Endoscopic and Keyhole Cranial Base Surgery

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To my wife, Deborah; my daughters, Sophia and Vivienne; my parents, Claire and Leonard Evans; and my parents-in-law, Barbara and Ronald Ahern: I greatly appreciate your endless support. You have all made my endeavors, as well as my entire life, more enjoyable and meaningful.

To my fellows and trainees: You have provided me the wonderful gift and great satisfaction of being able to share the intricacies of cranial base surgery with new, enthusiastic individuals each year. I am always impressed with your dedication, creativity, and passion to learn.

JJE

For Sarah, to whom I owe everything.

TJK

To my wonderful wife and children and to my mentors, colleagues, and patients, who have helped me to become a better, more thoughtful surgeon.

CJF

To Suchetha, my patients, and my mentors, all of whom inspire me to be my best. And to Professor Rhoton, whose work and character ignited my interest in cranial base surgery.

VRK

Foreword

Skull base surgery has its origins as an independent subspecialty of neurosurgery and otolaryngology in the late 1980s and early 1990s. In the early years, the emphasis was on the development of approaches providing wide exposure of difficult lesions to allow for complete resection, proper reconstruction, and good cosmetic and complication-free outcomes. Many of the approaches were centered on the concept that expanded bony removal allowed for tumor resection with minimal brain retraction or damage to surrounding neurovascular structures. In an effort to minimize damage to the brain, the approaches became “maximally invasive” in the eyes of some but could be accomplished with minimal morbidity and excellent outcomes in the hands of surgeons with subspecialized training. In this way, skull base surgery grew into a field of its own.

As part of the natural evolution of the field, the emphasis has shifted over the last 10 years to try to do more with less. The movement in favor of “minimally invasive neurosurgery” can perhaps be thought of more aptly as “minimal access neurosurgery,” since the purpose is to accomplish the same goal through a smaller opening. At the heart of this movement is the use of the endoscope as a primary means of visualization instead of the established workhorse of neurosurgery, the operating microscope. Endoscopic skull base surgery is still evolving, with newer technologies being applied and with the results and complications being evaluated in real time.

It is important to remember that, as with any evolutionary process, both the paths and the outcomes are rarely ever binary. While it is tempting for all of us to try to directly compare endoscopic skull base surgery to open skull base surgery and decide which is “best,” we must constantly remind ourselves that the answer will never be absolute. In fact, the very question of open versus endoscopic approach represents a spectrum of possibilities, with fully endoscopic procedures at one end and classic open approaches at the other. In between, there are many shades of gray that include keyhole surgery (surgery through a small craniotomy), endoscope-assisted microsurgery, and, most recently, exoscopic 3-dimensional visualization surgery.

There are many questions facing the field currently:

- What are the real benefits and risks of these approaches, in comparison to one another?
- What are the short-term morbidities (e.g., duration of hospital stay, number of visits to the doctor’s office, time to return to full-time work, etc.)?
- What are the total costs of treatment including adjunctive therapies such as radiosurgery, and what are the costs of long-term surveillance?
- Is there any effect on tumor recurrence rates?

These are all legitimate questions that surgeons must contend with when crafting a treatment plan. All of these questions would be best answered by a randomized trial, which is unlikely and in many cases impossible. As such, we are dependent on data that, in many cases, are difficult to be free from bias: institutional case series, meta-analyses, or registries. More practically, we must ask ourselves the relative merits and pitfalls of the various approaches with regard to the individual patient and lesion in front of us on any given day. A critical concept is that the various endoscopic, keyhole, and open approaches can be complementary and very effectively used in combination. The well-trained modern cranial base surgeon will have all of these approaches in his/her armamentarium as well as the knowledge of when and how to employ them.

In this exciting book*, Endoscopic and Keyhole Cranial Base Surgery,* the editors – Drs. Evans, Kenning, Farrell, and Kshettry – have gone through great efforts to synthesize contemporary knowledge in the field in order to provide a guide for surgeons who are trying to make these decisions. This work presents the basic concepts and techniques of neuroendoscopic and keyhole surgery, and then goes on to provide a point-counterpoint type treatment of various skull base targets and complex cranial lesions. The management and treatment of various lesions ranging from benign and malignant skull base tumors to intraventricular lesions to aneurysms are discussed from three perspectives: endoscopic transnasal approaches, keyhole transcranial approaches, and classical open transcranial approaches. This book is very well illustrated, and all the chapter authors are well known in their fields. As a summary, the editors have provided their own perspectives on the comparison of the various approaches.

Our advice to young surgeons at the beginning of their journey would be to get the best training possible, but also to learn continuously by watching other master surgeons in order to hone one's own skills and further develop one's own surgical instincts. While there are many changes occurring in the field, it is most important for a surgeon not to become completely dependent on one technique, but rather to become facile with the full spectrum of approaches so as to be able to choose the one that best fits the situation. Neuroendoscopic and keyhole cranial base approaches have undoubtedly revolutionized our field, but we would urge the younger generation not to forget the value of open approaches in the right context. In both directions, it is vitally important for us to distinguish between *being able* to do something in a certain way and whether we *should* do it that way. This important text addresses some of these broader questions, and this discussion is one that will (and should) continue indefinitely.

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Preface

Cranial base surgery is considered one of the most challenging surgical disciplines because of the difficult access to pathology that is invariably surrounded by critical neurovascular structures. Not only can the pathological processes cause cranial nerve, vascular, and brainstem dysfunction, but the cranial base approaches themselves may also be associated with significant morbidity from the compromise of surrounding normal and uninvolved structures. Furthermore, these invasive approaches often create defects that require extensive repair to prevent cerebrospinal fluid leakage, infection, and cosmetic deformity.

More recently, minimally invasive approaches to the brain and cranial base have been developed. Ideally, these approaches employ smaller incisions, natural pathways, and more limited craniotomies, with the hope of creating less approach-related morbidity while allowing optimal management of the pathology. The modern cranial base surgeon is faced with more options than ever, including the classical "open" approaches as well as a wide range of rapidly evolving keyhole and endoscopic procedures.

Although minimally invasive cranial base approaches have been described previously, this unique text provides a critical evaluation and comparison of these surgical techniques for accessing specific intracranial pathologies and anatomical targets. The text is divided into three sections designed to evaluate the relative merits and limitations of the open, keyhole, and endoscopic cranial base approaches. The first section of this book details endoscopic endonasal principles, anatomy, and specific approaches. The second section focuses on general keyhole surgery principles and specific keyhole procedures. The third section provides a critical review of modern cranial base surgery, with highly experienced surgeons presenting the open, keyhole, and endoscopic endonasal approaches to specific target pathologies. At the end of each chapter in this important *target-based* section, the authors discuss the management of a specific case example utilizing their surgical approach. This novel format offers a comprehensive overview of the various cranial base approaches and gives perspective into the expert's thought process behind utilizing a particular approach. The goal is to provide the reader with sufficient information to help select and tailor the optimal surgical approach to a specific cranial base pathology or anatomical target.

We have brought together the global leaders in open, keyhole, and endoscopic cranial base surgery in order to create a comprehensive resource for novice and experienced surgeons involved with the treatment of these difficult cases. We are extremely grateful to each of our contributing authors, all renowned experts in the field, for enthusiastically accepting our invitation. We also would like to extend a special thanks to Dr. Alan Siu for his efforts. Finally, we are indebted to Springer Science and their superb editorial staff for the guidance and assistance in producing this text. We hope that this will be a valuable resource to those involved in treating patients with disorders of the cranial base.

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Part I

Endoscopic Endonasal Surgery

1 Principles of Endoscopic Endonasal Surgery

Tyler J. Kenning, Varun R. Kshettry, Christopher J. Farrell, and James J. Evans

1.1 Introduction

Endoscopic endonasal surgery has revolutionized the field of cranial base surgery; however, these surgical approaches involve complex anatomy and require a unique set of skills which its practitioner must master to optimize their use. There is a steep learning curve with the techniques as the endoscopic endonasal approach (EEA) represents a crossroads between the rhinologic principles of otolaryngology and traditional cranial base surgery familiar to specialized neurosurgeons. As a result, EEA is best practiced as a joint venture between the disciplines. This chapter will focus on many of the principles of endoscopic endonasal surgery and the considerations necessary for its successful application to cranial base pathology.

The EEA has expanded from being used for purely sellar pathology to addressing lesions from the frontal sinus to the upper cervical spine and from the midline to the lateral aspects of the infratemporal fossa (Fig. 1.1). When applied to intradural pathology, the endonasal corridor is largely utilized to target ventral median and paramedian structures. Intradural pathology that extends lateral to the cranial nerves and internal carotid arteries is typically best approached through more "traditional" or "keyhole" open techniques, although these can be combined with extended endonasal approaches.

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1.2 Anatomy

The hallmark of any successful surgeon is proficiency with pertinent anatomy. As the sinonasal cavity is familiar territory to the rhinologist but typically less so to the traditional neurosurgeon, its use as a surgical corridor can appear daunting to those with less experience with sinonasal anatomy. Similarly, the intracranial space, as the purview of the neurosurgeon, can seem very foreign to the otolaryngologist. Again, we believe this further supports our assertion that EEA should be undertaken by a surgical team, consisting of both a neurosurgeon and a rhinologist in order to achieve optimal outcomes. The anatomy of the endoscopic endonasal cranial base approaches that must be mastered, as well as the many anatomical variants that can exist, is extensively covered in Chap. 2.

1.3 Patient Positioning and Surgical Planning

As the EEA addresses ventral lesions, patients are placed supine with the head in a neutral position. We prefer to use a navigation system (when necessary) that does not require rigid pin fixation of the head. This allows for easy repositioning of the head during surgery as well as prevents excessive pressure against sinonasal structures by allowing some slight movement if too much force is applied. When accessing pathology of the anterior cranial base, a varying extent of head extension is recommended, while slight flexion is helpful for clival and craniocervical junction lesions. The operative table should be placed in reverse Trendelenburg to enhance venous drainage and minimize bothersome oozing from the sinonasal mucosa of nasal structures. While some surgical teams will place the assistant driving the endoscope on the same side of the table as the operating surgeon

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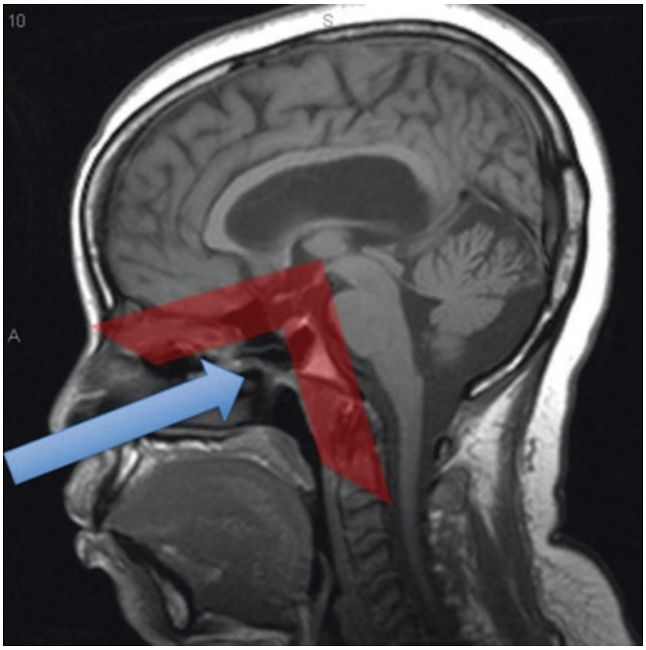

Fig. 1.1 Area of the cranial base able to be surgically accessed by the endonasal endoscopic approach

Fig. 1.2 Surgeon orientation on opposite sides of the patient's head for endonasal endoscopic surgery

(typically the right side for right-handed surgeons), we prefer that the neurosurgeon and the otolaryngologist stand on opposite sides of the patient's head so that they may work simultaneously in an ergonomic fashion (Fig. 1.2). This surgeon positioning is quite versatile as the operating surgeon can stand on either side of the table depending on his/her handedness or on the laterality of the pathology while the assistant driving the endoscope stands on the opposite side. The anesthesia team is placed at the foot of the patient as the surgeons, navigation system, and endoscopic monitors occupy the space at the head of the bed.

The cranial base exposure, opening, and access to the pertinent pathology must be performed with the closure in mind to minimize the risk of persistent CSF fistulas postoperatively. The various closure techniques and materials, both autografts and allografts, available are detailed in Chap. 3. Abdominal or thigh incisions can be prepared for fat, muscle, and fascia lata grafts, when appropriate, and should be planned for, both in terms of patient positioning and setup of the operative field. Although lumbar drainage is utilized at some centers for postoperative management of high-flow cerebrospinal fluid leaks, we do not feel they are necessary when a well-tailored, multilayered cranial base repair can be achieved. The exception to this rule is in patients with idiopathic intracranial hypertension and an associated cranial base CSF fistula or meningocele requiring repair. In these patients, we utilize lumbar drainage as a bridge until permanent CSF diversion (e.g., ventriculoperitoneal shunt) can be performed to treat the elevated intracranial pressures a few days after the cranial base repair.

1.4 Endonasal Access and Limitation of Postoperative Morbidity

Just prior to introduction of the endoscope, the nasal cavity is decongested with cotton pledgets soaked in 1:1000 epinephrine or oxymetazoline. Injection of the turbinates and nasal septum with 1% lidocaine with 1:100,000 epinephrine can also further reduce intraoperative blood loss. Antibiotic coverage is provided with a third-generation cephalosporin for purely sinonasal and sellar pathology, while the addition of vancomycin can be used for extended cases where there will be significant disruption of the subarachnoid space. The evidence for enhanced antibiotic coverage, however, is lacking and is typically employed at the preference of the surgical team with the consideration for preoperative bacterial nasal colonization assessment.

Typically, binarial access is utilized, although access through a single nare can be performed for more focal lesions such as pituitary adenomas and meningoencephaloceles. In addition to addressing the pathology and repairing any resultant cranial base defects, the goals of endonasal surgery should emphasize minimization of postoperative morbidity while maintaining normal sinonasal function. The nasal mucosa is highly vascularized and allows for rapid healing, which can be tremendously helpful to the endoscopic surgeon in terms of repair of the cranial base but also may promote the formation of postoperative adhesions and discomfort.

Preservation of normal sinonasal structures when possible, including all turbinates, and minimizing posterior nasal septectomy will avoid excessive cautery for hemostasis and the resultant significant nasal crusting postoperatively. In general, we follow the premise of preserving all structures unless they are directly involved by the pathology. We prefer to use a limited amount of absorbable packing for support of cranial base grafts and flaps and avoid the use of nonabsorbable packs or Foley catheter balloons. Finally, at the conclusion of the procedure, silastic or gelatin sheets are placed between the nasal septum and middle turbinates and potentially even in the middle meatus to limit the development of postoperative synechiae.

Starting on postoperative day 2, all patients are instructed to irrigate the nasal cavity with saline spray at least three times per day for 2–3 weeks. After that time, high-volume saline irrigation can be used to optimize mucosal healing and help debride the nasal cavity. When a nasoseptal flap is not raised, any septal splints or silastic sheets placed can be removed at 1 week; otherwise, they are removed at 2 weeks in cases where the septal mucosa has been elevated. Crusting in the nasal cavity can then be gently debrided in the clinic.

1.5 Instrumentation

EEA has largely replaced microscopic transnasal surgery due to the tremendous advantages in visualization and illumination provided by the endoscope. Standard endoscopes utilized for EEA are rigid, 4 mm in diameter, and 18 or 30 cm in length. Smaller 2.7 mm endoscopes can be used for pediatric patients. Although zero-degree scopes are predominantly used, angled scopes of 30, 45, 60, and even 90 degrees provide the ability to expand operative visualization (Fig. 1.3a–d).

Endoscopes traditionally have provided only a two-dimensional view, and therefore the optics can suffer compared to the 3D view of the operative microscope. While some surgeons may find this to be a tremendous disadvantage, it can be overcome by using the manual movement of the scope and operative instruments to provide a better appreciation of the three-dimensional anatomy. We therefore recommend the scope to be "driven" by a member of the operative team, although some surgeons do have a preference for a mechanical scope holder. More recently, 3D endoscopes have become available, but their widespread use is limited by cost, need for additional equipment (e.g., properly equipped monitors and eyeglasses), and in some cases eye strain.

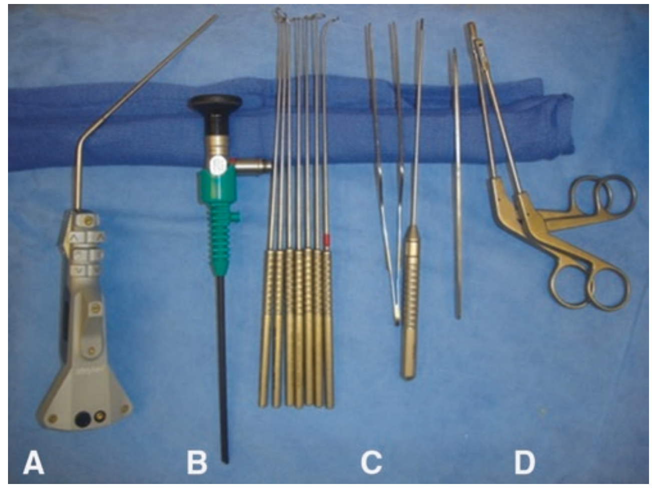

Fig. 1.3 (**a–d**) Sample of the equipment and instrumentation used in the endonasal endoscopic approach, including (**a**) navigation suctions, (**b**) endoscope with irrigation sheath, (**c**) straight endonasal instruments, and (**d**) sinonasal rongeurs

As the endoscope is advanced within the surgical field of the sinonasal cavity, unlike the operative microscope, it becomes imperative to have a mechanism for maintaining visualization and clearing the scope of blood and surgical debris. This can be done with either manual irrigation or a cleaning-irrigation system. The latter typically takes the form of a sheath around the endoscope that is controlled with a foot pedal or hand control. Although we find these systems highly efficient in limiting repeated entrances and exits from the nostril, surgeons who prefer manual irrigation criticize the added bulk and size of the irrigation sheath.

Unlike the bayoneted microscopic transsphenoidal instruments, the operative instruments used in EEA should be straight to avoid conflict with the endoscope (Fig. 1.4a, b). While their shaft is straight, the tips can be fitted with a variety of angled tips and shapes. Additionally, malleable instruments that can be shaped to accommodate the unique needs of each operative pathology are also of tremendous utility.

Hemostatic control can be obtained quickly and efficiently by a surgeon proficient in EEA. Sinonasal venous oozing can be reduced with elevating the head of the bed, preventing hypertension, total intravenous anesthesia, and warm irrigation fluid. Electrocautery is the mainstay of hemostasis with monopolar devices being quite effective. When used for mucosal graft and flap incisions, a fine needlepoint monopolar electrocautery should be used, while broad suction monopolar devices are better for obtaining hemostasis and vascular control within the sinonasal cavity. Bipolar electrocautery has proven more challenging in endonasal surgery; however, a number of devices, both in malleable suction and pistol-grip form, are available. Additional vascular control can be obtained with vascular clips, and

Fig. 1.4 (**a**, **b**) Endonasal instruments with straight shafts, including (**a**) ring currettes and (**b**) microdissectors

traditional microaneurysm clips for intracranial use can be placed with specially designed appliers that can also be malleable. In addition to electrocautery, other hemostatic agents useful in EEA are familiar to both neurosurgeons and otolaryngologists. These include flowable hemostatics, oxidized cellulose, thrombin-soaked gelatin sponges, and even bone wax for osseous bleeding.

For soft tissue removal and tumor debulking, tissue debriders developed for rhinologic surgery can be used and are available in both straight and angled forms. For more delicate and intracranial work, ultrasonic aspirators and side-cutting tumor aspiration devices are better utilized. These can be combined with traditional bimanual microsurgical techniques.

For exposure of the osseous cranial base and creating osteotomies, angled self-irrigating drills are available. These have a protective sheath along the drill shaft to prevent thermal injury to nasal structures. We prefer larger (3 or 4 mm) round cutting burrs for removal of sinonasal bone (e.g., sphenoid rostrum, pterygoid base, etc.). When drilling the cranial base or trying to skeletonize the bone surrounding neurovascular structures, diamond burrs are preferred in smaller sizes.

1.6 Imaging

To best assess the target pathology, high-quality preoperative imaging is essential for surgical planning and for the purposes of intraoperative neuronavigation. We recommend a thin slice CT with multiplanar reconstructions for analysis of the osseous cranial base and sinonasal anatomy. This demonstrates the degree of aeration of the paranasal sinuses and the osseous integrity of involved structures, location of hyperostosis, evaluation of intersinus septae, and the presence of anatomical variations (e.g., Onodi cells, pneumatized anterior clinoid processes, concha bullosa, etc.). Additionally, CT angiograms can be obtained to clarify the associated vascular anatomy.

High-field MRI is helpful to distinguish soft tissue and neurovascular structures (Figs. 1.5a–d and 1.6a–d). The interface between neural structures and pathology can be reviewed through the multiple MRI sequences. Any vascular abnormalities evident on CTA or MRA sequences can be further evaluated with digital subtraction angiography (DSA). The latter also provides the ability for embolization of a vascular lesion or tumor, when necessary.

1.7 Intraoperative Monitoring

When traversing the cranial base and managing the associated pathology and intracranial anatomy, multiple neurovascular structures may be at risk. Overall the rates of vascular injury and postoperative cranial neuropathies are relatively low, but these can be devastating complications. Any potential measures to further mitigate these risks should be utilized. Although standard transsphenoidal transsellar approaches may not require intraoperative monitoring, it is typically useful for all extended approaches and for any lesion extending intracranially beyond the sella and diaphragm sella.

Global cortical monitoring with electroencephalography (EEG), somatosensory evoked potentials (SSEPs), and transcranial electrical motor evoked potentials (MEPs) provides critical information when the internal carotid arteries or their branches are at risk. When addressing pathology within

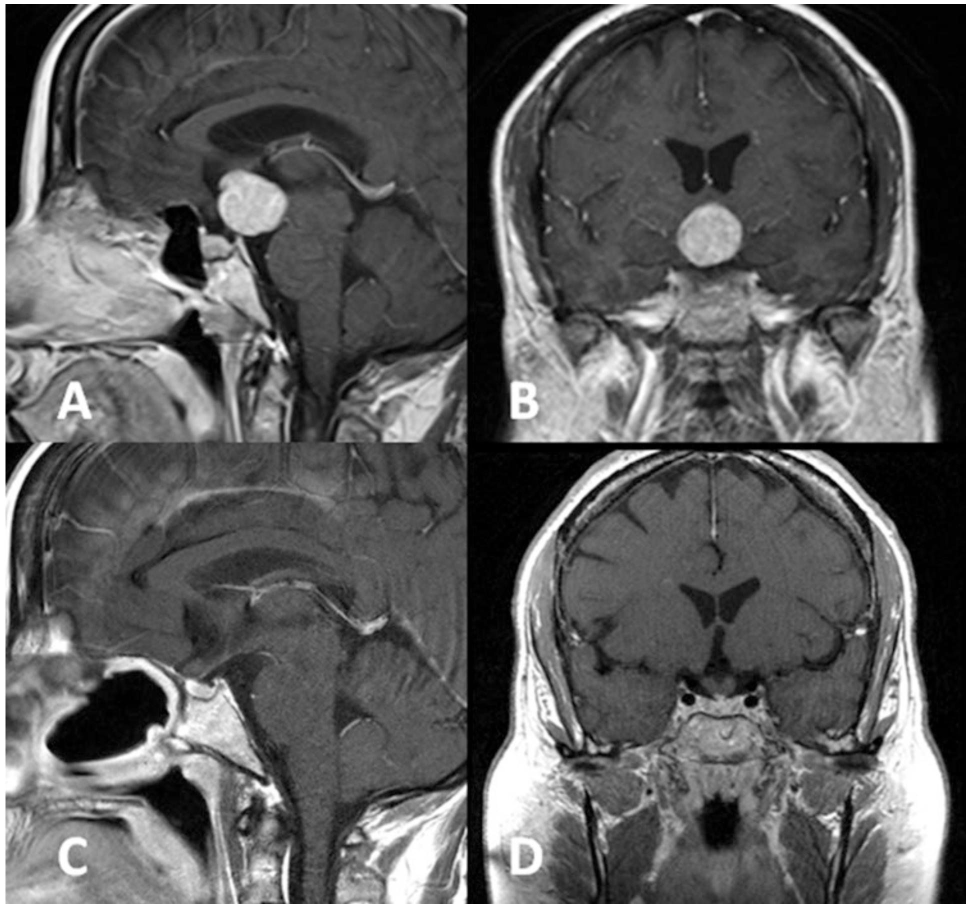

Fig. 1.5 (**a**–**d**) Suprasellar craniopharyngioma resected through an endoscopic endonasal approach. Preoperative (**a**) sagittal and (**b**) coronal MRI. Postoperative (**c**) sagittal and (**d**) coronal MRI

the posterior fossa or craniocervical junction through transclival approaches, brainstem ischemia can be detected with brainstem auditory evoked potentials (BAEPs). Cranial nerve monitoring can be performed through electromyography (EMG), which may either be spontaneous or triggered with a hand-held probe. Triggered EMG of extraocular muscles of the eye is very useful when performing significant work within the cavernous sinus.

1.8 Navigation

The endoscope provides excellent visualization of the sinonasal cavity and the cranial base, but image guidance can be an invaluable tool for endonasal surgery. It decreases surgical disorientation, optimizes outcomes, and lowers complications rates. Currently available navigation systems function via either optical or electromagnetic technology, and our preference is for an optical system that does not require head fixation for the reasons previously mentioned. While we advocate for the routine use of navigation to maximize familiarity with its use and to serve as a teaching tool for trainees, it is especially helpful in a number of scenarios:

- Presellar or conchal-type sphenoid sinuses and other anatomic variations
- When the normal anatomy has been distorted by the pathology
- Approaching an intracranial lesion
- Recurrent surgery

1.9 Conclusion

The endoscopic endonasal approach affords significant advantages including direct access to pathology, reduced postoperative pain, lower incidence of complications, and quicker patient recovery. As its use has extended beyond the more familiar transsphenoidal approach to sellar pathology, it is critical that the endonasal cranial base surgeon has full knowledge of all of the principles of endoscopic surgery. This will permit optimal outcomes while minimizing complications.

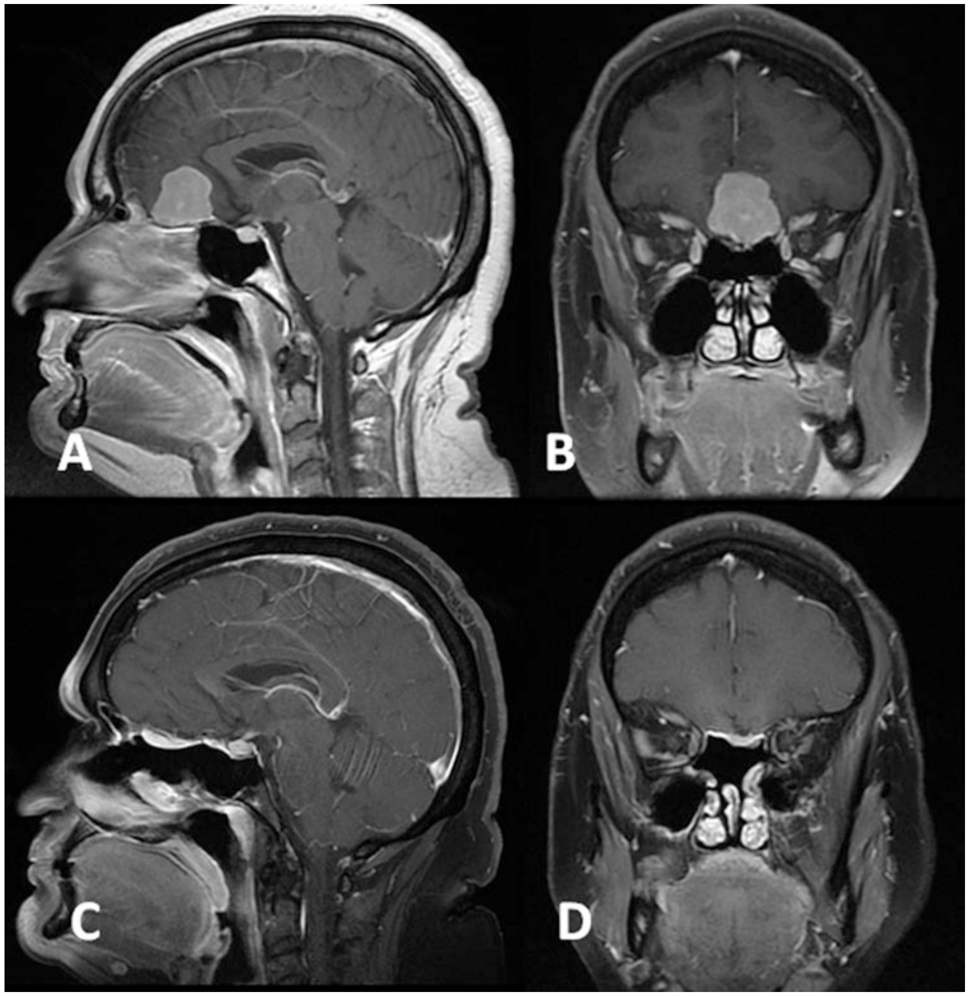

Fig. 1.6 (**a**–**d**) Olfactory groove meningioma resected through an endoscopic endonasal approach. Preoperative (**a**) sagittal and (**b**) coronal MRI. Postoperative (**c**) sagittal and (**d**) coronal MRI

2 Sinonasal and Parasellar Anatomy

Maria Peris-Celda, Carlos D. Pinheiro-Neto, Rowan Valentine, and Albert L. Rhoton

2.1 Sinonasal Anatomy

The Nose

The nose is part of the upper respiratory tract and can be broadly divided into the external nose (that emerges from the face) and the internal nasal cavity, both divided into right and left halves by the midline nasal septum. The nasal cavity is a truncated pyramid wider at its base that opens anteriorly onto the face through the external nose at the anterior nasal aperture and posteriorly onto the nasopharynx by way of the posterior nasal apertures, also called choanae.

Seven bones form the structure of the nose and paranasal sinuses: frontal, sphenoid, vomer, and ethmoid bones as unpaired and nasal, palatine, maxillae, and inferior concha as paired ones (Figs. 2.1a–f and 2.2a–g). The external nose is formed by the frontal processes of the maxilla laterally and the nasal bones anteriorly and superiorly forming the dorsum of the nose. The frontal processes of the maxillae separate the orbit from the nose anteriorly forming the medial orbital rim. The superior (lateral) and inferior (lower or alar) nasal cartilages keep the air pathway open at the aditus or nasal vestibules, externally seen as nostrils. The lateral boundary of the nostrils is formed by fibrofatty tissue and some small cartilages [1–4].

The posterior nasal aperture is formed by the anterior part of the sphenoidal body above, perpendicular plate of the palatine bone laterally, the horizontal plate of the palatine inferiorly, and the posterior edge of the vomer that divides this aperture in two halves [2, 3] (Figs. 2.1a–f and 2.2a–g). The lateral wall of the nasal cavity can be divided in a superior part that forms the medial wall of the orbit and an inferior part with the maxillary sinus laterally. The medial wall of the orbit is composed of the frontal process of the maxilla, lacrimal bone, and the orbital plate of the ethmoid bone whose air cells (ethmoidal labyrinth) separate the orbit from the nasal cavity. This thin part of the ethmoid bone that faces the orbit is also called the lamina papyracea for its "paper-thin" fragility. The inferior part of the lateral wall of the nasal cavity is formed from anterior to posterior by the maxilla, inferior concha, and perpendicular plate of the palatine bone. The palatine bone articulates posteriorly with the pterygoid plates of the sphenoid bone, and the pterygopalatine fossa lies in their junction bounded anteriorly by the posterior aspect of the maxilla. The floor of the nasal cavity is composed from anterior to posterior by the palatine process of the maxilla and the horizontal plate of the palatine bone. The roof from anterior to posterior is formed by the nasal bones, nasal spine of the frontal bone, cribriform plate (ethmoid), and sphenoidal body. The nasal septum divides the cavity into two halves, with its osseous posterior part formed superiorly by the perpendicular plate of the ethmoid and inferiorly by the vomer and its anterior cartilaginous part formed by the quadrangular cartilage [2–4] (Figs. 2.1a–f, 2.2a–g, and 2.3a–d).

There can be up to five osseous structures that protrude from the lateral wall of the nasal cavity – three constant: superior, middle, and inferior turbinates (conchae), the inconstant supreme turbinate (Santorini's), and in 1% of cases there is a fifth turbinate superior to the supreme (Zuckerkandl's). The supreme, superior, and middle turbinates belong to the ethmoid bone, while a separate bone,

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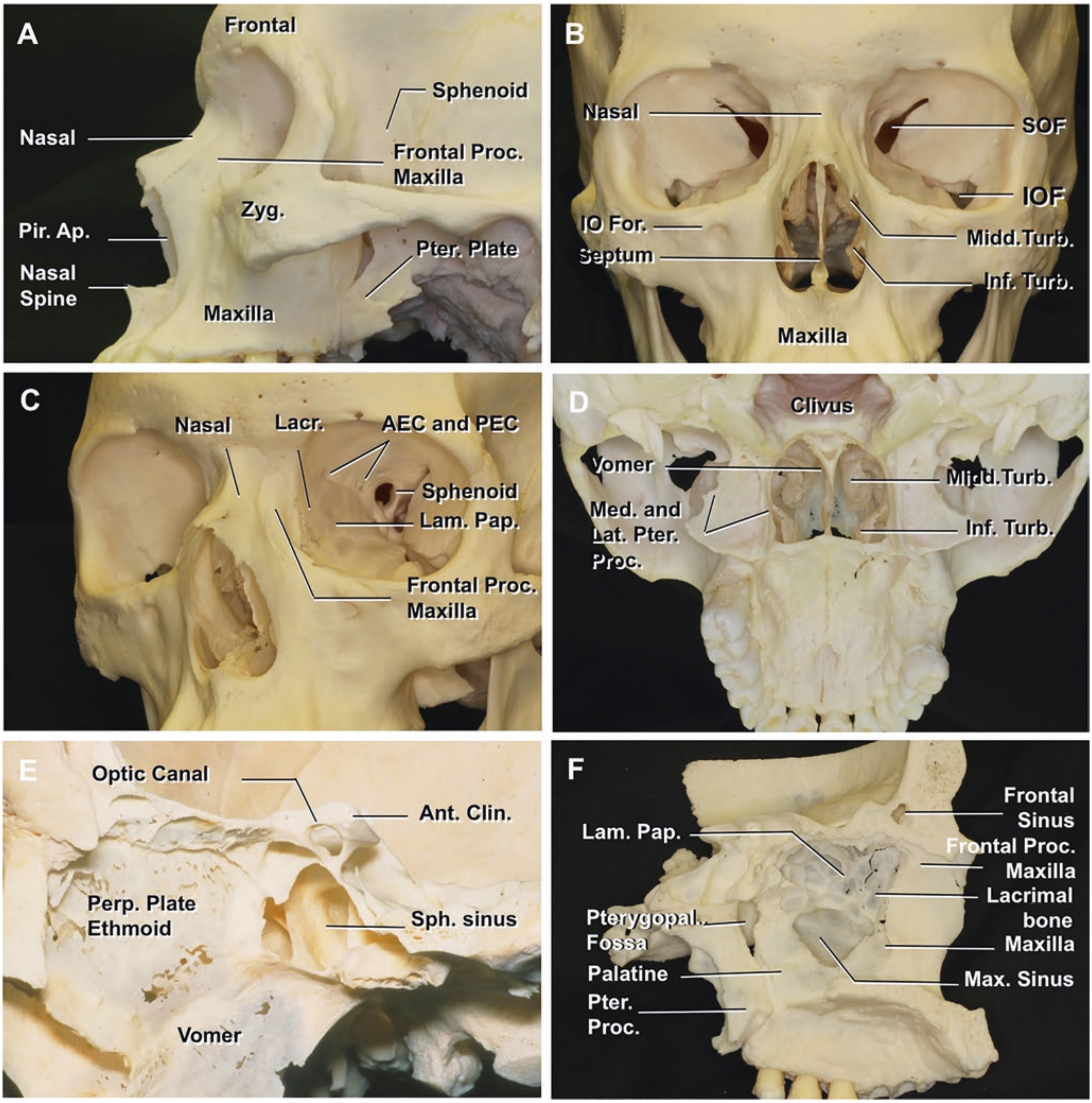

Fig. 2.1 (**a**–**f**) Structure and relationships of the nasal cavity in the articulated skull. (**a**, **b**) The piriform aperture constitutes the osseous anterior nasal aperture and is formed inferiorly and laterally by the maxillae and superiorly by the nasal bones. (**c**) The superolateral wall of the nasal cavity constitutes the medial wall of the orbit and is formed from anterior to posterior by the frontal process of the maxilla, lacrimal bone, lamina papyracea of the ethmoid bone. The anterior and posterior ethmoidal canals are formed by the frontal bone superiorly and ethmoid bone inferiorly. (**d**) Posterior view of the nasal cavity. The pterygoid processes of the sphenoid bone are posterior to the nasal cavity. The vomer divides the posterior nasal apertures called choanae, the inferior and middle turbinates are visualized posteriorly. (**e**) Skull split sagittally at the level of the septum. The septum is formed by the perpendicular plate of the ethmoid superiorly and the vomer inferiorly. (**f**) Medial view of the lateral nasal cavity and the medial orbital wall. The ethmoid labyrinth has been removed leaving the lamina papyracea, a thin layer of bone. The medial wall of the orbit is composed of the frontal process of the maxilla, lacrimal bone, and the orbital plate of the ethmoid bone whose air cells (ethmoid labyrinth) separate the orbit from the nasal cavity. Inferiorly, the lateral nasal cavity is formed by the maxilla and palatine bone. The palatine bone articulates posteriorly with the pterygoid processes, and the pterygopalatine fossa is located between them. *A* artery, *AEC* and *PEC* anterior and posterior ethmoidal canal, *For* foramen, *Inf* inferior, *IO* infraorbital, *IOF* inferior orbital fissure, *Lam* lamina, *Max* maxillary, *Med* medial, *Midd* middle, *Lat* lateral, *Pap* papyracea, *Pter* pterygoid, *Proc* process, *Pir* piriform, *Ap* aperture, *Pterygopal* pterygopalatine, *SOF* superior orbital fissure, *Turb* turbinate

Fig. 2.2 (continued) for the turbinates, meatus, and maxillary sinus. (**g**) Articulation of the maxilla, palatine, and ethmoid bones, medial view. The ethmoid bone has been divided in midline and the perpendicular plate removed. The supreme (if present), superior, and middle turbinates are part of the ethmoid bone, and the inferior turbinate is an independent bone. The posterior attachment of the middle turbinate is located below the sphenopalatine foramen. *Alv* alveolar, *Att* attachment, *Crib* cribriform, *Inf* inferior, *Hor* horizontal, *Inf* inferior, *Max* maxillary, *Midd* middle, *Perp* perpendicular, *Post* posterior, *Proc* process, *Sphen* sphenoidal, *Sphenopal* sphenopalatine, *Zyg* zygomatic bone, *Turb* turbinate

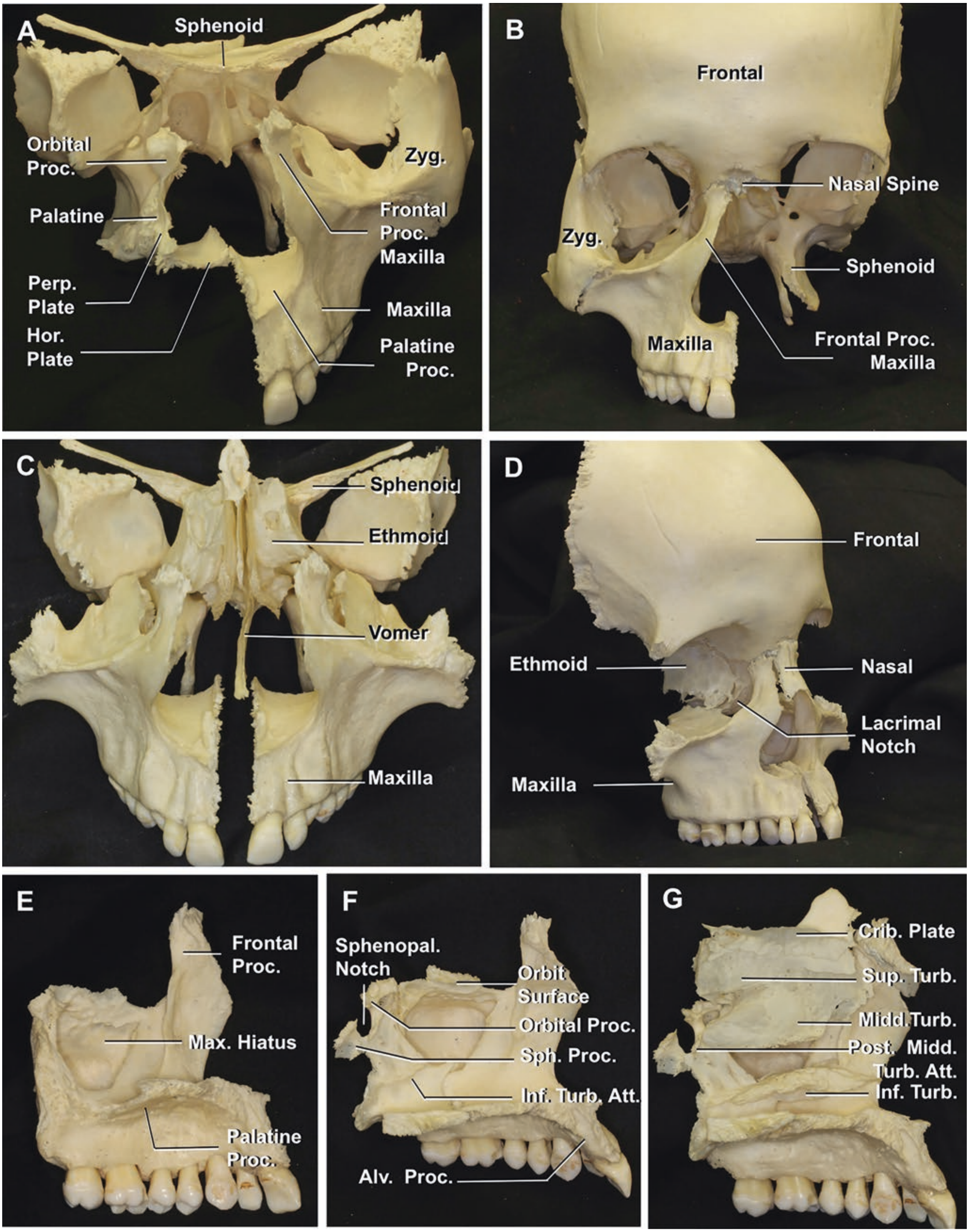

Fig. 2.2 (**a**–**g**) Bones that form the structure of the nasal cavity. (**a**) Anterior view of the maxilla, palatine bone, and sphenoid bone. The hard palate, floor of the nasal cavity, is formed by the palatine process of the maxilla anteriorly and horizontal plate of the palatine bone posteriorly. (**b**) The frontal bone articulates anteriorly with the frontal process of the maxilla, and its nasal spine forms part of the roof of the nasal cavity. (**c**) The ethmoid bone articulates posteriorly with the body of the sphenoid, forms part of the medial orbit wall, septum, and roof of the nose. (**d**) The roof of the nasal cavity is formed by the nasal bones, nasal spine of the frontal bones, cribriform plate of the ethmoid and sphenoidal body (not shown in this picture). (**e**) Medial view of the maxilla, the maxillary sinus opens medially into the nasal cavity. The frontal process of the maxilla articulates with the frontal bone and lacrimal bone. The palatine process articulates with the contralateral mate and posteriorly with the palatine bone. (**f**) Articulation with the palatine bone. The orbital process and the sphenoidal process of the palatine limit anteriorly and posteriorly the sphenopalatine foramen, which is completed superiorly by the sphenoid. The sphenopalatine artery, terminal branch of the maxillary artery, exits the pterygopalatine fossa through this foramen and enters the nasal cavity. It gives a medial or septal branch for the septum and the lateral branch

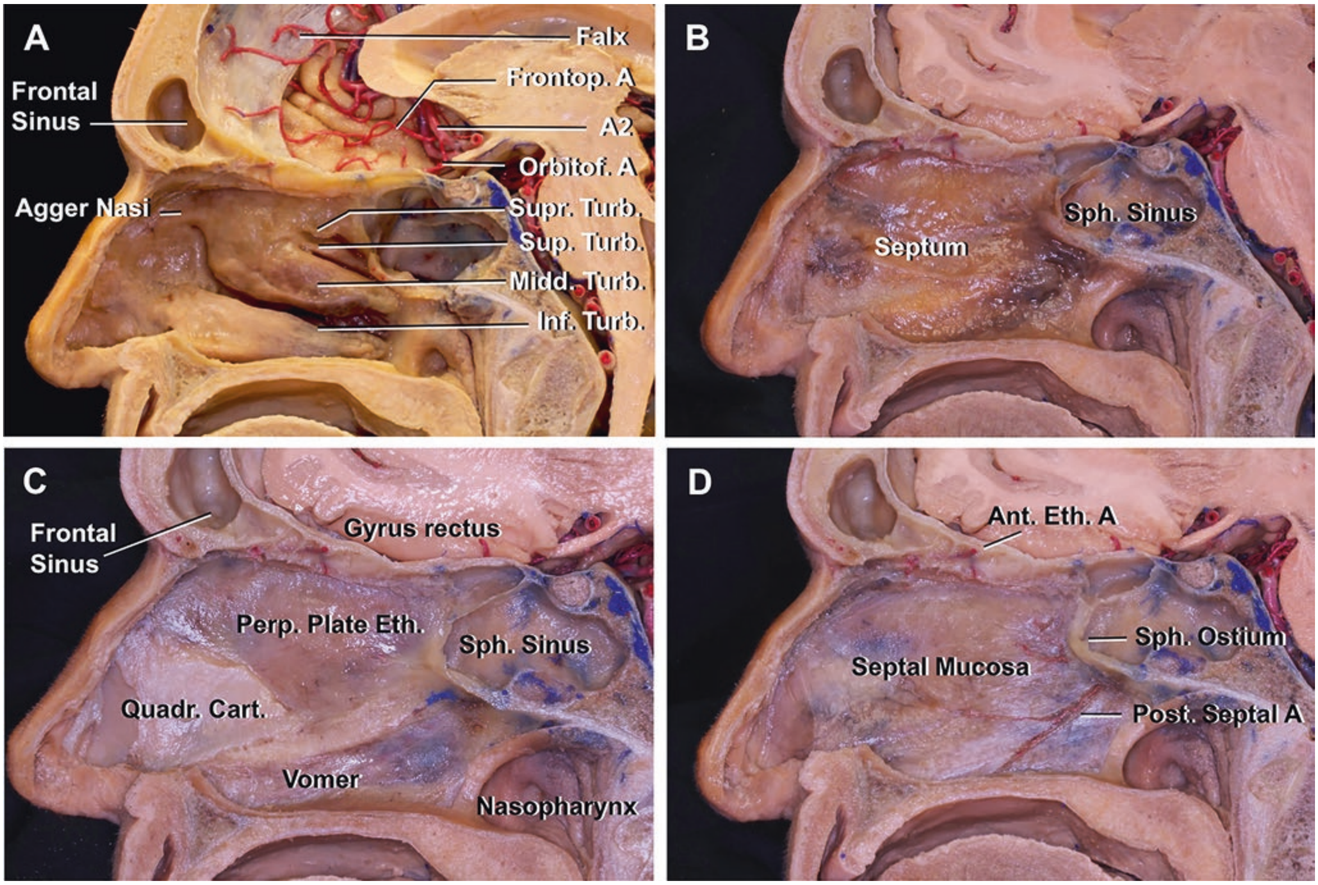

Fig. 2.3 (**a**–**d**) Anatomical specimen divided in the midsagittal plane. (**a**) Right nasal cavity with the supreme, superior, middle, and inferior turbinates. The left hemisphere has been removed keeping the A2 branches in midline. The orbitofrontal artery is the first branch of A2; it lies in the olfactory sulcus in the frontobasal surface and is the first branch encountered in an endoscopic craniofacial resection. The frontopolar artery arises superior to the orbitofrontal artery in midline. (**b**) The nasal septum mucosa. (**c**) The mucosa has been removed to show the structure of the septum with an anterior cartilaginous and posterior osseous portion formed by the perpendicular plate of the ethmoid bone superiorly and the vomer inferiorly. (**d**) The bone and cartilages have been removed and the periosteal-pericondreal surface of the septum dissected to show the main arterial supply: the posterior septal arteries, branches of the sphenopalatine artery, and the ethmoidal arteries branches of the ophthalmic artery. *A* artery, *Ant* anterior, *Eth* ethmoidal, *Frontop* frontopolar, *Orbitof* orbitofrontal, *Post* posterior, *Sphen* sphenoidal, *Sup* superior, *Supr* supreme, *Turb* turbinate

the inferior concha, forms the inferior turbinate. Below each turbinates' free inferior border, the corresponding spaces are called the superior, middle, and inferior meatuses. The middle meatus is the most complex in structure. Two important structures can be distinguished without further dissection, an elongated protrusion anteriorly called the uncinate process and a bulging structure posteriorly that corresponds to the ethmoidal bulla. The nasolacrimal groove and canal and the site of the lacrimal sac and nasolacrimal duct, respectively, are more frequently just posterior to the origin of the axilla of the middle nasal turbinate and end in the inferior meatus approximately 2 cm behind the nostril. The nasolacrimal canal is formed by the maxilla and lacrimal bone and protrudes vertically just anterior to the middle meatus forming the lacrimal crest. Approximately 1 cm behind the inferior turbinate and within the nasopharynx is the opening of the Eustachian tube [1]. The posterior ethmoidal cells drain into the superior meatus; the anterior ethmoidal cells, maxillary sinus, and frontal sinus into the middle meatus; and the nasolacrimal duct into the inferior meatus. The sphenoidal sinus drains directly into the nasal cavity (Fig. 2.4a–l).

