moderate movement</td>
<td></td>
</tr>
<tr>
<td>Eye: complete closure with effort</td>
<td></td>
</tr>
<tr>
<td>Mouth: slightly weak with maximum effort</td>
<td></td>
</tr>
<tr>
<td>Grade 4</td>
<td>Moderate to severe dysfunction</td>
</tr>
<tr>
<td>A. Gross: obvious weakness and/or asymmetry</td>
<td></td>
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<tr>
<td>B. Motion:</td>
<td></td>
</tr>
<tr>
<td>Forehead: none</td>
<td></td>
</tr>
<tr>
<td>Eye: incomplete closure</td>
<td></td>
</tr>
<tr>
<td>Mouth: asymmetry with maximum effort</td>
<td></td>
</tr>
<tr>
<td>Grade 5</td>
<td>Severe dysfunction</td>
</tr>
<tr>
<td>A. Gross: only barely perceptible motion</td>
<td></td>
</tr>
<tr>
<td>B. At rest: asymmetry</td>
<td></td>
</tr>
<tr>
<td>C. Motion:</td>
<td></td>
</tr>
<tr>
<td>Forehead: none</td>
<td></td>
</tr>
<tr>
<td>Eye: incomplete closure</td>
<td></td>
</tr>
<tr>
<td>Grade 6</td>
<td>Complete paralysis: no movement</td>
</tr>
<tr>
<td>Grades 1 to 3 are associated with acceptable function</td>
<td></td>
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</tbody>
</table>
thrombosis or disruption of blood vessels. Necrosis of the nerve cells follows acute and irreversible injury. In the early stages, the neurons shrink and the Nissl substance stains are relatively normal. Many necrotic neurons are then removed by phagocytes to form microglial nodules. Cavitation follows as the surrounding tissues become involved in the degeneration. Fragmentation and disappearance of the endoplasmic reticulum and disappearance of the Golgi apparatus follow dissolution of the Nissl bodies. Later, cytoplasmic membranes and ribosomes disappear and mitochondria are disrupted. These changes are accompanied by vascular congestion and perivascular inflammation. Phagocytes engulf these necrotic neurons, and soon the nerve fibers show changes of degeneration, usually within 3 days of the death of their corresponding cell bodies.

**Management**

High-resolution CT is indicated in all cases of facial nerve trauma. In the initial stages of trauma with cerebrospinal fluid (CSF) otorrhea, judicious use of diuretics and proper positioning help reduce the leak. Usually prophylactic antibiotics are not indicated, unless surgical intervention is planned. Electrophysiologic monitoring is preferred, as an excellent spontaneous recovery can be expected with delayed-onset paralysis. With nonsurgical management, 90% of patients experience good recovery within 6 months. Absent facial nerve stimulation after 4 days may indicate the need for surgical exploration, especially with transverse fractures of the temporal bone and a discontinuous fallopian canal. Available electrodagnostic tests that are used to evaluate facial nerve injury examine the nerve distal to the stylomastoid foramen. These tests cannot evaluate the nerve across the injury site because the nerve is mostly injured within the temporal bone. Harl and McPhee therefore suggested transcranial magnetic stimulation of the nerve, proximal to the injury site. They found that such stimulation was able to assess the integrity of the nerve after trauma and could predict the potential for functional recovery. These methods, however, are still at an experimental stage. The early return of voluntary motor potentials on standard electromyography carries a favorable prognosis.

Facial nerve decompression surgery should be considered for patients showing more than 90% denervation within 6 days after the onset of palsy. A mastoidectomy and decompression of the nerve under microsurgical techniques with or without repair using grafts may be beneficial. Decompression surgery usually has a beneficial effect when performed within 14 days of injury, and late surgery is recommended in bad head injury cases and in patients who do not exhibit recovery of facial nerve function. Beneficial effects of decompression may be seen even in patients who present 1 to 3 months after injury with more than 95% denervation on electromyography. The surgical approach depends on the hearing status. With preserved hearing, a transmastoid (extralabyrinthine) approach is indicated when the tympanic and mastoid segments of the facial nerve need to be inspected, whereas a middle cranial fossa approach is appropriate for the geniculate ganglion and labyrinthine regions. A translabyrinthine approach provides access to the entire intratemporal facial nerve, in cases with hearing loss. Adjuvant therapy entails the application of natural tears and Lacri-Lube and is indicated if eye closure is impaired. In cases of complete facial nerve palsy with fifth nerve impairment, early tarsorrhaphy would be preferable to prevent corneal ulcerations and subsequent complications leading to phthisis bulbi.

**Anesthesia**

Surgical treatment is required in selective acute situations, and anesthesia follows routine trauma protocol, except for an allowance for intraoperative neuroelectrophysiologic monitoring where ultra–short-acting muscle relaxants and inhalation anesthetics are favored.

**Prognosis**

A positive percutaneous stimulation after 4 days of facial nerve trauma indicates excellent prognosis for recovery. In most cases, facial palsy makes a good spontaneous recovery. In patients with deteriorating facial nerve injuries as demonstrated by electromyoneurography, surgical exploration is indicated. Primary end-to-end neurorrhaphy is the preferred management for transaction injuries, whereas facial nerve decompression may benefit other forms of high-grade nerve trauma. Recovery of satisfactory nerve function can be expected in 75% of cases treated within 3 months after trauma. Facial reanimation is performed (e.g., hypoglossal-facial anastomosis) after 1 to 2 months, if the facial nerve is divided or after 1 year if no function returns before that time. In these cases with delayed presentations, secondary facial reanimation procedures, such as cranial nerve crossovers, dynamic muscle slings, or various static procedures, are useful adjuncts.

**References**

Infections in the posterior fossa are ominous not only due to its small dimensions, but also due to the close proximity of the brainstem with the resultant mass effect, occlusion of the cerebrospinal fluid (CSF) pathways, and hydrocephalus. Because of the adjacent middle ear and mastoid air cells, any contiguous infections can easily spread to the posterior fossa. Infections can also spread hematogenously from a distant source. Rarely, penetrating trauma to the posterior fossa, such as depressed fractures and gunshot wounds, may cause infections by contiguous means. The risk of development of brain abscess after penetrating craniocerebral trauma is 3 to 4.2%.1 Bacterial brain abscess are rare in the United States and they occur approximately in 1500 to 2500 infections each year.2 Improvements in the management of ear, sinus, and orofacial infections were responsible for the reduction of intracranial infections.3

◆ Classification of Posterior Fossa Infections

1. Bacterial infections
   A. Pyogenic infections
   B. Mycobacterial infections
2. Fungal infections
3. Parasitic infections

◆ Bacterial Infections

Pyogenic Infections of Posterior Fossa

Cerebellar Abscess

Of all the sites in the posterior fossa, the cerebellum is the most common site for a brain abscess. Cerebellar abscesses comprise 6 to 35% of all brain abscesses. They are often ominously silent and carry a significant mortality.4 Middle ear infections are the commonest source of cerebellar abscess. The majority of otogenic brain abscesses are located in the temporal lobe followed by the cerebellum.1 The infection spreads to the posterior fossa in contiguity, leading to either abscess formation or subdural empyema. The cerebellar abscess usually has a connection to the petrous bone via a fibrous track consisting of adhesions between the arachnoid and the dura overlying the abscess. In 6% of otogenic abscesses, normal cerebellar tissue lies between the abscess and the petrous bone.4

◆ Causative Organisms of Otogenic Cerebellar Abscess

Understanding the pathogenesis of brain abscesses is important in determining the most likely causative microorganisms and subsequent treatment.2 The causative organisms of bacterial brain abscess depend on geographic distribution, patient age, underlying medical or surgical conditions, and the mode of infection.2 Aerobic or microaerophilic streptococci account for approximately 50% of the aerobic isolates and have been present in approximately 70% of all brain abscess patients. Anaerobic bacteria (e.g., Bacteroides spp., Prevotella spp., Peptostreptococcus, Fusobacterium spp., and Actinomyces spp.) are another major cause of brain abscesses.3 Staphylococcus aureus is the most common cause of traumatic and postoperative brain abscess.1 In 30 to 60% of cases pyogenic brain abscesses are due to mixed infections. Viridans streptococci and Klebsiella pneumoniae are associated with hematogenous spread. K. pneumoniae is a common cause of brain abscess in patients with diabetes mellitus.6 V. streptococci is the most prevalent pathogen in infection of the paranasal sinuses. Head trauma and postneurosurgical states have become important predisposing factors and nosocomial infections also played an important role.4 A majority of cerebellar abscesses are associated with ear or mastoid infections and are usually solitary.5
The Stages of Abscess Formation (Figs. 8.1, 8.2, 8.3, and 8.4)

Based on histology criteria, development and progression of the brain's abscess are divided into four stages: early cerebritis, late cerebritis, early capsule formation, and late capsule formation. Each stage can be identified on radiologic imaging.

Early Cerebritis (1–3 Days)
This stage is characterized by perivascular polymorphonuclear leukocyte infiltration with swollen endothelium, which results in focal inflammation and edema.

Late Cerebritis (4–9 Days)
A necrotic center develops during this stage. Necrotic debris is converted to pus by enzymatic digestion.

Early Capsule Formation (10–14 Days)
This stage is characterized by the appearance of reticulin fibers, which are converted into collagenous capsule. A vascularized capsule enhances with contrast, resulting in a ring-enhancing lesion.

Late Capsule Formation (Beyond Day 14)
At this stage the abscess develops a mature collagenous wall surrounded by a mild degree of cerebritis.

Clinical Features
Clinically the posterior fossa abscess presents with the classic triad of increased intracranial pressure, focal neurologic deficits, and fever. The history is of short duration. Presenting features include the syndrome of the cerebellopontine (CP) angle, cerebellar compression, and cranial nerve deficits including the Gradenigo syndrome, papilledema, and features of hydrocephalus. The most common symptom is headache, and the most common abnormality on physical examination is a decrease in the level of consciousness.

Laboratory Investigations
Blood cultures should be obtained in all patients, especially for the uncommon sources as in infective endocarditis. Cerebrospinal fluid (CSF) analysis is rarely diagnostic, and lumbar puncture is not indicated, especially in the setting of posterior fossa mass, which might result in herniation. Pus should
be obtained during surgery or by stereotactic aspiration and it should be cultured. Other laboratory studies, such as the leukocyte count, erythrocyte sedimentation, and C-reactive protein rate may be normal and not helpful in clinical management.

◆ Radiology

Radiologic findings vary with the stage of abscess. Repeat imaging at regular intervals is recommended to follow the response to therapy. In the cerebritis stage, a computed tomography (CT) scan shows an irregular area of low density. In the early cerebritis stage, there may or may not be contrast enhancement.8 Contrast enhancement starts to appear in the late cerebritis stage.9 Once the capsule is formed, it is well appreciated as a thick ring surrounding a central hypodense or hypointense core. Usually the capsule is thinner toward the ventricular surface. The earliest stages of cerebritis usually show some amount of vasogenic edema. Ependymal entrenchment of the ventricular surface. The earliest stages of cerebritis usually form a thick capsule because of the anaerobic nature of the bacteria and the ischemic origin of the cavity of the abscess. Diffusion-weighted (DW) magnetic resonance imaging (MRI) is helpful in diagnosing a brain abscess and determining the nature of the bacteria and the ischemic origin of the cavity of the abscess. Diffusion-weighted (DW) magnetic resonance imaging (MRI) is helpful in diagnosing a brain abscess and differentiating it from a neoplasm.10

◆ Treatment

A posterior fossa abscess with clinical features of compression is a surgical emergency, and unless treatment is initiated promptly, the neurologic condition may deteriorate. Radiologic findings are helpful in making the decision about the extent of the surgical procedure, which varies from a bur hole to a standard suboccipital craniotomy. Compared with primary aspiration, primary excision is the preferred method for treating a cerebellar abscess.11 Excision may not be possible in all cases. When the capsule is ill-formed, stereotactic or image-guided aspiration yields a specimen for diagnosis and achieves the decompression of the posterior fossa contents. Accompanying hydrocephalus may require external ventricular drainage. In cases of congenital cyanotic heart disease, a corticotomy must always be done under direct vision, to avoid dilated surface veins. There are reports where the endoscope was used in aspirating an intracranial abscess.12,13

In a chronic abscess or in cases of recurrent collections of pus requiring repeated aspiration, excision of the abscess is necessary. Excision of the abscess is also recommended in cases of abscess due to penetrating trauma. There is a well-defined plane of cleavage between the capsule and the adjacent brain parenchyma. Image guidance greatly helps in cases of abscesses that are deeply located. Care must be taken when excising the abscess close to the ventricular system so as not to rupture into the ventricular system. Small fragments of capsule adherent to the surrounding structures such as the neurovascular bundle in the CP angle or the fourth ventricle can be left behind. Persistent hydrocephalus is treated with a shunting procedure, once the infection is controlled.14

Treatment does not stop with the brain abscess. The underlying condition responsible for the abscess needs to be dealt with as well, otherwise the infection might recur again.

Subdural Empyema

Subdural empyema is defined as a purulent infection of the space between the dura and the arachnoid membrane.9 Subdural empyema accounts for approximately 13 to 23% of all intracranial infections.13 With the wide availability of computer-based imaging and good medical facilities, and with the possibility of prompt surgical intervention, the mortality rate has dropped from as high as 40% down to 12.2%.16 With prompt diagnosis and treatment, the mortality should be less than 10%.17,18 Macroscopically one can find thickened arachnoid and thrombosed meningeal veins. Microscopically, subdural empyema is characterized by various degrees of organized exudates, with infiltration of the arachnoid with mature polymorphonuclear leukocytes. Superficial layers of the cerebellum may show ischemic necrosis. The main causes of subdural empyema are chronic paranasal sinusitis, otitis media, and mastoiditis. Other causes include complications of cranial surgery and compound depressed fracture. Subdural empyema is a surgical emergency; while awaiting operative intervention, antibiotics should be chosen based on the suspected source of infection. Treatment consists of a suboccipital craniotomy and drainage of pus, and institution
of appropriate antibiotics. Prognostic factors include the preoperative level of consciousness, the timing, the aggressiveness of treatment, and the rapidity of progression of the disease. Poor prognostic factors are grade III and grade IV coma at admission and age younger than 2 years.

**Brainstem Abscess**

Solitary brainstem abscess accounts for less than 4% of all posterior cranial fossa abscesses. The pons is the most common location followed by the midbrain and medulla. Most infections reach the brainstem through hematogenous spread or from the middle ear by contiguous spread. In a third of cases, the origin of infection is uncertain, and without treatment the outlook is poor. The treatment of choice is surgery for aspiration of the abscess to confirm the diagnosis and to obtain pus for culture and sensitivity. Usually 6 to 8 weeks of parenteral administration of sensitive antibiotics is required.

**Tuberculous Infections**

Central nervous system (CNS) tuberculosis occurs in approximately 1% of all patients with active tuberculosis. Tuberculous infections of the posterior fossa usually manifest as meningitis or as a mass lesion (tuberculoma and tuberculous abscess formation). Tuberculosis of the CNS is most commonly seen in developing countries. In the developed world, infection with human immunodeficiency virus (HIV) has caused an increase in mycobacterium infection, especially of the atypical strains. Intracranial tuberculosis takes several forms:

1. Tuberculous meningitis
2. Tuberculoma
3. Tubercular abscess
4. Vasculitis

Tuberculous meningitis (TBM) is the most serious clinical form of extrapulmonary tuberculosis and is a medical emergency. In TBM, proliferative inflammatory arachnoiditis is most marked in the basal and perichiasmatic locations. In chronic forms, it resembles a fibrous mass engulfing the cranial nerves. Vasculitis of the posterior circulation is rare. Deranged dynamics of CSF results in communicating hydrocephalus due to inflammatory exudates blocking CSF absorption sites. In the majority of patients, CSF analysis shows pleocytosis, low sugar levels, and elevated proteins. The yield of *Mycobacterium tuberculosis* on Lowenstein-Jensen medium from CSF is around 50%. The diagnostic yield of CSF microscopy and culture for *M. tuberculosis* increases with the volume of CSF submitted, and the British Infection Society recommends repeated lumbar puncture if the diagnosis remains uncertain because the diagnosis of TBM is made with lumbar puncture and examination of the CSF. Less often, hydrocephalus may result from blockade of the aqueduct or the fourth ventricular outlet by inflammatory exudates. Treatment consists of the CSF diversion procedure to relieve raised intracranial pressure (ICP). Contrast CT may demonstrate basal exudates and other coexistent lesions such as tuberculomas. The role of neuroendoscopy in patients with TBM with hydrocephalus is not yet established. The raised ICP in TBM could be due to hydrocephalus, focal brain lesions, or cerebral edema. Mannitol is currently the most commonly used agent to treat the cerebral edema. The Cochrane Review recommends the use of corticosteroids in HIV-negative patients with TBM to reduce death and disability.

**Tuberculomas**

Tuberculomas are a conglomeration of caseous foci that develop from multiple tubercles seeding through hematogenous spread, with a centrally located active focus and fibrous capsule formation. When the host resistance is poor, this process may result in a focal area of cerebritis and tuberculomas. Tuberculomas mimic tumors both clinically and radiologically. The characteristic CT finding is a nodular enhancing lesion with central hypodensity and peripheral contrast ring enhancement. In later stages, well-encapsulated tuberculomas appear iso- to hyperdense on plain scans. The incidence of tuberculomas is high among children. In contrast to adults, posterior fossa tuberculomas are common in children.

Conservative management with antitubercular therapy (ATT) is indicated for intracranial tuberculomas because they respond very well. The indications for surgery are raised ICP and CT evidence of a mass effect. Lesions showing a poor response to ATT should be subjected to biopsy/surgery and histopathologic confirmation.

Posterior fossa tuberculomas most often require surgical intervention unless they are very small and peripherally located. A standard suboccipital craniotomy and excision of the lesion is performed for both diagnosis as well as treatment. As the lesions are avascular and well encapsulated, the plane of dissection is easy, and the lesions are totally excisable, although excision is not mandatory. The aim of surgery is to release the obstruction to the CSF pathways. Typically, the granulomas are characterized by the presence of epithelioid cells and Langhans’ cells with inflammatory exudates (Figs. 8.6 and 8.7). The caseous necrosis shows multiple granulomas with these cellular contents (Fig. 8.8).

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**Fig. 8.6** Caseous necrosis surrounded by epithelioid histiocytes and lymphocytes. (H&E ×100.)
Tuberculous Abscess

Dandy in 1932 noted the presence of pus in tuberculomas. Instead of forming caseating granuloma, the brain parenchyma liquefies and becomes an encapsulated sterile pocket of pus. The wall of the tuberculous abscess does not contain the epithelioid granulomas, but is made up of fibrous tissue, as in a pyogenic abscess. The pus of the tuberculous abscess shows the presence of numerous tubercle bacilli, as has been reported by various authors. Tuberculous brain abscesses commonly occur in patients with abnormal cell-mediated immunity and are mostly focal. The criteria for diagnosis of a tuberculous abscess are the presence of pus, isolation of acid-fast bacilli from pus, and histologic confirmation of the abscess.

◆ Bacteriology
The majority of intracranial tuberculomas are caused by the human strain of tubercle bacilli.

◆ Radiologic Findings

Computed Tomography Scan Findings
1. Isodense lesion or slightly hyperdense lesion with strong contrast enhancement
2. Disk-like shape
3. A nodular shape with irregular margins
4. Combination of disks and ring
5. Nonenhancing lesion
6. Target sign with central calcification
7. Approximately 50 to 60% of tuberculomas are multiple.

Magnetic Resonance Imaging
On T1-weighted MRI, the granulomas are usually hyperintense and the variable intensity on MRI represents layers of collagen fibers and inflammatory cellular infiltration surrounding the active caseous focus. MR spectroscopy demonstrates a characteristic lipid peak in the central liquefaction area.

Medical Treatment
The first-line antitubercular drugs that are most commonly used are isoniazid, rifampicin, and pyrazinamide; all are bactericidal. Ethambutol, a bacteriostatic drug, is included as a complement in some cases. In view of the lack of wide data from well-controlled trials, presently four drugs are administered for the initial 3 to 4 months and two drugs for another 14 to 16 months. Most intracranial tuberculomas start to regress after 4 to 6 weeks of antitubercular therapy, and most resolve within 12 to 14 months. In one study, more than two thirds of patients with partially excised or biopsied tuberculomas exhibited persistent lesions on CT scans even after 18 months of ATT. Some tuberculomas show an initial increase in size after starting ATT and then regress after the treatment continues for a time, before the onset of any neurologic deficits or altered sensorium; the outcome is excellent. An unusual phenomenon, paradoxical progression of lesions, has been observed in certain cases, but this does not represent treatment failure.

Surgery is indicated if the tuberculoma is obstructing CSF pathways in the posterior fossa, leading to hydrocephalus, or when the diagnosis is in doubt and in certain drug-resistant cases. Total excision of a tuberculoma may be advocated whenever feasible. As reported by Poonnoose et al, only 18.2% of patients demonstrated complete resolution of their tuberculomas after 9 months of ATT. Even after 18 months of ATT, approximately 70% of patients had residual lesions on imaging. By 24 months, only 54% of patients demonstrated complete resolution of their tuberculomas. Some authors feel that the duration of therapy should be based on the radiologic response of the lesion.
The causative agents include coccidioides, histoplasma, paracoccidioides, and blastomyces. Opportunistic fungi include aspergillus, candida, cryptococcus, and mucormycosis. In HIV-infected individuals, the incidence of cryptococcal infection in the era of highly active antiretroviral therapy (HAART) is between 0.1% and 1%.40 Central nervous system fungal infections are secondary to hematogenous dissemination from a focus elsewhere in the body. CNS fungal infections are increasing due to the use of immunosuppressive drugs in oncology, the high prevalence of HIV, poor control of diabetes mellitus, malignancy, and following organ transplantation.41 Fungal infections of the CNS may cause leptomeningitis, cerebral abscess, granuloma, and obstruction of vasculature by hyphae causing infarcts.

These fungi gain entry into body through the respiratory system, paranasal sinuses, the middle ear, and the skin. Invasion of the CNS may lead to granuloma formation or meningoencephalitis.

Fungal granulomas are most commonly due to aspergillus42 (Figs. 8.9 and 8.10). Aspergillus is highly angiotropic. It can also lead to a mycotic aneurysm. Aspergillus spp. have an angioinvasive nature due to their ability to produce the enzyme elastase. The elastin present in arterial walls and anatomic barriers offers resistance to bacterial invasion but cannot protect against fungi that produce elastase.43 The pseudomycetes are all capable of producing granulomas except for Cryptococcus. The exact incidence of isolated posterior fossa fungal abscesses or granulomas is not known. However, a high index of suspicion and the use of imaging can help in diagnosing them preoperatively.44 In the study by Gasparetto et al,45 the incidence of CNS paracoccidiomycosis affecting the cerebellar hemisphere was 35%

Surgical Treatment

There are three commonly performed procedures: (1) surgical excision to remove or reduce the mass effect, (2) ventricular placement to treat hydrocephalus, and 3) stereotactic biopsy as a means for establishing the diagnosis and for identifying the organism.

Hydrocephalus may result from basal arachnoiditis or intraventricular granulomas. The CSF diversion procedure is warranted if the raised intracranial pressure is symptomatic. The incidence of shunt obstruction is quite high following placement of a shunt. Occasionally, cryptococcal meningitis mimics pseudotumor cerebri, which necessitates a bilateral subtemporal decompression procedure.46

Fungal abscesses are encountered with aspergillosis, cladosporiosis, mucormycosis, candidiasis, and nocardiasis. Usually abscesses are multiple, preceded by vasculitis and hemorraghic infarction. A well-formed abscess requires open or stereotactic drainage, with institution of antifungal therapy. When accessible, a fungal abscess should be excised totally.

Fungal granulomas are produced by aspergillosis, histoplasmosis, blastomycosis, paracoccidiomycosis, and cryptococcus. They mimic tuberculomas but are firmer in consistency, and they often necessitate the use of a knife to cut open the lesion and excise it. The plane of cleavage is difficult, but, in contrast to tuberculomas, fungal granulomas require total excision whenever possible.

The treatment of choice in fungal granulomas or abscesses is antifungal therapy after surgical excision of the lesion, whenever feasible and safe for the patient. The most frequently used agents in this clinical setting are amphotericin B and flucytosine. The usual duration of therapy is 8 to 10 weeks. The recommended dosage of amphotericin B is 0.5 to 0.7 mg/kg. As with its use for other conditions, amphotericin B treatment requires monitoring of renal function and good hydration. The recent addition, liposomal amphotericin, is showing promising results.

Flucytosine is a synthetic oral drug useful in cryptococcus, candidiasis, and chromoblastomycosis. Within the fungal cell, flucytosine is converted to the antimetabolite 5-fluorouracil. Drug resistance appears rather rapidly when flucytosine is used alone. For this reason the drug is generally used in combination with amphotericin B. The usual dose of flucytosine is 25 to 37.5 mg/kg every 6 hours. It is well absorbed from the
gastrointestinal tract and has good penetration into the CSF. Normal renal functions are mandatory before this drug is administered.

Parasitic Infestation of Posterior Fossa

Parasitic infections involving the posterior fossa include cysticercosis, hydatidosis, schistosomiasis, paragonimiasis, trichinosis, filariasis, angiostrongyliasis, and gnathostomiasis. Most of these infections are not surgically relevant, but cysticercosis is the most prevalent.

Neurocysticercosis

Human cysticercosis results when a person serves as the intermediate host of *Taenia solium*, a tapeworm whose larvae develop in various body tissues including the brain. Neurocysticercosis (NCC) is a growing public health problem in the United States. It has four forms morphologically: racemose, cystic-intracerebral, intraventricular, and spinal. The racemose form of cysticercal cysts occupies the basal cisterns. It produces chronic meningitis, a process leading to communicating hydrocephalus. Intraventricular cysticercal cysts may block the outlet of the fourth ventricle, causing obstructive hydrocephalus, which may require surgical diversion procedure. They also induce aqueduct stenosis of an obstructive and inflammatory nature. Intraparenchymal cysticercosis of the cerebellum and brainstem occurs as part of a generalized disease spectrum (Fig. 8.11). Dead cysts undergo calcification.

Diagnosis

The diagnosis of NCC is based mainly on imaging studies and serologic tests. CT scans show features of various stages of the disease. A hyperdense cystic lesion, a hyperdense ring with ring enhancement, and a mural nodule suggestive of scolex are diagnostic. MRI features vary according to the stage. Sometimes intraventricular cysts are difficult to visualize on plain T1- and T2-weighted images. Magnetic resonance CSF flow studies may demonstrate flow obstruction at the site of the cyst. MRI can accurately visualize all types of NCC (Figs. 8.12 and 8.13). Currently, most centers use an enzyme-linked immunoelectrotransfer blot (EITB) with purified glycoprotein antigens (Western blot), which can be done in serum samples or in CSF.

Treatment

The most common surgical indication in NCC is hydrocephalus. Both obstructive and communicating hydrocephalus can occur. Neuroendoscopic surgery is an effective treatment modality for patients with intraventricular NCC. A large cyst of more than 2 cm in the cerebellar hemisphere may warrant surgical excision. Antihelminthic agents are the mainstay of definitive treatment when it is required. But controversy exists as to whether antiparasitic treatment of cysticercosis is necessary in most cases. The indiscriminate use of cysticidal agents in patients with a heavy parasitic load may cause lysis of the cysts, resulting in brain swelling and coma; in patients with intraventricular cysticercosis cysticidal agents may lead to hydrocephalus.

Albendazole is the most commonly used and time-tested drug for cysticercosis; it is prescribed at 15 mg/kg body weight per day in divided doses for 30 days. Albendazole is cost-effective and devoid of major side effects. It has a higher parasiticidal effect than praziquantel. Praziquantel dosage is 50 mg/kg/day orally divided three times a day for 2 weeks. It has to be administered under cover of steroids if the lesions are in the brainstem or orbit to avoid a flare up of edema due to the release of toxic products from the cyst contents.

Corticosteroids are the main form of therapy for cysticercotic encephalitis, angiitis, and chronic meningitis that causes progressive entrapment of cranial nerves. Patients with only calcification should not receive cysticidal drugs because the lesions represent dead parasites. New therapies with albendazole and praziquantel have reduced the duration of the anticysticercal therapy to 8- and 1-day courses, respectively.

Antiepileptic drugs (AEDs) are recommended to treat acute symptomatic seizures and can be withdrawn once the follow-up scans show resolution of the lesion. In most cases, the seizures can be managed with a single medication.

Hydatid Disease

Hydatid disease is a parasitic infestation by a tapeworm of the genus *Echinococcus*. The liver is the most common site for a hydatid cyst (65–75%), whereas hydatidosis of the brain accounts for 1 to 3% of all hydatid lesions. The infection spreads to the human brain primarily via the hematogenous route or by metastatic spread when a cyst ruptures in the heart or lung. Usually the hydatid cysts in the brain are single and most commonly present with features of raised ICP. About 50 to 75% of intracranial hydatid cysts are seen in children. They are mostly located in the frontal and parietal regions. The cysts tend to occur in the distribution of the middle cerebral artery.
A posterior fossa hydatid cyst is very rare. It has been demonstrated experimentally that it takes 5 to 16 months for the cyst to grow 1 cm in diameter in the brain. CT shows a well-defined round or oval cystic structure of CSF density. Rim enhancement is infrequent, but intravascular contrast material may minimally increase the attenuation value of the cystic fluid. There is no surrounding edema, but a considerable mass effect and hydrocephalus may be present. Calcification occurs in less than 1% of the brain hydatid cysts. A typical germinal layer and scolex are diagnostic pathology features (Fig. 8.14). Positive Casoni’s test is diagnostic, but a negative test does not rule out the diagnosis. MRI is superior to CT for evaluation of brain hydatid disease. In MRI, usually an intraaxial fluid-containing structure is seen with no associated edema or enhancement, unless cystic rupture and leakage occur and induce an inflammatory reaction. A diagnostic feature of cerebral hydatid disease that has been described is the low signal intensity of the cyst wall in T2-weighted images.

**Surgical Treatment**

Surgical excision of the hydatid cyst is the treatment of choice if it occurs in the posterior fossa. Intact delivery of the hydatid cyst should be the goal in all cases. Care must be taken in opening the dura to avoid premature rupture of the cyst. The pia-arachnoid is opened at the periphery of the cyst, and the cyst is allowed to deliver itself by the pulsations of the brain. The cyst can be delivered by lowering the head of the operating table and instilling warm saline between the cyst and the surrounding brain.

A recurrent hydatid cyst is far more complicated to excise due to the inflammatory adhesions between the surrounding neurovascular structures and the basal dura. Medical
management with albendazole is necessary in instances where complete excision is not possible and when spillage of contents occurs. Albendazole results in disappearance of up to 48% of cysts and a substantial reduction in size of the cysts in another 28%.60

Mycotic Aneurysms

Mycotic aneurysm are rare neurovascular lesions, constituting approximately 2 to 5% of total intracranial aneurysms.63 They are more commonly located on the middle cerebral artery (MCA) or its distal branches. Mycotic aneurysms involving the posterior circulation are rare.64 The causative organisms could be either bacterial or fungal. In the series published by Chun et al,65 the majority of the organisms were either streptococcus or staphylococcus. Fungal organisms such
as aspergillus, candida, and phacomycetes may also cause mycotic aneurysms. *Aspergillus* spp. have an amo-biotic nature due to the ability to produce the enzyme elastase. The elastin present in arterial walls and anatomic barriers offers resistance to bacterial invasion but cannot protect against fungi that produce elastase.43

Fungal mycotic aneurysms involve large intracranial vessels, in contrast to bacterial aneurysms. Though the exact incidence of fungal mycotic aneurysms is not known, usually they occur in approximately 50% of cases involving large vessels of the posterior fossa.66 The infectious emboli lodge in distal cerebral arteries and occlude the blood flow, and the resultant intense inflammation in the vessel wall weakens it.35

**Clinical Manifestations**

The clinical manifestations depend on the mode of presentation. A ruptured mycotic aneurysm usually presents with a hematomy, and the resultant mass effect might lead to neurologic deterioration or cerebellar signs and symptoms.

The gold standard for the diagnosis of peripherally located mycotic aneurysms is digital subtraction angiography. Serial angiograms may be necessary during the treatment process to monitor the efficacy of treatment.67

**Treatment**

Treatment of the underlying predisposing infection is an important component of therapy,68 and in a majority of the cases it is endocarditis.69 The aims of treatment include treating the underlying infectious process responsible for mycotic aneurysms and obliteration of the aneurysm. The management of unruptured mycotic aneurysms depends on their size and location, and on the risk of bleeding.68 Antibiotic treatment needs to continue for 4 to 6 weeks or until the cultures are consistently sterile.

**Indications for Surgery**

The indications for surgery are hematoma with mass effect and failure to respond to antimicrobial therapy. One of the problems in the obliteration of the aneurysm by surgery is the friable nature of the aneurysm wall and the parent artery. Prior antibiotic therapy might make the aneurysm less friable and facilitate surgical clipping. Mycotic aneurysms that are peripherally located can be excised.

The mycotic aneurysm on the proximal vessels that is otherwise surgically difficult to access may be treated conservatively with antibiotics. The use of stereotactic, angiographic guidance for localization and clipping of a small, distal intracranial bacterial aneurysm may be quite useful for surgically treating mycotic and other peripheral aneurysms.60 If the mycotic aneurysm involves the proximal intracranial vertebral artery, excision of the aneurysm with end-to-end anastomoses or extracranial-intracranial bypass may be attempted. In patients, harboring multiple mycotic aneurysms involving the posterior circulation, treatment strategy includes obliteration of the lesion most likely to have bled, as well as any easily accessible aneurysm where obliteration will not unnecessarily increase the risk of surgical complications.

Various reports have shown the safely and efficacy of endovascular treatment of cerebral mycotic aneurysms.70,71 The number of cases in the above series are small, and much larger studies are needed to confirm the role of endovascular treatment.

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The temporal bone is an area of convergence between neurosurgery and otolaryngology. Disease processes that begin in the mastoid can affect the intracranial contents, and vice versa. Treating the complications from disease extension or surgical intervention in the mastoid may require cooperation between the neurosurgeon and the otolaryngologist. An understanding of the anatomy, disease processes, and surgical procedures in the mastoid prepares the neurosurgeon for collaboration in this area.

◆ **Anatomy**

The mastoid cavity is bounded laterally by the bony cortex, superiorly by the middle cranial fossa, posteriorly by the sigmoid sinus and posterior fossa dura, and inferiorly by the cortex of the mastoid tip. The anterior limit is the posterior border of the external auditory canal, the middle ear, and facial nerve (Fig. 9.1). The medial border is more complex, and is made up of the cochlea, semicircular canals, and the petrous apex. The normal bony cortex between the mastoid cavity and the middle or posterior fossa dura is rarely more than 1 mm thick, and natural dehiscence in the bone is common.

The mastoid cavity is a complex system of interconnected air cells that communicate directly with the middle ear through the mastoid antrum. In the disease-free state, aeration of these cells is maintained by their connection with the nasopharynx through the eustachian tube, middle ear, and mastoid antrum. Understanding this system is crucial to understanding the pathology of the middle ear and mastoid cavity. In short, obstruction of this air cell system at any step can result in disease.

◆ **Pathology of the Mastoid**

**Infectious**

**Otitis Media**

Otitis media is an infection of the middle ear and often involves the mastoid cavity via its connection through the mastoid antrum. Otitis media is more common in patients with dysfunctional eustachian tubes. The function of the eustachian tube is to maintain an air-containing middle ear space by allowing air to pass intermittently from the nasopharynx to the middle ear with swallowing or yawning. The efficiency of the eustachian tube varies from person to person and throughout life. The reasons for this are not always clear, but hypotheses include the weakness of muscles that open the eustachian tube, upper respiratory tract infections, allergies, mucociliary disorders, and surgery or radiation treatment to the head and neck. Children are more prone to otitis media because their eustachian tube is shorter and the angle of the eustachian tube with the nasopharynx is less acute, making contamination of the middle ear by nasopharyngeal secretions more likely.

Acute otitis media is usually a self-limited infection caused by the common upper respiratory pathogens *Streptococcus pneumoniae*, *Haemophilus influenzae*, *Streptococcus pyogenes*, *Pseudomonas aeruginosa*, and *Moraxella catarrhalis*. The time to resolution is reduced with antibiotic treatment. Complications of otitis media are possible, even in the age of antibiotics, and they are important because otitis media is so common. Acute coalescent mastoiditis is the most common complication of otitis media manifested by severe infection of the pneumatic spaces and the mastoid cavity, anterior dis-
placement of the auricle, and tender postauricular swelling. Otitis media is still a common cause of meningitis in children. Facial nerve paralysis, petrous apicitis, labyrinthitis, epidural or subdural abscess, and lateral sinus thrombosis are all potential complications of otitis media² (Fig. 9.2).

**Otitis Externa**

Otitis externa (swimmer’s ear) is an infection of the external auditory canal that can have overlapping symptoms with otitis media. Otitis externa is an infection of the skin of the external auditory canal and causes pain, discharge, and hearing loss. The middle ear is not involved. Rarely, otitis externa can develop into malignant otitis externa, an osteomyelitis of the bony external auditory canal, mastoid cavity, and skull base. Malignant otitis externa is caused by *P. aeruginosa* in 98% of cases, and occurs most frequently in diabetics or other immunosuppressed patients. Cranial nerve paralysis, jugular vein thrombosis, and intracranial extension are possible complications³ (Fig. 9.3).

**Benign Neoplasms**

There are over 25 different benign neoplasms described in the temporal bone. We limit our discussion to the three most common: cholesteatoma, chemodectoma, and cholesterol granuloma.⁴

**Cholesteatoma**

A cholesteatoma (keratocyst) is an accumulation or mass of keratinizing squamous epithelium within the middle ear or mastoid. Primary acquired cholesteatomas develop from retraction pockets in the upper portion (pars flaccida) of the tympanic membrane. Secondary acquired cholesteatomas develop from skin migrating into the middle ear through marginal perforations of the tympanic membrane. Congenital cholesteatomas are rare, and appear to originate from
embryonic rests of ectodermal tissue found medial to the tympanic membrane.

Once inside the endodermal space of the middle ear or mastoid cavity, these ectodermal cells grow without contact inhibition. Unable to desquamate like normal epithelium, the cholesteatoma accumulates keratin and expands in size. Pressure from the growing cyst, and perhaps lytic enzymes produced by the cholesteatoma, causes the slow destruction of middle ear and mastoid structures.

The most common complication of cholesteatomas is conductive hearing loss from interference or erosion of the middle ear ossicles. Cholesteatomas may also erode into the semicircular canals, cochlea, or facial nerve causing vertigo, deafness, or facial paralysis, respectively. Invasion through the tegmen tympani into the middle cranial fossa can provide a route for the intracranial spread of infection including meningitis, epidural abscess, or intracranial abscess. The loss of supporting bone in the floor of the middle fossa can allow the development of mastoid meningocele, encephalocele, and cerebral spinal fluid leak5 (Fig. 9.4).

Glomus Tumors (Chemodectomas)

Glomus tumors (chemodectomas) are highly vascular, slow-growing tumors that originate from chemoreceptor cells along the parasympathetic nerves. They are named according to their location. If they originate in the bifurcation of the carotid artery, they are known as carotid body tumors. Glomus vagale tumors originate from the vagus nerve in the neck and may extend into the jugular foramen. Glomus tympanicum, the smallest of these tumors, is located in the middle ear, along “Jacobson’s” nerve of cranial nerve IX. Glomus jugulare tumors originate in the jugular bulb and fill or expand the jugular foramen. Glomus jugulare tumors are the most likely to have intracranial extension. Chemodectomas are locally invasive and invade both bone and soft tissue. They frequently present with single or multiple cranial nerve palsies, particularly the lower cranial nerves passing through the jugular foramen.6

Cholesterol Granuloma

A cholesterol granuloma (cholesterol cyst) occurs when an air cell becomes “disconnected” from the pneumatized system and the cell fills with inspissated material. Cholesterol crystals are seen histologically within these cysts and are thought to result from microhemorrhage. As material accumulates within the cell, it slowly expands, often destroying bone. The most common symptom cholesterol granuloma is headache, although large lesions can erode into the otic capsule causing sensorineural hearing loss or dizziness.

Small cholesterol granulomas in the petrous apex are common incidental findings on magnetic resonance imaging (MRI) scans (Fig. 9.5). Only large cysts threatening the inner ear or extending into the posterior fossa require treatment. Treatment consists of establishing a drainage path into the middle ear or mastoid space, but is often difficult to maintain patent. Complete excision of the cyst wall is more difficult because the cyst is often wrapped around the internal carotid or cranial nerves.7 Cholesterol cysts are usually managed by permanently draining them through the middle ear or occasionally through the petrous apex into the sphenoid sinus.

Fig. 9.4 Axial (A) and coronal (B) computed tomography (CT) images of right ear cholesteatoma with erosion through the tegmen tympani and horizontal semicircular canal.
Malignant Neoplasms

Malignant neoplasms of the mastoid cavity are very rare. They include primary cancers such as squamous cell carcinoma, adenocarcinoma, rhabdomyosarcoma, osteosarcoma, lymphoma, and leukemic infiltrates. Metastatic tumor is more common, most often from the lung and breast.8

Cerebral Spinal Fluid Leak and Meningoceles

Breaks in the tegmen tympani, whether secondary to trauma, pseudotumor cerebri, tumor growth, iatrogenic, or a congenital defect, can allow cerebrospinal fluid (CSF) leakage into the mastoid with or without an associated encephalocele (Fig. 9.6). “Spontaneous” CSF otorrhea or an encephalocele should raise the possibility of benign intracranial hypertension. The CSF may present as otorrhea if there is a breach in the tympanic membrane or as rhinorrhea if the tympanic membrane is intact and the CSF drains into the nasopharynx through the eustachian tube. This latter circumstance has led to the incorrect diagnosis of anterior fossa CSF rhinorrhea and unnecessary craniotomies. When the tympanic membrane is intact, the CSF behind the drum causes a bulging, clear tympanic membrane that may be quite difficult to distinguish from a normal tympanic membrane. Indications of the diagnosis include sluggishness in the movement of the drum on pneumatoscopy and the presence of bubbles behind the drum. CSF otorrhea may also be misdiagnosed as chronic serous otitis media. A tympanostomy tube placed in such an ear yields continuous clear watery drainage. In this instance the tympanostomy tube should be removed before bacteria from the ear canal contaminates the middle ear.

Conservative management with bed rest and lumbar drain is rarely successful except in acute traumatic injuries. Repair usually depends on surgical exploration and correction of the defect with bone and/or fascia graft repair either through the mastoid or small middle fossa craniotomy, depending on the location of the defect.9

Ménière’s Disease

Ménière’s disease is a clinical syndrome consisting of episodic vertigo, fluctuating hearing loss, tinnitus, and fullness in the ear. These symptoms may be variably present in a given patient. Although the etiology of Ménière’s disease is unknown, ultimately overproduction of endolymph and intermittent rupture of inner ear membranes leads to typical vertiginous attacks. Most patients with Ménière’s disease can be managed medically with a low-sodium (<2 g/day) diet and systemic diuretics. Patients failing medical management, depending on the severity of symptoms and the degree of hearing loss, may be candidates for endolymphatic sac shunt surgery, chemical or surgical labyrinthectomy, or vestibular nerve section.

Surgery of the Mastoid

The goals of otologic surgery are the eradication of disease, the establishment of a safe ear, and the restoration of hearing. The extent of disease in the mastoid dictates whether a conservative or radical approach must be used. A simple or intact canal wall mastoidectomy preserves the external auditory canal and the middle ear. The mastoid cavity remains connected to the middle ear and nasopharynx through the eustachian tube. More extensive disease requires removal of the external auditory canal wall (canal wall down) and marsupializes the mastoid into an enlarged external auditory canal. A modified radical mastoidectomy preserves a shallow middle ear space and conductive mechanism of the middle ear, whereas in a true radical mastoidectomy the eustachian tube is obliterated and the middle ear is sacrificed.
Simple Mastoidectomy (Intact Canal Wall Mastoidectomy)

The key to mastoid surgery is the identification of landmarks, most of which are buried in bone. The sigmoid sinus, facial nerve, and semicircular canals are routinely encountered in mastoid surgery and can be protected only by a thorough knowledge of the three-dimensional interrelationships of these structures in the temporal bone.

Facial nerve monitors are used in most mastoid surgery. Any manipulation or trauma to the nerve generates an auditory signal to the surgeon and operating room staff. A facial nerve stimulator can be used to confirm the identity or location of the nerve. However, monitors can fail or malfunction and are disabled when electrocautery is used; therefore, an absence of a signal from a monitor never absolutely ensures the safety of the nerve. A monitor is never a substitute for thorough knowledge of the anatomic landmarks of the temporal bone.

An incision is made 1 to 2 cm posterior to the postauricular crease and extends from behind the ear lobe to above the ear in the temporal scalp. A flap including the postauricular skin and pinna is developed just superficial to the superficial temporal fascia and mastoid periosteum. The periosteum is divided to expose the mastoid cortex. The upper limit of the mastoid cavity is approximated by the temporal line, which extends posteriorly from the roof of the external auditory canal and the root of the zygoma.

Once the mastoid cortex is exposed, several landmarks guide the dissection: a cribiform area of bone is seen in the postauricular area, the spine of Henle at the posterior border of the external auditory canal, and the external auditory canal itself. Drilling with high-speed drills and constant irrigation is started in a triangle outlined by the temporal line, which extends posteriorly from the roof of the external auditory canal and the root of the zygoma.

The opening in the mastoid is enlarged until three primary landmarks are identified: the tegmen tympani between the mastoid and the middle cranial fossa, the sigmoid sinus posteriorly, and the external auditory canal anteriorly. A large central mastoid air cell, the mastoid antrum, is the next landmark. The facial nerve, lateral semicircular canal, and the short process of the incus are found in the medial wall of the antrum. If the surgery is being performed for cholesteatoma, the antrum is frequently filled by the keratocyst, and it must be cleared away to accurately see the landmarks. The facial nerve enters the mastoid cavity inferior to the lateral semicircular canal and then turns inferiorly toward the stylomastoid foramen. The facial nerve may be skeletonized as much as necessary for removal of disease. If necessary, the air cells lying between the facial nerve and the chorda tympani can be removed to provide direct access to the middle ear, round window, and stapes. The opening so created is known as the facial recess. The middle ear can also be approached directly by elevating the external canal skin and tympanic membrane.

If the ossicular chain has been damaged by disease or removed to facilitate disease control, the ossicles must be restored to preserve hearing. When the surgery has been straightforward, hearing reconstruction may be performed at the initial setting; however, if the disease is extensive, and particularly when it is infiltrated into numerous mastoid air cells, it is often advisable to delay hearing reconstruction for a second-look procedure 6 to 9 months later. After this delay the tympanic membrane should be stable, and any recurrent disease will have gotten extensive enough to be identifiable.

Modified Radical Mastoidectomy

If the disease cannot be successfully removed with the external auditory canal intact, the posterior wall of the external auditory canal may be removed, resulting in a modified radical mastoidectomy. The only difference in this procedure is that the patient must have the mastoid cavity cleaned by appropriately trained personnel at regular intervals (6 to 12 months) forever; however, the operation has a higher probability of permanently eradicating disease.