The vascularization of the nasal cavity depends on the ophthalmic, maxillary, and facial arteries. They form anastomotic plexuses between them in the nasal mucosa. The anterior and posterior ethmoidal arteries which are branches of the ophthalmic artery supply the ethmoidal and frontal sinuses and the roof of the nose (including the septum). The sphenopalatine artery, a branch of the maxillary artery, supplies the mucosa of the conchae, meatus, and posteroinferior part of the nasal septum and is the principal vessel supplying the nasal mucosa. Of importance in endoscopic anatomy is the location of the sphenopalatine foramen where the sphenopalatine artery exits the pterygopalatine fossa and enters the nasal cavity. It gives a medial or septal branch for the

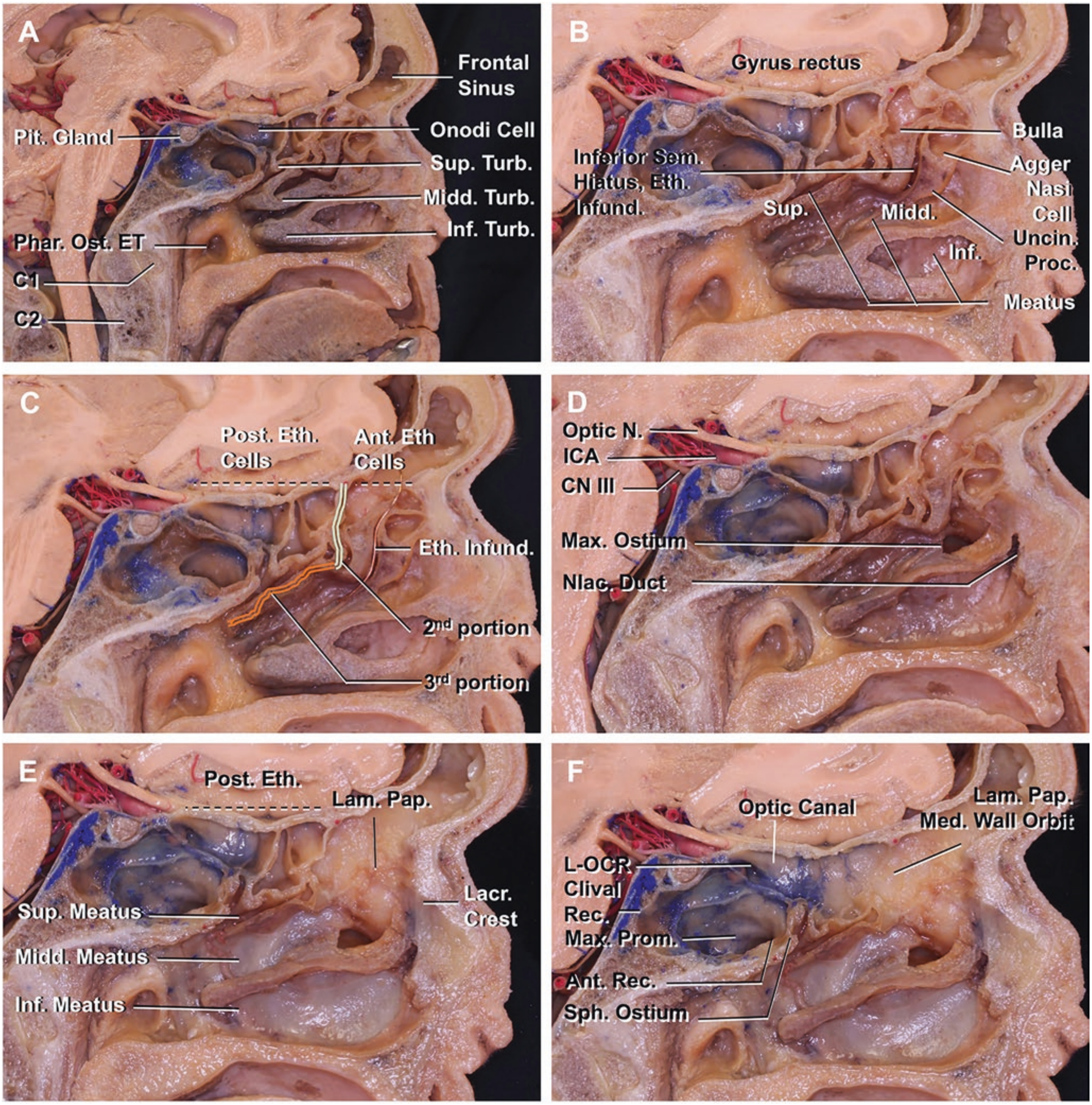

Fig. 2.4 (**a**–**l**) Left side of an anatomical specimen divided in the sagittal plane through the left nasal cavity, dissection from medial to lateral. (**a**) Note that the ethmoid labyrinth in this specimen pneumatizes posteriorly up to the optic nerve (Onodi cell). (**b**) The uncinate process mucosa has been dissected to show its inferior attachment to the inferior turbinate, hiding the maxillary ostium behind. The middle turbinate has been resected leaving its attachment to the lateral basal wall. The ethmoid infundibulum is located between the bulla, lamina papyracea, and uncinate process and is a three-dimensional space. The two-dimensional plane between these two structures is called inferior semilunar hiatus. The agger nasi cell is a pneumatization of the agger nasi region that is anterior to the middle turbinate attachment. (**c**) The second and third portions of the basal lamella of the middle turbinate attachment in the lateral nasal wall are shown in different colors. The vertical second portion divides the ethmoid cells in anterior and posterior ethmoid cells according to the surgical classification. (**d**) When the uncinate process is resected, the maxillary ostium is visualized. The nasolacrimal duct has been opened. (**e**) An anterior ethmoidectomy has been performed being its posterior limit the second portion of the middle turbinate attachment. (**f**) The medial wall of the orbit (lamina papyracea) is exposed up to the optic canal. (**g**) A medial maxillary antrostomy has been performed; the maxillary sinus is located below the orbit. Medial and inferior approaches to extraconal and intraconal pathologies of the orbit can be performed endonasally. (**h**) The maxillary opening has been fully completed medially, the lamina papyracea has been removed, and the periorbit is divided to expose the orbital fat. (**i**, **j**) Dissection of the orbital contents medially and inferiorly up to the infraorbital nerve. The central retinal artery usually enters the optic nerve in its inferior portion. (**k**) The sphenoidal sinus bone over the sella and cavernous sinus has been drilled and the periosteal layer of the dura mater exposed. (**l**) The dura mater has been removed, and the cavernous sinus dissected. The ophthalmic division of the trigeminal nerve and the VI cranial nerve enters the superior orbital fissure. The maxillary nerve passes through the foramen rotundum and forms anteriorly the infraorbital nerve. The floor of the sinus has been drilled to expose the Vidian nerve whose posterior aspect is in close relationship with the internal carotid artery at foramen lacerum. *Ant* anterior, *CN* cranial Nerve, *Eth* ethmoidal, *ICA* internal carotid artery, *Inf* inferior, *Infund* infundibulum, *Lam* lamina, *Lacr* lacrimal, *L-OCR* lateral opticocarotid recess, *Max* maxillary, *Med* medial, *Midd* middle, *N* nerve, *Nlac* nasolacrimal, *Pap* papyracea, *Pit* pituitary, *Post* posterior, *Prom* prominence, *Proc* process, *Rec* recess, *Sphen* sphenoidal, *Sup* superior, *Turb* turbinate, *Uncin* uncinate

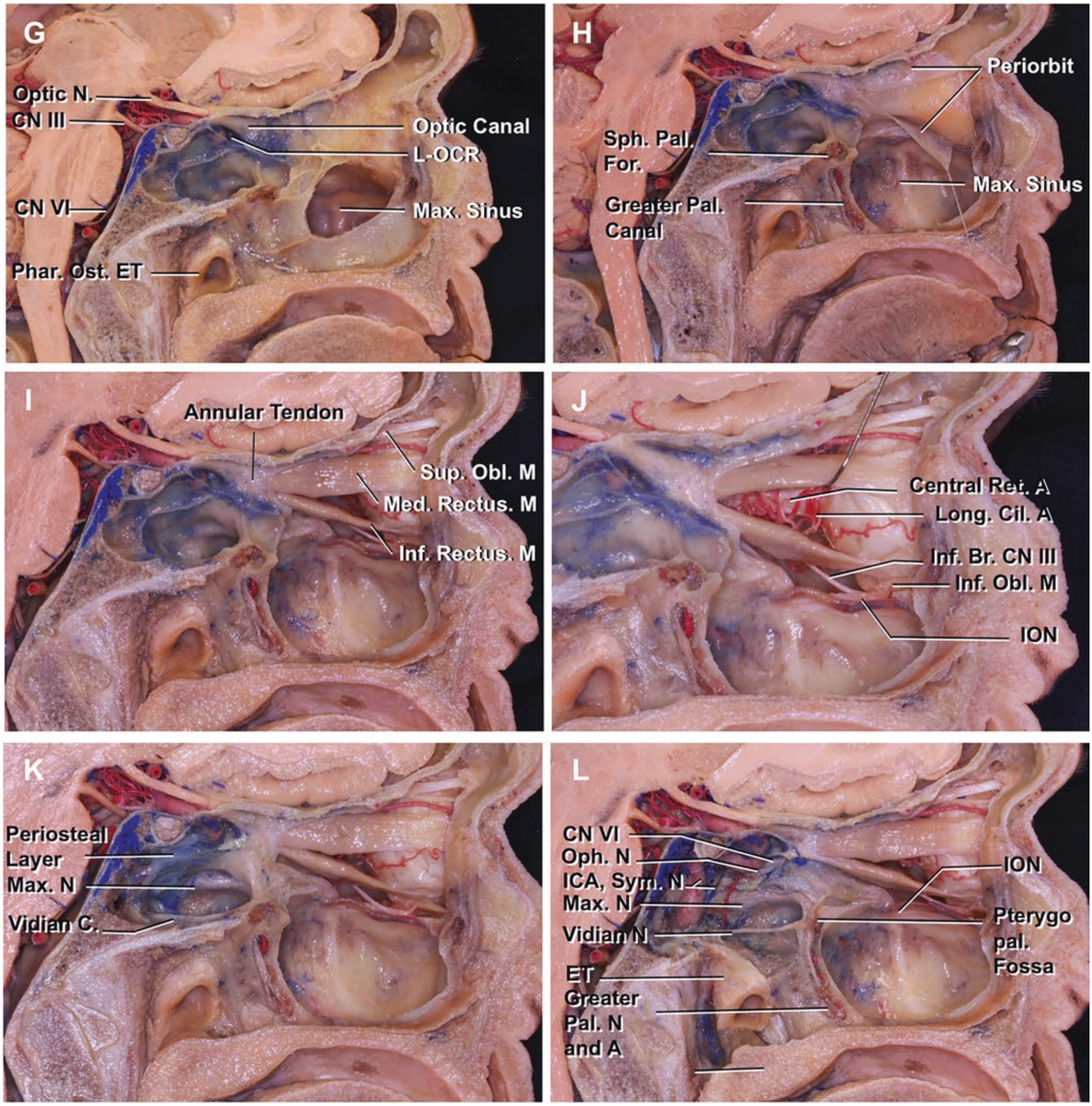

Fig. 2.4 (continued)

septum and a lateral branch for the turbinates, meatus, and maxillary sinus although the number and distribution of its branches are highly variable. The medial branch, or posterior septal artery, supplies the nasoseptal flap for skull base reconstruction [5, 6]. The sphenopalatine foramen is directly superior to the posterior aspect of the middle turbinate attachment and is formed anteriorly by the orbital process of the palatine bone, posteriorly by the sphenoidal process of the palatine bone, and superiorly by the sphenoidal body (Figs. 2.1a–f, 2.2a–g, and 2.3a–d). The greater palatine branch of the maxillary artery supplies the region of the inferior meatus. Its terminal part ascends through the incisive canal to anastomose on the septum with branches of the sphenopalatine and anterior ethmoidal arteries and with the septal branch of the superior labial artery. The mucosa of the maxillary sinus is supplied by the infraorbital artery and the superior, anterior, and posterior alveolar branches of the maxillary artery. The pharyngeal branch of the maxillary artery that runs through the palatovaginal canal supplies the sphenoidal sinus mucosa.

The trigeminal nerve carries the sensitive innervation roof of the nasal cavity and upper septum with the nasociliary nerves (ophthalmic division) that comes through the anterior and posterior ethmoidal canals. The anterior superior alveolar nerve supplies part of the septum, the floor near the anterior nasal spine and the anterior part of the lateral wall as high as the opening of the maxillary sinus. Branches of the greater palatine nerve supply the posterior three-quarters of the lateral wall, roof, and floor along with the inferior part of the nasal septum (medial posterior superior nasal nerves and the nasopalatine nerve).

Sympathetic postganglionic vasomotor fibers are distributed with the blood vessels. Parasympathetic fibers are carried through the Vidian nerve, synapse in the pterygopalatine ganglion, and are distributed via branches of the maxillary nerves in the nasal mucosa [7, 8].

Frontal Sinus and Related Structures

The frontal sinus is formed by the frontal bone and is closed inferiorly by the ethmoid bone (Fig. 2.5a–f). The brain surface facing the posterior wall of the frontal sinus is the medial aspect of the frontal pole. The frontal sinus drains through the frontal infundibulum that narrows to the frontal ostium. The secretions are directed through the ethmoid infundibulum to the middle meatus or directly into the middle meatus. The frontal infundibulum, frontal ostium, and ethmoid infundibulum are part of the ethmoid bone. The uncinate process attachment directs the drainage of the sinus to the middle meatus, being independent of the ethmoid infundibulum when the uncinate process is attached to the lamina papyracea. Variants in the course of the frontoethmoidal cells, uncinate process, and ethmoidal bulla can narrow the frontal recess and with it the natural sinus ostium [9]. There can be extended pneumatization of the frontal sinus forming interfrontal sinus septal cells that is associated with pneumatized crista galli. The frontal sinus drainage is directed by the ethmoid anatomy.

Ethmoidal Labyrinth and Related Structures

The ethmoid bone has a cuboidal shape. It lies anteriorly in the cranial base and contributes to the medial walls of the orbit, the nasal septum, and the roof and lateral walls of the nasal cavity. It has a horizontal perforated cribriform plate forming the roof, a median perpendicular plate, and two lateral labyrinths that contain the ethmoidal air cells. The middle, superior, and supreme (if present) turbinates form part of the ethmoid bone protruding medially into the nasal cavity (Fig. 2.6a–f).

The cribriform plate fills the ethmoidal notch of the frontal bone and forms part of the nasal roof. It is penetrated by numerous foramina that transmits branches of the olfactory nerves and their associated meninges. The crista galli is a thick appendix that projects upward from the anteromedial portion of the cribriform plate. The falx cerebri is attached to its thinner posterior border, while the shorter, thick anterior border articulates with the frontal bone by two small alae completing the foramen caecum. On both sides of the crista galli, the cribriform plate is narrow and depressed forming the olfactory fossa that is related to the olfactory bulb and gyrus rectus which lie above it. A groove runs forward to the foramen caecum from the anterior ethmoidal canal. The cribriform plate joins the ethmoidal labyrinth, whose roof is called the fovea ethmoidalis, through a thin piece of bone (lateral lamella) which is of great surgical interest. The coronal relationship of the fovea ethmoidalis with the lateral lamella and cribriform plate is subject to many variations. Keros identified three types of relationships: Keros type I (8% of cases), when the ethmoid roof is almost flat because the lateral lamella is no more than 3 mm high; Keros type II (80% of cases), when the lateral lamella is 4–7 mm; and Keros type III (12%), when it measures more than 7 mm [10]. When the cribriform plate is very low with respect to the ethmoid roof, surgical manipulation in these areas may cause CSF leaks easily [9]. The lateral lamella bone can be 0.2 mm thin, but the thinnest part is the site where the anterior ethmoidal artery enters the olfactory fossa in the ethmoid sulcus [9] (Figs. 2.6a–f and 2.7a, b).

The perpendicular plate of the ethmoid is located in the midline and descends from the cribriform plate to form the upper part of the nasal septum. It is thin superiorly and thicker inferiorly, quadrilateral in shape, and usually with some deviation. Its anterior border articulates with the nasal spine of the frontal bone and the crests of the nasal bones, and its posterior border articulates with the crest of the body of the sphenoid bone above and the vomer forming the inferior part of the septum below. The thick inferior border is attached to the nasal quadrangular cartilage (Figs. 2.1a–f, 2.3a–d, and 2.6a–f).

Embryologically the turbinates originate from their own basal lamellae. The highest point of the most superior turbinate is separated from the lamina cribrosa by approximately 5 mm anteriorly and 8 mm posteriorly [3] (Fig. 2.4a–l). The supreme, superior, and middle turbinates belong to the ethmoid bone, while the inferior turbinate is formed by a separate bone, the inferior concha. The direction of the major axes of the turbinates is oblique with the anterior part higher than the posterior part. The inferior turbinate, the longest and broadest, articulates from anterior to posterior with the maxilla, lacrimal, and palatine bone. The middle turbinate has its head very close to the nasal roof and the cribriform-plate

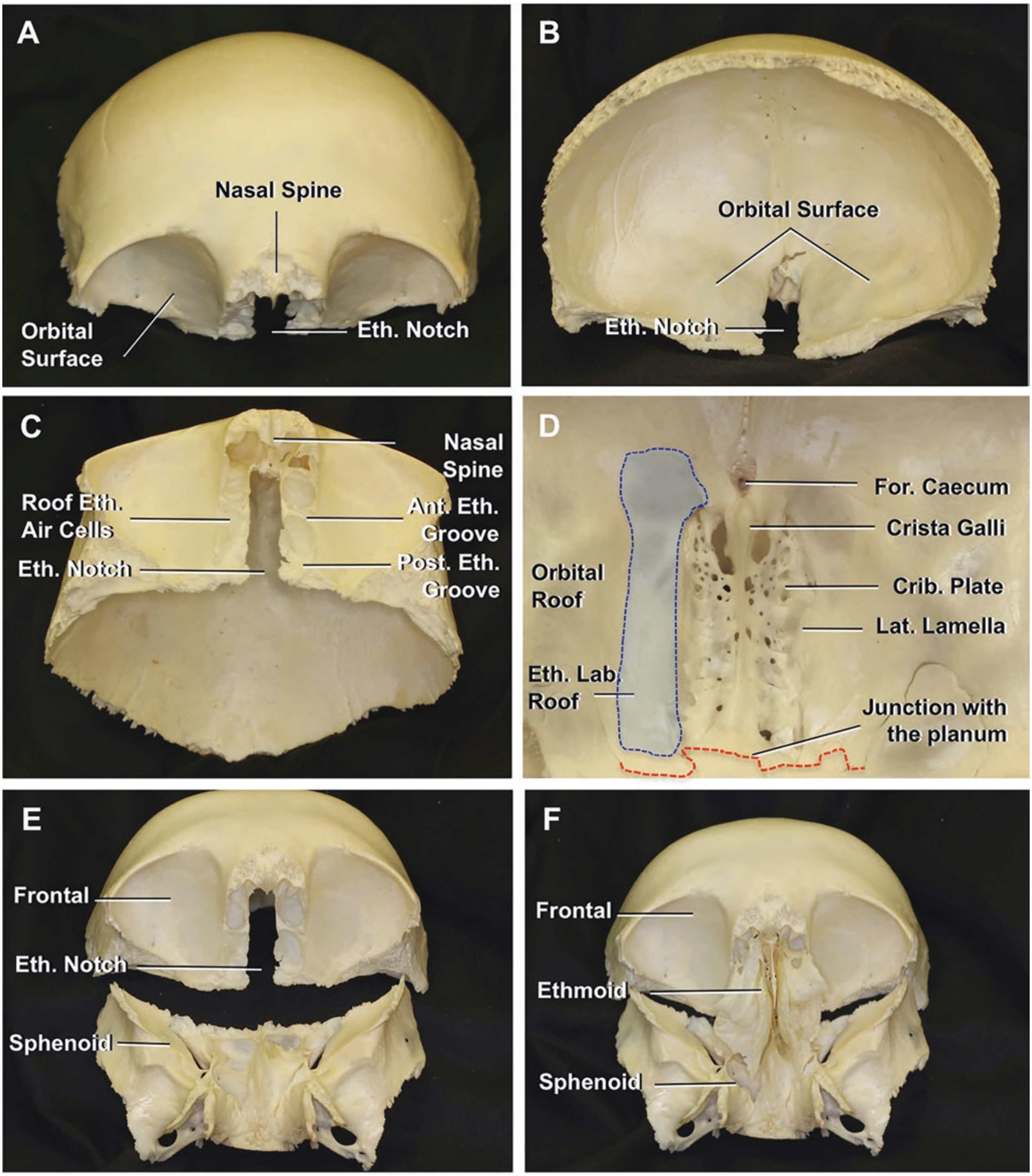

Fig. 2.5 (**a**–**f**) Frontal bone. (**a**, **b**) Anterior and posterior view of the frontal bone. It forms the roof of the orbit. Between both orbital surfaces, the ethmoid notch will be completed with the cribriform plate of the ethmoid bone. (**c**) The frontal bone from below. The ethmoid cells of the orbital plates are completed superiorly by the frontal bone as well as the ethmoid grooves that convert into the ethmoid canals. (**d**) Superior view of the cribriform area in an articulated skull. The crista galli is located anteriorly in midline and attaches the falx. The cribriform plate joins laterally the skull base through the lateral lamella with different lengths depending on the depth of the olfactory fossa. The *blue area* indicates the left ethmoid labyrinth roof in the anterior skull base. The *red dotted line* indicates the junction with the planum sphenoidale posteriorly. (**e**) Articulation with the sphenoid bone posteriorly, inferior view. (**f**) The ethmoid bone has been added completing the anterior skull base. *Ant* anterior, *Crib* cribriform, *Eth* ethmoidal, *For* foramen, *Lat* lateral, *Post* posterior

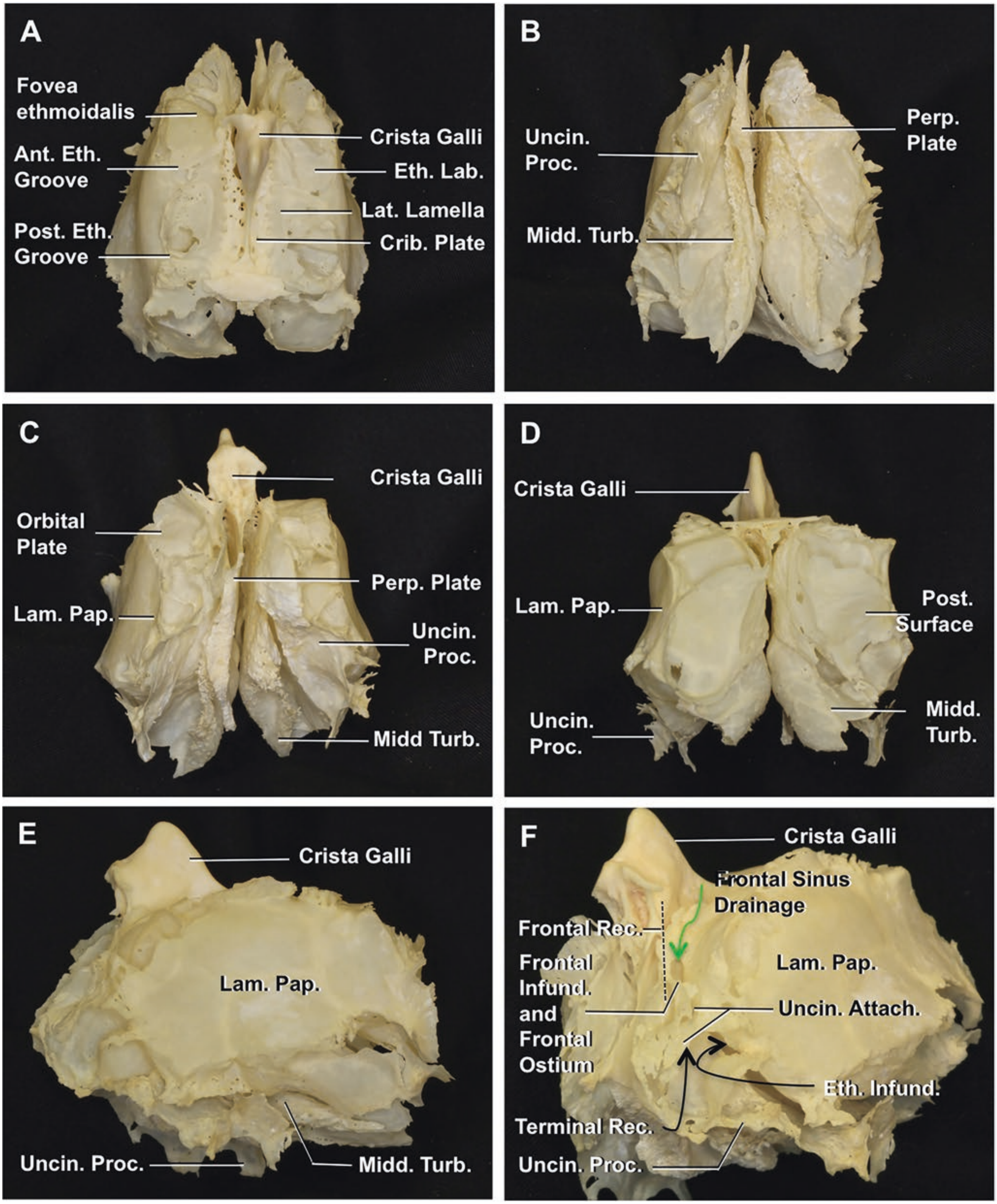

Fig. 2.6 (**a**–**f**) Ethmoid bone. (**a**) Superior view. (**b**) Inferior view. (**c**) Anterior view; the uncinate process is anterior to the orbital plate. In this case the perpendicular plate forming the superior surface of the septum is deviated to the right. (**d**) Posterior surface of the ethmoid that articulates with the sphenoid bone. (**e**) Lateral view of the ethmoid bone, the lamina papyracea forms the medial wall of the orbit. (**f**) Oblique view; the uncinate process is seen anterior to the orbital plate curving medially and inferiorly. The uncinate attaches to the lamina papyracea in 50% cases. In these cases, the frontal sinus drains into the middle meatus medially, independent to the ethmoid infundibulum which ends superiorly in the terminal recess. The frontal sinus drains through the frontal recess, narrowing gradually through the frontal infundibulum to the frontal ostium. From the level of the ostium, the frontal recess widens again in the inferior and posterior direction; this inferior segment is not seen in the picture as it is hidden by the uncinate process. *Ant* anterior, *Attach* attachment, *Eth* ethmoidal, *Infund* infundibulum, *Lab* labyrinth, *Lam* lamina, *Lat* lateral, *Midd* middle, *Pap* papyracea, *Perp* perpendicular, *Post* posterior, *Proc* process, *Rec* recess, *Turb* turbinate, *Uncin* uncinate

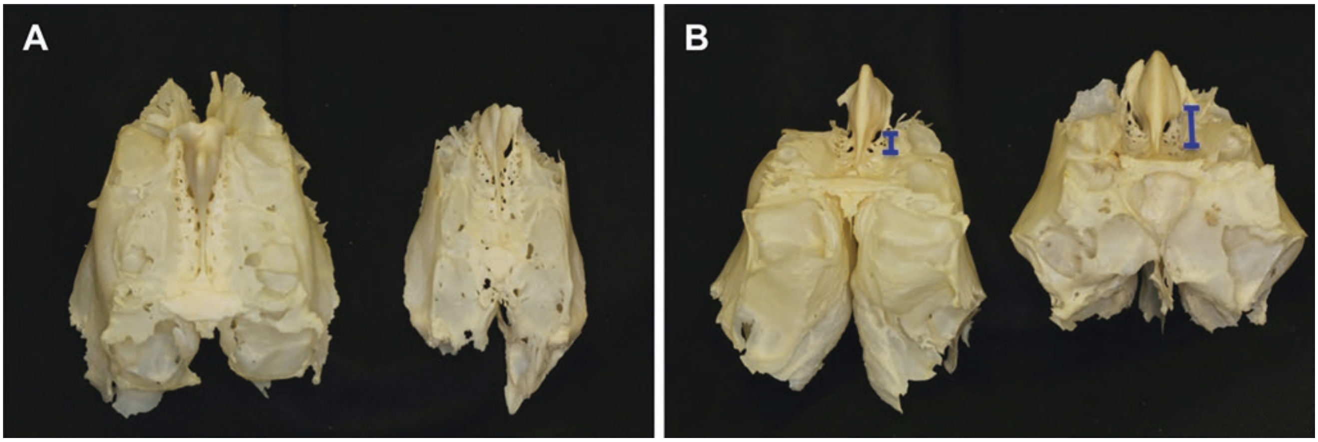

Fig. 2.7 (**a**, **b**) The ethmoid bone has great variability. (**a**) Superior surfaces of two different ethmoid bones; note the difference in anteroposterior and lateral dimensions. (**b**) Different depths of the olfactory fossae (*blue lines*) of importance in skull base approaches

area. The middle turbinate insertion lies in three different planes: the anterior third lies vertically attaching to the lateral end of the lamina cribrosa, the middle third is fixed to the lamina papyracea in an almost coronal plane, and the posterior segment is attached to the lamina papyracea, medial wall of the maxillary sinus, or both, turning inferiorly to become horizontal [11]. The middle turbinate can be pneumatized in 35% of cases [12] (concha bullosa), narrowing the surgical corridor. The posterior aspect of the middle turbinate articulates with the perpendicular plate of the palatine bone under the inferior edge of the sphenopalatine foramen. This attachment leaves an elevation at the anteroinferior margin of the sphenopalatine foramen. The endoscope can give a distorted view so that the attachment of the middle turbinate seems superior to the foramen (Fig. 2.8k).

The middle meatus is complex in its structure. In this area there is a bulging elongated osseous structure called the uncinate process which is located directly posterior to the nasolacrimal duct and functions as a guide for paranasal drainage. The uncinate process is a remnant of the descending portion of the first ethmoturbinal [11] (Figs. 2.4a–l and 2.6a–f). The uncinate may insert superiorly into the lamina papyracea (50%), skull base (25%), middle turbinate (25%), or a combination of these [8, 13], and it passes medial to the ostium of the maxillary sinus to join the ethmoidal process of the inferior nasal concha. In the dry skull, this sickle-shaped bone is located between the anterior and posterior fontanelles that are closed by connective tissue and mucosa in life and separates the maxillary sinus from the middle meatus. The closure of the anterior and posterior fontanelles results in the uncinate process only having a free posterior edge. Posterior and above the uncinate process is the ethmoidal bulla, formed by the bulging of one or more anterior ethmoidal cells, whose ostium opens posteriorly into the middle meatus (Fig. 2.8a–l). The bulla is formed by the pneumatization of the bulla lamella or second ethmoid basal lamella [11]. If the bulla does not reach the skull base, it results in the formation of the suprabullar recess and may extend into a retrobulbar space if the posterior wall of the bulla is not in contact with the basal lamella of the middle turbinate. Between the front face of the bulla and the uncinate process, there is a semicircular slit called inferior semilunar hiatus or semilunar hiatus (Figs. 2.8a–l and 2.9a–f). The superior semilunar hiatus is located between the ethmoidal bulla and the middle turbinate [11]. The ethmoid infundibulum is a three-dimensional space bordered medially by the uncinate process, laterally by the lamina papyracea and posteriorly by the ethmoidal bulla. Posteriorly, this infundibulum opens into the middle meatus through the inferior semilunar hiatus. The anterior ethmoidal cells and maxillary sinus drain into the ethmoidal infundibulum and, in 50% of cases, also the frontal sinus. Where the uncinate process is attached determines the drainage pathway of the frontal sinus. If the uncinate process is attached to the lamina papyracea, the infundibulum ends in a blind pouch called the terminal recess, and the frontal sinus drains independently into the middle meatus. If the uncinate attaches to the skull base or the middle turbinate, the frontal sinus will drain into the infundibulum and thus near or into the maxillary ostium [8] (Figs. 2.5a–f and 2.9a–f). The natural ostium of the maxillary sinus does not have osseous edges as the medial maxillary opening is wide as seen in a dry skull. The maxillary ostium is completely hidden from view by the middle portion of the uncinate process, which must be completely removed in order to visualize this opening (Figs. 2.4a–l and 2.8a–l). The term ostiomeatal complex is a functional unit that comprises the maxillary sinus ostium, anterior ethmoidal cells and their ostia, ethmoid infundibulum, hiatus semilunaris, and middle meatus [11].

The superior meatus under the superior turbinate is very small and is the space where the posterior ethmoidal cells ostia and recesses are located. The sphenoidal sinus ostium

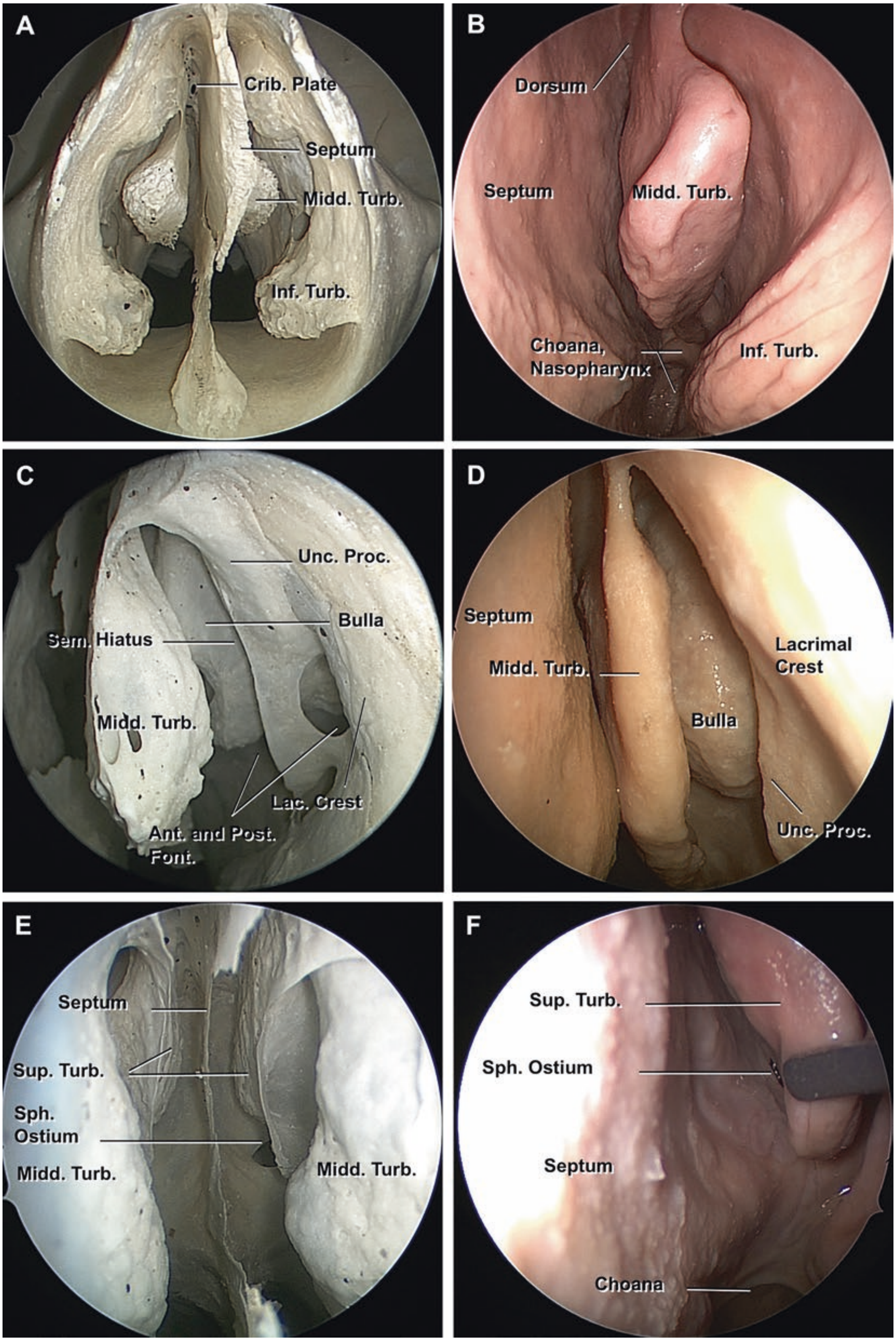

Fig. 2.8 (**a**–**l**) Images of the nasal cavity obtained with a 0° endoscope in a dry skull (left) and equivalent image in a formalin-fixed anatomical specimen (right). (**a**) General view introducing the endoscope through the piriform aperture, the septum separates the nasal cavity in midline; the inferior and middle turbinate can be identified. (**b**) The endoscope is introduced through the left nostril. The septum is located on the left side of the image, the middle and inferior turbinates as appendices protruding from the lateral wall of the nasal cavity. The choana or posterior nasal aperture opens into the nasopharynx. (**c**, **d**) Image of the middle meatus, anterior part. Below and lateral to the middle turbinate, the first prominence seen is the uncinate process and posteriorly the bulla ethmoidalis (anterior ethmoid cells). The inferior semilunar hiatus is the area between them; the space between them and the lamina papyracea is the ethmoid infundibulum, common drainage of the anterior ethmoid cells, maxillary sinus, and in some cases the frontal sinus depending on the uncinate process attachment. Anterior to the middle turbinate, there is a craneocaudal prominence called lacrimal crest, which covers the nasolacrimal duct. In the dry skull, the uncinate process is located between the anterior and posterior fontanelles that are closed by mucosa in life. The maxillary ostium is hidden by the uncinate process located between this and the bulla ethmoidalis.

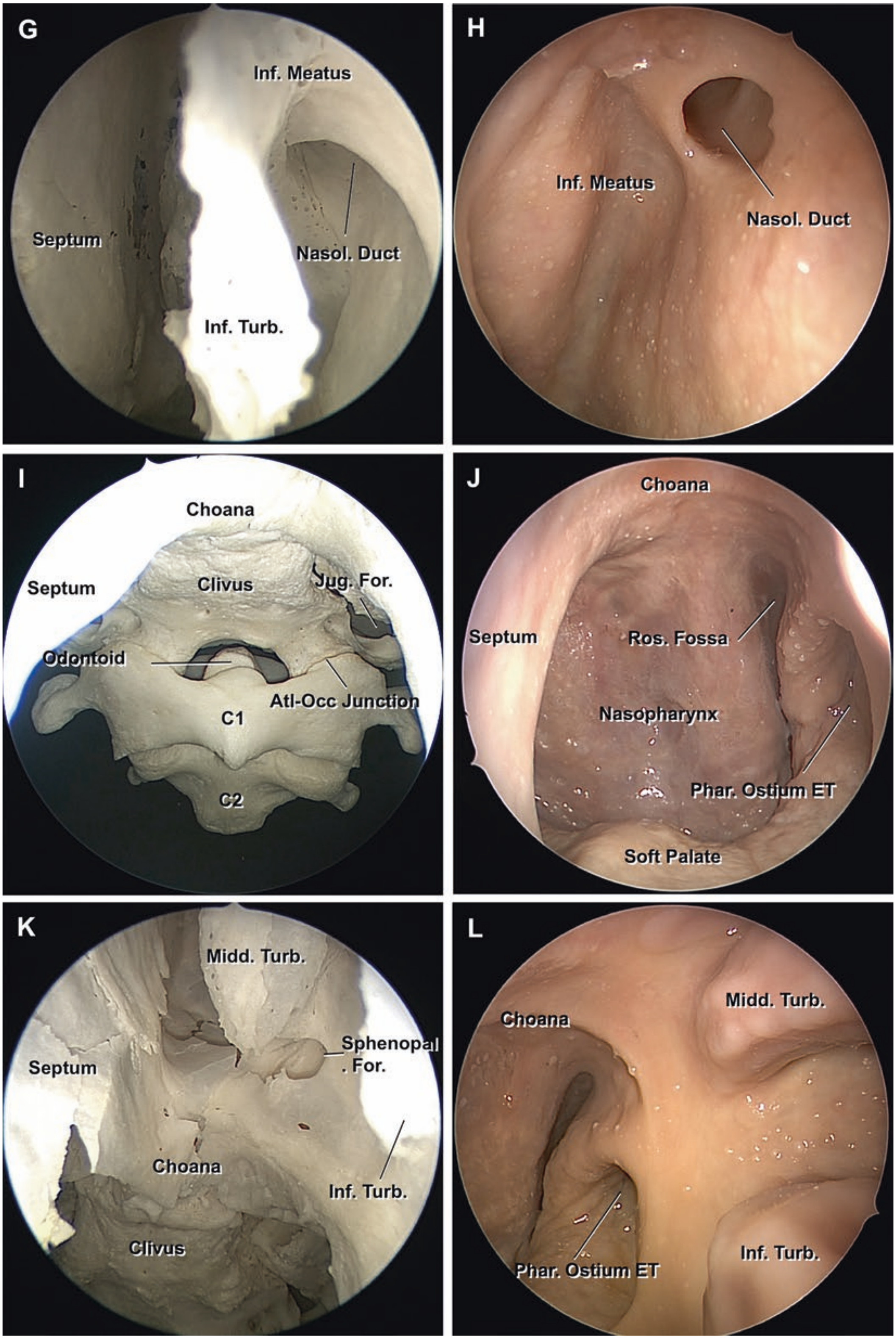

Fig. 2.8 (contined) (**e**, **f**) The sphenoidal ostium is found posteriorly, usually medial to the superior turbinate and lateral to the septum. (**g**, **h**) View of the anterior part of the left inferior meatus, the nasolacrimal duct ostium is seen with the 45° endoscope. (**i**, **j**) View of the left choana with the 0° endoscope. The nasal cavity opens onto the nasopharynx through the posterior nasal apertures called choanae. The nasopharynx faces the middle clivus, inferior clivus, and craniocervical junction. Endoscopic endonasal procedures give access to pathologies up to C2 body. The Eustachian tube opens into the nasopharynx. (**k**, **l**) Equivalent views in a dry skull and an anatomical specimen of the posterior aspect of the nasal cavity (45° endoscope). The sphenopalatine foramen is posterior and superior to the tail of the middle turbinate. The fish-eye view provided by the endoscope distorts this relationship. The posterior septal arteries that supply the septum and thus the nasoseptal flap, cross from lateral to medial coming from this foramen. *Ant* anterior, *Crib* cribriform, *ET* eustachian tube, *For* foramen, *Inf* inferior, *Jug* jugular, *Midd* middle, *Nasol* sasolacrimal, *Phar* pharyngeal, *Post* posterior, *Proc* process, *Sphen* sphenoidal, *Sphenopal* sphenopalatine, *Ros* Rosenmuller, *Sup* superior, *Turb* turbinate, *Uncin* uncinate (All: Used with permission from Peris-Celda et al. [33])

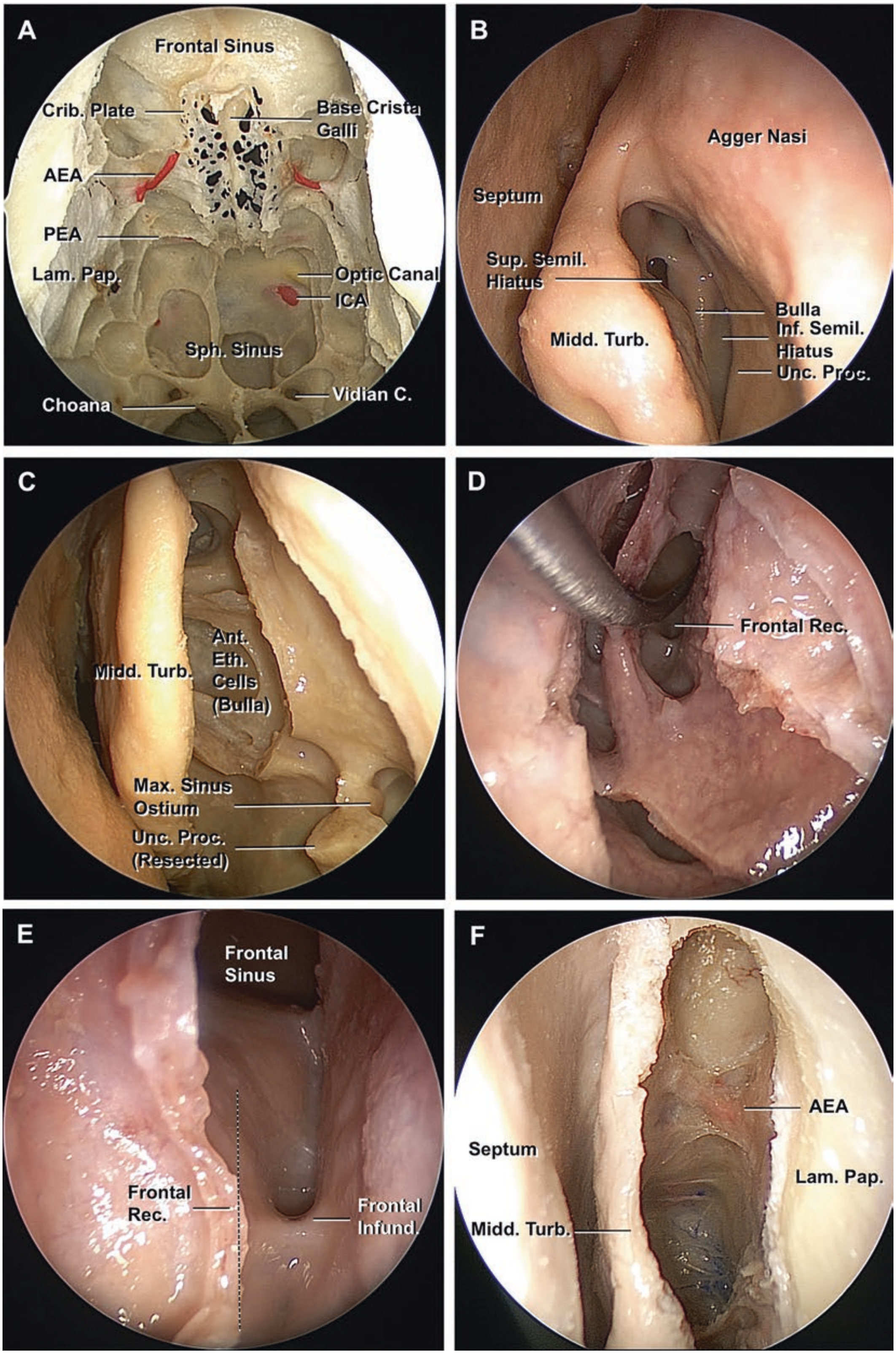

Fig. 2.9 (**a**–**f**) Anatomy and dissection of the frontal and ethmoid sinuses. (**a**) Complete sphenoidotomy, ethmoidectomy, and opening of the frontal sinus bilaterally in a dry skull. The osseous elements that form the anterior skull base are completely exposed; the lateral limits are represented by the lamina papyracea bilaterally. (**b**) Middle meatus superiorly. The agger nasi is bulging anterior to the middle turbinate attachment. The inferior semilunar hiatus is located between the uncinate and bulla, whereas the superior semilunar hiatus is located between the middle turbinate and the bulla. (**c**) Resection of the bulla ethmoidalis and the uncinate process. The maxillary ostium is visualized behind the uncinate process. (**d**) View with the 45° endoscope of the frontal recess. (**e**) Frontal recess. (**f**) Resection of the left ethmoid labyrinth, the middle turbinate is medial and lamina papyracea lateral. *AEA* anterior ethmoidal artery, *C* canal, *Crib* cribriform, *ICA* internal carotid artery, *Inf* inferior, *Infund* infundibulum, *Lam* lamina, *Pap* papyracea, *PEA* posterior ethmoidal artery, *Proc* process, *Rec* recess, *Sphen* sphenoidal, *Semil* semilunar, *Sup* superior, *Turb* turbinate, *Uncin* uncinate

opens medial and at the caudal level of the end of this turbinate approximately 12 mm superior to the choana (Fig. 2.4a–l). If there is a supreme turbinate, the sphenoidal ostium is located usually lateral to this structure.