Radical Mastoidectomy

A radical mastoidectomy is similar to a modified radical mastoidectomy, except that the eustachian tube is permanently

Fig. 9.7 Surface landmarks for mastoidectomy. (Courtesy of the Mayo Foundation.)
blocked, the ossicles are removed, and the remnant of the tympanic membrane is allowed to collapse on the medial wall of the middle ear. All remaining air cells and mucosa are removed up to the middle ear and mastoid to form one common cavity. This operation is reserved for the most recalcitrant middle ear and mastoid disease, and permanently closes the door on any kind of surgical hearing restoration, although the patient remains a candidate for a hearing aid (Fig. 9.9).

Transmastoid Approaches to the Posterior Fossa

Because a large portion of the mastoid is expendable, it can be removed for access to the deeper portions of the inner ear or posterior fossa. The retrolabyrinthine presigmoid approach preserves the inner ear and can be used for retrolabyrinthine nerve section. The translabyrinthine approach is used for removal of acoustic neuromas.

Retrolabyrinthine Presigmoid Approach

The retrolabyrinthine presigmoid approach provides direct access to the posterior fossa between the anterior edge of the sigmoid sinus and the posterior extent of the otic capsule, primarily the posterior semicircular canal. Because the otic capsule is preserved, there should be no significant change in hearing from this approach. After performing a routine mastoidectomy and identifying the facial nerve and semicircular canals, the anterior edge of the sigmoid sinus is carefully defined. If the access is too small, a “Bill’s island” of bone can be created by circumferentially drilling around the dome of the sigmoid with a diamond bur. The sigmoid can then be safely displaced posteriorly and held with a self-retaining retractor.

The bone is thinned and then removed from the posterior fossa dura between the sigmoid sinus and the otic capsule. The endolymphatic sac may be identified and preserved. The posterior fossa dura is incised lateral to the endolymphatic sac, and the posterior fossa entered. Access to the posterior fossa is adequate for vestibular nerve section and vascular loop decompression of cranial nerve VII or VIII.

Patient selection is important because occasionally a patient will have an anteriorly placed sigmoid, making the retrolabyrinthine approach impossible. This is usually seen in the context of chronic otitis media, but can also occur spontaneously. Axial computed tomography (CT) scans are the easiest way to evaluate the location of the sigmoid sinus.

Translabyrinthine Approach

The translabyrinthine approach has been the workhorse procedure for acoustic neuroma removal for many decades. It provides the closest and most direct route to the posterior fossa for the removal of acoustic neuromas, but with the complete sacrifice of hearing.

The procedure starts with a complete mastoidectomy, with removal of all the mastoid air cells from the middle fossa dura to the mastoid tip, and from the sigmoid sinus to the external auditory canal. The descending portion of the facial nerve is identified and its bony covering preserved. A Bill’s island may be created to allow retraction of the sigmoid sinus if additional exposure is needed.

A complete labyrinthectomy provides access to the internal auditory canal (IAC). The canals are systematically removed and followed to the vestibule. The IAC is located immediately medial to the vestibule; however, it is best approached after removing all bone from the posterior fossa dura medial to
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the sigmoid and following the dura to the reflection into the IAC. It is important that wide exposure of the IAC be obtained before it is opened; a full 180 degrees of the posterior surface should be exposed before the dura is opened. Inferiorly the exposure should extend well below the IAC, although a high-riding jugular bulb may limit this exposure. Superiorly, the exposure extends to the tegmen and middle fossa dura. Removal of bone behind the sigmoid and up the parietal skull may add exposure if removal of a large tumor is needed.

Middle Fossa Approach

The middle fossa approach affords access to the IAC while preserving the middle ear conductive mechanism, cochlea, and auditory nerve. The middle fossa approach is applicable to small tumors, generally limited to the IAC. To make the effort of attempted hearing preservation worthwhile, the patient must have hearing thresholds of 50 dB and discrimination score of 50% or better.

The middle fossa approach is started with a vertical skin incision above the external auditory canal. A small craniotomy enables extradural elevation of the temporal lobe. The floor of the middle fossa has few landmarks and the bony surface may be quite irregular. The goal is to identify the IAC without damage to the facial nerve, superior semicircular canal, and cochlea, all of which are more superficial and partially block the view of the canal. Reliable landmarks include the superior petrosal sinus along the posterior petrous ridge and the middle meningeal artery anteriorly. Several techniques have been described for identification of the IAC. House proposed following the superficial petrosal nerve retrograde to the geniculate ganglion and then tracing the facial nerve to the IAC. Fisch is a proponent of identifying the superior semicircular canal first. An angle of 60 degrees from the superior semicircular canal will fall on the long axis of the internal canal. Lastly, Garcia Ibanez prefers to dissect as far medially in the floor of the middle fossa as possible and then directly identify the IAC medial to all other vital structures.

Regardless of the technique used to find the IAC, it must be opened as widely as possible without violation of the cochlea (anterior) or superior semicircular canal (posterior). The bone around the porus acusticus can be removed to yield limited access to the posterior fossa, but tumors with any significant extension into the posterior fossa are best managed in other ways. Although hearing can be preserved with the middle fossa approach, it does present the technical challenge that the facial nerve lies superficial to the tumor, whereas in the posterior approaches the facial nerve is on the far side of the tumor where it remains relatively protected until the nerve dissection is actually started.

◆ Conclusion

The mastoid is a critical interface between the specialties of neurosurgery and otolaryngology. An understanding of disease processes, surgical techniques, and potential complications is important for both specialties if practitioners are to render optimum care to their patients.

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References

Cavernous Malformations

Cavernomas or cavernous malformations of the central nervous system are uncommon pathologic entities, accounting for approximately 16% of all intracranial vascular lesions. Histologically, they are composed of a dilated, single endothelial cell layer and sinusoidal veins with little or no intervening neural tissue. Ultrastructural studies have demonstrated that the absence of tight junctions between endothelial cells may account for the propensity of cavernous malformations to leech blood products into adjacent tissue. These lesions are usually angiographically occult because of their slow flow, which belies their proclivity for hemorrhage. Their magnetic resonance imaging (MRI) appearance is uniquely characteristic. T2-weighted images exhibit a dark, hemosiderin-stained rim and mixed-age blood products composing the nidus of the lesion. Patchy enhancement is observed occasionally, but otherwise cavernous malformations seldom enhance. Contrast administration usually unveils an associated venous angioma.

Posterior fossa cavernous malformations account for approximately 9 to 35% of intracranial cavernous malformations. In order of decreasing frequency, the most common locations involving these lesions are the cerebellum, pons, midbrain, and medulla. On rare occasion, cavernous malformations of the cranial nerves are discovered. Symptomatic annualized event rates for brainstem cavernous malformations have been reported to range from 0.25 to 22.9%. It is believed that hemorrhage rates rise significantly after the initial hemorrhage, with rebleed rates reported as high as 30 to 60% per patient per year. Posterior fossa cavernous malformations, particularly in the brainstem, are associated with a significantly worse clinical course than lesions in supratentorial locations. In the analysis of our institutional series, which is based on the assumption that the lesions were present since birth, the retrospective hemorrhage rate was 5%. In another prospective study, the annual event rate for deep lesions was 10.6%. Hemorrhage rates have been reported higher in women, implicating estrogen as a risk factor. With the widespread availability of MRI, posterior fossa cavernous malformations are being detected more frequently and often incidentally.

Because they are usually located in exquisitely eloquent parenchyma, posterior fossa cavernous malformations present a particularly challenging tactical quandary for neurosurgeons. The stochastic nature of brainstem cavernoma hemorrhages is problematic. A phenomenon known as “temporal clustering” of brainstem cavernomas refers to the clustering of hemorrhagic events in a relatively short time period, flanked by longer periods of relative quiescence. These active periods involving multiple hemorrhages can produce a significant stepwise decline in neurologic function. Hence, an untreated patient who suffers repeated hemorrhages should be strongly considered for surgery. However, surgical intervention risks iatrogenic neurologic deficits and poses a daunting technical challenge in this unforgiving region of the brain. Hence, the treatment algorithm for posterior fossa cavernous malformations can be unclear.

In our experience, we base the decision to operate on the presence of the following factors: (1) neurologic symptoms directly attributable to repeated hemorrhages; (2) documented intra- or extralesional hemorrhage associated with mass effect; and (3) posterior fossa cavernous malformations that approach a pial surface, are exophytic, or are adjacent to non-eloquent parenchyma that serves as a surgical avenue. Surgical intervention on acute or subacute hemorrhagic lesions can take advantage of the plane created by the hematoma between the cavernous malformation and parenchyma, thus favoring early operation. However, patients are often referred after significant or even complete recovery from initial deficits. Therefore, the timing of surgery has varied, depending on the severity of a patient’s clinical presentation and on referral patterns.

Because the natural history of posterior fossa cavernous malformations is not completely understood and symptoms can change dramatically over time, a careful discussion of...
treatment options with patients and family members is mandatory. The option of expectant observation must be explicitly addressed. Asymptomatic lesions or previously symptomatic patients who have recovered from initial deficits may be followed nonsurgically, particularly if the cavernous malformation is deep-seated or minute in size. If surgery is recommended, patients and family should be informed that direct intervention typically mimics the course of a hemorrhagic event. That is, patients should expect to suffer transient, mild but discernible, neurologic deficits following surgery. Most patients, however, return to their baseline preoperative function during follow-up.

◆ Surgical Approach

Once the decision to pursue surgery has been made, the surgical approach is chosen by using the two-point method (Fig. 10.3). Extending a line from the center of the lesion through the nearest point of contact with a pial surface or surgical corridor demonstrates the optimal surgical approach. This method maximizes exposure and minimizes transit through eloquent brain tissue. For posterolateral midbrain lesions, we prefer either a subtemporal or supracerebellar-infratentorial approach. Anterior pontine lesions are accessible through a transsylvian, orbitozygomatic, or retrosigmoid corridor. Lower brainstem lesions are approached via a midline suboccipital or far-lateral craniotomy depending on their precise location (Fig. 10.4).

◆ Microsurgical Technique

After the appropriate approach has been selected, several important technical issues involving microdissection of posterior fossa cavernous malformations must be considered. For lesions that reach a pial surface, the malformation is easily visualized before surgical manipulation. A simple corticectomy can be performed directly over the lesion, followed by microdissection around its borders. We favor microdissectors (Synergetics USA, St. Louis, MO) that offer a variety of instrument angles and microcurette tip sizes to facilitate resection. It is imperative to maintain the plane around the lesion itself without violating the thin-walled, sinusoidal veins of the posterior fossa cavernous malformation or disrupting normal adjacent (usually hemosiderin-stained) brain tissue.

For lesions just beneath the pial surface or buried by overlying parenchyma, we routinely use neuronavigation (Stealth Station; Medtronic SNT, Louisville, CO) as an essential adjunc-
tive tool for locating the lesion. Preoperative diffusion tensor imaging (DTI), which visualizes major white matter tracts, can aid in the selection of a surgical approach by determining which tracts are at risk during dissection and lesion removal. This technology can also be applied to intraoperative neuronavigation, with the hope of reducing postoperative neurologic deficits. Intraoperative nerve stimulation of the cranial nerve VII can be useful when treating lesions adjacent to the floor of the fourth ventricle.

For posterior fossa cavernous malformations deep to the pial surface, a fundamental understanding of access zones in the brainstem is required to enter lesions adjacent to eloquent brainstem structures while minimizing residual deficits (Fig. 10.5). We advocate an extraleral dissection and excision of the lesion when possible. If a cavernoma is entered inadvertently, the thin walls of the lesion will collapse, which renders them difficult to identify and dissect from the walls, and risks leaving residual malformation. Once the lesion has been resected, a thorough final inspection of the walls ensures that no residual remains. Bipolar cauterization for hemostasis is used only sparingly to minimize tissue damage. Procoagulant agents such as Nu-Knit (Johnson & Johnson, Arlington, TX) or FloSeal (Baxter Healthcare, Fremont, CA) are preferred.

Frequently, a venous angioma is encountered during microdissection (Fig. 10.6). This association is well described, and a popular theory places cavernous malformations, venous angiomas, and capillary telangiectasias along the same pathophysiologic spectrum. It is imperative to distinguish this entity from the cavernous malformations itself. Inadvertent injury or resection can lead to devastating venous infarction.

Typically, we obtain postoperative MRIs to serve as a baseline for future comparisons. A 1-year postoperative MRI is followed by regular scans at increasing intervals to monitor for the rare possibility of recurrence. After 10 years of follow-up, patients are advised to obtain imaging studies only if symptoms recur.

Our previously published surgical experience with 100 patients with brainstem cavernous malformations yielded 87% favorable outcomes, 9% worsening of neurologic deficits, and 4% mortality. These results are consistent with data from other series. Comparatively, our untreated cohort of 83 brainstem cavernous malformations have fared worse, with 65% favorable, 33% worse than at presentation, and 2% mortality. Permanent complications have ranged from 12 to 70%. These data stress the importance of careful patient selection before exercising surgical options. The surgical risks also must be weighed against the poor natural clinical history of brainstem cavernous malformations when compared with supratentorial lesions. Using judicious inclusion criteria and careful microsurgical technique, these lesions can be resected with acceptable results (Fig. 10.7).

Radiosurgery

The effect of stereotactic radiosurgery on cavernous malformations remains controversial. Several studies have demonstrated a potential beneficial effect of radiation therapy, stereotactic proton-beam therapy, or stereotactic radiosurgery on the hemorrhage rate associated with cavernous malformations. Hasegawa et al documented a dramatic reduction in hemorrhage rate from 33.9% per year before gamma knife radiosurgery (GKRS; mean pre-GKRS follow-up of 4.33 years) to 12.3% per year in the first 2 years after GKRS. This rate dropped to 0.76% per year during the 2nd through 12th years of follow-up. In this series, the radiosurgical morbidity rate was 13.4%. Similarly, Pollock et al reported a significant reduction in the annual hemorrhage rate from 40% to 2.9% after 2 years of follow-up. However, their rate of treatment morbidity was 41%. Finally, a large series by Lunsford et al showed that patients with hemorrhagic or symptomatic lesions in deep or highly eloquent structures treated with GKRS had a significant reduction in hemorrhage rate from 32.5% to 10.8% per year for the first 2 years after treatment. The rate was reduced to 1.06% annually thereafter with up to 20 years of follow-up. Treatment morbidity was 13.5%.

Whether GKRS really affects cavernous malformations is confounded by multiple variables, including the effect of temporal clustering of hemorrhages on outcome after GKRS, relatively short follow-up periods, population or referral selection biases toward GKRS, and the lack of histologic evidence of radiation-induced changes. It is important to recognize that radiosurgery, particularly in the posterior fossa, subjects

Fig. 10.3 Two-point method. A line connects the center of the lesions with the nearest point on the surface. This trajectory determines the optimal approach for resection. (Courtesy of Barrow Neurological Institute.)
Fig. 10.4 Surgical approaches to the brainstem. (Courtesy of Barrow Neurological Institute.)
patients to a modest but significant risk of morbidity, whereas the effect on the lesion itself is only poorly understood. Future long-term follow-up studies may better elucidate the potential benefits of radiosurgery. Based on current evidence and our clinical experience, we do not recommend radiosurgery as a primary treatment option to our patients.

◆ Dural Fistulas of the Posterior Fossa

Like cavernous malformations, dural arteriovenous fistulas (dAVFs) are uncommon entities of the posterior fossa that can manifest incidentally, secondary to neurologic symptoms, or due to frank hemorrhage. They account for 10 to 15% of all intracranial vascular lesions.37–41 The transverse-sigmoid sinus (38%) is the most common location of dAVFs, followed by the cavernous sinus (34%). Less common locations include the superior sagittal sinus (5%), ethmoidal sinus (4%), superior and inferior petrosal sinuses (5% and 3%, respectively), and marginal sinus (4%).42,43

The most common symptom at presentation is headache. However, bruits, visual symptoms, venous infarction, and intracranial hemorrhage are well described.44,45 dAVFs are also associated with trauma, chronic venous hypertension, middle ear infections, surgery of the venous sinus, or sinus occlusion.42 Dural sinus thrombosis is strongly correlated with the pathophysiology of dAVF formation and persistence. Once the venous system is subjected to arterialized pressures from the fistula, ensuing venous hypertension can lead to leptomeningeal venous drainage, venous varices, and subsequent

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**Fig. 10.5** Annotated lines indicate relative safe entry zones into the brainstem when cavernomas lie beneath the pial surface. (A) Lateral view, (B) Posterior view. (Courtesy of Barrow Neurological Institute.)

**Fig. 10.6** MRI of a classic venous angioma associated with a cavernous malformation (arrow). (Courtesy of Barrow Neurological Institute.)

**Fig. 10.7** Mesencephalic brainstem cavernous malformation. Two-point method favors either a subtemporal or lateral supracerbellar infratentorial approach; the latter was used in this case. A safe entry zone in the posterolateral midbrain, lateral to the tectal plate, was chosen as the entry point. (Courtesy of Barrow Neurological Institute.)
risk for hemorrhagic or ischemic events. The link with angiogenic growth factors has been described in animal models,\textsuperscript{32,46} the inhibition of which might serve as a potential treatment avenue in the future. Nonetheless, most patients have no antecedent history to explain the formation of the lesion.

A multivariate analysis of more than 100 cases\textsuperscript{47,48} demonstrated that leptomeningeal or galenic (deep) venous drainage pattern and venous varix formation significantly correlated with aggressive behavior and poor outcome. Using angioarchitectural features, grading scales have been formulated to predict the risk of neurologic symptoms or hemorrhage.\textsuperscript{49–52} The University of California–San Francisco (UCSF) scale describes four grades of dAVFs.\textsuperscript{49} Grade 1 lesions exhibit anterograde drainage via normal sinus pathways. Grade 2 lesions demonstrate antero- and retrograde venous drainage with or without cortical venous drainage. Grade 3 lesions have retrograde venous drainage related to an occluded sinus. Grade 4 lesions have only cortical venous drainage. According to the UCSF data, hemorrhage occurred in 31% of grade 3 and in 100% of grade 4 lesions.

The Cognard classification describes five types of dAVFs delineating location, direction of flow, and drainage patterns.\textsuperscript{50} Type 1 lesions have antegrade drainage into a dural venous sinus. Type 2 lesions are divided into three subtypes: type 2A lesions have direct retrograde drainage into a dural sinus; type 2B lesions flow antegrade into a dural sinus with retrograde cortical venous drainage; and type 2A+B lesions have retrograde flow into a dural sinus with retrograde cortical drainage. Type 3 lesions drain directly into nonectatic cortical veins. Type 4 lesions have direct ectatic cortical venous drainage. Type 5 lesions have retrograde drainage into spinal perimedullary veins and are classified further as spinal dAVFs.\textsuperscript{53}

Borden et al\textsuperscript{51} classified dAVFs into three types based on patterns of venous drainage. Type I lesions drain antegrade directly into a major venous dural sinus. Type II lesions drain into the venous dural sinus with retrograde drainage into subarachnoid veins. Type III lesions drain retrograde into cortical veins. Types II and III are associated with significant venous hypertension and are considered at high risk for hemorrhage. We typically use the Borden classification system and refer to dAVFs using this classification in this chapter’s discussion.

\section*{Natural History and Treatment Algorithm}

The decision to treat dAVFs can depend on a several factors, including the patient’s symptoms, presence of hemorrhage, location of the fistula, its angiographic features, and changes in the lesion over time.\textsuperscript{39,40,42,49,51,54–56} Borden type I dAVFs are thought to have a benign natural history, with one series\textsuperscript{57} showing only one hemorrhage in 68 untreated patients followed for at least 27 months. However, type I lesions carry a 2% risk of progression to a higher grade.\textsuperscript{57,58} This progression is not always heralded by new symptoms.

Higher-grade dAVFs are considered more dangerous. Van Dijk et al\textsuperscript{59} followed 20 patients with type II or III dAVFs over a total of 87 patient-years, calculating an 8% risk of hemorrhage and 10% risk of death on an annual basis. This and other studies have justified the aggressive treatment of type II or III lesions, especially when symptomatic.\textsuperscript{56,60,62} Study of asymptomatic patients with high-risk angiographic features, such as cortical venous drainage (CVD) with distal sinus stenosis or deep venous drainage, demonstrated a high annualized risk of hemorrhage.\textsuperscript{56,62,63} However, recent natural history data from Strom et al\textsuperscript{64} of type II or III dAVFs demonstrated that lesions exhibiting asymptomatic CVD follow a more benign course than similar lesions with symptomatic CVD (1.4% versus 19% per year event rate). Similarly, Söderman et al\textsuperscript{65} followed 85 patients with dAVF and CVD and found the annual risk of hemorrhage varied depending on hemorrhage at presentation: 7.5% per year for patients presenting with hemorrhage, and only 1.5% per year for patients without. The presence of high-risk features requires careful consideration when determining treatment strategy.

Patients with low-risk dAVFs and no associated neurologic symptoms can be observed and should undergo regular angiographic follow-up. Low-risk dAVFs associated with flow-related headaches, orbital symptoms, or bruises that impair the quality of life may be treated with carotid compressive therapy\textsuperscript{45,65–68} or palliative transarterial embolization. The goal of treatment of high-risk dAVFs is complete obliteration; however, high-grade lesions with complex or inaccessible features are sometimes treated with surgical or endovascular disconnection of the cortical venous drainage.\textsuperscript{50,70} This leaves the fistula intact, converting a high-risk dAVF to a more benign lesion, although this outcome is not as favorable as complete resection or obliteration. As with cavernous malformations, treatment recommendations should be based on both clinical presentation and angiographic features.

\section*{Surgical Approach and Microsurgical Technique}

The key to successful surgical management of posterior fossa dAVFs is appropriate angiographic identification of the fistulous connection. Careful evaluation of the primary source on angiographic images enables the surgeon to identify the exact point at which arterial blood flow from the extracranial circulation enters the venous circulation. Knowing this exact location, the fistula can be localized within the posterior fossa (i.e., transverse-sigmoid junction, torcular Herophili, superior petrosal sinus, etc.). Once the location is determined, the appropriate surgical approach can be chosen. Almost all posterior fossa dAVFs can be accessed through a retrosigmoid, supratentorial-infratentorial, or lateral suboccipital craniotomy (Figs. 10.8 and 10.9).\textsuperscript{71}

Intraoperatively, careful exploration allows the artery feeding the fistula to be identified. At this point, simple ligation of the fistula is performed with arteriovenous malformation (AVM) clips. Numerous tiny arterial feeding branches may be observed in the vicinity of the fistula. These branches may be ligated at surgery, but disruption of the main fistula is sufficient to obliterate the entire lesion.\textsuperscript{72,73} If multiple small-to-medium feeders that cannot be reached easily are present, preoperative transarterial embolization is helpful. When external ligation is not technically feasible, direct embolization...
10 Hemangiomas and Dural Fistulas

of the fistula with muslin packing is a useful technique. Intraoperative angiography is extremely useful to confirm complete obliteration before closure. Close clinical and angiographic follow-up is warranted to check for recurrences.

◆ Endovascular Treatment

At our institution, endovascular treatment options for dAVFs are usually explored first. Careful patient selection for endovascular therapy is paramount. Even with contemporary techniques, specific angiographic features make some dAVFs better suited for stand-alone endovascular treatment and others better suited for combined endovascular, surgical, and/or radiosurgical treatment. If a fistula has normal anterograde sinus drainage, every effort must be made to maintain the patency of the parent sinus. Inadvertent occlusion of the parent dural sinus may be poorly tolerated and can lead to severe venous outflow obstruction and venous infarction. Caragine et al\textsuperscript{74} presented a series of patients with transverse-sigmoid junction dAVFs associated with a venous pouch parallel to the parent sinus. By recognizing this separate, parallel channel, the endovascular surgeon can safely embolize the venous pouch, preserve the parent sinus, and cure the patient.

Lesions with parent sinus occlusion distal to the fistula can be obliterated with coiling of the sinus at the fistulous connection because there is a lower risk of losing important distal venous drainage. Local venous anatomy must be delineated for each case before executing parent sinus embolization. Alternatively, if the venous route to the dAVF is blocked by occlusion or if there is direct communication between the dAVF and a cortical vein, it is usually preferable to surgically obliterate the fistula directly.\textsuperscript{75} In such cases intraoperative direct puncture embolization of the fistula is another option.\textsuperscript{76}

Prior to the advent of nonadhesive liquid embolic agents (such as Onyx; see below), endovascular treatment of dAVFs typically involved staged transarterial embolization of dAVF feeders followed by transvenous coiling. Smaller feeding branches could be obliterated with liquid adhesives agents such as N-butyl cyanoacrylate (NBCA), whereas larger arterial feeders were left intact for “road-mapping” purposes and for angiographic verification that subsequent transvenous coiling or surgical ligation has obliterated the fistula. This technique is still commonly used as primary treatment in many centers and in cases where curative transarterial Onyx embolization is not feasible.\textsuperscript{77}

Onyx (ev3 Endovascular, Plymouth, MN), a nonadhesive liquid embolic agent originally approved for the endovascular occlusion of arteriovenous malformations, is an emerging endovascular treatment for dAVFs. Onyx is cohesive, rather than adhesive, and is considered a more controllable and less rapid form of embolization compared with NBCA. Stiefel et al\textsuperscript{78} treated 28 patients with dAVFs of various grades, achieving angiographic cure in 21. Similar results were obtained by Cognard et al\textsuperscript{79} in their series of 30 patients. Several other series have described excellent short- and long-term cure rates with Onyx treatment of dAVF.\textsuperscript{80–82} Long-term angiographic and clinical follow-up of such treatments is needed to confirm the durable efficacy of Onyx embolization.

◆ Surgical Treatment

For those dAVFs where definitive endovascular occlusion cannot be safely obtained, surgical ligation is necessary. Similar
to multimodality treatment in arteriovenous malformations, preoperative embolization of arterial connections, particularly in surgically inaccessible areas, can facilitate the surgeon’s task of dissecting and ligating the main fistula (Fig. 10.10). If a combined approach is warranted, embolization of arteries with significant scalp supply (superficial temporal, occipital, posterior auricular) is avoided to prevent possible skin necrosis after surgery. Instead, middle meningeal artery feeders are targeted and embolized.

Surgical obliteration of dAVFs has a high success rate. The best surgical approach is matched to the location of the lesion. Meticulous skeletonization of the associated dural sinus is required, including disconnection of feeders from the tentorium and falx where applicable. In 23 patients with high-grade dAVFs treated with surgery with or without preoperative embolization, Liu et al reported complete angiographic obliteration and no perioperative complications or further clinical events with up to 84 months of follow-up. Another series of 17 patients with high-grade dAVFs treated with surgery alone showed angiographic cure in 16. Collice et al described complete angiographic cure in 34 patients treated with surgery with or without embolization. Overall, surgery

Fig. 10.9 Anteroposterior (A) and lateral (B) angiograms showing a tentorial dAVF. (C) Postoperative angiogram after retrosigmoid craniotomy and direct clipping of the fistulous connection shows excellent obliteration of the lesion. (Courtesy of Barrow Neurological Institute.)
Fig. 10.10  (A) Left transverse-sigmoid sinus region dAVF with primary cortical venous drainage and obstruction of transverse sinus outflow. (B) Scout film after preoperative embolization. (C) Postembolization angiogram shows significant reduction of several of the occipital artery feeders. Residual feeders are present due to endovascular inaccessibility. (Courtesy of Barrow Neurological Institute.)
is considered a safe and effective treatment for dAVFs, particularly when embolization is not feasible or curative.

**Radiosurgery**

Recent data have suggested that GKRS, either with or without tandem transarterial embolization, may provide therapeutic relief of symptoms and potentially promote the obliteration of fistulas. Pan et al reported that over a median follow-up of 19 months, 58% of patients were angiographically cured after GKRS at a dose of 16.5 to 19 Gy at the 50% isodose line. Friedman et al described 25 radiosurgery patients, 22 of whom also received transarterial embolization. Of these patients, 17 ultimately underwent angiographic follow-up, and 11 patients demonstrated “total or near total (>90%) obliteration.” All but one patient experienced immediate relief of symptoms. Söderman et al studied GKRS in 53 patients with dAVFs and reported an angiographic cure rate of 68% at 2 years. Wu et al studied GKRS in 81 patients with dAVFs and found complete or partial symptomatic relief in 75% after treatment. However, angiographic cure at 24 months was only 50% and was slightly less in patients with high-grade lesions. Although the clinical outcome of patients treated with GKRS seems to be better than the natural history of untreated patients, relief of symptoms is not directly comparable to a surgical or endovascular cure. Nonetheless, this modality is recognized as a useful adjuvant or stand-alone treatment of dAVFs. Posterior fossa dAVFs commonly recruit contributions from the tentorial branch of the meningohypophyseal trunk. Typically, due to the high tortuosity of this vessel, embolization is not feasible. Therefore, GKRS is ideally suited for high-risk preretrosal or skull-based dAVFs that cannot be safely treated with embolization due to potential collateral arterial feeders to the posterior circulation and cranial nerves or that are poorly accessible surgically.

**References**


III

Surgical Techniques
The neural structures found within the posterior fossa are responsible for some of the most critical functions in the central nervous system. The limited space that encompasses this compartment of the brain leaves little room for a mass lesion to form before it has the potential to become dramatically symptomatic. Brainstem compression, herniation, and death are all risks of mass lesions that occur in this location. The ability to safely treat pathology of the posterior fossa has significantly evolved over the course of the modern neurosurgical era. Advances in anesthesia, aseptic technique, neurologic localization, and technical tools and skills have allowed for safe and effective surgical treatment options to be developed.

Two standard approaches are used to access the majority of posterior fossa pathology: the midline suboccipital approach and the retromastoid approach. Although some technical variability exists among neurosurgeons in regard to these procedures, the fundamental concepts remain the same. A thorough familiarity with these approaches is a critical skill for any neurosurgeon treating these lesions to possess.

◆ Preoperative Management

Preoperative considerations for posterior fossa surgeries should include a thorough history and physical examination, a cardiogram, urinalysis, chest x-ray, and laboratory tests that include a complete metabolic profile, complete blood count, and coagulation profile. All necessary neuroimaging should be obtained and carefully evaluated for vascular relationships to the lesion, ventricular size, and any distortion of normal anatomy. When appropriate, arrangements should be made for intraoperative neurophysiologic monitoring. Neuromonitoring is frequently done for lesions involving the cranial nerves.

Providing quality neuroanesthesia for the patient is a critical component of a smooth intraoperative course during a posterior fossa surgery. Prior to the surgery there should be clear communication with the anesthesiologist regarding the administration of antibiotics, osmotic agents, and corticosteroids. Wide swings in blood pressure should be avoided during head-pin placement, and for the duration of the surgical procedure and emergence from anesthesia. In cases involving neurophysiologic monitoring of cranial nerves, the anesthesiologist needs to avoid giving any pharmaceutical agents, such as paralytics, that can interfere with the monitoring.

◆ The Midline Suboccipital Approach

The midline suboccipital approach can be used to access vascular, neoplastic, and developmental pathologies of the cerebellar hemispheres, vermis, fourth ventricle, brainstem, and foramen magnum. The surgical goals of this approach should include sufficient exposure, adequate decompression of the foramen magnum, avoidance of injury to the venous sinuses and structures adjacent to the fourth ventricle, and prevention of avoidable complications.

Positioning

The patient can be positioned in either the prone or sitting position. For the prone position, the head is placed in three-point pin fixation and the patient is positioned on chest rolls with padding at all pressure points. The prone position minimizes the risk of a venous air embolism and facilitates controlled release of cerebrospinal fluid (CSF). For the sitting position, the head is again secured in three-point pin fixation and flexed forward. This position facilitates increased venous drainage and subsequent lowering of the intracranial pressure, gravitational drainage of blood and CSF from the surgical field, and unrestricted ventilation. The risk of venous air emboli and a biomechanical disadvantage for the surgeon that can result in arm fatigue are disadvantages of this position. Precordial Dopplers and a central venous line in the right atrium should be placed prior to beginning the
surgery in order to detect and evacuate any potential air emboli.\(^2,3\)

Landmarks

Several surface landmarks are important for planning the skin incision and craniotomy for the midline approach. The inion overlies the torcular, and the course of the transverse sinus can be approximated along the line between the inion and superior auditory meatus.\(^2–4\) (Fig. 11.2) A line between the inion and the spinous process of C2 should be approximated. A standard vertical incision extends along this line from approximately 2 cm above the inion to the spinous process of C2. The incision can be shortened or lengthened depending on the exact nature of the pathology. Approximately 10 to 15 mm inferior to the superior nuchal line the bone thickness decreases dramatically. This occipital bone thickness increases again to 5 to 6 mm within 1 cm of the foramen magnum.\(^5\)

Surgical Technique

The incision is made as described above, and deepened down to the bone along the midline avascular plane. The muscles are elevated from the underlying occipital bone in a subperiosteal fashion. The dorsal arch of C1 is exposed, taking care to remain medial to the vertebral arteries. Brisk bleeding from the surrounding venous plexus may serve as a warning sign that the vertebral artery is nearby. Self-retaining retractors are used to maintain exposure. A craniotomy is performed by placing two bur holes below the transverse sinus on either side of the midline septum. A craniotomy flap is turned and elevated. If necessary, a Midus Rex drill (Medtronic, Minneapolis, MN) with a B1 or C1 bit attachment can be used to score the edges of the craniotomy followed by a Kerrison rongeur to completely free the bone. Drilling and rongeurs should be used to create a wide opening at the foramen magnum, ensuring complete decompression of the brainstem. For lesions involving the foramen magnum and upper cervical spine, a laminectomy at C1 is performed. The dura is generally opened in a Y-shaped fashion with the straight limb of the Y extending down the midline to the region of the foramen magnum. The dural flaps are reflected superolaterally with sutures.\(^3,6\) (Fig. 11.3).

The cerebellar hemispheres will be visualized immediately. The arachnoid overlying the cisterna magna can be opened to drain CSF and allow for brain relaxation. The cerebellar tonsils should be identified as well as the posterior inferior cerebellar arteries (PICAs), noting that PICA anatomy can be variable. After freeing the arachnoid attachments, the tonsils can be gently retracted laterally, opening the foramen Magendie and leading to visualization of the floor of the fourth ventricle and cervicomedullary junction (Fig. 11.4).

Cerebellomedullary Fissure Approach to the Fourth Ventricle

If the upper portion of the fourth ventricle and cerebral aqueduct need to be visualized, the cerebellomedullary fissure approach to the fourth ventricle may be used. The cerebellomedullary fissure separates the tonsils from the medulla oblongata. The PICA and the vein of the cerebellomedullary fissure course together through this space. To expose the fissure, the tonsils are retracted superolaterally. The lateral recesses of the fourth ventricle are exposed and can be followed to the foramen of Luschka, communicating with the cerebellopontine angle. The tela choroidea is identified where it forms the roof of the fourth ventricle. The tela is incised, facilitating entrance to the fourth ventricle.\(^6–8\) This approach enables the exposure of fourth ventricular tumors while avoiding division of the vermis and its associated complications.

Image guidance can be of great utility for lesions involving the parenchyma of the cerebellum to minimize injury to normal parenchyma, especially the deep cerebellar nuclei. It is important to note that the cerebellar hemispheres relax and shift position after release of a significant amount of CSF. If using image-guided stereotaxy, it may benefit the surgeon to localize the lesion prior to releasing CSF.
Fig. 11.2  (A) Soft tissue is divided in the midline and elevated laterally off the suboccipital bone. (B) Suboccipital craniotomy is performed with wide opening of the foramen magnum. A C1 laminectomy is performed for lesions lying at or below the foramen magnum. (C) The dural is opened in a Y-shaped fashion and reflected superolaterally. (D) The cerebellar hemispheres, tonsils, and posterior inferior cerebellar artery (PICA) can be visualized following dural opening.

Fig. 11.3  (A) Posterior view of the cerebellum and PICA. (B) Floor of the fourth ventricle. Br., branch; Flocc., flocculus; Hem., hemisphere; Inf., inferior; Lat., lateral; Mid., middle; Ped., pedicle; PICA, posterior inferior cerebellar artery; Sup., superior; Verm., vermian. (Courtesy of Albert L. Rhoton, Jr., MD)
Closure

Strict hemostasis should always be achieved prior to closure of the dura. The most common complication associated with posterior fossa surgery is leakage of CSF. To minimize the risk of either a leak or a pseudomeningocele, a watertight closure of the dural opening should be achieved. This may require using dural substitutes or autologous grafts and can be augmented by a dural sealant or fibrin glue. The Valsalva maneuver can be performed by the anesthesiologist to search for CSF communication through the dural closure. If there is no notable swelling in the posterior fossa at the time of closure, the bone flap should be secured into place with titanium microplates. The muscle, fascia, and skin should be closed in multiple layers to act as a secondary barrier against a CSF leak.

Pitfalls and Complications

During positioning of the patient, excessive turning of the head and neck can obstruct venous outflow and create elevated intracranial pressure. One potential vascular injury that can occur with this approach typically happens with aggressive lateral dissection of muscle off of the C1 lamina, resulting in injury to the vertebral artery as it courses across the posterior arch of the atlas. Another involves occluding the PICA while retracting the tonsils. The location of the PICA should always be noted prior to placing the retractors, and care should be taken not to compress or injure the artery.

Surgical procedures involving the cerebellum and fourth ventricle carry a risk of injury to the surrounding neurologic structures. Although a significant amount of tissue in the cerebellar hemispheres can be sacrificed without any demonstrable loss of function, certain cerebellar structures are more symptomatic than others when compromised. Unilateral injury to the portion of the hemisphere lateral to the dentate nuclei may result in ataxia, hypotonia, and adiadochokinesia in the ipsilateral limbs. A direct injury to the dentate nucleus increases the severity and duration of these symptoms as well as results in an intention tremor with voluntary movements. Dysarthria can result from injury to the paravermian cerebellum. Injury to the middle cerebellar peduncle can lead to ipsilateral ataxia, dysmetria, and hypotonia. Dissection of tumors of the fourth ventricle can put the superior and inferior cerebellar peduncles at risk. Injury to the superior peduncle may result in intention tremor, dysmetria, and decomposition of movement, whereas injury to the inferior peduncle may result in ataxia and gait difficulties. Small lesions in the vermis are usually asymptomatic; however, injury to the portions containing vestibular fibers—namely the uvula, nodule, and flocculus—creates deficits of equilibrium, including truncal ataxia, staggering gait, head and trunk oscillation, and occasionally nystagmus. Cerebellar mutism is an occasional transient symptom seen in children after the removal of midline cerebellar lesions and is discussed in further detail below.

Resection of tumors or other lesions involving the floor of the fourth ventricle can create problems with facial movement, lateral gaze, hypotension, apnea, speech and swallowing, and cough reflex.

Postoperative swelling, bleeding, or arachnoid scarring can lead to obstruction of CSF flow and subsequent hydrocephalus. This can be diagnosed by neurologic examination and confirmed with computed tomography (CT) prior to placing an external ventricular drain. Aseptic meningitis can appear 5 to 7 days after the operation and is characterized by fever, photophobia, and nuchal rigidity. It is treated with corticosteroids; however, CSF cultures should be obtained and prophylactic antibiotics initiated until bacterial meningitis has been ruled out.

◆ The Retromastoid Approach

The retromastoid approach can be used to access tumors of the cerebellopontine angle, vascular lesions in the posterior circulation, and pathologies involving cranial nerves V through XII. The surgical goals of this approach should include adequate exposure, avoidance of injury to the dura or venous sinuses, and prevention of avoidable complications.
11 Basic Concepts in Posterior Fossa Surgery

Positioning and Incision

We prefer the lateral position for this procedure, but other options include the sitting, three-quarter lateral, or supine position with the head maximally rotated. For the lateral position, an axillary roll is placed and all pressure points are padded prior to securing the patient to the operative table. The ipsilateral shoulder is pulled down to maximize the angle between the head and shoulder. After securing the head in three-point pin fixation, the sagittal suture is aligned parallel to the floor prior to gently flexing and laterally tilting the head toward the contralateral shoulder (Fig. 11.5).

Landmarks

Several key landmarks should be identified. The line between the inion and the zygomatic root indicates the course of the transverse sinus. The digastric or mastoid groove indicates the position of the sigmoid sinus. The position of the asterion is variable in the cephalocaudal direction; however, it is located directly over the transverse-sigmoid junction in 74.4 to 86% of patients, and bur holes placed in this area risk damage to the underlying sinus. A retroauricular incision is made in either a curvilinear or lazy-S shape approximately 1 cm medial to the mastoid. The extent of the incision depends on the nature of the pathology, but typically it extends from the top of the pinna to the mastoid tip (Fig. 11.5).

Surgical Technique

The soft tissue is elevated in subperiosteal fashion from the underlying bone, and a self-retaining retractor is placed to maintain exposure. A bur hole is made approximately 2 cm inferior to the asterion and 1 to 2 cm medial to the mastoid groove. A craniotomy flap is then turned, keeping the superolateral aspect at the edge of the transverse-sigmoid sinus junction. If necessary, further bone can be removed with the drill or rongeurs until the anterior margin of the bony opening reaches the medial edge of the sigmoid sinus. For pathology that involves the lower cranial nerves, the foramen magnum may need to be opened as well.

The dura is opened in a K-shaped fashion and reflected with sutures. The cerebellum is then gently elevated until the arachnoid of the cisternal magna is visualized and opened, facilitating drainage of CSF and relaxation of the cerebellum. If necessary, a self-retaining retractor can be placed to gently retract the cerebellum downward to visualize the structures of the cerebellopontine angle. Angling the microscope toward the upper portion of the cerebellopontine angle facilitates visualization of the trigeminal nerve, superior cerebellar artery, and superior petrosal vein below the tentorium. The fourth cranial nerve may be seen along the medial edge of the tentorium behind the arachnoid plane. Moving the angle of the microscope slightly inferiorly reveals the cranial nerve VII/VIII complex entering the internal auditory canal, along with the anterior inferior cerebellar artery. Looking further inferiorly through the cranial opening reveals the lower cranial nerve complex (IX, X, and XI) along with the PICA, the vertebral artery, and the vertebrobasilar junction (Fig. 11.6).

Closure

Strict hemostasis should be ensured prior to beginning the closure, and as with the midline approach, the dura should be closed in a watertight fashion to minimize the higher risk of CSF leakage. The Valsalva maneuver is performed by the anesthetist to inspect the dural closure. Dural sealants are appropriate if any communication through the dural closure is suspect. If a craniotomy has been performed, the bone flap
Fig. 11.6 (A) Cranial nerve (CN) V and the superior cerebellar artery can be visualized at the superior aspect of the cerebellopontine angle. (B) The CN VII/VIII complex is visualized after resection of a small acoustic neuroma. CN VIII is retracted inferiorly, demonstrating CN VII. (C) The lower cranial nerves (IX, X, XI) and the posterior inferior cerebellar artery are visualized at the inferior aspect of the cerebellopontine angle.
should be secured into place. Patients who have undergone craniectomies are at risk for chronic craniectomy headaches, and a cranioplasty with bone substitute should be considered as part of the closure. The muscle, fascia, and skin should be closed in multiple layers to act as a secondary barrier against a CSF leak.

**Pitfalls and Complications**

Lacerations of the transverse or sigmoid sinus can lead to significant blood loss if not addressed expeditiously. When tamponading sinus bleeding, care must be taken not to occlude the vessel as this could lead to venous outflow obstruction and elevated intracranial pressure. If necessary, bony removal can be extended to repair a sinus laceration primarily. Any mastoid air cells that have been exposed during the opening should be sealed completely with bone wax to avoid CSF leakage.

Excessive manipulation or retraction of the cerebellum can lead to posterior fossa swelling, obstruction of CSF outflow, and subsequent hydrocephalus. Cerebellar contusions can be caused by prolonged retraction or overly aggressive retraction in the setting of cerebellar swelling. If noted during the surgery, the contusion should be resected to avoid associated swelling in the postoperative period.

Injury to the cranial nerves can have devastating consequences for the patient. Trigeminal and facial paralysis can lead to exposure keratopathy if not aggressively managed with lubricants. Facial paralysis can affect speech or lead to significant aesthetic consequences. Injury to cranial nerve VIII results in ipsilateral hearing loss. Injury to the lower cranial nerves (IX, X, and XI) can result in dysphagia and aspiration pneumonia. These injuries can generally be avoided with gentle retraction, careful microsurgical technique, gentle handling of the nerves and arteries, and neurophysiologic monitoring.

**Postoperative Care**

The postoperative care varies depending on the patient’s pathology and any complications that occur; however, any patient who has undergone a surgical procedure in the posterior fossa should be monitored in an intensive care unit or neurologic step-down unit during the immediate postoperative period. Most neurosurgical patients are kept on prophylactic intravenous antibiotics for 24 hours. Corticosteroids should be continued from the operating room and weaned as appropriate. Blood pressure should be strictly controlled and antihypertensive medications used when necessary. Relevant neuroimaging should be obtained, including magnetic resonance imaging (MRI) within 24 hours following a tumor resection, angiography following treatment of vascular pathologies, and head CT to address any suspicion of hematoma formation, brain swelling, or development of hydrocephalus. Most importantly, the neurologic examination should be followed closely. Sedating medications should be avoided whenever possible and any deterioration in neurologic status should be evaluated immediately.

◆ **Complications and Their Avoidance in Posterior Fossa Surgery**

Neurosurgical procedures addressing posterior fossa pathology carry a higher incidence of neurologic complication when compared with supratentorial procedures. Dubey et al.\(^4\) report an overall complication rate of 31.8% in all posterior fossa surgeries, with CSF leaks occurring with the highest frequency, followed by meningitis, wound infection, and cranial neuropathies. The increased risk for complication in this area is likely related to the close proximity of critical neurovascular structures as well as the CSF fluid dynamics present in the region of the posterior fossa. Tumors in close proximity to the cranial nerves and surrounding critical vascular structures make complete surgical excision challenging, if not impossible. It is especially important when operating in this region of the central nervous system to keep in mind that a preserved quality of life holds greater value than total tumor excision.

**Cerebrospinal Fluid Leaks**

Cerebrospinal fluid leakage after a surgical approach is the most common complication of posterior fossa surgery, with a reported incidence of 4 to 17%\(^9,14,15\). Factors that may predispose a patient to a higher risk of CSF leak include larger tumors, dural invasion of the tumor, abnormal underlying CSF hydrodynamics (i.e., hydrocephalus), intraventricular hemorrhage, brain edema, and elevated intracranial pressure. CSF leak following a retromastoid approach can drain through open mastoid air cells into the middle ear cavity, and then through the eustachian tube to present as CSF rhinorrhea. This type of leak has been referred to as paradoxical CSF rhinorrhea. Faithful wax occlusion of all mastoid air cells during the surgical approach successfully avoids this complication. Watertight dural closure should be the goal following any posterior fossa approach. If the dural edges cannot be completely approximated, the defects should be repaired with a form of autologous graft such as muscle or pericranium. Additionally, many forms of dural graft matrix are now produced and can also be used for dural defects. Dural sealants can be used to reinforce a closure and have demonstrated their value in the literature with significantly lower rates of associated CSF leakage.\(^14,16\)

If CSF leakage through the wound occurs, conservative management should be initiated immediately, including wound resutting, application of a pressure dressing, bed rest, and elevation of the patient’s head. Should the leak persist, the patient may need to be treated with lumbar drainage, or rarely surgical repair. Early recognition and treatment of known CSF leakage is important to avoid a secondary infection.