The ethmoidal labyrinths consist of numerous paper-thin ethmoidal air cells between two vertical plates. The vertical second portion of the basal lamella of the middle turbinate divides the ethmoidal cells in anterior and posterior ethmoidal cells according to the surgical classification. As opposed to the surgical classification, the anatomical classification considers a division of the ethmoidal cells in anterior, middle, and posterior groups. It is considered that, on average, there are 11 anterior ethmoidal air cells, 3 middle, and 6 posterior [8]. The anatomical classification considers the cells forming the bulla ethmoidalis as middle ethmoidal cells and has been criticized by some authors [11]. The lateral surface (orbital plate) of the labyrinth is part of the medial orbital wall. The medial surface of the labyrinth forms part of the superolateral nasal wall. It appears as a thin lamella that descends from the inferior surface of the cribriform plate and ends as the middle turbinate. In the isolated bone, many air cells are open, but in the articulated skull, they are closed by proximity to adjoining bones, except where they drain into the nasal cavity. The walls of the air cells anterior to the orbital plate are completed by the lacrimal bone and frontal process of the maxilla. The air cells superiorly are completed by the frontal bone that also converts the anterior and posterior ethmoidal grooves in ethmoidal canals. Both anterior and posterior ethmoidal arteries diverge medially from the ophthalmic artery in the orbit through the ethmoidal conducts or canals. These may be totally covered by the bone or hanged into the fovea ethmoidalis placing them at risk of inadvertent injury during surgery. Care must be taken during the approach to avoid tearing of these arteries as they may retract into the orbit where ongoing bleeding may cause intraorbital hematoma and even blindness due to high ocular pressure. One study found that in 43% of the cases, the anterior ethmoidal canal travels entirely through the ethmoidal labyrinth placing the artery at risk in surgery [14]. Preoperative assessment with CT scan is important to assess the location of the canal. The ethmoidal arteries travel to the lateral aspect of the cribriform plate where they give meningeal and nasal branches that later anastomose with the septal branches from the sphenopalatine artery. The posterior ethmoidal artery is usually smaller than the anterior and absent in 30% of the cases. The distance between the anterior lacrimal crest of the maxilla and the anterior ethmoidal foramen is 22–24 mm, between the anterior and posterior ethmoidal foramina is 12–15 mm, and from the posterior to the optic canal is 3–7 mm [15]. On the posterior surface of the ethmoid, open air cells are covered by the sphenoidal conchae and the orbital process of the palatine bone [8] (Figs. 2.5a–f and 2.9a–f).

The frontal recess is the most anterior and superior portion of the anterior ethmoid complex that leads to and communicates with the frontal sinus. It has an hourglass shape with the constricted region at the level of the frontal ostium. Its medial wall is the most anterior and superior part of the middle turbinate, the lateral wall is the lamina papyracea, and a posterior margin exists when the bulla reaches the skull base, separating the frontal recess from the suprabullar recess. The floor of the frontal recess is so variable that there is no definition. If an ethmoid bone is seen from above, the frontal recess narrows toward the frontal ostium through the frontal infundibulum. From the level of the ostium, the frontal recess widens again in the inferior and posterior direction [11] (Figs. 2.5a–f and 2.6a–f).

The ethmoidal cells in contact with the frontal sinus or related to the frontal recess are called frontoethmoidal cells. They vary greatly the anatomy of the frontal recess, the antechamber to the frontal sinus. The agger nasi is the most superior remnant of the first ethmoturbinal and persists as a mound or crest that can be seen anterior and superior to the middle turbinate [11]. When this area of the lateral nasal wall undergoes pneumatization, it is called an agger nasi cell (Figs. 2.4a–l and 2.9a–f). It is important as it dictates the frontal sinus drainage pathway. The agger nasi cell is the most anteriorly placed frontoethmoidal cell and is present in 98.5% of patients. In the simplest situation, the patient has a single agger nasi cell without other frontoethmoidal cells. If it is present, it can usually be seen on the CT scan anterior to the middle turbinate [13]. Most of the agger nasi cell is anterior to the uncinate, but the posterior half of the agger nasi cell has an intimate relationship with the upward extension of the uncinate process. In most cases, the uncinate and the medial wall of an agger nasi cell implant on the lamina papyracea [13]. In these cases, when the uncinate process inserts in the papyracea below the natural frontal sinus ostium, the frontal sinus drains directly into the middle meatus. There can be one (Kuhn type I) or multiple (Kuhn type II) ethmoidal cells above the agger nasi cell in the frontal recess. None of them pass the frontal beak. The frontal beak is formed by the internal bony indentation at the level of the nasofrontal suture [9]. Kuhn type III is a single cell in the frontal recess with pneumatization into the frontal sinus less than 50% of its height; the posterior wall is part of the frontal recess. Kuhn type IV cell is a rare isolated cell in the frontal sinus that extends more than 50% of the height of this sinus. If there are ethmoidal cells located above the ethmoidal bulla, they are called suprabullar cells, their cranial boundary is the skull base, and they do not extend to the frontal sinus. If these cells extend into the frontal sinus from behind the frontal recess, they are called frontal bulla cells. If a single or multiple ethmoidal cells extend into the orbit from the frontal recess, they are classified as supraorbital cells [16].

The medial wall of the orbit can be approached endoscopically as well as the inferior wall medial to the infraorbital nerve. Relationships of anatomical structures are of essential importance in performing endoscopic approaches to these areas. The orbit is anterior to the optic nerve prominence in the sphenoidal sinus, superior to the maxillary sinus, and lateral to the ethmoidal cells.

Of importance in surgery, the most posterior ethmoid cell may pneumatize laterally and superiorly into the sphenoidal sinus and then are called sphenoethmoidal cells or Onodi cells which are located between the sphenoidal sinus and the planum. This is important surgically to identify as the optic nerve and carotid artery may lie unprotected within this cell and if unrecognized may result in inadvertent injury (Figs. 2.4a–l and 2.11b). Its prevalence was 65.3% in one study [17].

Different degrees of ethmoid bone resection are required in the endoscopic endonasal approaches to the skull base and are tailored depending on the pathology. They can vary from a complete ethmoidectomy in an anterior craniofacial resection (Figs. 2.9a–f, 2.10a–f, and 2.11a–l) to a partial posterior ethmoidectomy in transsphenoidal approaches (Fig. 2.11a–l).

Sphenoidal Sinus and Related Structures

The sphenoid bone has three parts: body; two lesser wings, which spread outward from the superolateral part of the body; two greater wings, which spread upward from the lower part of the body; and two pterygoid processes with their medial and lateral pterygoid plates directed downward from the body (Fig. 2.12a–f). The body of the sphenoid bone is more or less cubical and contains the sphenoidal sinus. Approaches to the planum, ventral optic canal, tuberculum, upper clivus, and upper part of the middle clivus can be performed through the sphenoidal sinus in the midline. Most of the endoscopic skull base approaches involve opening of the sphenoidal sinus in order to have a clear reference of midline and the internal carotid arteries.

The neurovascular relationships of the sphenoid bone are among the most complex of any bone, and the intracranial relationships of the recesses of the sphenoidal sinus are keys to understand and accurately perform endoscopic endonasal approaches (Fig. 2.11a–l). The sphenoidal sinus is the center of the endonasal approaches to the sellar and parasellar areas.

The sphenoidal sinus separates the cavernous sinuses, the cavernous segments of the internal carotid arteries, and the optic, extraocular, and trigeminal nerves. The anterior wall faces the nasal cavity; the posterior wall of the sphenoidal sinus is formed by the part of the clivus between the dorsum sellae and the basilar part of the occipital bone. The lateral wall of the sphenoidal sinus usually borders the medial wall of the cavernous sinus and can extend laterally beyond the line connecting the Vidian canal and foramen rotundum. The superior wall of the sphenoidal sinus is formed by the planum and the sellar wall, whereas the inferior wall of the sphenoidal sinus faces the nasopharynx.

The sphenoidal sinus opens through the sphenoidal ostia into the nasal cavity. The anterior wall of the sphenoidal sinus is concave with both ostia medial and superior. The posterior part of the ethmoid bone, containing the posterior ethmoidal cells, articulates with the sphenoid laterally to the ostia. The recess formed at this articulation between the body of the sphenoid and the posterior aspect of the ethmoid bone is called the sphenoethmoidal recess. It is important to identify the presence of an Onodi cell in preoperative studies [18].

There are different patterns of pneumatization of the sphenoidal sinus of importance in surgery that can be summarized into three major types in the adult: conchal (the inferior part of the sella is solid bone, 1–5%), presellar (the air cavity does not penetrate beyond a vertical plane parallel to the anterior sellar wall, 23–24%) and, the most common, the sellar type (pneumatized below the sella up to the clivus, 67–76%). The sphenoidal sinus usually has septations that can vary greatly. In 68% of the cases, a single major septum separates the sinus in two, and the septations are often located off midline. The most common type of sphenoidal sinus has multiple small cavities in the large paired sinuses [15]. In 87% of the cases, there is an sphenoidal septation contacting the carotid [19].

Endonasally, in the sellar type of sphenoidal sinus, the pituitary fossa corresponds to the sellar prominence, a bulging oval area in the midline, surrounded laterally by the carotid prominence which corresponds to the anterior loop of the intracavernous carotid artery and superiorly to the clinoid segment of the carotid artery (Fig. 2.11a–l). The carotid artery can be prominent in the anterior part of the sella (presellar segment) in 98%, below the sellar floor (infrasellar segment) in 80%, and in the posterolateral part of the sinus (retrosellar segment or vertical segment) in 78% of cases. Any part of the prominences can be present and the others absent. Only the presellar segment is present in the presellar type of sphenoidal sinus. The bone over the cavernous carotid just below the tuberculum can be dehiscent in 10% of cases and less than 0.5 mm thick in 90%. The shortest distance between carotids in the sinus is located just below the tuberculum in 72% of cases, at the level of the floor of the sella in 20%, and at the clivus in 8% [2]. The segment of the carotid artery adjacent to the pituitary gland is also called parasellar and, the segment adjacent to the clivus, the paraclival segment. The optic nerves in the optic canals protrude into the superolateral part of the sinus above the carotid prominences and head medial to lateral in a posterior to anterior direction. The optic canals can be dehiscent in up to 8% cases [4]. The lateral opticocarotid recess is located lateral to the carotid prominence, between the optic and carotid prominences. It has a triangular

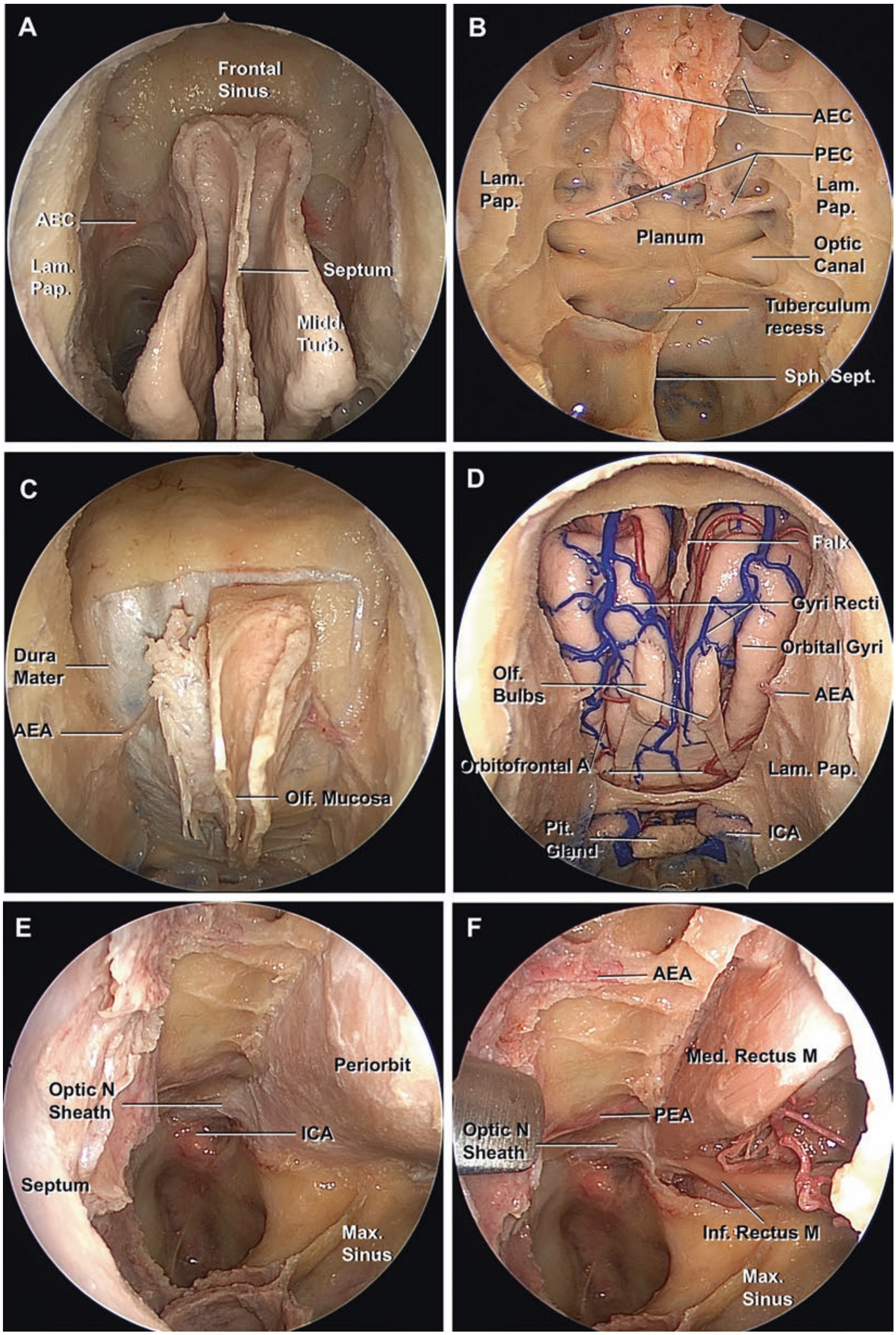

Fig. 2.10 (**a**–**f**) Anatomy of the anterior endoscopic craniofacial resection (**a**) Bilateral ethmoidectomy and Draf 3 of the frontal sinus. (**b**) The middle turbinate and superior part of the septum have been resected bilaterally and the sphenoidal sinus opened. (**c**) The bone of the anterior cranial fossa surrounding the right olfactory fossa has been drilled to expose the dura mater. (**d**) The dura mater of the anterior skull base has been resected and the falx divided anteriorly so that the olfactory bulbs, gyri recti, and part of the orbital gyri are exposed from orbit to orbit in an endoscopic craniofacial resection. The pituitary gland and both parasellar internal carotid artery are dissected posteriorly. (**e**, **f**) Endoscopic endonasal approach to the left orbit. To approach the orbit, medial maxillary antrostomy and unilateral ethmoid labyrinthectomy with lamina papyracea removal need to be performed. For intraconal pathologies, the periorbit needs to be incised, and the pathologies are commonly approached between the medial and inferior rectus muscles. *A* artery, *AEA* anterior ethmoidal artery, *C* canal, *Crib* cribriform, *ICA* internal carotid artery, *Inf* inferior, *Lam* lamina, *Max* maxillary, *N* nerve, *Olf* Olfactory, *Pap* papyracea, *PEA* posterior ethmoidal artery, *Pit* pituitary, *Sphen* sphenoidal, *Semil* semilunar, *Sup* superior, *Turb* turbinate, *Uncin* uncinate

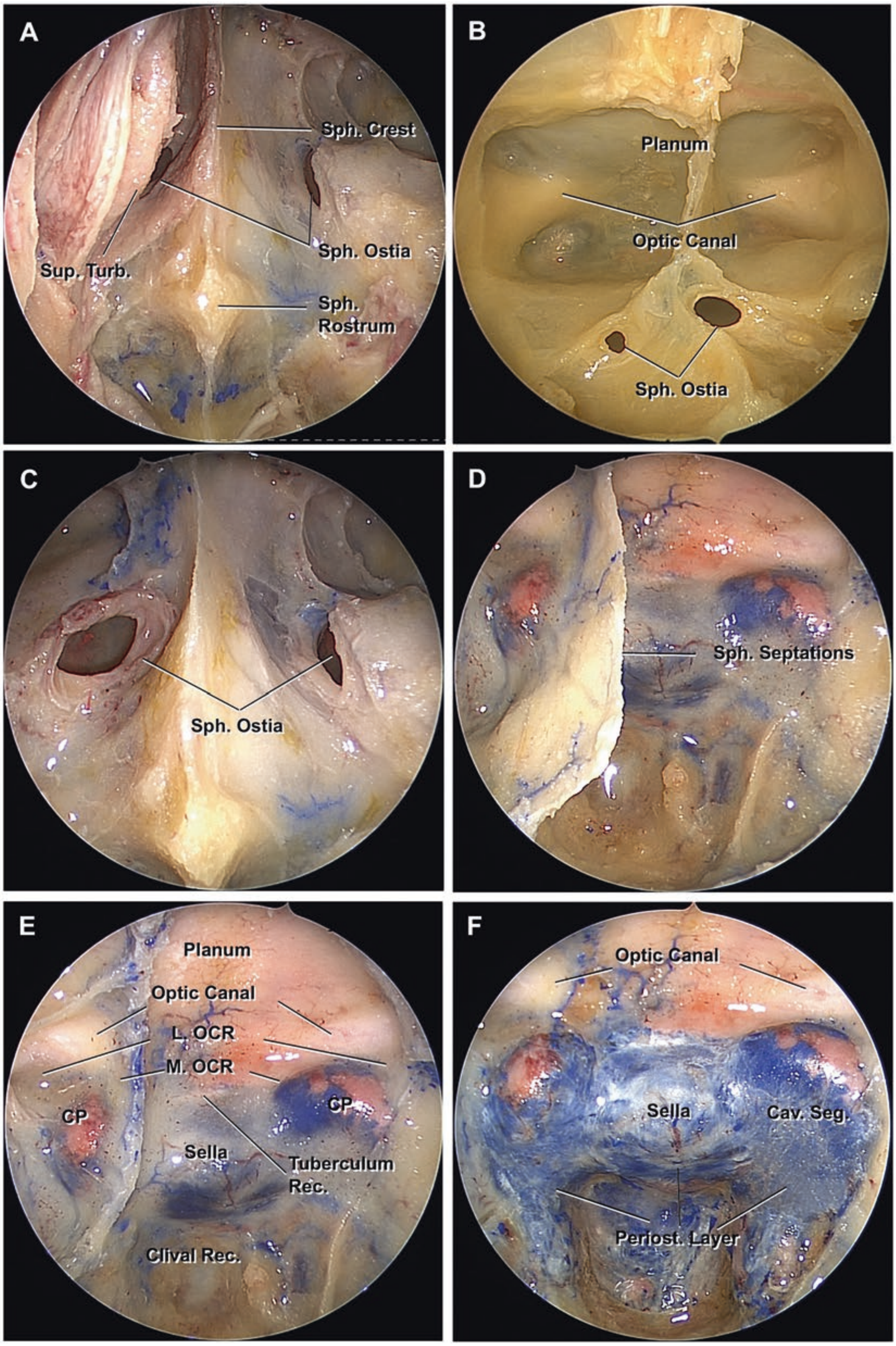

Fig. 2.11 (**a**–**l**) Transsphenoidal approach. (**a**) View of the sphenoidal rostrum and crest; reliable references for midline. The sphenoidal ostia are medial to the superior turbinates. Compare how the access to the sphenoidal sinus is enlarged with a posterior ethmoidectomy (left side). (**b**) Example of bilateral Onodi cell. The ethmoid sinus pneumatizes posteriorly over the sphenoidal sinus and involves both optic canals. (**c**) View of the anterior wall of the sphenoidal sinus once bilateral posterior ethmoidectomies are performed. (**d**) View of the sphenoidal sinus; usually the intrasphenoidal septations are related to the internal carotid artery posteriorly. (**e**) Anatomic references in the sphenoidal sinus, the sellar prominence in the center, carotid prominences laterally, and optic canals superiorly. Note the tuberculum, opticocarotid, and clival recesses. (**f**) After drilling the bone over the sella and cavernous sinus, the periosteal layer of the dura mater is exposed. (**g**) The periosteal layer of the dura mater is removed. The venous channels that connect both cavernous sinuses run between these two layers. (**h**) The dura mater has been resected to expose the anatomy of the sellar region, suprasellar region, and cavernous sinus. (**i**) Lateral view of the left cavernous sinus. The sympathetic plexus travels with the internal carotid artery, and a branch enters the orbit; the gasserian ganglion with the first and second division of the trigeminal

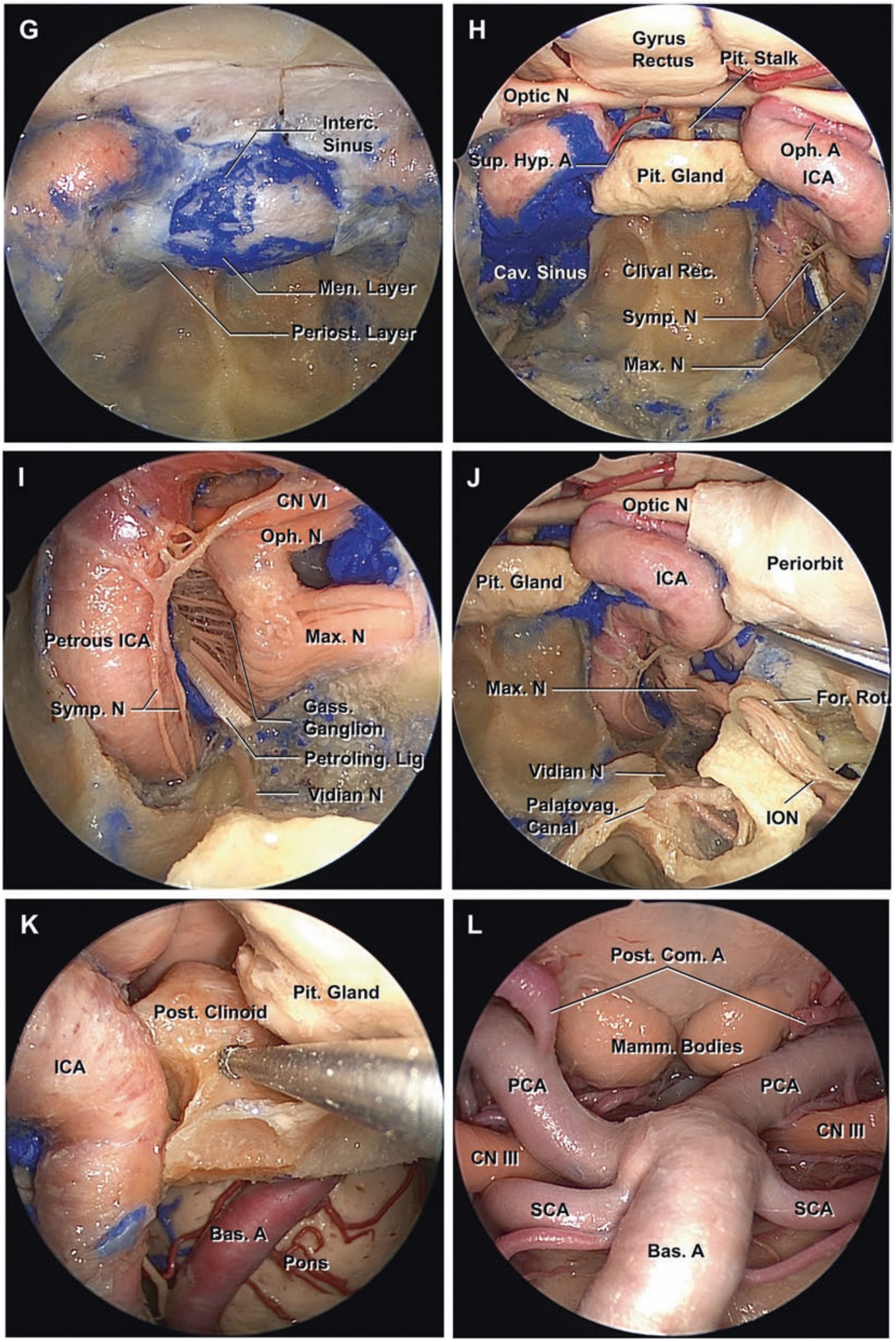

Fig. 2.11 (continued) nerve can be seen lateral to the internal carotid artery. The VI cranial nerve is located medial to the first division of the trigeminal nerve. The Vidian nerve is related to the lateral aspect of the lacerum segment of the internal carotid artery. (**j**) View of the left cavernous sinus. The maxillary nerve has been followed anteriorly passing through the foramen rotundum and in continuation with the infraorbital nerve. The Vidian nerve has been followed up to the pterygopalatine fossa anteriorly. In surgery, the foramen rotundum and maxillary nerve are references to the gasserian ganglion, whereas the Vidian nerve is a reference for the internal carotid artery. The palatovaginal canal is medial to the Vidian canal and runs medially and posteriorly. (**k**) When the posterior clinoids need to be approached, the pituitary gland has to be transposed medially and superiorly. (**l**) View of the interpeduncular fossa after removal of the dorsum sellae and posterior clinoids. The gland is located above and anterior to the endoscope. *A* artery, *Bas* basilar, *Cav* cavernous, *CN* cranial nerve, *Com* communicating, *CP* carotid prominence, *For* foramen, *Gass* Gasserian, *Hyp* hypophyseal, *ICA* internal carotid artery, *ION* infraorbital nerve, *Mamm* mammillary, *Max* maxillary, *M. OCR* medial opticocarotid recess, *L. OCR* lateral opticocarotid recess, *Men* meningeal, *N* nerve, *Oph* ophthalmic, *Palatovag* palatovaginal, *PCA* posterior cerebral artery, *Periost* periosteal, *Post* posterior, *Pit* pituitary, *Rec* recess, *Rot* rotundum, *SCA* superior cerebellar artery, *Seg* segment, *Sup* superior, *Symp* sympathetic, *Sph* sphenoidal (All: Used with permission from Peris-Celda et al. [33])

shape and corresponds to the optic strut base, and its medial projection corresponds to the clinoid segment of the internal carotid artery. This recess may completely pneumatize into the anterior clinoid process. This pneumatization is often responsible for the postoperative nasal cerebrospinal fluid leaks that can occur after performing an anterior clinoidectomy through a transcranial approach. The tuberculum recess is located between the sellar prominence and the planum and in most cases corresponds to the tuberculum endocranially. The diaphragma sellae attachment is located at the level of the tuberculum recess or slightly inferior. The lateral extensions of the tuberculum recess have been described as the medial opticocarotid recesses, where the optic prominence meets the carotid prominence medial to the carotid artery. Two junction points can be recognized in the medial opticocarotid recess: the medial opticocarotid point, where the optic nerve prominence meets the carotid prominence medially; and the caroticosellar point, where the carotid prominence meets the sellar prominence superiorly. A clinically relevant middle clinoid process (greater than 1.5 mm) is present in 21.1% of cases and can form a caroticoclinoid ring, a bony ring joining the anterior and middle clinoid processes, in 2.94%. The base of the middle clinoid process is most frequently located approximately 1 mm inferior and lateral to the caroticosellar junction point, and its highest point is further inferior and lateral. On average, the middle clinoid is 4.75 mm inferior to the medial opticocarotid point [20]. The average distance between the medial edges of the optic nerves entering the optic canal into the sinus is 15.75 mm and does not seem to be affected by the pneumatization of the recesses. There is no recess or prominence that represents the prechiasmatic sulcus inside the sphenoidal sinus, although its location can be predicted between both optic prominences medially [21].

Extensive pneumatization of the sphenoidal sinus may produce several recesses that help in recognizing structures during surgery or provide routes of access to specific areas. The sphenoidal recesses may be divided into anterior, lateral, and posterior groups. An anterior recess of the sphenoidal sinus can be encountered anterior to a line passing through the sphenoidal crest bulging into the maxillary sinus above the sphenopalatine artery and foramen (12% of cases). A lateral recess can be found in 46% of the cases when the pneumatization extends lateral to the Vidian-rotundum line. The lateral recess can be divided in three subtypes: pneumatization that extends into the greater wing of the sphenoid bone, inferiorly into the pterygoid, or as a combination of both. The posterior wall of the sphenoidal sinus is related to the basilar process of the sphenoidal body that if pneumatized constitutes the clival recess or posterior recess. Three subtypes of clival recesses can be found: the dorsum type, which extends above the horizontal plane of the floor of the pituitary and into the dorsum sellae; the subdorsum type, which lies between the horizontal plane of the floor of pituitary fossa and the horizontal plane passing through the anterior opening of Vidian canals; and the occipital type, which extends inferiorly below the horizontal plane crossing the anterior opening of the paired Vidian canals. In one study, of the 68 clival-type sinuses found on 100 CT images, the dorsum type was found in 16 (23.5%), the subdorsum type in 43 (63.2%), the occipital type in 1 (1.5%), and the combined dorsumoccipital type in 8 (11.8%). In the endonasal approach, the subdorsum type is encountered immediately as the sinus is entered, whereas the dorsum type may be partially hidden behind the pituitary fossa, and the clival type may be partially hidden below the floor of the sphenoidal sinus. There is controversy over whether the sphenoidal sinus extends into the occipital bone. Some authors reported that the sinus never extends below the spheno-occipital synchondrosis, whereas others have found that the sinus extends into the occipital bone [21].

The inferior wall of the sphenoidal sinus is usually at the level of the base of the pterygoid processes, with the pterygoid or Vidian canal located superior to the medial pterygoid plate. It can be frequently exposed and even dehiscent within the sinus which can put this structure at risk during surgery. The Vidian canal, with the Vidian nerve and artery, is a landmark to find the carotid artery genu in the foramen lacerum where it courses superiorly from the horizontal petrous segment to enter the cavernous sinus in the paraclival space. The posterior end of the Vidian canal is located inferolateral to the anterior end of the carotid canal and anterior genu of the petrous carotid. The anterior end of the Vidian canal, which opens into the medial part of the posterior wall of the pterygopalatine fossa, is funnel or trumpet shaped, with the anterior part facing the pterygopalatine fossa being wider than the posterior part. The anterior opening of the Vidian canal is located inferomedial to the foramen rotundum on the anterior surface of the pterygoid process of the sphenoid bone [21] (Figs. 2.12a–f and 2.13a–f). The Vidian nerve is formed by the greater superficial petrosal and deep petrosal nerves. The carotid nerve, a branch of the sympathetic ganglia, branches into two parts near the genu of the petrous carotid artery. One of them constitutes the deep petrosal nerve that joins the greater superficial nerve to form the Vidian nerve, and the other follows the internal carotid artery. Apart from an important landmark, the Vidian nerve is of importance for lacrimal and mucous secretion and must be preserved and transposed if needed [22–24]. The palatovaginal canal (Fig. 2.13a–f), also called palatosphenoidal canal [25] with the pharyngeal artery and nerve, is located between the sphenoidal apophysis of the palatine bone and the sphenoid. Its anterior opening is located in the pterygopalatine fossa medial to the Vidian canal. It opens posteriorly into the nasopharynx. It can be used as a medial landmark for the Vidian canal. To visualize this structure endoscopically, the sphenoidal process of the palatine bone has to be resected [22].

When a lateral recess is present, the second division of the trigeminal nerve can be distinguished as a prominence

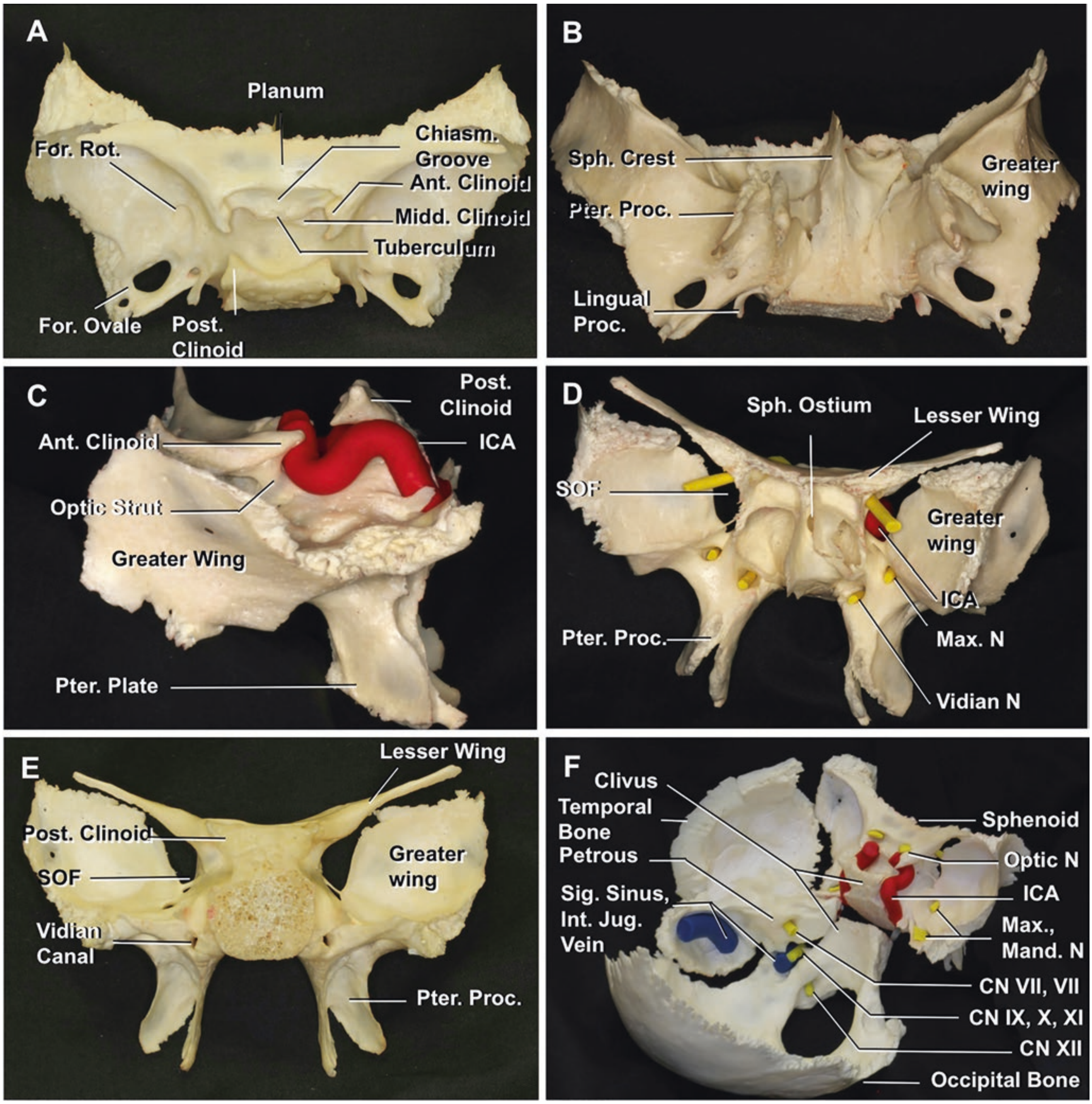

Fig. 2.12 (**a**–**f**) Sphenoid bone and related structures. (**a**) Superior view. (**b**) Inferior view. (**c**) Lateral view; the internal carotid artery has been represented in the carotid sulcus with molding material. (**d**) Anterior view of the sphenoid. Note the anatomical location of the optic, maxillary, and Vidian nerves and the internal carotid artery in the sphenoid bone. (**e**) Posterior view of the sphenoid. (**f**) The sphenoid articulates posteriorly with the occipital and temporal bones. The clivus is formed by the median segment of the sphenoid body superiorly and the median portion of the occipital bone inferiorly. The junction of the clivus and petrous segment of the temporal bone forms the petroclival region. *Ant* anterior, *Chiasm* chiasmatic, *CN* cranial nerve, *For* foramen, *ICA* internal carotid artery, *Int* internal, *Jug* jugular, *Mand* mandibular, *Max* maxillary, *Midd* middle, *N* nerve, *Post* posterior, *Proc* process, *Pter* pterygoid, *Rot* rotundum, *Sig* sigmoid, *Sph* sphenoidal, *SOF* superior orbital fissure (All: Used with permission from Peris-Celda et al. [33])

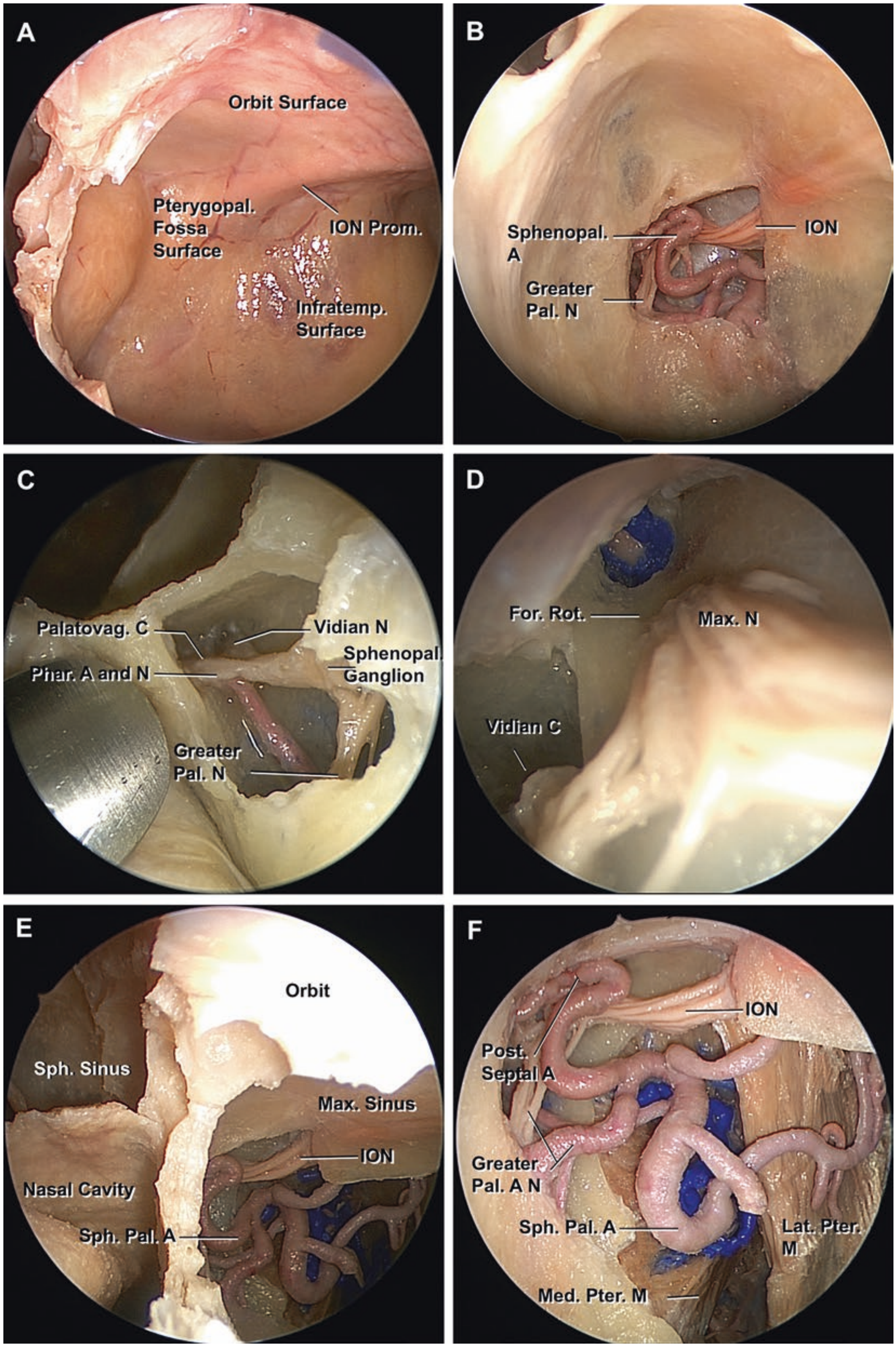

Fig. 2.13 (**a**–**f**) Anatomy of the maxillary sinus, left side. (**a**) View inside the maxillary sinus after an endoscopic endonasal medial maxillary antrostomy; the infraorbital prominence is located in the roof of the sinus, the inferior orbital wall. (**b**) Opening of the posterior wall of the maxillary sinus medial to the infraorbital nerve into the pterygopalatine fossa. (**c**) Medial aspect of the left pterygopalatine fossa; note the palatovaginal canal medially and the Vidian canal laterally. (**d**) In the pterygopalatine fossa, the maxillary nerve is superior and lateral to the Vidian canal. (**e**) General view of the left maxillary sinus and its relationships with the nose and paranasal sinuses, orbit above, nasal cavity laterally and sphenoid sinus superiorly, posteriorly, and laterally. The posterior wall of the maxillary sinus gives access to the pterygopalatine and infratemporal fossa. (**f**) Close-up view of the pterygopalatine fossa and infratemporal fossa. The terminal branch of the internal maxillary artery is the sphenopalatine artery. *A* artery, *C* canal, *For* foramen, *Infratemp* infratemporal, *ION* infraorbital nerve, *Lat* lateral, *M* muscle, *Max* maxillary, *Med* medial, *N* nerve, *Prom* prominence, *Pal* palatine, *Pter* pterygoid, *Pterygopal* pterygopalatine, *Sphenopal* sphenopalatine, *Rot* rotundum

7–15 mm long covered by thin bone that may be dehiscent. The trigeminal ganglion and the first and third divisions are separated from the lateral wall of the sphenoidal sinus by the carotid artery. Above and below the second division of the trigeminal nerve, through the anteromedial and anterolateral triangles of the skull base, the middle fossa can be reached endoscopically [15] (Fig. 2.11a–l). The third division of the trigeminal nerve is located lateral to the paraclival segment of the internal carotid artery and has an almost vertical direction through the foramen ovale and into the infratemporal fossa. Meckel's cave can be reached endoscopically through the quadrangular space defined between the paraclival carotid medially, the horizontal segment of the petrous carotid inferiorly, the second division laterally, and VI cranial nerve superiorly [26, 27] (Fig. 2.11a–l). Depending on the sphenoidal sinus pneumatization, the third division of the trigeminal nerve may be seen coursing inferiorly in the posterior aspect of the lateral recess.

Maxillary Sinus and Related Structures

The maxillary sinus when completely developed in the adult is a pyramidal cavity in the body of maxilla, having an apex that points laterally and extends into the zygomatic process. The base, on the lateral wall of the nasal cavity, is the site of the maxillary hiatus. The posterior wall of the sinus is pierced by the alveolar canals which transmit the posterosuperior alveolar nerves to the molar teeth. These canals may produce protrusions in the wall of the sinus. The floor is formed by the alveolar process of maxilla. The superior surface of the maxillary sinus forms the inferior wall of the orbit and contains the infraorbital nerve in its groove and canal. The middle and anterior superior alveolar nerves leave the infraorbital nerve laterally and follow the lateral wall of the sinus to reach premolars, canine, and incisive teeth anteriorly. The posterior wall of the maxillary sinus articulates with the palatine bone and in its lateral part gives access to the infratemporal fossa. The infraorbital nerve prominence inside the sinus guides the surgeon posteriorly to the pterygopalatine fossa and foramen rotundum behind the posterior wall of the maxillary sinus (Figs. 2.4g–l and 2.13a–f). On occasions, ethmoidal cells can develop into the maxillary sinus inferior to the orbit (Haller cells).

The maxillary sinus is a door that opens several important surgical corridors: medially and superiorly providing access to the inferomedial wall of the orbit, posteriorly and laterally to the infratemporal fossa, and posteriorly and medially to the pterygopalatine fossa and pterygoid processes. The transpterygoid approach is the first step in localizing the petrous internal carotid artery and provides access to the ventral skull base superior (suprapetrous space) and inferior to the petrous internal carotid artery (infrapetrous, parapharyngeal space) [28].

2.2 Sellar and Parasellar Regions

Pituitary Gland

The pituitary gland lies within the pituitary fossa of the sphenoid bone where it is covered superiorly by a circular diaphragma sellae of dura mater. The pituitary gland is a reddish-gray, ovoid body, about 12 mm in transverse and 8 mm in anteroposterior diameter, and with an average adult weight of 500 mg (Figs. 2.11a–f and 2.14a–e). It is continuous with the pituitary stalk and infundibulum, a hollow, conical inferior process arising from the tuber cinereum of the hypothalamus. The gland is covered entirely by the endosteal or meningeal layer of dura mater. The latter is pierced centrally by an aperture for the infundibulum and separates the anterior-superior aspect of the pituitary from the optic chiasm. Inferiorly, the pituitary is separated from the floor of the fossa by a venous sinus that communicates with the circular sinus. The infundibulum has a central infundibular stem, which contains neural hypophyseal connections, and is continuous with the median eminence of the tuber cinereum. The anterior lobe wraps around the lower part of the pituitary stalk to form the pars tuberalis. The posterior lobe is softer, almost gelatinous, and is more densely adherent to the sellar wall. The anterior lobe is firmer and is more easily separated from the sellar walls. The gland's width is the same or more than either its depth or its length in most patients. Its inferior surface usually conforms to the shape of the sellar floor, but its lateral and superior margins vary in shape because these walls are composed of soft tissue. If there is a large opening in the diaphragm, the gland tends to be concave superiorly in the area around the stalk. The superior surface may become triangular as a result of being compressed laterally and posteriorly by the carotid arteries. As the anterior lobe is separated from the posterior lobe, there is a tendency for the pars tuberalis to be retained with the posterior lobe. Small intermediate lobe cysts are frequently encountered during separation of the anterior and posterior lobes [2, 29].

Cavernous Sinus

The cavernous sinuses are paired parasellar venous sinus structures. They are encased between the meningeal and periosteal layers of dura mater. The internal carotid artery runs through the cavernous sinus, and it gives the name to this segment. The III, IV, and VI cranial nerves and the first division of the trigeminal nerve pass through the cavernous

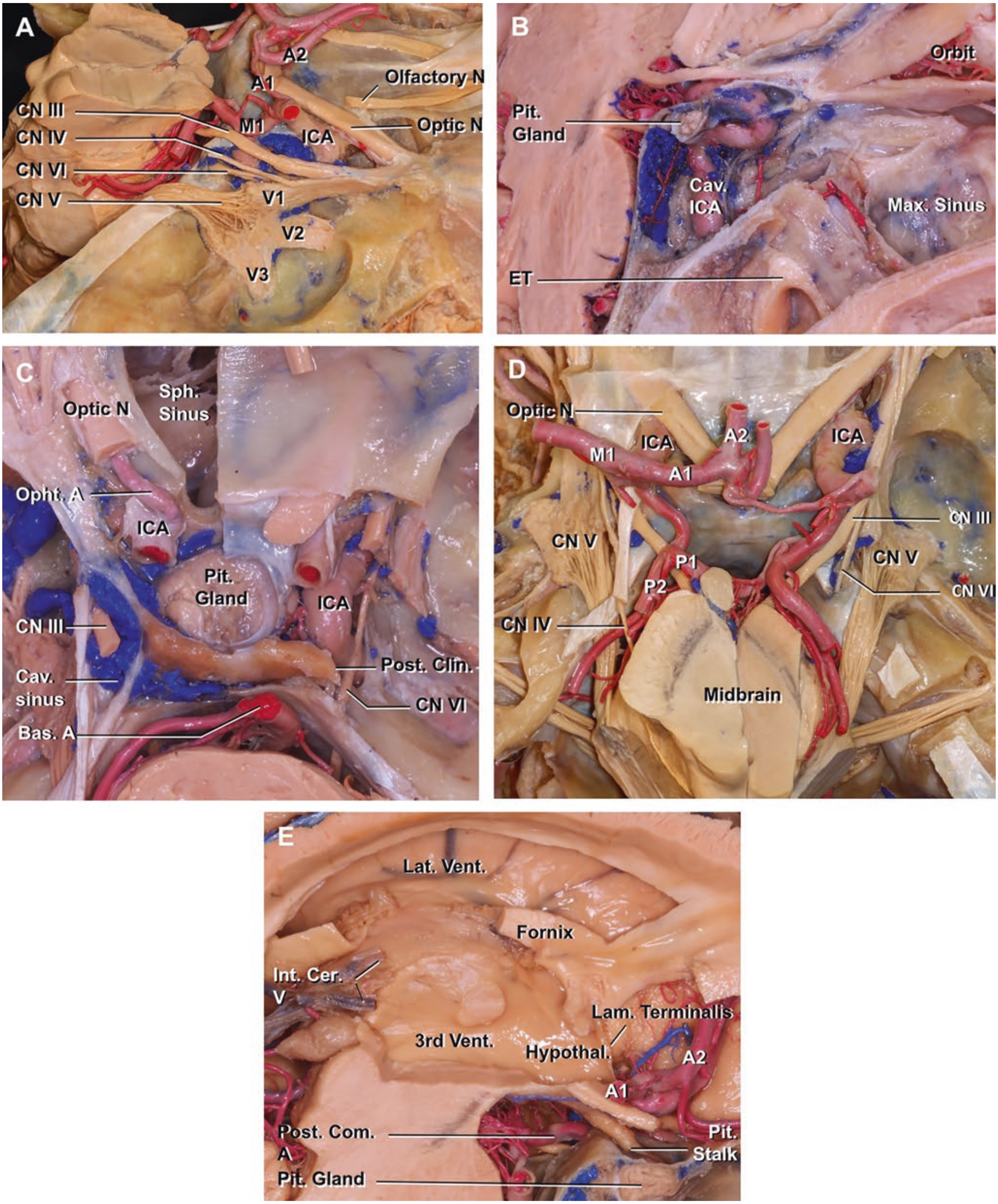

Fig. 2.14 (**a**–**e**) Anatomy of the parasellar and suprasellar areas. (**a**) Lateral view of the right middle fossa and cavernous sinus. The dura mater has been resected and the anterior clinoid process removed. (**b**) Specimen divided in the midsagittal plane, left side, medial view. The bone and the dura mater covering the left cavernous sinus have been removed; the cavernous segment of the carotid artery is exposed. (**c**) Superior view of the sellar and parasellar areas. The dura mater has been removed and the cavernous sinuses opened. The internal carotid artery is exposed on the right side. (**d**) Superior view of the cranial base with Willi's polygon and cranial nerves. (**e**) Specimen divided in the midsagittal plane; image of the ventricles, infundibulum, chiasm, and pituitary gland. *Cav* cavernous, *Cer* cerebral, *CN* cranial nerve *ICA* internal carotid artery, *Hypothal* hypothalamus, *Infund* infundibulum, *Int* internal, *Lam* lamina, *Lat* lateral, *Max* maxillary, *N* nerve, *Opht* ophthalmic, *Pit* pituitary, *Post* posterior, *Sph* sphenoid, *V* vein, *Vent* ventricle

sinus. The abducens nerve is located completely inside the cavernous sinus, whereas the others lie embedded within the periosteal layer of the lateral wall of the cavernous sinus on their way to the superior orbital fissure [30, 31] (Figs. 2.11a–f and 2.14a–e). The nerves are lateral to the internal carotid artery in the cavernous sinus.

The internal carotid artery exits the carotid canal above the foramen lacerum and lateral to the dorsum sellae where it passes under the petrolingual ligament and enters the cavernous sinus. It then turns abruptly forward to course along the carotid sulcus and lateral part of the body of the sphenoid. It forms a vertical posterior bend, passes forward in a horizontal direction for about 2 cm, and forms a horizontal anterior bend. It terminates by passing upward along the medial side of the anterior clinoid process and posterior surface of the optic strut where it penetrates the roof of the cavernous sinus. The clinoid segment of the carotid artery is tightly surrounded by the anterior clinoid process laterally, the optic strut anteriorly, and the carotid sulcus medially. The dura lining the surface of these osseous structures facing the clinoid segment forms the carotid collar around the clinoid segment. In 19 of 20 paraclinoid areas, a carotid cave has been found as an intradural pouch that extends below the level of the distal dural ring between the wall of the internal carotid artery and the dural collar surrounding the internal carotid artery [30]. The superior hypophyseal artery frequently arises in the carotid cave [32].

When viewed from laterally, the cavernous and intracranial portions of the carotid artery have several curves that form an S shape. Together, these portions are called the carotid siphon. The intracavernous internal carotid artery gives rise to branches that supply the walls and enclosed structures of the sella, cavernous sinus, and tentorium. The meningohypophyseal trunk and the inferolateral trunk are the most consistent branches of the intracavernous internal carotid artery.

2.3 Suprasellar Anatomy

The optic nerves emerge from the optic canals medial to the attachment of the free edges of the anterior clinoid processes and are directed posteriorly, superiorly, and medially toward the optic chiasm (Fig. 2.14a–e). The anterior cerebral and anterior communicating arteries, the lamina terminalis, and the third ventricle are located above the chiasm. The tuber cinereum and the infundibulum are posterior to, the internal carotid arteries are lateral to, and the diaphragm sellae and pituitary gland are below the optic chiasm. The suprachiasmatic recess of the third ventricle is located between the chiasm and lamina terminalis. The infundibular recess extends into the base of the pituitary stalk behind the optic chiasm. The normal chiasm overlies the diaphragma sellae and the pituitary gland (70%), the prefixed chiasm overlies the tuberculum (15%), and the postfixed chiasm overlies the dorsum (15%). The carotid artery and the optic nerve are medial to the anterior clinoid process. The artery exits the cavernous sinus beneath and slightly lateral to the optic nerve. The optic nerve pursues a posteromedial course toward the chiasm, and the carotid artery pursues a posterolateral course toward its bifurcation below the anterior perforated substance into the anterior and middle cerebral arteries [2]. The supraclinoid portion of the internal carotid artery is divided into three segments based on the site of origin of the ophthalmic, posterior communicating, and anterior choroidal arteries. The internal carotid, anterior choroidal, anterior and posterior cerebral, and anterior and posterior communicating arteries give rise to perforating branches that reach the walls of the third ventricle and anterior incisural space. All the arterial components of the circle of Willis and the adjacent segments of the carotid and basilar arteries and their perforating branches run in the suprasellar space.

Table 2.1 Anatomical variations of importance in endoscopic endonasal approaches

Anatomical structure	Approach	Variation	Anatomical structure at risk
Anterior ethmoidal canal	Transethmoidal procedures	Canal courses freely into the ethmoidal cells (43%) [14]	Anterior ethmoidal artery, possible intraorbital hematoma/blindness
Height of the lateral lamella	Transcribriform Transethmoidal procedures	Keros type I (8%), II (80%), III (12%) [10, 12]	Base of the frontal lobes, anterior skull; greater risk when type III
Concha bullosa	All endonasal approaches	Present (35%) or absent [12]	Increases difficulty of the approach (narrows nasal corridor)
Onodi cell	Transethmoidal Transsphenoidal	Present (65%) [17]or absent	Optic nerve, internal carotid artery
Pneumatization sphenoidal sinus	Transsphenoidal	Conchal (1–5%), presellar (23–24%), sellar (67–76%) [2]	The internal carotid artery, optic nerve, pituitary gland. Conchal and presellar higher risk, prominences in sphenoidal sinus completely or partially absent
Parasellar carotid distance	Transsphenoidal	Closest at tuberculum (72%), at the floor of the sella (20%), at the clivus (8%) [22]	The internal carotid artery, shorter distance, higher risk

Gentle and accurate dissection is needed to preserve them when operating in these areas.

2.4 Conclusion

Endoscopic skull base surgery requires extensive knowledge of the ventral skull base anatomy as related to endonasal anatomy. Knowledge of the anatomical variations and their preoperative recognition (Table 2.1) is essential to minimize complications.

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Nasal Flaps and Reconstruction

3

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3.1 Introduction

Endoscopic skull base surgery has become a rapidly changing and innovative area of otolaryngology and neurosurgery. Over recent years, approaches and reconstructive options have dramatically diversified. A broadened anatomic understanding has allowed expansion of endoscopic surgery to include approaches to the median and paramedian skull base, orbit, and upper cervical spine. Techniques are now widely used to treat both extradural and intradural pathology. With these more extensive approaches, reconstructive techniques have concurrently evolved with wide representation in the literature. Previously used cellular and acellular grafts have now been replaced or used in conjunction with a multitude of vascularized reconstruction techniques to continue to improve surgical outcomes. Initially, the bulk of skull base defects were repaired using cellular or acellular free grafts. A meta-analysis of CSF leak repair resulting from trauma or endoscopic sinus surgery using such grafts was studied by Hegazy et al. in 2000 [1]. In this study, 289 patients with CSF leaks were assessed. Patients were repaired with multiple different techniques with 90% success, supporting the success of multiple non-vascularized techniques. Reconstruction techniques then evolved to encompass vascularized flaps for reconstruction. In a 2014 study by Thorp et al., 152 patients were identified that had undergone vascularized skull base reconstructions. This study assessed multiple reconstructive techniques including nasoseptal flap, pericranial flap, facial artery buccinator flap, and inferior turbinate flap reconstruction. Overall, the CSF leak rate in this study was found to be 3.3%, supporting the robust nature of these reconstructions [2]. This chapter will discuss all of these techniques and their role in this rapidly expanding field.

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3.2 Rationale

Skull base surgery has dramatically evolved over recent years. Since its inception, the goals of the surgical team have included not only tumor resection but also watertight skull base reconstruction to separate the nasal cavity from the intradural space. These efforts are directly aimed at preventing postoperative cerebrospinal fluid (CSF) leaks, which have been associated with severe consequences including meningitis, pneumocephalus, and even death.

3.3 Patient Selection

The continued advancement of the field of endoscopic skull base surgery has resulted in diversified patients and pathologies amenable to endoscopic techniques. Expanded approaches necessitate careful preoperative planning for each case. Tumor characteristics are extensively assessed, including the type of tumor and its location in regard to surrounding structures. With these factors in mind, an expected surgical defect is then estimated, as is careful planning of possible reconstructive options. In addition, patient-specific factors affecting wound healing must be identified preoperatively and managed with care. Such factors include underlying medical comorbidities, obesity, prior radiation, or smoking. Select patients with increased risks of CSF leak including those with elevated intracranial pressures or morbid obesity should be identified and optimized prior to surgical resection. While management of patients being treated with endonasal skull base surgery has generally become standardized, exceptions may be made on a case-by-case basis.

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Patients identified as having increased risk for CSF leak may require lumbar drain diversion preoperatively at the discretion of the skull base surgery team. Stoken et al. demonstrated that early in the evolution of skull base surgery over the last decade, lumbar drains were frequently used preoperatively for CSF diversion [3]. Previously discussed in the neurosurgery literature stemming from open cranial cases, the drain was thought to decrease postoperative inflammation and swelling. Once thought to be vital to preserve skull base reconstructions, lumbar drains are not without potential complications. As described by Governale et al., the literature reports a 3% risk of major complications associated with lumbar drains, while the minor complication rate increases to 5% [4]. Recent studies in the literature have shown that lumbar drains in the preoperative period are not required in endoscopic skull base reconstructions. Garcia-Navarro et al. reviewed 46 cases undergoing endoscopic skull base surgery, and in this study, 67% of patients had lumbar drains placed. Of these, only two patients had postoperative CSF leaks. The study determined that there was no significant correlation between the use of lumbar drain and CSF leaks [5]. In a separate study by Ransom et al., 65 patients were retrospectively studied and demonstrated a postoperative CSF leak rate of 6.2%. They also found that the lumbar drain complication rate was 12.3%, leading to a recommendation that lumbar drains should be used judiciously in the setting of skull base surgery [6]. There are populations of patients, however, that may significantly benefit from the intraoperative and even postoperative use of lumbar drains. These include patients with chronic intracranial hypertension and/or patients with recurrent leaks requiring revision reconstruction.

The location and size of the tumor also helps identify patients at risk for CSF leak. Zanation et al. also identified that patients with anterior skull base defects were more likely to leak than clival defects [7]. Furthermore, Patel et al. demonstrated that there was a significant correlation between intraoperative high-flow CSF leaks and postoperative leaks [8]. While there is no consensus on the use of lumbar drains in the perioperative period for endoscopic skull base surgery, the surgical team should use their discretion in the setting of each individual patient's factors to determine when the benefits of CSF diversion outweigh the risks.

3.4 Skull Base Pathology

See Table 3.1.

3.5 Surgical Technique

Careful presurgical planning and preparation is vital to the success of these operations. Patients and their tumors must be evaluated on a case-by-case basis. Underlying health problems should be addressed and optimized prior to surgery. Previous radiation therapy, prior or current smoking, as well as underlying medical comorbidities can significantly affect surgical outcomes and must be identified before surgery. With that in mind, standardization of the process with few modifications allows for consistent and successful surgical outcomes.

Endoscopic skull base surgery begins with a smooth induction of general anesthesia. Due to possible tumor impingement on vascular structures, it is vital to avoid extremes of hypotension or hypertension. Once an adequate plane of anesthesia is established, attention is turned to surgical preparation and meticulous positioning. Skull base procedures involve the use of image guidance; therefore, stereotactic systems with fine cut CT and MRI data are loaded and checked to be functioning. In select patients with risk factors,

Table 3.1 Skull base lesions/conditions

Malignant	Benign	Sellar	Fibro-osseous	Other
Esthesioneuroblastoma	Schwannoma	Pituitary adenoma	Ossifying fibroma	Cerebrospinal fluid leak
Adenocarcinoma	Chondroma	Meningioma	Fibrous dysplasia	Encephalocele
Salivary gland carcinoma (adenoid cystic)	Meningioma	Craniopharyngioma	Osteoma	
Squamous cell carcinoma	Nasopharyngeal angiofibroma	Rathke's cleft cyst		
Sinonasal undifferentiated carcinoma	Hemangiopericytoma	Paraganglioma		
Rhabdomyosarcoma	Hemangioma			
Chondrosarcoma	Petrous apex lesions			
Melanoma	Epidermoid			
Lymphoma				
Metastasis				
Osteosarcoma				

a lumbar drain would be placed at this time. Once confirmation of adequate lumbar drain function is confirmed, the surgical bed is turned 90° away from the care of anesthesia. Depending on surgeon preference, the bed position may be adjusted into a modified beach chair position with the head up or remain flat. Different degrees of reverse Trendelenburg and bed tilt may be used to maximize surgeon comfort. During patient positioning, careful attention is placed on padding pressure points such as elbows and heels, as long surgical procedures can lead to peripheral neuropathies and sores. In order to ensure adequate exposure for reconstruction, additional surgical sites (scalp, abdomen, and/or thigh) are prepped and draped in the standard sterile fashion prior to commencement of the surgical procedure. Immobilization with pins is not commonly used for endoscopic endonasal surgeries unless a concurrent transcranial approach is also expected or if required by the navigation system utilized. Once all positioning and surgical preparation is complete, the image guidance system is brought onto the field, and patient registration is completed in the standard fashion.

Bolstering Technique

Reconstruction of the skull base using a combination of cellular and acellular grafts and vascularized flap techniques requires multilayer closure and proper bolstering to ensure successful outcomes. The repair is most vulnerable during the initial phases of healing, immediately following repair. A standardized approach to bolstering helps produce reliable and predictable results. First, Surgicel (Ethicon US, LLC) is typically used to keep any cellular graft or flap in place. It is placed around the margins of the reconstruction, once the reconstructive tissues are in place. Following placement of Surgicel, the most vital areas of the repair and those most prone to leakage are bolstered using firm NasoPore® (Polyganics, Groningen, the Netherlands). This is typically cut into smaller pieces in order to improve manipulation. Once complete, a biologic glue such as DuraSeal® (Confluent Surgical Inc., Waltham, MA, USA) is placed over the entire repair. NasoPore is then used in multiple layers over the reconstruction to further bolster the repair. The goal of this packing is to prevent any movement at the repair site for optimal healing to occur. This must also remain in place when non-dissolvable packs or the Foley balloon are removed after surgery. Once NasoPore is in adequate position, a 14-French Coude Foley catheter is placed under direct visualization to further bolster the repair. Occasionally, expandable tampon-like sponges may be used instead of a Foley catheter. Such sponges are used when the vector of support lies in the vertical plane when the patient is in standing position. The sponges are placed bilaterally under direct visualization in order to further bolster the underlying repair. Dependent upon patient-specific factors, the Foley catheter or nonabsorbable packs are removed 3–7 days after surgery. Patients require antibiotic therapy while nonabsorbable packing is in place. Nasal irrigations are typically started 1 week postoperatively.

Grafts

Acellular Grafts

Acellular grafts play a large role in skull base reconstruction. These materials may be used in conjunction with a multitude of other techniques, making them vital to the success of endoscopic surgery. As noted by Kim et al., the key for all skull base reconstruction should be a multilayer approach for closure [9]. Should the reconstructive surgeon prefer to use an inlay technique, acellular dermal matrix (AlloDerm®, LifeCell, Branchburg, NJ, USA) is effective, placed in the subdural or epidural plane. The use of onlay technique requires that all surrounding mucosa be removed to prevent delayed mucocele formation. The graft requires adequate saline hydration prior to its use. Should the resection require removal of dura, acellular dermal matrix may be used in conjunction with a collagen matrix (Duragen®, Integra Life Sciences, Plainsboro, NJ, USA). This material is used as an inlay graft either between brain and dura in the subdural plane or between dura and the bony skull in the epidural plane. The placement of this graft in this plane many times eliminates CSF leakage resulting from tumor resection. It must be placed beyond the dural margin with adequate (5–10 mm) margins in order to be effective in CSF leak repair. In certain cases involving the sphenoid or clivus, bony ledges may be limited, thus only supporting an onlay graft. With all acellular techniques, the material must be bolstered into place using a combination of packing as previously detailed.

Cellular Grafts

Cellular grafts encompass a vast array of techniques that may be used in combination or independent of the acellular techniques described above. As noted with acellular grafts, emphasis is placed on a multilayer technique with elimination of CSF leak intraoperatively. In a study by Harvey et al., smaller defects (<1 cm) repaired with multilayer closure using free tissue was found to have a success rate of greater than 90% [10]. Several options for free mucosal grafts are available and are described below.

Free Mucosal Graft

The use of a free mucosal graft in skull base reconstruction is a widely applied technique. Tissue is readily available throughout the nasal cavity for harvest and use, eliminating the need for a second surgical site. Mucosal grafts may be taken from the nasal floor, middle turbinate, or septum for use in skull base reconstruction. Many surgeons prefer to preserve the middle turbinate to optimize postoperative sinonasal function and to reserve the nasal septum mucosa for more complex reconstructions, thus making nasal floor mucosa the most readily available cellular reconstructive option.

Using a needle tip bovie that has been bent 45°, the mucosa along the nasal floor is incised. The incision may be carried onto the inferior nasal septum and extended under the inferior turbinate in order to maximize size. Attention must be noted to the location of the soft palate to avoid an incision in this area. The graft incisions are carried anteriorly along the nasal floor until the desired size is achieved. Careful elevation of the graft is performed with a Cottle elevator, ensuring the mucosal side remains identified throughout the dissection. After removal of the graft, the mucosal surface can be inked with a surgical marker to allow for proper placement during repair. Once completely removed from its donor site, it may be used for reconstruction. As emphasized with acellular techniques, removal of all underlying mucosa at the site of reconstruction must be performed in order to avoid the formation of a mucocele. Likewise, it should be ensured that the periosteal aspect of the graft faces the dural and osseous defect. The free graft may be used with previously discussed acellular techniques and bolstered in a similar fashion. Meticulous hemostasis along the donor site should be achieved prior to the conclusion of surgery. If the middle turbinate is removed as part of the approach for skull base reconstruction, the mucosa lining of this structure may be used as a mucosal graft. Careful removal of the underlying bone must be performed prior to its use. Both techniques serve as an excellent option for smaller defects.

Abdominal Fat

The use of abdominal fat in skull base reconstructions is also widely used. It is typically placed to help obliterate space prior to a multilayer reconstruction, in order to create a less irregular defect for reconstruction. To harvest, the incision is made at a second surgical site in either the periumbilical region, lower abdominal area, or lateral hip. This area is prepped and draped separately at the beginning of the procedure in order to maintain sterility. The fat is then harvested through circumferential dissection. Once an adequate volume has been collected, the wound is irrigated, hemostasis obtained, and it is closed in a multilayer fashion. If the donor defect is large due to the need for significant fat harvest, a suction drain may be used to prevent fluid collection or hematoma. The fat is then typically placed intradurally prior to additional dural and skull base resection. Recently, the use of dermal fat grafts has gained popularity. Contrary to normal fat grafts, typically an elliptical incision is created, but the fat is not immediately harvested. The epidermis is removed leaving dermis attached to fat, and the two are harvested together as a composite graft. A larger volume of fat may be dissected circumferentially around the piece of dermis; however, this must be kept in continuity. Once an adequate volume is collected, the two are removed together and used in the skull base reconstruction. Careful sizing of the graft must be done, in order for the dermal component to rest at the level of the skull base defect, thus allowing a more laminar reconstruction. The use of dermis allows for a more robust bolster for reconstruction and facilitates manipulation or the graft during placement. Both free fat and dermal fat grafts are frequently used with a combination of other techniques, both acellular and/or vascularized reconstructions. They are bolstered with a combination of absorbable packing, biologic glue, and expandable sponges/Foley catheter as previously described. It is important to note that while cellular and acellular techniques provide a robust reconstruction for small skull base defects, Hadad et al. found that in resections greater than 3 cm, multilayered free tissue grafts resulted in unacceptably high rates of postoperative CSF leaks at 20–30%, and consideration of additional techniques, namely, vascularized reconstruction, was recommended [11].

Flaps

Vascularized flap reconstruction has become the mainstay in endoscopic skull base surgery with the ever-expanding complexity of cases and pathology amenable to this approach. Initially described by Hadad et al. in 2006, the nasoseptal flap, comprised of mucoperiosteum and mucoperichondrium from the nasal septum and pedicled on the posterior septal artery, has become the preferred technique for skull base reconstruction [11]. It is characterized by a long robust pedicle that makes it ideal for use in a multitude of defects and locations. The harvest of the nasoseptal flap can be extended onto the nasal floor, allowing it to expand from orbit to orbit and from sella to frontal sinus [7]. In addition to its robust pedicle and expansive reach, the endoscopic harvest of the nasoseptal flap prevents the need for a second surgical site and the potential associated morbidity. The primary disadvantage of the nasoseptal flap is that its use must be expected preoperatively. Because of the location of the pedicle, its harvest must be performed prior to sphenoidotomy and posterior septectomy in order to ensure its viability. The “rescue” technique will be further described below and has largely offset this disadvantage. In certain settings, the nasoseptal flap may be unavailable for use due to tumor involvement or vascular compromise due to prior surgery. Prior septoplasty is not a complete contraindication to nasoseptal flap use, although the flap must be elevated with great care to preserve its integrity without tears. If it is unclear as to whether the vascular pedicle to the nasal septum is viable, a Doppler probe can be utilized to confirm its presence.

Technique for the Nasoseptal Flap

The patient is positioned in the standard fashion as previously described. To improve visualization, the inferior turbinates may be outfractured bilaterally, and the middle turbinate on the side of the nasoseptal flap (NSF) harvest is removed at the discretion of the surgeon. Meticulous hemostasis is performed after middle turbinate removal to have clear visualization; however, caution is used posteriorly in order to preserve the pedicle from nondirected cautery. The superior turbinate is then gently lateralized until the natural os of the sphenoid sinus is identified. The flap is harvested based on a prediction of the surgical resection made prior to surgery. When in question, an overestimation of size is always preferred. Using needle tip extended length monopolar cautery that is bent 45°, two parallel incisions are made. The inferior incision is made across the posterior choana margin and then onto the nasal septum at its junction with the nasal floor. The superior incision begins at the natural os of the sphenoid sinus and then extends superiorly. In order to preserve the olfactory epithelium, the incision is placed 1–2 cm from the superior portion of the septum. A vertical incision is then placed anteriorly connecting the previously performed inferior and superior limbs and can extend as far as the mucocutaneous junction. A Cottle elevator is then used to begin the elevation. Careful separation of all incisions anteriorly should be performed before proceeding with more posterior elevation, as incomplete elevation at the incision lines can lead to tearing, reducing the functionality of the flap. Once the flap is elevated, it is then placed into the nasopharynx or ipsilateral maxillary sinus until the extirpative portion of the procedure is complete (Fig. 3.1a–d).

Once attention is turned to the reconstruction, the flap is lifted out and rotated into place along the skull base ensuring the pedicle remains untwisted. It is important to ensure that the perichondrial aspect of the flap is in contact with the cranial base rather than its mucosal aspect to allow the graft to adhere and prevent delayed mucocele formation. As these can sometimes be difficult to distinguish at the end of the procedure, it is sometimes helpful to mark the mucosal aspect of the flap with a surgical marker immediately after harvesting. After placement over the cranial base defect, the flap is then used as part of a multilayer reconstruction of the skull base defect in combination with other cellular and acellular techniques. Careful attention to placing the flap in direct contact with the bony margins of the defect is critical. In addition, as described with free mucosal grafts, all underlying mucosa must be removed to prevent mucocele formation. The flap is then bolstered into place as previously described. As discussed before, patient characteristics including underlying medical problems, previous radiation, presence of high-flow CSF leak, or revision procedure affect the duration of the use of nonabsorbable packs and/or Foley catheters postoperatively. Reconstruction bolsters including nonabsorbable packs and/or Foley catheters may remain in place anywhere from 3 to 7 days.

Nasoseptal “Rescue” Flap Technique

Extirpative procedures that require large skull base defects allow for the preoperative expectation that a nasoseptal flap or other pedicled flap will be required for reconstruction. However, in some cases, the need for the nasoseptal flap may not be known at the beginning of the case. In this specific patient population, a nasoseptal “rescue” flap may be elevated at the beginning of the case. This procedure allows for the preservation of the vascular pedicle but does not eliminate the opportunity to leave the flap in its native position should its use not be necessary. Rivera-Serrano et al. describe a technique in which partial harvest is done at the beginning of the case. In this technique, the superior incision is performed as with the nasoseptal flap. It begins at the sphenoid os and extends approximately 2 cm anteriorly along the same incision a traditional NSF would follow [12]. Rawal et al. further describe the technique in which a Cottle elevator is then used to expose the sphenoid rostrum by reflecting the flap inferiorly. By displacing this area prior to sphenoidotomy and septectomy, the vascular pedicle of the flap is protected for further reconstruction [13]. If a nasoseptal flap is required for reconstruction, the remainder of the NSF incisions are then placed, and the flap is harvested in the same fashion. The flap is then placed as part of a multilayer closure as previously described. If a nasoseptal flap is not required, the inferior reflected “rescue” flap is then returned to its native position (Fig. 3.2a–c).

Endoscopic-Assisted Pericranial Flap

While the nasoseptal flap remains the workhorse of skull base reconstructions, the pericranial flap (PCF) provides an option for reconstruction when this is either not available or inadequate. As described by Zanation et al., the PCF is a very robust flap pedicled on the supraorbital and supratrochlear arteries [14]. Because of its large size, it allows for reconstruction of the entire skull base especially in defects following more extensive and difficult resections. The reconstructive surgeon may choose to harvest the flap through an array of techniques including an endoscopic-assisted, hemicoronal, or coronal approach with or without a small glabellar incision for intranasal introduction. The noted glabellar incision allows for a bony window through the nasion to be utilized for nasal introduction of the flap [14]. The PCF also provides an important extranasal option for reconstruction, a valuable alternative for previously radiated patients requiring extensive

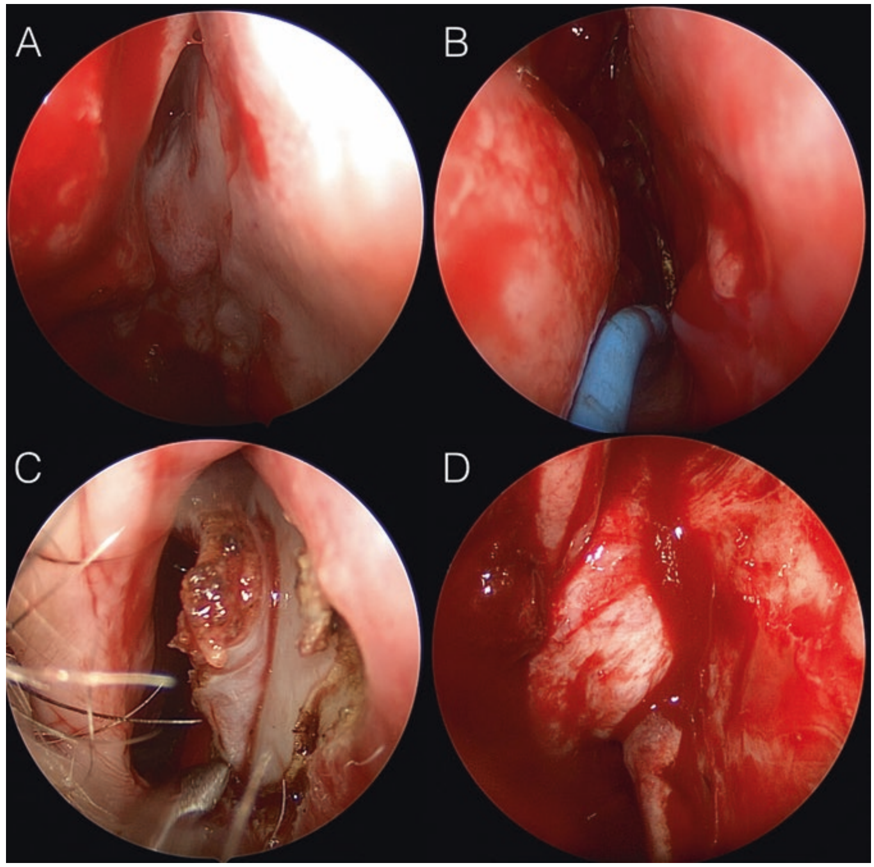

Fig. 3.1 (**a**–**d**) Nasoseptal flap. (**a**) Prior to incision, the superior turbinate is lateralized to visualize both the choana and the natural ostium of the sphenoid sinus. These landmarks are confirmed prior to incision. (**b**) Inferior incision made with the needle tip bovie. This incision can be modified to widen the flap. (**c**) Careful elevation with a cottle in the mucoperichondrial plane protects the integrity of the flap. (**d**) Nasoseptal flap elevated off the septum and tucked into the nasopharynx

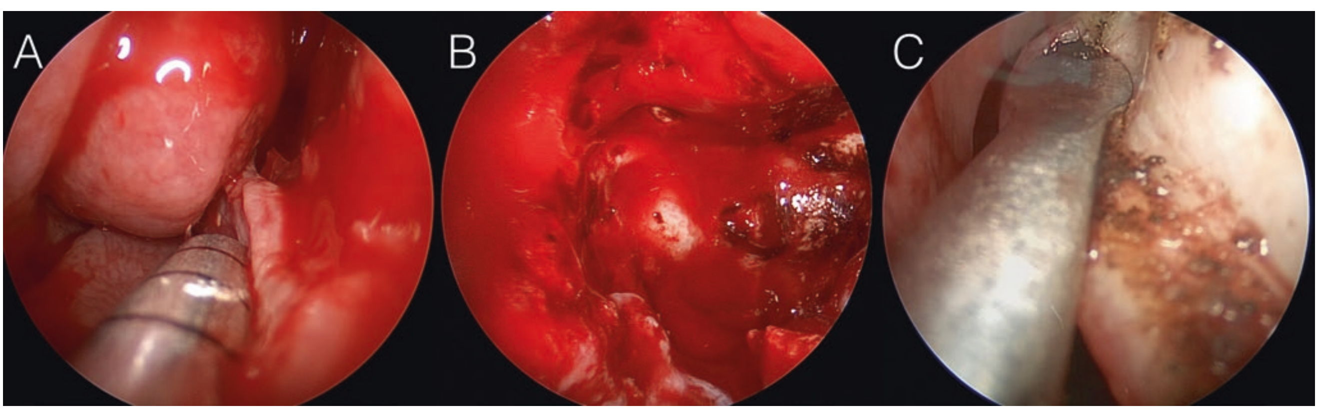

Fig. 3.2 (**a**–**c**) Rescue flap. (**a**) Middle turbinate is removed, and superior turbinate is lateralized prior to incisions. Confirm the location of the natural ostium of the sphenoid sinus. Superior longitudinal incision beginning at the sphenoid ostium is extending anteriorly along the expected nasoseptal flap trajectory without full extension anteriorly. (**b**) Rescue flap use still allows for wide exposure and access for tumor dissection. (**c**) After placement of the incision, release is performed in the mucoperichondrial plane

resection or those for whom the nasal septum is unavailable due to tumor involvement or prior surgery.

In a radioanatomic study by Patel et al., imaging studies for ten patients were assessed preoperatively to determine ideal PCF incisions. From this study, the average length from the nasion to the sphenoid sinus was 4.51 cm, nasion to posterior wall of the sella was 7.57 cm, and nasion to the inferior clivus was 12.10 cm. The average external pedicle length was measured to be 4.36 cm, a combination of distances measured from lateral supraorbital notch to the mid-forehead plus the mid-forehead to the nasion. These values were used to obtain average PCF lengths needed for reconstruction of defects in these areas. The following measurements were obtained with a 3 cm correction factor accounting for flap transposition through the nasionectomy: 11.31–12.44 cm for anterior fossa defects, 14.31–15.57 cm for sellar defects, and 18.3–20.42 for clival defects. All patients in this study had no evidence of postoperative CSF leak [15].

Endoscopic-Assisted Pericranial Flap Technique

The patient is prepped and draped in the same fashion as previously described for skull base surgery. The hair is stapled away from the planned surgical site but not shaved. The planned scalp incision is then marked, and its location is at the discretion of the reconstructive surgeon. The supraorbital notch is then identified, and 1.5 cm are marked on either side. During planning, the midline should be marked as the contralateral PCF may be used for revision surgery. After the extirpative portion of the procedure is complete, attention is turned to elevation of the PCF. The skin incision is performed, and dissection is then carried out in a subgaleal plane using direct visualization aided by the endoscope. The endoscope allows improved visualization anteriorly and posteriorly to ensure a large flap may be harvested (Fig. 3.3a–c).

Once this dissection is complete, a needle tip extended length cautery is used to make an incision in the pericranium at the most posterior aspect of the field. Bilateral lateral incisions are then placed ensuring the pedicle is preserved. Once this is complete, the pericranial flap is elevated with endoscopic assistance to help prevent tearing in the flap. As noted with the nasoseptal flap, it is important that all incisions are complete prior to elevating to prevent damage to the integrity of the flap. Attention is then turned to the glabella incision. This is dissected down to the periosteum, which is incised using bovie electrocautery at the level of the nasion. A drill is used to transgress the bone of the nasion and enter the nasal cavity. Once this area is opened adequately to prevent pressure on the flap or its pedicle, the flap is introduced through this area into the nasal cavity. Caution must be taken during this translocation to not twist the pedicle, as the vascular supply could be compromised. Once within the nasal cavity, the PCF is moved into position, ensuring direct contact with defect margins to ensure adequate healing (Fig. 3.4a–f). Patency of the frontal sinus outflow can be maintained with steroid-eluting stents (PROPEL Sinus Implant, Intersect ENT, Menlo Park, CA) placed alongside the PCF, but the primary concern should be cranial base repair. Once healed, the PCF can always be surgically dissected/divided in a delayed fashion to reestablish sinus drainage and prevent mucocele formation. After placing the PCF, a multilayer reconstruction and bolstering is used as previously noted. The external incisions are then copiously irrigated with normal saline and closed in a multilayer fashion. The scalp wound typically requires a small suction drain to prevent the formation of hematoma or seroma in the wound bed. The surgical drain is monitored until appropriate for removal prior to hospital discharge.

Temporoparietal Fascia Flap

Traditionally, the temporoparietal fascia flap (TPFF) was used extensively in head and neck cancer reconstructions. As reviewed by Patel et al., this widely versatile flap is based on the more anterior branch of the superficial temporal artery (STA), a terminal branch of the external carotid system [16]. This fan-shaped flap provides a reliable extranasal reconstructive option, when the flaps listed above are not available [7, 8]. Like the PCF, the TPFF provides a valuable extranasal option for patients with previous skull base or sinonasal radiation. Due to the location of its pedicle, it is best suited for reconstruction of the sellar region extending down into the clivus. As noted by Patel et al., this limits its use for anterior cranial fossa defects [8]. Careful patient selection must also be used as history of temporal artery biopsy or scalp radiation can lead to vascular compromise or donor site morbidity. An advantage to this flap is it provides large size and bulk for patients requiring extensive resection. Disadvantages include donor site necrosis, alopecia, scarring, frontal branch of the facial nerve weakness or dysfunction, and cosmetic deformity. The dissection for transposition of the flap also involves the infratemporal fossa, putting the internal maxillary artery at risk.

Temporoparietal Fascia Flap Technique

The patient is positioned and prepped for endoscopic skull base surgery in the standard fashion as previously described. In addition, the patient is also prepped for an ipsilateral scalp hemicoronal incision. The TPFF harvest is not started until completion of tumor resection and wide ipsilateral maxillary antrostomy and complete ethmoidectomy. Following adequate exposure, the sphenopalatine artery (SPA) and posterior septal artery are identified and clipped endoscopically. The SPA is then followed into the pterygopalatine fossa (PPF), and the posterior and lateral walls of the maxillary sinus are removed to expose the infratemporal fossa. Once the internal maxillary artery (IMA) is fully visualized, the descending palatine artery may be identified and dissected. Once these vessels are identified and dissected, the contents of the PPF may be protected and moved laterally until the pterygoid plates are fully visualized. At this location, the vidian nerve is typically sacrificed to allow displacement, but the pterygopalatine ganglion may be preserved. Attention is then turned to the external flap harvest. An ipsilateral hemicoronal incision is made with care to ensure preservation of the STA, which lies in the subcutaneous tissues. The incision and elevation should be in the subfollicular plane. An aggressive incision in this area can lead to compromise of the vascular pedicle. Once exposure of the flap of desired size is complete, an incision is placed through the fascia laterally. The flap is then elevated from the temporalis muscle fascia superiorly and superficial layer of the deep temporal fascia below the level of the temporal line of fusion including elevation of the periosteum from the zygomatic arch.

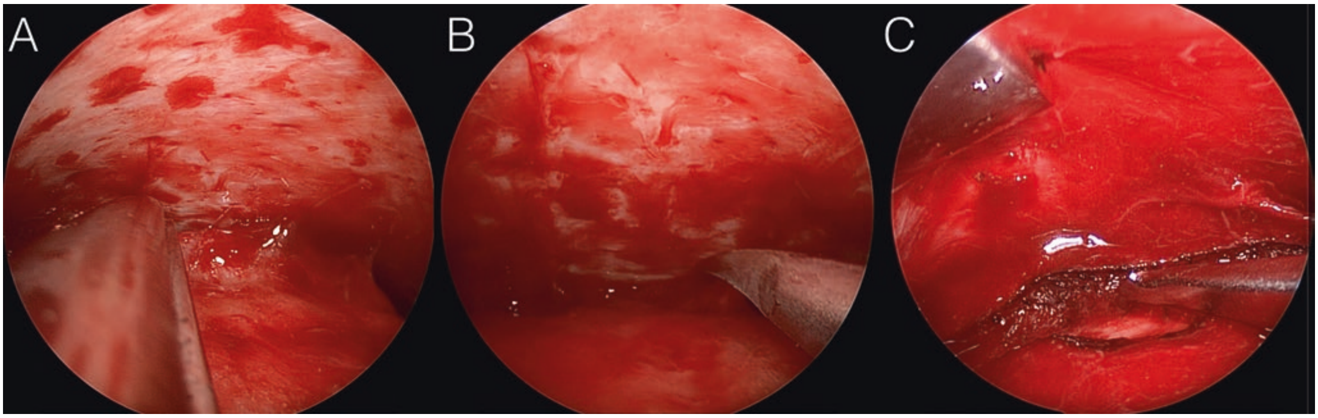

Fig. 3.3 (**a**–**c**) (**a**) After skin incisions, elevation is performed in the subgaleal plane. This is done with sharp instrumentation to ensure preservation of a thick pericranial flap. (**b**) Generous elevation is performed prior to incisions with monopoly cautery. (**c**) After incisions have been placed with preservation of a 3 cm vascular pedicle, progressive elevation of the flap is performed with a periosteal elevator

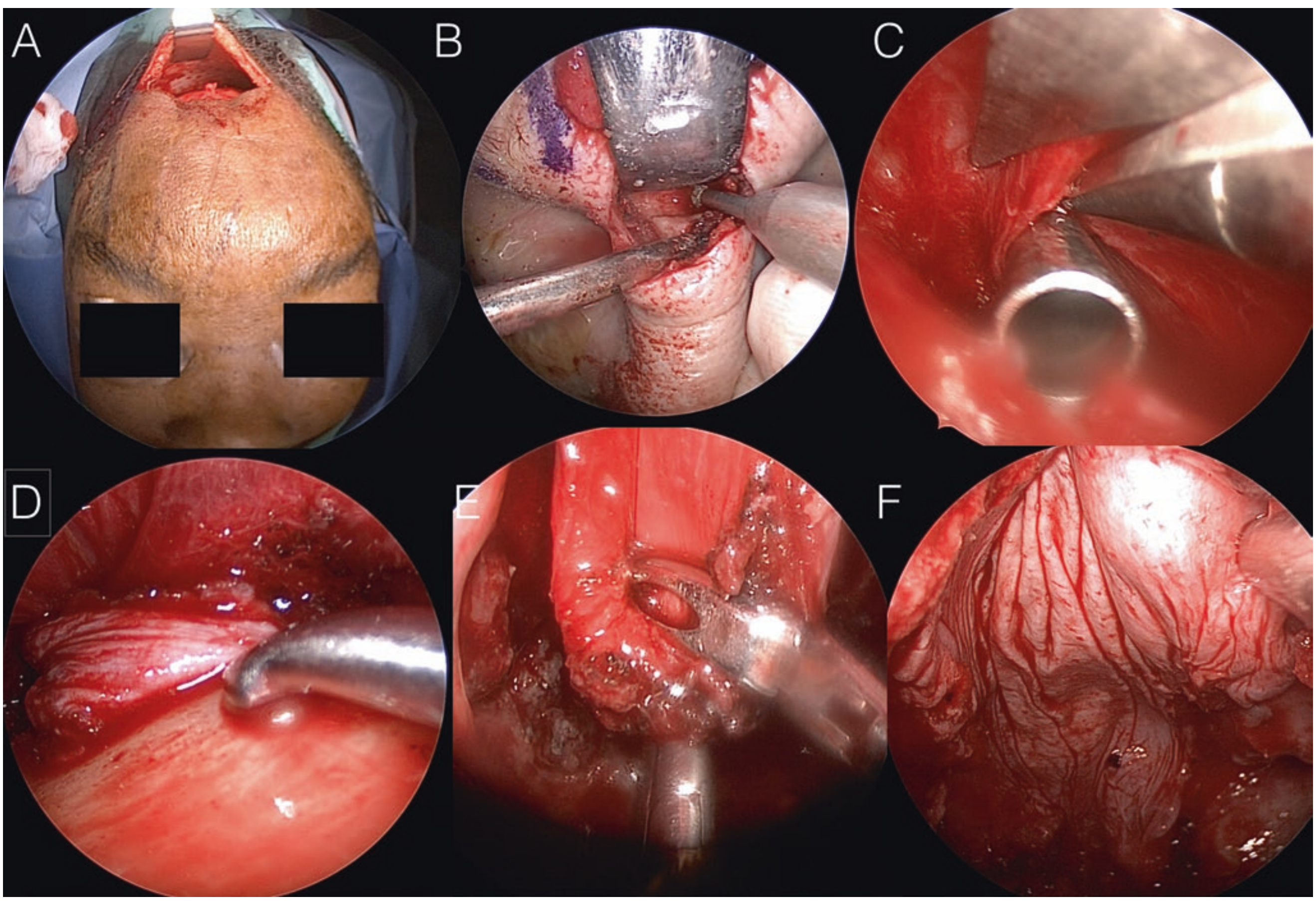

Fig. 3.4 (**a**–**f**) Progressive pericranial flap elevation. (**a**) Trichophytic superficial incision used without need for complete hemicoronal. Elevation performed through this incision in the subgaleal plane. (**b**) Horizontal glabellar incision carried to the level of the nasal bones. The periosteum is dissected, and a nasionectomy is performed to obtain access to the nasal cavity. (**c**) Communication between the subgaleal and subperiosteal planes are confirmed prior to movement of the flap. (**d**) Progressive elevation of the flap down to the nasionectomy is performed to ensure preservation of the pedicle. (**e**) Transposition of the pericranial flap into the nasal cavity performed under direct visualization. (**f**) Pericranial flap used for anterior skull base defect prior to packing

A wide tunnel is then formed in this area down to the infratemporal fossa. This can typically be done with commercially available percutaneous tracheostomy dilators. A lateral canthotomy incision can occasionally help at this point to help expose the pterygomaxillary fissure to help in full transposition of the flap. Once the tract is complete, a guide wire is placed through the largest tracheal dilator. The dilator is then removed, and the flap is attached to the guidewire and pulled into the nasal cavity. Meticulous attention to over-rotation of the flap in this area is important to minimize the potential for vascular compromise to the flap. Once in the nose, the TPFF is placed overlying the defect. This can be used in conjunction with other reconstruction techniques and is bolstered into the place in the standard fashion. External incisions are again copiously irrigated and closed in a multilayer fashion with a surgical drain in place.