**Wound Infection and Meningitis**

The incidence of postoperative infection following posterior fossa surgery is far higher than infection following supratentorial craniotomies. This is likely due to the increased incidence of CSF leakage. Dubey et al.\(^4\) reported a 13.6% incidence
of all postoperative infections, including superficial, deep, and bone flap infections as well as meningitis. The majority of postoperative infections result from contamination of the sterile field during the surgical procedure. Other risk factors for this complication include persistent CSF leak, presence of a foreign body, prolonged surgical time, long-term steroid usage, diabetes, reoperation, and cytotoxic therapy. If meningitis occurs, early diagnosis and treatment with appropriate antibiotics is critical to avoid neurologic sequelae.

To minimize the risk of postoperative infection, a broad-spectrum antibiotic should be given prior to making the skin incision during the surgical procedure and for the immediate postoperative period. Standard sterile techniques, meticulous wound closure, and proper postoperative wound care minimize the risk of infection.

### Aseptic Meningitis

Aseptic meningitis may mimic the clinical presentation of bacterial meningitis but does not demonstrate CSF pleocytosis, hypoglycemia, and hyperproteinemia. Unlike with bacterial meningitis, however, the cultures and Gram stain will be negative. Aseptic meningitis is a diagnosis of exclusion and is treated with steroids. Antibiotic treatment is unnecessary once bacterial meningitis has been ruled out.

The etiology of this complication is unclear, but associations have been made between its development and the presence of blood or tumor cyst fluid in the basilar cisterns. It typically presents 4 to 7 days postoperatively, and there have been reports of resolution of symptoms following the closure of a concurrent pseudomeningocele.

### Cerebellar Mutism

Cerebellar mutism is an uncommon but well-recognized complication after posterior fossa surgery; however, the precise mechanism and anatomic basis for this deficit remains unclear. This complication occurs most commonly in children after removal of tumors involving the cerebellar vermis. The patient is typically mute from day one to several days following surgery, and the symptom subsequently resolves over a period of weeks to months. Suspected causes include damage to the dentate nucleus, disruption of the dentatothalamocortical pathway, injury to the median structures of the cerebellum, and postoperative vasospasm of the cerebellar arteries. Avoiding this complication entails implementing surgical approaches that do not involve incision of the vermis, and instead use other techniques such as approach through the cerebellomedullary fissure for access to the fourth ventricle. Minimizing retraction on the cerebellar hemispheres may also help to avoid this deficit.

### Pseudomeningocele

The incidence of pseudomeningocele following posterior fossa surgery ranges from 15 to 28%. CSF leaks through the dural closure and collects under the skin and tissue planes, resulting in a fluctuating fluid collection. If the incision breaks down, a CSF leak will result. An enlarging pseudomeningocele may indicate the development of hydrocephalus and warrant CSF diversion. An increased risk for developing postoperative pseudomeningocele has been associated with posterior fossa craniectomy rather than craniotomy.

### Cranial Nerve Palsies

Cranial nerve morbidity has been reported to occur in 4.8% of patients who undergo posterior fossa surgery. A postoperative cranial nerve deficit can result from nerve retraction, direct injury to the nerve, compromise of its blood supply, or postoperative vasospasm. Cranial nerves III, IV, and VI are more tolerant of manipulation and recover more easily than cranial nerves VII to XII. An injury to cranial nerves III, IV, and VI results in diplopia, and may require subsequent oculoplastic procedures. Damage to cranial nerve V is generally well tolerated with the exception of damage to V1, which can cause corneal ulcerations if the eye is not kept adequately lubricated. Cranial nerve VII is extremely sensitive to manipulation and prone to injury. Profound hearing loss can occur as a result, even in the presence of anatomic preservation of the nerve. Deficits of the lower cranial nerves (IX, X, XI, XII) are less frequent but can be associated with significant deficits, including difficulty swallowing and inability to protect the airway, resulting in the need for feeding tube placement and tracheostomy to avoid aspiration pneumonia. Maintaining the continuity of the nerve during the surgical procedure provides the best chance of functional recovery should the nerve be otherwise injured. Intraoperative neurophysiologic monitoring of the cranial nerves enables the surgeon to minimize trauma to the nerves and maintain anatomic preservation. Several studies have demonstrated the importance of neurophysiologic monitoring in reducing the incidence of postoperative cranial nerve deficits.

### Postoperative Hematoma

Postoperative hematoma has been reported to occur in 3% of posterior fossa surgeries. This complication is usually associated with resection of parenchymal lesions and inadequate tumor bed hemostasis. Patients typically present with depressed consciousness or focal neurologic deficit. Mass lesions are poorly tolerated in the posterior fossa, and early recognition with postoperative neuroimaging and surgical evacuation are critical to preventing permanent or devastating neurologic damage. Meticulous operative technique, control of coagulopathy, and tight perioperative blood pressure control minimize the occurrence of this complication.

### Posterior Fossa Edema

Posterior fossa edema is a postoperative complication that results from direct manipulation of the brain tissue. The amount of edema seen in the brain tissue is directly related to the length and force of tissue retraction that occurred during the
surgery. If excessive retraction is used to obtain exposure, tissue damage will occur, leading to subsequent edema. Brain edema is a serious complication and can lead to neurologic deficits. Proper patient positioning, hyperventilation, high-dose corticosteroids, adequate bone removal, CSF drainage, diuretics, and intermittent retraction can help minimize the occurrence of this complication. Preservation of vascularure whenever possible and the use of limited coagulation and delicate handling of nervous tissue will also reduce the incidence of postoperative edema.

**Hydrocephalus**

The incidence of new-onset postoperative hydrocephalus following a posterior fossa surgery has been reported at 4.5%. The majority of these patients developed hydrocephalus as a result of another postoperative complication, such as edema, postoperative hematoma, infection, direct CSF obstruction, or impaired CSF absorption after spillage of blood into the CSF cisterns and subarachnoid space. Prior cranial surgery or prior radiation may also increase the risk of developing this complication. Postoperative hydrocephalus can present with headache, nausea/vomiting, gait disturbance, abducens nerve palsy, or an enlarging cranium and bulging fontanel in young children. The development of hydrocephalus may manifest as CSF leakage, and should always be suspected in the setting of a leaking wound. This complication can be minimized by preventing other forms of postoperative complications such as hematoma, edema, and infection. The treatment is CSF diversion via shunting procedure.

**Conclusion**

The basic approaches to the posterior fossa can be used to treat a wide range of pathologies. The frequency of complications is dependent on the type, size, and location of the pathologic lesion as well as the careful handling of neurovascular structures and the fastidious exposure and closure of the surgical wound. Meticulous microsurgical technique is critical to keep morbidity to a minimum. Learning the nuances and pitfalls associated with these procedures enables the neurosurgeon to treat posterior fossa pathology safely and effectively.

**References**

Petroclival meningiomas are among the most difficult cranial base lesions to treat. They are located deep in the skull base, have an unpredictable growth pattern, and often envelope multiple cranial nerves as well as important venous and arterial structures. Although the advent of skull base approaches has heralded a new era in the treatment of these lesions, the perioperative morbidity and mortality of such tumors remains significant. Several classification schemes have been proposed for petroclival meningiomas. Sekhar et al divide these lesions based on their site of origin and anatomic extension. Tumors are placed into three categories: those involving the upper clivus, those involving the middle clivus, and those involving the lower clivus. Based on the site of origin of the meningioma, the appropriate surgical approach may be chosen. This chapter presents four surgical options for removal of a petroclival tumor, along with their respective advantages and disadvantages.

**Posterior Transpetrosal Approach**

**Indication**

The posterior transpetrosal approach is the most versatile approach to petroclival meningiomas. It is the preferred surgical option for large petroclival tumors, especially meningiomas. This procedure provides access to the entire clivus as well as to tumor extension into Meckel’s cave and the posterior cavernous sinus. This approach combines a mastoidectomy with a supra- and infratentorial craniotomy. The petrosectomy options include a retrolabyrinthine resection, which preserves hearing but provides a small avenue in which to work. The translabyrinthine technique sacrifices hearing and provides a somewhat larger working area. The transcochlear technique, which provides the largest working area via maximal petrous resection, sacrifices hearing and transposes the facial nerve.

The drawbacks of the posterior transpetrosal approach are the additional surgical time required for the exposure, possible injury to the vein of Labbé, the risk of temporal lobe contusion, and cerebrospinal fluid (CSF) leak. Prior to performing this approach, the entrance point of the vein of Labbé into the transverse sinus should be noted on the preoperative gadolinium-enhanced magnetic resonance imaging (MRI) scans. A more posterior entrance into the transverse sinus is favorable. If the vein of Labbé enters the dural sinus anteriorly at the transverse-sigmoid sinus junction, the exposure provided by this approach will be limited unless the sigmoid sinus can be divided, or the mastoidectomy is extended to include a translabyrinthine or transcochlear exposure. A preoperative arteriogram is sometimes needed to determine the location of these venous structures when their size and anatomic course is not apparent on the MRI. The patency of the torcular should be assessed before surgery to determine the relative importance of the ipsilateral sigmoid sinus–jugular vein drainage system.

**Procedure**

The patient is placed supine with the head turned 60 to 80 degrees to the side opposite the tumor. A roll is placed under the ipsilateral shoulder. The neck is slightly extended such that the temporal lobe will drop away from the middle fossa floor without much retraction. The abdomen is prepped for obtaining a fat graft to fill the mastoidectomy defect. Electrodes are placed for monitoring the facial nerve and sixth cranial nerve if desired. Auditory evoked brainstem recordings can also be performed. The incision should be varied depending on the anterior extent of the tumor. If the tumor extends significantly into the middle fossa, Meckel’s cave, or cavernous sinus, the incision should start in front of the tragus, extend superiorly behind the temporal hairline, and then curve posteriorly just below the origin of the temporals.
muscle. The incision should then curve inferiorly across the region of the transverse sinus and then down behind the mastoid process (Fig. 12.1A). If the tumor does not extend into the middle fossa, Meckel’s cave, or cavernous sinus, then a C-shaped incision can be used (Fig. 12.1B). The skin flap is elevated anteriorly above the plane of the temporalis fascia and periosteum of the mastoid process. An anterior-based temporalis muscle–parietal periosteum flap is elevated. A mastoid periosteum–sternocleidomastoid muscle flap is elevated and retracted inferiorly (Fig. 12.2).

The sequence of bone work varies among surgeons. We prefer to start with the mastoidectomy. This enables us to unequivocally identify the location of the transverse sinus, which decreases the likelihood of this structure being injured while turning the craniotomy. The mastoidectomy must include removal of the bone along the dura of the middle and posterior fossa down to the level of the labyrinth, as well as the bone over the sigmoid sinus. Every millimeter of bone around the labyrinth should be removed, as this significantly improves the exposure of the base of the tumor. Exposing the distal sigmoid sinus just proximal to the jugular bulb is required to allow adequate posterior displacement of the sigmoid sinus. The dura is stripped off the undersurface of the convexity temporal, parietal, and occipital bones with a Penfield 3 dissector. The transverse sinus is stripped off the overlying calvaria. A trough of bone is removed with a cutting bur in the line of the planned craniotomy cut, over the posterior transverse sinus, exposing the dura both above and below the sinus. If the convexity dura is adherent to the skull, an additional trough-shaped bur hole is made over the superior temporal lobe. A one-piece supra- and infratentorial craniotomy is turned and the bone flap elevated (Fig. 12.3). Intravenous mannitol can be administered to aid in brain relaxation.

The amount of temporal bone removed can be varied to improve the exposure as needed. There are several options, as described in the following subsections.

**Retro labyrinthine Petrosectomy**

To preserve hearing, the integrity of the labyrinth is maintained. In addition to skeletonizing the lateral semicircular canal as already described, the posterior and superior canals are also skeletonized. Drilling is taken as far anterior as possible, exposing the maximal area of dura. This route provides good access to the cerebellopontine angle and lower clivus but does not provide direct visualization of the bony surface of the middle and upper rostral clivus.

**Trans labyrinthine Petrosectomy**

The translabyrinthine approach, which sacrifices hearing, is accomplished via the same basic mastoidectomy already described, but the semicircular canals are also removed. The three canals are skeletonized and drilled away. Drilling then continues forward so that the posterior aspect of the internal auditory canal also can be skeletonized. The bone overlying
the labyrinthine and mastoid segments of the facial nerve is also thinned. These maneuvers should be performed under continuous facial nerve monitoring.

Transcochlear Petroectomy

This technique provides the maximal area of exposure by completely removing the petrous bone. The external auditory canal is transected and oversewn. The initial drilling is as described for the translabyrinthine technique. The facial nerve canal is completely skeletonized and opened to allow the removal of the facial nerve. The greater superficial petrosal nerve (GSPN) can be divided to permit the posterior transposition of the facial nerve. This maneuver moves the facial nerve out of the surgical corridor and allows the complete removal of the internal auditory canal and cochlear apparatus. Alternatively, the GSPN can be preserved and the course of the facial nerve skeletonized. This maneuver preserves the blood supply of the facial nerve and usually does not produce a postoperative facial paralysis. However, the facial nerve then remains directly in the surgeon’s working space throughout the procedure. Drilling then continues until

**Fig. 12.3** (A) Mastoidectomy and small troughs drilled over the transverse sinus and along the outline of the bone flap. (B) Bone flap turned. (C) Posterior transpetrosal approach bone flap removed. (D) Dural opening. (E) Exposure of a petroclival meningioma medial to cranial nerves V and VIII.
the internal carotid artery has been skeletonized up to the siphon. This technique provides extensive exposure of the petroclival region at the cost of sacrificing hearing and a facial nerve paresis or paralysis if the GSPN is sectioned. This route is also associated with a significant risk of CSF fistula.

Once the bony exposure is complete, the dura is opened along the inferior edge of the temporal lobe and along the presigmoid sinus dura. The superior petrosal sinus is divided with the bipolar coagulator if it is small and tantalum vascular clips if large. The dural opening then proceeds posteriorly along the superior aspect of the transverse–sigmoid sinus junction, to the point just anterior to the entrance of the vein of Labbé into the dura near the transverse sinus. Great care must be taken to avoid injury to the vein of Labbé. The tentorium is then divided from a lateral to medial direction just behind the posterior edge of the superior petrosal sinus. Temporal bridging veins entering the tentorium should be saved. The medial edge of the tentorium is divided just behind the entrance of cranial nerve IV into the tentorial edge. Once the tentorium is divided, the sigmoid sinus can be displaced posteriorly, and the exposure improves substantially.

After the dura and arachnoid have been opened to permit CSF drainage and relaxation of the brain, the tumor is exposed. The surgeon must orient himself or herself because the normal appearance of the anatomy can be significantly distorted. Tumor removal proceeds according to general neurosurgical principles. Primarily, early devascularization is combined with central debulking. Tumor removal is accomplished by working in the spaces between the cranial nerves. Most petroclival meningiomas receive their vascular supply from the meningocephalophyseal trunk off the internal carotid artery. This area of the tumor along the petrous apex should be attacked first. After the initial debulking, dissection proceeds along the tumor capsule. Dissection must remain within the arachnoid planes if the tumor is to be separated successfully from critical vascular and neural structures.

A combination of suction, bipolar coagulation, microscissors, ultrasonic aspiration, and/or CO₂ laser are used for the major debulking. During the debulking, the surgeon must maintain a clear mental picture of the expected anatomy and possible location of displaced structures. In particular, the abducens nerve and the basilar artery can be embedded within the tumor and difficult to identify. Tumor debulking proceeds from less critical superficial areas to deeper regions where critical structures may be found.

After generous debulking and relaxation, the tumor capsule is dissected free from the surrounding structures. Meticulous care must be taken to maintain the dissection within the arachnoid planes, however difficult it may be to achieve this goal. Frequently, one must alternate between further debulking and capsule removal. When most of the tumor has been debulked, the thinned-out capsule is carefully dissected free of surrounding structures.

With large extensive tumors, the lower cranial nerves are often involved along the lower pole of the tumor. The facial and vestibulocochlear nerves are usually displaced laterally and inferiorly. Dissection along these nerves can be enhanced with the use of monitoring and a facial nerve stimulator. The abducens nerve is typically within the tumor or stretched anteriorly. It is best to identify this nerve proximally and follow it distally into its dural entrance. Similarly, the trigeminal and trochlear nerves are identified. Occasionally, the dura over Meckel’s cave is opened to remove tumor that has grown into this region.

The basilar artery is usually within the tumor or displaced contralaterally. Utmost care must be exercised when dissecting this vessel from the tumor capsule, particularly its critical perforator vessels. It is often wiser to leave a remnant of firmly attached capsule to the basilar artery than to risk neurologic devastation from injuring the perforators or the basilar artery itself.

Closure begins with an attempt to repair the dura in a watertight fashion. If a dural cuff exists deep in the exposure, around the labyrinth, then a free piece of parietal periosteum or temporalis fascia is used to repair the dura primarily. If there is no deep dura to sew to, the temporalis muscle–peristomial periosteum flap is rotated down into the mastoid defect and tucked to the dural edges as possible. An additional 2 cm by 2 cm patch of temporalis fascia can be placed over the mastoid antrum, to prevent the temporalis muscle or abdominal fat graft from bulging into the middle ear onto the incus. Some surgeons prefer to use hydroxyapatite bone cement to seal the mastoid antrum. The mastoid defect is then filled with an abdominal fat graft. Titanium mesh can be placed over the fat graft to prevent a sunken mastoid area (Fig. 12.4). Placing mesh over a fat graft in the mastoid must be done with caution. Any sharp edges on the mesh must be bent inward, or over time they will protrude through the scalp as the fat graft atrophies and surrounding skin retracts. There is also anecdotal evidence that mesh, when used over a fat graft in the mastoid, can contribute to late postoperative CSF leak. As the fat graft in the mastoid atrophies, it can adhere to the mesh. This causes the fat to pull away from the presigmoid dura as well as the antrum/middle ear, allowing CSF to leak down the eustachian tube. If mesh is used, the sternocleidomastoid periosteum flap is then sutured back into its normal position over the mesh. The skin is closed in the usual fashion.
If hearing is impaired preoperatively, this approach can be combined with removal of the labyrinth and/or cochlea. This improves access to the base of the tumor and the clivus. In patients with good hearing preoperatively, some surgeons advocate a partial posterior labyrinthectomy to improve basal exposure, with reasonable hearing preservation rates.

**Frontotemporal/Orbitozygomatic Craniotomy**

**Indication**

This approach is reserved for petroclival meningiomas with primarily supratentorial extension. Its strength lies with its familiarity to the neurosurgical community, the width of exposure, and the ability to work from multiple angles. The drawbacks of this approach are the potential for injury of the frontal branch of the facial nerve, the danger to orbital contents, the potential need for extensive arachnoid dissection with manipulation of the frontal and temporal lobes, the risk of injury to sylvian vessels, and poor access to the middle and lower clivus.

**Procedure**

The head is rotated from 30 to 60 degrees depending on the location of the lesion. Positioning is such that the malar eminence represents the zenith of the operative field. The incision starts at the inferior border of the zygoma and extends in a gentle curve to the level of the contralateral mid-pupillary line, terminating behind the hair line. The skin is elevated anteriorly until the superficial temporal fat pad is encountered. Further dissection anteriorly may lead to injury of the frontalis branch of the facial nerve. The temporal fascia is incised in the line of the skin incision to the level of the superior temporal line. Next the fascia is incised anteriorly approximately 0.5 cm below the superior temporal line, terminating just behind the lateral orbital rim. This leaves a generous cuff of muscle for reattachment of the temporalis muscle at the end of the operation and protects the frontal branch of the facial nerve. The temporal fascia is dissected off the underlying muscle so as to expose the zygoma, the malar eminence, and supraorbital rim. The underlying muscle is then dissected from the cranium and is retracted inferriorly. The periorbita is dissected beginning at the lateral edge of the orbital rim and proceeding medially. At the end of the dissection the entire orbital rim lateral to the supraorbital nerve, the malar eminence, and the zygoma should be completely exposed. An orbitozygomatic (OZ) osteotomy significantly increases the exposure. It can be performed as one or two pieces. In our practice it is completed in two separate steps (Fig. 12.5). First a standard pterional craniotomy is performed. The reciprocating saw is used to perform the OZ osteotomy. Six cuts are required for the removal of the OZ rim. The first cut is made at the base of the zygomatic arch. The second cut begins at the inferolateral edge of the zygoma near the malar region and proceeds halfway along the malar eminence to the lateral orbital rim. A third cut begins intraorbitally in the inferior orbital fissure and extends posterolaterally until it meets the edge of the second cut. The fourth cut begins intraorbitally 1 to 2 mm lateral to the supraorbital notch and proceeds posteriorly along the orbital roof approximately 4 cm and medially to the superior orbital fissure. A fifth cut is made from the inferior orbital fissure posteriorly to the temporal region. The final cut is made from the superior orbital fissure inferiorly to meet the previous cut. After the OZ osteotomy is completed, the sphenoid wing is drilled so as to allow for an unobstructed view. The dura is then opened in a standard frontotemporal fashion. After the intradural resection is completed the dura is closed and the bony components replaced and fixed into their anatomic location. The muscle, fascia, and skin are reapproximated in the standard fashion.

**Middle Fossa Approach**

**Indication**

This approach is reserved for small lesions that extend into the middle fossa and posterior cavernous sinus, with an inferior extent that is above the internal auditory canal. The main advantage of this approach is that it is fast and is primarily extradural in nature. The drawbacks are the risks of CSF leak, injury to the carotid artery, retraction injury to the temporal lobe, and limited inferior exposure along the clivus.

**Procedure**

The head is placed so that the zygoma is parallel to the floor. The vertex of the head may be tilted 10 to 20 degrees below the horizontal so that gravity allows the temporal lobe to fall away from the temporal fossa floor. An incision is made from in front of the tragus extending superiorly and slightly posteriorly, until it is above the origin of the temporalis muscle. A temporal craniotomy is turned that is flush with the middle fossa floor (Fig. 12.6). The inferior exposure usually enters the mastoid air cells. Some authors recommend resection of the root of the zygomatic arch including the glenoid fossa, the roof of the external ear, and the superior third of the mastoid process. The approach is extradural and is assisted greatly by the use of a dedicated middle fossa retractor. There are six key structures that must be identified during this exposure. The first is the posterior petrous ridge. This ridge determines the posterior aspect of the exposure. The second is the arcuate eminence, which overlies the superior semicircular canal. The third is the GSPN, which is left in its place along the middle fossa floor by dissecting the dura off the temporal bone in a posterior to anterior direction, starting along the posterior petrous ridge. Care should be taken to avoid traction on the greater superficial nerve and geniculate ganglion. Next the middle meningeal artery is identified and divided. This maneuver allows additional anterior extradural dissection exposing the mandibular division of the trigeminal nerve as it travels into the foramen ovale. The fifth structure identified...
is the petrous carotid artery. This is usually found medial and just behind the third division of the trigeminal nerve as it enters the foramen ovale. It is often incompletely covered by dehiscent bone or cartilage. The final structure to be identified is known as the trigeminal impression. This bony depression over the petrous apex is made by the trigeminal nerve as it enters Meckel's cave, and helps to define the top of the medial petrous apex.

Fig. 12.5 The two-piece orbitozygomatic approach. (A) Scalp incision. (B) Osteotomy cuts. (C) Two-piece orbitozygomatic bone flaps. (D) Exposure of a petroclival meningioma by the orbitozygomatic bone flap. (E) View after resection of a petroclival meningioma.
All drilling should be performed carefully, usually with a high-speed diamond drill. First the petrous apex is removed exposing the posterior fossa dura. The bone removal proceeds until the internal auditory canal is exposed posteriorly, and the posterior edge of the horizontal segment of the internal carotid artery is exposed anteriorly. Care must be taken to avoid entrance into the cochlea, if this structure is to be preserved. This results in exposure through Kawase's triangle, which provides access to the superior clivus. This region is defined anteriorly by the mandibular division of the trigeminal nerve and the internal carotid artery, posteriorly by the internal auditory canal and the superior semicircular canal, laterally by the cochlea and GSPN, and medially by the petrous ridge. The dura along the lateral edge of the third
division of the trigeminal nerve is opened and the dura elevated up off of V3. The dura above and below the superior petrosal sinus is opened next. The superior petrosal sinus is ligated and divided. The tumor is then removed, generally by internal debulking, followed by microsurgical capsular dissection. At the end of the intradural resection, the dura, craniotomy, muscle, and skin are closed in the standard fashion. Particular attention must be paid to any exposed air cells. If necessary, a temporalis muscle with attached parietal periosteum can be rotated in under the temporal lobe to seal the mastoid and petrous apex air cells. Meticulous use of bone wax or bone cement may also help minimize the risk of postoperative CSF leak.

◆ Suboccipital Approach

Indication

This approach is reserved primarily for small tumors limited to the infratentorial space that are more petrous than clival in origin. It also can be used in older patients who have some degree of cerebellar atrophy, thus limiting the need for cerebellar retraction. It allows excellent visualization of the petrous bone in a relatively rapid fashion. When modified to a far lateral approach, it extends the operative field to include the lower clivus, including the foramen magnum.23,24 Drawbacks of this approach are a smaller working space, the risk of injury to critical arteriovenous structures and cranial nerves, and CSF leak. The far lateral approach may also result in atlanto-occipital instability if the majority of the occipital condyle is removed.

Procedure

The patient is placed in the lateral decubitus position with a roll under the ipsilateral shoulder. The neck is placed in slight flexion with rotation of the head, away from the surgical site. Care is taken to avoid overrotation and obstruction of the contralateral jugular venous outflow. A linear incision is placed approximately 2 to 3 cm behind the ear. The mastoid emissary vein is occluded with bone wax. A trough of bone is removed along the inferior border of the transverse sinus, as well as along the posterior border of the sigmoid sinus until the mastoid emissary veins entrance into the sigmoid is identified and divided (Fig. 12.7). The dura over the posterior fossa dura is then stripped off the bone with a Penfield 3. A footplate may then be used to complete the craniotomy. All mastoid air cells should be waxed. The dura is then opened along the sigmoid and transverse sinus, leaving a cuff of dura for resuturing at the end of the case. The cerebellar hemisphere is left covered with dura as much as possible.
The arachnoid over the cisterna magna is opened. This allows for removal of CSF, which provides decompression of the CSF spaces, increases the initial exposure, and limits the need for cerebellar retraction. The cerebellar cortex should be draped with a layer of cottonoid or Telfa. The surgeon can then proceed with arachnoid dissection and removal of the tumor. If intradural drilling is performed, packing of all exposed CSF spaces with Gelfoam and copious irrigation may help reduce the incidence of postoperative headache and nausea. Special care should be taken during the closing stages of this procedure. Durational closure should be watertight. Air cells should be meticulously waxed. Fascial and muscle closure should be performed in multiple layers. The skin is closed with a running stitch. All of these maneuvers are intended to minimize the risk of postoperative CSF leak.

Clinical Pearls

1. Turning a combined supra- and infratentorial bone flap without tearing the convexity dura or injuring the underlying venous sinuses can be difficult. Performing the mastoidectomy first aids in the exact identification of the location of the transverse sinus and makes the turning of this bone flap much easier.

2. When performing the posterior transpetrosal approach, elevating both an anterior-based temporalis muscle–parietal peristeum flap and an inferiorly based sternocleidomastoid peristeum flap during the initial exposure is very useful in performing a closure that minimizes the risk of postoperative CSF leak.

3. When performing the posterior transpetrosal approach, the entrance point of the vein of Labbé into the transverse sinus should be determined preoperatively. This can usually be determined from an MR/magnetic resonance venogram (MRV), but in some cases will require a preoperative arteriogram. The sigmoid sinus should be preserved, especially if the torcular is not patent.

4. When performing the posterior transpetrosal approach, temporal bridging veins entering the tentorium can be preserved by sectioning the tentorium anterior to their entrance into the tentorium and placing the temporal lobe retractor under both the tentorium and the temporal lobe.

5. A cosmetic mastoidectomy can be performed via removal of the outer table of the mastoid bone for later replacement or by covering the abdominal fat graft placed in the mastoid defect with titanium mesh. However, if mesh is used, all sharp edges must be turned inward, or they will protrude through the skin months or years later as the fat atrophies.

6. Preoperative embolization of a petroclival tumor can decrease intraoperative blood loss, allow for better visualization, and facilitate radical excision of the tumor while reducing the risk of neurologic morbidity. However, embolization of the meningohypophyseal trunk is difficult and should be performed only by the most skilled interventionalist. If the patient is not embolized, the area of the tumor closest to the meningohypophyseal trunk should be attacked first to devascularize the tumor as much as possible.

7. Drilling a trough through the skull along the inferior transverse sinus and posterior sigmoid sinus until the entrance of the mastoid emissary vein is ligated decreases the incidence of significant bleeding when turning a suboccipital bone flap with a footprint attachment.

8. Stereotactic radiosurgery allows the surgeon to compromise resection in favor of preservation of function. When critical structures are involved by the tumor that cannot be dissected free, a carpet of tumor may be left on these structures and the remnant tumor irradiated in the postoperative period. Radiosurgery has a very low morbidity rate and enables good tumor control of such remnants.

References


Suggested Reading

II Surgical Techniques


The jugular foramen at the base of the skull is an anatomically complex region. It is divided into two separate compartments. The posterior lateral compartment is the pars vascularis, which contains the sigmoid sinus, jugular vein, and the posterior meningeal artery. The anterior medial compartment is the pars nervosa, through which the glossopharyngeal, vagal, and accessory cranial nerves, and the inferior petrosal sinus course (Fig. 13.1). Successful treatment of tumors involving the jugular foramen mandates a clear understanding of the anatomy of the jugular foramen and the natural history of the tumors involved.

Since the first reported exploration of the jugular bulb by Seiffert in 1934,1 surgeons have been striving for the optimal approach and technique to facilitate complete extirpation of tumors of the jugular foramen. Earlier treatments focused on limited resection combined with radiation therapy or radiation therapy alone. The relative inaccessibility of the jugular bulb region, the potential for harming the surrounding neurovascular structures, and the extremely vascular nature of these tumors dictated treatment that was, by and large, palliative. As experience was gained, innovative surgical procedures were developed, most notably the combined lateral skull base approaches and the infratemporal approach. In addition to the advancements in surgical approaches, the implementation of adjunctive therapies, such as superselective embolization, gamma knife radiosurgery, and the aggressive treatment of postoperative neurologic deficits, has resulted in dramatic improvements in long-term outcomes.

Historical Overview

The development of surgical techniques aimed at the removal of jugular bulb tumors is intimately tied to the history of the surgical management of glomus jugulare tumors. The seminal work on the glomus jugulare, or jugular body, was reported in 1941 by Guild2 at the American Association of Anatomists meeting in Chicago. He described nests of “blood vessels of capillary or of pre-capillary caliber with numerous epithelioid cells between the vessels” located near or in the wall of the jugular bulb. During the 1950s, several authors communicated their efforts in treating glomus jugulare tumors with mostly discouraging results.3,4 The intricate anatomy of the jugular bulb region and the risk of hemorrhage from the tumor, in combination with the lack of high-definition imaging studies to elucidate tumor margins, were significantly limiting.

In the early 1960s, the introduction of imaging techniques, with advances in diagnostic technology, enabled one to detail the surgical anatomy of the jugular foramen affected by neoplasia. A significant reduction in the morbidity and mortality with improved surgical accuracy resulted.3 Arteriography,5 polytomography,6 and retrograde venography7 allowed the surgeon to “view” the tumor and select the most judicious approach for treatment. Shapiro and Neues8 in 1964 reported their experience with a patient with a recurrent glomus jugulare tumor. They accomplished complete removal of the tumor involving the jugular bulb region with translocation of the facial nerve. Unlike earlier reports, there was minimal blood loss, and neurologically the patient did well. Gejrot9 described a similar procedure in 1965 in a series of four patients. These reports were important in that they established the foundation for current surgical techniques. They proved that tumors in the jugular bulb area could be removed with preservation of neural function. One critical contribution in Gejrot’s approach, which persists as a crucial step of modern surgical treatment of jugular bulb tumors, was that he stressed the importance of maintaining the medial wall of the sigmoid sinus at the level of the jugular bulb to protect the lower cranial nerves.

Further interest and refinement of surgical technique continued through the ensuing years. Gardner et al10 in 1977 detailed an operative technique with a combined lateral skull base approach utilizing a multidisciplinary team. The approach consisted of three phases. Exposure of the skull base through the neck was performed first, followed by bone re-

Historical Overview

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moval within the temporal bone and the jugular fossa. Lastly, tumor removal was accomplished with wound reconstruction to follow. In 1977, Fisch introduced the infratemporal approach to gain complete access to the internal carotid artery within the temporal bone, which was a major limitation of the earlier approaches. The exposure and control of the carotid artery allowed for larger glomus jugulare tumors to be treated with increased safety. Furthermore, Fisch added a classification scheme for glomus tumors of the temporal bone, which he revised in 1981 (Table 13.1).

A major advancement in the surgical treatment of glomus jugulare tumors occurred with the development of preoperative superselective embolization of the external carotid arterial supply of these highly vascular tumors. First introduced by Hilal and Michelsen and Brismar and Cronqvist, Simpson et al reported in 1979 the use of preoperative embolization in glomus jugulare tumors in an effort to reduce operative blood loss. Murphy and Brackmann substantiated the use of preoperative embolization in a 1989 report analyzing 35 patients. They concluded that there was a significant reduction in the operative blood loss and operating time, and embolization led to a higher rate of complete resection of tumors. However, there did not appear to be a reduction in the risk of injury to the lower cranial nerves. As for the morbidity of the embolization procedure, the current state of technology and expertise in interventional radiology has significantly reduced the incidence of stroke and cranial nerve injury experienced during the earlier years of application.

**Tumor Types**

The most common tumors involving the jugular foramen are paragangliomas, schwannomas, and meningiomas. Although appearing histologically benign, glomus jugulare tumors are characterized by unpredictable biologic behavior. Typically arising from glomus bodies positioned in the dome of the jugular bulb, the neoplasm is intimately associated with critical neurovascular structures at the skull base.

Schwannomas of the jugular foramen arise from cranial nerves IX, X, and XI. These nerves exit the brainstem as a series of rootlets from the retro-olivary sulcus and then traverse to the jugular foramen in an inferolateral direction. Symptoms depend on the cranial nerve involved, the site of origin, and the extent of spread. Jugular foramen schwannomas can be classified according to their growth patterns as described by Kaye et al (Table 13.2).

Meningiomas are the third most common tumor of the jugular foramen. Arising from the arachnoid cap cells, they typically originate from the dura overlying the posterior aspect of the petrous bone. Growth often occurs intracranially with subsequent involvement of the jugular foramen. Less common, they may originate within the jugular foramen itself. Intracranial and extracranial spread is possible.

**Patient Selection**

Achieving an optimal treatment outcome is directly related to the proper choice of a management strategy unique for each patient. A multidisciplinary team utilizes a comprehensive and selective approach in defining the appropriate treatment regimen for a particular lesion. Risk factors considered include the patient’s age and medical condition, the extent of tumor, and the presence of neurologic deficits.

Symptomatic elderly patients or those with significant medical conditions are best treated with palliative radiotherapy, despite the lack of conclusive data establishing its efficacy. In extensive tumors in which it is felt that significant postoperative morbidity will occur, subtotal resection with or without postoperative radiation may be the treatment of

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**Table 13.1** Fisch Classification of Glomus Tumors of the Temporal Region

| Type A | Tumors limited to the middle ear space |
| Type B | Tumors limited to the middle ear or mastoid without involvement of the infralabyrinthine space of the temporal bone |
| Type C | Tumors involving the infralabyrinthine and apical spaces of the temporal bone, with extension into the apex |
| Type D1 | Tumors with intracranial extension less than 2 cm in diameter |
| Type D2 | Tumors with intracranial extension greater than 2 cm in diameter |

**Table 13.2** Growth Patterns of Jugular Foramen Schwannomas

| Type A | Tumors that are primarily intracranial with only a small extension into bone |
| Type B | Tumors in which the main mass is within bone, with or without an intracranial component |
| Type C | Tumors that are primarily extracranial with only a minor extension into bone or into the posterior fossa |
The aggressiveness of surgical management has advanced with selective embolization of large vascular tumors and is heavily dependent on the experience and skill of the surgeon. In selected patients with static cranial nerve deficits and no evidence of brainstem compression, observation alone may be warranted.

Once the decision is made for surgical treatment, judgment must be used with regard to subtotal versus gross total resection of a tumor. Subtotal resection of a tumor is the choice of treatment if infiltration of surrounding neurovascular structures is present or anatomic planes are absent. This is especially true in patients with no preoperative cranial nerve deficits. If surgical planes can be established between the tumor and the surrounding anatomy, total resection of a tumor may be accomplished with minimal morbidity.

◆ Preoperative Management

The surgical removal of tumors of the jugular foramen requires a complete and thorough preoperative evaluation involving a multidisciplinary team. Careful attention is given to the medical condition of the patient, tumor size, involvement of the surrounding neurovascular structures, and vascular supply of the tumor. In assessing glomus tumors, one must determine the presence or absence of catecholamine secretion and the presence of multiple tumors.

Plain skull x-rays are of limited benefit, but they may show enlargement of the jugular foramen. A computed tomography scan with thin cuts defines the extent of temporal bone involvement by the tumor and the altered anatomy of the jugular foramen. Glomus jugulare tumors typically result in permeative, erosive margins (Fig. 13.2). Schwannomas of the jugular foramen produce smooth, bony margins through an expansile process (Fig. 13.3). Meningiomas may be calcified, and there can be associated hyperostosis of the bone (Fig. 13.4). Magnetic resonance imaging with and without contrast reveals excellent definition of the soft tissue component of the tumor at the level of the skull base and extracranial space (Fig. 13.5).

Four-vessel cerebral angiography should be performed in all cases suspicious for glomus jugulare tumors as a defini-
One often finds rich arterial contributions to the tumor, mostly from the ascending pharyngeal and caroticotympanic arteries (Fig. 13.6). In large tumors, a significant arterial contribution may be found from the internal carotid artery vessel wall when the artery is encased by tumor growth. An examination of the late venous phase is mandatory to determine the patency of the sigmoid-jugular venous system and to identify unusual collateral venous outflow that has developed in response to the tumor. The contralateral jugular vein should be visualized, as it may be congenitally small or absent and blocked in cases of bilateral glomus jugulare tumors. The sigmoid-jugular venous system may be obstructed at any level from compression or intraluminal invasion by tumor. Evaluating the intraluminal extent of the tumor aids in determining the proper level for ligation of the sigmoid sinus and jugular vein to prevent venous tumor embolization when surgically removing a glomus jugulare tumor.

Patients with tumors that encase or displace the carotid artery may be evaluated for the possibility of arterial sacrifice or bypass (Fig. 13.7). An assessment of the collateral circulation during the arterial phase of the angiogram gives one initial insight into whether or not the patient would tolerate permanent occlusion of the internal carotid artery. The gold standard for determination is temporary balloon occlusion, which may be performed during the angiographic study. Temporary occlusion of the internal carotid artery identifies the majority of individuals tolerant of carotid artery sacrifice intraoperatively. However, there is no single test or combination of tests that can absolutely ensure the absence of a cerebrovascular accident following occlusion of an internal carotid artery. If the patient fails the test occlusion, an elective arterial bypass prior to tumor removal would be recommended.

The effectiveness of radiosurgery during the past decade for treatment of the majority of the common tumors that occur in the jugular foramen region has resulted in the rare sacrifice of the internal carotid artery to completely remove a benign, slow-growing tumor of the jugular foramen region. In the majority of skull base centers that manage tumors of the jugular foramen that encase the internal carotid artery, surgical subtotal removal of the tumor is the preferred approach followed by radiosurgical treatment of residual tumor involving the internal carotid artery.

A major advancement in the surgical treatment of glomus tumors over the past two decades has been the development and application of preoperative superselective embolization.
Performed by the interventional radiologists 24 to 48 hours prior to surgery, embolic materials or coils are used to selectively occlude the arterial feeders of these complex vascular tumors. The current state of technology and expertise in interventional radiology has significantly reduced the incidence of stroke and cranial nerve injury experienced during the early years of application.

In a minority of cases, glomus jugulare tumors secrete catecholamines. An excess of catecholamines can elevate the blood pressure. Manipulation of the tumor during surgery may result in significant intraoperative hypertension that may be uncontrollable. A preoperative 24-hour urine specimen is performed to detect the presence of vanillylmandelic acid, metanephrines, and free catecholamines. If present, a pharmacologic blockade is performed with the use of an alpha-blocker, and this is usually initiated 2 weeks before the operation. A beta-blocker is given approximately 24 hours before surgery to avoid tachycardia.

Other testing completed during the preoperative period includes routine audiometry to establish the function of the hearing apparatus. Routine laboratory studies should include a peripheral blood count and coagulation panel because of the highly vascular nature of these tumors with anticipated blood loss.

**Operative Treatment**

**Anesthesia and Monitoring**

The goal of general anesthesia for glomus jugulare tumors is to provide a quiet, surgical field and ample working space for multiple surgical teams. One must be prepared for a lengthy operation involving multiple stages that may involve considerable blood loss and cranial nerve dysfunction. A thorough discussion of the anesthesia plan, to include prolonged intubation and invasive monitoring, should precede surgery.

A smooth induction with a one-time dose of nondepolarizing muscle relaxant facilitates intubation with a reinforced endotracheal tube. After the airway is secured, a Dobbhoff tube (Covidien, Norwalk, CT) is placed. Continuous systemic monitoring is essential and includes placement of an arterial line (in the contralateral radial artery), a central intravenous (IV) line (placed in the femoral vein), a Foley catheter, and a rectal temperature probe. Central IV line insertion into the jugular vein of the neck is avoided because of the unilateral obstruction of the sigmoid-jugular venous system related to the tumor process. Facial nerve monitoring is performed routinely. Glossopharyngeal and vagal nerves are monitored in selective cases only if the size of the tumor suggests a potential for injury. Blood gases and electrolytes are evaluated periodically, as well as blood sugars when indicated. Maintenance of anesthesia is accomplished with a continuous infusion of narcotic (Sufenta) plus a lowered concentration of inhalational agent.

Emergence from anesthesia should include rigid blood pressure control. Strict maintenance of blood pressure control should occur in the first 12 hours in the postoperative period. We commonly use a continuous infusion of the calcium channel blocker Cardene for its ease of titration and short-acting effects.

In tumors involving the jugular fossa, in the absence of preexisting lower cranial nerve deficit, conservative airway management immediately postoperatively with continued intubation during the initial 12 to 24 hours is encouraged. In patients with long-standing cranial nerve deficits, with a surgery ending in less than 8 hours and no comorbidity considerations, a more aggressive approach to extubation may be considered. Patients with brainstem compression should be managed conservatively with regard to extubation.

If uncertainty of lower cranial nerve injury exists, direct fiber-optic visualization of the vocal cords at the time of extubation should be performed. The vocal cords may be swollen and edematous from extended intubation related to a surgical procedure exceeding 8 hours. In such cases, a vocal cord paralysis may not be obvious clinically, and the patient will initially appear to ventilate adequately. Within a 12- to 24-hour period following extubation, aspiration becomes apparent and the patient’s pulmonary status deteriorates.

**Positioning (Fig. 13.8)**

Positioning of the patient has two main goals: optimal surgical access by multiple surgical teams, and facilitation of venous drainage. We accomplish these goals by placing the patient supine in a flexed position, head up, with the legs slightly bent at the knees. The head is not placed in a fixed headrest; instead, the head and neck are available for turning as the surgeon desires. The arms are tucked at the patient’s side after placing each arm in slight flexion at the elbow with the hands open and flexed. Straps are padded with cushioning and placed across the patient’s chest, hips, and legs to allow for full tilt of the table in any direction.

**Operative Approaches**

The operative approach is dictated by the location and size of the tumor mass. The size of the tumor and the need for control of the carotid artery mandates individualization of the surgical technique. Three techniques will be described here: the combined lateral skull base approach, the modified lateral skull base approach, and the preauricular infratemporal fossa approach. Modifications of these approaches may be used for tumors with intracranial extension. These include retrosigmoid, presigmoid, retrolabyrinthine, translabyrinthine, and transcoclear approaches.

**Operative Procedures**

**Combined Lateral Skull Base Approach**

**High Cervical Exposure**

This approach is the procedure of choice for small or medium-size tumors that extend up to the level of the petrous internal carotid artery. The first portion of this procedure consists of a postauricular C-shaped incision that begins ap-
proximately 3 cm above the pinna and proceeds posteriorly 3 cm behind the helix of the ear. This incision is continued inferiorly onto the neck along the anterior border of the sternocleidomastoid muscle. The greater occipital nerve is often exposed in the operative field and may be harvested to be used later in the operative procedure as a nerve graft. The skin flap, in addition to the musculoperiosteal flap covering the suboccipital bone, is then rotated anteriorly. The external auditory canal is transected at the bony-cartilaginous junction, and the musculoperiosteal flap is used to close the meatus and create a blind pouch. After identification of the sternocleidomastoid muscle, dissection proceeds anteriorly. The facial vein is ligated and transected, and the common carotid artery, internal and external carotid arteries, internal jugular vein, cranial nerves X and XII, and the ansa cervicalis are isolated. These structures are followed superiorly, where cranial nerve XI is identified (Fig. 13.9). Transection of the sternocleidomastoid muscle from the mastoid tip and the posterior belly of the digastric muscle from the digastric groove are then completed. Elevating the tail of the parotid gland, the main trunk of the facial nerve can be identified at the stylomastoid foramen. Removal of the styloid process follows after transection of the stylopharyngeal, styloglossus, and stylohyoid muscles. The base of the styloid is preserved for anatomic orientation, and direct access to the high cervical carotid artery in the carotid space immediately below the skull base. Cranial nerve IX will be seen crossing over the anterior surface of the internal carotid artery at this level.

Palpation of the transverse process of the first cervical vertebra, the base of the styloid process, and the mastoid tip provides constant bony anatomic landmarks for orientation with respect to the jugular foramen. The jugular vein is found resting on the rectus capitis lateralis muscle, which extends from the transverse process of C1 to the posterior bony margin of the jugular foramen. The transverse process of the first cervical vertebra may be removed for exposure; however, care should be taken to identify and protect the vertebral artery. Cranial nerve XI is usually identified as it crosses over the anterior surface of the jugular vein at or above the level of the transverse process of C1 to enter the posterior margin of the sternocleidomastoid muscle. Soft tissue surrounding the jugular foramen may be removed, allowing a 270-degree control of the jugular vein (Fig. 13.10). The medial margin of the jugular foramen is not violated in the dissection to protect the lower cranial nerves as they exit the skull base.

◆ Temporal Bone Exposure

The initial step involves a simple mastoidectomy. The bone of the external ear canal is removed followed by disarticulation of the incostapedial joint. This facilitates removal of the
III Surgical Techniques

Fig. 13.9 High cervical exposure. The suction device is on the transverse process of the first cervical vertebra.

Fig. 13.10 Proximal and distal control of the jugular vein is obtained. A 270-degree exposure of the jugular foramen provides the surgeon excellent visualization of the neurovascular anatomy.