Turbinate Flaps

Less commonly used intranasal options for patients in which the NSF is not available for skull base reconstruction exist. Zanation et al. discussed two flaps, the inferior turbinate flap (ITF) and the middle turbinate flap (MTF), each pedicled on their respective arteries and branches of the posterior lateral nasal artery, a terminal branch of the sphenopalatine [7]. The ITF has a shorter length and arc of rotation when compared to the NSF but provides an option for smaller skull base defects. Zanation et al. discussed its use for the sellar, parasellar, and midclival areas [7]. This flap can be combined with a contralateral ITF for better skull base coverage. To harvest, incisions should include the entire medial surface of the turbinate and can be extended onto the lateral wall mucoperiosteum for wider coverage. With a posterior pedicle, care must be taken to not avulse this area during harvest or transposition. After use, nasal splints are used to prevent scarring within the nose. Patel et al. described the elevation of the MTF but noted limited utility given its technically difficult harvest and thin mucosa [8].

3.6 Postoperative Care and Complications

Extensive endoscopic endonasal skull base resections and reconstructions are not completed without risk for complications. Cautious standardized postoperative care following these procedures helps ensure the optimal results. Immediately after surgery, patients have packing in their nose supporting complex multilayer closures. This typically includes nonabsorbable packing or a Foley catheter used to bolster the repair and depends on intraoperative factors. As previously discussed by Patel et al., intraoperative high-flow CSF leak is the most reliable predictor of developing a postoperative leak [8]. Following skull base surgery, all patients keep a urinary Foley catheter overnight to monitor urine output and rule out concerns for diabetes insipidus. Patients with extensive tumor resections, intradural or intra-arachnoidal resections go to the neurosurgical intensive care unit overnight for close observation. In patients with high risk of postoperative leak, bedrest may be implemented for up to 48 h, at which time activity is still significantly limited. An aggressive bowel regimen is also implored to prevent significant straining postoperatively. Patients with no to low-flow intraoperative CSF leaks have nonabsorbable packing or Foley catheter removed prior to discharge on postoperative day 3. Those with high-flow leaks do not have packing removed until postoperative day 5 and sometimes postoperative day 7. When used, lumbar drains are initially kept open and then slowly tapered until appropriate for removal. All patients require antibiotic therapy while packing is in place for prophylaxis to reduce the risk of meningitis. Commencement of nasal saline irrigations is typically determined by intraoperative leak status reconstruction type.

As with all surgeries, complications exist with endoscopic endonasal skull base surgery reconstruction. Kassam et al. describe these as postoperative CSF leak, meningitis, pneumocephalus, and/or graft failure or displacement [17]. To further assess these complications, a meta-analysis by Harvey et al. reviewed 38 studies. Overall the study found that endonasal skull base reconstruction techniques revealed a postoperative CSF leak rate of 11.5% (70/609) [18]. In this study, analysis of patients reconstructed with free grafts revealed a CSF leak rate of 15.6% (51/326), while the vascularized flap rate was 6.7% [18]. In a separate study by Thorp et al., 152 flaps were assessed and only 5 (3.3%) were found to have CSF leaks (3 NSF, 1 PCF, 1 ITF) [2]. In this study, the majority of leaks were in patients with high-flow CSF leaks. The average time to leak in this study was 43.6 days, as CSF leaks typically occur in the immediate postoperative period. This study did not find an association between complications and radiation therapy. Pneumocephalus is typically an immediate postoperative concern that presents with mental status changes, headache, and vomiting. Expeditious clinical and radiologic evaluation is vital for identification and management of this complication. Reconstructive flaps that have tears present the possibility of CSF leakage as well. With the exception of large tears, small areas of damage typically do not restrict the use of the flap due to the underlying multilayer closure. However, if the surgeon has concerns for intraoperative CSF leak due to lack of flap integrity, a separate reconstruction technique should be used at the time of surgery. Even with the meticulous use of bolstering materials intraoperatively, very rarely flaps may shift. Should this occur and result in CSF leakage, expedited surgical revision should be undertaken. If changes to flap position are only noted on postoperative imaging but no clinical concerns are present, no further revision should be performed.

Overall, review of studies, such as Zanation et al., reveal excellent results using a variety of techniques for skull base reconstruction [7, 18]. In a separate study by Zanation et al., 70 skull base reconstructions for high-flow CSF leaks were assessed, and the CSF leak rate was found to be 5.7% [18]. In a further study by Zanation et al., NSF previously used were revised, taken down, and reused, and postoperative CSF leak rates were no higher. There was also no evidence of flap death in this group [19]. With expanding skull base surgery techniques, the NSF remains the primary option for vascularized skull base repair. However, a study by Patel et al. demonstrated that secondary vascularized flaps beyond the NSF (PCF and TPFF) had a success rate of 97%, compared to that of the NSF (95%) [15]. In a separate study, Patel et al. assessed 34 patients in which the NSF was not available for use. Here, the success rate was greater than 95%, demonstrating new techniques provide consistent and robust repairs in the face of ever-increasing complex pathology [16].

3.7 Surgical Pearls

Nasoseptal Flap

- Meticulous surgical planning is vital to adequate NSF harvest. Overestimation is always preferred.
- The superior incision must be placed 1–2 cm below the superior border of the septum to save olfaction.
- When making nasoseptal flap cuts, do not move too quickly as this will leave areas attached, risking flap tearing. Ensure all anterior elevation has occurred along all three incision lines prior to posterior elevation.
- Standardized multilayer closure including cellular, acellular, and flap techniques is vital to consistent outcomes.

Pericranial Flap

- Very robust flap that offers reconstruction of the entire skull base, especially anterior skull base defects.
- Do not taper the pedicle too much as this can lead to vascular compromise.
- Perform extensive dissection circumferentially prior to incising pericranium or the size of the flap can be significantly truncated.

Temporoparietal Facial Flap

- This flap provides robust reconstruction in patients with very limited options for further skull base repair.
- Dissection in the infratemporal fossa is extensive and has a different risk profile than other reconstructive efforts.
- Over-rotation of the pedicle can lead to flap compromise.

Postoperative Care

- Postoperative activity should be very conservatively managed.
- Patients at increased risk for CSF leak should progress through postoperative care with extreme caution.
- Any concerns for early CSF leak should be revised and repaired if possible before significant complications arise.

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4 Functional and Nonfunctional Pituitary Adenomas

Ankush Chandra and Manish K. Aghi

4.1 Introduction

Pituitary adenomas are slow-growing, benign, monoclonal tumors that arise from the cells of the pituitary gland. Due to their low rate of recurrence and rare transformation to higher-grade tumors, pituitary adenomas are classified as WHO (World Health Organization) grade I tumors [1]. While pituitary adenomas were once considered a rare occurrence, recent studies have shown that their prevalence is much higher due to the advent of better diagnostic methods and increased utilization of imaging techniques. Pituitary adenomas account for 15–18% of all intracranial tumors and are the third most common primary brain tumor, following meningiomas and gliomas [2–6]. They are divided into two categories, namely, functional pituitary adenomas (FPAs) and nonfunctional pituitary adenomas (NFPAs).

4.2 Rationale

In general, the rationale for treatment of pituitary adenomas is alleviation of presenting symptoms and prevention of future symptoms. FPAs typically present with hypersecretion of one or more hormones of the pituitary gland detectable in the serum, leading to clinical syndromes whose symptoms are summarized in detail in Table 4.1. NFPAs are endocrine-inactive tumors and can be incidental findings on radiologic exams or can present with symptoms of mass effect (Table 4.1, Fig. 4.1) [23–26]. Due to physiologic effects of FPAs, they typically present at smaller sizes than NFPAs [26]. When treatment is indicated for symptomatic adenomas or incidentally found adenomas large enough to warrant preemptive treatment, surgical resection is the first-line treatment for most pituitary adenomas.

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4.3 Patient Selection

Appropriate patient selection requires a complete diagnostic workup. If a pituitary adenoma is clinically suspected, radiological investigation should be performed. *Magnetic resonance imaging* (MRI) is the **gold standard** for imaging most sellar tumors (Fig. 4.2) [7, 27]. It is important to note that imaging is not only important for confirming a pituitary mass but also to differentially diagnose other sellar and extrasellar masses such as craniopharyngioma, Rathke's cleft cyst, pituitary abscess, epidermoid cyst, chordoma, meningioma, metastatic tumor, aneurysm, lymphocytic hypophysitis, arachnoid cyst, mucocele, lymphoma, or sarcoidosis [7].

In parallel, appropriate laboratory biochemical tests should be done. Routine endocrine evaluation of all anterior pituitary axes to assess for hypopituitarism is recommended because, beyond revealing a significant rate of deficits surpassing the level of clinical suspicion for all pituitary axes, the cutoff values to initiate thyroid and adrenal replacement might be different in a patient with panhypopituitarism versus isolated deficiencies [7, 28]. Such evaluation will also distinguish NFPAs from specific types of FPAs [7, 28]. Different pathologies associated with pituitary adenomas are summarized in Table 4.2.

Prolactinomas

According to the guidelines set by the Pituitary Society and the Endocrine Society, the diagnosis of prolactinoma requires both biochemical and radiographic evidences. A single measurement of serum prolactin is recommended. If serum prolactin levels are above 200 μg/L, a prolactinoma is almost certainly the underlying cause [8, 11, 12, 36]. However, if the serum prolactin levels are present between the upper limit of normal prolactin levels and 200 μg/L in a patient with imaging suggestive of pituitary adenoma, then stalk effect from a nonfunctional adenoma is a possible diagnosis rather than prolactinoma. Since prolactinomas are very responsive to

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Table 4.1 Symptoms associated with different types of pituitary adenomas. Shown are symptoms associated with different types of functional and nonfunctional pituitary adenomas

Type of pituitary adenoma	Associated symptoms
Prolactinoma	*High prolactin*: Amenorrhea (females), hypogonadism, chronic kidney disease, galactorrhea, infertility, osteoporosis, hirsutism [7] *Macroadenoma (mass effect)*: Headaches, visual impairments, and hypopituitarism [7, 8]
NFPA	*Symptoms of mass effect*: Chronic headache, visual impairments, and pituitary insufficiency [9, 10]
Somatotroph adenoma (acromegaly)	*Classical presentation*: Acral and soft tissue overgrowth, bony overgrowth, coarse facial appearance, skin thickening, macrognathia, enlarged hands and feet [11–14] *Uncontrolled acromegaly*: Carpel tunnel syndrome, metabolic dysregulation (diabetes mellitus, dyslipidemia, and insulin resistance), cardiovascular disorders (hypertension, cardiac hypertrophy, cardiac myopathy, and arrhythmias), and increased risk of colon polyps, colon cancer, and other tumors [12–18] *Macroadenomas (mass effect)*: Disordered sleep, hypopituitarism, headaches, and visual disturbances [13, 15, 16, 19, 20]
Corticotroph adenoma (Cushing's disease)	*Characteristic symptoms*: Central obesity, moon face, facial plethora, diabetes mellitus, hirsutism, easy bruising, proximal myopathy, and purple striae [21, 22] *Other symptoms*: Metabolic, neuropsychological, musculoskeletal, cardiovascular, dermatological, hormonal, and immunological abnormalities [21]

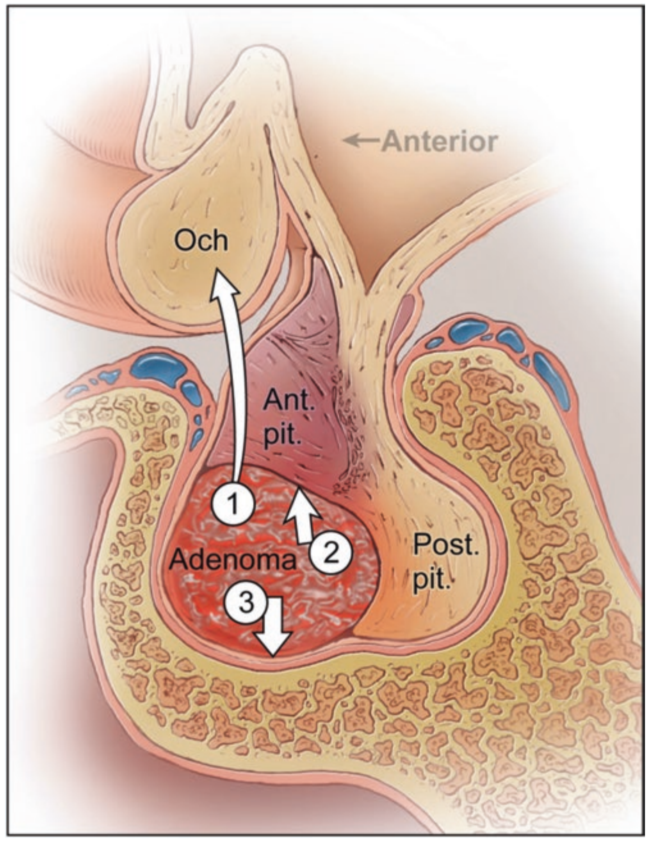

Fig. 4.1 Symptoms of mass effect from pituitary adenomas. These symptoms (headache, visual, or endocrine dysfunction) are classic symptoms of NFPAs and FPAs, with FPAs also having symptoms of hormonal hypersecretion that often predominate over these mass effect symptoms

medical treatment, surgery is reserved for those prolactinomas that have a definitive surgical indication (acute hemorrhage, cystic components, or resistance to or intolerance of dopamine agonists) or for patients who make an informed choice to pursue surgery after seeing both an endocrinologist to discuss medical therapy and a neurosurgeon with extensive experience with pituitary tumor surgery. Figures 4.3b and 4.4a, b show the histology of prolactinomas.

Acromegaly

According to the clinical practice guidelines set by the Endocrine Society, once the physician identifies clinical features of acromegaly, biochemical screening should be performed for confirmed diagnosis [13, 37]. Serum IGF-1, the preferred screening tool over random GH level, is measured and matched to age and sex of the patient [11, 13]. If IGF-1 levels are discordant with normal range, an oral glucose tolerance test (OGTT) with 75 g of glucose is performed: a lack of fall of serum GH levels below 1 ug/L within 2 h would confirm acromegaly [13, 15, 38]. First-line therapy for acromegaly is transsphenoidal surgery (details of which are described below) [13, 37]. In cases where there is residual tumor following surgery, repeat surgery should be performed to achieve full resection if the residual mass is surgically accessible [13]. If residual tumor is not surgically accessible, radiation therapy or radiosurgery, or medical therapy with somatostatin analogues or the GH receptor antagonist, pegvisomant, should be implemented. Figures 4.3a and 4.5a, b show the histology of these tumors.

Cushing's Disease

The Endocrine Society recommends exclusion of exogenous glucocorticoid use prior to performing any laboratory test to evaluate for suspected Cushing's disease [39, 40]. For initial diagnosis, one of the four tests should be used for patients with low index of suspicion, and two tests should be done for patients with high index of suspicion: (1) urine free cortisol

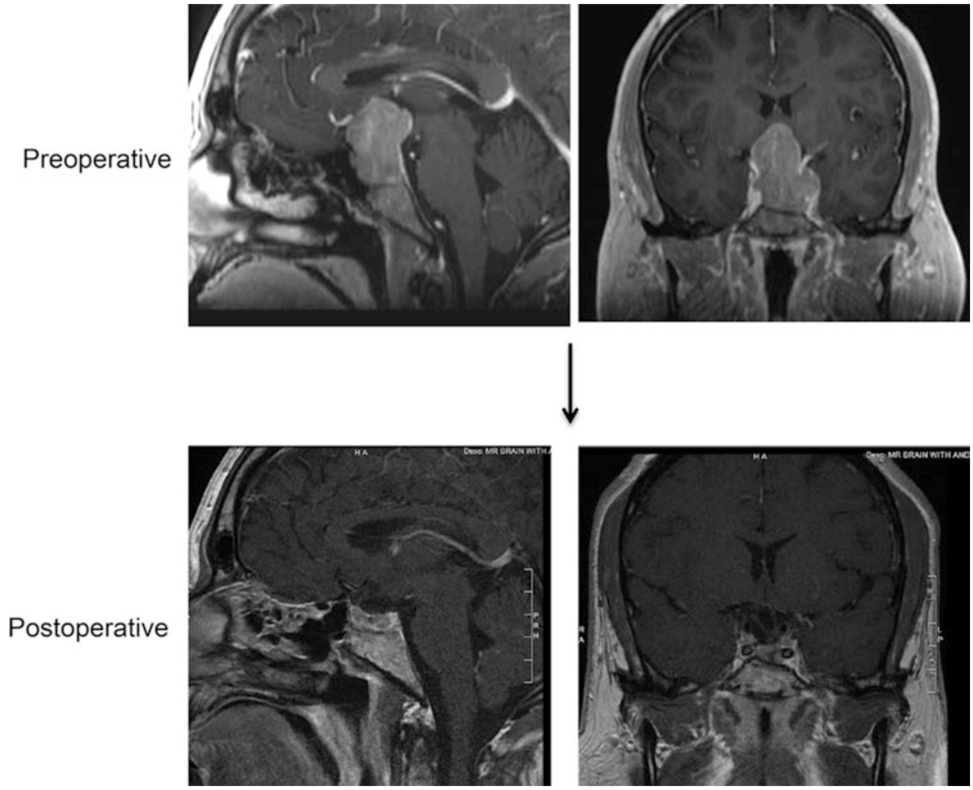

Fig. 4.2 MRI images of pituitary adenoma. Shown are sagittal (*left*) and coronal (*right*) T1 post-gadolinium images of a newly diagnosed giant (larger than 4 cm in maximal diameter) nonfunctional pituitary adenoma with the *upper images* representing preoperative images and *lower images* representing postoperative images after endoscopic endonasal resection with no residual tumor seen on postoperative imaging

Table 4.2 Summary of different pathologic types of pituitary adenomas and their prevalence. Shown are the prevalence, common age groups, and gender predilections of nonfunctional pituitary adenomas (NFPAs) and four types of functional adenomas

Type of lesion	Prevalence (%)	Common age group	Most prevalent gender
Prolactinoma	50[a]	20–50 years old[b]	Females > males[c]
NFPA (endocrine-silent)	25–35[a]	>40 years old[b]	Males > females[c]
Somatotroph adenoma (acromegaly)	10–15[a]	Bimodal: ~20 years old and 50–65 years old[b]	Males > females[c]
Corticotroph adenoma (Cushing's disease)	10–15[a]	<30 years old (M)[b] >37 years old (F)[b]	Females > males[c]
TSH-releasing adenoma	0.5–3[a]	50–60 years old[b]	Females > males[c]

[a]Refs. [29–31]
[b]Refs. [29, 32–34]
[c]Ref. [35]

(at least two measurements); (2) nocturnal salivary cortisol (two measurements); (3) 1-mg overnight dexamethasone suppression test (DST); and (4) longer low-dose DST (2 mg/d for 48 h) [40]. While these tests are sensitive to elevated levels of glucocorticoids, they are not specific [41]. If the results from the initial tests are positive, then additional tests need to be done to differentiate between Cushing's syndrome and pseudo-Cushing's syndrome, which can be achieved by dexamethasone-corticotrophin releasing hormone (CRH) test [11]. Furthermore, if ACTH is elevated, a combination of CRH/desmopressin tests, high-dose dexamethasone test, and pituitary MRI may be used to confirm pituitary source [11]. In order to distinguish from an ectopic ACTH tumor, inferior petrosal sinus sampling (IPSS) using desmopressin or CRH is the single most important test [39, 42, 43]. The first-line treatment for Cushing's disease is microscopic or endoscopic transsphenoidal adenomectomy [44, 45] by an experienced surgeon as it is associated with a complete remission rate of 65–90% for microadenomas and 65% for macroadenomas [39, 46–52]. In the event there is incomplete resection of the tumor, a repeat transsphenoidal surgery is recommended provided there is radiological evidence of the tumor [47, 53]. Should transsphenoidal surgery fail to achieve biochemical remission, additional treatment should be pursued either with radiosurgery or medical management with agents inhibiting ACTH release

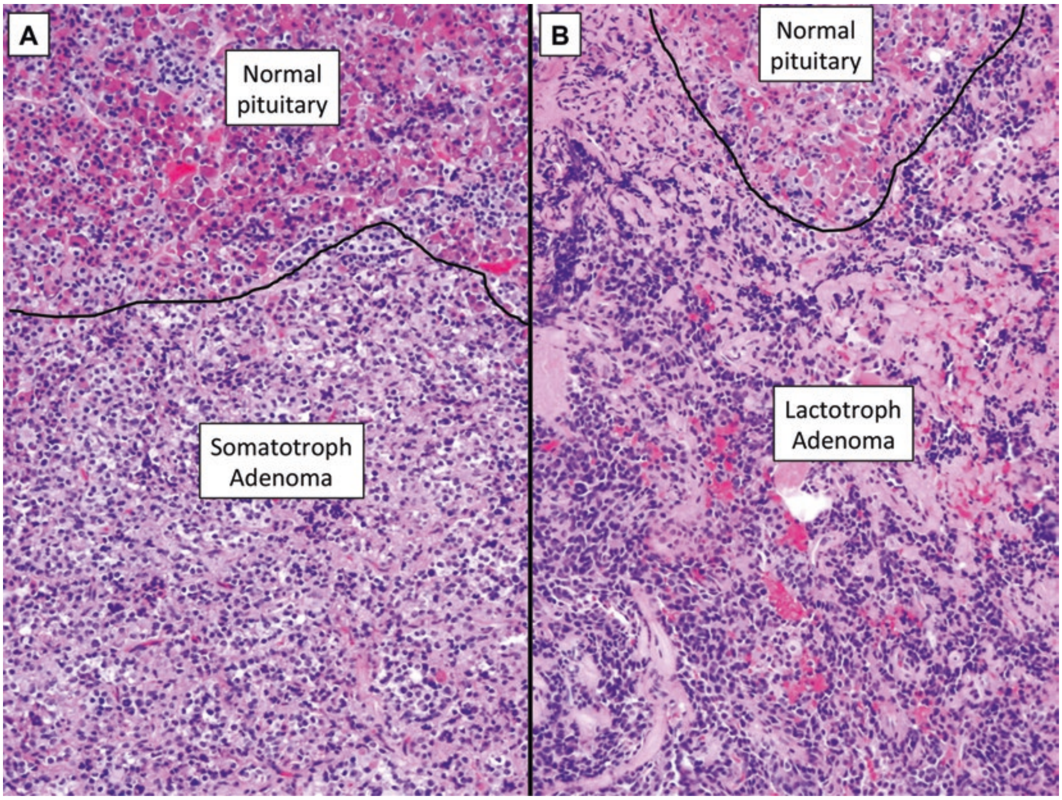

Fig. 4.3 (**a**, **b**) **Histological appearance of gland-tumor interface**. Shown are hematoxylin and eosin stains of (**a**) somatotroph and (**b**) lactotroph adenomas resected from a single patient with two microadenomas. A sharp gland-tumor interface is seen with both adenomas

in the pituitary gland, agents targeting steroid biosynthesis in the adrenal glands, or agents targeting the cortisol receptor in target tissues.

Nonfunctional Pituitary Adenomas

Due to their lack of hormone production and insidious growth pattern, NFPAs (Figs. 4.2a, b) are diagnosed either incidentally on unrelated radiologic evaluations or due to symptoms of mass effect of the lesion on surrounding structures [23–26]. According to the recent guidelines established by the Congress of Neurological Surgeons, high-resolution MRI is recommended for radiologic assessment of NFPA, while routine endocrine evaluation of all anterior pituitary axes for hypothyroidism, prolactin testing for high levels, and high levels of IGF-1 to rule out growth hormone hypersecretion, are recommended [28, 54]. Ophthalmological tests, such as automated static perimetry and visual evoked potentials, are used to determine early visual deficits and optic nerve functioning, respectively [55]. First-line treatment for NPFA is surgical resection by microsurgical or endoscopic transsphenoidal surgery, with the endoscopic approach providing greater visualization of the surgical field [56]. In invasive NFPA cases, where there is significant suprasellar, temporal, and frontal extension, a combined transcranial and transsphenoidal surgical technique can be used [56]. In the event of residual or recurrent NFPA, radiosurgery or radiation therapy is recommended to lower the risk of tumor progression. In cases where there is no residual or only small residual adenoma postoperatively, observation through serial imaging can be followed [57].

4.4 Surgical Anatomy

In general, treatment options include observation through serial imaging for smaller asymptomatic NFPAs, medical management with dopamine agonists for prolactinomas, or surgical resection through a transsphenoidal approach followed by medical management or radiation for residual tumor as needed based on whether the tumor is an NFPA or FPA. Medical treatment options are reviewed elsewhere [7, 8, 13, 37, 39, 47, 58–64]. Before understanding the nuances of transsphenoidal surgery, pertinent surgical anatomy of the transsphenoidal corridor must be understood.

The pituitary gland sits in the sella turcica of the sphenoid bone, which is present in the center of skull base in the

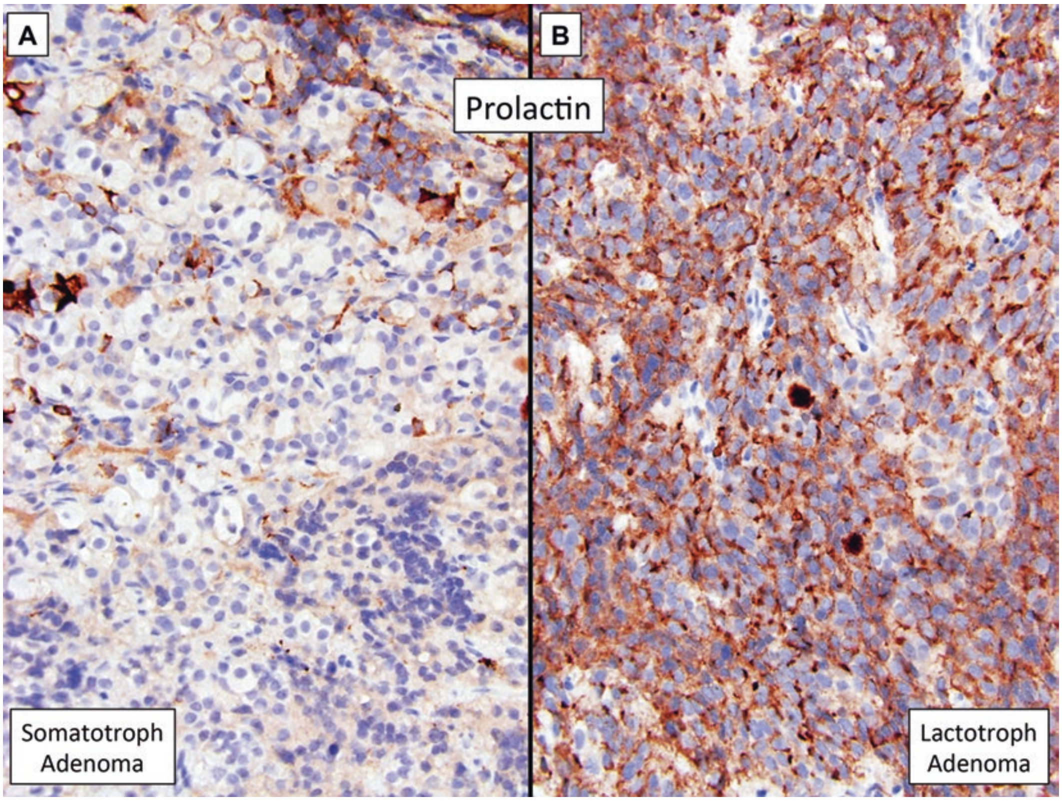

Fig. 4.4 (**a**, **b**) **Histological appearance of prolactin-staining of pituitary adenomas**. Shown is peroxidase-based prolactin immunostaining of (**a**) somatotroph and (**b**) lactotroph adenomas resected from a single patient with two microadenomas (same patient as in Fig. 4.3a, b). *Brown stain* represents areas of prolactin immunostaining. Note that there is prolactin immunostaining in the normal gland, but the density of this staining is higher in the prolactinoma

middle cranial fossa [65–67]. Gaining neurosurgical access to the sella requires understanding the vital anatomical structures surrounding the sella superiorly, posteriorly, and laterally [68–70]. Superior to the sella, the diaphragm sellae separates the pituitary gland from the CSF of the suprasellar cistern, which contains the optic nerves and chiasm. Posterior to the sella, the bony dorsum sella, and the inferior extension of the posterior clinoid processes, separates the pituitary gland from the dura overlying the prepontine cistern, which contains the basilar artery anterior to the brain stem. The cavernous sinus and internal carotid arteries are lateral to the sella. These anatomic constraints, superior, posterior, and lateral to the sella, make the endonasal route through the sphenoid sinus the preferred surgical route to access pathologies in the pituitary gland.

Sellar tumors often extend upward into the suprasellar region, leading to clinical consequences as a result of compression of the optic chiasm. If the optic chiasm is prefixed and located more anteriorly over the bony tuberculum sellae, a variant occurring in 10% of cadaveric specimens, a growing pituitary tumor can cause mass effect on the retrochiasmatic optic tracts. If the chiasm is postfixed and located more posteriorly over the bony dorsum sellae, a variant occurring in 10% of cadaveric specimens, a growing pituitary tumor can exert mass effect on the prechiasmatic optic nerves (Fig. 4.6) [70].

The suprasellar compartment houses the anterior incisural space, which is located between the free edges of the tentorium and the front of the midbrain [65, 71]. This area is clinically significant because the infundibulum crosses this region to reach the diaphragma sellae. The optic nerve and chiasm, anterior part of the optic tract, oculomotor nerve, and the posterior part of the olfactory tracts also pass through the suprasellar region. Arterial structures in the suprasellar region include the Circle of Willis, basilar, and internal carotid arteries. Specifically, the posterior portion of the Circle of Willis and the apex of the basilar artery are present in the anterior incisural space below the floor of the third ventricle, while the anterior and posterior cerebral arteries and the perforating branches from the internal carotid, anterior choroidal, anterior and posterior cerebral, and anterior and posterior communicating arteries send branches to the walls of the third ventricle and anterior incisural space. A suprasellar lesion or suprasellar extension of a pituitary adenoma can

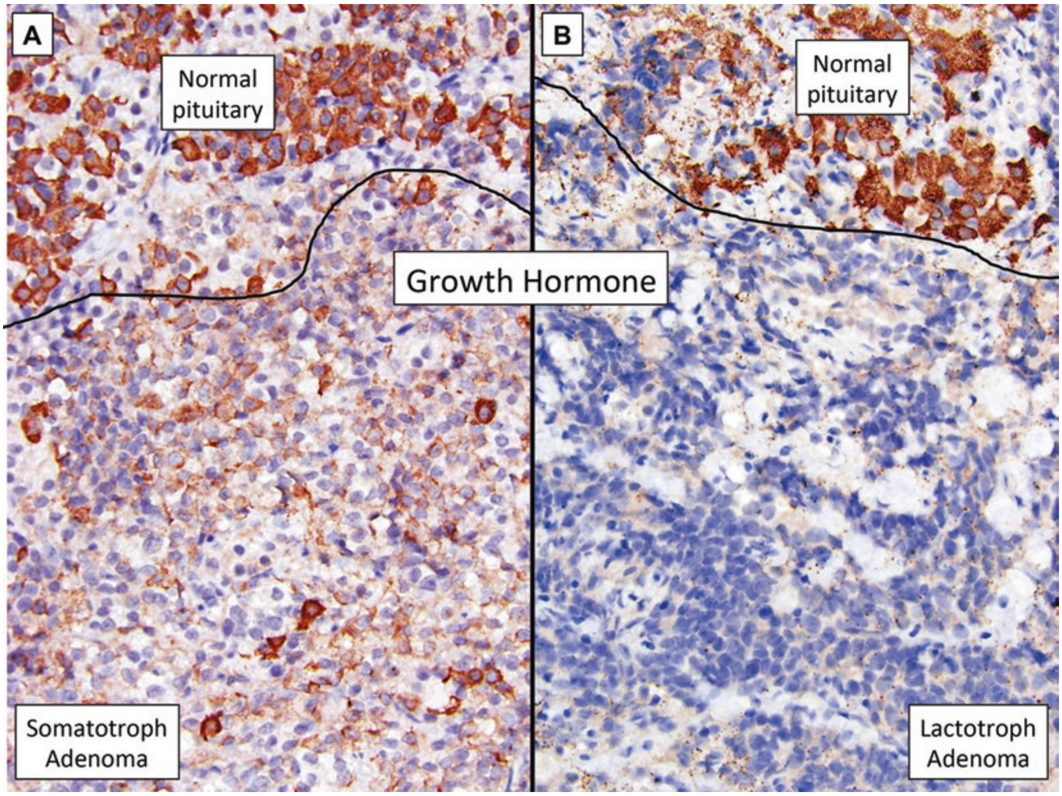

Fig. 4.5 (**a**, **b**) **Histological appearance of growth hormone (GH) staining of pituitary adenomas.** Shown is peroxidase-based GH immunostaining of (**a**) somatotroph and (**b**) lactotroph adenomas resected from a single patient with two microadenomas (same case as shown in Fig. 4.3a, b). *Brown stain* represents areas of GH immunostaining. Note that there is GH immunostaining in the normal gland, but the density of this staining is higher in the somatotroph adenoma

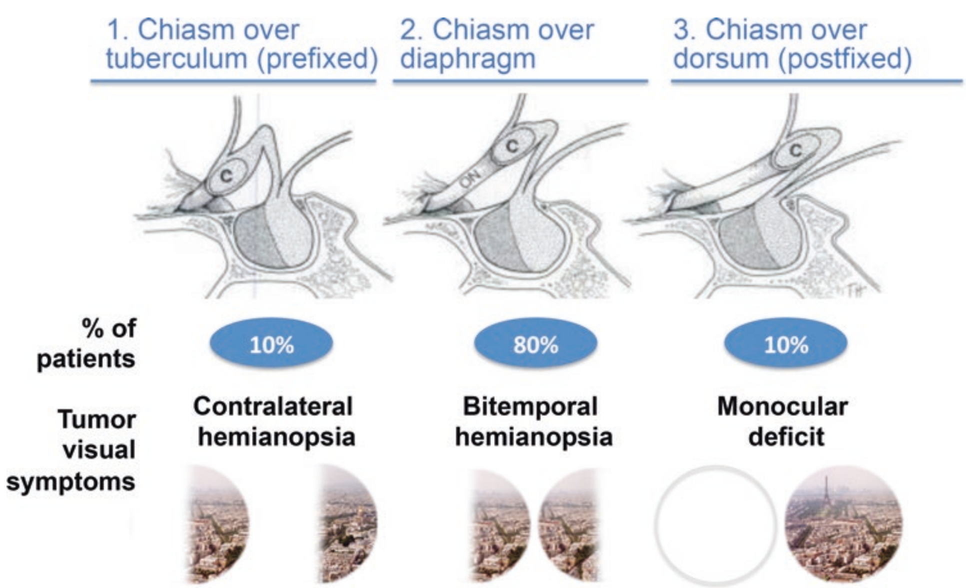

Fig. 4.6 **Three possible optic chiasm locations in patients.** Shown are three potential locations of the optic chiasm in the midsagittal plane: an anterior location for a prefixed chiasm over the bony tuberculum sellae, a central location over the diaphragm sellae, and a posterior location for a postfixed chiasm over the bony dorsum sellae. This impacts the potential visual field deficits that arise from a growing adenoma – an adenoma growing in a patient with a prefixed chiasm will exert mass effect on the retrochiasmatic fibers and could therefore cause a contralateral hemianopsia; an adenoma growing in a patient with a central chiasm will exert mass effect on the chiasm and could cause a bitemporal hemianopsia; and an adenoma growing in a patient with a postfixed chiasm will exert mass effect on the prechiasmatic optic nerve fibers and could therefore cause a monocular deficit

distort or encase these arteries and their perforating branches, making it vital to review preoperative imaging for these vessels before surgery [71–76].

4.5 Surgical Technique

Endoscopic transsphenoidal surgery is a versatile approach for resection of both FPA and NFPAs, combining the minimal invasiveness of endonasal approaches with the superior panoramic visualization offered by the endoscope relative to the microscope [13, 36, 37, 47, 56]. While many studies have reported similar to no difference in the effectiveness of endoscopic versus microsurgical transsphenoidal surgery, a recent extensive meta-analysis comparing the outcomes of endoscopic transsphenoidal and microsurgical technique found the endoscopic approach to be superior with higher safety and efficacy as well as higher gross total resection of the tumor, lower rate of postoperative septal perforation, and shorter postoperative hospital stays [77–79].

Operative Setup

Endoscopic endonasal pituitary surgery can be done by a neurosurgeon working alone or by a neurosurgeon working in partnership with an otolaryngologist. Prior to the operation, a surgical plan should be made, including decisions about whether to place a lumbar drain preoperatively, whether a fat graft will be harvested, and understanding of the tumor's involvement of vascular structures. Considerations specific to the patient such as airway issues that might arise with acromegaly, including the potential need for awake fiber-optic intubation in patients with excessive oropharyngeal soft tissue, and blood pressure and electrolyte issues that can arise with Cushing's disease patients should be discussed with the anesthesiologist before proceeding with surgery. Once the patient is under general anesthesia, an orotracheal intubation is performed. An antibiotic to cover nasal flora and stress-dose steroids are typically given. Many institutions give stress-dose steroids for all patients regardless of the size of the pituitary tumor, even when the tumor is causing preoperative hypopituitarism. For surgery, the patient is positioned supine with the trunk elevated to 10–20° and the head turned toward the surgeon with their head fixed in a Mayfield headrest or in a three-pin head-fixation system, with neuronavigation as a useful tool [80–82]. Special care should be taken to avoid obstruction of the jugular outflow and/or stretch injury to the brachial plexus. Depending on the anatomy of the lesion, the vertical position of the patient's head is manipulated. If the lesion is present in the clivus, sphenoid sinus, or is a microadenoma, the head is slightly flexed such that the bridge of the nose is parallel to the floor of the operating room. If the lesion extends into the suprasellar region or is a macroadenoma, the head is inclined toward the floor of the operating room. Next, the nasal cavity and surrounding facial region are disinfected with a 5% povidone-iodine solution [83].

Instrumentation

There are six components that enable visualization during endoscopic pituitary surgery: the light source, the endoscope, fiber-optic cable, camera, monitor, and video recording equipment [80]. Most endoscopes are rigid with 4 mm diameter, 18 or 30 cm length, and a 0°, 30°, or 45° lens [80]. For children or patients with very narrow nasal passages, smaller endoscopes ranging from 1.9 to 2.7 mm diameter are available. In order to enable the endoscope to be brought in and out of the nasal passages with minimal accumulation of debris on the lens, the endoscope is inserted through a sheath connected to an irrigation system for clearing the lens during the procedure [82]. When the procedure is being performed by a surgeon working alone, an endoscope holder can be used to provide stability to the endoscope for a fixed image of the surgical field and to free the surgeon's hand.

The endoscope is connected to the light source by a high-quality fiber-optic cable. The light source commonly used is a Xenon cold light as this light source has a color temperature similar to that of solar light and provides lower heat dispersion with greater illumination in contrast to halogen lights [80, 81].

Surgical tools used in endoscopic transsphenoidal surgery are distinct from those used in microsurgical techniques, which require bayonet-shaped tools. Instead, straight tools are used in the endoscopic technique as they can be inserted close to the endoscope along its axis and can be equipped with different angled tips to manage all areas of the surgical site. Several surgical tools are available in the market and are used variably by surgeons [84, 85].

The endoscopic light source, monitor, video camera, and recorder are placed behind the head of the patient and in front of the operator. The lead surgeon is on the right of the patient, while the scrub nurse and assisting surgeon is to the left of the patient. The anesthesiologist with his equipment is usually at the level of the patient's legs (Fig. 4.7).

Technique

The endoscopic endonasal transsphenoidal surgical technique can be divided into three phases: *nasal phase*, *sphenoid phase*, and *sellar phase* [80, 84]. During the nasal phase, three clinically significant goals are met: exposure of nasal cavity anatomy, formation of a surgical pathway for tools and navigation, and gaining access to the sphenoid

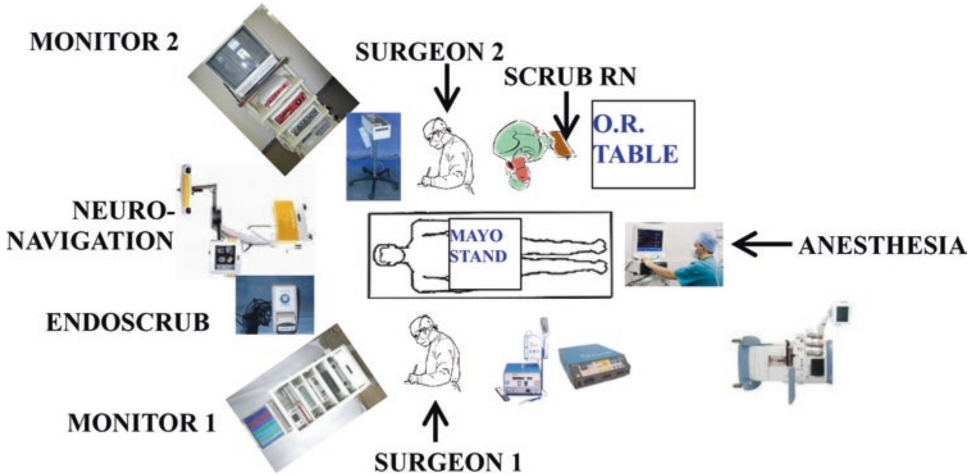

Fig. 4.7 Operating room setup for endoscopic endonasal pituitary surgery. Shown is a diagram of a typical operating room setup for endoscopic endonasal pituitary surgery. The anesthesiologist is at the foot of the bed with an extended endotracheal tube secured to the patient's side and running to the ventilator. Endoscopic monitors are placed at 45° angles to the left and right of the patient allowing surgeons working on each side of the patient to visualize the monitors. The *photo* shows surgeons operating across from each other, but they can also both operate on the patient's right side, with the otolaryngologist in front of the neurosurgeon

sinus. A 0- or 30º endoscope with 4 mm diameter is inserted into one nostril. Typically, the right nostril is preferred as a surgical route because of easy access to the surgeon standing to the right of the patient. But in certain scenarios, the left nostril may be used due to narrowing of the right nostril as a result of scarring, hypertrophy of turbinates (e.g., in acromegaly), septal deviation, and synechiae due to prior sinonasal surgery [82]. The first structure to be identified is the inferior turbinate laterally and the nasal septum medially. At the level of the head of the middle turbinate, cottonoids soaked in epinephrine (1:100,000) are placed between the nasal septum and middle turbinate to vasoconstrict the area and widen the nasal space [80, 83]. To further widen the nasal space for an adequate surgical route, the head of the middle turbinate is dislocated laterally where, if needed, it can be protected by gauze or a cottonoid to prevent abrasive injury during the procedure. As the endoscope is moved along the floor of the nasal cavity, it reaches the choana, an orifice which communicates with the nasopharynx. The medial margin of the choana is the vomer, which also marks the midline of the approach. At this point the endoscope is angled upward along the roof of the choana, and the sphenoethmoidal recess until the sphenoid ostium (usually 1.5 cm above the roof of choana) is reached to access the sphenoid sinus.

The sphenoid phase involves anterior sphenoidotomy for accessing the sella and pituitary gland. In order to prevent bleeding from the septal branches of the sphenopalatine artery, once the endoscope reaches the sphenoid cavity, the septal mucosa is coagulated, beginning 0.5 cm from the top of choana to the superior border of the nasal cavity [80, 82]. Next, the septal mucosa is incised and mobilized to the side with a microdissector following which the nasal septum is broken at the sphenoid rostrum to achieve wide exposure of the sphenoid floor [86]. Anterior sphenoidotomy can be performed using either Kerrison rongeurs or a microdrill by proceeding in a circumferential direction. Next, the sphenoid rostrum is removed in fragments and not en bloc to prevent any laceration and bleeding in the nasal mucosa during retraction from the nasal cavity [80, 82]. In order to ensure surgical tools reach the sella within the visible field of the endoscope, it is necessary to make sure the removal of the anterior wall is wide. Caution must be taken in the inferolateral direction to prevent injury to the sphenopalatine artery or its branches as discussed in "Surgical Anatomy." One or more septa can be seen inside the sphenoid sinus, and special care must be taken when removing septa implanted over carotid prominences. Septa are removed using nasal forceps or cutting bone punches without detaching the sphenoidal mucosa, except when mucosa is too prominent or adenomatous infiltration is seen or suspected [80, 81]. Following removal of the septa, the posterior and lateral walls of the sphenoid sinus are endoscopically visible, with the sellar floor at the center, planum sphenoidale above it, clival indentation below it, and bony prominences of the intracavernous carotid artery and optic nerve lateral [80, 81].

The sellar phase involves exposing and resecting the lesion (Fig. 4.8a–d). Once the sphenoid sinus roof is completely cleared and visible, a longer endoscope (4 mm in diameter, 0º angled lens, 30 cm length) is used and, if desired, can be stabilized by fixing it to an adjustable endoscope holder to free both hands of the surgeon. In the case of most macroadenomas, the sellar floor can be opened using a dissector and enlarged with a rongeur [81]. However, if the sellar floor is very thick, as is often the case for microadenomas,

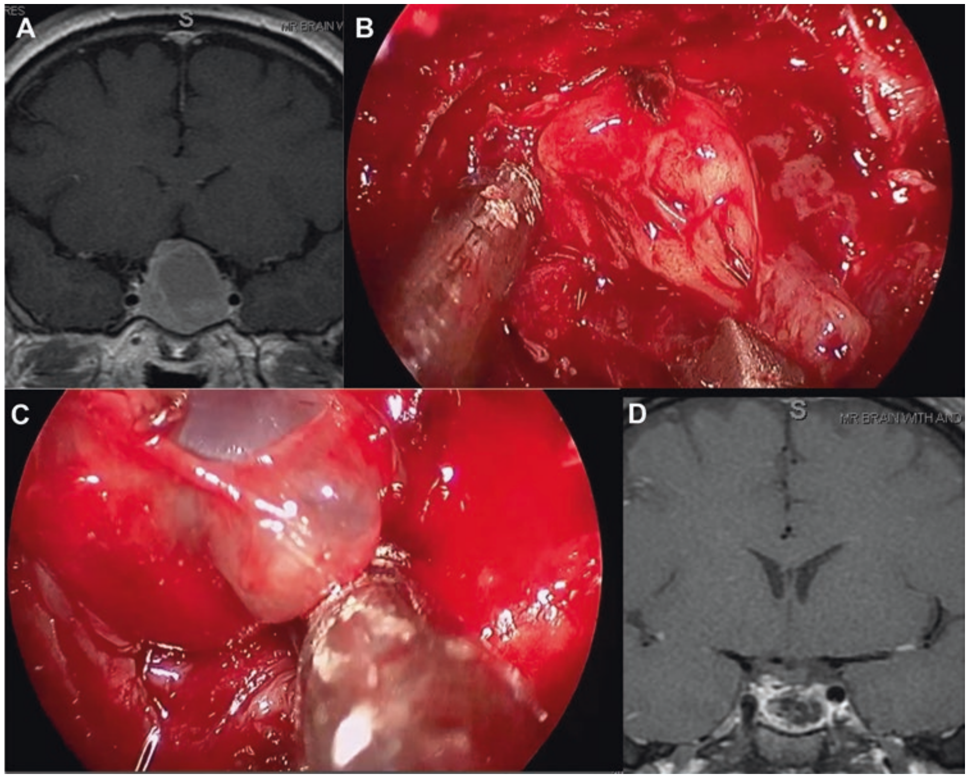

Fig. 4.8 **(a–d) View through the endoscope of a pituitary adenoma being resected.** Shown are MRI images and endoscopic view from a particular case of (**a**) nonfunctional pituitary macroadenoma with internal blood products in the tumor in a patient with vision loss with tumor to the left and some normal gland to the right as seen on a coronal T1 post-gadolinium MRI; (**b**) view through a zero degree endoscope during endoscopic transsphenoidal resection of the yellow hemorrhagic tumor being resected from the sella; (**c**) view through a zero degree endoscope of the last bit of adenoma being removed with the translucent diaphragma sellae prolapsing down into the sella; and (**d**) postoperative MRI showing no residual adenoma

a high-speed microdrill or bone punchers are used. In either case, the opening in the sellar bone is enlarged according to the pathology of the lesion. Next, the exposed dura is incised in a cruciate fashion or in a linear rectangle [80, 81, 86]. In the case of macroadenomas, the dural incision is often bloodless due to the compression and obliteration of superior and inferior intercavernous sinuses, while, in the case of microadenomas and specifically in Cushing's disease, the entire sellar dura may be covered by one or two venous channels and can lead to dural bleeding on incision [80, 82]. To circumvent this problem, the intercavernous sinuses can be cauterized or secured and sealed using surgical clips around them [80, 82]. Extra caution must be taken when incising dura in microadenomas because sometimes an ectatic carotid artery may also be located within the sella in acromegalic patients [87, 88].