Fig. 13.11 The facial nerve (arrow) is obviously infiltrated with tumor as it descends through the fallopian canal.

tympanic membrane, malleus, and incus. Care is taken to protect the stapes. If tumor is present in the middle ear, it is mobilized inferiorly. One must take caution to remain below the labyrinth to avoid loss of hearing. In patients with non-serviceable hearing, removal of the cochlea and labyrinth may be necessary for tumor exposure. Skeletonization of the mastoid and tympanic segments of the facial nerve ensues, and the nerve is traced superiorly to the geniculate ganglion. Fibrous attachments along the course of the nerve are then cut sharply. Depending on the size and extent of the tumor, the greater superficial petrosal nerve and the chorda tympani may be sectioned, and the facial nerve mobilized anteriorly. Drilling of the anterior epitympanum to the soft tissue depth forms a tangential notch where the facial nerve may be placed. The proximal nerve lies within this groove and the distal portion within the parotid gland. Preservation of soft tissue at the stylomastoid foramen is critical to preserving the arterial blood supply from the stylomastoid artery to the facial nerve. Care is taken for the remainder of the procedure to protect the facial nerve for manipulation. If it is found that the nerve is directly involved with tumor during the exposure (Fig. 13.11), the diseased portion may be resected, and an interposition nerve graft (greater occipital nerve) may be used as a cable graft.

Bone overlying the sigmoid sinus is drilled away, and the presigmoid and retrosigmoid bone is removed to expose the posterior fossa dura. With the skull base cleared of soft tis-
sue attachments (as performed during the high cervical exposure), drilling is continued to reveal the tumor, as well as the jugular bulb. This involves removal of all bone between the posterior end of the digastric groove and the jugular foramen. Attention is then turned to the high cervical carotid artery within the carotid space and the petrous segment of the internal carotid artery within the temporal bone. The bony tympanum, hypotympanum and roof of the glenoid fossa may be drilled away. The mandibular condyle may be displaced anterior-inferiorly or resected, depending on the need for exposure. In our experience, less postoperative pain with little restriction of range of motion of the jaw joint is more common with resection of the mandibular condyle compared with disarticulation of the jaw joint. Drilling continues, beginning at the entrance of the internal carotid artery into the carotid canal. The artery is followed superiorly to the genu adjacent to the eustachian tube and cochlea. If the horizontal segment needs to be exposed, the eustachian tube is divided, inverted, and obliterated with muscle graft, bone wax, and fibrin glue.

◆ Tumor Removal

The jugular vein inferior to intraluminal tumor extension is ligated with a silk suture and sharply divided (Fig. 13.12). This maneuver is necessary to prevent embolization of tumor present in the jugular vein distally into the superior vena cava as the tumor is manipulated in its removal. The sigmoid sinus is closed with a silk suture ligature distal to the entry of the superior petrosal sinus into the junction of the transverse-sigmoid sinus (Fig. 13.13). Closure of the sigmoid sinus in this manner enables venous drainage from the superior petrosal sinus in a retrograde fashion. The lateral wall of the sigmoid sinus is resected and mobilized toward the jugular bulb. Working from above and below the bulb, the tumor is removed in a piecemeal fashion. The lower cranial nerves passing through the jugular foramen are protected by the medial wall of the jugular bulb, which must be preserved for the cranial nerves to be protected. Diffuse bleeding can occur within the jugular bulb from the inferior petrosal sinus, which is often represented by multiple small venous channels along the medial wall of the jugular bulb. Gentle packing with Surgicel or soft bone wax controls this venous bleeding. Care must be taken to avoid using bipolar cautery or excessive compressive with cotonoid pledges to control bleeding along the medial wall of the jugular bulb. Failure to do so may result in a traumatic injury of the lower cranial nerves. During the final steps of tumor dissection, one must directly

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Fig. 13.12  The facial nerve has been transposed anteriorly. Ligation of the distal jugular vein is performed inferior to the level of tumor extension.

Fig. 13.13  Following ligation of the jugular vein, a 2–0 silk suture is passed circumferential to the sigmoid sinus (distal to the superior petrosal sinus).
visualize the carotid artery and the lower cranial nerves to avoid injury (Fig. 13.14).

Most tumors invading the posterior fossa do so in an extradural fashion with displacement of the brainstem and cerebellum. Taking care to maintain this dural layer can help reduce the risk of postoperative spinal fluid leakage. Only with very large tumors will there be absence of a layer of dura between the tumor and the contents of the posterior fossa.

◆ Wound Closure (Fig. 13.15)

If a dural defect has been created by removal of the tumor, this may be repaired primarily with the use of a fascial graft in a watertight fashion. A fat graft harvested from the anterior abdominal wall is placed into the dead space created by the temporal bone dissection. A vascularized musculoperiosteal flap is then mobilized inferiorly over the fat graft and tacked down to the soft tissue edges. Incomplete closure of the dural defect requires the use of a lumbar spinal fluid drain postoperatively to reduce the incidence of spinal fluid leakage.

Modified Lateral Skull Base Approach (Fig. 13.16)

This approach is used for patients with small tumors localized to the region of the jugular bulb. Often with this select group of patients has functional preoperative hearing, and it is necessary to preserve middle ear function.

The initial phase of the operation involves the high cervical dissection as described earlier. The temporal bone dissection involves a simple mastoidectomy but limits the bone removal to preserve the posterior bony ear canal wall, tympanic membrane, and the ossicles of the middle ear space. The mastoid segment of the facial nerve rarely would require translocation for tumor removal. Tumor that is present in the middle ear may be removed through the facial recess alone.

Preauricular Infratemporal Fossa Approach

The preauricular infratemporal fossa approach is used for larger tumors with extensive involvement of the petrous internal carotid artery. Further dissection is performed anterior to the ear canal to expose the infratemporal fossa. Disarticulation of the jaw joint is necessary. Bone removal is similar to that in the combined lateral skull base approach, but continued work is focused anteriorly to expose the petrous internal carotid artery to the level of the cavernous sinus.

◆ Management of Major Complications

Cranial Nerve Palsy

Growth of a tumor involving the jugular bulb may result in direct compression or invasion of cranial nerves in the region.
of the jugular foramen. Most commonly affected are the lower cranial nerves and, less frequently, the facial nerve. Lack of preoperative cranial nerve dysfunction may not correlate with the finding at surgery of tumor invasion of a particular cranial nerve. An acute loss of lower cranial nerve function may be devastating if not properly managed, whereas those patients with chronic loss of function can usually compensate for the deficit. The aggressive management of postoperative cranial neuropathies is absolutely critical to a successful surgical outcome.

Injury to the lower cranial nerves is one of the primary risks when operating on tumors of the jugular bulb region. Acute loss of a single lower cranial nerve is usually tolerated, but loss of multiple lower cranial nerves or the vagus nerve may cause a sudden incoordination of swallowing and lead to life-threatening aspiration pneumonia. An acute glossopharyngeal and vagal cranial nerve palsy should be aggressively treated with early placement of a gastrostomy tube. Immediately following the operation, the patient may not manifest symptoms of aspiration secondary to swelling of the vocal cords from the endotracheal tube. However, as the swelling resolves, the patient is at increased risk for aspiration. If the nerve is expected to recover, injection of the vocal cord with Gelfoam will suffice to temporarily protect the patient. If it is not expected to recover, permanent medialization of the vocal cord is recommended. Most patients with preservation of the anatomic continuity of the cranial nerves compensate for swallowing dysfunction and improved phonation over a 3-month period, at which time the gastrostomy tube can be removed. There is a rare patient who presents with regurgitation through the nose secondary to paralysis of the palate, and a palatoplasty effectively corrects this. Persistent dysphagia resulting in recurrent aspiration despite aggressive therapy may require a cricopharyngeal myotomy.

Protection of the eye in a patient with facial cranial nerve palsy is paramount. In the acute postoperative period, a protective eye patch and lubrication is necessary. If the patient’s facial palsy is anticipated to persist beyond 2 months, augmentation of eyelid closure with implantation of a gold weight is recommended. Recovery of facial nerve function postoperatively is almost certain if the anatomic continuity of the facial nerve is preserved during the operative procedure, and the facial nerve responds favorably to electrical stimulation. In patients in whom the continuity of the facial nerve could not be preserved or repaired primarily, a facial-to-hypoglossal anastomosis is performed during the early postoperative period as the patient’s condition permits.

Cerebrospinal Fluid Leak/Wound Healing

Cerebrospinal fluid (CSF) leakage following resection of tumors involving the jugular foramen is related to several factors: the size of the tumor and involvement of the dura, the nutritional status of the patient, and the state of cerebrospinal fluid dynamics.

Careful attention is paid to achieving a watertight closure of the dural with either a primary repair or with a fascial graft. The dural defect is usually present with resection of a large tumor at the level of the jugular foramen. This dural defect is repaired primarily with a fascial graft if possible, placement of a fat graft to obliterate the dead space of the surgical wound, and rotation of a vascularized graft over the adipose tissue of the surgical defect followed by a multilayered tissue closure. A postoperative lumbar spinal drain is advised if dural closure is not achieved. The prevention of spinal fluid rhinorrhea and otorrhea is accomplished by closure of the eustachian tube and external auditory canal during the surgical procedure.

Early nutritional supplementation is mandatory to promote tissue healing. If the patient is not able to tolerate feedings by mouth, a small-diameter feeding tube is inserted within the first 24 hours. Alimentation is immediately begun. If oral feedings are not possible beyond 10 to 14 days postoperatively, a percutaneous endoscopic gastrostomy (PEG) is recommended.

Altered CSF dynamics may lead to postoperative hydrocephalus and result in CSF leakage. The altered spinal fluid dynamics may result from contamination of the wound by blood products, bone dust, or from occlusion of the venous circulation at the level of the jugular bulb. The use of continuous lumbar spinal fluid drainage for 3 to 7 days post-
operatively aids in the normalization of intracranial pressure, clearing of the spinal fluid circulation, and prevention of the accumulation of fluid under the wound flap. This promotes sealing of the dural defect and healing of the wound. We have found that the impairment of CSF absorption is usually transient, thus obviating the need for permanent cerebrospinal fluid diversion.

**Vascular Injury**

In larger tumors that encase the internal carotid artery, there is a risk of injury to the artery. Every patient that demonstrates involvement of the carotid artery undergoes temporary balloon test occlusion to determine the adequacy of collateral circulation. Surgical removal of tumor proceeds with the intent of always preserving the internal carotid artery. If injury to the carotid artery should occur, temporary occlusion of the artery followed by a primary repair is attempted using 9–0 Prolene vascular suture. If this cannot be performed, the results of the balloon occlusion test dictate whether the artery is sacrificed or an arterial bypass is utilized. A saphenous vein interposition graft is recommended if technically possible. A superficial temporal artery–middle cerebral artery bypass may not be an option because of preoperative embolization of the external carotid artery circulation.

**Conclusion**

Although appearing histologically benign, the majority of tumors involving the jugular foramen are characterized by unpredictable biologic behavior. Tumors originating in the jugular bulb region present a significant surgical challenge. Successful treatment of jugular bulb tumors mandates a clear understanding of the anatomy of the jugular foramen and the natural history of the pathology of the tumor. With the significant advances in surgical approaches to the lateral skull base, surgeons can now accomplish complete extirpation of the majority of jugular foramen tumors with low morbidity rates and good long-term outcomes.

**References**

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The Retromastoid Approach: Techniques and Results

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Since its development at the turn of the 20th century, the retromastoid-retrosigmoid suboccipital approach remains, after several modifications, an exquisite route for neurosurgical access to different pathologic processes in various locations, such as the cerebellar hemisphere, the petroclival region, the foramen magnum, the jugular foramen, and in particular the cerebellopontine angle (CPA). Numerous pathologic entities, including glossopharyngeal, trigeminal, vagal, and vestibular schwannomas; meningiomas; epidermoids; glomus tumors; trigeminal and glossopharyngeal neuralgia; hemifacial spasm; and vertebrobasilar aneurysms may be approached via this route. The senior author’s (M.S.) personal experience is based on microsurgery of various CPA lesions since 1968. With the growing knowledge of CPA and internal auditory canal (IAC) microanatomy, the more recent surgical experience of the senior author with over 3000 vestibular schwannomas, over 1000 posterior fossa meningiomas, over 100 CPA epidermoids, and over 1000 vascular decompressions in the CPA since 1978 has shown that, regardless of the lesion size, the lateral suboccipital retrosigmoid approach offers the best possible exposure for identifying cranial nerves from their exit zone at the brainstem up to their bony entry points in the skull base. This approach, therefore, optimizes the preservation of these structures.

General Principles of the Surgical Technique

The procedure may be performed with the patient in a semi-sitting (lounging) position for the resection of posterior fossa tumors or in the supine position (with the patient’s head turned 60 degrees contralaterally) for vascular decompression. For the semi-sitting position, the back of the operating table is elevated to 30 degrees. The head is turned 30 degrees to the side of the craniotomy and placed in a three-point head-fixation device, and then flexed slightly. The head and neck are in a natural anatomic position, without unnecessary strain or compromise of cervical venous drainage. The patient is padded appropriately. Arterial blood pressure is monitored, a central venous catheter is placed in the right atrium, and a precordial Doppler is used to monitor signs of air embolism. In the semi-sitting position, the risk of air embolism is decreased by elevating the legs to the level of the right atrium of the heart.

Bone window computed tomography (CT), in addition to magnetic resonance imaging (MRI), reveals individual anatomic variations of the posterior fossa, such as emissary veins, location of the jugular bulb, as well as the relationship of the tumor to the IAC and other bony structures (Figs. 14.1 and 14.2). X-ray studies of cervical spine are also obtained routinely to detect possible spinal abnormalities and prevent medullary compression. Sensory evoked potentials (SEPs) are also obtained routinely. The mastoid eminence, digastric groove, and inion should be palpated and identified. A curvilinear skin incision is then placed behind the ear, approximately 1.5 to 2 cm medial to the mastoid process. The excision reaches from just above the superior nuchal line to the level of C1. The trapezius and splenius capitis muscles are detached from the superior nuchal line. The occipital artery and the greater occipital nerve are preserved. Care should be taken to avoid injury to the vertebral artery. A suboccipital craniectomy is then executed. Important landmarks are the external occipital protuberance that overlies the confluence sinuum, and the superior nuchal line overlying the transverse sinus. A single bur hole is placed below the Frankfurt horizontal line and 3 cm behind the external auditory canal. The dura is separated and the bone removed with rongeurs to enlarge the craniectomy to 3 to 4 cm square, exposing the transverse sinus and its junction with the sigmoid sinus.
Fig. 14.1 (A) Preoperative axial 1.5-mm bone window computed tomography (CT) scan shows widening of the left internal auditory canal (IAC) caused by a medium-sized (T3a) vestibular schwannoma with intra- and extrameatal extension seen on the magnetic resonance imaging (MRI) scan (B).

Fig. 14.2 (A) Preoperative T1-weighted axial MRI shows a primarily intracanalicular vestibular schwannoma with some extrameatal extension (T2 tumor). (B,C) The axial 1.5-mm bone window CT scans show a highly positioned jugular bulb (JB) and extensive widening and bone destruction of the right IAC by the tumor. (D,E) Intraoperatively, drilling of the posterior wall of the IAC was performed and the jugular bulb (JB) skeletonized and pushed downward so that the tumor (Tu) could be exposed. The tumor was then completely removed. (F) The postoperative CT scan demonstrates the reconstruction of the bone defect after right suboccipital retromastoid craniectomy and the extent of the drilling of the posterior lip of the IAC.
The Retromastoid Approach: Techniques and Results

Tumors

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sinus. Emissary veins are exposed carefully with a diamond drill and subsequently coagulated to prevent air embolism. The dura is usually opened along the sigmoid and transverse sinuses, and is covered with an overlying cottonoid on top of the cerebellar hemisphere. To provide extra space, avoid compression of the cerebellum, and improve exposure, the lateral cerebellomedullary cistern is opened and cerebrospinal fluid is withdrawn. Following tumor removal, hemostasis is carefully attained. The mastoid air cells are sealed with autologous muscle or fat and fibrin glue. The dura is closed in a watertight manner and the wound is closed in multiple layers (Fig. 14.3).

◆ Indications, Clinical Features, Special Techniques, and Outcomes

It is beyond the scope of this chapter to discuss all the aforementioned pathologic entities that apply to the CPA. We thus focus our attention on the most common CPA lesions. Vestibular schwannomas account for the vast majority of these lesions, followed by meningiomas and epidermoids. Modifications of this approach for the resection of a variety of lesions are also presented.

◆ Vestibular Schwannomas

With a prevalence of 80 to 90%, vestibular schwannomas represent the most common lesions in the CPA. Throughout the history of vestibular schwannoma management the surgical goals remain the same: completeness of resection, preservation of the facial nerve, and, if possible, preservation of cochlear nerve function, which has an impact on the patient’s quality of life. Based on experience with vestibular schwannoma surgery since 1968, and having tested the various approaches and their combinations, the senior author has found the lateral suboccipital retromastoid approach to be the best approach for the achievement of these goals, regardless of tumor size. He has operated successfully on more than 3000 patients with vestibular schwannomas since 1978 via this route.

Vestibular schwannomas arise from the intracanalicular portion of the vestibular nerve, with initial growth within the IAC and extension into the CPA. This is the basis for the Hanover Tumor Extension System, which classifies tumors as follows: class T1, intrameatal tumors; class T2, intra- and extrameatal tumors; class T3a, tumors that fill the cerebello-pontine cistern; class T3b, tumors that reach the brainstem; class T4a, tumors that compress the brainstem; and class T4b,
tumors that severely dislocate the brainstem and compress and occlude the fourth ventricle. Of special anatomic interest in vestibular schwannoma surgery is the anterior inferior cerebellar artery. Disturbance of this artery, which gives rise to the internal auditory (labyrinthine) artery, can result in infarction of the lateral brainstem with catastrophic consequences.

Beginning with the opening of the dura, the entire operative procedure is performed under the operating microscope. The lower cranial nerves are identified and protected with a wet cottonoid. The cerebellum is also covered with a moist cottonoid and a brain retractor placed to offer support and provide exposure. The portion of the tumor within the CPA is now visualized (Fig. 14.4A). In cases of a small intracanalicular vestibular schwannoma (T1), however, no tumor may be seen in the CPA. In medium-sized tumors (T3), the facial and vestibulocochlear nerves are in general identified easily medially at the brainstem. In larger vestibular schwannomas (T4), the tumor has to be debulked before attempting to identify the cranial nerves. The IAC is opened as a first step to establish the lateral tumor extension in all tumors (T1 to T4). This is begun with the circular excision of dura from the petrous bone posterior to the IAC followed by drilling of the posterior wall of the meatus with a high-speed diamond bur under continuous irrigation with saline solution. Initial drilling is performed with a 4- to 5-mm bur; as the fundus is approached progressively, smaller diamond burs are employed.

It is important to know the position of the jugular bulb from a review of the preoperative CT scans before drilling the porus (Fig. 14.2B,C). In cases of a highly positioned jugular bulb, drilling is done cautiously to avoid entry into the bulb. The jugular bulb is skeletonized by gentle drilling of the posterior wall of the IAC (Fig. 14.2D). After this, the bulb can be pushed downward and the remaining bone of the posterior wall of the IAC can be drilled away and the intracanalicular tumor portion exposed (Fig. 14.2E,F). Care must be taken to avoid opening or injuring the semicircular canal and vestibule, especially if the tumor extends far laterally. The remaining distance to the fundus should be assessed continuously by using an angulated microinstrument. If the semicircular canals should become fenestrated, suctioning of the perilymph and endolymph must be avoided, and the fenestration closed with fascia and sealed with fibrin glue immediately.

The drilling should proceed in a medial-to-lateral direction as the fundus is approached. In this way the bone plate between the IAC, the ampulla of the semicircular canal, and the jugular bulb can be thinned in 1/10-mm increments with the diamond bur without causing injury to the peripheral vestibular apparatus or the jugular bulb. Drilling should leave at least 2 mm of bony distance from the fundus of the IAC to avoid entering the labyrinth. The depth should be palpated with a microneurve hook. Once the porus has been drilled, the decision regarding the procedure for tumor removal depends on tumor size. With small, primarily intracanalicular tumors (T1), the next step is to identify the facial and cochlear nerves medially near the brainstem. The tumor is dissected gently from the cranial nerves, with the surgeon grasping the tumor with the right hand and using the microforceps with the left hand to push the arachnoid away. This maneuver progressively separates the tumor capsule from the cranial nerves. Permanent irrigation of the surgical field should be performed at this moment to optimize the view. The critical final step is removal of the tumor at its last area of adherence close to the porus. This is the point of greatest concern because traction or other injury to the cranial nerves may occur.

When dissecting the tumor capsule from the cranial nerves, it is helpful to dissect from all sides toward the point at which the capsule is most firmly attached to the adjacent cranial nerves. For larger tumors (Figs. 14.4, 14.5, and 14.6) partial debulking of the tumor may be necessary before beginning...
further dissection. Thus, a part of the tumor capsule that can be entered without traction or potential injury to the cranial nerves is identified and an intracapsular piecemeal enucleation is then performed. It is important to exercise great care not to penetrate the anterior wall of the tumor capsule. It is also extremely important not to cause traction pressure on the nerves.

As the intracapsular decompression progresses, the tumor capsule becomes lax. It is then possible to identify the lateral extent of the tumor and also the points at which the tumor is especially adherent to the facial and cochlear nerves and the brainstem. The tumor capsule may then be gently dissected from these structures. Again, it is important to stay within the arachnoid plane, grasping the tumor capsule with microforceps held in the surgeon's right hand, pushing the arachnoid away and separating it from the tumor capsule with forceps held in the left hand under permanent irrigation (Fig. 14.4B–D).

Finally, as the tumor mass becomes reduced in size, the intracanalicular portion of the tumor is carefully dissected from the facial and cochlear nerves within the IAC, and the last remnants of the tumor are removed. Use of bipolar cautery is not needed during tumor removal and the operative field is irrigated continuously with saline solution to maintain good visualization. Monitoring of auditory evoked potentials and direct brainstem recording of auditory evoked potentials should be performed routinely. In case of any changes in the wave amplitude, the surgeon has to stop the procedure and change the strategy of removal.

Key features for achieving complete tumor removal and preservation of the facial and cochlear nerves are careful opening of the IAC, primary debulking of the tumor mass, identification of the cranial nerves at the brainstem and at the fundus of the IAC, and constant respect for the arachnoid plane. A small autologous muscle or fat graft has to be applied to the opened portion of the IAC to prevent internal cerebrospinal fluid fistula. It should be emphasized that the suboccipital route offers the best opportunity for preservation as well as reconstruction of the facial nerve. Reconstructive procedures may be performed in the same operation preventing further surgeries (Fig. 14.6). Considering the results of the first 1000 surgeries of the senior author that were presented in 1997,4,5 anatomic preservation of the facial nerve was achieved in 93% and of the cochlear nerve in 68% of the cases. Functional preservation of the cochlear nerve was achieved in 39% and complete removal in 979 of the 1000 patients. In a current evaluation of the most recent 200 cases, the rate of facial nerve anatomic preservation has currently improved to 98.4% in the hands of the senior author. In smaller tumors (T1 to T3 tumors), the rate of facial nerve preservation has been found to be as high as 100% recently.

Some authors state that hearing preservation surgery should be undertaken only in selected cases, depending on tumor size and preoperative hearing level. Our philosophy,
however, is to attempt hearing preservation in all cases. Our results regarding hearing preservation consider the total patient population, regardless of tumor size, and not just a group of selected cases. In 2002 our group analyzed hearing function in 1800 cases with vestibular schwannomas. Although the overall preservation rate was 40%, there were considerable differences depending on the preoperative hearing quality and tumor extension. The best results were achieved in small intrameatal tumors (T1, 56% preservation rate) and small intra-extrameatal tumors (T2, 57%). Preservation rate was 44% in medium-sized tumors (T3) and 20% for T4 tumors. Considering cases with normal or good preoperative hearing specifically, on average a 54% preservation rate was accomplished (71% in T1, 69% in T2, 58% in T3, and 29% in T4). Evaluating the most recent 200 cases, we found that the senior author now achieves increased rates of anatomic cochlear nerve preservation—as high as 84%—and functional cochlear nerve preservation is now achieved in 51% of the cases. Thus, the most important factors predicting hearing preservation include tumor size and extension, the preoperative hearing level, and the surgeon’s skills.

**Meningiomas**

With a prevalence of 10 to 15%, CPA meningiomas represent the second most common CPA entity. Meningiomas may develop anteriorly or posteriorly to the IAC and may extend rostrally or caudally. Due to this variable growth direction, the relation to the neurovascular structures is also highly variable. In the experience of the senior author with the removal of over 1000 meningiomas since 1968 in this region, including CPA, craniocervical, tentorial and petroclival meningiomas, most of these tumors, even with extension into the middle cranial fossa, may be removed through a simple retrosigmoid approach or through modifications of this approach. It should be emphasized that as for most nonacoustic lesions, even profound preoperative hearing deficiency may recover almost completely following decompression and removal of the meningioma.

Using these approaches to the CPA and the petroclival region, we distinguish different surgical “floors,” which are determined by the location of the cranial nerves within the CPA. They create the surgical opening to the tumor. One such
available opening is between the caudal cranial nerves and the seventh and eighth cranial nerves. Another opening is between the facial, vestibulocochlear, and trigeminal nerves. For the supratentorial portion of the tumor, the opening is between the trigeminal and the trochlear nerves and the tentorial edges. To avoid mechanical or thermal injury to these structures, moist cottonoids for protection should be used. Great attention must be paid to prevent injury to the sixth cranial nerve, which is not in the field of vision until late in the procedure. Therefore, coagulation of tumor tissue in the vicinity of the abducens nerve must be avoided. Once the tumor is reduced in size and the brainstem is decompressed, the abducens nerve should be identified at the brainstem and followed to its entry point into Dorello’s canal. The foremost dissection technique consists of piecemeal tumor removal with the use of bipolar cautery and the Cavitron ultrasonic surgical aspirator (CUSA). If the tumor fills the entire CPA from the trigeminal down to the caudal cranial nerves, the lateral tumor parts should be removed first, exposing the caudal cranial nerves followed by the facial and vestibulocochlear nerves. Care must be taken not to damage vascular structures that may be encased in the tumor. When removing the area of tumor attachment on the bony skull base, the surgeon should first coagulate the residual tumor bipolar and then circumscribe it with a knife to separate it from the bone. It is not unusual for this area to have a rich blood supply. The diamond bur is indeed very useful because it provides hemostasis as well as complete removal of tumor matrix.

In a series of 134 CPA meningiomas we used the simple lateral suboccipital route in 90% of the cases. A combined presigmoidal or combined suboccipital and subtemporal approach was used in 10% of the cases. Overall, 95% complete tumor removal was accomplished. Hearing preservation rate was 82% and hearing improvement in 6% of the cases in this study. Facial nerve paresis or paralysis was encountered in 17% of the cases and facial nerve reconstruction was necessary in 7%. CPA meningiomas may originate from or extend into the IAC (Fig. 14.7), sometimes reaching the fundus. When this occurs complete opening of the canal is necessary to achieve total tumor removal. In a study involving 421 patients with CPA meningiomas, we have recently shown that in all patients with IAC involvement the retrosigmoid approach may be performed, regardless of whether the major portion of the meningioma resides within the CPA. Total tumor removal was achieved in 86.1% of the cases. Opening of the IAC using a diamond drill does not influence cranial nerve outcome. Depending on the origin of the meningioma and IAC involvement, facial and cochlear nerve preservation rates of 75 to 94% were achieved. In all cases where the IAC is drilled, a small autologous muscle or fat graft has to be

![Fig. 14.7](image-url) Axial and coronal MRI scans of a large CPA meningioma with growth into the right IAC (A). The tumor was completely removed through a simple right retrosigmoid suboccipital approach with preservation of all cranial nerves.
applied. Tentorial meningiomas with supra- and/or infratentorial extensions may also be removed through a lateral retrosigmoid approach. The first steps of the approach to tentorial meningiomas are performed in the same manner as described above. After opening of the cerebellomedullary cistern and cerebrospinal fluid drainage, the cerebellum is free to be shifted downward, exposing the supracerebellar space. The entire length of the involved tentorium is exposed in this way. Without brain compression, the tentorium may be resected in a safe area and the tumor portion that extends into the supratentorial area can be safely removed.

◆ Modifications of the Retromastoid Approach

Different tumors may be encountered in the petroclival region, the most common being meningiomas affecting Meckel’s cave and schwannomas of the trigeminal nerve. Other lesions include epidermoid cysts, chondrosarcomas, and chordomas. Petroclival tumors usually extend into both the middle and posterior fossae. The choice of surgical approach depends largely on the type of tumor extension. In the past, many of these tumors were thought to be inoperable; however, in the last two decades, major efforts have been made to improve surgical results. Different approaches employing varying degrees of petrosectomy and tentorium division have been introduced for a better exposure and resection of tumors in the petroclival region with extension into multiple cranial fossae.9–12

In 1988 the senior author described the presigmoid approach to the petroclival region.2,9,12 This approach includes a classic suboccipital craniotomy combined with a temporal craniotomy and extensive drilling of the mastoid and petrous bone.2,9,12 The decision to use a presigmoid approach depends on involvement of the supratentorial region through the tentorial notch into the middle fossa and posterior portion of the cavernous sinus. The complete transection of the tentorium extends the view to the middle fossa, Meckel’s cave, and posterior aspect of the cavernous sinus. Various lesions including petroclival meningiomas and extended glomus tumors and jugular foramen schwannomas can be approached via this route. With extensive experience in managing petroclival lesions, however, the senior author also learned that several of these tumors could be safely removed by the simple retrosigmoid approach without major exposure of petrous bone structures. Avoidance of large petrous bone resections was found to reduce operative risks, such as hearing loss, as well as operative complications, such as cerebrospinal fluid leaks and particularly venous complications associated with retraction of the vein of Labbé and coagulation of basal veins. In 1982, therefore, the senior author introduced a modification of the classic retrosigmoid approach, the retrosigmoid intradural suprameatal approach (RISA) to treat certain petroclival lesions.1,2 This approach includes the classic retromastoid, retrosigmoid craniotomy, and intradural drilling of the bone located above and anterior to the internal auditory meatus.

Petroclival meningiomas are usually located ventral to the cranial nerve VII-VIII complex, displacing these nerves posteriorly and caudally. The tumor may fill the entire CPA and engulf the neurovascular structures, and thus tumor removal is performed in the lateromedial direction, starting from the bone and moving toward the brainstem. Tumor invasion into the cranial nerve foramina may make visualization of the cranial nerves difficult. Therefore, piecemeal tumor resection should be performed instead of en bloc tumor removal. Tumor dissection should be performed while respecting the arachnoid planes. If the pia mater of the brainstem and neurovascular structures is infiltrated, complete tumor removal should not be performed at the price of catastrophic consequences. Once the part of the tumor located in the CPA is removed, drilling involves the suprameatal area and the posterior suprameatal portion of the petrous apex, which is located over the cranial nerve VII-VIII complex and dorsolateral to the trigeminal nerve. With this, Meckel’s cave is exposed and opened. This facilitates increased exposure and mobilization of the trigeminal nerve and access to the middle cranial fossa. The combination of this step with incision and division of the tentorium increases the corridor to the middle fossa. Due to the variation in bone anatomy in this region, the amount of bone resection may vary. The enlarged approach permits visualization of the oculomotor nerves, the posterior clinoid, the internal carotid arteries, and even the optic nerves. Thus, this approach delivers a useful modification of the retromastoid approach for the resection of large petroclival tumors, without need for supratentorial craniotomies.

A standard suboccipital approach combined with the opening of the foramen magnum and laminectomy of the involved cervical segments may be sufficient for the majority of cases with cranio cervical meningiomas or other lesions that involve the foramen magnum, lower clivus, jugular, and hypoglossal foramen.

◆ Epidermoid Cysts

Epidermoid tumors are dysontogenic lesions and represent 0.2 to 1.8% of intracranial tumors. Their most common location in the central nervous system is the CPA. Epidermoids represent 4.6 to 6.3% of all CPA lesions. The clinical presentation and symptoms of CPA epidermoids depend on the close adhesions to and compression of neurovascular structures. Facial nerve involvement and unilateral hearing loss are the most common presenting symptoms. However, facial nerve symptoms occur earlier than in vestibular schwannomas. Some patients may present with a classic trigeminal neuralgia or a constant neuralgic pain.

Even large tumors that extend beyond the CPA can be managed with the retrosigmoid approach. Due to the fact that hearing may improve or recur, a translabyrinthine approach should not be chosen in a patient with a CPA epidermoid whenever possible. However, the approach must be individually tailored to the location and extent of the lesion to ensure optimal exposure. Bilateral lesions that involve the cranial nerves on both sides constitute a surgical challenge.
Resection of these masses in a stepwise manner with significant time between the two procedures to allow recovery of possible deficits, especially of the lower cranial nerves, is emphasized. Thus, whereas epidermoids confined unilaterally to the CPA can be best exposed through a lateral suboccipital approach, in the case of significant supratentorial or bilateral growth it may be necessary to proceed to staged surgeries.

The lesion presents as a soft, pearly globular mass with irregular borders (Fig. 14.9). The capsule is thin and adherent to the arachnoid at numerous sites. The strategy is to remove the tumor in one region completely before moving to the next region. In this way small tumor remnants are not left behind, and thus the risk of regrowth is lowered. An effective technique is to open the capsule and debulk the tumor first. Then the matrix can be separated from the neurovascular structures under optimal vision. As a final step, all the corners of the CPA should be inspected, and the surgical field should be irrigated to ensure removal of remnants. In the experience of the senior author with the surgical resection of over 100 CPA epidermoids through the suboccipital retromastoid approach, total removal can be achieved safely in over 75% of cases. Postoperative deficits are mainly related to the degree of adhesion of the tumor to the neurovascular structures. Consequently, it is advisable to leave small parts of the capsule rather than risk a surgical catastrophe.
References

Skull base surgeons owe a great debt to Dr. William House, who, in the face of significant opposition, popularized the translabyrinthine approach nearly 50 years ago. The term *translabyrinthine* is something of a misnomer, implying as it does a narrow approach via only the posterior and superior parts of the otic capsule. A more accurate if somewhat ponderous description would be transoccipital, transtemporal, transmastoid, and translabyrinthine, emphasizing the need to remove a significant portion of the temporal and occipital squama. This point is well demonstrated by the size of the bony defect evident in the postoperative computed tomography (CT) scan shown in Fig. 15.1. Provided that these steps are taken, the authors believe that no tumor is too large to be successfully dealt with by this approach.

Although there has been concern in the past that this approach provides inadequate access to the cerebellopontine angle, it has been shown to compare favorably with the retrosigmoid approach in this regard. In addition, it gives a very clear view of the facial nerve in the lateral end of the internal auditory canal. Lastly, brain retraction is extradural.

The only significant drawback is that there is no prospect of saving hearing, although the issue of hearing preservation surgery remains contentious. Very occasionally an anatomic aberration such as a very high or very large dominant jugular bulb or extreme forward position of the sigmoid sinus will make the approach technically difficult. However, this is a rare event if sufficient bone is removed from behind the sigmoid sinus and from around the bulb itself.

With experience, this approach can be performed very rapidly. In the authors’ hands the time taken from the skin incision until the dura is opened is less than 90 minutes.

◆ **Patient Selection**

In the authors’ unit, the translabyrinthine approach is now the choice for acoustic neuroma surgery, other than in patients with small tumors and good hearing, when formerly the middle fossa route was chosen. In recent years, tumors smaller than 15 mm in the cerebellopontine angle have been managed by the so-called wait-and-scan technique. The retrosigmoid approach is reserved for meningiomas and other cerebellopontine angle tumors where hearing preservation is more predictable and where visualization of the lateral end of the internal auditory canal is not required.

◆ **Positioning and Preparation**

After induction of anesthesia, the area of the incision is shaved and the incision marked and infiltrated generously with a local anesthetic and vasoconstrictor mixture. The patient is positioned supine on the operating table with the head turned away from the side of surgery and the face supported by a soft pad. The use of a head ring is avoided as it flexes the neck, closing the angle between the ear and shoulder and hampering access, as well as limiting head rotation. Rigid head fixation is not required. The patient is strapped to the operating table, as this allows the table to be rolled toward or away from the surgeon as the operation proceeds, changing the angle of the head accurately and predictably, and without disturbing the drapes. Facial nerve monitoring is mandatory, and both passive electromyogram (EMG) monitoring and an active electrode are required. The anesthetist must be made aware of the use of the monitor, as muscle relaxants will abolish EMG potentials. Short-acting relaxants, however, may be used for induction of anesthesia.

Regarding draping, an adhesive clear-plastic drape allows the pinna to be held forward and out of the operative field. A plastic pouch protects the surgeon’s lap from irrigation fluid. The surgeon sits and is positioned as for tympanomastoid surgery, with the microscope base at the head of the table and the scrub nurse on the opposite side of the patient from the surgeon (Fig. 15.2).
◆ Incision and Exposure

The incision is shown in Fig. 15.3. It is important to make the incision sufficiently posteriorly to allow bone removal behind the sigmoid sinus and sufficiently superiorly to be able to expose at least 3 to 4 cm of bone above the temporal line. Anteriorly it is carried to a point in line with the tragus, as this provides access to the posterior root of the zygoma, deep to which lies the anterior part of the epitympanum. Below the temporal line the incision continues straight down to the periosteum; above it the plane of dissection is immediately superficial to the temporalis fascia. This is facilitated by an incision down to bone along the temporal line. A large piece of temporalis fascia, for dural closure, is harvested and a 3-cm-long vertical incision is made through the posteroinferior part of the temporalis muscle, continuous with the previous incision along the temporal line. The muscle is then mobilized in an anterosuperior direction so that the posterior root of the zygoma and the temporal squama are exposed (Fig. 15.4). It is important to minimize mobilization of the membranous ear canal to reduce the chance of subsequent stenosis and recurrent otitis externa. The soft tissues may be held with either self-retaining retractors or hooks connected via elastic bands to towel clips attached to the mattress of the operating table.

◆ Mastoidectomy

A very wide intact canal wall mastoidectomy is performed. Cortical bone removal is continued both 2 cm (and occasionally 4 cm) behind the sigmoid sinus (the occipital squama) and on to the squamous temporal bone to expose a crescentic area of vertical middle fossa dura approximately 3 to 4 cm in maximum height (Fig. 15.5). This is best achieved with a large (8 or 9 mm) bur, which will reduce both the time taken and the chance of injury to the underlying dura. Safety is further enhanced by changing from a cutting to a diamond bur as the dura is approached. It is particularly important to saucerize the edges of the mastoidectomy and not to leave vertical edges or overhangs that will hamper access later in the procedure.
Bone is then removed from over the sigmoid sinus, the posterior fossa plate, and the middle fossa plate. These latter areas join over the superior petrosal sinus (the sinodural angle). Bone is removed from the angle, and from above and behind the point at which the sigmoid, transverse, and superior petrosal sinuses meet. This maneuver produces a “sac of dura,” which facilitates extradural retraction of the temporal lobe and cerebellum later in the procedure.

Any bleeding at this stage comes either from mastoid emissary veins or, rarely, a torn venous sinus. The former is readily controlled with bone wax. The latter can be stopped by gentle pressure over a large piece of a collagen-based absorbable hemostat supported by a neurosurgical cottonoid.

The mastoidectomy is continued under the operating microscope. The mastoid antrum is entered and the horizontal semicircular canal identified. The superior aspect of the body of the incus is exposed and the epitympanum opened anteriorly so that the head of the malleus is clearly visible. It is important at this stage to remember that the middle fossa dura curves sharply downward over the anterior part of the epitympanum, exposing it to a drill injury in this area.

An important step is positive identification of the facial nerve. It is found in its descending portion, well inferior to the external genu. In the region of the horizontal semicircular canal the nerve is changing direction and therefore at risk of injury. Inferiorly its straighter course means that the nerve can be identified here quite safely. A cutting bur is used in the manner of a paintbrush, carefully brushing off layers of bone along the expected course of the nerve until the fine blood vessels of the epineurium or a more diffuse “blush” can be seen through the bone. In this way a layer of bone can be left over the nerve. If the position of the sigmoid sinus allows, the nerve should be approached from posteriorly or posterolaterally rather than laterally, as this enables the surgeon to use the equator rather than the pole of the bur, improving visualization. Copious irrigation is essential to prevent thermal damage to the nerve, especially if a diamond bur is used at this stage. Once the nerve has been found, it is followed superiorly in the same fashion until the entire descending portion is clearly on view.

A facial recess opening (posterior tympanotomy) is then created (Fig. 15.6). This is achieved by drilling between the facial nerve and chorda tympani to fashion an oval-shaped opening into the middle ear. The incudostapedial joint is disarticulated and the incus removed. These maneuvers allow the eustachian tube to be packed with muscle under direct vision to eliminate one source of postoperative cerebrospinal fluid (CSF) leakage. The bridge of bone over the fossa incudis is removed and the horizontal segment of the facial nerve can clearly be seen.

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**Fig. 15.4** The temporal squama and the posterior root of the zygoma (RZ) are exposed after soft tissue mobilization.

**Fig. 15.5** The sigmoid sinus (SS) and vertical middle fossa dura (MFD) are exposed. PCW, posterior canal wall.
**Perilabyrinthectomy**

The approach to the internal auditory canal, of which labyrinthectomy is a vital step, is most safely done in a wide field. The authors have coined the term *perilabyrinthectomy* to describe the removal of bone above and below the labyrinth prior to labyrinthectomy.

Inferiorly this involves creating a space bounded by the jugular bulb below, the posterior semicircular canal above, the posterior fossa dura behind and in front, and laterally the facial nerve and more medially the solid bone in which runs the cochlear aqueduct. The bone is removed from over the jugular bulb and from the angle between it and the sigmoid sinus. If the bulb is particularly high and limiting access, this maneuver allows it to be depressed inferiorly and held in position with bone wax.4

Above the labyrinth, the superior semicircular canal is skeletonized by removing bone from the middle fossa plate and from the angle between the superior and lateral semicircular canals, the so-called solid angle of Citelli.

These essential steps result in the labyrinth projecting into space (Fig. 15.7), as it were, allowing for meticulous removal of bone in the exposure of the internal auditory canal.

**Labyrinthectomy**

The labyrinthectomy is commenced by opening the horizontal semicircular canal. Because the external genu and horizontal segments of the facial nerve are clearly on view, they should be safe from injury. The dissection continues anteriorly until the ampulla is reached. Directly superior to this is the ampulla of the superior semicircular canal, which is now opened. This is an extremely important landmark and delineates the anterior limit of the superior part of the dissection, as anterior to the ampulla lies the labyrinthine segment of the facial nerve.

The superior semicircular canal is then opened along its entire length until the crus commune is reached. This leads the surgeon naturally into opening the posterior canal. The upper part of the vertical segment of the facial nerve can be seen lateral to the ampulla of the posterior canal. All three semicircular canals should now be clearly on view and open (Fig. 15.8), as is the vestibule, in which the membranous sacule and utricle can often be seen. During the labyrinthectomy it is usually possible to identify the endolymphatic sac on the posterior fossa dura. This can be a useful teaching exercise, and it facilitates identification of the intraosseous portion of the endolymphatic duct. The rugose portion of the sac is divided as it enters the bone, enabling the posterior fossa dura to fall back, away from any residual bone, which needs to be drilled out.

**Approach to the Internal Auditory Canal**

When the labyrinthectomy has been completed, only two landmarks remain in a mass of bone: the stump of the ampulla of the superior semicircular canal, and the open vestibule. The former is superior to the lateral end of the internal auditory canal (IAC), and the latter is directly lateral to the lateral end of the canal.

The IAC is approached by drilling through this mass of bone on a broad front, aiming initially to identify it posteriorly and inferiorly, furthest away from the facial nerve.
The anterior limit of the inferior dissection is the aforementioned cochlear aqueduct. Beyond it lie the glossopharyngeal, vagus, and accessory nerves in the neural compartment of the jugular foramen. The cochlear aqueduct is not always readily seen, but when encountered usually rewards the surgeon with a flow of CSF. This helps to decompress the posterior fossa, facilitating both drilling and extradural retraction.

As the IAC, full of tumor, is approached, it is seen as a color change in the bone. Once it has been clearly identified, posterior and superior bone removal can proceed with a degree of confidence. The aim is to expose the canal around 270 degrees of its circumference, leaving a thin eggshell of bone overlying it for protection (Fig. 15.9). The bone in the angle between the IAC and the posterior fossa dura, overlying the porus, forms something of a collar that eventually can be lifted free without tearing the underlying dura. It is also important at this stage to ensure that the medial bone removal is continued far enough anteriorly round both the superior and inferior aspects of the IAC.

Once the IAC has been fully skeletonized, the remaining bone is removed using a diamond bur and picks or elevators. It is safer to start medially, at the porus end of the IAC, concentrating again initially on the inferior aspect of the canal, which is furthest away from the facial nerve. Medially and superiorly it is essential to ensure that bone removal is complete, as later dissection of tumor from the facial nerve in this area will be compromised by any residual bone that interferes with visualization of the nerve. Exposure of the IAC continues until the crista falciformis (horizontal crest) is exposed. This is an important landmark for the facial nerve at the lateral end of the IAC, the nerve lying anterosuperior to the crest.

The other landmark at the lateral end of the IAC is Bill’s bar, which is a spur of bone lying in the coronal plane separating the facial nerve in front from the superior vestibular nerve behind. Bill’s bar, however, is much more difficult to identify with certainty than is the horizontal crest.

All bone removal must be completed before the dura is opened to prevent contaminating the CSF with bone dust or chips. The suction should also be changed to a Brackmann-type side fenestrated sucker-irrigator. The posterior fossa dura is initially incised using a scalpel, in line with the IAC, taking care not to injure the underlying cerebellum. The incision is continued anteriorly and laterally above and below the porus acusticus using dural scissors. The tumor should now be on view. Depending on the size of the tumor, the dural opening may need to be enlarged by making cuts at right angles to the original incision.

These authors believe that the dura of the IAC should not be opened until the end of the tumor dissection, nor should positive identification of the facial nerve at the lateral end of the IAC take place until that time. This prevents undue mobilization of (and traction on) the facial nerve while tumor debulking is taking place medially.

◆ Tumor Removal

Tumor removal is a gradual, stepwise process. Unlike surgery of malignant neoplasms, the tumor is removed piecemeal, and extensive intracapsular debulking is the key to the removal. The posterior surface of the tumor is first tested with the facial nerve stimulator to ensure that there are no facial nerve fibers stretched across this surface, and that the rare diagnosis of a facial nerve neuroma has not been missed. An opening is then made in the tumor after cautery with the bipolar diathermy. The debulking is then started though this opening, using a combination of cupped tumor forceps and bipolar diathermy.
As the debulking proceeds it becomes possible to mobilize the capsule of the tumor, separating it from the underlying cerebellum and brainstem before removal. It is important that intracapsular debulking proceeds as far as possible before a search is made for the facial nerve at the brainstem. This minimizes the need for later retraction on the nerve.