Endoscopic tumor resection uses techniques identical to those of conventional microsurgical endonasal approaches, specifically internal debulking, capsular mobilization, and extracapsular dissection of neurovascular structures, along with coagulation and capsule resection [83]. Using various curettes, suction, and grasping forceps, the lesion is removed in fractions. Following removal of the lesion, the 0º endoscope can be replaced with a 30 or 45º endoscope if needed to visualize the lateral surgical borders to ensure maximal extent of resection. If an intraoperative CSF leak occurs during the resection, repair can be done using a variety of techniques, including autologous devascularized tissue graft such as a periumbilical fat graft or fascia lata, as well as autologous vascularized tissue graft such as a nasoseptal flap [81, 89–92]. The sellar and sphenoid floor can be reconstructed with bone pieces, although the importance of such repair remains subject to debate. At the end of the procedure, hemostasis is obtained and the middle turbinate is restored gently to its normal anatomical position. The amount of packing to place in the nares is subject to the discretion of the surgeon, with a septal splint sutured to the site of nasoseptal flap harvest offering value in preventing postoperative nasal crusting and bioresorbable nasal dressing such as NasoPore™ (Stryker Instruments) providing some tamponade to support the healing of the surgical site.

Special points of emphasis for larger tumors are discussed below:

1. **Macroadenomas**: Resection of macroadenomas is accomplished sequentially with the inferior and lateral regions of the lesion removed first followed by the superior aspect [80]. Following removal of visible tumor, if the diaphragma sellae does not descend, the surgeon can ask the anesthesia team to perform a Valsalva maneuver to force the diaphragma downward and any remaining unresected tumor to protrude into the sellar cavity [80, 81]. A thorough inspection of the sellar cavity and suprasellar cistern, if it was explored, must be done after resection of all visible tumor, and an angled endoscope can be used for this step, if needed. If the adenoma invades the

medial wall of the cavernous sinus, the endoscopic approach can allow chasing of the tumor through the focus of invasion or through a more lateral transethmoid-transpterygoid path through the bulla ethmoidalis of the middle turbinate followed by ethmoidectomy of the anterior and posterior ethmoid cells [84]. For such a lateral approach, the use of intraoperative Doppler probe is necessary to prevent carotid artery injury in the cavernous sinuses. Resection of tumor in the cavernous sinus requires considerable experience and judgment and may be more appropriate for functional adenomas where biochemical remission will require complete tumor resection or where maximizing extent of resection can improve the efficacy of any necessary postoperative medical therapy.

2. **Giant pituitary adenomas**: While the endoscopic endonasal transsphenoidal surgical technique has allowed resection of large adenomas [93–96], a transsphenoidal approach will leave significant enough residual for some giant adenomas (adenomas larger than 40 mm in diameter) to warrant a craniotomy either after initial transsphenoidal surgery (Fig. 4.9) or instead of transsphenoidal surgery [97, 98]. For staged approaches, it is important to minimize the time gap between the initial transsphenoidal surgery and the craniotomy due to the risk of apoplectic events occurring in residual tumor after the first surgery. For select tumors where the amount of residual adenoma is small enough to not require a second stage craniotomy, follow-up MRI in 4–6 weeks may be done to evaluate the residual tumor burden and reveal its collapse into the sella, in which case repeat transsphenoidal surgery can be considered.

4.6 Postoperative Care and Complications

Postoperative pain and discomfort has been reported to be minimal and patients rarely need analgesics [82, 86]. Due to potential occurrence of diabetes insipidus, patients are kept in the hospital at least for overnight observation [86, 99]. Serum sodium and urine output are measured the night after surgery as well as the following morning. For hypersecreting adenomas, the levels of the hypersecreted hormone can be checked postoperatively to assess for biochemical remission, recognizing that IGF-1 takes up to 6 months to normalize after surgery for acromegaly; immediate prolactin normalization typically occurs after prolactinoma resection; and cortisol sub-normalization is anticipated to occur with biochemical remission of Cushing's disease [100]. At discharge, patients are encouraged to engage in copious irrigation of the nasal vestibule and cavity three times a day for 1 week [80]. Irrigation of the nasal cavities serves to protect from infections, wash out small clots in the nasal cavities, and prevent possible endonasal synechiae. Patients are discharged on maintenance dexamethasone if there is preoperative evidence of adrenal insufficiency [82, 86]. At 12 weeks, in addition to clinical evaluation, an MRI is done to evaluate the extent of

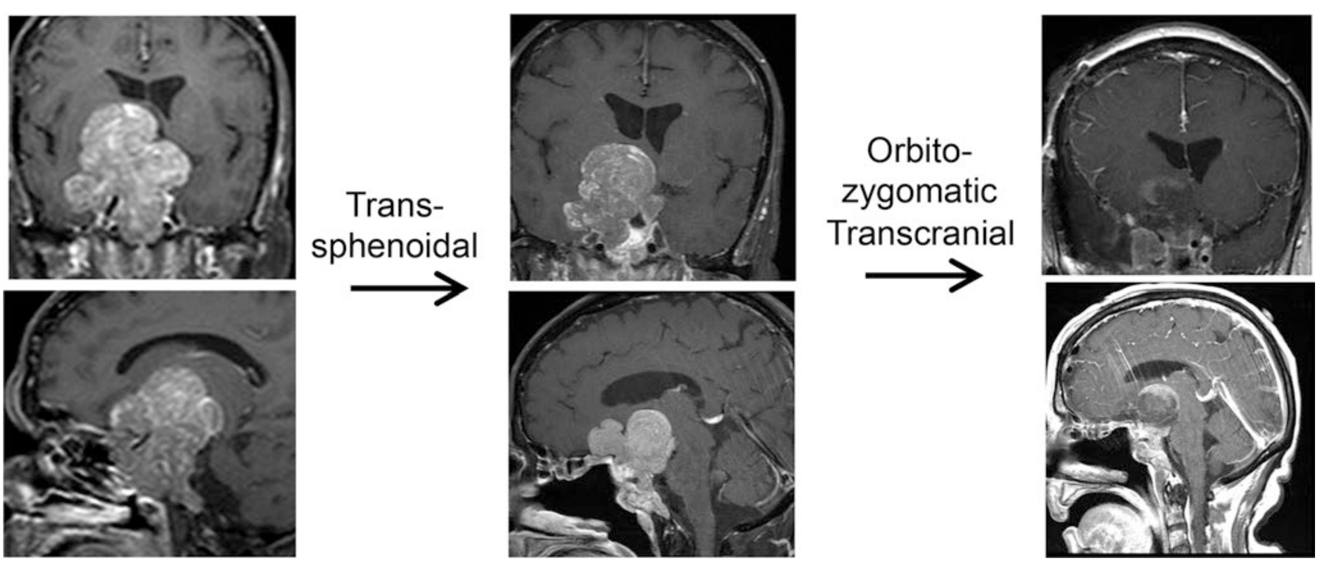

Fig. 4.9 Example of a giant pituitary adenoma requiring a two-staged approach. Shown is an example of a giant nonfunctional pituitary macroadenoma in a patient with two decades of vision loss. MRI revealed tumor eroding into the right Sylvian fissure with significant inferior extension into the sphenoid sinus. A staged approach was performed, with an endoscopic transsphenoidal resection of the sphenoid sinus and sellar components of the tumor done first, with intraoperative pathologic analysis confirming the diagnosis of pituitary adenoma. The next morning, the patient was brought back to the operating room where a right orbitozygomatic craniotomy was performed for resection of remaining tumor

surgical resection. For functional adenomas, the hormone being hypersecreted is also assessed at the 12-week follow-up [8, 13, 47, 53, 57, 60, 101, 102].

Complications of endoscopic transsphenoidal surgery, albeit uncommon, arise as a result of tumor characteristics (size and extension) and surgical approach and manipulation of the pituitary gland and surrounding structures [100, 103]. Complications associated with the endoscopic approach may be divided into four groups based on the anatomic location from which the complication arises: (1) nasofacial, (2) sphenoid sinus, (3) parasellar, and (4) endocrine [104]. Nasofacial complications primarily include epistaxis due to damage to vessels in the nasal cavity such as sphenopalatine artery and its branches [100]. Sphenoid sinus complications involve sphenoid sinusitis occurring in reaction to the transnasal portion of the procedure [104]. Parasellar complications include CSF rhinorrhea, swelling or infarction of residual tumor, meningitis, hematoma in the resection cavity, visual defects and transient CN VI palsy due to postoperative edema, and carotid artery injury [100, 104–106]. Endocrine complications include diabetes insipidus and new postoperative hypopituitarism [97, 104].

4.7 Surgical Pearls

- For newer neurosurgeons or those new to endoscopic transsphenoidal techniques, it is best to start with smaller cases before applying the approach to larger, riskier cases.
- Collaborating with an experienced otolaryngologist with skull base training can allow a two-surgeon teamwork concept to be applied to endoscopic endonasal pituitary surgery, which can reduce operative time and improve outcomes.
- It is important for the neurosurgeon operating on pituitary tumors to understand the delicate aspects of neuroendocrine surgery, such as identifying and preserving the normal gland by defining the gland-tumor interface and using an extracapsular dissection whenever possible.

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5 Suprasellar Pathology

Sacit Bulent Omay, Vijay K. Anand, and Theodore H. Schwartz

5.1 Introduction

The suprasellar space is defined by the diaphragma sellae inferiorly and the floor of the third ventricle superiorly. A "craniectomy" achieved by removing the tuberculum sella, prechiasmatic sulcus, and posterior planum sphenoidale opens the suprasellar space. It can be divided into the infrachiasmatic, suprachiasmatic, and retrochiasmatic areas. The infrachiasmatic space is between the optic chiasm and the pituitary gland. The infundibulum is the key structure in this area and is covered by the suprasellar cistern arachnoid anteriorly and the membrane of Liliequist posteriorly. Tuberculum sellae meningiomas that extend into this area displace the infundibulum and superior hypophyseal arteries posteriorly. The suprachiasmatic space is above the optic chiasm as the name implies. The posterior olfactory tract, the A1 segments, the anterior communicating artery, and A2 segments of the anterior cerebral artery are the key structures in this area. Planum meningiomas arise in this space and generally displace the optic chiasm and anterior cerebral artery complexes as well as recurrent artery of Heubner and the fronto-orbital artery posteriorly and/or inferiorly. The retrochiasmatic space extends from the infundibulum to the posterior perforated substance, cerebral peduncles, and interpeduncular cistern posteriorly and to the third ventricle superiorly. Craniopharyngiomas and Rathke's cleft cysts often extend into this space [1].

5.2 Rationale

Surgical lesions of the suprasellar region are varied in pathology (Table 5.1), but they commonly present a challenge to the neurosurgeon due to the proximity to the optic nerves and chiasm, pituitary gland, infundibulum, hypothalamus, and third ventricle. Lesions in this region are frequently benign, but pathologies and surgical interventions to remove these may result in visual field loss, endocrine dysfunction, and hydrocephalus. Surgical decision-making becomes a complex process of balancing aggressive resection strategies with the goal of minimizing cognitive, ophthalmologic, and endocrine morbidity [2].

Until recently, lesions of the suprasellar space were treated solely with open transcranial approaches from above. These approaches involve brain retraction; require work through small triangles defined by the cranial nerves, the internal carotid artery (ICA), and its branches; and cause cosmetic compromise [3–6]. The last decade has been marked by the advancement and the popularization of the endonasal endoscopic approaches (EEA) that use minimal access strategies such as a natural orifice like the nostrils but also facilitate aggressive oncologic surgery. The endoscopic endonasal transplanum and transtuberculum approach is the defining EEA to the suprasellar cistern and is the most direct route to the region that avoids manipulation of the optic nerves and carotid artery [5]. It provides a direct route to suprasellar lesions that obviates the need for brain retraction. Also, unlike a transcranial approach, it does not place critical neurovascular structures such as the optic nerves and carotid arteries between the surgeon and the tumor. It facilitates complete, bilateral optic canal decompression without manipulation of a compressed optic nerve. Moreover, approaching these

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Table 5.1 Suprasellar pathology

Pituitary macroadenoma
Rathke's cleft cyst
Meningioma
Craniopharyngioma
Epidermoid and dermoid tumors
Glioma
Arachnoid cyst
Metastasis
Germ cell tumors
Hypothalamic hamartoma
Aneurysm
Abscess
Lymphoma
Chordoma

tumors from below enables the surgeon to remove the bone at the base of the tumor, which is a common site for meningioma recurrence, and to interrupt the dural vascular supply early in the operation, minimizing blood loss [7].

With joint effort from the disciplines of neurosurgery and otolaryngology, endoscopic endonasal procedures can now deliver a unique view above the sella into the suprasellar cistern, even to the roof of the third ventricle and the interpeduncular cistern [8]. The key to the success of this "minimal access" but maximally aggressive approach is in careful case selection. Due to its limitations and capabilities, EEA requires specific understanding of the pathology from an endoscopic perspective and the acquisition of the key methods that are required to endoscopically access and resect these lesions [5]. The case selection and the surgical decision-making for an endonasal endoscopic surgeon involve three questions that need to be answered [5]: A target (where?), the nature of the lesion (what?), and the approach (how?). For the purpose of this chapter, the target is the suprasellar cistern, and we will go over the approach required to get there and discuss the nuances for specific pathologies.

5.3 Patient Selection

Indications

Any lesion located or extended into the suprasellar space exerting mass effect, causing neurologic, endocrinologic, or ophthalmologic symptoms, or any lesion which requires a tissue diagnosis is an indication for a suprasellar approach to either resect or biopsy these lesions as necessary**.** Large tumors that cannot be completely resected using an endonasal approach are not always a contraindication. Depending on the age of the patient and the surgical goals, internal decompression or staged resection with an additional transcranial or cranioendoscopic approach may be appropriate [9].

Patients with any visual symptoms or compression of the optic apparatus on imaging should undergo a detailed neuro-ophthalmologic examination with visual field testing. Full endocrinologic workup is usually indicated to establish baseline hormone deficiencies. A comprehensive neuroradiological evaluation of the lesion is of paramount importance. MRI of the brain and specifically a pituitary protocol MRI with fine coronal cuts is required for tumors of the sella. The location of the bony septations within the sphenoid sinus and the location of carotid arteries should be determined by CT or CTA as needed [7].

The pneumatization level of the sphenoid sinus should be evaluated. A detailed plan for the extent of the craniectomy to adequately expose a surgical corridor should be determined. Septal perforations and spurs should be identified to establish whether a nasoseptal flap can be raised and from which sides of the septum or whether another type of flap will be required for closure.

Contraindications and Considerations

The endonasal endoscopic approach is often not suitable for malignant tumors that might require en bloc resection. However, there are no data showing that en bloc resection is any better with a piecemeal gross-total resection with negative margins, followed with fractionated radiation or radiosurgery [7, 10–14]. Pathology which extends laterally over the orbits or lateral to or behind the carotid arteries is difficult to remove using EEA. The width of the planum sphenoidale, between the laminae papyracea, has been measured in cadaver studies at 26 ± 4 mm, which narrows to 16 ± 3 mm at the posterior aspect of the tuberculum sellae [7, 15]. This width defines the preferred corridor by the EEA and optimally exposes the medial aspect of the optic canals [7]. Beyond these boundaries, even though visualization is achieved with the use of angled scopes, resection of a lesion around a corner may not be technically feasible [9]. Encasement of neurovascular structures is not a contraindication to this approach, but the surgeon will have to consider his or her ability to safely dissect a tumor off a vital structure and should keep in mind the possibility of radiosurgery and fractionated radiation to control the growth of residual unresectable tumor [9].

Although, cavernous sinus invasion is not an absolute contraindication, it requires the surgeon to be cognizant of surgical goals. The additional risks to the neurovascular structures should be considered [16]. Similarly, vascular encasement of the A2 branches of the anterior cerebral artery is not an absolute contraindication but deserves a similar consideration. Edema within the brain or floor of the third ventricle is not an absolute contraindication, but leaving the tumor invading into the hypothalamus, which commonly occurs in craniopharyngiomas, may be appropriate to preserve function. It is important to remember the differential diagnosis of large macroadenomas. These may include suprasellar lesions such as hypothalamic hamartomas, large intracranial aneurysms, and germ cell tumors, which may not be suitable for this approach. Appropriate preoperative imaging and screening should be undertaken to rule out such lesions preoperatively [16].

5.4 Surgical Technique

Instruments

EEA instruments and the standard transcranial microsurgical instruments are different in design. Long straight, rather than bayoneted, or pistol-grip instruments are better for endoscopic approaches. Monopolar cautery is favored during the approach for nasal and sphenoid mucosal bleeding, while bipolar coagulation is used on the dura and intracranial structures. A tissue shaver or micro-debrider is useful for resection of intranasal pathology, whereas intracranial pathology requires instruments of precision such as a NICO Myriad (Indianapolis, IN) or gentle bimanual suction. 18″ and 30″ endoscopes with 0°, 30°, and 45° lenses are required. A sheath around the scope can be used to irrigate and clean the lens during the operation for maximized efficiency. A scope holder helps maintain a fixed, steady field of view during the case, which is very valuable during fine dissection although having a second individual drive the scope also has its advantages. High-definition, large monitors enhance visibility and surgeon's comfort [16].

The Approach

The operation is done under general anesthesia. We routinely use cefazolin or triple antibiotics (vancomycin, a second-generation cephalosporin and Flagyl) when the arachnoid is expected to be opened. A lumbar drain is placed, and 0.25 ml of 10% fluorescein (AK-Fluor®; Akorn, Buffalo Grove, IL, USA) is injected in 10 ml of CSF to help visualize CSF leaks [17, 18]. With the use of cottonoids soaked in 4 ml of 4% cocaine, the nasal mucosa is vasoconstricted early during the preoperative preparations. The patient's head is pinned in a Mayfield skull clamp, slightly extended (15°) and turned toward the patient's right (15°) to facilitate exposure of the subfrontal anterior cranial compartment. The head is elevated above the heart to enable better venous drainage. The abdomen and/or lateral thigh are prepped for autologous fat and fascia lata grafts. By using a 0°, 18-cm, 4-mm rigid endoscope (Karl Storz, Tuttlingen, Germany), we inject the mucosa adjacent to the sphenopalatine, anterior and posterior ethmoidal arteries, and the middle turbinates with a mixture of 1% lidocaine and epinephrine (1:100,000) [19]. A bilateral approach is generally preferred for resection of meningiomas to improve visualization and working space. Since the endoscope sits in the top of the left nostril, the superior turbinate is often sacrificed on this side to create room. For tumors in which large dural and skull base defects are anticipated, we favor harvesting single or bilateral nasal septal flaps before proceeding with the intranasal exposure (Fig. 5.1). This maximizes the size of these mucosal grafts and preserves the vascular pedicle(s) until rotation and placement at the end of the surgery. The flaps are stored in the oropharynx during the remainder of the procedure [19].

The ostium of the sphenoid sinus is located and then enlarged to expose the sphenoid sinus (Fig. 5.2). The posterior third of the nasal septum adjacent to the vomer and maxillary crest is resected with a tissue shaver. At this point,

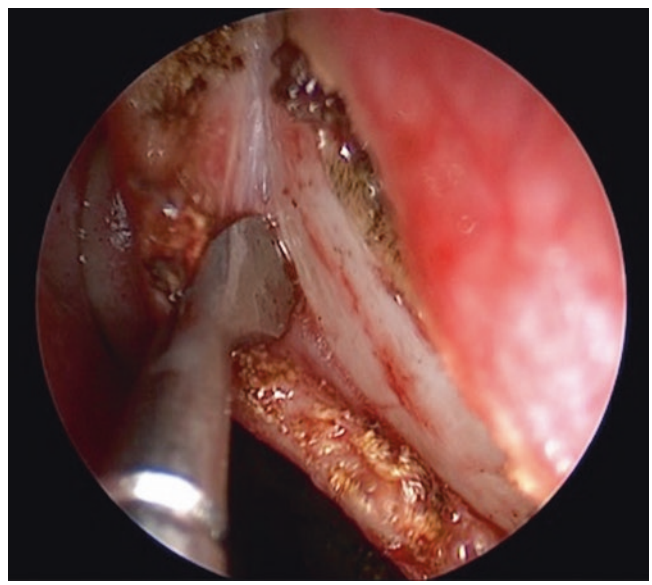

Fig. 5.1 Creation of the nasoseptal flap. Nasoseptal flap is dissected off the septum

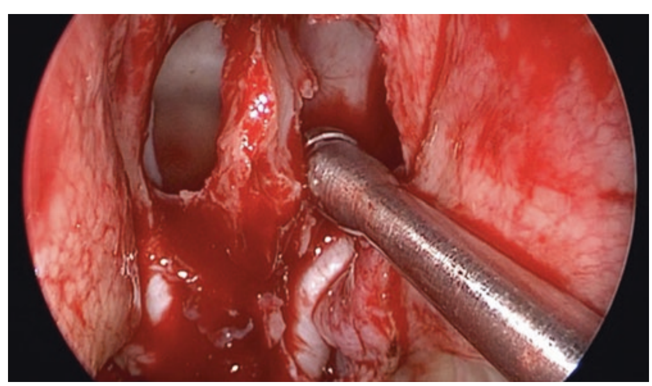

Fig. 5.2 The ostium of the sphenoid sinus is localized and then enlarged to expose the sphenoid sinus

a panoramic view of the sphenoid sinus is achieved, and bimanual surgery with four separate instruments becomes possible. The sphenoid sinus rostrum is fully exposed, and the floor and lateral wall of the sphenoid sinus are drilled down to facilitate placement of the nasoseptal flap at the end of the operation on a flush surface. Removing the anterior wall of the sphenoid sinus is recommended to provide enough room for the endoscope and instruments to fit within the sphenoid sinus during the procedure. Fracturing the thin cribriform plate should be avoided; it is a common site of iatrogenic CSF leak. Sphenoid septa are removed, and the mucosa of the sphenoid sinus is completely removed to prevent mucocele formation under the nasoseptal flap. The keel or rostrum of the sphenoid is removed so the flap will lay smoothly on the sinus floor and not have to stretch to reach the defect.

Hemostasis is achieved with the use of warm saline irrigation and/or thrombin-soaked gelfoam with gentle pressure. At this point a 0°, 30-cm, 4-mm rigid endoscope is introduced through the nostril and stabilized with a scope holder. Anatomical landmarks like the carotid protuberance, optic protuberance, and medial and lateral opticocarotid recesses are identified (Fig. 5.3). With the use of neuronavigation, the anterior and lateral extent of the sphenoid opening is evaluated. The borders of the opening are the medial opticocarotid recesses laterally and the clival recess inferiorly. At this point onward, the use of a diamond drill and copious irrigation is recommended for an accurate, atraumatic bone removal. The upper third of the sella is opened by using a high-speed drill, curette, and Kerrison rongeur. The opening is extended anteriorly above the level of the diaphragm sellae, and the planum sphenoidale is removed, the anterior extent of which is determined by the pathology. Certain cases may require bilateral posterior ethmoidectomies to optimally visualize the anterior part of the planum sphenoidale. Injury to the posterior ethmoidal arteries should be avoided by identifying and coagulating them. The use of image guidance is recommended at this stage. If necessary for the exposure, depending on the pathology, the bone overlying the medial opticocarotid recess is then drilled thin in an eggshell manner, and curettes are used to unroof the optic canals. Kerrison rongeurs should be avoided in osseous decompression of the medial optic nerves as the instrument's footplate when placed within the tight confines of the optic canal can injure the nerve and result in vision loss. It is recommended that the location of the carotid and ophthalmic arteries are verified with the use of a Doppler ultrasound probe [19, 20].

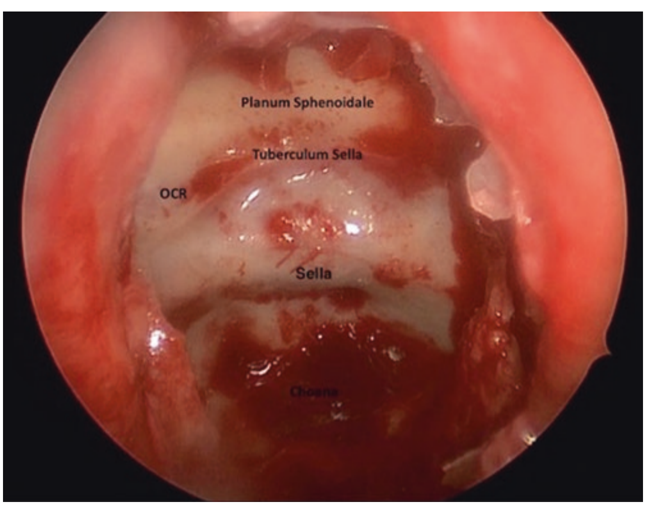

Fig. 5.3 Panoramic view of the choana, opticocarotid recess (*OCR*), sella, tuberculum sella, and planum sphenoidale. Detailed understanding of the patient's individual bony anatomy of the skull base is required

Intradural Surgery

Prechiasmal tumors, represented most commonly by meningiomas of the planum and tuberculum sella, can be visualized once the dura is opened. The micro-Doppler should be used to identify the location of the carotid artery prior to opening the dura. In the case of carotid injury at any point in the operation, hemostasis should be achieved either by coagulation or packing, and the patient should be taken for emergent endovascular assessment and treatment [16]. The dura above and below the superior intercavernous sinus is opened with a sickle knife horizontally, and the sinus is coagulated and cut just medial to the cavernous sinus bilaterally. It is a safe habit to use the sickle knife only along the midline and extend the dural opening by a blunt dissector to prevent accidental carotid injury. Dura can be cut and coagulated and can be removed with a Kerrison since it will not be used during the closure. Internal decompression is performed, depending on the consistency of the tumor using suction, a NICO Myriad® (Indianapolis, IN, USA), monopolar ring cautery (Elliquence, Baldwin, NY, USA), or microscissors. Tumor decompression with the use of a pituitary rongeur is also possible, although care must be taken to avoid placing excessive traction on the tumor with unnecessary tension on neurovascular structures. The blood supply is interrupted by the nature of the transtuberculum transplanum approach, and the tumor could be internally decompressed without having to operate around the optic nerves or carotid arteries. Once decompression is achieved, the tumor capsule could be mobilized easily; then the ACoA complex and perforating arteries can be dissected sharply off the tumor. The optic nerves and pituitary stalk can be clearly seen posterior and inferior to the tumor and can be dissected off the back of the tumor, respecting the arachnoidal plane. The remaining capsule should be removed completely if possible. The resection cavity and the course of the optic nerve into the optic canal can be examined for residual tumor using a 45°, 18-cm long, 4-mm diameter rigid endoscope. For tuberculum meningiomas, the diaphragm, which is often the site of origin of the tumor, is removed in total at the end of the operation to ensure a complete resection [20].

Postchiasmal tumors are generally represented by craniopharyngiomas or Rathke's cleft cysts, which arise from the back of the pituitary stalk and extend into the third ventricle behind the optic chiasm. Because the lesions usually extend posteriorly into the third ventricle, removal of the entire planum is usually not necessary to access them. Although the 30° scope may be useful as needed, most of the dissection can often be performed with a 0°, 30-cm, 4-mm rigid endoscope. After the dura is opened above and below the intercavernous sinus as described above, the arachnoid of the suprasellar cistern is incised, and a corridor is opened between the pituitary gland below and the optic chiasm above. This chiasm-pituitary corridor (CPC) is the gateway to access these lesions, and the size of this corridor does not usually affect the ability to access and safely resect them. If the tumor is immediately apparent and is pushing up the chiasm, this may increase the CPC. If this is not the case, slight upward pressure on the chiasm and downward pressure on the pituitary gland can be tolerated.

The stalk can be encountered in the front, in the middle, or behind the tumor. The patient's preoperative hypothalamic-pituitary function and operative goal dictates the strategy of dealing with the stalk. If the patient already has hypopituitarism with DI, the surgeon can sacrifice and resect the stalk. It is possible to mobilize the pituitary gland or work through a corridor below the pituitary gland via another small bony opening [6]. Internal decompression of the tumor, especially in large tumors, with drainage of the associated cysts may facilitate mobilization of the capsule and sharp dissection. The solid components are carefully dissected free from the optic chiasm and stalk. It is extremely important to visualize and preserve critical neurovascular structures such as the carotid arteries, the anterior communicating artery complex, and hypothalamic and chiasmal perforators. In the face of hypothalamic invasions, it is recommended to leave capsule/tumor behind for adjuvant radiotherapy rather than risk hypothalamic injury. The tumor bed including the third ventricle and interpeduncular cistern can be visualized with a 45°, 18-cm, 4-mm rigid endoscope, and residual tumor may be removed if found [20].

Closure

After resecting large tumors, the defect in the skull base may be quite large, and there may be excessive dead space under the brain. We do not recommend using fat to fill this space as it is unnecessary if a good closure is done and the postoperative imaging will be confusing. Intradural Duraform (DePuy Synthes, West Chester, PA, USA) can be used although this is not required. We recommend the use of the "gasket-seal closure" after EEA [21, 22]. A fascia lata graft that is close to double the size of the defect in the skull base is laid over the craniectomy site. A piece of Medpor® (Porex, Fairburn, GA, USA) that is cut to fit the defect is countersunk over the graft and wedged in place (Fig. 5.4). The nasoseptal flap, kept in the nasopharynx during the procedure, is placed on the defect making sure that it is placed on top and beyond the fascia lata touching skull base at the edges. Finally, the closure is covered with polymerized hydrogel (DuraSeal®; Confluent Surgical, MA, USA) on top of the nasoseptal flap to keep it in place. The sphenoid sinus, ethmoid sinuses, and roof of the nose are filled with thrombin-infused gelatin matrix (FloSeal; Baxter, IL, USA) to facilitate hemostasis [19, 20]. If a large sellar opening is performed as would be done to resect a giant macroadenoma, we do not use the gasket-seal technique since the curvature from the bottom of the sella to the front of the planum does not accommodate a rigid inlay. In this situation, we place fat in the sella, buttress the fat with Medpor, and cover it with a nasoseptal flap and DuraSeal [16].

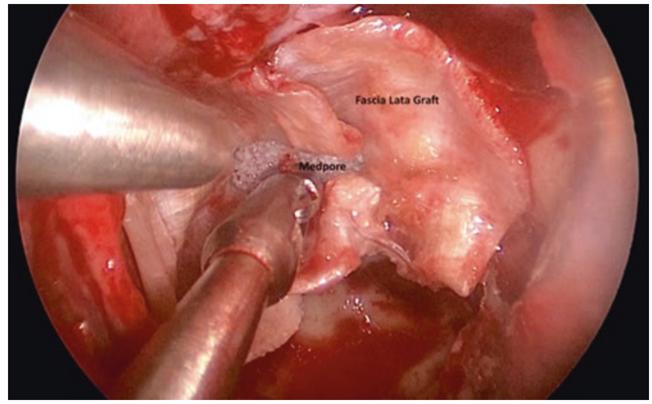

Fig. 5.4 Closure. A "Gasket seal" is the preferred method for closing skull base defects after EEA. It involves a fascia lata autograft and Medpor cut to size buttressing it

5.5 Common Lesions

Pituitary Adenomas

Pituitary adenomas represent approximately 7–17% of all intracranial tumors. They arise from adenohypophyseal cells in the anterior pituitary, and despite their benign histologic nature, they may enlarge and invade surrounding structures [23, 24]. They are categorized as functional or nonfunctional depending on whether they are hormonally active or not. Prolactinomas, the most common functional adenoma, are treated with dopamine-agonist medical therapy, with surgical treatment reserved for patients who fail or do not tolerate the medical therapy. Transsphenoidal surgery remains the primary treatment for adenomas secreting adrenocorticotropic hormone (ACTH, Cushing's disease) and growth hormone (acromegaly) with biochemical remission rates significantly correlated with tumor size and invasiveness [25]. Nonfunctioning microadenomas (<1 cm) are usually clinically silent, macroadenomas commonly present with mass effect causing visual and hormonal deficits, and cranial nerve malfunction caused by involvement of the cavernous sinus. Surgery is generally indicated for patients with visual compromise or tumor growth documented on serial imaging studies [25, 26].

With significant suprasellar extension, especially with tumors that extend above the planum sphenoidale, there is often benefit of removing the tuberculum sellae and a part of the planum sphenoidale. This allows an extracapsular dissection of the tumor to ensure a complete resection. Although some authors have recommended a combined transcranial-transsphenoidal approach to these tumors, the view is adequate with an endoscopic transtuberculum, transplanum approach to completely remove the tumor without a craniotomy [9]. EEA provides a view from the ventral midline corridor, and the pathology is encountered prior to critical nerves and vessels, minimizing the risk of damage to these structures during tumor resection. Likewise, removal of the tuberculum sella and part of the planum sphenoidale, as well as exposure of the ventral and medial cavernous sinus, allows the surgeon to reach around the capsule of the tumor and not just perform an internal decompression but a gross-total resection [27, 28].

EEA can be safe and effective for the resection of pituitary adenomas of any size or invasiveness without significant lateral extension beyond the carotid bifurcation. Endonasal approaches to these lesions have been shown to have a significantly higher rate of gross-total resection, better visual outcome, and less recurrences. CSF leak rates and meningitis were found to be lower for endoscopic approaches than for transcranial approaches [28]. The advantages of endonasal approaches are most profound in tumors with suprasellar extension and CS invasion [29].

Craniopharyngiomas

Craniopharyngiomas arise from the pituitary stalk and generally are located posterior to the chiasm extending into the third ventricle. They can cause hydrocephalus, visual loss, pituitary dysfunction, and diabetes insipidus (DI). The extended transplanum approach can provide an ideal corridor between the optic chiasm and the normal pituitary gland for their removal (Fig. 5.5). Although some authors have advocated subtotal resection followed by radiation due to

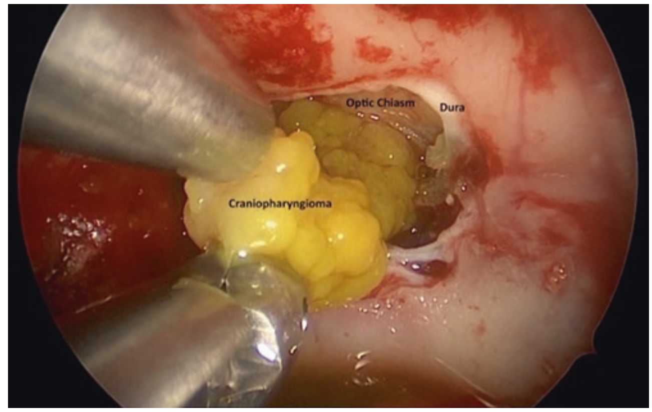

Fig. 5.5 Removal of a craniopharyngioma; please note that the size of the mass removed may be larger than the opening provided by the optic chiasm and the pituitary gland. Optic chiasm tolerates slight upward manipulation for the dissection and delivery of the tumor

high likelihood of causing deficits, gross-total resection offers a reasonably high likelihood of cure, particularly if performed transsphenoidally [20, 30, 31]. The transplanum, transtuberculum approach does place the stalk at high risk of injury due to the anatomical nature of the approach, with a potential of postoperative DI and hypopituitarism, but transcranial approaches equally place pituitary function at risk of injury, with rates of hypopituitarism reaching 70–80% [32, 33]. In addition, it is possible to minimize injury to the pituitary gland and stalk either by mobilizing or partially resecting the pituitary gland to enlarge the surgical corridor [9]. Another advantage of the endoscopic transplanum, transtuberculum approach is the unobstructed view of the entire third ventricle from below, which cannot be obtained with a transcranial/microscope-based approach, increasing the chances of gross-total resection with less morbidity with EEA [9] (Fig. 5.6). The risk of hyperphagia and hypothalamic injury with gross-total resection is largely dependent on the extent of tumor invasion, which is unchanged by the method of approach or visualization. Therefore, the risks of hyperphagia after gross-total resection are likely to be roughly equivalent after endoscopic transsphenoidal as after microscope-based transcranial approaches. If the goal of surgery is partial resection and cyst decompression, this can also be easily achieved with an endoscopic, transsphenoidal, transtuberculum approach [20].

Meningiomas

Meningiomas of the planum sphenoidale and tuberculum sella and even some carefully selected small olfactory groove meningiomas are amenable to the endoscopic, endonasal transtuberculum, transplanum approach for resection. Tumors that extend lateral to the carotid may not be suitable

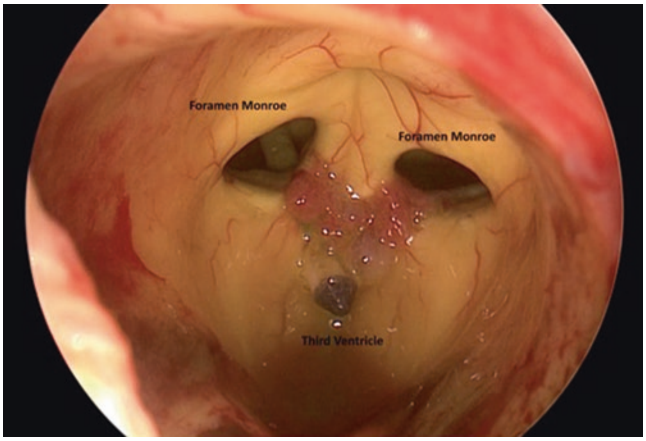

Fig. 5.6 A 45° endoscope provides a detailed view of the third ventricle below. Foramen of Monro (FM) visible bilaterally. Please note the ventriculostomy catheter visible in the lateral ventricle on the right through the FM, and it penetrates into the third ventricle close to midline. For patients with hydrocephalus, a ventriculostomy is preferred over lumbar drain

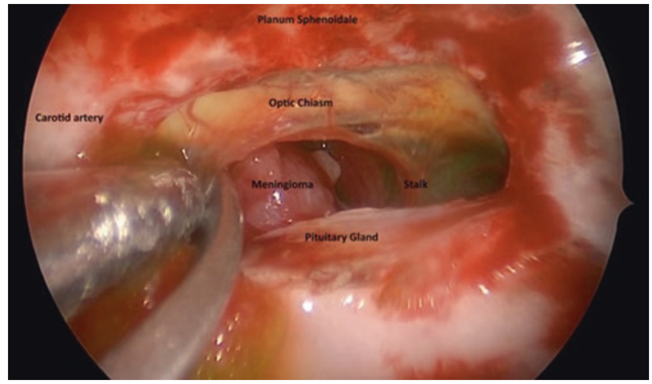

Fig. 5.7 After the craniectomy, and debulking of the tumor, optic chiasm and the pituitary gland are well exposed; meningioma is visible on the right and stalk on the left

to EES. Meningiomas of the planum sphenoidale and tuberculum sella are mostly prechiasmal, and the most common presentation is visual loss. Subfrontal and pterional transcranial approaches do not only require brain retraction but also place optic nerves and the carotid arteries between the tumor and the surgeon [34, 35]. On the contrary, the EEA permits the surgeon to be able to immediately internally debulk the tumor before encountering any of the critical neurovascular structures [9, 20]. Although gross-total resection is possible with this approach and is generally the goal, the transtuberculum, transplanum approach also makes possible a less morbid decompression of the optic apparatus in frail or elderly patients who are losing vision in preparation for post resection radiation or observation. Another advantage of approaching these tumors from below is the ability to interrupt the dural vascular supply to the tumor early in the operation. This option is particularly important if preoperative embolization is not possible. In addition, the transtuberculum, transplanum approach potentially offers a higher likelihood of curing these meningiomas. Not only the dura, but also the bone at the base of the tumor, is removed during the approach. These are precisely the areas that may be infiltrated by tumor that are most difficult to remove from a transcranial route. In addition, wide opening of the optic canals bilaterally can ensure removal of tumor remnants that lie below the optic nerve, which would be very difficult to remove with a transcranial approach [9] (Fig. 5.7).

Potential disadvantages of endonasal resection of meningiomas include a narrow working space and reduced degrees of freedom with the dissecting instruments. Additionally, the entire dural attachment and dural tail may not be completely exposed and removed. However, with increasing understanding of the adjacent anatomy from the endoscopic perspective and with angled instruments, appropriate access corridors can be established enabling bimanual, microscope-like dissection around the indwelling endoscope. Careful patient selection and knowledge of the limitations of the endonasal corridor can reduce the incidence of residual tumor. In addition, operating through the nose carries with it the theoretical risk of intracranial infection and CSF leak, given the challenge of reconstructing the dura and skull base from below. Somewhat surprisingly, however, the rate of postoperative infection with EEA is extremely low. Surgeon familiarity with endoscopic techniques greatly affects operative time, which can significantly impact patient outcomes [19, 36].

Rathke's Cleft Cysts

Rathke's cleft cysts (RCCs) are benign congenital lesions believed to derive from remnants of Rathke's pouch. Rathke's cleft is the residual lumen between the anterior and intermediate lobe constitutes found after the formation of the pituitary gland. Further enlargement of Rathke's cleft with proliferation of the cells lining it and accumulation of its secretions leads to RCCs [37].

These cysts can be encountered in 12–33% of normal pituitary glands in routine autopsies [38]. These cysts generally are sellar, although in one-third of cases, they can have a suprasellar component. Pure suprasellar lesions are rare at the time of diagnosis. They present with headaches, visual impairment, and endocrine dysfunction. On MRI, enhancement of the pituitary around the cyst wall may be observed leading to the classical aspect of "the egg in the cup" sign [39]. The treatment of Rathke's cleft cysts mainly is surgical. A transsphenoidal approach, together with the drainage of the cyst accompanied by a biopsy of the cyst wall, usually is the preferred treatment to avoid any postoperative hypopituitarism or DI [38, 40].

Chordomas

Chordomas are rare (0.1% of skull base tumors) neoplasms originating from the notochord cells [37, 41]. Clinical presentation depends on the size and extension of the lesion, but the most common presenting symptoms are visual complaints, mainly diplopia. Intermittent or partial abducens nerve palsy is a common initial symptom of clival chordomas, related either to posterior cavernous sinus extension or invasion into Dorello's canal. Chordomas tend to recur and metastasize, and their long-term prognosis is poor. Radical resection and aggressive adjuvant radiation therapy is the recommended treatment for these lesions [37, 41, 42]. EEA is becoming the gold standard to resect skull base chordomas [37, 42–45]. Isolated suprasellar chordomas are very rare; instead, they usually arise in the clivus and may have significant suprasellar extension which may require a transplanum approach to supplement the transclival approach to resect them [16].

5.6 Postoperative Care

Once the surgery is complete, extubation must be smooth to avoid dislodgment of the closure. During closure we routinely utilize CSF drainage to keep ICP low. CSF drainage via the lumbar drain is continued for 24 h for with a rate of 5 cc/hour. Patients are kept under surveillance for postoperative diabetes insipidus (DI). Patients may be administered a small dose of glucocorticoids postoperatively. If there is clearly damage to the pituitary-hypothalamic axis or known preoperative hypocortisolism, glucocorticoids are continued for the patient's hospital stay and are tapered by the endocrinologist after discharge. Otherwise, a fasting morning cortisol level is obtained on the morning of the second postoperative day, and cortisol replacement is initiated only if the level is abnormally low. Infectious complications are rare, especially in the absence of a postoperative CSF leak. Antibiotics are delivered in the perioperative for 24–48 h. A postoperative MRI scan usually is obtained on the second postoperative day and then 3 months after surgery [9, 19].

5.7 Surgical Pearls

- Early visual identification of the pituitary and stalk is important. Meticulous surgical dissection to preserve the pituitary stalk and supporting vasculature is important to prevent postoperative diabetes insipidus. Devascularization of the stalk can cause hypopituitarism and the superior hypophyseal arteries should be identified and preserved if possible. Branches to the chiasm should also be preserved if possible to avoid visual deterioration although collaterals from above exist.
- For EEA, it is important not to make the bone opening too small. Once the surgeon is confident in their ability to close large skull base defects, larger bone openings are possible. Navigation is useful to determine the required size of the opening to expose the extent of the tumor. If the bone opening is too small, the surgeon is forced to pull the pathology into the field of view which risks injury to vessels attached to the back of the tumor.
- For very large tumors and for extended approaches, the middle turbinate may be removed to provide additional visualization. Frequent internal decompression and extracapsular dissection permits the surgeon to avoid blindly pulling the tumor into the surgical field [16].

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6 Anterior Cranial Base

Luigi Maria Cavallo, Domenico Solari, Alberto Di Somma, Waleed A. Azab, and Paolo Cappabianca

6.1 Introduction

Anterior cranial base lesions have been traditionally approached through classical frontal or fronto-lateral approaches [1], although several variations of these approaches have been defined in an attempt to limit brain retraction and approach-related morbidity. Craniotomies for midline anterior cranial base pathologies, moreover, carry a concrete risk of damaging the optic apparatus, mainly due to the intimate association of these tumors with the optic nerves and chiasm.

Recently, expanded endonasal approaches have been developed in an attempt of reducing brain retraction as well as improving ophthalmological outcomes by approaching the tumors from an anteromedial and inferior trajectory [2–11].

The versatility of endoscopic endonasal surgery for managing anterior cranial base lesions has been increasingly recognized over the past decade. Despite initial discussion about the possibility of accessing the anterior skull base and eventually managing lesions involving this area via the nose, the pure endoscopic endonasal approach extended to the suprasellar area and anterior cranial fossa has proved to be both effective and safe [12].

Nowadays, different pathologies involving the anterior cranial base can be managed via the endonasal route, with this approach considered the favorite pathway for treating cerebrospinal fluid leaks [13] and, among the oncological diseases, the esthesioneuroblastoma (olfactory neuroblastoma) [14]. Furthermore, the endoscopic endonasal route can be effective for the management of idiopathic or secondary meningoencephaloceles, especially for the latter when a wide osteo-dural skull base breach has been created during surgery [15].

However, it has to be stressed that meningiomas are the most common pathologies located in the anterior cranial fossa and the endonasal route can be considered for the management of certain cases of such lesions [16–22]. This chapter will focus on the endoscopic endonasal approach for the management of tuberculum sellae and planum sphenoidale meningiomas.

6.2 Rationale

Anterior skull base meningiomas can be approached through transcranial (coronal subfrontal, oblique subfrontal, interhemispheric, or classic pterional transsylvian approaches) or extended endonasal approaches, either with the microscopic or endoscopic.

With the concept of minimally invasive neurosurgery and development of modern neuroimaging techniques, neurosurgeons gained the possibility of a proper characterization of brain tumors and their relation to surrounding structures. The introduction of the endoscope into the realm of neurosurgery led to successful management of deep-seated lesions without brain retraction with access to the anterior skull base gained via a ventral route from below using the endonasal approach. This pathway for the treatment of anterior cranial fossa meningiomas has been increasingly reported during the last decade, with several contributions from different groups [22–26].

Tuberculum sellae meningiomas present a surgical excision challenge, in part because of the adjacent critical structures involved by the tumor including the optic nerves, carotid and anterior cerebral arteries, pituitary gland and stalk, and the hypothalamus. An endoscopic endonasal approach theoretically avoids manipulation by entering the meningioma directly through its dural base, thereby minimizing retraction

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of the optic nerves and decreasing the risk for postoperative visual deterioration. However, this route is burdened by a higher rate of CSF leakage. Modern closure techniques, when performed by experienced hands, result in very low rates of CSF leakage, and recent data suggest that the endoscopic endonasal approach might be optimal for a select group of lesions and could further expand its applications in the near future [27, 28].

In its general principles, the ideal surgical approach should provide enough exposure of the tumor, including its dural attachment, to interrupt its blood supply as early as possible in the procedure. In addition, brain retraction and manipulation of critical neurovascular structures should be minimized in order to avoid procedure-related morbidity.

The selection of the most appropriate approach depends on multiple factors, including surgeon preference and experience, tumor size and location, extent of dural attachment, and relationship with the surrounding neurovascular structures.

6.3 Patient Selection

Given the narrow corridor provided by the endonasal pathway, patient selection is of utmost importance. In the sagittal plane, endoscopic endonasal surgery can reach from the frontal sinus down to the craniocervical junction, whereas anatomical limitations could hinder this route moving off the midline.

Indications and Contraindications with Anatomical Limitations of Approach

There are no absolute limitations in terms of lesion size for endoscopic endonasal surgery. The lateral limits of the corridor are formed within the nasal cavities by the lamina papyracea and within the sphenoid sinus inferiorly at the level of the sella by the carotid siphons in the cavernous sinus and more superiorly by the lateral opticocarotid recesses and carotid bifurcation, although tumors extending slightly beyond these limits can be rolled toward the center after adequate internal debulking (Fig. 6.1a–d).

Key considerations for endoscopic endonasal surgery of anterior cranial base meningiomas include sphenoid sinus and tuberculum/suprasellar notch anatomy [29], relative position of the optic chiasm and pituitary stalk, parasellar extension of the tumor or encasement of neurovascular structures, and degree of tumor calcification. Although involvement of the optic canals was initially thought to be a contraindication for endoscopic endonasal surgery, adequate and safe exploration and resection of intracanalicular meningiomas has been demonstrated [30]. Indeed, exploration and graded opening of the medial optic canal is mandatory for ensuring complete tumor removal when canal involvement is demonstrated on preoperative high-resolution MR images (Fig. 6.2a–d).

Debate exists over the need for a cortical cuff around the tumor, defined as brain tissue separating the tumor capsule from vessels. The presence of a cortical cuff certainly facilitates tumor dissection, but its presence is not mandatory, and careful extracapsular microdissection of vessels off the tumor capsule is possible. Generally, the most amenable lesions are those that are smaller and mostly midline, maintain their arachnoid plane as evidenced by a lack of edema in the brain, do not encase neurovascular structures, and have a well-aerated sphenoid sinus for access and closure.

Table 6.1 presents a list of potential pathologies that can be addressed via extended endonasal approach.

6.4 Surgical Anatomy

Once the endoscope is inserted into the nasal cavity, the first two anatomic landmarks that should be identified are the nasal septum and the lateral wall of the nasal cavity that is lined by three nasal turbinates. Moving posteriorly, between the tail of the inferior turbinate and the nasal septum, it is possible to see the choana. The choana represents the most important landmark during the nasal phase of the surgical procedure, as it drives the approach toward the anterior sphenoidal wall. The subsequent landmark that should be exposed is the head of the middle turbinate. Its tail usually lies at the level of the sphenopalatine foramen through which the sphenopalatine artery enters into the nasal cavity; thereafter, it divides in two branches, the nasopalatine artery medially and the posterior nasal artery laterally. The tail of the middle turbinate represents an important landmark for the control of eventual arterial bleeding during the anterior sphenoidotomy. Moreover, whether or not a pedicled nasoseptal flap will be used for reconstruction of the anterior cranial base, its anatomy has to be understood. The flap consists of the mucoperiosteum and mucoperichondrium of the nasal septum and it is pedicled on the posterior septal artery, a branch of the sphenopalatine artery. The posterior septal artery arises from the sphenopalatine artery, a branch of the internal maxillary artery, and bifurcates into a superior and inferior branch with the latter being the larger one. The branches of the posterior septal artery then form a dense network along the septum to supply the inferior two thirds of the septum and a large portion of the nasal floor. Posterior, superior, and medial to the middle turbinate, it is possible to identify the superior turbinate.

Above the choana, the sphenoethmoid recess forms the posterior wall of the nasal cavity. In its upper portion, the sphenoid ostium can be identified, variable in shape, dimen-

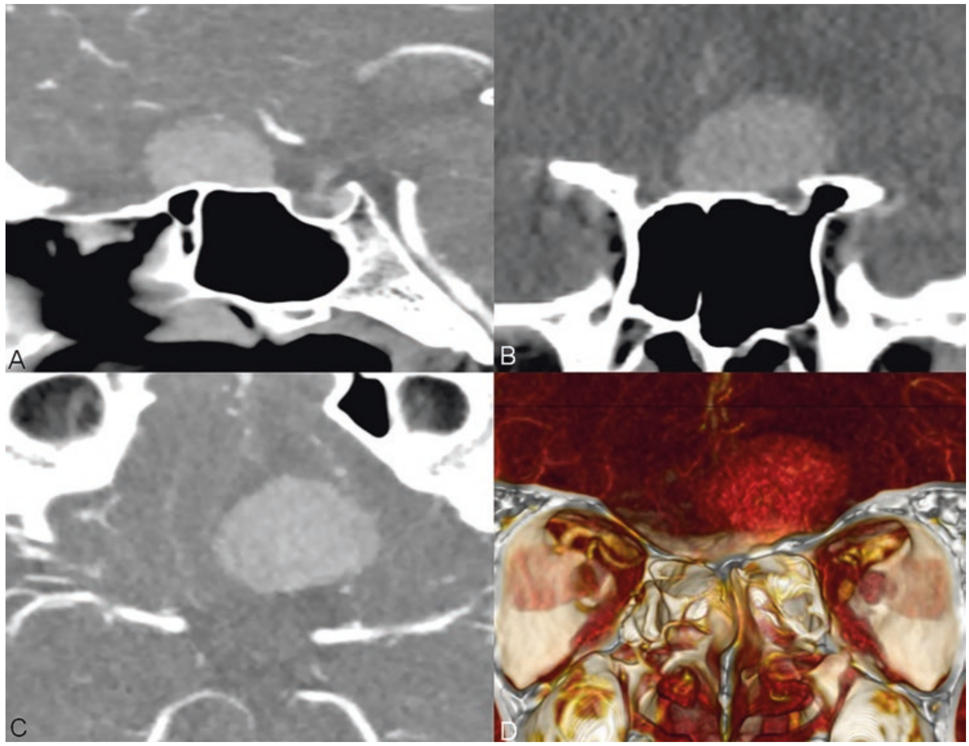

Fig. 6.1 (**a**–**d**) Sagittal (**a**), coronal (**b**), and axial (**c**) contrast-enhanced CT scans with a 3D volume-rendered reconstruction (**d**) showing a planum sphenoidale meningioma extending laterally to the left side

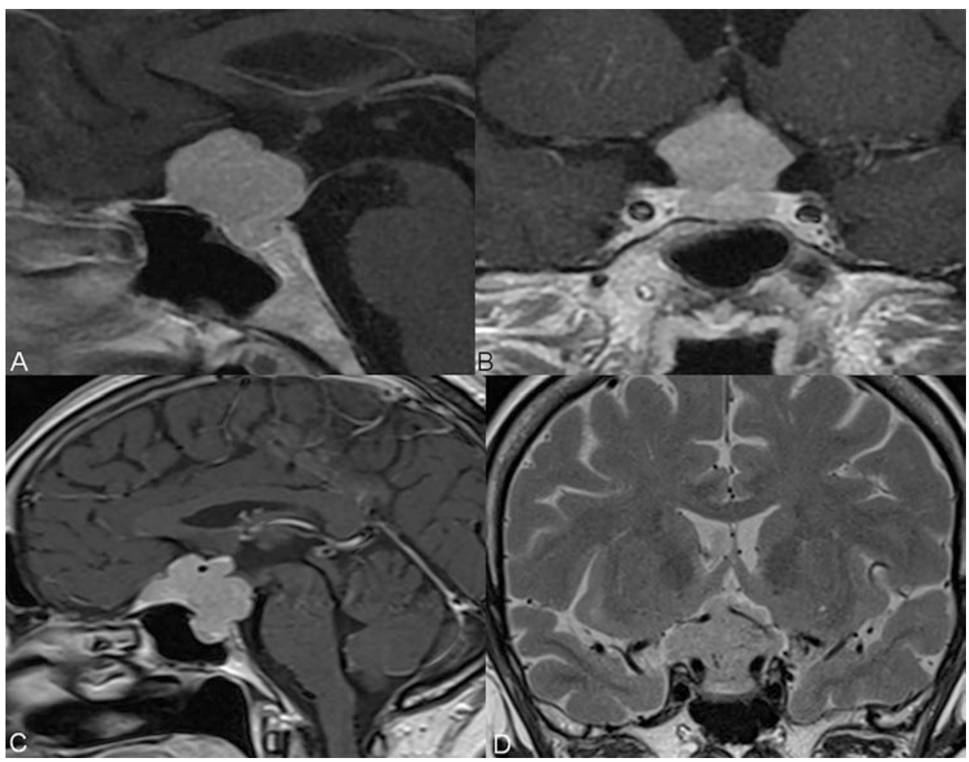

Fig. 6.2 (**a**–**d**) Sagittal (**a**) and coronal (**b**) T1-weighted MRI scans showing a tuberculum sellae meningioma displacing the optic apparatus and pituitary stalk posteriorly. Sagittal (**c**) T1-weighted and coronal (**d**) T2-weighted MRI showing a tuberculum sellae meningioma encasing the anterior cerebral artery complex

sion, and location, being sometimes covered by the tail of the superior turbinate. It represents the natural communication between the nasal cavity and the sphenoid sinus.

Hence, at the level of the sphenoethmoid recess, the nasal septum is detached from the prow of the anterior wall of the sphenoid bone which is then opened circumferentially. Opening of the anterior wall allows the endoscope to enter into the sphenoid cavity which is often divided by one or more septa.

The degree of pneumatization of the sphenoid bone is an important factor for the identification of the bony protuberances and depressions inside. As well, the configuration of

Table 6.1 Potential pathologies that can be addressed via extended endonasal approach

	Potential pathologies
Primary brain tumors	Meningiomas
	Craniopharyngiomas
	Pituitary adenomas
Others	CSF fistula of the anterior cranial base
	Meningoencephaloceles
	Olfactory neuroblastomas
	Metastasis
	Malignant cranial base lesions

the sphenoid sinus has to be addressed properly. The sphenoid sinus could present extreme variability in terms of size, shape, and, above all, degree of pneumatization. Depending on the degree of its pneumatization, a series of protuberances and depressions molded on its posterior and lateral walls can be identified. The sellar floor is at the center, the sphenoid planum above, and the clival indentation below. Lateral to the clivus and sellar floor, the bony prominences of the intracavernous carotid arteries (paraclival and parasellar ICA) and optic nerves can be observed. Between them, the lateral opticocarotid recess lies, molded by the pneumatization of the optic strut of the anterior clinoid process. The intracranial aspect of the upper border of the lateral opticocarotid recess is covered by a thickening of the dura and periosteum that forms the distal dural ring, which separates the optic nerve from the clinoidal segment of the internal carotid artery (ICA). The inferior border of the lateral opticocarotid recess also presents a thickening of the dura and periosteum, which forms the proximal dural ring, separating the intracavernous portion of the carotid artery from the clinoidal segment. The lateral aspect of the tuberculum sellae represents the point where the bony prominences of the carotid artery and optic nerve join medially (medial opticocarotid recess). This recess is less evident than the lateral recess but represents the lateral limit of bony exposure to unlock the suprasellar area.

The sella turcica is limited superiorly by the diaphragma sellae, a fold of dura with a central opening which is pierced by the pituitary stalk and its blood supply. The diaphragma sellae separates the anterior lobe of the pituitary gland from the optic chiasm and the suprasellar cistern. Once the bone of the sella and the planum sphenoidale has been removed, venous sinuses that interconnect the cavernous sinuses appear. The intercavernous connections are named on the basis of their relationship to the pituitary gland – the anterior, or superior, intercavernous sinus passes anterior to the hypophysis, while the posterior, or inferior, intercavernous sinus passes behind the gland. However, these intercavernous connections can run at any site along the anterior, inferior, or posterior surface of the gland or may be absent. There is also a large intercavernous venous connection (i.e., the basilar sinus) that passes posterior to the dorsum sellae and upper clivus and connects the inferior and posterior aspects of both cavernous sinuses.

Above the sella, the angle formed by the convergence of the sphenoid planum with the sellar floor corresponds to the tuberculum sellae, recently renamed in accord with the endonasal visualization, the "suprasellar notch" (SSN) meaning "angular or V-shaped cut indentation" [29]. Moving anteriorly, we can recognize the sphenoid planum, laterally delimited by the protuberances of the optic nerves. At this point, the bone of the suprasellar notch and the planum sphenoidale can be removed 1.5–2 cm in a posteroanterior direction and laterally up to the optic protuberances. The sellar and suprasellar dura are then opened to permit the exploration of the neurovascular structures localized above the diaphragma sellae.

From the endoscopic endonasal perspective, the suprasellar area can be divided into four areas by two ideal planes, one passing through the inferior surface of the chiasm and the mammillary bodies and another passing through the posterior margin of the chiasm and dorsum sellae. These two lines define four regions – the suprachiasmatic, subchiasmatic, retrosellar, and ventricular regions [31]. For the interest of the approach, we will consider in detail only the supra- and the infrachiasmatic region anatomy.

In the *suprachiasmatic region*, the chiasmatic and lamina terminalis cisterns with related contents are accessible. The anterior margin of the chiasm and medial portion of the optic nerves along with the anterior cerebral arteries, anterior communicating artery, and recurrent Heubner arteries, and gyri recti of the frontal lobes are identified.

The anatomical variability of the optic chiasm as related to the anterior skull base and surrounding structures should be highlighted. Several anatomical studies have demonstrated in about 80% of patients, the optic chiasm overlies the diaphragm sellae, so defined as normal positioning. The remaining 20% were equally distributed between the prefixed variant (chiasm located above the tuberculum sellae) and the postfixed variant (chiasm located over the dorsum sellae).

In the *subchiasmatic space*, the pituitary stalk is at the center of the view below the chiasm, with the superior hypophyseal arteries and its perforating branches supplying the inferior surface of the chiasm and optic nerves. The superior aspect of the pituitary gland and dorsum sellae is also visible. The superior hypophyseal arteries supply the optic chiasm, floor of the hypothalamus, and median eminence. Inferiorly, the inferior hypophyseal artery divides into medial and lateral branches which anastomose with the corresponding vessels of the opposite side forming an arterial ring around the hypophysis (Fig. 6.3a–d).

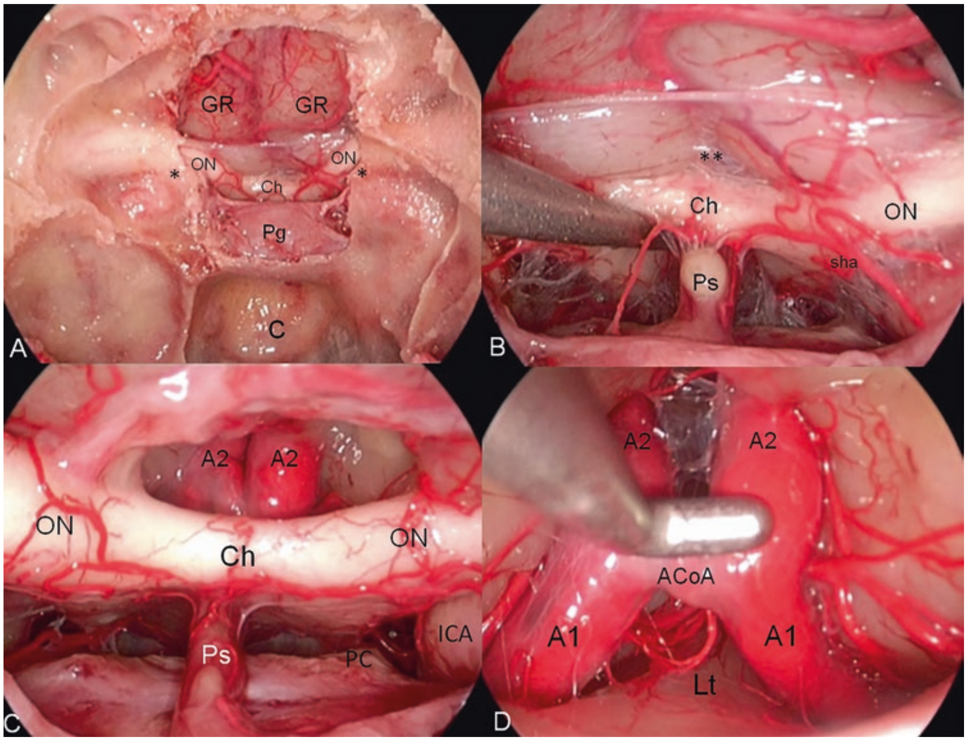

Fig. 6.3 (**a**–**d**) Endoscopic endonasal anatomic picture showing the transtuberculum-transplanum approach; the medial opticocarotid recess has been used as the most lateral limit of such route (**a**). Closeup view of the subchiasmatic area (**b**) and opening of the lamina terminalis cistern (**c**) with visualization of the main neurovascular structures of the suprachiasmatic region (**d**). *A1* pre-communicating segment of the anterior cerebral artery, *A2* post-communicating segment of the anterior cerebral artery, *ACoA* anterior communicating artery, *C* clivus, *Ch* optic chiasm, *GR* gyrus rectus, *ICA* internal carotid artery, *ON* optic nerve, *Pg* pituitary gland, *Ps* pituitary stalk, *sha* superior hypophyseal artery, * medial opticocarotid recess, ** lamina terminalis cistern, *PC* posterior clinoid process, *Lt* lamina terminalis

6.5 Surgical Technique with Nuances in Nasal Approach Including Management of Turbinates, Nasal Septum, and Reconstruction

Operative Setup

An integrated operating room helps to optimize teamwork and improve patient care. In the operating room, all of the equipment (i.e., cold light source, video camera, monitor, and video recording system) are placed ergonomically behind the head of the patient and in front of the first surgeon, who is at the right side of the patient. The anesthesiologist is positioned with his/her equipment at the left side of the patient at the level of the head. The second surgeon is at the left side of the patient, and the scrub nurse is positioned at the level of the patient's legs.

Instrumentation

In our institution, during a fully endoscopic endonasal transsphenoidal approach, rigid diagnostic Hopkins® telescopes (Karl Storz, Tuttlingen, Germany) – *without* a working channel – are used. There are three types of telescopes available that vary in length, diameter, and direction of view: 0° and 30° telescopes, length 18 cm, diameter 4 mm; 0° and 30° telescopes, length 18 cm, diameter 2.7 mm; and 0° and 30° telescopes, length 30 cm, diameter 4 mm. The 0° scope, length 18 cm, diameter 4 mm, is the one most frequently used. The endoscope is connected to the Karl Storz images spies (Storz Professional Image Enhancement System, Tuttlingen, Germany) camera platform.

In view of the fact that the scope is mainly an optical device, it is usually not equipped with an operating channel. Accordingly, the surgical instruments are introduced alongside the scope.

A special outer sheath and irrigation system (Clearvision® II, Karl Storz, Tuttlingen, Germany) is used to rinse the distal objective lens, obviating the need for repeated withdrawal and reinsertion of the scope into the nasal cavity during surgery.

The most commonly used 0° scope (diameter 4 mm, length 18 cm) is usually *operated freehand throughout the entire surgical procedure.* During the sellar step of the procedure, the endoscope is held dynamically by a second surgeon, allowing the first surgeon to work bimanually with two instruments. 30° or 45° telescopes are used in selected cases or in specific phases of the surgical procedure, e.g., the exploration of the sellar cavity after tumor removal.

Two or three operating instruments – depending on the specific needs and circumstances – plus the endoscope can be inserted through both nostrils, thus providing increased working space and improved maneuverability.

The use of neuronavigation during a standard endoscopic approach is currently reserved for selected cases only, e.g., in the presence of a conchal-type sphenoid sinus, certain cases of recurrences with a previous history of transsphenoidal surgery, and patients with large lesions involving the para- and suprasellar areas. Furthermore, the use of a micro-Doppler probe can be useful to localize the course of the ICA during removal of pituitary adenomas with lateral extension into the cavernous sinus.

6.6 Technique

The details of the approach are well described and will be only briefly reviewed here to emphasize variations of the technique. The patient is positioned supine, either in pin fixation or a horseshoe headrest, depending on whether cranial manipulation is desired. After induction of general anesthesia, if required, frameless neuronavigation is registered by using CT and/or MRI. Routine nasal mucosal preparation is used and a suitable donor site (fascia lata and/or periumbilical fat) is prepared.

As a general rule, the neurosurgeon operates bimanually through both nares in a classical microsurgical attitude, while an assistant provides dynamic endoscopic visualization using a combination of 0° and 30° endoscopes.

The harvesting of a Hadad-Bassagasteguy vascularized nasoseptal flap is usually performed during the nasal step of the procedure. We prefer to just define the incision and then raise the flap at the end of the procedure.

For extended procedures to the anterior cranial base, the middle turbinate of one side should be removed to gain more space in the nasal cavity. A wide sphenoidotomy is performed with complete removal of the rostrum and flattening of the floor of the sphenoid to aid the placement of the nasoseptal flap. Bilateral posterior ethmoidectomies are essential for providing a clear view of the anterior extent of the planum sphenoidale. A high-speed drill with a round diamond bit is used to remove the bone over the anterosuperior sella just below the superior intercavernous sinus, extending up to the tuberculum, lateral to the medial opticocarotid recesses, and forward along the planum to the anterior extent of the tumor and its dural tail.

Therefore, bone removal over the uninvolved pituitary gland is minimized unless the tumor invades into the sella. If optic canal invasion has been identified on preoperative imaging, the medial optic canals should be drilled away to reach the anterior extent of the tumor within the canals.

In cases of tuberculum sellae and planum sphenoidale meningiomas, the coagulation of the dural attachment is achieved as the first maneuver, so that early tumor devascularization can be obtained (Fig. 6.4a–d). The tumor is therefore debulked safely and its capsule finally dissected from the surrounding microvascular structures via an extra-arachnoidal route (Figs. 6.5a, b and 6.6a–d). In this particular case, the main advantages of the endoscopic endonasal technique come from the early tumor devascularization and dissection of the tumor with minimal manipulation of the optic pathways.

In cases of planum sphenoidale meningiomas, the bone opening has to be extended more anteriorly and laterally above the orbital roof and should include also the isolation of the posterior ethmoidal artery in order to identify the cleavage plane between the lesion and the brain (Fig. 6.4c, d).

For olfactory groove meningiomas, bone removal and exposure of the target area can be tailored to each case according to lesion extension. In cases of olfactory groove meningiomas, middle turbinectomy is performed bilaterally followed by a radical anterior and posterior ethmoidectomy. The superior half of the nasal septum is also removed with extension anteriorly to identify the posterior border of the frontal sinus. The bone of the anterior skull base enclosed between the two orbits is removed, thus creating a wide surgical corridor which can be extended laterally between the two medial orbital walls and anteroposteriorly from the frontal sinus to the sella according to tumor extension. Once the dura has been opened, the lesion can be removed following the steps described previously. The endoscopic approach again allows coagulation of the dural attachment and early devascularization of the tumor.

During an extended transsphenoidal approach, especially to the suprasellar area, a large osteo-dural opening has to be created and the cisternal space is often widely dissected. A conspicuous intraoperative CSF leak should therefore be anticipated. An effective watertight closure, however, is mandatory to prevent postoperative CSF leakage.

We prefer the so-called sandwich technique in which the cistern is covered with a layer of collagen sponge coated with fibrinogen and thrombin and the surgical cavity is filled with fat graft sutured to the inner layer of three layers of fascia lata or dural substitute. The first layer is then positioned intradurally, the second between the dura and the bone, and the third is applied to cover the bone. In order to support the materials used for reconstruction at the level of the skull base defect, a vascular flap of septal mucosa is created by cutting the septal mucosa along the inferior edge of the septum from the choana to the cartilage portion of the septum and superiorly to the level of the rostral portion of the middle turbinate [32, 33].

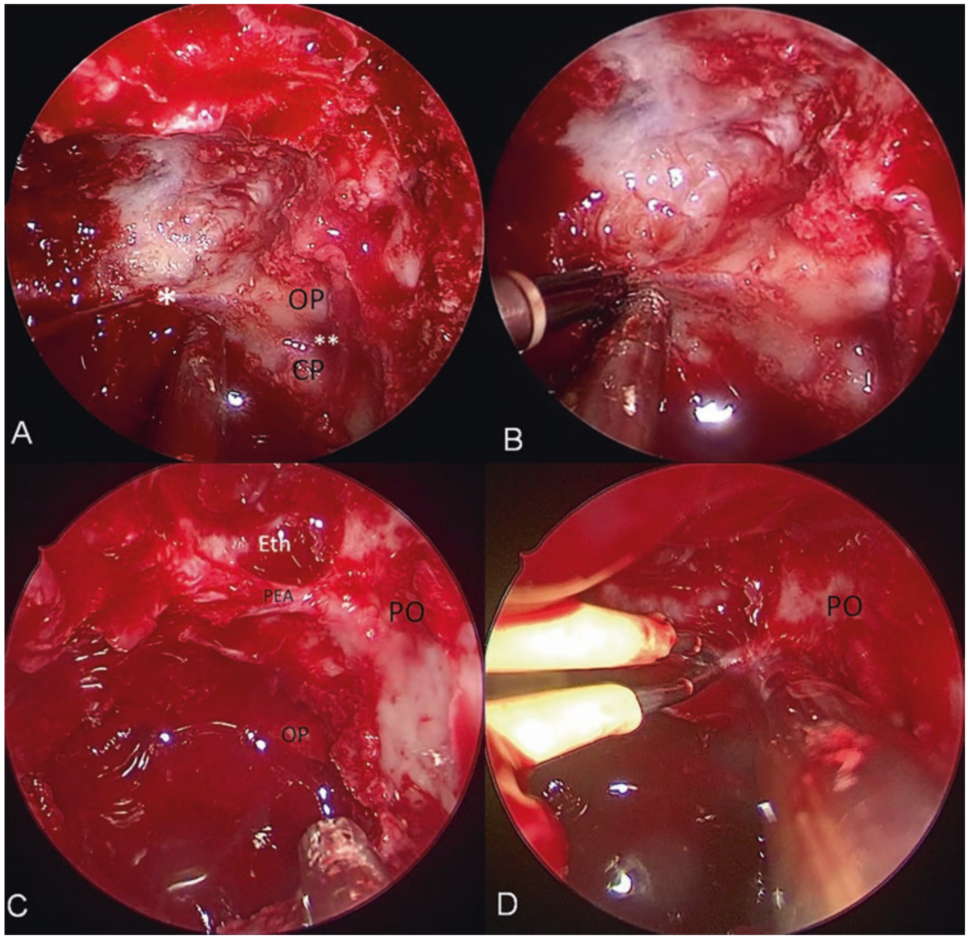

Fig. 6.4 (**a**–**d**) Endoscopic endonasal transtuberculum-transplanum approach for anterior cranial base meningiomas. A feeding artery of the tumor dural attachment is coagulated before opening the dura. Exposition of the periorbita and posterior ethmoidal artery which is coagulated before debulking the tumor. * bleeding from a feeding artery of the tumor, ** medial opticocarotid recess, *CP* carotid protuberance, *Eth* ethmoid cell, *OP* optic protuberance, *PEA* posterior ethmoidal artery, *PO* periorbita

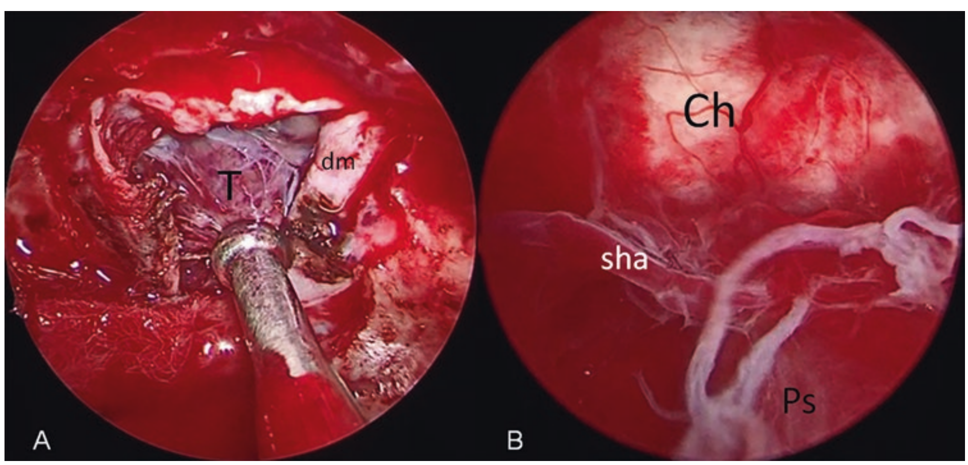

Fig. 6.5 (**a**, **b**) Endoscopic endonasal approach to a tuberculum sellae meningioma showing the dural involvement (**a**) and a closeup view of the subchiasmatic space obtained with diving technique (**b**). Once the dura has been opened, the tumor is visualized. *Ch* optic chiasm, *dm* dura mater, *Ps* pituitary stalk, *sha* superior hypophyseal artery, *T* tumor

Following mucosal dissection from the septal bone, the flap is pedicled laterally around the sphenopalatine foramen and positioned in the choana during the operation. At the end of the procedure, the flap is used to cover the posterior wall of the sphenoid sinus. An inflated Foley balloon catheter, filled with 7–8 ml of saline solution, could be placed in the sphenoid sinus to support the reconstruction (Fig. 6.7a–c).

6.7 Postoperative Care/Complications

Lesions treated via an endoscopic endonasal approach require intensive and watchful postoperative care. New-onset diabetes insipidus (DI) should be diagnosed promptly and treated with desmopressin (DDAVP).

The patient should rest in bed with the head 30° raised in order to facilitate venous return and CSF flow toward the

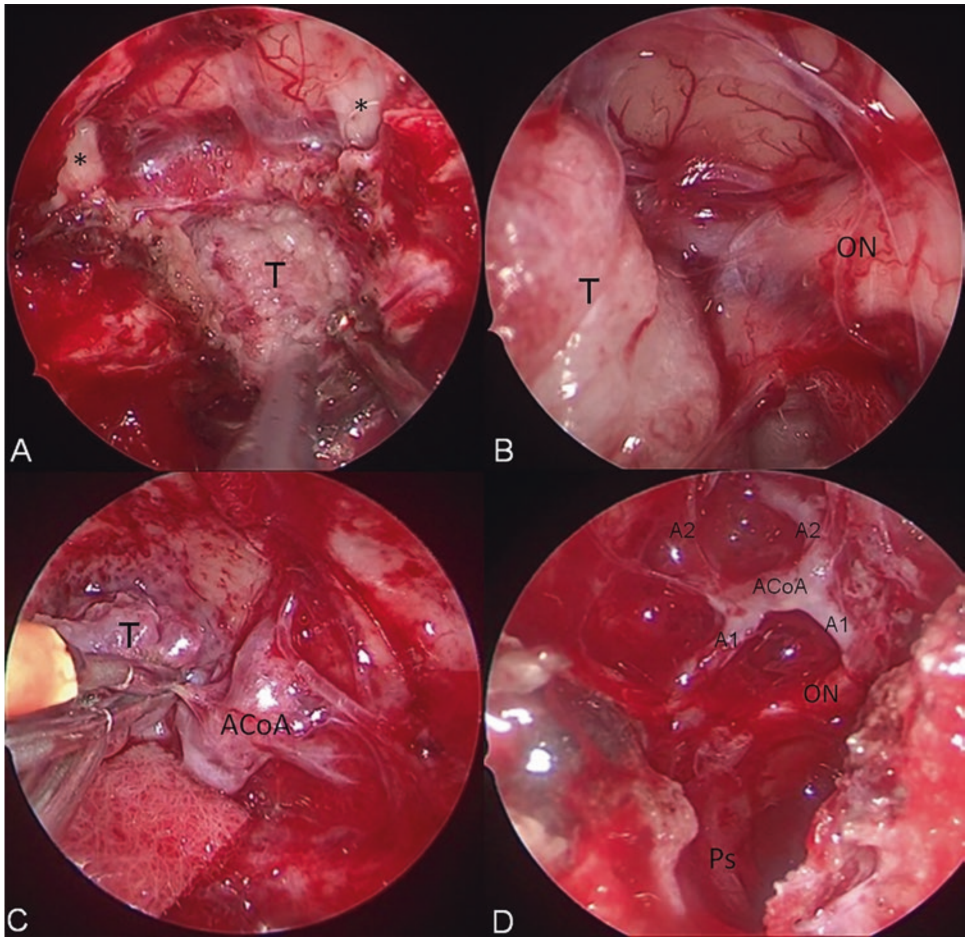

Fig. 6.6 (**a**–**d**) Intraoperative endoscopic pictures showing details of the tumor dissection from the main neurovascular structures. The tumor is debulked with the aid of ultrasound aspiration (**a**). Closeup view of the dissection from the left optic nerve (**b**) and anterior cerebral artery complex (**c**). Panoramic view after tumor removal (**d**). * olfactory tract, *A1* pre-communicating segment of the anterior cerebral artery, *A2* post-communicating segment of the anterior cerebral artery, *ACoA* anterior communicating artery, *ON* optic nerve, *Ps* pituitary stalk, *T* tumor

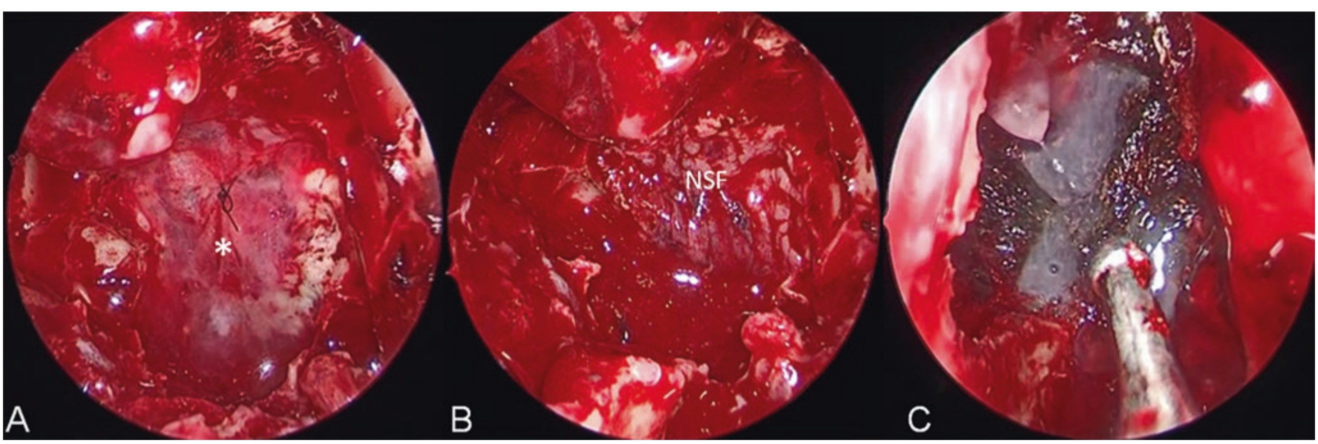

Fig. 6.7 (**a**–**c**) Reconstruction of the skull base defect obtained with different materials. The skull base defect is initially closed with dural substitute and fibrin glue (**a**). The naso-septal flap is used to cover the osteo-dural defect (**b**), and it is held in place by Surgicel® and fibrin glue (**c**). *NSF* naso-septal flap, * "sandwich technique" used to cover the skull base defect

spine. Additionally, patients are asked to adopt postoperative habits in order to prevent any ICP increase and the eventual displacement of the skull base reconstruction. It is preferable to cough and/or sneeze with the mouth open, assume as early as possible a stand-up position, and start walking. Bending and squatting should be avoided and stool softeners should be prescribed. Computed tomography (CT) scan is performed routinely at postoperative day 1 in order to evaluate any neurosurgical complications and assess the amount of pneumocephalus. According to a recent contribution, frontal and intraventricular pneumocephalus is not necessarily associated with a postoperative CSF leak; however, a "suspicious" pattern of air, namely, pneumocephalus in the convexity, interhemispheric fissure, and sella, parasellar, or perimesencephalic locations, may be significantly associated with a postoperative CSF leak occurrence, and for such reason these patients require closer observation [34].

Based on our experience, we noticed that the sudden onset of elevated patient temperature starting from postoperative day 2 could be suggestive of a not yet visible leakage. The endoscopic endonasal inspection of the surgical site can be easily performed at patient's bed with the portable Tele Pack X Led (Karl Storz, Tuttlingen, Germany) to assess the status of the surgical wound and resiliency of the reconstruction.

In these terms, patients with minimal postoperative CSF leak can be managed without reoperation. If necessary, repeated endoscope-guided fibrin glue injections at the bedside or in the outpatient ward, i.e., according to the so-called "awake sealant technique," can be performed [35].

However, in cases of severe CSF leak, displacement of the reconstruction materials, and/or evident communication of the sphenoid sinus with the intradural compartment, immediate transsphenoidal reoperation is needed.

6.8 Surgical Pearls

- Large opening of the planum sphenoidale extending above the optic prominence is essential to properly manage the dural attachment and identify a clear cleavage plane in cases of tuberculum sellae meningiomas.
- An effective watertight closure of the anterior cranial base defect with the so-called sandwich technique is mandatory to prevent postoperative CSF leaks.
- The "awake sealant technique" can be useful in the management of minor to moderate postoperative CSF leaks and eventually to reinforce the reconstruction in the immediate postoperative period.

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7 Cavernous Sinus and Meckel's Cave

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Abbreviations

CN Cranial nerve
CS Cavernous sinus
EEA Endoscopic endonasal approach
ICA Internal carotid artery

7.1 Introduction and Rationale

The cavernous sinus and Meckel's cave have traditionally been considered some of the most difficult areas to access surgically. With the advent of endoscopic and minimally invasive approaches, *medial-to-lateral* and *anterior-to-posterior* surgical corridors are added to the surgeon's armamentarium. Although extensive literature exists discussing the relevant surgical anatomy and surgical approaches from a lateral/open perspective, the experience with endonasal approaches is still growing. In this chapter, we analyze our criteria when selecting the endonasal approach for pathology involving the cavernous sinus and Meckel's cave, examine the intricate surgical anatomy of these areas focusing on the endonasal perspective, and provide relevant surgical pearls based on our experience. Furthermore, we discuss our operative setup and instrumentation, as well as postoperative care and complications.

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7.2 Patient Selection: Indications/Contraindications

As with all surgical approaches, appropriate selection of patients is paramount for minimizing morbidity and maximizing the chances of definitively addressing the pathology at hand. The cavernous sinus and Meckel's cave are very closely interrelated anatomically. However, despite this close spatial relationship, their compartmentalization makes it so that, more often than not, they are affected by distinct pathologies which remain confined to one anatomical area without invasion into the other. As such, these are considered separately (Table 7.1). Patient selection is heavily dependent on (1) the presumed pathology and (2) anatomy of the lesion and surrounding structures.

Considering Pathology for Patient Selection

The most important components of patient selection are the presumed diagnosis and definition of the goal of surgery. If tissue diagnosis is not available, a differential diagnosis is made based on the available imaging and history. The details of formulating such a differential diagnosis are beyond the scope of this chapter. However, when diagnosis is unclear, a tissue biopsy may be the primary goal of surgery. The medial-to-lateral surgical corridor of the endoscopic approaches is often ideal for obtaining tissue diagnosis of lesions within the cavernous sinus (e.g., when Tolosa-Hunt syndrome and sarcoid are in question) or Meckel's cave, while minimizing the need for a more invasive procedure.

By far the most common indication for surgery within the cavernous sinus is the resection of pituitary adenomas [1–3]. Although in the era of microscopic pituitary surgery, significant cavernous sinus invasion was regarded a considerable challenge, with the advent of the endoscopic approaches, resections of tumors with pronounced extensions into the

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cavernous sinus have become more effective. This becomes especially important when treating secreting pituitary adenomas. However, even with significantly improved optics and instrumentation, tumor consistency remains one of the most important factors determining its resectability. As such, soft tumors can often be removed completely from the cavernous sinus, while avoiding neurovascular injury (e.g., most pituitary adenomas, chordomas, chondrosarcomas). On the other hand, infiltration of the cavernous sinusoids with fibrous tumors (such as adenomas that have undergone medical treatment/radiation or meningiomas) makes tumor resection much more difficult and significantly increases the risk for neurovascular injury (especially injury to the abducens nerve and carotid artery). In these situations, the goals of surgery usually become maximal safe debulking of the tumor and anatomic separation of the lesion from the pituitary gland so as to decrease the risk of hypopituitarism with subsequent radiosurgery/radiotherapy. The latter is usually achieved by placing a fat graft between the pituitary gland and the lesion within the cavernous sinus (Fig. 7.1a–c) [4].

Primary Meckel's cave pathology usually involves schwannomas of the trigeminal nerve [5]. Although meningiomas can primarily involve the meningeal envelope of Meckel's cave, they usually extend from the posterior fossa into the cave through the porus trigeminus, without actually involving the cavernous sinus. These meningiomas often present with trigeminal neuralgia or neuropathy, which can be effectively relieved with surgical removal.

Pathologies that can affect both the cavernous sinus as well as Meckel's cave include petroclival meningiomas as described above as well as chordomas. The tendency of the latter to spread through the basilar plexus and interdural venous channels provides a portal to the cavernous sinus, while extension outside the venous sinusoids leads to the porus trigeminus and Meckel's cave. Chordomas tend to be softer and easier to resect, in contrast to petroclival meningiomas, which can often be more fibrous. Additionally, malignancies that arise more anteriorly, such as in the paranasal sinuses or the pterygopalatine fossa, can extend poste-

Table 7.1 Pathologies for cavernous sinus and Meckel's cave

Cavernous sinus pathology	
Primary	*Secondary*
Meningioma Schwannoma Aneurysm Hemangioma Inflammatory (e.g., sarcoidosis, Tolosa-hunt syndrome) Infectious (e.g., neurocysticercosis, septic thrombophlebitis) Thrombosis Arteriovenous fistulas Lymphoma Dermoids	Pituitary adenoma (by far most common) Meningioma Spread of malignancies from paranasal sinuses (e.g., adenoid cystic carcinoma, squamous cell carcinoma, adenocarcinoma, etc.) Chordoma Chondrosarcoma Craniopharyngioma Juvenile nasopharyngeal angiofibroma
Meckel's cave pathology	
Primary	*Secondary*
Schwannoma (by far most common) Meningioma CAPNON Inflammatory states (e.g., sarcoidosis) Infections (e.g., neurocysticercosis) Lymphoma	Spread of malignancies from paranasal sinuses (e.g., adenoid cystic carcinoma, squamous cell carcinoma, adenocarcinoma, etc.) Meningioma Chordoma Chondrosarcoma Juvenile nasopharyngeal angiofibroma
Pathology potentially affecting both the cavernous sinus and Meckel's cave	
Meningiomas (most commonly petroclival) Chordomas Spread of malignancies from paranasal sinuses (e.g., adenoid cystic carcinoma, squamous cell carcinoma, adenocarcinoma, etc.) Chondrosarcomas	

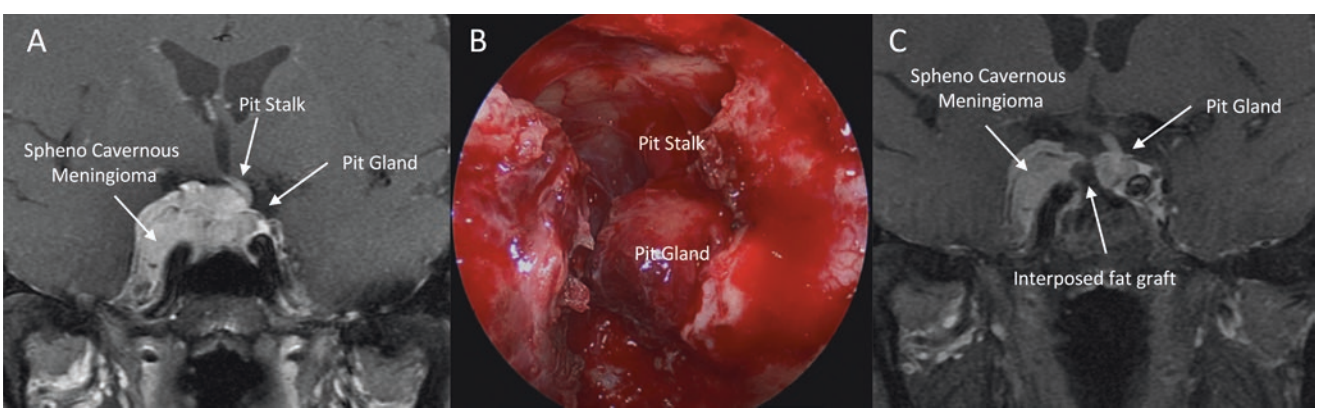

Fig. 7.1 (**a**–**c**) Separation of the pituitary gland from the cavernous sinus. A case of a sphenocavernous meningioma invading the sella, with displacement of the pituitary gland (pit gland) and its stalk (pit stalk) to the left (**a**). After debulking of the medial cavernous sinus and the sella (**b**), a fat graft was placed between the remainder of the tumor within the cavernous sinus and the pituitary gland so as to decrease the risk of hypopituitarism with subsequent radiation therapy (**c**)

riorly and involve both areas through the foramen rotundum (leading into Meckel's cave) and the superior orbital fissure (leading into the cavernous sinus). In these cases, the goal of surgery is usually palliative, aiming to address either refractory trigeminal neuropathy or tumor debulking prior to radiation treatment, but in very selective occasions, surgery is performed with curative intent which requires orbital enucleation and sacrifice of trigeminal nerve branches.

Anatomical Considerations

Regardless of the pathology at hand, the anatomy of each lesion is crucial in determining the best approach and its limitations. The medial-to-lateral surgical corridor provided by the endonasal approaches allows the surgeon to work medial to the lateral wall of the cavernous sinus, where cranial nerves III, IV, and V1 run toward the superior orbital fissure. This makes these approaches ideal for addressing midline pathology with lateral extension. Although such procedures place cranial nerve VI at risk, the nerve itself does not constitute a limitation of the approach. The compartment lateral to the horizontal carotid artery (lateral cavernous sinus compartment – discussed in the anatomy section) is the most challenging to expose through endoscopic approaches. However, adequate bone removal can allow manipulation of the cavernous internal carotid and enhance the surgical corridor. In rare occasions tumors extending beyond the lateral and superior walls of the cavernous sinus can still be approached through endoscopic approaches, but generally this becomes very challenging. Tumors with extension into the cavernous sinus from lateral to medial, such as with medial sphenoid wing meningiomas, place the cranial nerves between the tumor and midline and thus are better approached through lateral approaches, either conventional (e.g., cranio-orbitozygomatic) or minimally invasive/keyhole approaches (e.g., lateral orbitotomy through a lateral canthus incision). In our experience, the roof of the cavernous sinus does not constitute a limitation for endoscopic approaches, and tumors extending through the roof into the peduncular and ambient cisterns can safely and effectively be addressed through this ventral surgical corridor. A detailed understanding of the anatomy allows the surgeon to develop the surgical corridor provided by the tumor while preserving key neurovascular structures.