The facial nerve is likely to lie anteriorly and inferiorly to the bulk of the tumor. The tumor will almost certainly distort the course of the nerve, which may be attenuated significantly near the porus. The nerve is nearly always found on the brainstem anterior and slightly inferior to the vestibulocochlear nerve, which is a regularly identified landmark as it runs into and blends with the tumor. The nerve ascends along the brainstem, deep to the tumor until it takes a near right-angled bend to run across the cerebellopontine angle to the porus. It is at the porus itself that the facial nerve is most at risk, which emphasizes the need for maximal bone removal at this point. The nerve monitor and stimulator are invaluable in the presence of such altered anatomy, and the authors find the use of an insulated Fisch-type middle ear disector particularly helpful, as it reduces the need to change between instruments and provides early warning should the nerve be encountered unexpectedly during dissection (Fig. 15.10).

Debulking and mobilization continue from medial to lateral. The anterior inferior cerebellar artery runs in or outside of the arachnoid mesh that surrounds the tumor. Generally a ready plane of cleavage is found deep to the arachnoid, allowing the artery to drop away out of harm’s way. The vestibulocochlear nerve is best left intact at the brainstem until such time as this compromises access, as it will protect the facial nerve from some of the traction caused by tumor mobilization. Dissection of the tumor from the lateral end of the IAC is performed last. As the facial nerve is firmly fixed in its labyrinthine segment, even mild traction here may result in some loss of function; if there is still a significant bulk of tumor present at this stage, the pull of its weight alone may be enough to damage the nerve. This again emphasizes the need for extensive debulking before formal tumor removal. Once the facial nerve has been identified here, the final remaining fragments or tumor are removed, along with the vestibular nerves. When tumor removal is complete, the integrity of the facial nerve is once again tested with the stimulator, as this has been shown to be of valuable prognostic significance; evidence of conduction at a stimulator current of 0.05 mA is a favorable sign.7

◆ Closure and Postoperative Care

After meticulous hemostasis the cerebellopontine angle is filled with Hartmann’s solution to prevent pneumocephalus,8 and the dural defect is covered with a temporalis fascia graft. If not already done, the eustachian tube and middle ear are packed firmly with muscle. Subcutaneous fat is harvested from the left iliac fossa (to prevent confusion with an appendicectomy scar) and used to fill the mastoid cavity. The wound is closed in layers, aiming for a watertight closure. The combination of interrupted muscle and subcutaneous layers of an absorbable braided suture material (2–0 Vicryl) and a continuous subcuticular layer of absorbable monofilament (4–0 Monocryl) generally produces highly satisfactory results. A firm head bandage is applied and left in situ for 3 days.

If the total duration of surgery has been less than 5 hours, the patient does not usually go to the intensive care unit unless dictated by comorbidity. The urinary catheter is removed and the patient encouraged to mobilize the second day after surgery. Prophylactic antibiotics are given for 24 hours, but steroids or mannitol are not used. An aperient is routinely prescribed to avoid straining and the risk of precipitating a cerebrospinal fluid leak.

Acknowledgments

The senior author (P.A.F.) wishes to pay tribute to three surgeons whose experience and generosity encouraged and guided him to an interest in translabyrinthine surgery and one hopes a certain degree of skill. They are Bill House, who began it all; Derald Brackmann; and the late and much missed Antonio de la Cruz.

References

William House refined the middle fossa approach in 1961. Initially, he used this approach to decompress the internal auditory canal (IAC) in cases of extensive otosclerosis with sensorineural hearing loss. Although not beneficial for that condition, it quickly became evident that this approach was suitable for small acoustic tumors in patients with good hearing. The middle fossa procedure is a hearing-preservation approach. For tumors, it allows complete exposure of the lateral end of the IAC, so no blind dissection is necessary at the fundus. This is a definite advantage over the retrosigmoid approach, which does not provide exposure of the lateral third of the IAC without loss of hearing. Disadvantages include the fact that this is a technically difficult approach. Because of the superior location of the facial nerve in the canal, dissection of the tumor may subject the facial nerve to more manipulation than in other approaches. But with the advent of facial nerve monitoring, problems of the facial nerve are kept to a minimum and facial nerve outcome is no different than when using the translabyrinthine approach for tumors of similar size, at least in experienced hands. Retraction of the temporal lobe is required for the duration of the drilling and tumor removal, which usually does not exceed 1 to 1½ hours.

In the treatment of acoustic tumors, the translabyrinthine and middle fossa approaches are often compared with the retrosigmoid approach. The translabyrinthine approach provides wide access to the cerebellopontine angle (CPA) with little cerebellar retraction, and permits exposure of the entire facial nerve from the brainstem to the stylomastoid foramen. The exposure is mostly extradural, minimizing possible injuries to the brain and to the cerebellum. In addition, it provides a more direct and anterior perspective. A shorter distance separates the surgeon from the contents of the CPA. The translabyrinthine approach enables identification of the facial nerve at the lateral end of the IAC before tumor dissection. The main disadvantages of the translabyrinthine approach are the need to sacrifice hearing in the operated ear, the limited exposure of the lower part of the CPA, and the limited access to the neural contents of the foramen magnum and foramen jugulare.

The retrosigmoid approach provides a panoramic view of the posterior fossa from the tentorium to the foramen magnum. Access is provided to the cerebellar hemisphere, the lateral aspect of the pons and medulla, and the root entry zone and cisternal course of cranial nerves V to XI. Although exposure superiorly is limited by the tentorium, this approach could be combined with a middle fossa or transtentorial exposure. The retrosigmoid approach represents a modification of the classical suboccipital approach. Krause and others first employed the suboccipital route during the latter portion of the 19th century. In this procedure, a large bone flap is removed from the suboccipital area, with the anterior limit of the dissection being the first mastoid cell. Superiorly, bone is removed up to the inferior margin of the transverse sinus. The retrosigmoid approach offers a more favorable angle of view into the CPA and a markedly reduced need for cerebellar retraction than the classic suboccipital approach. It does not pose a risk of air embolism or quadriplegia, as does the classic suboccipital approach, which places the patient in the sitting position. Advantages are wide access to the CPA and the potential for hearing preservation. The retrosigmoid approach is capable of addressing most lesions of the CPA. Disadvantages include a higher incidence of postoperative headaches and cerebrospinal fluid (CSF) leaks when compared with the translabyrinthine approach. The lower incidence of postcraniotomy headaches in the translabyrinthine approach may be due to lesser cerebellar retraction than the classic suboccipital approach. The retrosigmoid approach is capable of addressing most lesions of the CPA. Disadvantages include a higher incidence of postoperative headaches and cerebrospinal fluid (CSF) leaks when compared with the translabyrinthine approach. The lower incidence of postcraniotomy headaches in the translabyrinthine approach may be due to lesser cerebellar retraction than the classic suboccipital approach.
approaches of choice for removal of most acoustic tumors because they provide access to the whole length of the IAC. This chapter describes the middle fossa craniotomy, its indications, and the results.

**Indications**

The main indication for the middle fossa craniotomy is removal of intracanalicular vestibular schwannomas in patients with good hearing. Arbitrarily, good hearing has been defined as a speech reception threshold of at least 30 dB and a word recognition score of at least 70%. Exceptions to this rule do exist, as in the case of poor hearing in the contralateral ear or in bilateral tumors. Typically we use the middle fossa approach for tumors up to 2 cm in size in patients with at least serviceable hearing. In selected cases, the extended middle fossa craniotomy is used. In general, this approach is reserved for patients younger than 65 years of age because of the fragility of the dura and the underlying temporal lobe in older patients. Another indication for the middle fossa craniotomy is decompression of the IAC in patients with neurofibromatosis type 2 to allow the tumor room for expansion to preserve the hearing on that side. The middle fossa craniotomy also can be used to perform vestibular neurectomies when indicated, although we prefer the retrolabyrinthine craniotomy for vestibular neurectomy.

The middle fossa approach is also used to repair symptomatic superior canal dehiscence and in patients with large tegmen defects with encephaloceles and CSF leaks.

**Preoperative Studies**

Magnetic resonance imaging (MRI) is necessary to show the exact location and size of the tumor. Tumor size is thought to be an important prognostic factor, with some studies showing that the smaller the tumor, the better the chances at hearing preservation. MRI determines the relationship of the tumor to the brainstem, cerebellum, and IAC. Particular attention should be paid to a major vessel loop crossing within the tumor. The nature of the tumor and its consistency, cystic versus solid, can also be evaluated. Other predictive factors are provided by audiometry, auditory brainstem response (ABR), and electroneurogram (ENG). The better the hearing in the ear, the better the chance of preserving useful hearing. Patients with a normal ABR have a better chance of postoperative hearing preservation. Superior vestibular nerve tumors indicated by abnormal caloric responses are also thought to be favorable prognostic factors. However, in some studies, tumor size and ENG results were not predictive.

**Surgical Technique**

A standard endotracheal anesthesia induced with thiopental, and a short-acting muscle relaxant is used. Intravenous furosemide 40 mg and mannitol 1 g/kg body weight are administered for brain relaxation when the skin incision is made. Preoperative antibiotics are also administered. The blood pressure is monitored using an arterial line.

Facial nerve monitoring is done for the entire period of the operation. Cochlear nerve function is monitored using ABR; once the tumor is exposed, direct cochlear nerve potentials are obtained just prior to tumor removal.

The patient is placed in the supine position, with the head rotated. We do not use a headholder, although one may be used if greater posterior exposure is required. The operating room table is reversed so that the patient’s head is located on the foot of the bed; this allows the surgeon to sit and work comfortably during the procedure without any obstruction under the table. A long anesthesia circuit permits the anesthesiologist to stay at the other end of the surgical site. An electrically controlled table enables the frequent turning from side to side needed during neurotologic procedures.

The middle fossa approach facilitates the unroofing of the IAC and the exposure of the fundus of the canal. The facial nerve is located at the lateral end of the canal, where it enters the temporal bone and becomes the labyrinthine segment between the cochlea and the superior semicircular canal. This approach makes possible the removal of laterally located tumors in the IAC without the need for blind dissection.

The incision starts in the pretragal area, curves initially posteriorly above the ear, and then runs vertically for 4 cm before curving at a right angle anteriorly in the temporal area. The shape of the incision resembles a question mark (Fig. 16.1). Once the skin is elevated, an incision is made along the insertion of the temporalis muscle and fascia, and the muscle is reflected anterior inferiorly.

**Fig. 16.1** The incision starts in the pretragal area, curves initially posteriorly above the ear, and then runs vertically for 4 cm before curving at a right angle anteriorly in the temporal area.
Using a cutting bur, an opening is made in the squamous portion of the temporal bone. The craniotomy measures $5 \times 5$ cm and is located two thirds anterior and one third posterior in relation to the external auditory canal (Fig. 16.2). The bone flap is kept in antibiotic solution and is placed back at the conclusion of the surgery. The dura is now elevated from posteriorly to anteriorly from the floor of the middle fossa, and any remaining bone over the root of the zygoma is drilled away as close as possible to the floor of the middle fossa (Fig. 16.3). The initial landmark, the middle meningeal artery, marks the anterior extent of dural elevation. Frequently, bleeding is encountered in this area and is controlled by packing Surgicel into the foramen spinosum. Dissection of the dura continues until the petrous ridge is identified. Once the dura is completely elevated, the House-Urban retractor is placed into position over the porus acusticus (Fig. 16.4). At this point in time, the arcuate eminence and the greater superficial petrosal nerve have been identified. In a small proportion of patients, the geniculate ganglion is dehiscent, and care is taken not to injure it while elevating the dura. Posterior to anterior elevation avoids elevating the ganglion.

Using suction irrigation and diamond burs, dissection of the IAC is started medially. The IAC bisects the angle formed by the greater superficial petrosal nerve and the arcuate eminence as described by Bocca and García-Ibáñez. Identifying the IAC medially and anteriorly is safest because medially there are no important anatomic structures. Once the IAC is identified, bone surrounding it in the area of the porus is removed. Bone removal extends posteriorly to the level of the arcuate eminence and the common crus, anteriorly to Kawase's triangle. Bone is removed 270 degrees around the canal, including the entire posterior lip. Lateral dissection of the IAC then proceeds. The exposure narrows laterally be-
cause of the presence of the cochlea anteriorly and the am-
pullated end of the superior semicircular canal posteriorly.
At the lateral end of the canal, Bill’s bar is identified (Fig.
16.5). The facial nerve is followed into its labyrinthine por-
tion. The ligament surrounding it at the beginning of the laby-
rinthine segment is cut to allow for decompression of the
nerve in this portion.

The dura of the IAC is opened along the posterior aspect
(Fig. 16.6). The facial nerve is identified clearly and stimu-
lated. The superior vestibular nerve is cut at the end of the
IAC. Following this, the vestibulofacial anastomotic fibers are
cut. The tumor is then separated from the end of the IAC and
from the facial nerve (Fig. 16.7). The goal is to free the tumor
from the facial nerve and to deliver it from under the nerve.
Dissection of the lateral end of the inferior compartment of
the IAC can be very difficult. It is best to cut both superior
and inferior vestibular nerves to avoid postoperative unstead-
iness. Once the lateral end of the tumor has been delivered,
the plane between the cochlear and facial nerves and tumor
become apparent. This plane is developed using fine hooks.
Tumor dissection proceeds from medial to lateral to avoid
stretching the facial and cochlear nerves. At this point, a
search for the anterior inferior cerebellar artery begins. Great
care is taken to identify and not injure this important artery.
At this point, the tumor is separated gently from this vessel.
Debulking of the tumor begins, using small cup forceps. At
all times, care is taken not to injure the facial nerve with the
suction or by stretching it. Finally, the medial end of the
tumor is freed with small hooks.
Papaverine pledgets are placed over the nerve and in the
dissection field if changes in monitoring potentials are noted.

Fig. 16.4 Placement of the House-Urban retractor and exposure of
the internal auditory canal.

Fig. 16.5 The vertical crest or Bill’s bar separates the facial nerve
from the superior vestibular nerve at the fundus of the internal
auditory canal.

Fig. 16.6 The dura is incised along the posterior border of the
dissection.
This is thought to prevent vasoconstriction of the vessels feeding the inner ear and therefore enhancing chances at hearing preservation.

Once tumor removal is completed, hemostasis is obtained (Fig. 16.8). The tumor bed is irrigated copiously. Abdominal fat is obtained and is used to close the dural defect. The temporal lobe retractor is removed. The dura is suspended on either side of the craniotomy to limit the dead space. A Penrose drain is placed into the wound. The bone flap is repositioned and secured in place using three titanium microplates. The wound is closed in layers, and a mastoid-type pressure dressing is applied.

◆ Postoperative Care

The patient is observed in the intensive care unit for a period of 24 hours. Steroids and antibiotics are routinely used. For the middle fossa approach, the Penrose drain is removed from the wound on the first postoperative day. A new pressure dressing is applied. The wound is inspected every day thereafter. The mastoid dressing remains in place for 4 days and the patient is instructed not to lift or strain during the early postoperative period.

◆ Perioperative Complications

Slattery et al.11 reviewed the perioperative morbidity of acoustic tumor surgery for a series of 1687 patients having surgery at the House Ear Institute (Los Angeles, CA) between 1987 and 1997. This included tumors of all sizes and using all surgical approaches. A CSF leak was the most common problem, occurring in 9.4% overall. Looking at the data from that study for just those tumors that underwent a middle fossa ($n = 432$) approach (mean size, 1.1 cm; min-max, 0.3–4.0), CSF leak occurred in 24 of the cases (5.6%), with reoperation required for treatment in only four cases (0.9% of all). In tumors operated with the middle fossa approach from 1998 through 2004 at the institute ($n = 692$), 4.4% had a CSF leak. Most leaks can be stopped with a pressure head dressing and bed rest with the patient’s head elevated. If the leak continues despite having the dressing in place, a lumbar spinal drain is inserted and kept in place for 3 to 4 days. Reexploration of the wound and repacking of the wound with additional fat is done if the leak persists despite the above steps. If the hearing is lost, the leak is controlled by performing a blind sac closure of the external auditory canal and obliteration of the middle ear space and eustachian tube using Surgicel and muscle.

Meningitis is an uncommon complication and is managed with appropriate antibiotics following culture and identification of the offending organisms. Middle fossa cases from the Slattery et al study showed a prevalence of meningitis of 1.2%. For middle fossa cases of the past 7 years at the institute, meningitis occurred in 0.3%.

Although rare, the most common early postoperative complication is a hematoma in the CPA, occurring in 0.9% of all size tumors in the Slattery et al study and 0.7% of the middle fossa surgeries. This is manifested by signs of increased CPA pressure, and is managed by immediate opening of the wound and removal of the fat in the intensive care unit. The patient is then taken to surgery to secure hemostasis and repack the wound. The incidence of this complication is lowered by leaving a Penrose drain in the wound for the first 24 hours, and then removing it on the first postoperative day.
by obtaining meticulous hemostasis prior to closure of the wound.

Other perioperative complications occurring in the Slattery et al study for tumors removed by the middle fossa approach include craniotomy wound infection (1.7%) and cerebral edema (1.4%). There were no cases of hydrocephalus, embolus, thrombosis, abdominal wound infection, stroke, or death.

If facial weakness occurs, the eye is protected by using conservative measures first. These include artificial tears, moisture chambers, and soft contact lens. In certain situations, the insertion of a gold weight or a palpebral spring may be necessary. In middle fossa cases from the Slattery et al study, 1.4% required one of these surgical procedures, as did 0.9% of the cases in the 1998–2004 series.

Possible problems related to temporal lobe retraction include memory loss, auditory hallucinations, and speech disturbances. These are rarely significant clinical problems. Seizures are often cited as a possible complication of the middle fossa approach, but in reality this problem is rarely encountered. Older patients do not tolerate the middle fossa approach as well as younger patients due to the fragility of the dura and retraction of the temporal lobe. Fortunately, all of the above complications have been very rare in our experience.

◆ Results

Complete tumor removal has been demonstrated in more than 95% of patients undergoing microsurgical resection, and was accomplished in 94.6% of all middle fossa surgeries at the House Ear Institute (n = 1183) and 95.1% of the 1998–2004 series. Tumor recurrence has been shown to occur in 0.3% or less of cases with long-term follow-up. Cranial nerve morbidity, particularly of cranial nerves VII and VIII, is of particular concern when judging the success of any form of treatment modality for vestibular schwannomas.

Facial Nerve

Previous publications from the House Ear Institute have reported a follow-up (1-year) rate of 95.8% good facial function (House-Brackmann grades I or II) in tumors less than or equal to 1.5 cm in size and removed using the middle fossa approach, a rate that was not statistically significantly different from that of the translabyrinthine cases. For tumors 1 cm or less in size, results were similar. For the recent 7-year period, 692 tumors underwent middle fossa removal, with a good facial function rate of 90.2%, including all tumor sizes (up to 6 cm, with 16>1.5 cm).

Others have reported facial nerve results for small tumors removed using the middle fossa approach. For example, Satar et al reported rates of good facial function (House-Brackmann grades I or II) in 93.7% of 64 intracanalicular tumors and 97.6% of 42 tumors with 1 to 9 mm CPA extension.

Hearing Preservation

For middle fossa procedures, hearing preservation is a goal. Slattery and Brackmann reviewed the published results of hearing preservation for both the suboccipital/retrosigmoid and the middle fossa approaches. As they note, there are different definitions of hearing preservation, including simply preserving the cochlear nerve, any measurable hearing, serviceable hearing, or lack of change in hearing from the preoperative level. Hearing preservation rates, as reported by the various authors reviewed, ranged from 16.5% to 65% for the suboccipital approach and 36% to 71% for the middle fossa approach.

A report from the House Ear Institute on all tumors operated using the middle fossa approach over a 2½-year period from 1993 to 1995 found measurable hearing preserved in 68% of 143 patients. Hearing was preserved within 15 dB pure tone average (PTA) and 15% speech discrimination score (SDS) of preoperative levels in 52%; 47% did not change or improved their hearing class using the American Academy of Otolaryngology–Head and Neck Surgery (AAO-HNS) classification system. In a more recent paper from the institute, hearing preservation was evaluated by tumor size for patients operated using the middle fossa approach between 1992 and 1998. Again, no effect of tumor size on hearing outcome was found. For 107 tumors of 1 cm or less in size, some measurable hearing was preserved in 79%, and serviceable hearing (equal to or better than 50 dB PTA and 50% SDS) was preserved in 60%. Hearing was preserved to within 15 dB and 15% of preoperative levels in 52%. For the most recent 7-year series, 59.1% of the 428 patients with audiologic data available to compute the AAO-HNS class had class A or B results (50 dB/50%), whereas 73.2% had measurable hearing. Long-term stability of hearing preservation in our middle fossa vestibular schwannoma cases has also been examined. Friedman et al found that 70% of patients with immediate postoperative serviceable hearing maintained serviceable hearing at more than 5 years after surgery.

Others have also reported hearing preservation rates in small tumors. Staecker et al reported a hearing preservation rate of 57% in mainly intracanalicular tumors removed by the middle fossa approach and 47% in similar tumors removed using the retrosigmoid approach. Satar et al reported rates of functional hearing preservation in intracanalicular tumors and those with up to 9 mm CPA extension of 62% and 63%, respectively. These studies, as well as our own data, all show preservation of serviceable hearing in approximately 60% of patients undergoing middle fossa acoustic tumor resection for small tumors.

◆ Conclusion

The middle cranial fossa approach is a technically demanding approach. It remains our approach of choice for patients with small acoustic tumors and serviceable hearing. Hearing preservation rates are approximately 60 to 70%. Results on the facial nerve outcomes are no different from those for other more traditional approaches. The safety and efficacy of the middle fossa approach in the area of acoustic tumor resection, our main indication, has been clearly demonstrated over the years.
References

Microvascular Decompression for Hemifacial Spasm and Trigeminal Neuralgia

Tetsuo Kanno and Iyer Viswanathan

The microvascular decompression of cranial nerves has become an accepted surgical technique for the treatment of trigeminal neuralgia, hemifacial spasm (HFS), glossopharyngeal neuralgia, and other cranial nerve neuropathies. It is a surgery that requires sound anatomic knowledge and surgical finesse. The cerebellopontine angle has important vessels and nerves that may be damaged, leaving the patient with severe deficits. The patients are generally healthy except for the neuralgia; therefore, such deficits are unacceptable.

This chapter is based on the surgical experience of over 837 cases since 1980, and we discuss the lessons we have learned from our complications and failures.

◆ History

Observations of vascular compression of the seventh cranial nerve in patients with HFS were first reported by Campbell and Keedy in 1947 and by Laine and Nayrac in 1948. Gardner and Sava in 1962 proposed decompression of the facial nerve for HFS. Nicolas André coined the term tic dououreux, and in 1756 discussed surgical treatments in five patients who suffered with a “cruel and obscure illness, which causes . . . in the face some violent motions, some hideous grimaces, which are an insurmountable obstacle to the reception of food, which put off sleep.”4 In 1966, Jannetta and Rand proposed that the etiology of trigeminal neuralgia (TN) lies in the compression of the trigeminal nerve by small vessels near the brainstem. Since that time, microvascular decompression (MVD) has become a mainstay in the management of this painful condition.

◆ Procedure

The operative steps are described and illustrated in the following subsections. The steps are designed to achieve clinical pain relief while avoiding potential complications.

Intraoperative Monitoring

Facial nerve monitoring is done in cases of HFS, but we have found that the results of monitoring do not always correlate with clinical results.6 Intraoperative monitoring of brainstem auditory evoked potentials (BAEPs) is a useful tool to decrease the danger of hearing loss, and we use it in most cases.

Positioning of the Patient

The success of the microvascular decompression procedure is significantly determined by patient positioning. The authors prefer the modified park-bench position (Fig. 17.1). Following anesthesia and intubation, a Sugita head fixation device is applied and the patient is placed in the lateral decubitus position with appropriate padding of pressure points and an axillary roll. The neck is flexed slightly with the chin approximately two fingerbreadths from the sternum. The head is rotated approximately 10 degrees away from the affected side. If a trigeminal or cochlear approach is planned, the vertex is kept parallel to the floor to keep the cranial nerve VII-VIII complex at a more inferior position in relation to the trigeminal nerve. The head holder is secured in the proper position. The patient is fixed securely with belts to the table at the hip and chest to allow rotation of the table during the operation. The patient’s shoulder is taped down and placed out of the way.

Surgical Incision

A limited area behind the ear is shaved, and bony landmarks are identified by palpation before the patient is prepared for surgery. The surgeon should have an idea of the position of the transverse and sigmoid sinus for proper bur-hole placement. An S-shaped incision beginning approximately 5 cm above the midmastoid is taken down 3 cm posterior to the inferior edge of the mastoid tip (Fig. 17.2). Three quarters of
the incision is drawn inferior to the junction of the transverse and sigmoid sinuses and one quarter above. This curved incision lowers the height of reflected skin and subcutaneous tissue on the surgeon’s side (Fig. 17.3).

The incision is extended down to the occipital bone by using monopolar cautery. The soft tissues are dissected using a periosteal elevator and electrocautery where necessary. The nuchal muscles are dissected with electrocautery and the transverse occipital artery is ligated and divided. The mastoid emissary vein usually bleeds and is waxed in its fossa. An angled mastoid retractor is put in place. The loose fatty tissue at the inferior aspect of the digastric groove is preserved to avoid injury to the vertebral artery and condylar vein.

**Craniotomy**

Before bur-hole placement, bone landmarks should be well exposed. The bony exposure should be enough to identify the edge of the junction of the transverse and sigmoid sinuses. A small craniectomy can then be tailored according to the cranial nerve approach desired. The craniotomy for HFS is shaped like an inverted triangle with a 3-cm maximum dimension. The craniotomy for trigeminal neuralgia is more elliptical and also more cranial by approximately 1 cm (Fig. 17.4). The advantage of the inverted triangle is that the retractor can be moved along the sigmoid sinus to visualize from the cranial nerve VII-VIII complex to the IX-X-XI complex. Mastoid air cells should be thoroughly waxed. The bone work must be anterior enough to allow dural incision and reflection (Fig. 17.5).

**Dural Opening**

The surgeon should allow some drainage of cerebrospinal fluid (CSF) before placing the microretractor. As the CSF is being drained, the cerebellum will begin to fall away from the cerebellopontine angle. Some unpleasant bleeding from the torn bridging veins may be encountered, which can be managed by suction over a cottonoid and bipolar diathermy. We usually try to identify the VII-VIII complex through the arachnoid. In cases of HFS, we cut the arachnoid down to the lower cranial nerves and in TN we cut it up to cranial nerve V and the petrous vein. It is important to open the arachnoid with sharp dissection to see the root entry zone (REZ). In HFS, the arachnoid is cut over the VII-VIII complex, the IX-XI complex, and choroid plexus (Fig. 17.6).
Fig. 17.3  The advantage of the curved incision is that it lowers the height of reflected skin and subcutaneous tissue on the surgeon’s side.

Fig. 17.4  The schematic relation of the craniotomy to the bony landmarks and venous sinuses. (A) posterior view; (B) inferior view.
The senior author does not hesitate to use retractors until the REZ becomes visible. It is also easy to elevate the arachnoid membrane with suction. The large size of the flocculus in some patients makes it necessary to use a retractor to see the REZ (Fig. 17.7). After this, retractors can be avoided altogether to prevent pull on cranial nerves VII and VIII. In cases of TN we prefer the lateral approach of identifying the VII-VIII complex and then moving cranially to cranial nerve V. It is better to apply retractors after the petrosal vein is lax.

Identification of Offending Vessel and Nerve Decompression

The basis of surgery in HFS is the vessel compression at the REZ where the oligodendroglia changes to the myelin sheath (Fig. 17.8). The common vessels causing compression of the facial nerve are the posterior inferior cerebellar artery (PICA) and the anterior inferior cerebellar artery (AICA). The vessel must be sharply dissected free from the arachnoid and mobilized laterally away from the nerve so that a Teflon implant can be placed (Fig. 17.9). Fibrin glue is applied over the Teflon to prevent slippage. In cases of atypical HFS, the pathologic vascular entity is almost always located rostral to the nerve or between cranial nerves VII and VIII. When the anterior inferior cerebellar artery is transposed laterally, a small branch to the pons may get stretched.

Compression of cranial nerve VII by the vertebral artery is a more challenging job for the surgeon. The artery may compress the nerve all along the course or at the REZ behind the cranial nerve IX (Fig. 17.10). The AICA also may run along the nerve, but decompression of only the AICA does not relieve
the facial spasm. Vertebral artery compression has to be treated by transposition and not Teflon alone. The vertebral artery is transposed laterally, wrapped in Surgicel and Teflon, and coated with fibrin glue or cyanoacrylate (Fig. 17.11). Microvascular decompression of the trigeminal nerve requires sharp dissection of all arachnoid around the trigeminal nerve and superior cerebellar artery. The most common vessel found is a superior cerebellar artery loop, which compresses the trigeminal nerve either at the brainstem or distally (Fig. 17.12). The nerve should be inspected from its origin at the brainstem laterally to its exit from the cerebellopontine angle, and all vessels should be treated. After the arachnoid is dissected and the vessel is freed, the loop can be mobilized to the lateral aspect of the nerve, and a piece of shredded Teflon felt can be placed between the vessel and the nerve.

In some cases, no offending artery is found but a vein is identified running over the REZ of cranial nerve V. This vein must be ablated. Sometimes a branch of the petrosal vein may be the offending vessel and needs to be ablated (Fig. 17.13). It is important not to ablate all the petrosal vein branches. Brain retraction must be avoided after petrosal vein ablation to prevent hemorrhagic venous infarction. In certain cases, despite performing a thorough search, no compressing vessel is found but cranial nerve V looks kinked. This kinking itself can produce neuralgia and needs to be corrected.
Closure

At the conclusion of the procedure, repetitive Valsalva maneuvers are performed to ensure hemostasis. The retractor is then removed and the cerebellar surface is carefully inspected. The area is gently irrigated with warm saline and the dura is closed. We insist on watertight dural closure in which fascia/muscle grafts from the inferior portion of the incision are used when necessary. The bone edges of the mastoid air cells are thoroughly waxed for a second time. A small strip of polyglycolic acid is placed over the suture line and fibrin glue is applied over it. Gelfoam is then placed over the durotomy. The bone defect may be either repaired by titanium mesh or left alone. The deep and superficial muscles

![Fig. 17.10](image1.png) (A) Partial compression on cranial nerve VII by the vertebral artery. (B) Complete compression on cranial nerve VII by the vertebral artery.

![Fig. 17.11](image2.png) Transposition of the vertebral artery (VA). Alon α, fibrin glue.

![Fig. 17.12](image3.png) Compression of cranial nerve V by the superior cerebellar artery (Sup. Cerebellar A.).
are approximated with interrupted No. 2-0 absorbable sutures, and the fascia is closed with the same type of sutures. The fascial closure has to be good to prevent any CSF leakage. We prefer interrupted sutures. The subcutaneous tissues are approximated with No. 3-0 interrupted absorbable sutures. A well-approximated subcutaneous layer can also afford protection from CSF leakage. The skin is closed with No. 4-0 nylon or staples.

Use of Endoscope in Microvascular Decompression

Why do some microvascular decompression procedures fail? One reason is misidentification of the compressing vessels. The aim of the endoscope is to increase the yield of compressing vessels. With the endoscope, we can visualize areas behind the nerves, vessels, flocculus, and cerebellum that are not visible to the microscope. We use the Machida right-angled endoscope, which is easy to handle and safe (Fig. 17.14). In two or three cases out of 50 it provides the neurosurgeon with additional information. The space behind cranial nerve V can be deep and large veins traverse the space. Endoscopy is useful to look at the details of this space. It is of particular importance when the nerve is compressed from behind, so that the compression point can still be seen clearly and tackled (Fig. 17.15).

Failure of Microvascular Decompression

Even for experienced surgeons, it is still impossible to accomplish a 100% cure rate. Most series report 85 to 96% cure rates after microvascular decompression surgery. Recurrence is defined as persistent or diminished facial spasm after a period of complete resolution after microvascular decompression. The authors have performed 837 procedures of microvascular decompression, of which 550 were for HFS and 287 for trigeminal neuralgia. We assess the cure rates for microvascular decompression in cases of HFS 12 months after surgery. We had a cure rate of 95.3% up through 2001, and have analyzed the cases of recurrence that we managed and did a literature review to postulate certain causes of recurrence.

Wrong Insertion of Teflon Patch

It is important to insert the Teflon patch at the REZ of cranial nerve VII, which is located behind cranial nerve IX. Peripheral insertion may lead to recurrence. At the second surgery, lower cranial nerves were carefully dissected and the REZ was well exposed. Decompressions were made at the level of REZ with good results.

Missed Compressive Element

Sometimes the vascular compressive element may be missed on initial surgery. The initial pain relief may be because of partial nerve injury. We have had two cases in which the internal auditory artery was found as the compressive element at reexploration (Fig. 17.16). Sometimes both the AICA and the vertebral artery may be compressing the nerve (Fig. 17.17). One of them may be missed at the initial surgery. Here once again, we insist on the need to look at the nerve from the REZ to the periphery. Venous compression is generally undertreated and may lead to recurrence.

Slippage or Decomposition of the Teflon Sponge

Many times at reexploration, the muscle plug used during the previous surgery was found to have decomposed. This is rarely seen with Teflon. To prevent slippage we use Surgicel and glue on the Teflon patch.
Scarring

The implant itself may produce severe scarring in some cases, causing nerve deficits and recurrence. Hence, it is important to use thin-shredded Teflon in optimum quantities. We have had a case in which a large patch was demonstrated on postoperative computed tomography (CT) scan causing facial palsy. The patient was reexplored and the patch was removed.

Fig. 17.15  Diagrammatic (A) and endoscopic (B) views of the vessel compressing cranial nerve V.

Demyelination

Bederson and Wilson reported in 1989 that prolonged extrinsic compression may lead to intrinsic demyelinating lesion, which will not respond to decompression. Reexploration of such patients may not lead to any compressive element, and a partial rhizotomy or other ablative procedures may have to be considered.

Fig. 17.16  (A) At first surgery, only the anterior inferior cerebellar artery (AICA) was transposed. (B) At the second surgery, the internal auditory artery was also found compressing the nerve and was decompressed. The arrows indicate the insertion of the Telfa (cottonoid) in the plane between the artery and the nerve.
Complications

The complications encountered in our series included cerebellar injury and CSF leakage. Other complications of the procedure such as facial weakness or anesthesia, hearing loss, and lower cranial nerve dysfunction were rare. Most cases with postoperative facial weakness improved within 3 months. Unfortunately, one patient required plastic surgery for the facial palsy. Another patient lost hearing completely.

The following surgical errors may lead to facial paresis or palsy:
- Direct nerve injury
- Extensive coagulation near the facial nerve
- Wrong tangent of retraction: retraction should never pull on the cranial nerve VII-VIII complex and should be avoided after the REZ is exposed
- Large Teflon patch, as already mentioned, may itself produce neuralgia
- Postoperative viral infection, commonly varicella zoster virus

Conclusion

Microvascular decompression is one of the most gratifying procedures in neurosurgery. Proper surgical techniques can alleviate the patient’s symptoms and make it an excellent operation for a painful disease.

References

Radiosurgery is a minimally invasive technique designed to deliver a high dose of ionizing radiation in a single treatment session using multiple beams precisely focused at the intracranial target, eliciting a specific radiobiologic response. The long-term results of radiosurgery have proven it to be an effective, minimally invasive tool in the armamentarium for the treatment of complex neurosurgical conditions. In radiosurgery, the target is not immediately destroyed or extirpated from the brain, but is instead exposed to a high dose of radiation, which ultimately results in a specific and toxic radiobiologic response, leading to arrest of the cell division, irrespective of the individual cell's mitotic activity, oxygenation, and radiosensitivity. Radiosurgery can be performed using the gamma knife, a linear accelerator-based system, or a heavy-charged particle beam.

◆ Gamma Knife

The pioneering work on the development of the gamma knife was done at the Karolinska in Stockholm, Sweden. The most recent model is the Leksell Gamma Knife Perfexion (Elekta Instruments AB, Stockholm, Sweden) (Fig. 18.1). This model differs from previous models B and C of the Leksell gamma knife (Fig. 18.2). The 4- and 8-mm collimators remain, but the 14- and 18-mm collimators have been replaced with a 16-mm collimator. There is no collimator helmet, and the couch now serves as the patient positioning system. Doses to critical structures can be limited by a process called dynamic shaping. Extracranial doses are significantly lower than the previous units. It is now possible to treat pathology up to 26 cm caudal to the vertex of the cranium; as a result, head and neck cancers are now within the scope of gamma knife radiosurgery (GKRS). The previous versions of the gamma knife (models C and 4C) uses an automated positioning system (APS), eliminating the need for manual setting of the coordinates for each isocenter. Facility for image fusion is present in Leksell gamma knife model 4C.

◆ Gamma Knife Radiosurgical Procedure (Models B and C)

The gamma knife radiosurgical procedure involves four steps:
1. Application of the Leksell frame to the patient’s head
2. Stereotactic brain imaging
3. Dose planning
4. Stereotactic radiation delivery

Application of Leksell Frame

The procedure begins with rigid fixation of a magnetic resonance imaging (MRI)-compatible Leksell stereotactic frame (model G; Elekta Instruments, Atlanta, GA) to the patient’s head after careful review of the preoperative images. Frame placement is one of the most important aspects of the procedure (Fig. 18.3). The frame should be placed in such a way that the lesion should be as close to the center of the frame as possible. The scalp at the pin sites is infiltrated with local anesthesia, and the frame is secured rigidly to the outer table of the skull with pins. For lateral lesions, the frame is shifted laterally (right or left) toward the lesion. It is important to make sure that there is enough space on the contralateral side to allow positioning of the fiducial box on the frame before sending the patient for stereotactic imaging.

Stereotactic brain imaging

Magnetic resonance imaging is the preferred modality of imaging available at present for radiosurgery planning. High-
resolution MRI is used for stereotactic radiosurgical planning in almost all eligible cases, except arteriovenous malformations (AVMs). The accuracy of images is checked for each image sequence by comparing the known frame measurement with image measurements. The accuracy of target localization is ensured with a properly shimmed magnet, regular servicing, and strict quality assurance on the MRI unit. A computed tomography (CT) scan is occasionally used for radiosurgical planning when MRI is contraindicated for any reason. For AVM planning, both digital subtraction angiography (DSA) and MRI images are acquired. CT cisternography can be used for treatment planning for trigeminal neuralgia if an MRI is contraindicated.

Dose Planning

Stereotactic images are transported via a fiber optic Ethernet to the Gamma Plan (Elekta Instruments, Atlanta, GA) computer, where images are first checked for any distortion or inaccuracy. Planning is performed on axial MRI scans with coronal and sagittal reconstructions. Depending on the size and shape of the target volume, the appropriate collimator is used for each shot placement. The position and weightage of the shots can be adjusted to match the target volume. Every shot or isocenter has three coordinates (x, y, and z). Proper dose selection is very important for achieving conformity and for yielding a good response without any adverse effects. Shields can be used to block the radiation dose to critical structures. Various plugging patterns can be used to modify the shape. After completion of the dose-planning portion of the GKRS, it is important to perform a collision check on all shots to determine shot positions that may not be put into effect (unachievable due to the possibility of a collision between the collimator helmet, the patient’s skull, and/or any...
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part of the frame). A neurosurgeon, radiation oncologist, and medical physicist jointly decide on the maximum dose and the dose to the margin.

Stereotactic Radiation Delivery

Once the radiosurgical plan is finalized, it is exported to the control console computer and checked by the physicist. The patient is then placed comfortably on the couch in the GK-C unit, the frame is attached to the APS, and the physicist and one physician of the team perform a position and collision test run. The APS-based treatment allows the administration of a series of multiple shots (called a run) in a sequential automated session. Not all shots (shots with extreme coordinates) can be treated with APS. One may have to use the manual mode by using trunnions to treat the shots or iso-center.

Postoperative Care

All patients receive an intravenous 40-mg dose of methylprednisolone at the conclusion of the procedure. The stereotactic frame is removed after the procedure. Patients are observed for a few hours in the same-day surgical unit and are usually discharged within 24 hours.

◆ Indications for Radiosurgery in Posterior Cranial Fossa Lesions

Vestibular Schwannoma

Vestibular schwannoma are benign, slow-growing tumors that constitute approximately 10% of all primary intracranial tumors. A majority of the vestibular schwannoma grow at a rate <2 mm per year.2 Microsurgery and stereotactic radiosurgery (SRS) are well-established management options that are available to treat vestibular schwannomas. Conservative management is also an option when the tumor size is very small, when the patient states a preference for it, in patients of advanced age, and in cases in which the tumor is in the only hearing ear. The best management option for patients with small to moderate-sized vestibular schwannoma is controversial.3 However, the long-term results of SRS have established it as an important minimally invasive alternative to microsurgery.4-8 Along with long-term tumor control, other goals of GKRS treatment are hearing preservation and preservation of facial nerve function and trigeminal nerve function. Facial nerve preservation rates of approximately 95 to 100% have been reported with SRS.3,6,8,9 Hearing preservation rates of 50 to 75% have been reported with SRS.6,10,11

Radiosurgical Technique for Acoustic Tumors

◆ Preradiosurgical Evaluations

Radiologic evaluation should include high-resolution thin-section MRI scans. Hearing is graded using the Gardner-Robertson modification of the Silverstein and Norell classification,12 and facial nerve function is assessed according to the House-Brackmann grading system.13 Serviceable hearing (classes I and II) is defined as a pure tone average (PTA) or speech reception threshold lower than 50 dB and an speech discrimination score (SDS) better than 50%. Patients need to be counseled about various treatment options including conservative treatment. Occasionally in patients with neurofibromatosis, one may need to do a trial fitting of the frame.

◆ Radiosurgical Technique: Frame Placement

The aim of the frame placement is to bring the lesion as close to the center as possible. This can be achieved with placing the frame as low as possible and shifting it backward. The anterior posts are positioned as low as possible on the supraorbital region to avoid collision of the frontal post or pin with the collimator helmet. Sometimes in muscular individuals, it might be difficult to secure the frame with short posterior posts; in such cases long posts can be used to secure the frame posteriorly. This might minimize the pain, as the pins will travel through less suboccipital muscles.

◆ Dose Planning

The dose plan should be highly conformal especially at the anterior margin of the tumor, where the facial nerve and the cochlear nerve complex generally are located.

◆ Dose Prescription

The accepted marginal dose is 13 Gy. With this dose lower complication rates with good tumor control have been reported.14 Lower marginal doses are prescribed for patients with bilateral acoustic neura (neurofibromatosis type 2
[NF2]) or for patients with contralateral deafness, for whom hearing preservation may be more critical.

- Outcome of Gamma Knife Radiosurgery of Acoustic Schwannoma

In the study by Pollock et al,3 early outcomes were better for patients having SRS compared with surgical resection (level 2 evidence). The need for later tumor resection following SRS is less than 3%.3 In the study by Banerjee et al,15 radiosurgery is less expensive than microsurgical resection. Facial nerve preservation rates of approximately 95 to 100% have been reported with SRS.3,6,8,9 The treatment dose appears to be much more important than the treatment plan quality in the prevention of facial numness or weakness after radiosurgery for vestibular schwannoma.16 Hearing preservation rates of 50 to 75% have been reported with SRS.6,10,11 A high dose of radiation to the inner ear structures could be a cause for hearing loss after radiosurgery.17 In the study by Masager et al,18 the mean dose of radiation to the cochlea was 4.33 Gy (range 1.30 to 10.00 Gy), and they found that there is a significant correlation between the radiation dose received by the cochlea and the audiologic outcome. In another study, a maximum radiotherapy dose to the cochlear nucleus of >10 Gy was associated with hearing deterioration.19 Unlike microsurgery, hearing loss is gradual over 6 to 24 months. The hearing deterioration after radiosurgery could be due to radiation damage to the cochlear apparatus, disruption in the blood supply to the cochlea, adhesion between the tumor and the perineural tissue, a volume change of the tumor located in the internal auditory canal (IAC), and the compression effect on the cochlear nerve.20–22

- Surgery After Gamma Knife Radiosurgery of Acoustic Schwannoma

The need for later tumor resection following SRS is less than 3%.3 There are reports in which tumor removal after failed SRS is difficult due to fibrosis and adhesions.20,23 Others did not find any adhesion between the tumor and facial nerve after SRS.24 Transient swelling following SRS is observed in 62% of cases treated with SRS, and surgical excision needs to be carefully considered because of the natural regression of transient tumor swelling over time.25 Because of the high rates of facial nerve dysfunction following surgery for failed SRS treatment,20,26 subtotal resection of the tumor may facilitate preservation of cranial nerve function.21,25

- Acoustic Schwannoma and Neurofibromatosis Type 2

The local control following SRS of vestibular schwannoma in the context of NF2 varies between 74% and 100%.27–30 In two of the larger series there were no concerns regarding radiosurgery-associated secondary tumors in patients with vestibular schwannoma in the context of NF2.28,30 However, all patients need to be informed about such events.

- Trigeminal Schwannoma

Trigeminal nerve schwannomas account for 0.2 to 1% of all intracranial tumors and 0.8 to 8% of intracranial schwannomas. Schwannomas are rare and slow growing, and surgery is the primary treatment option. In a recent review the rate of trigeminal nerve dysfunction (new/worsened) after lesion removal varies from 13–86%.31 Radiosurgery may be a preferable alternative to surgical removal, especially for small to medium-sized tumors, because of its low morbidity. Tumor recurrence following microsurgery is similar to the tumor progression rate after radiosurgery.32–34 The accepted marginal dose is between 13 and 14 Gy, which is adequate to control trigeminal schwannoma locally. The overall tumor control reported in various studies is 90 to 100%,33,34,36 with a relatively low incidence of new or worsening of symptoms compared with microsurgical approach. The incidence of new-onset cranial neuropathies range between 0 and 30%,33,34,36 and the lack of central enhancement could be a warning sign of cranial neuropathies.36

- Meningioma

Posterior cranial fossa meningiomas constitute approximately 9% of all intracranial meningiomas. The most common surgical sequela is new or worsening cranial nerve deficit. The incidence of facial nerve weakness following posterior fossa meningioma surgery varies from 10.5 to 30%.37 The incidence of cerebrospinal fluid (CSF) leak following posterior fossa meningioma excision varies from 2.5 to 13.6%, and gross total resection varies from 57 to 94%.38–40 Within the past few years, radiosurgery has played an increasingly important role as a minimally invasive therapeutic modality, not only as an adjuvant but also as a primary treatment. The optimal dose for meningiomas is still under debate.41 In recently published studies the marginal dose varied between 12 and 20 Gy.31,42–45 In some of the above studies the 5-year progression-free survival after GKRS for meningiomas varied from 93 to 98%,41,46,47 with a marginal dose of 12 to 13 Gy. In one of the largest study of GKRS for meningiomas by Kollova et al,41 a marginal dose greater than 16 Gy was associated with edema, and with marginal dose of less than 12 Gy there was a higher incidence of an increase in tumor volume after radiosurgery. In older patients, in patients with significant concomitant medical problems or a high-risk tumor location, and in patients who are not willing to undergo an open microsurgical procedure, we would recommend performing radiosurgery as a safe and effective alternative primary treatment modality. Figure 18.4 shows the radiosurgical planning for a petroclival meningioma. The plan needs to be highly conformal toward the brainstem.

- Chordoma and Chondrosarcoma

Seven percent of chondrosarcomas48 originate from the posterior fossa and 35% of chordomas originate from the spheno-occipital area. Surgical excision is the primary modality to treat these lesions. Because of the close proximity to critical neurovascular structures, gross total resection is rarely achieved. In addition, most patients require postoperative radiotherapy. Tzortzidis et al49 reported a 42% recurrence-free survival for primary cases (microsurgery) and 26% recurrence-free survival for reoperated cases at 10-year
follow-up for chordoma. In one of the studies with the longest follow-up period for cranial chondrosarcomas, the authors reported a recurrence-free survival of 32% at 10-year follow-up following microsurgical resection. In one of the largest radiosurgical series, the 5- and 10-year local tumor control rates were 87% and 76%, respectively, in patients with small to medium-sized tumors of less than 20 mL. These authors recommend a marginal dose of 15 Gy to achieve long-term control. When a tumor residual volume is 20 mL or greater, even after maximum debulking efforts, charged-particle radiotherapy is recommended. Charged-particle radiotherapy (proton-beam radiotherapy) can achieve a substantial reduction in the volume of irradiated nontarget brain tissues. The beneficial impact of this radiotherapeutic modality can be attributed to the known Bragg peak effect: protons lose energy at an increasing rate as they slow down in traversed tissue, yielding an enhanced region of energy deposition—the Bragg peak—just before they stop.

**Glomus Tumor**

One of the practical problems in treating the glomus jugulare tumors is that the frame should be fixed as low as possible. The mainstay of treatment is surgical resection or external beam radiotherapy or both. The results of GKRS have encouraged centers to use SRS in the treatment of glomus jugulare tumors. GKRS can be used to treat these lesions primarily or can be used to treat any residual or recurrent lesions. In one study with a median follow-up of 7.2 years, tumor control rate was 94.7%. Pollock et al reported a progression-free survival of 75% at 10 years. Even the short-term results are very encouraging. In recently published reports, tumor control rates of 100% have been reported at a median follow-up of greater than 24 months. Proper selection of the patients is important. A marginal dose of 16 Gy (range 12–18 Gy) is generally well tolerated and effective. Proximity to brainstem and cranial nerves might dictate a reduction in the marginal dose.
Hemangioblastoma

Microsurgical resection offers cure in the majority of cases. Vascularity of the tumors and location in critical areas of the brain are the common reasons why in some patients completion resection is impossible. In multiple lesions, the advantage of the gamma knife is that the lesions can all be treated simultaneously. GKRS may not be an ideal choice for cystic hemangioblastoma, especially with large cysts. The most accepted marginal dose is approximately 18 Gy.

Malignant Glioma

All the studies so far had evaluated the role of GKRS as an adjuvant treatment rather than primary modality. Souhami et al. in their multicentre randomized trial, did not find any beneficial effect in adding SRS to the initial management of glioblastoma multiforme (GBM). Though the results of some studies are encouraging, caution needs to be exercised as the patients treated in the above series were a highly selective group. In the published literature, there was no uniformity in dose selection.

Low-Grade Glioma

Experiences in treating low-grade glioma with radiosurgery are limited. Several small studies have demonstrated early efficacy of radiosurgery in the setting of low-grade glioma. However, prolonged follow-up assessing long-term control in a large series is required.

Brainstem Glioma

For diffuse brainstem glioma, radiosurgery is not appropriate. Typically, such largely dispersed lesions manifest an aggressive biologic course and indicate a most serious prognosis. Neither surgery nor radiotherapy including GKRS is considered fitting. Radiosurgery is one of the options for small focal lesions of the brainstem as a primary treatment or following surgical decompression, but these studies have limitations such as small numbers of patients, no uniform consensus regarding the dose, and short follow-up period. The usual accepted brainstem tolerance dose is 12 Gy.

Medulloblastoma

The evidence for using GKRS in the management of unresectable residual or recurrent tumor comes from very few studies. With the growing experience with radiosurgery, more long-term outcome data may become available in the future to support its role in the management of these tumors.

Metastasis

The regional distribution of the metastasis in the brain is reported to be 80 to 85% in the cerebral hemispheres, 10 to 15% in the cerebellum, and 3 to 5% in the brainstem. According to the review by Mintz et al., two randomized trials demonstrated a significant survival benefit for patients who received surgery plus whole-brain radiation therapy (WBRT) as compared with patients who received WBRT alone, and one randomized trial detected no significant survival difference between the treatment groups.

Surgery followed by WBRT is recommended for patients with good performance status, minimal or no evidence of extracranial disease, and a surgically accessible single brain metastasis amenable to complete excision. The optimal dose and fractionation schedule for WBRT is 3000 cGy in 10 fractions. In a recently published randomized controlled trial (RCT) there was no difference in survival rates between GKRS alone and surgery plus WBRT for a single small brain metastasis.

For a single surgically unresectable metastasis, the results of a radiosurgery boost after WBRT are better than WBRT alone. For one to four brain metastases, in one RCT there was no improvement in survival, with WBRT plus SRS when compared with SRS alone. According to the Radiation Therapy Oncology Group (RTOG) trial 9508, the radiosurgery boost after WBRT for a single unresectable metastasis was better than WBRT alone, and this strategy is to be considered for two or three brain metastases as well.

Two factors that influence the marginal dose are the tumor size and previous radiotherapy. In the literature the prescribed marginal dose varied between 13 and 20 Gy. The most frequently prescribed marginal dose is 16 Gy.

Arteriovenous Malformations

Posterior cranial fossa AVMs, which receive arterial feeders from the verteobasilar system, constitute approximately 10% of all AVMs. Patients presenting with hemorrhage from posterior fossa AVMs have a significant risk of rehemorrhage (8.4–9.4% per year). GKRS is a well-established technique to cure AVMs of the brain. The disadvantages with GKRS are that the results are not instantaneous, unlike with surgical excision. There is always a risk of rebleeding during the waiting period. The risk of rebleeding after radiosurgery varied from 3.4 to 10%. Radiosurgery is appropriate for patients with small AVMs, especially when they are located in eloquent brain locations. The volume of the nidus is a more important factor than the maximum diameter. Treatment planning requires acquisition of thin-section MRI scans and digital subtraction angiograms. In cases where MRI is contraindicated, a CT angiogram can be performed. Conformity should be based on both MRI scans and DSA. The optimal marginal dose frequently prescribed is 25 Gy. A dose reduction may be necessary when the AVM is large or the nidus is not compact and located in the eloquent region of the brain. Radiosurgery can eliminate the risk of hemorrhage in approximately 75% of all AVM patients within 3 years of the procedure and can be repeated after 3 years. Individual obliteration rates vary from 50 to 88% depending on the marginal dose. Even for larger AVMs, staged volume radiosurgery is
Radiosurgical planning for a small AVM in the vermian region is shown in Fig. 18.5.

**Brainstem Arteriovenous Malformations**

Although surgery is advisable for brainstem AVMs reaching the pial surface, there is always a risk of causing neurologic damage in performing such an excision. Deeply located brainstem AVMs are best treated with radiosurgery. The compact nature of brainstem parenchyma, having critical neurologic function, undoubtedly increases both the radiosurgical and microsurgical complication rates. Conformal dose planning and appropriate dose selections are keys to the reduction of adverse radiation effects. Because the optimal dose is determined on the balance between the expected obliteration rates and the corresponding risks of radiation injury, knowledge of tolerable doses is essential in planning radiosurgical treatment.

**Cavernous Malformations**

Asymptomatic cavernous malformations can be treated conservatively. Intervention is required in cases with intractable seizures, recurrent hemorrhages, and hemorrhage causing severe neurologic deficits. For lesions that are located superficially and in noneloquent regions, surgical excision is safe and the risk of rebleeding can be eliminated. The role of radiosurgery in the management of cavernomas is controversial. Radiosurgery of the cavernomas is indicated for poorly accessible lesions such as brainstem cavernomas, where microsurgery is considered risky.

Currently, the marginal dose for cavernous hemangioma in our practice does not exceed 15 Gy, even for small lesions. The theoretical goal of radiosurgery is to obliterate the lesion completely and thereby to prevent subsequent rebleeding. Unlike cerebral AVMs, direct proof of the postradiosurgical obliteration of these lesions is not available. There are diffi-
culties in defining what constitutes a cavernous hemangioma bleeding and evaluating the results of GKRS. GKRS does not remove the intracranial pathologic process completely. The lesion regression and a decrease in volume can be regarded as a positive treatment response. The only way to evaluate the risk of rebleeding is clinical observation during a longer follow-up period. Thus, a longer follow-up period is needed to verify the protective effect of the radiosurgery on cavernous hemangioma. Lesion regression or failure to enlarge is an important parameter in relation to the effect of GKRS on a cavernous hemangioma.

**Trigeminal Neuralgia**

More recently, radiosurgery has emerged as a viable treatment option for trigeminal neuralgia. Patients are often offered medical therapy as the initial line of treatment. However, many patients fail or cannot tolerate medical therapy, and eventually require surgical intervention. Although often associated with initial pain relief, all surgical procedures have variable but definite rates of recurrence and morbidity. The addition of radiosurgery to the treatment armamentarium offers patients another less invasive alternative. Radiosurgery has been advocated as a minimally invasive alternative surgical approach to microvascular decompression (MVD) or percutaneous procedures. A single 4-mm isocenter is used for targeting the trigeminal root entry zone. The target is usually 3- to 6-mm anterior to the pontine edge (Fig. 18.6). The most accepted maximal dose is 80 Gy. Excellent outcomes were achieved within the first several weeks after the procedure in 35 to 74% of patients. The latency interval to pain relief is approximately 1 to 2 months. One of the most significant advantages of radiosurgery is the low risk of new trigeminal dysfunction, compared with other percutaneous surgical techniques. The incidence of this complication after radiosurgery has been reported to be between 0 and 17%. Radiosurgery may be appropriate for a subset of patients who

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**Fig. 18.6** Radiosurgical planning for a trigeminal neuralgia. A single 4-mm isocenter is used for targeting the trigeminal root entry zone. The target is usually 3 to 6 mm anterior to the pontine edge.
are not good surgical candidates. In addition, radiosurgery could be considered a good choice for patients with recurrent pain after failure of MVD or percutaneous surgery. Radiosurgery can be repeated if pain returns after initial relief. At the second procedure the maximum dose is reduced (60–70 Gy) and the nerve is targeted anterior to the prior target.190

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Aneurysms of the Basilar Trunk: Far Lateral and Petrosal Approaches

Anil Nanda, Haim Ezer, and Prasad Vannemreddy

Saccular aneurysms of the basilar artery have remained aloof, because of the presumed limitations of accessibility and fear of the consequences of changes in the basilar circulation. The surgical treatment of these aneurysms will undoubtedly develop in some degree and it would appear that in certain instances in which expectancy of life is poor, as with repeated hemorrhages, a direct attack is justifiable.

—Charles Drake, 1960

In the latter half of the 18th century, anatomist Cruveilhier described aneurysms of the vertebrobasilar system in his illustrations, and an angiographic demonstration of such aneurysms was first performed by Krayenbuhl. Though Dandy described his experiences with vertebrobasilar aneurysms, it was Schwartz who was credited with the first-ever preoperative diagnosis and direct surgical treatment of basilar artery aneurysm using a standard suboccipital craniectomy. Methods have included ligation of the neck of the aneurysm with or without excision of the aneurysm, trapping by ligation of arterial supply to the aneurysm, reinforcement of the aneurysmal wall, or ligation of the proximal vertebral artery in its intracranial or extracranial position. Standard subtemporal and suboccipital approaches have been used for many years. In 1959, Logue described the frontotemporal approach to basilar artery aneurysms. Drake compiled the experiences of neurosurgeons of the earlier years, and added his personal series of four cases in 1960. In this compiled series with 30 cases, there were only four deaths! Utilizing anatomic dissections, Drake exposed the upper third of the basilar artery and its bifurcation by an anterior subtemporal approach through the tentorial opening into the interpeduncular cistern. Brief periods of circulatory arrest helped to complete the dissection of the aneurysm neck. He also practiced division of the posterior communicating artery in some cases to improve access; however, not in all cases, as proposed by Gillingham. Kenneth Jamieson reported another series of 19 cases of vertebrobasilar aneurysms in 1964 with a mortality of 10 out of the 19. Taking their cue from Mullan et al. and Stevenson et al., who approached anterior brainstem lesions via the transclival route, John Fox and Sano et al. simultaneously described basilar craniectomy and transphenoidal-transclival approaches for obliteration of a midline vertebral artery aneurysm. The technical difficulties of a narrow operating space combined with a high risk of infection made the approach less popular. Meanwhile, a large number of patients were operated upon by Drake utilizing the subtemporal route, and later Yasargil et al. added the microsurgical pterional/transsylvian approach to reach the basilar tip. Access to a midbasilar or low-lying basilar bifurcation, however, remained difficult, and Kasdon and Stein in 1979 proposed a combined subtemporal and suboccipital approach. Solomon and Stein reported their experience with this combined approach to reach low basilar and vertebral artery aneurysms with results superior to the natural history. The sigmoid sinus was divided inferior to the vein of Labbé, enabling retraction of the temporal lobe, whereas the cerebellum was lifted medially and superiorly to visualize the lateral side of brainstem below the petrous apex. This combined approach facilitated visualization of cranial nerves III through XI and the lateral side of the cerebral peduncle and pons. Kawase et al. reduced retraction damage to the temporal lobe by advocating the extradural transpetrosal technique. However, this corridor remains constricted between the trigeminal nerve and the cochlea and has a potentially high risk of hearing loss and cerebrospinal fluid otorrhea. During the same period, Giannotta and Maceri reported performing a retrolabyrinthine transsphenoidal cranietomy to reach aneurysms arising from the basilar artery trunk. The incision extended from the asterion to below the rim of the foramen magnum, thus reducing the more extensive incisions of the combined approach. The air cells are removed from an area...
bordered superiorly by the floor of the middle fossa and the superior petrosal sinus, inferiorly by the jugular bulb, and anteriorly by the posterior semicircular canal (SCC). Bone overlying the sigmoid sinus was removed, thus exposing the posterior fossa dura anterior and posterior to the sigmoid sinus. The sinus was later divided. The positive aspect of this approach is the familiarity of most surgeons with the suboccipital exposure of posterior fossa. For a shallower posterior approach to the clival region, Sekhar and Estonillo\textsuperscript{15} proposed a combined transcoclear and infratemporal approach to reach basilar trunk. Several modifications to the standard suboccipital route followed, and more so to the traditional approaches to cerebellopontine angle tumors in the form of transcoclear, translabyrinthine, transotic, and transmastoid extensions. Miller et al\textsuperscript{16} introduced a common nomenclature for all of these modifications: anterior and posterior transpetrosal approaches that would lead to lesions at the brainstem or clivus or the basilar trunk. The conventional suboccipital approach also received some extensions as Heros in 1986 described a lateral suboccipital approach for vertebral and vertebrobasilar artery lesions. Further modifications by the extent of excision of the occipital condyle resulted in far-lateral and extreme lateral approaches.\textsuperscript{17,18}

\begin{itemize}
\item **Transpetrosal Approaches**
\end{itemize}

An imaginary line drawn between the two internal auditory meatus would make a plane for two different topographic groups of lesions: anterior plus superior and posterior plus inferior. The anterosuperior would be lesions of the petrous apex and the superior half of the clivus that can be accessed by an anterior/temporal petrosal approach, and the postero-inferior would be lesions of the cerebellopontine angle and the petroclival and jugular fossa, which are approachable via a posterior transpetrosal route. The anterior approach could be combined with a temporal approach, whereas the posterior approach could be combined with a lateral suboccipital approach to improve visibility and access to the midline lesions.

\begin{itemize}
\item **Anterior Petrosal Approach**
\end{itemize}

This approach is performed with the patient in the supine position with shoulder elevation and the head kept in a horizontal plane. As a routine, a lumbar spinal puncture and cerebrospinal fluid (CSF) drainage facilitate brain relaxation and decrease the chances of a postoperative CSF fistula.

An anterior petrosectomy is performed through a subtemporal craniotomy, with a zygomatic osteotomy utilizing a Falcorner’s flap preserving the superficial temporal artery. Wide exposure toward the midline is not necessary, whereas a wide basal tangent helps in manipulating instruments (Fig. 19.1).

An extradural dissection (described by Kawase in detail) exposes the middle meningeal artery, lesser superficial petrosal nerve, and greater superficial petrosal nerve (GSPN) (Fig. 19.2). All these structures are divided by some surgeons, to further the dissection and avoid traction on the facial nerve. Drilling of the GSPN with diamond bur reveals the internal carotid artery in the Glasscock triangle. The geniculate ganglion can be identified by tracing the GSPN posteriorly or by opening the epitympanum, which lies posterolateral to the geniculate ganglion.

The arcuate eminence is a landmark for the superior SCC, which lies perpendicular to the petrosal ridge. The flat area of bone, between the geniculate ganglion and the arcuate eminence, which is the important landmark in the exposure.
eminence, forms the meatal plane, overlying the internal auditory canal (IAC). Initially, this soft bone is drilled to reach the hard conch of bone overlying the bony SCC inside the arcuate eminence. This drilling exposes the IAC anterior to the SCC and is continued anteriorly to open the internal auditory meatus to expose the posterior fossa dura (Fig. 19.3). At the opening of the IAC, a vertical bony spicule called Bill’s bar separates the superior vestibular and facial nerves. Bill’s bar is an important anatomic landmark to identify the cochlea, which is separated by the facial nerve from the ampulla of the SCC.

The cochlea represents the posterolateral limit of the bony exposure through Kawase’s triangle and need not be exposed (Fig. 19.4). The hard otic capsule encasing the cochlea is a compact bone and looks distinctly lighter than the surrounding bone of the petrous apex. This remaining bone at the petrous apex bounded by the carotid artery, cochlea, and SCC/IAC constitutes the Kawase’s triangle and can be removed until the inferior petrosal sinus is exposed along the posterior fossa dura (Fig. 19.5).

The surgeon must keep in mind that extensive drilling along the petrous bone does not increase the visibility or exposure of the operative field and only increases the risk of injury to the cochlea, vestibule, internal carotid artery, and facial nerve, with subsequent neurologic deficits as well as CSF fistula and postoperative infection.

The middle fossa dura is usually opened along the temporal lobe. The tentorium is opened posterior to the trochlear nerve and lateral to the superior petrosal sinus. A perpendicular incision continues from here into the posterior fossa inferiorly.

**Posterior Petrosal Approach**

The patient is placed in the lateral position with the head rotated horizontally to reduce venous obstruction at the neck. Placement of a lumbar drain precedes the positioning of the patient. The basic technique utilizes the mastoidectomy approach along the periauricular region with a sinuous curvilinear incision that can be extended superiorly, inferiorly, or in an inverted U-shaped incision (Fig. 19.6). A standard craniotomy with four bur holes located over the asterion, both sides of the superior nuchal line, and at the junction of the inferior temporal line and pariøtomastoid suture is performed followed by a mastoidectomy (Fig. 19.7). All the cortical bone overlying the mastoid is removed superiorly to the supramastoid crest, inferiorly to the tip of mastoid process, and anteriorly to the posterior wall of the external auditory canal, thus delineating the compact sigmoid plate overlying the sigmoid sinus and sinodural angle (Fig. 19.8). The mastoid antrum is identified following the middle fossa plate and external auditory canal posterior wall. Medially, the cortical floor of antrum is formed by the lateral semicircular canal (LSCC) and requires preservation. The inferior surface of the LSCC is close to the external genu of the facial nerve and does not need to be exposed. Removal of the mastoid air cells inferiorly would expose the digastric ridge, which would lead anteriorly to the facial nerve at the stylomastoid foramen and extracranially. Removal of air cells in the inferior part of the mastoid and retrofacial air cells between the antrum and digastric ridge exposes the fallopian canal (encasing the facial nerve). The posterior SCC runs parallel to the posterior fossa plate of bone and is bisected by the LSCC. It is located between the LSCC and the posterior fossa plate.

The sigmoid plate is followed inferiorly through the infralabyrinthine air cells to expose the jugular bulb. Only the beginning of the jugular bulb is exposed during the posterior petrosectomy, and exposure of the jugular bulb can be improved by skeletonization of the facial nerve in the fallopian canal. The superior SCC is perpendicular to the LSCC and can be exposed by following the sinodural angle through the supralabyrinthine air cells and through the dense cortical bone located along the petrous ridge at the juncture of the posterior...
and middle fossa bone plates (Fig. 19.9). In a combined approach, this step can be facilitated by removing the middle fossa bone flap and identifying the arcuate eminence from a subtemporal trajectory (vide supra).

This area bounded by the posterior fossa dura, middle fossa dura, and posterior canal wall forms the Trautmann’s triangle, which actually represents the area of bone removed by the posterior petrosectomy approach (Fig. 19.10). The

**Fig. 19.5** Petrosal approaches. Intradural dissection in a cadaver, showing the drilled labyrinth and the petrous apex. The dotted area represents Glasscock’s triangle bounded by the greater superficial petrosal nerve (GSPN), the V3 nerve, and a line from the foramen spinosum to the facial hiatus. The thick blue arrowheads represent the extent of bone removed. The empty triangle represents Kawase’s triangle. C, internal carotid artery; CP, chorda (tympany) posterior; Ext., external; GG, geniculate ganglion; GAS, G., Gasserian ganglion; I, incus; IAM, internal acoustic meatus; mma, middle meningeal artery; Mal., malleus; Mid. F., middle foramen; Post. fossa, posterior fossa; S, stapes; Styl. F., stylomastoid foramen.

**Fig. 19.6** A periauricular incision can be fashioned in a curvilinear or inverted-U shape, basically to meet the mastoidectomy requirement to reach the petrous apex along the internal auditory canal.

**Fig. 19.7** The area of the mastoidectomy is marked on the surface of the bone.
The plates of bone overlying the middle fossa, posterior fossa, and sigmoid sinus are left behind until the end of the procedure to protect the dura and venous sinuses and guide the dissection. Elevating these bone plates completes petrosectomy. The medial portion of the petrous ridge remains deep to the superior SCC and posterior SCC, which can be drilled off as required, carrying the dissection across the IAC into the petrous apex.

The dural incision is made along the jugular bulb to the superior petrosal sinus, sparing the endolymphatic sac, and then along the posterior temporal lobe and anteriorly along the inferior temporal lobe. After dividing the superior petrosal sinus, the tentorium is cut posterior to the trochlear nerve (Fig. 19.11). Now the temporal lobe can be gently retracted along with the cut edge of the tentorium, avoiding injury to the cortex. The bulky nervous structure seen at this point is the trigeminal nerve, which is exposed while it enters Meckel’s cave (Fig. 19.12). Illustrative case angiograms are shown in Fig. 19.13.

An anterior petrosectomy provides access to the posterior fossa between the internal carotid artery, trigeminal root, and facial nerve. Lesions posterior to the IAC are better approached...
through a posterior petrosectomy or suboccipital craniotomy with preserved hearing. A posterior petrosectomy, in combination with an occipital-subtemporal craniotomy, opens up the entire posterior face of the petrous bone, the upper two thirds of the clivus, the anterior cerebellum, and the brainstem. Inferiorly, the anterior lip of the foramen magnum can be visualized. This lowermost exposure can be obtained by combining the posterior petrosectomy and the far lateral suboccipital approach.

◆ Far Lateral Suboccipital Approach

Drake and Peerless used either the subtemporal transtentorial or the suboccipital approach for aneurysms of the vertebrobasilar (VB) system and enjoyed great success. The lateral suboccipital approach has been in vogue for vertebral artery and VB junction aneurysms. Conventional occipital bone removal supplemented by excision of the posterior arch of the atlas up to the sulcus arteriosus laterally provides adequate exposure for most lesions at the foramen magnum.

Fig. 19.12 Gentle retraction of the temporal lobe and the tacked-up tentorium now exposed the bulky fifth nerve entering Meckel's cave. Dissecting the nerve along with the arachnoid and gentle retraction expose the basilar trunk, along with the anterior inferior cerebellar artery (AICA).

Fig. 19.13 (A–D) Preoperative and intraoperative angiograms showing successful obliteration of the basilar trunk aneurysms.
whereas the vertebral artery, for the most part, is identified early in the operative procedure.

In his modest report, Heros\textsuperscript{22} described a gradual evolution of lateral suboccipital approach, later renamed and modified by many, as the far lateral, transcondylar suboccipital, and extreme lateral suboccipital approach with removal of the occipital condyle to a variable extent.\textsuperscript{17,18,22–25}

The far lateral suboccipital craniotomy is performed in the lateral position. The patient is positioned under general anesthesia after placement of a lumbar drain. The incision is modified to a C-shaped retroauricular incision, one situated 2 cm medial to the mastoid, and reaching up to the C2 spinous process in the midline. This incision facilitates exposure of the mastoid and foramen magnum and the arch of C1 below. The muscles are divided with electrocautery along the line of the skin incision down to the suboccipital bone, where a subperiosteal dissection is performed. As the flap is reflected inferiorly, a sharp dissection is preferred to avoid injury to the vertebral artery at the foramen magnum (Fig. 19.14). The posterior arch of the atlas is exposed by continued subperiosteal dissection. A suboccipital craniotomy is performed after a single bur hole is placed over the occipital bone (to be replaced at the end of the procedure). Bone removal is now continued after excision of the posterior arch of the atlas toward the sulcus arteriosus. Bleeding from the robust venous plexus in this region can be avoided by placing cotton pledgets and bipolar dissection along the periosteum. Preferably now, the vertebral artery is identified with finger palpation along the lateral end of the foramen magnum. Thus, the most important aspect of this approach is adequate removal of bone in the area of the foramen magnum going laterally as far as the condylar fossa, posterior to the occipital condyle in close proximity to the entry of the vertebral artery into the dura (Fig. 19.15). At this lateralmost part of the foramen magnum, troublesome venous bleeding may be encountered, which can be controlled with bipolar coagulation and packing with oxidized cellulose. At the lateral extent of the exposure, the bone ridge becomes more vertical, and a high-speed drill is used to remove the last few millimeters of bone along the foramen magnum. This key point of the drilling enables exposure of anterior aspects of the medulla and upper cervical spinal cord from a lateral view without any retraction. As described by Heros, this lateral rim of the foramen magnum represents to the suboccipital exposure what the pterion does to the frontotemporal approach.

The dura is opened with an oblique incision toward the midline at C1 and taking a cuff around the vertebral artery. The cuff of dura around the vertebral artery is essential for a watertight closure of the dura. The lateral flap of dura is tented up with sutures secured to the lateral muscle mass. Opening the cisterna magna and dividing the denticulate ligament assist in relaxation and gravity-aided retraction of the brainstem. Gentle retraction on the cerebellar tonsils, in the early phases, is required to visualize more medial structures. As the CSF drainage continues, the retractor may be removed soon, to reduce retraction injury of the cranial nerves and brainstem.

The vertebrobasilar junction can be approached inferolaterally, between the lower cranial nerves while tracing the vertebral artery superiorly. For a higher location, a working space can be safely created between the lower cranial nerves and the cranial nerve VII-VIII complex (Figs. 19.16 and 19.17). Case illustrations of the pre- and post-clipping angiograms can be seen in Fig. 19.18.

An extreme lateral transcondylar approach was described by Sen and Sekhar\textsuperscript{27} in 1990 for tumors in the region of the foramen magnum and later advocated for aneurysms of the vertebrobasilar junction by Sekhar et al\textsuperscript{26} in 1994. The authors recommended extensive excision of the occipital condyle and in some instances of the jugular tubercle to enhance visibility. In 2002, we found in our cadaver study, as applied to a clinical series, that more extensive bone removal of the occipital condyle did not significantly improve visibility.\textsuperscript{18} It has been the experience of many authors that the nature,
extent, and location of the lesion determines the resectability rather than the extent of the condylar excision.\textsuperscript{27–30}

It is very appropriate to recall the philosophical statement by Robert Smith\textsuperscript{31} while performing cranial base surgery:

\begin{quote}
Only so much retraction of these structures (brainstem, cranial nerves and brain) is permissible. It is not the bony opening that limits an unobstructed view. Much of our current enthusiasm for cranial base approaches is based on what we can get away with, rather than what is needed, this too will pass.
\end{quote}

\begin{figure}[h]
\centering
\includegraphics[width=\textwidth]{image1}
\caption{The far lateral approach and intradural anatomy showing the lower cranial nerves (CN) and vertebral artery (VA).}
\end{figure}

\begin{figure}[h]
\centering
\includegraphics[width=\textwidth]{image2}
\caption{The far lateral approach. An intraoperative photograph shows the vertebral artery (VA) and lower cranial nerves (CN) in a case of a posterior inferior cerebellar artery (PICA) aneurysm. LD, ligamentum denticulatum.}
\end{figure}

\begin{figure}[h]
\centering
\includegraphics[width=\textwidth]{image3}
\caption{Preoperative (A) and intraoperative (B) pictures of a successful obliteration of a PICA aneurysm.}
\end{figure}
Fig. 19.18 (Continued)

References

Aneurysms of the basilar artery apex carry the highest natural history risk of rupture of all aneurysm locations. In the International Study of Unruptured Intracranial Aneurysms Update, the basilar apex location was the strongest predictor of hemorrhage (relative risk 2.3; 95% confidence interval 1.1–4.8; \( p = .025 \)). In that same study, the posterior circulation location was a significant predictor of a worse clinical outcome after surgical and endovascular treatment (for surgery: relative risk 1.6; 95% confidence interval 1.1–2.4; \( p = .025 \); for endovascular treatment: relative risk 2.25; 95% confidence interval 1.1–4.4; \( p = .02 \)). Others have also described the challenging nature of basilar tip aneurysm surgery. Thus, the cerebrovascular surgeon is faced with an aneurysm that has the highest risk of hemorrhage of all aneurysm locations if left untreated, but also the highest risk of morbidity associated with treatment.

◆ Combined Neurovascular Team Approach

At the Massachusetts General Hospital, we have adopted a combined neurovascular team approach of surgical and endovascular treatment to deal with the challenges of these lesions—a treatment philosophy that we have previously described. The cerebrovascular surgeons and the neurointerventionists on the team discuss each patient and determine jointly the treatment strategy on a case-by-case basis. The Massachusetts General Hospital (MGH) grade is a comprehensive aneurysm grading system, previously described, that accurately predicts the risks of surgical aneurysm treatment based on the Hunt-Hess grade (1 point for Hunt-Hess grade 4–5), the Fisher Scale score (1 point for Fisher Scale score 3–4), patient age (1 point for age >50), aneurysm size (1 point for size >10 mm), and if the aneurysm is a giant posterior circulation lesion (1 point for posterior circulation aneurysm \( \geq 25 \) mm). Good MGH grade patients tend to be treated with surgery, because of the higher rate of complete and durable occlusion, and poor MGH grade patients tend to be treated endovascularly.

Factors that are also important to treatment decision making are aneurysm size, neck size, aneurysm morphology, direction of the aneurysm, and aneurysm location in relation to the clivus (Table 20.1). Giant aneurysms (>25 mm) of the posterior circulation have been previously demonstrated by our group to be a higher surgical risk; therefore, they tend to be treated by our group endovascularly rather than surgically. Wide neck sizes (>4 mm) tend to be unfavorable for endovascular treatment; therefore, our group has preferred surgical reconstruction for wide-necked aneurysms or aneurysms with an unfavorable dome-to-neck ratio. Similarly, aneurysm morphology is important as endovascular therapy is less favorable for aneurysms that incorporate either or both P1 arteries, in which case surgical reconstruction is performed. Drake defined the importance of aneurysm direction, and described forward-projecting aneurysm domes as tending to be free of perforators, whereas backward sacs are at risk of the clip blades closing on the perforators on the opposite side. Thus, we have tended to treat backward-projecting aneurysms endovascularly. Finally, we have previously described a classification system of basilar artery aneurysms based on the location of the aneurysm in relation to the clivus and the posterior clinoid. Aneurysms that are below the posterior clinoid and are low in relation to the top of the clivus are more challenging to approach surgically, and thus we have tended to treat these aneurysms endovascularly.

◆ Surgical Approaches

We have previously described surgical approaches to basilar artery aneurysms using a classification system based on the
### Table 20.1  Factors for Deciding on Surgical or Endovascular Treatment for Basilar Apex Aneurysms

<table>
<thead>
<tr>
<th>Factor</th>
<th>Treatment Modality</th>
</tr>
</thead>
<tbody>
<tr>
<td>MGH grade</td>
<td>Surgery &gt; endovascular</td>
</tr>
<tr>
<td>Good MGH grade (0–2)</td>
<td>Surgery &gt; endovascular</td>
</tr>
<tr>
<td>Poor MGH grade (3–5)</td>
<td>Endovascular</td>
</tr>
<tr>
<td>Aneurysm size</td>
<td></td>
</tr>
<tr>
<td>&lt;15 mm</td>
<td>Surgery or endovascular</td>
</tr>
<tr>
<td>≥15 mm</td>
<td>Endovascular</td>
</tr>
<tr>
<td>Aneurysm neck size</td>
<td></td>
</tr>
<tr>
<td>&lt;4 mm</td>
<td>Surgery or endovascular</td>
</tr>
<tr>
<td>≥4 mm</td>
<td>Surgery</td>
</tr>
<tr>
<td>Aneurysm morphology</td>
<td></td>
</tr>
<tr>
<td>Distinct from P1(s)</td>
<td>Surgery or endovascular</td>
</tr>
<tr>
<td>Incorporates P1(s)</td>
<td>Surgery</td>
</tr>
<tr>
<td>Aneurysm direction</td>
<td></td>
</tr>
<tr>
<td>Projects forward</td>
<td>Surgery or endovascular</td>
</tr>
<tr>
<td>Projects backward</td>
<td>Endovascular</td>
</tr>
<tr>
<td>Aneurysm location to clivus</td>
<td></td>
</tr>
<tr>
<td>High supraclival</td>
<td>Surgery or endovascular</td>
</tr>
<tr>
<td>Supraclival</td>
<td>Surgery or endovascular</td>
</tr>
<tr>
<td>Upper clival</td>
<td>Surgery or endovascular</td>
</tr>
<tr>
<td>Middle or low clival</td>
<td>Endovascular</td>
</tr>
</tbody>
</table>

P1, first segment of the posterior cerebral artery (PCA).

### Table 20.2  Surgical Approaches to Basilar Apex Aneurysms Based on the Clival Location

<table>
<thead>
<tr>
<th>Clival Location</th>
<th>Surgical Approach</th>
</tr>
</thead>
<tbody>
<tr>
<td>High supraclival</td>
<td>Pterional</td>
</tr>
<tr>
<td></td>
<td>Pterional with orbitozygomatic resection</td>
</tr>
<tr>
<td>Supraclival</td>
<td>Pterional</td>
</tr>
<tr>
<td></td>
<td>Subtemporal</td>
</tr>
<tr>
<td></td>
<td>Combined “half-and-half”</td>
</tr>
<tr>
<td>Upper clival</td>
<td>Subtemporal</td>
</tr>
<tr>
<td></td>
<td>Subtemporal transtentorial</td>
</tr>
</tbody>
</table>

The location of the aneurysm in relation to the clivus. High supraclival aneurysms are approached via a pterional or pterional with orbitozygomatic resection approach, supraclival aneurysms via a pterional or subtemporal or combined “half-and-half” approach, and upper clival aneurysms via a subtemporal transtentorial approach (Table 20.2).

The pterional approach was popularized by Yasargil et al. and offers the advantage of less brain retraction than in the subtemporal approach. However, with the pterional approach, the line of sight is oblique, the working distance is longer, and low-lying lesions are obscured by the posterior clinoid process. The surgeon must work in a deep and narrow field in the opticocarotid triangle, or in a small space just lateral to the internal carotid artery. Adding orbitozygomatic resection to the pterional approach provides a view from the inferior direction with less retraction. We use a standard pterional approach used by many cerebrovascular surgeons. The patient is positioned supine with the head turned. We have preferred to use a lumbar cerebrospinal fluid drainage catheter. A standard pterional skin incision starts at the level of the zygoma just in front of the tragus and curves gently to the midline behind the hairline. The temporalis muscle is divided and flapped forward. A standard pterional craniotomy is performed, with further bony removal of the lateral sphenoid wing. The dura is opened in a curvilinear manner. The brain is relaxed by the administration of mannitol and by removal of cerebrospinal fluid from the lumbar drain. Further relaxation is achieved by suctioning of cerebrospinal fluid from above the optic nerve. The frontal lobe can then be gently and gradually retracted. The microscope is brought into view and the medial cisterns are then opened by arachnoid dissection. Two corridors for working are opened: the opticocarotid triangle, and a small space just lateral to the internal carotid artery. The membrane of Liliequist is opened, allowing access to the interpeduncular cistern. Isolation of the basilar artery is necessary for proximal vessel control. After temporary clip occlusion, dissection around the aneurysm can be performed, with utmost attention paid to brainstem and thalamic perforators. Final clip placement is performed with inspection of all sides of the aneurysm to ensure no perforating vessels are injured.

Drake popularized the subtemporal approach. The advantages are a short working distance and a perpendicular view of the aneurysm neck, but a disadvantage is the need for deep temporal retraction, particularly for high aneurysms. The approach is best for small basilar apex aneurysms or ones that project posteriorly. Our technique has been described previously. The patient is placed in the lateral position with the head elevated and the vertex dipped slightly to allow the temporal lobe to fall away from the floor of the middle fossa. A lumbar cerebrospinal fluid drainage catheter is used. We use a modified pterional type of skin incision, separate the temporalis muscle, and make a wide-based craniofacial osteotomy with additional bone removed to maximize exposure of the floor of the middle fossa (Fig. 20.1). The dura is opened in a T- or I-fashion with the vertical limb extended down to the skull base. The temporal lobe is gently and gradually elevated and retracted, which is aided by temporal lobe relaxation from the administration of mannitol and removal of cerebrospinal fluid from the lumbar drain. The microscope is brought into view. The third cranial nerve is identified, and the approach to the aneurysm can then follow along a corridor inferior to the third nerve; however, we have found it helpful also to expose a corridor superior to the third nerve and use both corridors. The edge of the tentorium can be retracted laterally by tacking it up with sutures between the third and fourth cranial nerves. A wide arachnoid opening is performed between the third nerve and the superior cerebellar artery. Working along these corridors, dissection is...
performed to identify the basilar artery for proximal vessel control. Temporary clip occlusion of the basilar artery is performed before dissecting around the aneurysm. The utmost attention is paid to brainstem and thalamic perforators, with a view on all sides of the aneurysm necessary before and after final clip placement to avoid potential injury to these vessels.

The combined “half-and-half” approach had been first described by Drake⁹ and by Sano,¹⁰ who called it the “temporo-polar approach,” but it is used by many cerebrovascular surgeons for basilar tip aneurysms. We use the technique described by Heros et al.⁶,¹¹ The patient is positioned supine with the head turned to the opposite side. A lumbar cerebrospinal fluid drainage catheter is used. We use a modified pterional-type skin incision down to the zygoma, which facilitates a larger craniotomy (Fig. 20.2). The temporalis muscle is divided and retracted posteriorly, leaving a cuff of temporalis fascia attached to the periosteum to suture to at closure. A large wide-based craniotomy is made, and bone is removed down to the floor of the middle fossa, and the posterior pterion and greater sphenoid wing are removed to completely expose the dura over the anterior aspect of the temporal pole. The dura is opened. The pole of the temporal lobe is then gently and gradually retracted posteriorly, which is facilitated by temporal lobe relaxation by the administration of mannitol and removal of cerebrospinal fluid from the lumbar drain. The microscope is brought into view, at which point, the medial cisterns and sylvian fissure are opened by arachnoid dissection. The third cranial nerve is then identified, and two corridors are opened through which to work,
medial and lateral to the nerve. The membrane of Liliequist is opened, providing access to the interpeduncular cistern. The basilar artery is identified for proximal vessel control. After temporary clip occlusion, dissection is performed around the aneurysm, and then final clip placement is performed.

The subtemporal transtentorial approach provides exposure for the upper clival laterally positioned basilar tip aneurysms. The technique we use has been described previously. The patient is placed in the full lateral position. A lumbar cerebrospinal fluid drainage catheter is used. The skin incision is in a horseshoe shape from the zygoma arching above the ear and down approximately 2 cm behind the mastoid, with the allowance for extending the incision 3 cm if a suboccipital craniectomy is later needed. A full wide temporal craniotomy is performed with additional bony removal as needed to expose the floor of the middle fossa. Openings into the mastoid air cells are waxed to prevent cerebrospinal fluid leakage. The dura is opened with an inferiorly based flap.

Fig. 20.1 (continued) (D) Tentorial retraction and arachnoid opening. (E) Initial dissection of basilar artery. PICA, posterior inferior cerebellar artery. (From Ogilvy CS, Crowell RM, Heros RC. Basilar and posterior cerebral artery aneurysms. In: Ojemann RG, Ogilvy CS, Crowell RM, Heros RC, eds. Surgical Management of Neurovascular Disease, 3rd ed. Baltimore: Williams & Wilkins. 1995, reprinted by permission.)
Fig. 20.2 Combined "half-and-half" approach. (A) Head position and skin incision. The dotted line indicates the level of the zygomatic arch. (B) The dura has been opened with a curvilinear inferior incision. A, retractor under the frontal lobe; B, retractor over the anterior temporal tip; C, inferior dural flap over the orbital roof. (C) Microsurgical exposure. The arachnoid has been opened widely and the sylvian fissure has been opened. A, right optic nerve; B, left optic nerve; C, left A1 artery; D, right A1 artery; E, right internal carotid artery; F, right middle cerebral artery; G, right posterior communicating artery; H, right posterior cerebral artery (P2); J, right ocular motor nerve; K, arachnoid over giant basilar artery; L, tentorial edge retracted with a suture. (From Ogilvy CS, Crowell RM, Heros RC. Basilar and posterior cerebral artery aneurysms. In: Ojemann RG, Ogilvy CS, Crowell RM, Heros RC, eds. Surgical Management of Neurovascular Disease, 3rd ed. Baltimore: Williams & Wilkins, 1995.)
The temporal lobe, which has been relaxed by the administration of mannitol and removal of cerebrospinal fluid from the lumbar drain, is gently and gradually elevated upward, as in the basic subtemporal approach; however, in this case, a more posterior edge of the tentorium is exposed. The microscope is brought into view and the fourth cranial nerve is identified. The tentorium is cut just behind the fourth cranial nerve and is then sectioned approximately 2 mm off the superior petrosal sinus and extending 3 to 4 cm posterolaterally, sparing the lateral sinus. This triangular flap is then excised, providing an excellent view of the clivus. Gentle retraction of the cerebellum exposes the cerebellopontine angle. The resulting view, which is best seen by angling the microscope from the posterior direction to see around the posterior clinoid process, is wide enough for temporary clip placement, aneurysm dissection, and final clipping (Fig. 20.3). In rare cases, a suboccipital craniectomy may be needed to further the exposure. The incision is extended 3 cm inferiorly, and a suboccipital craniectomy is performed over the sigmoid–transverse sinus junction. The tentorium is completely sectioned, including lateral sinus ligation (as long as the contralateral lateral sinus is patent), allowing for elevation of the entire tempo-occipital hemisphere, which produces maximal view onto the clivus.

◆ Results

From 1990 to 1998, the combined neurovascular team at the Massachusetts General Hospital treated 100 basilar tip aneurysms. Treatment was clipping in 72 patients and coiling in 28 patients. As mentioned above and in Table 20.1, our treatment strategy has been to treat good MGH grade patients with clipping and poor MGH grade patients with coiling. Aneurysm size, neck size, morphology, direction, and location in relation to the clivus also influence the selection of the treatment modality (Table 20.1). An angiographically complete occlusion was achieved with clipping in 60 of 64 patients with postoperative angiography (94%) (eight patients did not undergo postoperative angiography). Angiographic efficacy with coiling was 100% in eight patients (29%), ≥95% in eight patients (29%), and <95% in 12 patients (43%). Clinical outcomes as assessed by a slight modification of the Glasgow Outcome Scale (GOS) in the surgical group were excellent (GOS 5) in 46 patients (64%), good (GOS 4) in 14 patients (19%), fair (GOS 3) in six patients (8%), poor (GOS 2) in three patients (4%), and death (GOS 1) in three patients (4%). Clinical outcomes in the coiling group were excellent (GOS 5) in 17 patients (61%), good (GOS 4) in four patients (14%), fair (GOS 3) in four patients (14%), poor (GOS 2) in no patients, and death (GOS 1) in three patients (11%). There were no long-term posttreatment hemorrhages in the surgical group, but one coiled patient had a fatal posttreatment hemorrhage 5 years after initial coiling (the patients underwent a total of three coiling procedures for aneurysm recurrences).

◆ Conclusion

Basilar tip aneurysms have the worst natural history risk of all intracranial aneurysms and are associated with the
highest treatment-related morbidity. They are challenging aneurysms for the cerebrovascular surgeon and the neuro-interventionist. We have found that a combined neurovascular team approach offers the maximal chance for achieving the best clinical outcome and angiographic result while minimizing the risks of treatment-related morbidity.

References

Endovascular Treatment of Posterior Circulation Aneurysms


Aneurysms of the posterior circulation account for roughly 20% of all intracranial aneurysms, with the majority occurring at the basilar apex followed by the origin of the posterior inferior cerebellar artery (PICA). Less common are aneurysms of the posterior cerebral arteries (PCAs), the anterior inferior cerebellar arteries (AICAs), the superior cerebellar arteries (SCAs), the verteobasilar junction, and the basilar artery (BA) trunk. The deep location of these aneurysms and intimate relationships to critical neurovascular structures explain the relatively high morbidity and mortality related to their surgical treatment. Over the past 15 years, endovascular techniques have evolved from an unproven surgical alternative to the treatment of choice for posterior circulation aneurysms at many centers. As we shall discuss in this chapter, endovascular treatment of aneurysms is no longer limited to simple “coiling” of an aneurysm. New techniques have expanded the role of endovascular treatment for intracranial aneurysms and have established endovascular therapy as the first-line treatment for many posterior circulation lesions. That this bias is not simply unique to our institution but rather a disseminated concept is evident by the recent International Subarachnoid Aneurysm Trial (ISAT), in which only 2.7% of the enrolled cases involved aneurysms of the posterior circulation.

Selection of Patients

As mentioned above, the location of an aneurysm in the posterior fossa by itself presents favorably for endovascular therapy over surgery. At our institution, we consider coil embolization as first-line therapy for most posterior circulation aneurysms that warrant treatment. Surgery is considered only if all current endovascular techniques are exhausted and unsuccessful. This approach is much different than our approach to anterior circulation aneurysms, where we consider initially both treatment options. Clinical as well as anatomic factors must be considered in the selection of posterior circulation aneurysms for endovascular therapy. Advanced patient age, significant medical comorbidities, poor clinical grade of the ruptured lesions, and associated vasospasm also weigh favorably toward endovascular therapy.

Fundamentals of Endovascular Technique

A complete review of endovascular technique is beyond the scope of this chapter. This section outlines the major steps of coil embolization of aneurysms as performed at our institution. Direct coil embolization without the use of any supporting devices such as balloons or stents can be accomplished in the vast majority of intracranial aneurysms. The ideal aneurysm morphology for coiling is a dome-to-neck ratio of >2 and a neck diameter of less than 5 mm. Endovascular treatment of aneurysms ideally should be quick and straightforward to minimize complications. In contrast to open surgery, the endovascular approach to most intracranial aneurysms is generally the same, regardless of anatomic location. Of course, there are some institutional variations in technique, but the general principles of endovascular therapy are uniform. Endovascular procedures are routinely performed in our neuroangiography suite, with the patient under light intravenous sedation and analgesia. In this fashion, we can assess the patient’s neurologic status periodically. We reserve general anesthesia for patients with altered mental status or for those in whom light sedation is insufficient to reduce anxiety or movement. We are also more likely to use general anesthesia in patients with subarachnoid hemorrhage (SAH) because of their headache or altered mental status. Owing to the risk of intraprocedural aneurysm rupture, we do not hesitate to revert to general anesthesia for patients in whom conscious
Sedation is insufficient. All endovascular procedures are performed under continuous hemodynamic monitoring.

Ventriculostomy is used commonly to treat acute hydrocephalus in patients with aneurysmal SAH. The routine use of anticoagulants (see below) and antiplatelet agents during and after endovascular therapy may increase the hemorrhagic complications of such bedside procedures. Therefore, our threshold to perform a ventriculostomy is lower in patients who undergo coiling. If ventriculostomy is indicated, it is best done before the endovascular procedure rather than after it.

To reduce thromboembolic complications during aneurysm coiling, systemic anticoagulation is induced with intravenous heparin dosed according to body weight. An activated coagulation time of 250 to 300 seconds is achieved before we advance the microcatheters. For coiling of ruptured aneurysms, heparinization is usually delayed until the first coil has been deployed and the aneurysm dome is secured. In some cases where microcatheterization is anticipated to be difficult and prolonged, the patient initially receives a half dose of heparin, with the other half given immediately after placement of the first coil.

Arterial access is usually obtained through the common femoral artery where a sheath is introduced with a modified Seldinger technique. A guiding catheter is used to navigate the aorta and engage the extracranial vertebral arteries (VAs). Occasionally, these arteries may be difficult to access via the aortic arch because of tortuosity of the subclavian arteries, diffuse atherosclerotic disease, or oblique origins from the subclavian arteries. In these cases, we do not hesitate to use a radial or brachial artery approach. Digital cerebral angiography is then performed, and projections are found that optimally delineate the aneurysm neck with the relation to the parent blood vessel and neighboring branches. In recent years, rotational three-dimensional views have been useful in finding the optimum projection.

Using road-map guidance, the cerebral vessels are navigated with a microcatheter over a microwire, and the aneurysm is catheterized either directly with the microcatheter or over the microwire. In general, the verteobasilar system is less tortuous than the anterior circulation, and endovascular navigation is easier. This is particularly true for the BA, thus making microcatheterization of most basilar tip aneurysms relatively straightforward. After verification on two projections of the location of the microcatheter, platinum microcoils of different conformation, shapes, sizes, and stiffness are selected. The selected coils are delivered through the microcatheter, and the coils are detached when the position is evaluated to be appropriate and stable. If a chosen coil does not conform to the aneurysm shape or herniates into the parent vessel after several attempts, the coil is removed and a different one is chosen.

Before detachment, attention is focused to confirm the absence of parent vessel occlusion, thrombus, stenosis, or contrast extravasation. The endpoint of the procedure is dense packing of the aneurysm until additional coils cannot be placed. After microcatheter removal, final views should confirm the absence of aneurysm filling and any vascular complication related to the therapy (e.g., parent vessel stenosis, distal embolization). Final cervical angiographic runs should also be acquired to search for any iatrogenic dissection occurring during the guide-catheter positioning.

**Expanded Techniques**

The vast majority of embolized aneurysms can be coiled primarily as described above. However, certain large and broad-necked lesions cannot be treated with coiling alone. The unfavorable morphology of these aneurysms reduces the possibility of dense coil packing. The use of three-dimensional coils, balloons, and stents has expanded the endovascular treatment of large and complex aneurysms.

Three-dimensional coils were introduced in 1999. These soft coils have a three-dimensional memory so that they bend randomly during their deployment to conform to the shape of the aneurysm.9 This design allows these coils to bridge the neck of the aneurysm and provide a scaffold within which additional coils can be packed. As a result, the coil packing density can be increased in wide-necked aneurysms without prolapse of the coils into the parent vessel. After placement of several three-dimensional coils, the remaining open spaces of the aneurysm can be filled with two-dimensional coils. Complete aneurysm occlusion can be achieved in more than two thirds of aneurysms with wide necks, provided the dome-to-neck ratio is greater than 1.5.9 For aneurysms with very wide necks or dome-to-neck ratios less than 1.5, balloon or stent assistance may be necessary.

Balloon-assisted coiling involves intermittent inflation of a balloon positioned across the wide neck of an aneurysm.10 Because the inflated balloon is occlusive of the parent vessel, it is deflated after coil detachment. The purpose of the inflated balloon is to prevent herniation of the coil as it is deposited into the aneurysm (Fig. 21.1). It must be emphasized that balloon assistance may not affect packing densities and may not improve aneurysm recurrence rates. The complications of balloon-assisted coiling range were initially thought to be few, but complication rates as high as 14% have been described recently.11 Despite the increased complication rates with balloon as well as stent assistance, these adjunctive techniques still compare favorably to surgical results for posterior fossa aneurysms. Some of the increased complication rate is inherent to the complexity of the class of aneurysms that these adjuncts are designed to treat. The increased risks of using these adjunctive techniques must be weighed against the risk of treating these aneurysms surgically.

The concept of stent-assisted coiling is relatively straightforward and elegant. The stent is deployed across the wide neck of the aneurysm; microcatheterization of the aneurysm itself is then accomplished through the stent mesh, which prevents coil herniation into the parent vessel. Initially, balloon-mounted coronary stents were the only ones available for intracranial use. However, these stents can be difficult to navigate in the tortuous intracranial vessels. Furthermore, their stiffness and pressure-dependent deployment carries substantial risk of injury to the thin-walled cerebral vessels.

Recently, self-expanding stents have been introduced for intracranial use. The Neuroform (Boston Scientific, Fremont,
CA), a nickel-titanium alloy stent with a low profile and high porosity, is currently the only stent approved in the United States for intracranial aneurysm treatment. Although this stent is more navigable than the balloon-mounted stents, extremely tortuous arteries still present a formidable challenge for delivery. Stents have advantages over balloon assistance in that they offer a permanent scaffold to prevent coil herniation. The struts of the Neuroform stent do not appear to occlude adjacent branch arteries due to the open cell design of the stent.12

Experimentally, stents have been shown to divert blood from the inflow zone of the aneurysm, thus promoting thrombosis within the aneurysm.13 This property can be applied directly to side-wall aneurysms, such as basilar trunk or vertebrobasilar junction aneurysms (Fig. 21.2).14 For such lesions, stenting alone across the aneurysm neck without coiling may be enough to treat the lesion. Despite the problems of balloon-mounted stents, coronary stents have low porosity and have an advantage of impeding aneurysm filling and promoting thrombosis of the lesion. We have also used various combinations of stents within stents to decrease the aneurysm inflow further.

Potential complications of stenting include suboptimal positioning, increased thromboembolism and vessel injury inherent to placing additional devices into the intracranial vasculature, and delayed thrombosis of the stent. Furthermore, the natural history of stented intracranial lesions is not known, and clinical and radiologic surveillance for in-stent stenosis is necessary. We typically maintain patients treated with stents on both aspirin (325 mg daily) and clopidogrel (75 mg daily) for 1 month and then aspirin alone indefinitely thereafter. Overall, the results from stent-assisted coiling are favorable. The rate of technical success associated with stent-assisted coiling has been very high, approaching or greater than 90% in our experience and in published reports.15–17 Complication rates, mostly thromboembolic, have approached
25%, but once again this is partially reflective of the high-risk aneurysms for which this treatment modality has been developed. Even with stent-assistance, a recanalization rate of approximately 25% has been observed, but this is comparable to the rate of recanalization observed with direct coiling of small aneurysms.

Practical considerations may restrict the role of stenting in the setting of an SAH (Fig. 21.3). The rigorous use of antiplatelet agents required by stenting may impede adjunctive procedures such as ventriculostomy (as described previously), cerebrospinal fluid shunting, tracheostomy, and gastrostomy. These issues are not theoretical because the patients needing these procedures are usually the poor-grade patients selected for endovascular therapy in the first place. One strategy in these patients may be to secure the fundus of the wide-necked aneurysm with direct coil embolization acutely, and then completing the embolization in a delayed fashion with stent assistance. It has been shown that occlusion of the dome and fundus of a ruptured aneurysm may be protective in the short term after an SAH. A small neck remnant after initial coiling may lead to complete aneurysm occlusion due to progressive thrombosis in some cases. Of course, residual aneurysms need to be followed closely, as the risk of delayed rerupture can be as much as 5%. Our follow-up strategy for coiled aneurysms is control angiography after 6 months and then at 12 to 18 months if a residual neck is noted. The size of the residual neck determines the timing of the second follow-up angiogram. Aneurysms that remain completely occluded at the first follow-up are usually evaluated thereafter with annual magnetic resonance angiography.

**Deconstructive Approach**

If the above strategies can be considered reconstructive endovascular techniques, then, for completion, we shall mention deconstructive techniques. This technique involves the endovascular sacrifice of the parent vessel. This strategy is usually reserved for giant or fusiform aneurysms that cannot be treated using conventional endovascular techniques (see
Occlusion of the VA (or, rarely, the BA) may alter the hemodynamics of the aneurysm and promote aneurysm thrombosis. Endovascular vessel occlusion, originally done with detachable balloons, is now accomplished usually with detachable coils.

Prior to permanent vessel occlusion, balloon test occlusion is usually performed as a diagnostic procedure. An understanding of the collateral supply to the cerebral vascular territory at risk is a prerequisite to vessel sacrifice. A successful balloon test occlusion offers reassurance, but it does not guarantee against stroke after vessel sacrifice. Nonetheless, deconstructive approaches are effective in over 60% of inoperable aneurysms in the posterior circulation. Ischemic complications that result from deconstructive approaches are probably due to thromboembolism or failure of collateral circulation. To reduce this risk, endovascular occlusion may be an adjunct to surgical bypass. Despite parent vessel occlusion, the aneurysm may continue to fill owing to collateral blood supply.

Giant aneurysms of the vertebrobasilar circulation are rare lesions without any optimal treatment. Despite advances with skull base approaches to these aneurysms, the surgical morbidity remains high. Coiling of giant aneurysms usually does not result in complete occlusion, and recanalization rates are high. The two goals for treatment of giant vertebrobasilar aneurysms are reduction of mass effect and persistent thrombosis of the lesion. Endovascular treatment of these challenging aneurysms is mostly limited to deconstructive approaches. For selected patients, unilateral or bilateral VA occlusion may be effective in excluding the aneurysm from the circulation or reducing the lesion size. Less commonly, the BA itself can be occluded proximal to the aneurysm, provided the patient has good collateral filling through the posterior communicating arteries (PCoAs). In general, the closer the parent vessel occlusion to the aneurysm, the higher the chances of complete aneurysm thrombosis. To this end, the endovascular approach may be more successful than surgical occlusion of the parent vessel. Endovascular trapping of the aneurysm is also possible in which the parent vessel is occluded proximal and distal to the aneurysm to prevent retrograde filling. Parent vessel occlusion that leads to partial thrombosis of giant aneurysms may be disastrous. Partially...
Subsets of Posterior Fossa Aneurysms

Basilar Apex Aneurysms

The paradigm for the success of neuroendovascular therapy is the basilar apex aneurysm. Coil embolization undoubtedly has made the greatest impact on the treatment of this subset of intracranial aneurysms. Basilar apex aneurysms are, by far, the most common aneurysms of the posterior circulation.1 At many centers, including our own, these aneurysms are treated almost always by coiling. Our attitude is that endovascular options should be exhausted for this group of aneurysms before considering open surgical treatment. Stent and balloon-assisted coiling have allowed us to expand the indications for endovascular treatment to wide-necked and complex aneurysms. These advances, along with superior endovascular results, have made surgery for basilar apex aneurysms obsolete at many centers. Microcatheterization of these aneurysms is usually uneventful, as the BA usually has a straight course. The main challenge is preventing coil herniation and avoiding occlusion of the PCA.

Results from large endovascular series are favorable regarding technical success and occlusion rates for these aneurysms.18,26–28 Occlusion rates exceeding 90% can be achieved with primary coiling. In the largest series to date, complications occurred in 20% of 316 treated basilar apex aneurysms.29 Most of the complications were thromboembolic. Although many patients were lost to follow-up in this series, coil compaction and aneurysm recanalization was demonstrated in 24% at a mean follow-up of 19 months. Forty-eight patients (15%) required a second coiling session due to recanalization. These results seem at first glance to be mediocre, but they include preliminary experience from the early 1990s.

Direct comparison between surgical and endovascular results for these aneurysms is difficult, as most published reports include selection bias toward a certain modality, based on aneurysm morphology and size. The reported morbidity in most surgical series for ruptured basilar apex aneurysms approaches 20%, with mortality approaching 10%.1,25,30 By comparison, endovascular morbidity and mortality is in the range of 10% and less than 5%, respectively.29,31,32 Stenting strategies usually involve placement of a single stent into the PCA that is at greater risk of occlusion. Stent delivery can be difficult if the PCA originates at a 90-degree angle from the BA. In rare cases, more creative configurations such as “Y-stenting” may be used if both PCAs are at risk of occlusion. In this strategy, the first stent is delivered into one PCA, and the second stent is delivered into the other PCA through the struts of the first stent.33 Another elegant yet potentially difficult technique is navigating a stent through the PCoA into the ipsilateral P1 segment and crossing the basilar apex into the contralateral P1 segment.34 The result is a stent that spans the neck of the aneurysm and protects both PCAs. Obviously, this strategy requires a relatively large, straight PCoA. If the stent placement is successful, a microcatheter can then be advanced via the vertebrobasilar route, through the horizontally oriented stent, and into the aneurysm for coiling. However, these extra vessel manipulations carry increased risk, and we favor the simplest strategies possible to accomplish aneurysm obliteration and parent vessel preservation.

Posterior Cerebral Artery Aneurysms

Aneurysms of the PCA represent approximately 1% of all intracranial aneurysms.1,35 These aneurysms are classified according to the segment of the PCA involved, that is, P1, P1–PCoA junction, P2A, P2B, P3, or P4. Most PCA aneurysms arise from the P1 and P2 segments. PCA aneurysms tend to occur in younger patients, have a higher incidence of being large or giant than aneurysms at other locations, and often occur in the setting of other cerebrovascular lesions.36,37 Aneurysms of the P2 segment and beyond do not carry the same surgical prognosis as do those of the P1 segment.37 The P2 segment, which extends from the PCoA–PCA junction to the origin of the inferior temporal arteries in the quadrigeminal cistern, may give rise to aneurysms less frequently. Most reports of peripheral PCA aneurysms are comprised of small series or single cases (Fig. 21.4). P2 aneurysms are usually saccular, but fusiform and dissecting aneurysms are also common in this location.36,37 Compared with surgery, endovascular treatment of P2 aneurysms offers several advantages. The P2 segment is roughly 3 mm in diameter, making endovascular navigation possible.38 Because aneurysms of the P2 segment tend to be wide-necked, serpentine, or fusiform, endovascular preservation of the parent vessel is usually not possible. Balloon test occlusion of the P2 segment does not seem to be reliable. Instead, evaluation of the collateral network between the PCA and other vascular territories is more predictive of the safety of P2 sacrifice. On the basis of small series, it appears that sacrifice of the P2 segment, including the P2A subsegment, is safe and does not result in visual field deficits.37 In fact, occlusion appears to be safer in the proximal P2A segment because of preserved collateral supply distally from the anterior choroidal artery, splenial branches of the anterior cerebral artery, and leptomeningeal collaterals from the middle cerebral artery.37

Posterior Inferior Cerebellar Artery Aneurysms

Aneurysms of the PICA account for roughly 0.5% of all intracranial aneurysms.39 The morbidity associated with open surgery for these lesions has been reported to be as high as 66%, with most complications due to lower cranial nerve injuries.2,4 Although the vast majority of patients with unruptured PICA aneurysms or good-grade SAH ultimately recover well after surgery, endovascular therapy of these aneurysms...
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has proven to reduce the treatment-associated morbidity, and long-term results are comparable to those with clipping. Technical success with coiling of these aneurysms typically exceeds 95% in our experience and that of others (Fig. 21.5). Procedure-related morbidity, in the form of thromboembolism or intraprocedural aneurysm rupture, ranges from 10 to 15%. Endovascular results for ruptured PICA aneurysms are especially favorable, as almost 87% of patients with Hunt-Hess grades I, II, or III have good long-term clinical outcomes. In contrast to surgical results, half of the patients with poor Hunt-Hess grades had good clinical outcomes in one recent report. At our institution, the proximal PICA aneurysm is the main posterior circulation aneurysm that is still occasionally treated with surgery, but even this practice pattern is shifting toward endovascular therapy, considering these good angiographic and clinical results.

Most PICA aneurysms occur proximal to the choroidal point. The most important technical pearl regarding coiling of these proximal PICA aneurysms is preservation of the parent vessel because of brainstem perforators that arise from the anterior and lateral medullary segments of the parent vessel. Distal to the choroidal point, the PICA can usually be sacrificed with impunity. Endovascular treatment of unruptured proximal PICA aneurysms with wide necks usually requires stent assistance or partial coiling in the acute SAH setting, followed by stent assistance in a delayed fashion.

Peripheral Aneurysms of the Posterior Circulation

Aneurysms of the peripheral cerebellar arteries represent less than 1% of all aneurysms, with the majority located on the PICA. Most peripheral aneurysms in the posterior circulation tend to be traumatic or mycotic. Direct surgical or endovascular treatment of these lesions with preservation of the parent artery is possible but can be very difficult. These
aneurysms can often be treated safely and effectively with proximal occlusion of the parent vessel or trapping. These indirect techniques may be necessary in treating mycotic or traumatic aneurysms and saccular aneurysms with wide necks. An endovascular approach is attractive and appears to be as effective as surgical sacrifice of the parent artery. Test occlusion is often not technically feasible or practical, as it does not appear to be a reliable predictor of expected deficits from parent artery sacrifice in the peripheral cerebellar arteries because collateral circulation is usually adequate.43

Peripheral aneurysms of the SCA are extremely rare. Patients with these aneurysms usually present with SAH or
fourth cranial nerve palsy. Occlusion of the SCA seems to be well tolerated because of adequate collateral supply to the distal territory of this artery. Furthermore, there are few perforators to the brainstem originating from the peripheral segments of the SCA. The endovascular approach to these lesions involves selective microcatheterization of the distal SCA. Although direct treatment of these aneurysms with coils is optimal, this is usually not possible because of their lack of discrete necks. Endovascular occlusion of the parent artery should be done as close to the aneurysm as possible to minimize the length of arterial occlusion and to reduce the risk of occlusion of uninvolved branches.

Aneurysms of the distal AICA are equally rare with fewer than 100 reported cases. The AICA is the least likely of the major posterior circulation arteries to harbor an aneurysm. In addition to producing SAH, these lesions may produce symptoms such as tinnitus, vertigo, facial weakness, and hearing loss due to their location in the cerebellopontine angle. Endovascular treatment of these aneurysms involves either direct coil embolization of the lesion or occlusion of the distal AICA. A recurrent theme in treating peripheral aneurysms of the posterior fossa is that parent artery occlusion is safe as long as the aneurysm is distal to brainstem perforators. As with all endovascular arterial occlusions, there is a theoretical risk of retrograde thrombosis.

◆ Fusiform and Dissecting Aneurysms

Dissecting aneurysms of the posterior circulation are also uncommon, but they accounted for 4.5% of nontraumatic SAH in one autopsy series. However, the true incidence is not known and likely much lower than this figure. Due to its thin media and adventitia, the intradural VA is more susceptible to dissections than its extradural counterpart. Recurrent hemorrhages from dissecting VA aneurysms range from 30 to 70%. Mortality from untreated lesions has been reported to be 50%. Lesions with the highest risk involve the dominant VA with inadequate collateral supply from the contralateral VA or the PCoAs. Originally, endovascular treatment for these lesions was limited to deconstructive techniques (see above). These techniques include proximal occlusion of the affected segment of artery by coil embolization or endovascular trapping of the diseased segment. As with giant aneurysms, surgical bypass adjuncts may be necessary. One series of these deconstructive techniques yielded good or excellent clinical results in 61% and angiographic cure in 79% of patients.

Endovascular parent artery occlusion was evaluated in a series of 13 patients with dissecting or fusiform vertebrobasilar aneurysms. The clinical results were favorable for aneurysms in which complete thrombosis was achieved. Aneurysms involving only one VA were in this group. However, mortality exceeded 50% in the group in which only partial thrombosis occurred because of involvement of the BA or both VAs. The most secure deconstructive treatment may be aneurysm trapping, as proximal occlusion alone may result in recanalization from retrograde filling.

Reconstructive techniques with stenting could be an attractive alternative for treating these difficult lesions. Stenting of these lesions with or without coil may cause altered hemodynamics, resulting in aneurysm thrombosis and preservation of the parent vessel. Essentially, the stent reconstructs the original parent vessel while the aneurysm outside of the stent becomes thrombosed. The addition of coils may promote aneurysm thrombosis (Fig. 21.6). Preliminary experience indicates a high technical success rate and good short-term results.

Fig. 21.6  A 43-year-old woman presented with dull headaches for 2 weeks. There was no evidence of SAH on noninvasive studies. Angiography of a left VA injection (A, AP; and B, lateral), confirmed the presence of a giant, seemingly fusiform left PCA aneurysm. The right PCA did not fill from the anterior circulation. There was vasospasm of the PCAs. (continued)
Fig. 21.6 (continued)  Microcatheterization of the aneurysm (C, AP; and D, lateral) confirmed its fusiform morphology. Endovascular treatment was done in two stages. (E,F) First, a Neuroform stent (Boston Scientific) was placed through the fusiform aneurysm. (F) The arrows point to the stent markers. (G) The patient returned later for coil embolization through the stent. This is the unsubtracted view; the arrows point to the stent markers. One week later, the patient developed right hemiparesis from vasospasm of the left middle cerebral artery, suggesting that the aneurysm had ruptured previously. (H,I) Four months later, the patient had a very mild right hemiparesis; angiography showed a small, stable aneurysm neck remnant.
Endovascular treatment of posterior circulation aneurysms has supplanted surgical therapy as a first choice in many centers. Although certain aneurysms are not amenable to direct coil embolization, the expansion of endovascular techniques has allowed for treatment of broad-necked lesions. Although deconstructive approaches are appropriate for some fusiform and dissecting aneurysms, endovascular stent reconstruction seems promising for small, difficult lesions. Increased aneurysm recurrence rates and thromboembolic complications are the main drawbacks to endovascular treatment, and these considerations must be weighed against the morbidity of surgical treatment.

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In the United States, pineal-region tumors constitute approximately 1 to 3% of the intracranial masses. Victor Horsley attempted the first direct removal of a pineal tumor in 1905. This was the first attempt to remove any lesion in the region of the posterior third ventricle recorded in the modern era. Cushing proposed the idea of surgical treatment of pineal tumors but viewed this region as surgically inaccessible. In 1926, Krause reported three patients with a pineal tumor on whom he operated using a posterior fossa approach above the cerebellum (infratentorial, supracerebellar) with the patient in a sitting position.

In 1931, W.P. Van Wagenen proposed the transcortical transventricular approach, which never gained wide acceptance due to the difficulties associated with this approach. Benet Stein later improved upon the approach proposed by Krause, using microsurgical techniques and reported several series on his improved infratentorial, supracerebellar approach with the patient in the sitting position.

Walter Dandy was accredited with the first scholarly analysis with his supratentorial parafalcine approach. He started this approach after an initial trial on animals. He placed the patient in either a semi-sitting position or semiprone position with the side of the approach uppermost, thus retracting the occipital lobe against the gravity. He dissected down the medial side of the occipital lobe. On reaching the tentorial incisura, he sectioned the splenium of the corpus callosum and dissected the venous confluence.

Technical variations for similar lesions include the occipital transtentorial approach described by James Poppen in 1966, and the combined supratentorial and infratentorial transsinus approach described by Ziyal et al.

Not only have the surgical approaches evolved, but also advances in the neuroimaging, endoscopic techniques, and neuronavigation have greatly helped in the successful management of the pineal tumors with an associated decline in mortality.

Posterior Interhemispheric Approach to Pineal Region and Brainstem

Prasad Vannemreddy, Haim Ezer, and Anil Nanda

Posterior Approaches to the Pineal Region

Posterior approaches to pineal tumors can be divided into supratentorial and infratentorial. Supratentorial approaches include the occipital transtentorial, interhemispheric transcallosal, and interhemispheric retrocallosal. Infratentorial approaches include the infratentorial supracerebellar and infratentorial paramedian supracerebellar.

Two approaches most commonly performed for pineal region tumors are the occipital interhemispheric transtentorial and infratentorial supracerebellar. The wide variety of surgical positions applied to reach this location reflect the difficulties in accessing the lesions. The positions utilized for these approaches vary from prone to sitting to semiprone, semi-sitting, three-quarter prone/park-bench, and Concorde. The availability of newer imaging techniques and intraoperative neuronavigation methods have further improved the outlook and the operative results remarkably. Studies have reported minimal to zero mortality. Intraoperative image-guided surgery has been utilized as a routine at present in our experience.

Indications

Traditionally the posterior interhemispheric approach was applied to access tumors located at the posterior third ventricle, especially pineal tumors growing superiorly or laterally to the trigone of the lateral ventricle, or tumors growing into the ambient cistern. This approach was also used for tumors above the venous structures, around the vein of Galen. Tumors arising in the splenium of the corpus callosum or the medial occipital lobe are easily removed via this route. Extended indications include tumors arising from the brainstem, vascular malformations, or aneurysms of the
P2/3 segment of the posterior cerebral artery. Other extended indications include vascular malformations of the vein of Galen and lesions of the superior vermian/cerebellar area and cavernomas of the dorsal midbrain.\textsuperscript{20–28}

A lesion in the pineal gland location is easily accessed by the occipital interhemispheric approach, without resection of the tentorium or resection of the splenium of the corpus callosum.\textsuperscript{20–28}

Fig. 22.1 A standard pineal region tumor can be directly accessed by the occipital interhemispheric approach. This does not require sectioning of the tentorium or resection of the splenium of the corpus callosum.

Fig. 22.2 Lesions growing anteriorly and superiorly reaching under the splenium of the corpus callosum or infiltrating it would require resection of the corpus callosum. Sometimes endoscopic application is useful in avoiding the resection of splenium. Similarly, tumors that can be sucked out, such as those in the dermoid and epidermoid, can be removed without causing damage to splenium.

Fig. 22.3 Large tumors like meningiomas or pinealoblastoma or a low-level posterior cerebral artery (PCA) aneurysms and cerebellar vermin tumor are better managed by opening the tentorium cerebelli.

Preoperative Evaluation

Apart from routine evaluation of the patient for fitness for anesthesia, the surgical workup includes good-quality multiplanar magnetic resonance imaging (MRI) studies with and without contrast for precise localization of the tumor.\textsuperscript{6} Angiography may be beneficial in selected instances, such as vascular pathologies and selected vascular tumors. In these instances, an angiogram is helpful in delineating important surrounding vascular structures.\textsuperscript{29} MRI is mostly adequate for the diagnosis of cavernous angomas of the brainstem as well as posterior third ventricular region tumors. MR spectroscopy may help in preoperative histologic diagnosis.
standard digital subtraction angiography (DSA) for vascular malformations and aneurysms provides the location and helps in surgical strategies. It is important to note that a vascular nidus located lateral to the P2/P3 segments of the posterior cerebral artery (PCA) on the anteroposterior angiographic views may not be a suitable pathology to approach using the interhemispheric approach. This would necessitate severe occipital lobe retraction to reach the nidus, and it can produce irreversible hemianopsia.29

Frameless stereotaxy was utilized initially to plan the optimal skin incision in relation to the underlying superior sagittal sinus as well as determine the optimal trajectory. If the radiologic studies are suggestive of a germ cell tumor, then an MRI scan of the whole craniospinal axis is indicated. In these instances, measurement of tumor markers, by using, for example, tumor marked chorionic gonadotropin (hCG), carcinoembryonic antigen (CEA), or α-fetoprotein, is needed.1 The distinction between germinomatous and nongerminomatous tumors affects the patient’s management. Germinomatous tumors show good response to gamma knife radiosurgery.30 Nongerminomatous germ cell tumors are resistant to radiotherapy, but may respond to chemotherapy.30a,30b

◆ Positioning of the Patient

Various positions have been described for this approach, as the semi-sitting position described by Yasargil et al.26 and also, the Concorde position. The lateral position has been found to be suitable in our experience.17 Significant air embolism might result from the sitting or semi-sitting positions, which sometimes produces clinically relevant changes.20

In the operating suite, the endotracheal tube is placed in the operating area, and a central venous catheter and an arterial line are also placed. Following lumbar drain placement, the patient is then placed in the lateral position with the head turned 30 degrees toward the floor (which places the side of access in the dependent position), allowing the occipital lobe to fall inferiorly, to facilitate minimal retraction. A beanbag in conjunction with cloth tape and padding secures and maintains the body position; the superior shoulder is gently pulled using a strip of tape with foam under it, attached to the foot of the bed. The lower knee has to be bent with foam padding placed between the knees and malleoli, under the axilla, and around the elbows. After placement of the head in a Mayfield head-holder and subsequent fixation to the bed, image registration using fiducial markers and the Stealth neuronavigation system (Medtronic Sofamor Danek, Memphis, TN) performed.

◆ Operative Technique

Frameless stereotaxy helps in planning the optimal skin incision, in relation to the underlying superior sagittal sinus, as well as in determining the optimal trajectory (avoiding apparent bridging veins and the shortest distance to the lesion). A very thin strip of hair (approximately 2 cm wide) is shaved, and then a horseshoe-shaped incision is placed on the side of the craniotomy, in the occipital region. The base of the skin flap goes along the superior nuchal line, and the medial limb of the incision is just past the midline. The skin flap is elevated along with the galea and pericranium, and reflected inferiorly. A single bur hole is made near the midline, and a rectangular bone flap (approximately 5 cm at midline × 3 cm) is elevated using a high-speed drill. The edge of the superior sagittal sinus is barely visualized. The dural incision is made utilizing the entire space of the craniotomy and turned medially with the base on the sinus (Fig. 22.4).

The utmost care is required to avoid damage to the sinus. Bridging veins are scarce in the occipital region, and those present can be easily avoided.21 Having the occipital lobe in a dependent position facilitates a surgical corridor, which requires minimal arachnoidal dissection to visualize the white posterior corpus callosum, not to be confused with the yellow cingulated gyrus, which has a resemblance to the corpus callosum (Fig. 22.5A). Opening the lumbar drain at this time also facilitates brain relaxation. At this stage particularly, we have found it very useful to confirm our position and trajectory using frameless stereotaxy, and alter it accordingly to provide optimal access. The posterior end of the tentorial notch is typically encountered next, where the falx cerebi joins with the tentorium cerebelli. A thick layer of arachnoid, forming the posterior boundary of the quadrigeminal cistern, is now sharply incised to enter the quadrigeminal cistern.

Surgical Procedure to Remove the Lesion

The major venous network (veins associated with the vein of Galen) is usually seen covering a tumor in the pineal region. Although some authors believe that this is problematic by presenting a barrier between the surgeon and the lesion, we have found that direct visualization of the venous structures at this early stage of dissection actually aided us in protect-
ing the venous structures (Fig. 22.5B). Using meticulous,
sharp dissection, these structures can be gently separated
and retracted to provide direct access to the lesion.

Different lesions, depending on the size, gross consistency,
origin, and location in relation to the venous structures, can
be resected using different strategies. For example, in tento-
rial meningiomas, the venous system is usually largely dis-
placed ventrally, and the tumor is encountered before these
veins; thus tumors are decompressed piecemeal, and the
capsule can be removed at the end. Pineal region cysts are
punctured and the contents are aspirated or sucked out so
that the walls of the cysts collapse and can be excised com-
pletely after gentle separation from surrounding structures.

For arteriovenous malformations (AVMs), it is important
to mobilize the medial occipital and the deep venous veins
to expose the medial surface of the occipital lobe. Mobil-
ization of these veins enables the dependent occipital lobe
to fall away further. Moreover, this enables the surgeon to
visualize the posterior end of choroidal fissure and cisterna
ambiens. Also, a clear view of the P2 and P3 segments of
the posterior cerebral artery is obtained. By following the
branches of the posterior cerebral artery, the nidus of the
AVM can be reached. In addition, this exposure also provides
entry into the trigone of the lateral ventricle via the choroi-
dal fissure. Anterior dissection, just below the formation of
the vein of Galen, reveals a dense network of the arachnoid
mater (the velum interpositum cistern), which leads into the
third ventricle when divided. Coagulation of the tela choroi-
dea of the third ventricle provides entry into this cavity. Just
inferior to this, the pineal gland is visualized, along with the
posterior choroidal artery entering the gland. Below the pi-
neal gland, the quadrigeminal plate is seen, and just posterior,
the superior vermis of the cerebellum can be visualized.

◆ Case Illustrations

Case 1

A 53-year-old woman presented with recurrent episodes of
bleeding from a midbrain cavernoma (Fig. 22.6). She also
had one episode of bleeding from a cavernoma in the occipi-
tal lobe, and previously underwent multiple operations for
cavernomas located elsewhere in the brain. She presented
this time with an altered sensorium, oculomotor palsy, and
hemiparesis, following a bleed from a midbrain lesion. The
patient underwent surgery, using the occipital inter-hemi-
spheric approach (OIH) with frameless stereotaxy guidance.
Neither the tentorium nor the splenium was cut; the lesion
was completely resected (Fig. 22.7). The patient’s neurologic
deficits gradually resolved completely, and she has returned
to normal.

Case 2

A 32-year-old man presented with headache and altered
sensorium. Computed tomography (CT) of the head showed
a hematoma in the medial occipital lobe. Angiography re-
vealed a medial occipital AVM (Fig. 22.8A,B). The patient was
operated on with the occipital interhemispheric approach.
The AVM was totally resected (Fig. 22.8C). At 6-year follow-up, he is leading a fully active life, with no apparent new neurologic deficit.

**Case 3**

A 77-year-old woman presented with subarachnoid hemorrhage (Hunt-Hess grade I). Angiography revealed a distal posterior cerebral artery aneurysm (P3 segment) and a small AVM in the cerebellum. The distribution of blood and the appearance of the aneurysm suggested that the aneurysm had likely bled. Using OIHA without frameless stereotaxy, finding the aneurysm was considerably difficult. Ultimately, it was visualized after the tentorium was incised; the posterior cerebral artery had a somewhat anomalous course, dipping below the tentorium. The aneurysm was successfully clipped, and the patient had an uneventful recovery.

**Modifications**

Shirane et al. utilized the occipital transtentorial approach for pineal region tumors in 31 patients, and the applied navi-
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An image-guided system with endoscopy was used in 16 of these patients. All 16 patients had excellent outcomes without any complications. In the other 15 patients, neuronavigation could not be used, and complications were seen in five of the 15 patients in the form of seizures and hemianopsia. In contrast, none of the 16 patients who had image-guided surgery and endoscopic assistance developed any new deficits. The authors feel that the use of assisted systems, such as neuronavigation and endoscopy, during microsurgery increases the surgery's safety and accuracy.

Nazzaro et al. operated on 12 patients, who were placed in the semi-sitting position; none of them had an air embolism or hypotension. However, a detailed neuro-ophthalmology workup was performed in all cases, which revealed a transient visual field defect in all patients, and it persisted in two patients. Lapras et al. suggested that the incidence of visual field defects may be reduced by retracting the occipital lobe laterally instead of the superolateral lifting of the lobe with the patient in the sitting or semi-sitting position.

Konovalov et al. placed patients in the semi-sitting or three-quarter positions when operating on meningiomas of the pineal region, and performed a craniotomy from the nondominant side. An angiographic evaluation was obtained in seven of 10 patients for topography of the blood vessels around the tumor, especially the great vein of Galen and the straight sinus. According to the authors, the three-quarter prone position provides better visualization and less retraction of occipital lobe, albeit with unfamiliar anatomic detail.

Yasargil proposed the occipital interhemispheric approach as the first choice for aneurysms arising from the P3 segment of the posterior cerebral artery. The selection of the approach for the P2 segment, however, depends on the surgeon's choice. The P2p segment is located between the cerebral peduncle and the quadrigeminal cistern, and it traverses the ambient cistern. Terasaka et al. found that this segment in the ambient cistern is more easily reached by the occipital interhemispheric approach than by the subtemporal or pterional transsylvian approach. Based on their cadaver

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Fig. 22.7 Intraoperative pictures for the patient in Case 1. (A) Exposure of the posterior surface of the midbrain by OIHA. (B) The midbrain surface shows a yellowish tinge, which suggests hemorrhage. (C) Postresection cavity in the midbrain.
study and clinical experience, these authors suggest that high P2p segment aneurysms are better approached using this approach, especially when an angiogram in the lateral view shows the aneurysm to be close to the anterior choroidal artery in the temporal horn. However, just as in cases in which the vein of Labbé restricts temporal lobe retraction in the subtemporal approach, the occipital interhemispheric route also would face limitations due to the internal occipital vein, especially when the parent artery is located behind the aneurysm. These authors also believe that vascular reconstruction involving the PCA would be nearly impossible. Touho et al reported on an anastomosis of the occipital artery to the PCA, with interposition of a graft from the superficial temporal artery for severe stenosis of the distal basilar artery, with a good outcome. They propose this approach as a safer one compared with other routes utilized for an anastomosis between the superficial temporal artery and the superior cerebellar artery or the PCA. Most of these complications are attributed to severe temporal lobe retraction.

Ausman et al suggested the possibility of reaching the cerebellar vermis from above by cutting the tentorium via the occipital interhemispheric approach. The possibility of reaching the cerebellum by this route is especially interesting because the vascular supply of lesions located in this part of the cerebellum comes from the superior cerebellar artery, and this artery is visualized at the initial stages of surgery when the tentorium is incised from above. Kurokawa et al reported a series of six cases with cerebellar vermician tumors excised via the occipital interhemispheric transtentorial approach. A preliminary report of the transtentorial visualization of the cerebellar region appears in James Poppen’s report. We have utilized the transtentorial access to reach the P2/3 segment of the PCA with an aneurysm. Kurokawa et al found easy access to unilateral lesions of the cerebellar vermis.

Fig. 22.8  Intraoperative (A,B) and postoperative (C) angiograms of the patient in Case 2, who had a medial occipital arteriovenous malformation.
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Complications

The main surgical complications following this procedure are hemorrhage, brainstem damage, and visual field defects. Intra- and postoperative hemorrhages originate mainly from the veins around the tumor, the most difficult task being separation of the internal cerebral vein from the tumor. A small portion of the splenium may require excision to expose the internal cerebral vein safely and to preserve it. The Valsalva maneuver at the end of tumor excision confirms complete hemostasis.

Neural structures and the brainstem may be damaged by direct surgical trauma or due to interrupted vascular supply. Fukui et al. reported central hearing loss following surgery, which was attributed to damage to the auditory fibers from the inferior colliculus to the medial geniculate body, passing across the dorsolateral surface of the midbrain. Obstruction of the medial posterior choroidal artery may cause ischemia to the medial thalamus.

The occipital interhemispheric approach has the potential to damage the occipital lobe, caused by retraction. The internal occipital vein usually originates from the inferior medial surface of the occipital lobe and runs anteromedially toward the pineal region, entering the vein of Galen. Injury to this vein often results in either transient or permanent hemianopia.16,51 An additional advantage of utilizing an endoscope is the feasibility of performing a third ventriculostomy in cases of malignant tumors with infiltration of the surrounding structures.49 Endoscopy is sometimes complementary to neuronavigation to compensate for possible brain shifts.

Conclusion

Different approaches to the posterior third ventricular and pineal regions have been described. The key points in the application of the occipital interhemispheric approach are as follows: (1) Surgeons must be familiar with patient positioning, which can provide brain retraction by gravity. Surgeons choose from the various available surgical positions, depending on their experience. Appropriate positioning and orientation to the anatomic detail not only improve the surgical corridor but also reduce the retraction on the occipital lobe and the complications associated with it. (2) Preoperative planning includes the acquisition of a neuronavigational scan, using posterior fiducial markers to achieve greater accuracy. (3) Neuronavigation is important in the following situations: (a) in planning the skin incision and trajectory (to allow the shortest trajectory while avoiding apparent bridging veins); (b) while traversing the corridor between the occipital lobe and falx, to ensure the trajectory toward the target; (c) once near the target, in determining the shortest and safest route of entry toward an intraparenchymal lesion; and (d) in defining the extent of the resection at the end. In the event of a brain shift following opening, the landmarks on the navigation system will assist in providing accuracy during the procedure.

References


Posterior Fossa Arteriovenous Malformations and Cavernous Angiomas

Ramachandra P. Tummala, Mustafa K. Başkaya, and Roberto C. Heros

◆ General Surgical Strategies

The planning of a surgical approach in the posterior fossa is dependent on several factors. The shortest path from the surface of the patient to the epicenter of the lesion is determined. The margin of the lesion that is closest to a pial or ependymal surface must be identified to avoid or minimize the amount of dissection through normal tissue. After determining which of the cerebellar surfaces (tentorial, petrous, or suboccipital) offers the maximal visualization of the lesion, a group of approaches is chosen as potential candidates. A final surgical plan is chosen based on the surgeon’s preference and familiarity with a certain approach and a thorough knowledge of the regional anatomy. Of course, the most direct path to the lesion may not be the safest or most practical, and compromises in the approach may be necessary. One additional critical factor that affects the choice of approach in arteriovenous malformation (AVM) surgery is the location of arterial feeders. This is in contrast to cavernous malformation surgery in which the critical factor in the approach is the surface representation of the lesion.

Lesions of the inferior vermis, cerebellar tonsils, medial cerebellar hemispheres, and brainstem that extend through the floor of the fourth ventricle are best approached through a midline suboccipital craniotomy. More laterally situated cerebellar hemispheric lesions can be approached through a paramedian variant (Fig. 23.1). More laterally situated cerebellar hemispheric lesions can be approached through a paramedian variant. Lesions involving the tentorial surface of the cerebellum, tectal plate, or the pineal region can be reached through two main approaches. The supracerebellar, infratentorial approach provides a tangential view of the rostral vermis, the quadra-rangular lobules, and the superior semilunar lobule (Fig. 23.2). This approach can be performed with the patient in either the sitting or the Concorde position. Although precautions must be taken to detect and treat air embolism, we prefer the sitting position because the effects of gravity cause the cerebellum to fall inferiorly. Another potential disadvantage of this position is its initial awkwardness and long working distance to the lesion, given the fact that the operative microscope must be placed between the surgeon’s head and the patient. The body of the microscope is centered between the surgeon’s arms as opposed to the more natural position of the surgeon’s arms underneath the microscope. These conditions may increase fatigue in the upper extremities of the surgeon. We believe these inconveniences are offset by the increased exposure obtained from the effects of gravity on the cerebellum. We obtain additional exposure by routinely extending the craniotomy 2 cm above the torcular and transverse sinuses, allowing for maximal retraction of the superior leaf of dura based along the transverse sinuses. A paramedian or more lateral variant of this approach offers a more direct view of the superior cerebellar peduncle and is optimal for unilateral lesions between the trigeminal nerve and lateral border of the tectal plate.

If the cerebellar malformation is located more anteriorly and remains unilateral, the occipital transtentorial approach should be considered; we prefer this approach for lesions supplied by the superior cerebellar artery and its branches from one side only (Fig. 23.3). This approach is also satisfactory for lesions in the retromesencephalic region that extend inferiorly along the cerebellomesencephalic fissure. The patient is secured in the lateral position with the ipsilateral oc-
cipital lobe down. This positioning allows the occipital lobe to fall away from the falx, and the additional use of lumbar drainage results in minimal brain retraction. The craniotomy crosses the sagittal and transverse sinuses as in the supracerebellar, infratentorial approach. We divide the tentorium parallel to the straight sinus to expose the tentorial surface of the cerebellar hemispheres and the rostral vermis. Great care must be used to dissect through the thick layer of arachnoid overlying the vein of Galen and its tributaries. These veins obscure the contralateral structures. The occipital lobe has little tolerance for retraction, and despite the precautions taken to minimize the retraction, visual field deficits remain a serious potential consequence of this operation.

The other frequent approach for posterior fossa AVMs and cavernous malformations is the retrosigmoid approach (also referred to as retromastoid and lateral suboccipital approaches). Originally designed for cerebellopontine angle lesions, this approach can also be applied to vascular malformations of the petrous surface of the cerebellum and the lateral pons. We prefer the lateral position with the head parallel to the floor. Flexion of the neck provides increased visualization of inferiorly situated lesions and increases the

Fig. 23.1  (A) Axial T1-weighted magnetic resonance imaging (MRI) shows a left cerebellar arteriovenous malformation (AVM) in a 42-year-old man who presented initially with lethargy and ataxia from hemorrhage. Anteroposterior (B) and lateral (C) views of the left vertebral injection during cerebral angiography demonstrated the AVM. After recovering from the hemorrhage and the associated hydrocephalus, the AVM was resected through a left paramedian suboccipital craniotomy. (D) Postoperative angiography confirmed complete resection of the lesion.
working space above the shoulder. We supplement a conservative retrosigmoid craniotomy with a craniectomy to skeletonize the sigmoid sinus up to its junction with the transverse sinus. The dura is then opened in either a Y or a lambda (\(\lambda\)) pattern. The release of CSF from the cisterna magna or from the lateral cerebellomedullary cistern provides relaxation of the cerebellum and facilitates retraction to visualize the petrous surface. The middle cerebellar peduncle is also seen through this tangential exposure.

For more lateral approaches that minimize cerebellar retraction, the posterior temporal bone is resected to varying degrees. This family of presigmoid approaches includes the retrolabyrinthine, translabyrinthine, transcochlear, and transotic.\(^1\) For AVMs, we have never used more than a simple presigmoid, retrolabyrinthine approach combined with a small subtemporal craniotomy and division of the superior petrosal sinus and the tentorium all the way through the incisura.

This combined approach enables the safe division of the tentorium, preserving the temporal lobe draining veins, and provides a more direct view of the anterior and lateral surface of the brainstem and the region of the superior cerebellar peduncle. Finally, lesions involving the lateral and posterolateral medulla as well as the anterior cerebellar tonsils are best approached by the far lateral transcondylar approach.\(^2\)

**Microsurgery for Posterior Fossa Arteriovenous Malformations**

**Technical Points**

Arteriovenous malformations of the posterior fossa represent approximately 20% of all intracranial AVMs.\(^3\) Brainstem and cerebellar AVMs were considered historically to be simi-
lar entities because of their common vascular supplies. It is now clear that AVMs of the cerebellum and brainstem have distinct natural histories and different presentations. For the present discussion, we shall consider them separately.

Cerebellar AVMs have been classified by their location in the cerebellar hemispheres, tonsils, and vermis. Brainstem AVMs have been categorized as superficial or deep. Superficial lesions are pial or subpial and derive all their arterial feeders from circumferential arteries coursing around the brainstem with no perforating arterial supply. Reports of successful resection of brainstem AVMs have been limited to these superficial lesions. Deep AVMs are surrounded by brainstem parenchyma and are supplied by perforating arteries; this makes them inoperable for all practical purposes, and we shall not discuss them further.

As implied earlier, one of the key considerations in planning the surgical approach to any AVM is the early control of the arterial feeding pedicles. In general, one should respect the principle of taking the arterial supply only at the point where it reaches the malformation to avoid excessive parenchymal damage from occlusion of branches to normal brain before the feeder reaches the AVM. However, at times, “safety” supersedes “fineness,” and there are instances where we occlude the arterial supply to cerebellar AVMs as these feeders enter the cerebellum. This is particularly true with large AVMs involving one cerebellar hemisphere. In general, we have found that it is safe to occlude superior cerebellar branches that are presumed to go to a large cerebellar hemispheric AVM as they enter the cerebellum after coursing around the brainstem and giving all the important perforators to the brainstem. This arterial supply can be safely reached by either the supracerebellar, infratentorial approach or the occipital, transtentorial approach in smaller lesions that have unilateral supply, as indicated above. For unilateral large cerebellar hemispheric lesions that reach the petrous surface, we prefer the retrosigmoid approach, which can be extended superior to essentially a far lateral, supracerebellar, infratentorial approach and inferiorly to a far lateral, suboccipital, transcondylar approach. With these extensions, one can control all three main arterial supplies to a unilateral cerebellar lesion; the superior cerebellar supply can be controlled by going over the superior lateral surface of the cerebellum. The anterior inferior cerebellar supply can be controlled by following the posterior inferior cerebellar artery (PICA) between the tonsils to the choroidal point, one can sacrifice the branches that come back as they enter the cerebellar hemisphere in cases of large AVMs. Again, all the important

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**Fig. 23.3** Coronal contrasted T1-weighted (A) and axial T2-weighted (B) MRI of a 46-year-old woman who presented with progressive ataxia. The imaging characteristics are consistent with a cavernous malformation involving the anterior tentorial surface of the left cerebellar hemisphere. Because of its anterior, unilateral location, the lesion was resected through an occipital, transtentorial approach with the left side down. The patient developed a right-sided hemianopsia postoperatively, which resolved within 2 weeks.
branches of the PICA to the brainstem and to the deep gray matter of the cerebellum are given off before or at the choroidal point.

To reiterate, we do not recommend surgery for intrinsic AVMs of the brainstem fed by perforating vessels. The only AVMs we have approached are very small superficial lesions that are mostly pial or subpial and where the arterial supply is presumed to come around and not through the brainstem. Even when these conditions are met, we still find these lesions very difficult to treat, and on more than one occasion we have abandoned the attempt after not being able to identify clearly which small arterial branches go to the AVM and which go to the brainstem. Frequently, we place small temporary clips in all the feeders that we think go to the AVM and observe the vein for a significant change in color indicating dearterialization. If this does not occur, and the draining vein remains tense and red, we prefer to abandon the attempt at surgical excision and consider radiosurgery.

After controlling the main arterial supply, the AVM is usually not completely dearterialized and the vein almost always remains red. It is a matter of surgical judgment for the surgeon to decide when the lesion is ready for the surgeon to begin circumferential dissection, which should proceed always in a plane as close to the malformation as possible. However, hemispheric cerebellar AVMs afford the surgeon a generous dissection plane that may be a few millimeters from the AVM. Almost invariably, deep perforating vessels keep the lesion arterialized after control of the major arterial supply and have to be controlled at the very end of the dissection. These are very fragile vessels that are difficult to coagulate and retract easily into the parenchyma, causing further damage as the surgeon attempts to occlude them. We have found Sundt microclips (Codman and Shurtleff, Raynham, MA) invaluable in controlling these small deep perforators. Most frequently, the ependyma of the ventricle needs to be reached before full control of this perforating blood supply, and it is at that time that the vein or veins finally become blue and the lesion can be removed after ligation these vessels. With brainstem AVMs, as implied above, the surgeon does not have the luxury of dissecting around the AVM into the brainstem parenchyma if the lesion is still arterialized. One simply cannot afford to produce any significant damage of the brainstem by trying to control perforating arterial supply; therefore, unless the lesion can be dearterialized completely by control of feeders on the surface of the brainstem, it is best to abandon the attempt at excision.

**Intraoperative Complications**

As we review our results, our most common cause of intraoperative morbidity has been what we generally classify under “poor judgment.” This essentially refers to operating on AVMs either in the brainstem or extending into the brainstem through the cerebellar peduncles. This mistake was easier to make before the days of magnetic resonance imaging (MRI) when we frequently operated on cerebellar AVMs that involved the cerebellar peduncles or AVMs that we thought were primarily in the peduncles. It was a difficult matter to discern that the AVM did not extend into the brainstem or obtain a deep perforating supply through the peduncles that had to be followed into the brainstem. Nowadays, with a good MRI, it is relatively straightforward to know when such extension exists. Deep bleeding from perforating vessels that had to be followed through the deep gray matter of the cerebellum has been another source of morbidity, although that morbidity generally consists only of cerebellar ataxia that is not a major source of functional disability, and these patients are almost always independent. On a few occasions, we have encountered significant intraoperative bleeding from beginning the dissection of the AVM before sufficient control of the arterial supply either at surgery or by preoperative embolization. 

Generally, we have been cautious about using preoperative embolization because of its associated significant morbidity; however, clearly there are a few instances where we wish we had used preoperative embolization because the intraoperative bleeding encountered resulted in significant disability that probably could have been avoided. The other type of significant problem that we have encountered in a few instances is cranial nerve damage with lesions that extend into the cerebellopontine angle or around the lower cranial nerves. Needless to say, with more careful microsurgical technique, these complications could have been avoided.

**Postoperative Complications**

**Hemorrhage**

The most serious complication after AVM surgery is hemorrhage from residual fragments of an AVM or from insecure hemostasis. An unrecognized small piece of residual AVM is most frequently the source of bleeding because the necessity to excise an AVM on a plane very close to its margin creates the potential for leaving behind small remnants of AVM, which represent a significant risk of hemorrhage because they are still arterialized and frequently disconnected from its venous drainage. Additionally, at the end of the resection, in the deeper portion of the AVM, the surgeon frequently has difficulty differentiating true AVM from fragile deep feeding and draining vessels. Intraoperative angiography is very useful in detecting residual portions of AVM. However, certain operative positions may pose some difficulties obtaining proper images. In these instances, the patient should undergo immediate postoperative angiography before awakening from anesthesia, and if any residual AVM is found, the patient should be taken back to surgery for resection of the remaining AVM. The exception to the need for intraoperative or immediate postoperative angiography is with simple, smaller surface AVMs when the experienced surgeon can be relatively sure that the AVM has been completely removed.

To avoid the second error, insecure hemostasis, we perform the entire procedure under normotensive blood pressure. The use of hypotension can reduce the amount of bleeding during surgical dissection but may increase the risk of postoperative hemorrhage from insecure hemostasis. After resection of the nidus, we routinely elevate the blood pressure by approximately 20 to 30 mm Hg and observe the resection cavity for 10 minutes. With this maneuver, we have
encountered spontaneous bleeding within a few minutes in several patients. Needless to say, meticulous control of the blood pressure to parameters below the level at which hemostasis was achieved is essential during the wound closure, emergence from anesthesia, and the immediate postoperative period.

Venous Thrombosis

There is a theoretical risk of inducing stasis in long segments of veins where flow is suddenly interrupted after resection of high-flow AVMs. We have had one case of retrograde thrombosis and postoperative venous infarction after a straightforward resection of a high-flow AVM of the cerebellar vermis. At the end of the excision, we noticed that the markedly dilated internal cerebral veins, basal veins, and vein of Galen were relatively collapsed. The patient was comatose postoperatively, and a postoperative angiogram showed no filling of the deep venous system. The patient remained in a coma for several days but gradually began to improve to the point that he had an incomplete, but remarkable, recovery.

Common features of the previously reported cases and the one presented above are high flow to the AVM, extensive retrograde venous drainage, and frequent occlusion or stenosis of the antegrade venous drainage. According to a study of 33 patients in whom flow velocities and CO2 reactivities were measured, the flow velocity in the draining vessels was close to zero after removal of the AVM nidus. This may lead to venous thrombosis in the draining vessels because pathologic changes in the draining veins have already taken place, as a normal vein with normal endothelium is not likely to thrombose.

When faced with a high-flow AVM where the venous drainage is retrograde into normal veins that ordinarily drain parenchyma, it is desirable to reduce the flow gradually in the malformation with staged embolization or staged ligation of feeding arteries. Profound neurologic deficits due to venous thrombosis can have a much better prognosis for eventual recovery than similar neurologic deficits due to arterial occlusive disease, as in our case. An important clue that this type of complication would result in a good recovery is that the computed tomography (CT) scans may not show significant, irreversible brain damage in spite of the profound neurologic deficit during the initial postoperative period. Finally, these patients should be kept well hydrated during the intraoperative and postoperative periods to avoid further collapse of veins.

◆ Microsurgery for Posterior Fossa Cavernous Malformations

Indications

Infratentorial cavernous malformations (CMs) cause symptoms and neurologic deficits from hemorrhages, from obstructive hydrocephalus, or from mass effect from the growth of the lesion from repeated small hemorrhages. Relatively small brainstem lesions may result in significant deficits due to their critical location. Seizures are not a feature of posterior fossa lesions. Although headaches should not be a significant feature unless associated with hydrocephalus, we have seen several patients presenting with an acute headache associated with neurologic deficits. The technical points of surgery for cerebellar CMs are not significantly different from resection of supratentorial CMs. By virtue of their critical location, we shall focus here on the technical points of surgery for brainstem CMs.

The natural history of CMs is not as well defined as that of AVMs; however, an approximately 0.6 to 0.7% yearly risk of hemorrhage has been estimated for asymptomatic lesions. The rate of rehemorrhage in lesions that present with a clinically significant hemorrhage has been estimated to be approximately 4.5%. This rate depends on how hemorrhage is defined (i.e., by interval changes on MRI or, more strictly, by a significant clinical worsening accompanied by a clear indication of new hemorrhage on MRI). It has been suggested by several groups that brainstem CMs, which account for approximately 20% of all intracranial CMs, have a higher rate of bleeding. A large review of brainstem CMs estimated the rate of hemorrhage per year per lesion at 2.7%. However, this may be observational bias because brainstem lesions are more likely to present clinically when they hemorrhage compared with CMs in less eloquent areas of the brain.

The surgical resection of CMs contained within normal brainstem parenchyma carries a high morbidity. Recurrent hemorrhages may cause enlargement of the lesion and more pronounced neurologic deficits, but they rarely result in catastrophic deficits or death. As the lesion enlarges from repeated hemorrhages, it may dissect slowly through the parenchyma to the pial or ependymal surface. Once this occurs, they become more surgically accessible and obviate the need for incising the brainstem, thus carrying less surgical morbidity than their deeper counterparts.

Technical Points

Patients with an acute brainstem hemorrhage may look severely disabled, with poor prognosis; however, considerable neurologic improvement is to be expected. Thus, the surgeon must be cautious about operating acutely under the assumption that the patient already has a major deficit, and that surgery will not worsen the condition of the patient. Frequently, in this setting, a deficit that may likely have improved spontaneously may become irreversible as a consequence of ill-advised surgery.

The general surgical techniques for posterior fossa CM resection are no different from those for supratentorial CMs. Conventional imaging may be supplemented by frameless stereotactic guidance to plan the surgical approach. We routinely monitor motor, somatosensory, and brainstem evoked potentials during the resection. The only sound surgical strategy to remove a CM is an “all-or-none” commitment, although we must emphasize that the end point with large brainstem CMs is not as clear-cut as with AVMs. In the case of the latter, bleeding usually continues until the AVM is removed completely with intraoperative angiographic confirmation. On the other hand, the surgeon may attack the interior of a
brainstem cavernoma, coagulating lobules of an irregular lesion to “shrink” it away from the parenchyma. Under these circumstances, it is easy to resect a cavernous angioma incompletely, as we have done in two cases. Ideally, resection should be done by respecting the gliotic plane induced by hemorrhage. It is intuitive that the brainstem is unforgiving of spatial and technical errors.

Surgery of cavernous angiomas is very different from surgery for AVMs. The key step in AVM surgery is to occlude as much of the arterial supply to the lesion as possible before beginning resection of the lesion. In contrast, we preserve all significant (more than half a millimeter approximately) arterial or venous branches of cavernous angiomas. Our observations concur with others’ that many of these cavernous angiomas of the brainstem exist in close relationship to venous angiomas, and one of the keys to successful surgery of cavernous angiomas is full preservation of the frequently associated venous angiomas. Clearly, the cavernoma receives small arterial branches and is drained by very small venules, but, as a rule, no major arterial branches feed the lesion nor do major veins drain it. In this respect, cavernoma surgery is perhaps more similar to tumor surgery and can be likened to removal of a well-circumscribed cerebral metastasis. One can frequently work within the cavernoma because these lesions are under relatively low flow. Although this is not necessary in other locations in less eloquent brain, we frequently find it necessary in cavernomas of the brainstem, as indicated above, to get within the lesion, decompress it and gradually coagulate it away from the brain. This is very different from the strategy with cerebral AVMs.

The key to successful cavernoma surgery, then, is to avoid parenchymal damage in the trajectory toward the lesion. Because there is practically no “silent” brainstem parenchyma for the surgeon to traverse, we generally avoid surgery of deeply located brainstem cavernomas. Only when they come to the surface do we approach these lesions, and then the key is to choose an appropriate surgical approach to get us as close to the lesion as possible and to obtain an orthogonal view of the lesion. To this effect, CMs that reach the surface of the fourth ventricle are approached by a suboccipital craniotomy by either splitting the tonsils or, if they are more laterally located, through a velotonsillar approach. Whenever we operate in the floor of the fourth ventricle, we use facial nerve monitoring to be able to localize with precision the facial colliculus. Except for relatively small lesions that come to the floor of the fourth ventricle, we have been unable to remove large cavernomas of thepons through this approach without either producing a facial palsy or exacerbating a pre-existing palsy. Therefore, in patients with normal facial function, we are very reluctant to use this approach, and prefer to wait until the cavernoma has already produced significant facial weakness (Fig. 23.4). For lesions that come to the surface of the postero-lateral or lateral medulla or lower pons, we use the far lateral suboccipital approach. Higher pontine lesions that come to the surface laterally can be approached through a straightforward retromastoid cranietomy as used for microvascular decompression of the trigeminal nerve. More anterolateral pontine lesions can be approached through the presigmoid retrolabyrinthine exposure discussed above. For lateral mesencephalic lesions, we prefer the subtemporal transtentorial approach; however, the fourth nerve is encountered with this approach and must be preserved (Fig. 23.5). Lesions of the tectal place are best resected through the supracerebellar infratentorial exposure, an approach that can be modified to a more lateral exposure if necessary.18

Postoperative Outcomes

Because of their critical location, brainstem CMs represent a formidable neurosurgical challenge. The consensus among most neurosurgeons is that patients presenting with significant neurologic deficits should be treated surgically if the lesion comes to an “accessible” pial surface, such as the floor of the fourth ventricle or the lateral aspect of the brainstem. Patients with deeper lesions are generally monitored and surgically treated only if they exhibit severe progressive symptoms or if they develop permanent neurologic deficits that would result by default from surgery. This conservative attitude is justified because, as discussed previously, hemorrhages from these low flow lesions are rarely devastating and are usually confined by a gliotic plane, resulting in small increments of neurologic morbidity.

Several recent series have proven the feasibility of successful microsurgical resection of brainstem CMs.19–24 A retrospective analysis of 36 patients who underwent surgical management of brainstem CMs demonstrated postoperative complications including new cranial nerve deficits in 17, motor deficits in three, and new sensory complaints in 12 patients; postoperative Karnofsky Performance Scale (KPS) scores ranged from 80 to 100 compared with an average preoperative KPS score of 70.19 Outcomes for patients treated surgically and conservatively have been compared in retrospective studies. Of the patients treated surgically, approximately 84% had no or minimal deficits at follow-up. This compared favorably with the 66% of conservatively managed patients with a similar outcome.25

The encouraging results from the above series should be interpreted with caution and should not be regarded as an open invitation to resect all brainstem CMs. For example, a study comparing a group of surgical and nonsurgical patients with brainstem CMs showed that 38% of the surgical patients were disabled permanently after surgery, whereas 38% improved; none in the nonoperated group had worsened at 47-month follow-up. Only patients with multiple deficits and progressive neurologic deterioration improved with surgery.26 It is clear that successful surgical outcome depends on appropriate patient selection, optimal timing in relation to hemorrhage, careful surgical planning, meticulous surgical technique, and completeness of resection.
Fig. 23.4  Axial (A), sagittal (B), and coronal (C) contrasted T1-weighted MRI of a large pontine cavernous malformation in a 55-year-old woman presenting with left sixth and seventh nerve palsies and rapid worsening of ataxia and right-sided hypoesthesia. The cavernous malformation reaches the floor of the fourth ventricle. This lesion was resected completely through a midline suboccipital approach along with monitoring of the facial nerves, brainstem auditory evoked responses, somatosensory evoked potentials, and motor evoked potentials. The patient’s sixth and seventh nerve palsies remained unchanged after surgery.
Fig. 23.5  Axial T2-weighted (A), contrasted coronal (B), and contrasted sagittal (C) MRI reveals a large right pontomesencephalic cavernous malformation in this 30-year-old man presenting with complete right third nerve palsy and left spastic hemiparesis. The patient’s symptoms progressively worsened. (D) A gross total resection was achieved through a subtemporal, transpetrous approach. The patient’s hemiparesis was worse initially after surgery but gradually returned to the preoperative level.

References

IV

Miscellaneous
کتاب پزشکی

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Shunt Surgery for Posterior Fossa Lesions

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Numerous forms of treatment have been suggested and tried, but, as the number of methods indicates, they have been almost uniformly unsuccessful. The etiology being so obscure, any treatment is necessarily empirical and consequently unsatisfactory. Successful therapy must depend on the identification and the treatment of cause of the disease.

The above statement appears in “Internal Hydrocephalus: An Experimental, Clinical, and Pathological Study,” by Dandy and Blackfan, who studied hydrocephalus extensively in both clinical and experimental settings. Historically, the existence of a foramen (foramen of Magendie), leading from the fourth ventricle to the subarachnoid space, was suggested by Haller and Cotugno. In 1764, Cotugno first proved the existence of the subarachnoid space and also found fluid in this space in aquatic animals. Magendie made an important contribution to the understanding of cerebrospinal fluid (CSF) pathways by demonstrating in animals that fluid normally fills the ventricles and the subarachnoid space. He showed that a free communication exists between the ventricles and the subarachnoid space by means of the foramen, which was named after him. The central and spinal subarachnoid space forms a single freely communicating space. Magendie observed that the aqueduct of Sylvius or the foramen of Magendie was obstructed in several cases of hydrocephalus. The important issue of formation and absorption of CSF has been eloquently described by Key and Retzius and Dandy and Blackfan. Even today, the majority of their experimental results remain mostly valid.

Interestingly, the treatment proposed prior to the introduction of shunts for hydrocephalus in the early 1950s was to tackle the primary disease effectively. Present-day trends seem to consider this philosophy as well. Several studies have been performed to establish clear indications for ventricular shunts, and many are still trying to do so.

In 1881, Wernicke performed the first ventricular puncture and external CSF drainage. Quincke introduced lumbar puncture a decade later. Mikulicz reported the first ventriculoperitoneal shunt in 1893. A lumboureteral shunt was used successfully based on the concept of Heile, who sutured the renal pelvis to the dura and arachnoid. In 1918, Dandy introduced choroid plexectomy, and Torkildsen described ventricullocisternostomy.

Based on the pioneering work of John Holter and Eugene Spitz, valve regulated shunt systems were introduced, and the first such shunt surgery was reported in 1949 by Nulsen and Spitz. Several modifications and technological advances resulted in more than 130 different designs.

Posterior fossa lesions contribute to the development of the majority of hydrocephalus seen in younger patients. The lesions could be congenital, infective/inflammatory, or neoplastic. This chapter discusses the role of shunts in these different pathologies affecting posterior fossa structures (Fig. 24.1).

Dandy-Walker Cyst

In 1914, Walter Dandy and his pediatrician colleague Kenneth Blackfan described a group of congenital anomalies associated with cerebellar agenesis and a cystic lesion of the posterior fossa. Although Sutton described such an entity earlier, the details of this syndrome, demonstrated succinctly in autopsy specimens by Dandy and Blackfan, and later by Taggart and Walker, came to be known as the Dandy-Walker syndrome (DWS). The term was coined by Benda, who also pointed out that DWS is primarily a developmental anomaly of the fourth ventricle with membrane alterations and a cerebellar cleft. Benda also ruled out atresia of the foramina of Luschka and Magendie as originally put forth by Dandy. Gardner emphasized the disparity between the rate of formation and rate of egress of cerebrospinal fluid (CSF) from the neural tube at an early stage prior to the opening of the foramina of the fourth ventricle. DWS is characterized by cystic dilatation of the fourth ventricle, hypoplasia of the
cerebellar vermis, and hydrocephalus. The presence of a posterior fossa cyst with varying degrees of dysgenesis is essential for the diagnosis of this disorder (Fig. 24.2).

For treatment purposes, DWS can be similar to retrocerebellar arachnoid cyst (Blake's pouch cyst) and other variants, although the developmental origin differs. It is also noteworthy that hydrocephalus may not be an accompanying component of all cases of DWS.12,13

Dandy14 and Sahs15 advocated direct fenestration and communication of the cyst with the subarachnoid space, but with high mortality. The vast majority of these patients in later reports ended up with the placement of shunts.16,17 The primary mode of treatment is shunting. Various shunt procedures, such as ventriculoperitoneal (VP) shunt, cystoperitoneal (CP) shunt, and a combination of VP and CP shunts, are considered in the management options. Carmel et al18 suggested that a free communication exists between the cyst and ventricular system and proposed shunt placement in the lateral ventricle. They also proposed that the cyst-ventricle relationship may not remain constant throughout life, and that the free communication may be lost in the natural history of the disease as well as in the ventricular shunting procedure. For this reason, Raimondi et al19 advocated a combined ventricular and cyst shunt procedure (VP and CP shunts). Based on extensive manometric, radiologic, and pathologic exami-

Fig. 24.1 Axial (A) and sagittal (B) magnetic resonance imaging (MRI) scans of an arachnoid cyst, which is a more common entity in the posterior fossa, showing the compressed fourth ventricle and cerebellar verm. The cerebellar hemispheres are also seen to be normal but compressed on the ipsilateral side.

Fig. 24.2 Axial section of posterior fossa computed tomography (CT) scan showing a typical Dandy-Walker cyst. There is agenesis of the vermis, and major portions of the cerebellar hemisphere and the cyst may be in communication with the fourth ventricle.
nations, a Y connecting system was also recommended for equalization of pressures inside the ventricles and the cyst. This procedure found support from many others. In case of patency of cisternal communication, a cystoperitoneal shunt alone is suggested to be adequate because it drains both the cyst and the ventricles. The experience from the Hospital for Sick Children, Toronto, Canada, demonstrated that stenosis of the aqueduct comes as an acquired sequela because the interval between an initial VP shunt and the next CP shunt varied between 6 months and 15 years. Two separate earlier reports by Foltz and Shurtleff and Hawkins et al also pointed out that inserting a shunt into the lateral ventricle can lead to the development of an acquired aqueduct stenosis. Domingo and Peter suggested from their experience with 50 cases of posterior fossa cysts that CP shunts encourage normal flow of CSF through the aqueduct and consequently reduce the incidence of aqueduct stenosis.

Apart from stenosis of the aqueduct, several complications are reported with shunting procedures to decompress a Dandy-Walker cyst (DWC). Common complications include posterior fossa subdural hematomas, cranial nerve palsy with malpositioning of a shunt catheter, CSF leak, intracystic hemorrhage, and a blocked shunt. Lee et al reported complications related to the catheter placement due to injury to the fourth ventricle floor. One patient had a catheter tip in the brainstem. To minimize the complications resulting from catheter placement, Lee et al proposed a small 2 × 3 cm craniotomy inferior and medial to the junction of transverse and sigmoid sinuses. They also proposed that the placement of the catheter be done under ultrasound guidance. Montes et al advocated stereotactic transtentorial hiatus placement of a catheter. Recent studies have evaluated the role of endoscopic third ventriculostomy in selected cases of DWC.

**Case Illustration**

A 41-year-old man presented with headache, dizziness, and balance problems. He had a past history of hypertension and trauma to the head that was followed by worsening headaches 3 years ago. Neurologic examination revealed ataxia. Magnetic resonance imaging (MRI) showed a large DWC with a small cerebellum and absent/rudimentary vermis. He underwent surgery for resection of the cyst through a suboccipital craniectomy. An incision on the midline of the occipital region down to C2 was made, and under microscopic guidance an arachnoid cyst was drained completely. There was no postoperative complication, and the patient was discharged 3 days later. He had good relief of his symptoms and was able to ambulate well at the time of discharge. Two-year follow-up showed no neurologic deterioration or recurrence of symptoms other than mild headaches. It is very rare to have DWC presenting in adulthood.

**Trapped Fourth Ventricle**

When there is a proximal obstruction at the cerebral aqueduct and distal obstruction at the foramina of Magendie and Luschka, the fourth ventricle is trapped and CSF production by the choroid plexus results in progressive dilatation.

The fourth ventricle can be isolated in up to 17% of children with VP shunts who have had previous intraventricular hemorrhage, inflammatory processes, infection, previous intraventricular hemorrhage, or neoplastic disease (carcinomatous meningitis). The pressure gradient created by lateral ventricular shunts across the aqueduct and congenital malformations can result in obstruction of the aqueduct and fourth ventricular foramina. Progressive dilatation of the fourth ventricle results from CSF production by the choroid plexus or by a ball-valve one-way mechanism through the aqueduct. CSF diversion procedures for this complicated entity include shunt placement (transcerebellar, transaqueductal, transforminal across the foramen of Magendie and the transcortical transtentorial hiatus) in various directions. Recent advances have proposed endoscopic placement of catheters/stents, aqueductoplasty, and internal shunting procedures. As in the case of a DWC, a fourth ventricular shunt alone can be placed if imaging studies demonstrate that the aqueduct is closed, or a single catheter in lateral ventricle in the case of a patent aqueduct. However, the risk of upward or downward herniation exists with both types of shunting procedures, along with failure of either system, necessitating placement of the second shunt system. Raimondi’s Y system can help in equalization of the pressures, although not without the risk of shunt malfunction.

The midline transvermian and lateral transcerebellar approaches have been used for catheter placement. Intraoperative ultrasound guidance is useful in placement of the catheter inside the cyst. However, after decompression, as the brainstem and cerebellum expand, there is a risk of brainstem injury with the catheter tip and resultant focal neurologic deficits. An enlarged fourth ventricle can be tapped from supratenorial entry either by the transcortical transtentorial hiatus route or the transventricular transaqueductal method. Regardless of the approach, complications and revision rates remain high.

The transventricular transaqueductal approach is attractive because the catheter fenestrations can be fashioned as to drain both the lateral and fourth ventricle simultaneously. This also would synchronize the pressures inside these compartments. The transcortical transtentorial hiatus approach is appropriate for those large cysts that reach cranially across the tentorial hiatus into the supratentorial compartment. Both of these techniques require interactive digital imaging techniques and/or endoscopy.

Endoscopic techniques help in opening up the aqueduct by performing aqueductoplasty, placement of stenting along with third ventriculostomy, or shunt placement. The reported complication rate in this series was 25%. Third ventriculostomy or aqueductoplasty alone without a shunt also had failures and complications.

Aqueductoplasty is reported to have reclosure, requiring additional treatment. Stents also have been used in small numbers, and, like shunts, they are vulnerable to infection, migration, and obstruction. The management results and complications of a trapped fourth ventricle are similar to those of a DWC, although with a much wider range.
Neoplastic Lesions of Posterior Fossa

Tumors of the central nervous system (CNS) are the most common solid neoplasms found in children, and the majority of these tumors are infratentorial. The proximity to the fourth ventricle and thus to the CSF pathways predisposes these children to the development of obstructive hydrocephalus. At some point, these children require a CSF diversion procedure during the course of their illness.

Since the report published by Abraham and Chandy describing ventriculoatrial (VA) shunts in the management of posterior fossa (PF) tumors with hydrocephalus, many centers utilized either VP or VA shunts to reduce intracranial hypertension in PF tumors with hydrocephalus. Because VA or VP shunts are not devoid of problems, or more appropriately have high rates of malfunction and revisions, the routine placement of shunts has come under criticism. Major complications, including shunt malfunction, infection, migration, tumor hemorrhage, tentorial herniation, and a variety of abdominal complications, necessitated a review of the indications for routine shunt surgery in PF tumors. The American Society for Pediatric Neurosurgery performed a survey in 1985 on precraniotomy shunts and found that there was no clear evidence of advantage in routine shunting before PF tumor surgery.

A recent conference of experienced pediatric neurosurgeons reported that hydrocephalus existed in 80% of patients with PF tumors, and 25 to 30% required postoperative treatment for persisting hydrocephalus. However, there was no consensus on the way hydrocephalus should be treated before, during, or after the posterior fossa surgery. Undoubtedly, there is a group of patients with PF tumors requiring CSF diversion, but there are no reliable parameters to identify this group. The incidence of progressive postoperative hydrocephalus has been reported to be between 15% and 45% of patients with PF tumors, with an even higher incidence among children younger than 3 years of age. The addition of external ventricular drainage (EVD) either preoperatively or intraoperatively (continued postoperatively in some patients), endoscopic third ventriculostomy, and placement of subcutaneous reservoir have increased the available alternatives to a permanent shunting procedure.

Culley et al. did a retrospective analysis of 117 pediatric patients with PF tumors to identify the factors that determine the need for VP shunts. Among the various factors studied, younger age, midline tumors, subtotal tumor excision, prolonged EVD placement, cadaver dural grafts, formation of pseudomeningocele, and CSF infection were statistically significant predictors for the need for postoperative shunt placement. This study recommends total removal of tumors, especially those of the midline, with meticulous closure of the wound, prevention of postoperative CSF leak or infection, and the avoidance of cadaver dura graft for closure. Another retrospective study of 180 children with PF tumors confirmed similar factors that predispose to persistent postoperative hydrocephalus. Only 12 out of 180 patients (6.7%) required direct primary postoperative shunts within 6 postoperative weeks, eight patients (4.4%) were shunted due to recurrence of tumor, and eight more (4.4%) had late shunt surgery. This 15.5% shunt insertion rate appears to be lower than the 17 to 40% rate reported in the literature. This also suggests that a routine endoscopic third ventriculostomy may not be required in the majority of these cases. In another study, 29% of the patients with PF tumors required ventriculostomy, and 21% required a permanent shunt.

Endoscopic third ventriculostomy is gaining popularity in the management of hydrocephalus with posterior fossa lesions. Tamburrini et al. reported a 90% success rate with endoscopic third ventriculostomy. Morelli et al. reported an 81% success rate in controlling hydrocephalus with endoscopic third ventriculostomy.

Case Illustration

A 2-year-old girl was referred to us with the diagnosis of pilocytic astrocytoma. She underwent resection of the tumor and placement of a ventriculoperitoneal shunt. Three years later she had a recurrence of the tumor, which was again operated on. When she was 9 years old she presented with headache and visual problems. Neurologic examination revealed no deficits. An MRI scan showed a large posterior fossa cyst. She underwent a suboccipital craniotomy and resection of the tumor. The postoperative course was uneventful and she was discharged without any new neurologic deficit.

Nine months later she developed severe headaches along with nausea and vomiting. Physical and neurologic examinations were normal as before. An MRI scan showed a small cystic lesion with the possible diagnosis of recurrence. After a suboccipital craniotomy, microsurgical resection of the tumor was performed. Five-year follow-up showed no recurrence of tumor based on clinical and imaging exams.

Conclusion

In general, most reports suggest that younger child with a midline tumor (medulloblastoma or ependymoma) and preoperative dilated ventricles is a candidate for preoperative CSF diversion procedure and might require a permanent shunt placement in due course.
Fig. 24.3 Preoperative MRI scans in axial (A) and sagittal (B) views showing a large mixed-density lesion in the midline posterior fossa. The lesion is predominantly low density. Because of its midline position and compression of the cerebrospinal fluid (CSF) outlet, a ventriculoperitoneal shunt was placed initially. (C) Postoperative contrast-enhanced MRI reveals a residual tumor and posterior fossa pneumocephalus. The fourth ventricle is opened up.

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Following accidents, the leading cause of death in children is cancer. The most common solid cancer in childhood is medulloblastoma. That being said, the incidence of brain tumors in children is 2.6 to 4 per 100,000.1 It is thus no wonder that the average pediatrician, seeing 40 to 60 patients per day in his or her office, may overlook the possibility of a brain tumor in the child who presents with vomiting or dizziness early in the clinical course. In fact, the average child diagnosed with a brain tumor has been to three physicians and has been symptomatic for 6 months by the time the diagnosis is made. In an unpublished review of our earlier series, the author found that 24% of children diagnosed with a posterior fossa tumor had had an upper gastrointestinal (GI) series as part of their workup for vomiting prior to the diagnosis of a brain tumor.

Half of the brain tumors found in childhood occur in the posterior fossa, with medulloblastoma, juvenile pilocytic astrocytoma (JPA), and ependymoma being the big three.2 Multiple reviews of outcomes of children with posterior fossa tumors have demonstrated that, regardless of histology, the extent of resection is the most important predictor of outcome. In trials of ependymoma and medulloblastoma, a gross total resection or near-total resection doubles the 5-year survival of the child compared with a lesser resection.3–5 Thus, the work we do as neurosurgeons is more important to the survival and the quality of the survival of the child with a posterior fossa tumor than anything the radiation oncologist or the medical oncologist can add to the care. There are also studies showing that a fellowship-trained pediatric neurosurgeon is more likely to obtain a gross total resection of a child’s posterior fossa tumor than a generalist. It is paramount that the neurosurgeon who decides to operate on a child’s posterior fossa tumor be prepared to completely excise the tumor if feasible. To do so safely requires not only a skilled neurosurgeon but also a skilled pediatric anesthesiologist and skilled pediatric intensivists to assist with postoperative care. If this team is not available, it is better to refer the child to a specialty center. Fifty percent of the children referred to us with posterior fossa tumors have undergone inadequate surgery and have to undergo a second posterior fossa surgery to optimize their outcome.

Whereas in years past there were few textbooks to assist the neurosurgeon in preparing for surgery in the infant or child, there are now several good reference texts that can be referred to for specific tumors and their management.6–9

**Clinical Presentation**

The mean age of presentation of children with medulloblastoma or ependymoma is 5 years or less, whereas the mean age of presentation of the child with a juvenile pilocytic astrocytoma is 9 years. Furthermore, children with malignant brain tumors are more likely to present with weight loss and constitutional symptoms as compared with children with benign tumors. Remember that a 2- to 4-pound weight loss in a 5-year-old may represent 10 to 15% of the body mass and is significant. Finally, children with malignant tumors are generally symptomatic for a shorter period of time than are children with benign tumors. The exception to this is the child with a large cystic component to a pilocytic tumor or a hemangioblastoma, as these cysts can sometimes enlarge at a rapid rate.

Infants with posterior fossa tumors present with a tense fontanelle and progressive head growth. It is surprising how often a head crossing percentiles on a growth curve is the only clinical clue of an underlying posterior fossa tumor. As the tumors progress, the infants become irritable, have associated vomiting, and may have altered sensorium. They may at times present in extremis, with decerebrate posturing or coma.

Once children reach an age where they can communicate and ambulate, they typically present with morning headaches
and vomiting and gait ataxia. They are commonly referred to the ear, nose, and throat (ENT) department for evaluation of vertigo or labyrinthitis. Papilledema and the inability to walk tandem are common clinical signs. If the vermian tumor is eccentric, they may have dysdiadochokinesia as well. Our practice is to image the entire spine preoperatively in children with posterior fossa tumors, as postoperative imaging of the spine may be artifactual for weeks thereafter. In the child who has back pain, tenderness, or hyperreflexia, consideration of drop metastases from a malignant tumor should be entertained. Similarly, if the child with a posterior fossa tumor presents with seizures, cortical metastases or leptomeningeal disease should be considered.10

◆ Imaging

At the present time, there is no imaging modality that can predict the histology of a posterior fossa tumor with certainty. We have seen densely enhancing nodules associated with large cysts that were presumed to be pilocytic nodules and yet at surgery turned out to be hemangioblastomas (Fig. 25.1). We have seen multicystic tumors that we thought were JPAs but proved to be medulloblastomas. We have seen tumors with calcifications that we thought would be ependymomas turn out to be pilocytic astrocytomas. Having said this, there are some clues that can help predict what the tumors will most likely turn out to be. Our experienced neuroradiologists are able to predict tumor histology in nine out of 10 cases. Tumors that appear to be hyperdense on noncontrast computed tomography (CT) are most likely to be primitive neuroectodermal tumors (Fig. 25.2), whereas the ependymomas and astrocytomas are hypodense on noncontrast CT. A large vermian tumor that extends out of the foramen of Magendie or extends into the cervical spinal canal is more likely be an ependymoma (Fig. 25.3). Likewise, a tumor of the fourth ventricle that grows out the foramen of Luschka and fills the cerebellopontine angle is likely to prove to be an ependymoma (Fig. 25.4).11

◆ Preoperative Management

Although these children present with ventricular obstruction and symptoms of raised intracranial pressure, the vast majority can be observed in an intensive care setting and do
cases in which a child declines neurologically and has to be operated on emergently, but such cases are uncommon. Most children with posterior fossa tumors do not require permanent shunts. In our experience, only a fourth of the children with posterior fossa tumors ultimately require ventriculoperitoneal shunts. The use of external ventricular drainage is determined at the time of surgery based on the turgor of the dura after the craniectomy. Again, the majority of children do not require external ventricular drainage even though the ventricles may appear quite enlarged on preoperative imaging.

◆ Operative Management

Several age-dependent factors enter into the decision making of positioning, anesthesia, and postoperative care. They are discussed in the following subsections.

Anesthesia

The greatest challenge of tumor surgery in children younger than 2 years of age is that of blood loss. The circulating blood volume of a young child is estimated at 70 cc per kilogram body weight. Loss of more than 1.5 blood volumes runs the risk of a coagulopathy. The anesthesiologist and the surgeon must both pay close attention to blood loss, particularly that hidden under the drapes or in drainage bags. The anesthesiologist should begin replacement early when it becomes apparent that a transfusion will be necessary. Washed red blood cells are less likely to cause intraoperative problems with hyperkalemia in the child requiring large volumes of blood. In children with malignant tumors who have been receiving chemotherapy, irradiated red blood cells may be given to reduce the likelihood of viral transmission to a compromised host.

For tumors of the cerebellar vermis, the surgeon should, just after opening the dura, dissect out the cerebellar tonsils, identify each posterior inferior cerebellar artery (PICA) as it loops below the tonsils, and dissect them rostrally until the vermian branches of the PICA are identified. These branches give the main blood supply to vermian tumors and can be ligated early in the procedure as they enter the tumor, thus dramatically reducing the vascularity of the tumor.

Positioning and Fixation

Most pediatric neurosurgeons have stopped placing children in the sitting position, as the Concorde (prone, neck flexed) position offers just as good an exposure with greater surgeon comfort and little risk of air embolism (Fig. 25.5). That being said, posterior fossa surgery in children younger than 2 is usually done with the patient face down on a horseshoe headrest, as the risk of pin fixation in the young skull is obviated. Because of this, the risk of pressure sores on the malar prominence or forehead requires meticulous attention. The authors place Rest-on Foam (3M Corporation, St. Paul, MN) over the face, with the adhesive side to the skin. This helps pad
the face and also serves to anchor the endotracheal tube into place. For children over 3 years of age, the pediatric Mayfield pins are utilized but tightened to only 40 pounds of pressure until the pins grab the outer table of the skull.

Approaches

Although medulloblastomas can be found within the cerebellar hemisphere, most arise from the roof of the fourth ventricle, pushing downward into the ventricle; 35% invade the brainstem, often at the obex or the floor of the fourth ventricle. This can often be identified preoperatively by close inspection of the magnetic resonance imaging (MRI). Alternatively, upon exposing the cerebellum, the microscope is brought into the field and the cerebellar tonsils separated as the vermic branches of the PICA are dissected out. This approach facilitates concomitant inspection of the floor of the fourth ventricle. If it is invaded by tumor, caution must be taken in manipulation of the tumor as it is debulked, as this could lead to a “floor of the fourth syndrome” which includes an ipsilateral palsy of cranial nerves VI and VII and a contralateral hemiparesis.

Juvenile pilocytic astrocytomas arise from within the cerebellar hemispheres and are usually separated from the ventricle by the ependyma. On some occasions, however, they also invade the floor of the fourth ventricle. A variant of the cerebellar astrocytoma actually arises from the brainstem and is dorsally exophytic into the ventricle or out laterally into the cerebellopontine angle. Such tumors exit the brainstem, pulling functional tissue up with them much like the sides of a volcano. The inexperienced surgeon may be inclined to shave these tumors off flush with the floor of the ventricle or with the side of the stem, inadvertently injuring the brainstem in the process.

Ependymomas, by definition, take origin from the walls of the ventricle. They must carefully be debulked as the capsule is dissected away from neural tissue. Those arising from the floor of the fourth ventricle derive blood supply from multiple small perforating vessels arising from the brainstem. These vessels must be meticulously coagulated and cut, as avulsing them may cause them to retract and bleed into the brainstem. If they do, they should not be pursued. Gentle suction with a regulated suction, gentle irrigation, and time will allow them to stop oozing without injuring the brainstem.

A rare variant of the ependymoma arises from ependymal rests at the outer margin of the foramen of Luschka and grows out the foramen into the cerebellopontine angle as well as into the fourth ventricle. These tumors often grow to be quite large before obstructing the ventricle and causing hydrocephalus, their most common presenting symptom. By this time, these tumors often encase the lower cranial nerves as well as the vertebrobasilar complex, and may have invaded the side of the pons. This tumor is one of the most formidable posterior fossa tumors for the surgeon. Ependymomas have a predilection to grow out the porus acusticus or jugular foramen. To approach these tumors requires a modified Concorde position (Fig. 25.5) with the infant’s chin tucked and turned over to the shoulder as the infant is flexed. The skin incision begins midline but curves up behind the ear on the involved side. This allows bony removal across the midline, up to the torcular Herophil and around to the sigmoid sinus of the involved side. By gently elevating the involved cerebellar hemisphere and opening the telovelar space (cerebellomedullary fissure), one can dissect out the entire tumor and accomplish a gross total resection. Over half of these children require temporary tracheostomies and gastrostomies, but our experience has been that most of them can be decannulated by 6 months postoperatively.

In caring for children with phakomatoses, such as von Hippel-Lindau disease, one must recognize that the child will likely have recurrent disease elsewhere in the posterior fossa over time. As such, the surgeon performing the skin incision and approach to a single tumor should keep in mind the need for other approaches over time, as new tumors arise. One of three entries into the posterior fossa will, over time, allow access to the entire posterior fossa (Fig. 25.6).

Intraoperative Monitoring

Space limitations preclude a detailed discussion of intraoperative monitoring. The interested reader should consult an excellent study by Sala et al. For the purposes of our discussion, it is important to mention that intraoperative monitoring tends to cause the surgeon to leave more tumor behind. Recognizing that the most important predictor of survival in pediatric posterior fossa tumors is a gross or near total resection, the neurosurgeon relying on intraoperative monitoring...
must still accomplish this goal or the tumor will likely progress and the child will die.

◆ Postoperative Management

Imaging

It has been our practice to inform the parents preoperatively that the child will undergo a postoperative MRI within 48 hours of surgery, and that if this scan demonstrates any resectable residual tumor, the child will be returned to the operating room to remove that remnant. At times these scans may be equivocal, at which point review of the intraoperative video may be useful.

 Syndrome of Inappropriate Secretion of Antidiuretic Hormone

All pediatric patients experience a drop in their serum sodium the night after surgery. The exception is the child who develops diabetes insipidus and is volume depleted. Most of the time the sodium level does not drop below 132 mg%. This drop must be closely monitored, as in some children the sodium level drops below 125 mg%, which may precipitate a seizure. Such a seizure may cause bleeding into the fresh wound cavity.

Hydrocephalus

Eighty percent of children with posterior fossa tumors have hydrocephalus at the time of presentation. Most will improve with 24 to 48 hours of intravenous steroids and will not require cerebrospinal fluid (CSF) diversion. In the past, some authors have advocated shunting at presentation and elective tumor resection. St. Rose et al.13 have advocated endoscopic third ventriculostomy at presentation. Most of the time, CSF diversion, either temporary or permanent, is not required and the hydrocephalus resolves if the tumor is removed. This may take several days to resolve, however, and the patient must be monitored carefully. In our experience, about one fourth of children ultimately require a shunt for unremitting hydrocephalus.

Swallowing

In instances such as the dorsally exophytic brainstem tumors or the cerebellopontine angle ependymomas, in which the tumor invades the inferior floor of the fourth ventricle or involves the lower cranial nerves, the children are at risk for aspiration pneumonitis in the acute postoperative period. In these instances, it has been our practice to keep the children intubated and sedated overnight following surgery. On the day after surgery, if the child is wide awake, the ENT team is present in the intensive care unit and inspects the vocal cords and pharyngeal motility by fiberoptic endoscopy at the time of extubation. Children with an abnormal examination are maintained on nasogastric feeds and receive nothing by mouth until a formal swallowing study can be performed. For those with vocal cord paralysis or insensate pharynges, we have been quick to recommend tracheostomy and gastrostomy. This routine has prevented all but one death in affected children. That death occurred in a child who was ambulatory following resection of a dorsally exophytic brainstem tumor arising at the obex. She had a tracheostomy and gastrostomy and was in rehabilitation when her parents took her home for a weekend pass. She convinced them to let her drink a soft drink, at which time she aspirated, promptly arrested, and died.

Posterior Fossa Syndrome

One of the earlier reports of posterior fossa syndrome was by Wisoff and Epstein.14 It is a delayed syndrome that occurs 12 to 72 hours following resection of a posterior fossa tumor. The syndrome appears to be a severe dyspraxia in a child who initially awakens from surgery speaking and moving well, but then becomes mute, dystonic, is irritable, and has cognitive impairment. Many have trouble swallowing and have trouble with visual pursuit. The cause of the syndrome is poorly understood. Postoperative imaging is rarely helpful. Some authors have theorized that the syndrome develops from ischemia or edema of the deep cerebellar nuclei. Our current theory, given that it seems to occur most commonly after resection of large midline tumors, is that it is due to sudden decompression of the stretched cerebellar peduncles. The syndrome has been said to occur in 5 to 24% of children following posterior fossa tumor resection. Earlier authors felt that the syndrome always resolved within 6 months or a year of surgery. More recent studies have demonstrated that all of these children have long-term cognitive impairments and emotional problems.15 The posterior fossa syndrome is now
recognized as the most common cause of long-term impairment following posterior fossa tumor surgery in children.

◆ Conclusion

Removal of a posterior fossa tumor in a child is one of the most rewarding operations a neurosurgeon can perform. The children present deathly ill, and generally are discharged in good condition. For those with a JPA, the children are cured and can lead a normal life. Even for those with medulloblastoma or ependymoma, major advances have occurred in therapy over the last decade such that 5-year progression-free survival has improved from 35% to 75%. Many of the cognitive impairments seen a decade ago can now be avoided, so that the survivors have a decent quality of life.

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