With regard to Meckel's cave, when pathology primarily involves the root of the trigeminal nerve with extension into the Gasserian ganglion (such as with trigeminal schwannomas), a lateral approach (e.g., retrosigmoid, middle fossa with or without anterior petrosectomy, or posterior transpetrosal) is usually indicated as the endoscopic approaches usually do not provide optimal exposure. Conversely, when Meckel's cave is involved by extension from any of its three subdivisions (V1–3), the endoscopic approaches can provide great access, while avoiding the need for temporal lobe retraction. This may be even more important when addressing left-sided (i.e., dominant) lesions. Another important component in deciding the most appropriate approach is the presumed displacement of the trigeminal ganglion and its three trigeminal divisions. In patients presenting with benign lesions that occupy the medial inferior compartment of Meckel's cave, the trigeminal nerve is most often displaced superolaterally against the middle fossa dura. In these instances, the medial-to-lateral endoscopic corridor is more appropriate. Conversely, the ganglion may occasionally be displaced medially from a laterally located lesion. In these situations, either a lateral approach is indicated, or in selected cases, the anteromedial (between V1 and V2) and anterolateral (between V2 and V3) middle fossa triangles can be exploited through an endoscopic corridor to reach the lateral aspect of Meckel's cave [5].

7.3 Surgical Anatomy

The intricate anatomy of the cavernous sinus and Meckel's cave is generally regarded as a challenging topic to master. Although there is extensive literature detailing this anatomy from a lateral surgical perspective, familiarity with this same anatomy from an endoscopic perspective is still growing. Here we only briefly touch upon the classic anatomy of the cavernous sinus and Meckel's cave but emphasize anatomical details that become more relevant through the anterior-to-posterior and medial-to-lateral surgical corridors provided by the endoscopic approaches. Some of the basic landmarks of endoscopic endonasal surgery are shown in Fig. 7.2a–h.

Cavernous Sinus

Transcranial skull base approaches have provided safe access zones through its superior and lateral walls, with four different triangles: clinoidal/oculomotor for the superior wall and supratrochlear/infratrochlear for the lateral wall [6–11]. The classic cavernous sinus anatomy is shown in Fig. 7.3a–i. In contrast, the endonasal route provides access through the medial (sellar) and/or anterior (sphenoidal) walls of the cavernous sinus. Here we describe a practical and surgically relevant endonasal classification scheme in which the cavernous sinus is divided into four compartments according to their relation to the intracavernous internal carotid artery (Fig. 7.4a–d) [12]. The classification proposed here is partially modified from the venous spaces ("compartments") described by Harris and Rhoton in 1976 [6].

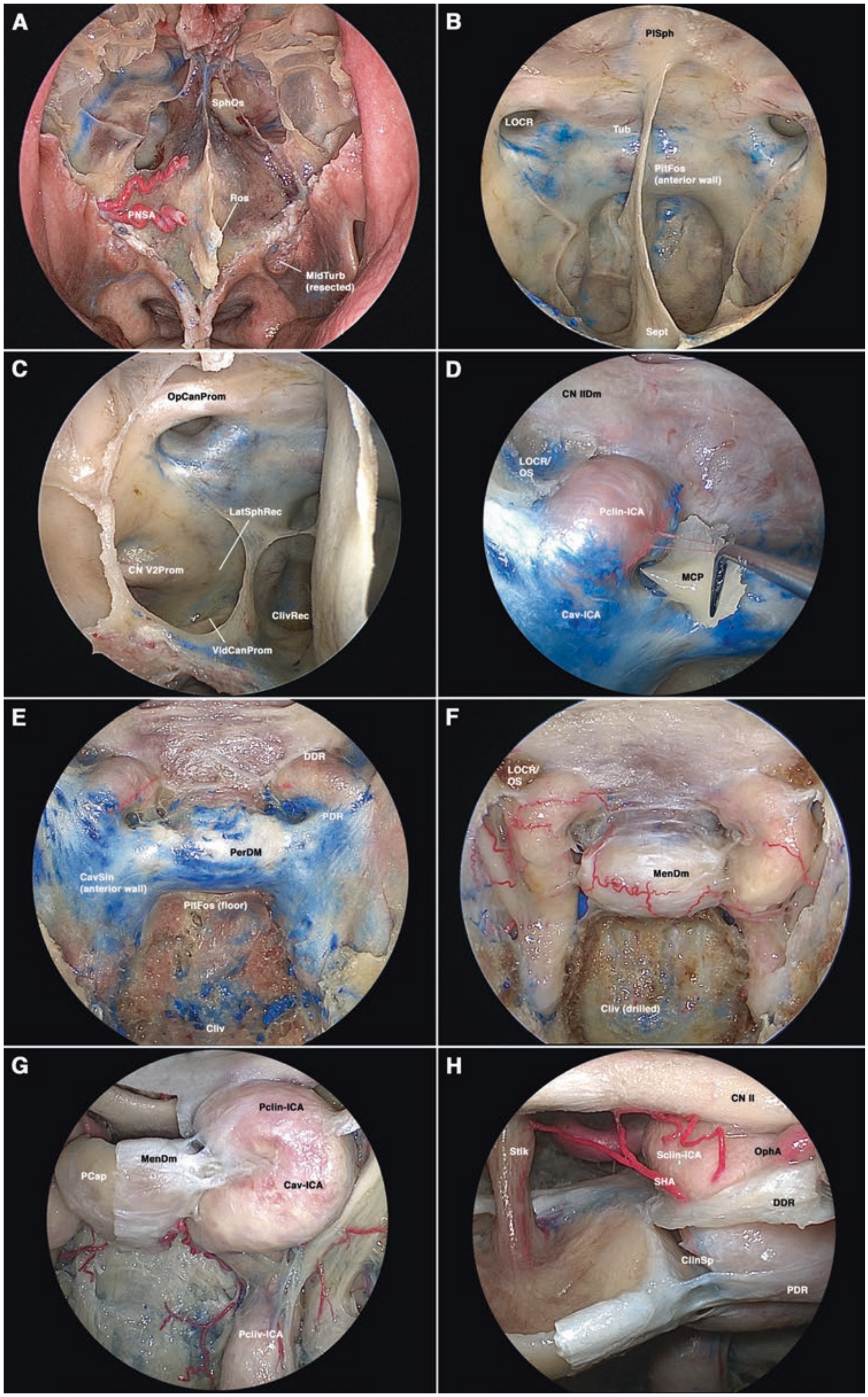

Fig. 7.2 (**a**–**h**) Overview of sphenoidal and perisellar endoscopic anatomy. (**a**) View onto the sphenoid bone following septectomy, middle turbinectomy, and ethmoidectomy for visualization. Note the posterior nasoseptal artery branches providing vascular supply for the nasoseptal flap. (**b**, **c**) Intrasphenoidal septations and protrusions of important neurovascular structures in its wall. (**d**) Detail of the middle clinoid process, a landmark for the cavernous sinus roof and transition from cavernous to paraclinoidal ICA segment. (**e**) Exposure of periosteal dura, (**f**) meningeal dura, and (**g**) pituitary capsule, the three distinct layers covering the pituitary gland anteriorly. (**h**) Detail of the clinoidal space, anteromedially to the cavernous sinus roof (**a**–**h**, formalin-fixed, silicone-injected cadaveric specimen). *MidTurb* middle turbinate, *Ros* rostrum, *SphOs* sphenoidal ostium, *PNSA* posterior nasoseptal artery, *PlSph* planum sphenoidale, *Tub* tuberculum sellae, *PitFos* pituitary fossa, *Sept* intrasphenoidal septations, *LOCR* lateral opticocarotid recess, *OpCanProm* optic canal prominence, *ClivRec* clival recess, *CN V2Prom* prominence of the maxillary division of trigeminal nerve, *VidCanProm* Vidian canal prominence, *LatSphRec* lateral recess of the sphenoid sinus, *CN IIDm* optic nerve within dura mater, *MCP* middle clinoid process, *Pclin-ICA* paraclinoidal internal carotid artery, *Cav-ICA* cavernous internal carotid artery, *LOCR/OS* lateral opticocarotid recess/optic strut, *CavSin* cavernous sinus, *Cliv* clivus, *PerDm* periosteal dura mater, *PDR* proximal dural ring, *DDR* distal dural ring, *MenDm* meningeal dura mater, *PCap* pituitary capsule, *Pcliv-ICA* paraclival internal carotid artery, *Stlk* pituitary stalk, *SHA* superior hypophyseal artery, *ClinSp* clinoidal space, *Sclin-ICA* supraclinoidal internal carotid artery, *CN II* optic nerve, *OphA* ophthalmic artery

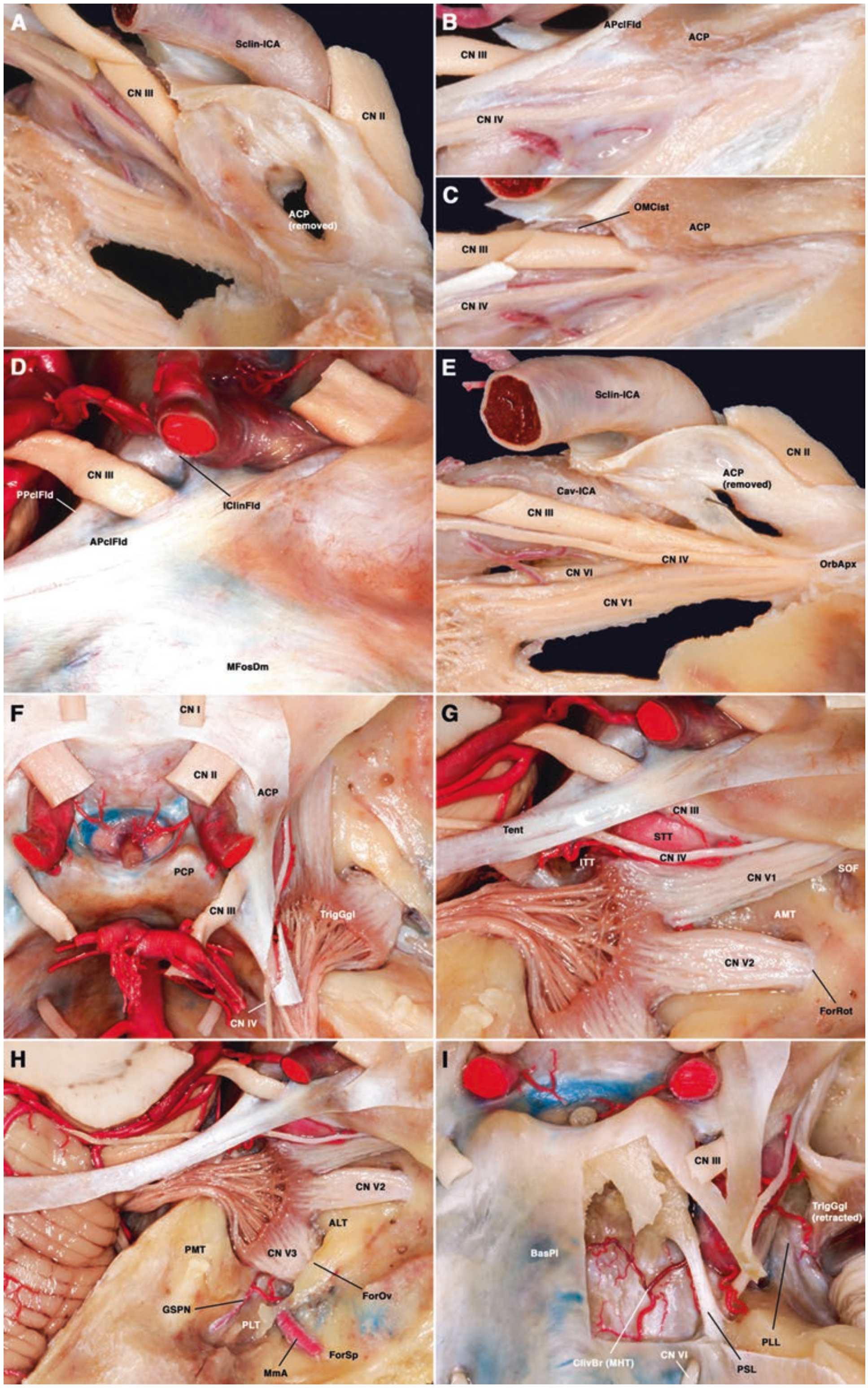

Fig. 7.3 (**a**–**i**) Surgical triangles of the cavernous sinus and middle cranial fossa. (**a**) Clinoidal/anteromedial triangle, bordered by the optic and oculomotor nerves and the tentorial edge, containing the anterior clinoid process and the clinoidal ICA. (**b**, **c**) Stepwise exposure of the oculomotor cistern underneath the anterior petroclinoidal fold. (**d**) Oculomotor/medial/Hakuba's triangle, bordered by the anterior and posterior petroclinoidal folds and the interclinoidal fold harboring the oculomotor nerve and the horizontal cavernous ICA. (**e**) Detail of the transition from cavernous sinus to orbital apex. (**f**) Axial view onto pituitary, middle, and posterior cranial fossae. (**g**) Supratrochlear/paramedian triangle, bordered by the oculomotor and trochlear nerves and the tentorial edge. Infratrochlear/Parkinson's triangle, bordered by the trochlear nerve, the ophthalmic division of the trigeminal nerve, and the tentorial edge, containing the cavernous ICA and the abducens nerve. Anteromedial/Mullan's triangle, bordered by the ophthalmic and maxillary divisions of the trigeminal nerve and a line connecting the superior orbital fissure with the foramen rotundum, containing the abducens nerve and ophthalmic vein. (**h**) Anterolateral triangle, bordered by the

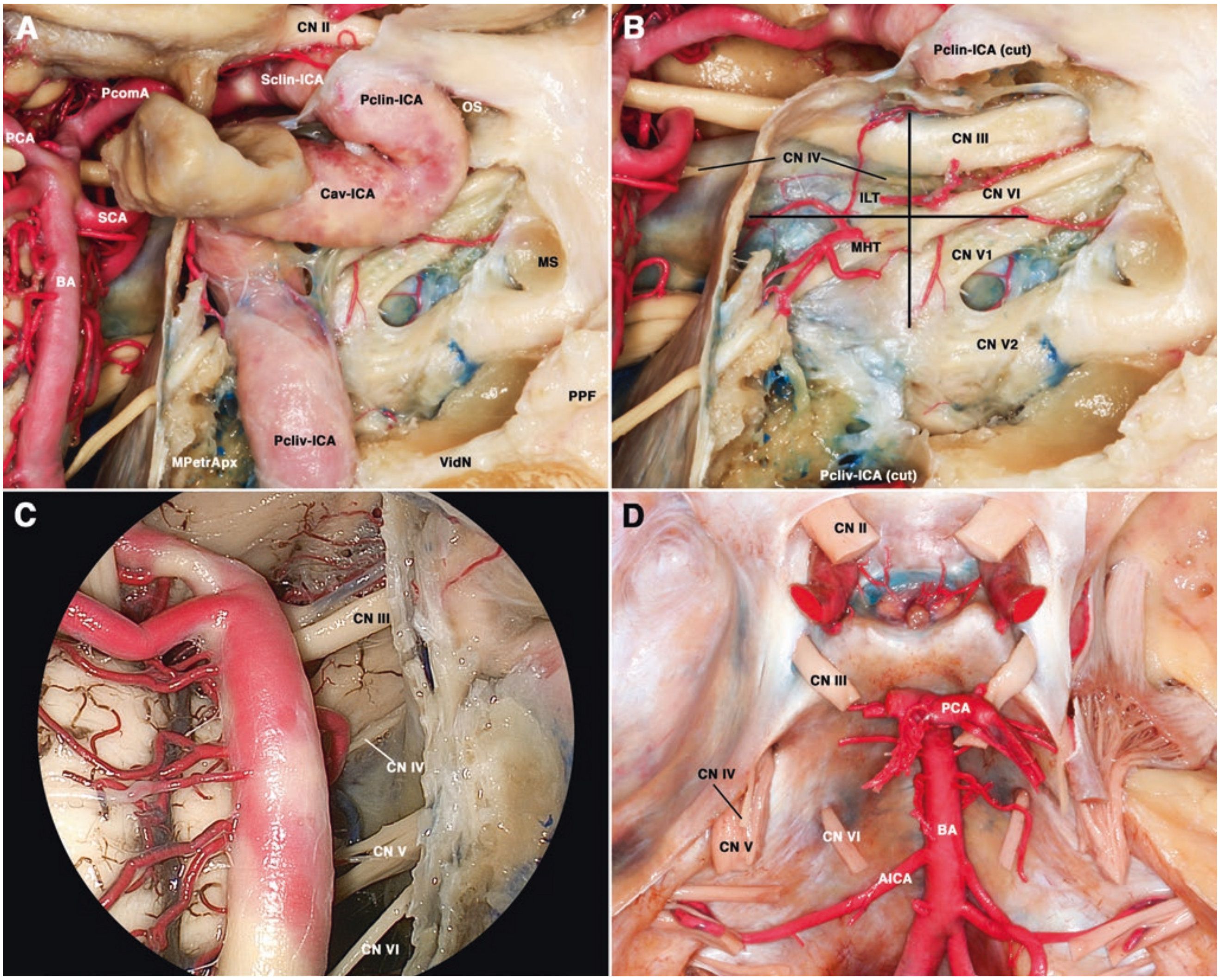

Fig. 7.4 (**a**–**d**) Endoscopic compartments of the cavernous sinus, overview. (**a**) Oblique view onto the cavernous sinus and adjacent areas medially in a hemisected specimen (left side). In (**b**), the ICA has been removed for visualization of the neurovascular structures in the respective compartments (for details see text). (**c**) Transclival endoscopic view, demonstrating the cisternal cranial nerves entering the dura/cavernous sinus. (**d**) Posterior microscopic view for comparison (**a**–**d**, formalin-fixed, silicone-injected cadaveric specimen). *BA* basilar artery, *PCA* posterior cerebral artery, *SCA* superior cerebellar artery, *PcomA* posterior communicating artery, *Sclin-ICA* supraclinoidal internal carotid artery, *Pclin-ICA* paraclinoidal internal carotid artery, *Cav-ICA* cavernous internal carotid artery, *Pcliv-ICA* paraclival internal carotid artery, *CN II* optic nerve, *MPetrApx* medial petrous apex, *MS* maxillary strut, *OS* optic strut, *VidN* Vidian nerve, *PPF* pterygopalatine fossa, *CN III* oculomotor nerve, *CN IV* trochlear nerve, *CN V1* ophthalmic division of the trigeminal nerve, *CN V2* maxillary division of the trigeminal nerve, *CN VI* abducens nerve, *MHT* meningohypophyseal trunk, *ILT* inferolateral trunk, *AICA* anterior inferior cerebellar artery

Fig. 7.3 (continued) maxillary and mandibular divisions of the trigeminal nerve and a line connecting the foramen rotundum and ovale. Posterolateral/Glasscock's triangle, bordered by the mandibular division of the trigeminal nerve, the greater superficial petrosal nerve, and a line from the foramen spinosum to the arcuate eminence. Posteromedial/Kawase's triangle, also known as the rhomboid, bordered by the mandibular division of the trigeminal nerve, the greater superficial petrosal nerve, the arcuate eminence, and the superior petrosal sinus. (**i**) Posterior aspect of the superior clivus and trigeminal impression; note the petrolingual and petrosphenoidal ligaments and the course of the abducens nerve (**a**–**h**, formalin-fixed, silicone-injected cadaveric specimen). *CN II* optic nerve, *CN III* oculomotor nerve, *ACP* anterior clinoid process, *Sclin-ICA* supraclinoidal internal carotid artery, *CN IV* trochlear nerve, *APclFld* anterior petroclinoidal fold, *OMCist* oculomotor cistern, *PPclFld* posterior petroclinoidal fold, *MFosDm* middle cranial fossa dura mater, *IClinFld* interclinoidal fold, *OrbApx* orbital apex, *Cav-ICA* cavernous internal carotid artery, *CN V1* ophthalmic division of the trigeminal nerve, *CN V2* maxillary division of the trigeminal nerve, *CN VI* abducens nerve, *CN I* olfactory nerve, *PCP* posterior clinoid process, *TrigGgl* trigeminal ganglion, *Tent* tentorium cerebelli, *STT* supratrochlear triangle, *ITT* infratrochlear triangle, *SOF* superior orbital fissure, *AMT* anteromedial triangle, *ForRot* foramen rotundum, *ForOv* foramen ovale, *CN V3* mandibular division of the trigeminal nerve, *ForSp* foramen spinosum, *MmA* middle meningeal artery, *ALT* anterolateral triangle, *GSPN* greater superficial petrosal nerve, *PLT* posterolateral triangle, *PMT* posteromedial triangle, *BasPl* basilar plexus, *ClivBr (MHT)* clival branches of the meningohypophyseal trunk, *PSL* petrosphenoidal ligament (Gruber's), *PLL* petrolingual ligament

Internal Carotid Artery Anatomy from an Endonasal Perspective

Understanding the parasellar and paraclival internal carotid artery anatomy is essential for understanding the anatomy of the cavernous sinus from an endonasal perspective [13] (Figs. 7.2a–h and 7.4a–d). It is divided into two segments: *cavernous and paraclinoidal.* The subsegments of the *cavernous ICA* from proximal to distal are short vertical (continuation of paraclival ICA), horizontal, and anterior genu. The posterior genu is located between the short vertical and horizontal subsegments. The *paraclinoidal ICA* is a continuation of the anterior genu of the cavernous ICA as it emerges from the cavernous sinus. It is located within the clinoidal space at the roof of the cavernous sinus and is limited by the proximal and distal dural rings. It is bounded superolaterally by the lateral opticocarotid recess (or optic strut). The middle clinoid, when present, marks the transition between the cavernous and paraclinoidal ICA, while the medial opticocarotid recess at the lateral aspect of the tuberculum sella is located just medial to the paraclinoidal-supraclinoidal ICA transition [14, 15].

Superior Compartment of the Cavernous Sinus (Figs. 7.5a–d and 7.6a–d)

This compartment lies above the horizontal cavernous carotid artery and posterior to its anterior genu. Superiorly and laterally, it is limited by the roof of the cavernous sinus, anterolaterally by the ventral surface of the paraclinoidal

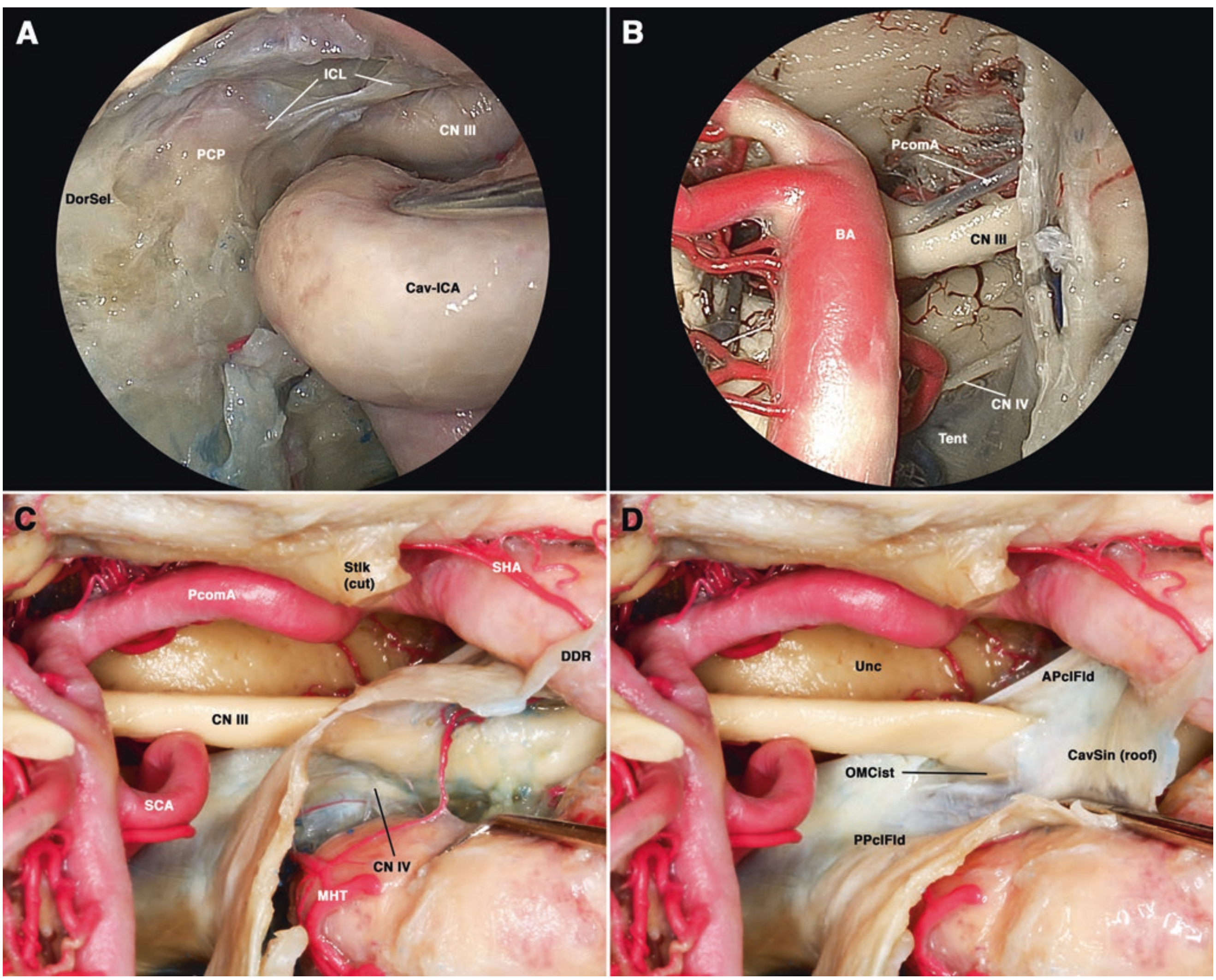

Fig. 7.5 (**a**–**d**) Superior compartment of the cavernous sinus anatomy. (**a**, **b**) Endoscopic and (**c**, **d**) microscopic anatomy (left side; for details see text). (**a**–**d**, formalin-fixed, silicone-injected cadaveric specimen). *DorSel* dorsum sellae, *PCP* posterior clinoid process, *CN III* oculomotor nerve, *Cav-ICA* cavernous internal carotid artery, *ICL* interclinoidal ligament, *BA* basilar artery, *Tent* tentorium cerebelli, *PcomA* posterior communicating artery, *CN IV* trochlear nerve, *Stlk* pituitary stalk, *SHA* superior hypophyseal artery, *MHT* meningohypophyseal trunk, *SCA* superior cerebellar artery, *DDR* distal dural ring, *APclFld* anterior petroclinoidal fold, *PPclFld* posterior petroclinoidal fold, *OMCist* oculomotor cistern, *Unc* uncus, *CavSin* cavernous sinus

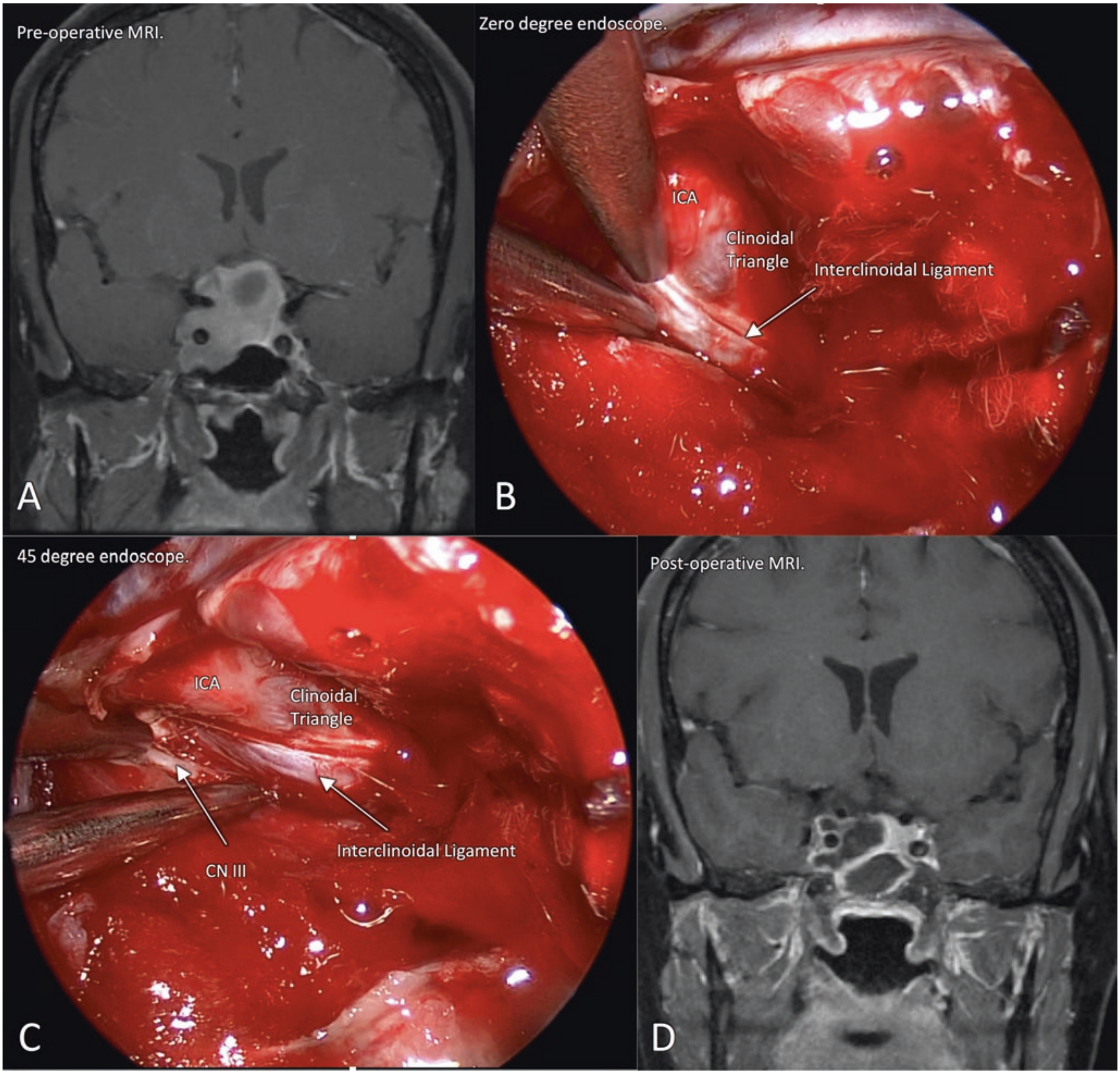

Fig. 7.6 (**a**–**d**) Clinical example of pituitary macroadenoma invading the superior compartment. (**a**) Preoperative MRI showing large adenoma, with cavernous sinus invasion (both superior and inferior compartments are invaded). (**b**) Intraoperative view using a zero-degree endoscope showing the interclinoidal ligament – an important landmark in identifying the oculomotor nerve (CNIII). (**c**) Using a 45-degree endoscope, CNIII is identified. (**d**) Postoperative MRI showing complete resection. The patient had no cranial neuropathies postoperatively

ICA within the clinoidal triangle, and posterolaterally by the dura of the oculomotor triangle. Posteriorly, its inferior border which marks the transition to the posterior compartment described below is the posterior genu of the cavernous internal carotid artery.

The key structure within this compartment is the *oculomotor nerve,* which enters through its posterior roof, corresponding to the oculomotor triangle, and then runs within its lateral wall. At its entrance into the cavernous sinus, the dura of the oculomotor triangle invaginates for a short distance before adhering to the nerve. This dural invagination creates the CSF-filled cistern of the oculomotor nerve. This segment of the oculomotor nerve running between the two layers of dura forming the walls of the oculomotor cistern is also called the *inter*dural segment [16]. Traveling anteriorly, inferiorly, and laterally, the nerve is incorporated in the lateral wall of the cavernous sinus. Notably, this transition into the lateral cavernous sinus wall roughly lies lateral to the anterior

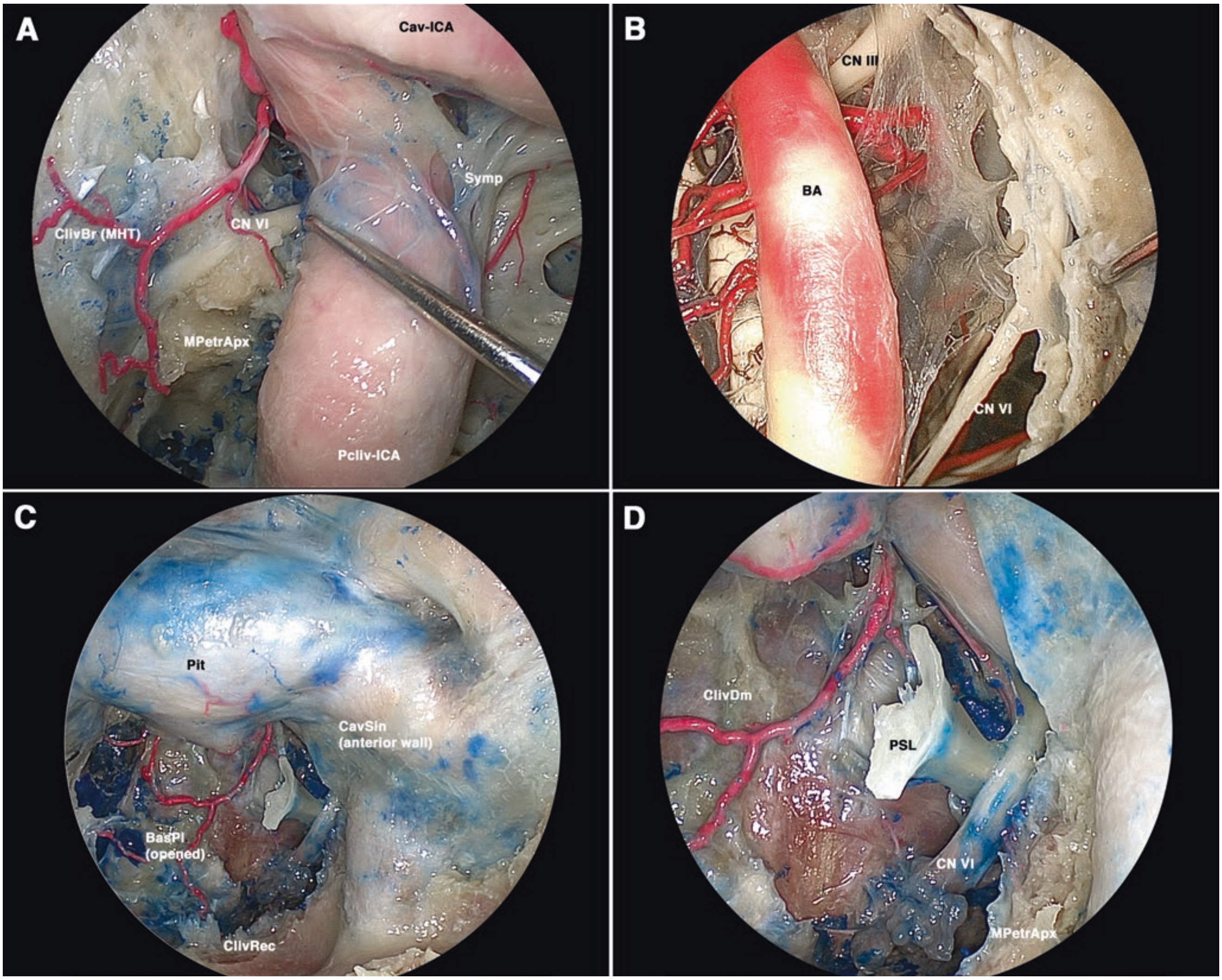

Fig. 7.7 (**a**–**d**) Posterior compartment of the cavernous sinus. (**a**–**d**) Endoscopic anatomy of the cisternal, intradural, and cavernous course of the abducens nerve (left side; for details see text) (**a**–**d**, formalin-fixed, silicone-injected cadaveric specimen). *Cav-ICA* cavernous internal carotid artery, *Pcliv-ICA* paraclival internal carotid artery, *ClivBr (MHT)* clival branches of the meningohypophyseal trunk, *MPetrApx* medial petrous apex, *CN VI* abducens nerve, *Symp* sympathetic plexus of the ICA, *BA* basilar artery, *CN III* oculomotor nerve, *Pit* pituitary gland, *BasPl* basilar plexus, *ClivRec* clival recess, *CavSin* cavernous sinus, *ClivDm* clival dura mater, *PSL* petrosphenoidal ligament (Gruber's)

genu of the cavernous internal carotid artery. A very important anatomic relationship from an endoscopic standpoint is that of the *interclinoidal ligament* and the oculomotor nerve. This robust dural band, extending between the anterior and posterior clinoid processes, corresponds to the medial border of the posterior roof of the cavernous sinus (the oculomotor triangle) and separates it from the clinoidal triangle anteriorly and medially, which forms the anterior roof of the cavernous sinus. When entering the cavernous sinus from a medial to lateral trajectory, as is the case with endoscopic approaches, the *interclinoidal ligament* is encountered first and can be often confused with the oculomotor nerve as it also forms a well-defined whitish band running in the same direction with the nerve. It is, therefore, important to remember that the oculomotor nerve lies just lateral and roughly parallel to the interclinoidal ligament, whereas the paraclinoidal ICA will run just medial and anterior to the ligament.

Posterior Compartment of the Cavernous Sinus (Figs. 7.7a–d and 7.8a–d)

This compartment lies posterior to the short vertical cavernous ICA and anterior to the lateral petroclival dura, which forms the posterior wall of the CS. As described previously, the posterior genu of the cavernous internal carotid artery, separating the short ascending segment from the horizontal segment, marks its superior border and the transition into the superior compartment.

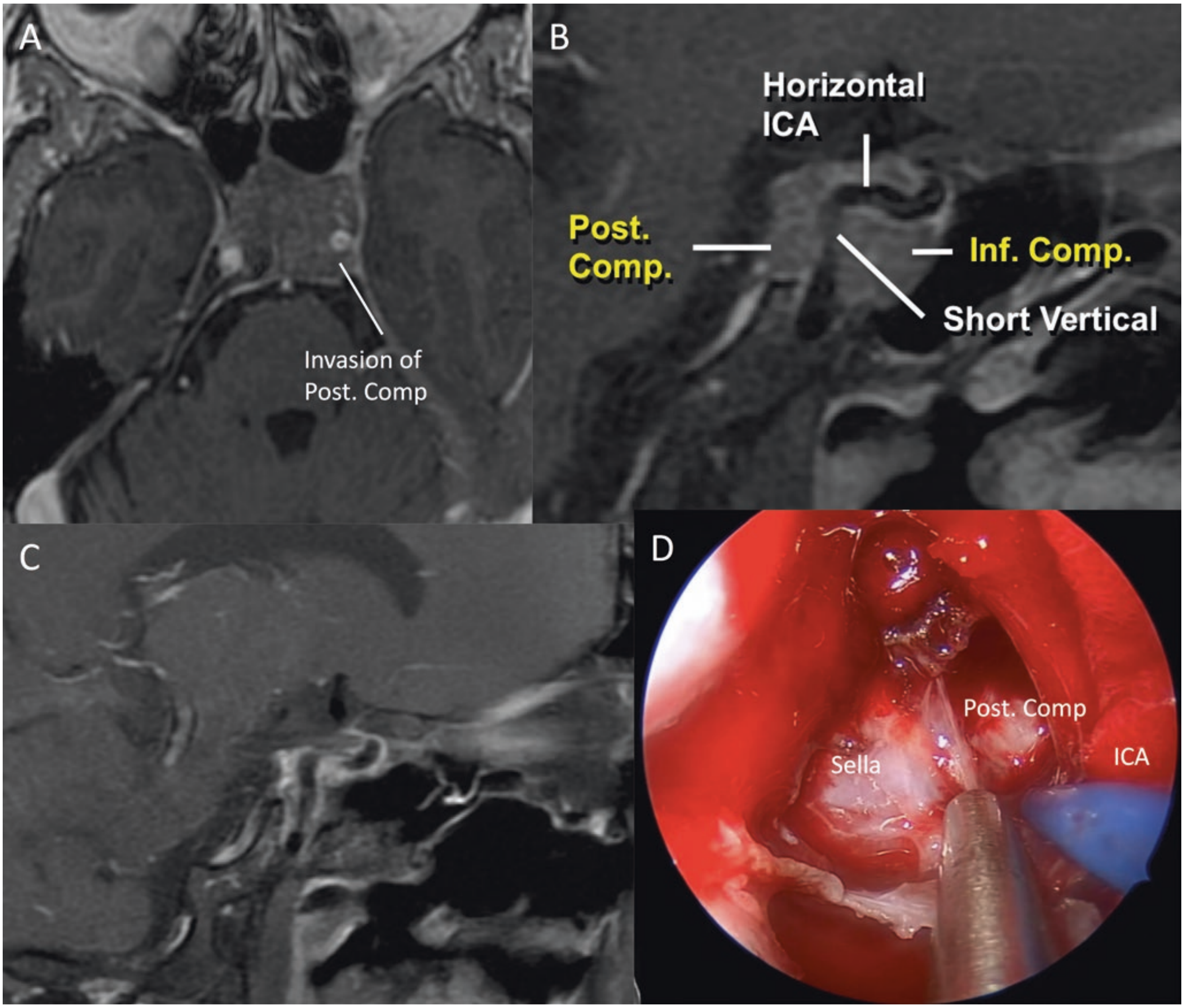

Fig. 7.8 (**a**–**d**) Clinical example of pituitary adenoma invading the posterior compartment. A large macroadenoma with invasion of the posterior (*Post Comp*) and inferior compartments (*Inf Comp*) (**a**, **b**). A complete resection was achieved without evidence of cranial neuropathy (**c**). Intraoperative image of the posterior compartment after resection of the tumor, with neurophysiologic confirmation of the location of cranial nerve VI (**d**)

Some of the key structures related to this compartment are the *abducens* nerve and the *meningohypophyseal trunk*. The *meningohypophyseal trunk* arises from the posterior wall of the posterior genu of the cavernous internal carotid artery and typically has three named branches, all of which can arise from the carotid artery directly and have relevance to endoscopic endonasal approaches: the inferior hypophyseal artery, the dorsal meningeal artery, and the tentorial artery (also known as the *artery of Bernasconi and Cassinari*). The tentorial artery(ies) can be significantly enlarged when providing blood supply to tumors such as petroclival meningiomas, and early identification and coagulation can aid in devascularizing the tumor. The inferior hypophyseal arteries course medially from the posterior genu toward the dura of the floor of the sella. While these arteries are thought to contribute to the vascular supply of the posterior gland, bilateral selective ligation of these vessels in a series of three patients undergoing interdural pituitary transpositions did not result in any permanent pituitary dysfunction [17], suggesting that the vascular supply of the human pituitary gland is less dependent on these arteries. Finally, the dorsal meningeal artery courses posteriorly, inferiorly, and medially toward the dura of the dorsum sellae. Notably, this artery and its branches usually lie just medial to the interdural segment of cranial nerve VI, and, therefore, extreme caution should be exercised when coagulating dural bleeders during middle and upper transclival approaches.

The *gulfar segment* of the *abducens nerve* is located at the most inferior portion of this compartment as it passes behind the internal carotid, through Dorello's canal to enter the cavernous sinus [18]. This nerve segment runs above the medial petrous apex and under the petrosphenoidal or Gruber's ligament, sitting at the confluence of the *superior* and *inferior petrosal sinuses* with the *basilar plexus* (also referred to as the gulf) [18]. Very importantly, it is critical to know that although at the point where the abducens nerve pierces the meningeal layer of the clival dura, it is ensheathed within two layers of dura created by an invagination of the meningeal layer (much like the oculomotor nerve in the oculomotor cistern), by the time the nerve reaches the posterior compartment, it is not covered by any dural layer and is more susceptible to injury [18].

Inferior Compartment of the Cavernous Sinus (Fig. 7.9a–f)

This compartment lies inferior to the horizontal segment of the cavernous internal carotid artery and its anterior genu and anterior to its short vertical segment. The anterior wall of this compartment is the anterior wall of the cavernous sinus.

The *sympathetic plexus* surrounding the adventitia of the internal carotid artery coalesces into one or more well-defined fascicles referred to as the *sympathetic nerve(s)*, which travel from the short vertical segment of the cavernous carotid to merge with the *distal cavernous segment of the abducens nerve*, as they run just inferior and lateral to the horizontal segment of the cavernous carotid at the transition between the inferior and lateral compartments. The sympathetic nerve follows an arcuate trajectory from an initial vertical to one that is more horizontal. Notably, the abducens nerve runs just medial to the first division of the trigeminal nerve (V1), which lies in the lateral compartment described below.

Lateral Compartment of the Cavernous Sinus (Figs. 7.9a–f and 7.10a–h)

This compartment occupies the area lateral to the cavernous carotid artery and anterior to its posterior genu. Craniocaudally, the lateral compartment extends from the proximal dural ring that covers the inferior surface of the optic strut superiorly to the V2 prominence and maxillary strut inferiorly. The optic and maxillary struts separate the superior orbital fissure from the optic canal and foramen rotundum, respectively. At the anterior limit of the optic and maxillary struts, the cranial nerves have exited the cavernous sinus and entered the superior orbital fissure.

This compartment contains the *third and fourth cranial nerve* and the *first division of the trigeminal nerve (V1)*, which are located at the lateral wall of the cavernous sinus. As mentioned above, the distal cavernous segment of the abducens nerve is located at the transition between inferior and lateral compartments and lies just medial to V1. The arterial branches of the *inferolateral trunk,* arising from the inferolateral surface of the horizontal cavernous ICA, course inferiorly and laterally between the abducens nerve and V1 to ultimately distribute along the lateral wall of the cavernous sinus.

Meckel's Cave and Trigeminal Nerve (Fig. 7.11a–h)

The posterior trigeminal root, arising from the lateral midpons, passes forward to Meckel's cave and the Gasserian ganglion in the middle fossa through the porus trigeminus, which has the trigeminal impression as a floor, the medial petrous apex and the inferior petrosal sinus as the medial wall, the trigeminal eminence as the lateral wall, and the superior petrosal sinus as a roof [5, 19]. Invagination of the meningeal layer of the dura through the porus trigeminus allows the subarachnoid space to extend forward within Meckel's cave to approximately the midportion of the Gasserian ganglion. The nerve splits into the three divisions at the anterior edge of the ganglion. The ophthalmic division courses in the lower anterior part of the cavernous sinus. The maxillary nerve courses just below the cavernous sinus, where its medial side produces a prominence in the lateral wall of the sphenoid sinus just before exiting the foramen rotundum to enter the pterygopalatine fossa. The mandibular nerve courses toward the foramen ovale to exit to the infratemporal fossa. Notably, only the upper third of the medial wall of Meckel's cave is located directly lateral to the cavernous sinus. The maxillary nerve does not course in the lateral wall of the dural envelope of the sinus as does the ophthalmic nerve. It courses beneath the dura of the middle fossa, below the level where the medial and lateral walls of the cavernous sinus join at the lower edge of the ophthalmic nerve. In effect, almost all of Meckel's cave is located inferior and lateral to the posterior part of the cavernous sinus [19].

An important anatomic relationship is that of Meckel's cave and the petrous, lacerum, and ascending cavernous segments of the internal carotid artery. The horizontal segment of the petrous carotid runs just medial to Meckel's cave. At the anterior petrous carotid genu and transition to the lacerum carotid segment, the carotid is separated from Meckel's cave by the lingual process of the sphenoid bone and petrolingual ligament. Removal of this usually thin bony structure provides access to Meckel's cave. The Vidian nerve, which is created by (a) the parasympathetic fibers of the greater superficial petrosal nerve coming from posteriorly and laterally under Meckel's cave and (b) the sympathetic fibers of the deep petrosal nerve coming from the petrous carotid sympathetic plexus, is found just lateral to the anterior genu of the petrous carotid. The Vidian artery, an inconstant branch of the petrous carotid entering the Vidian canal, can be significantly

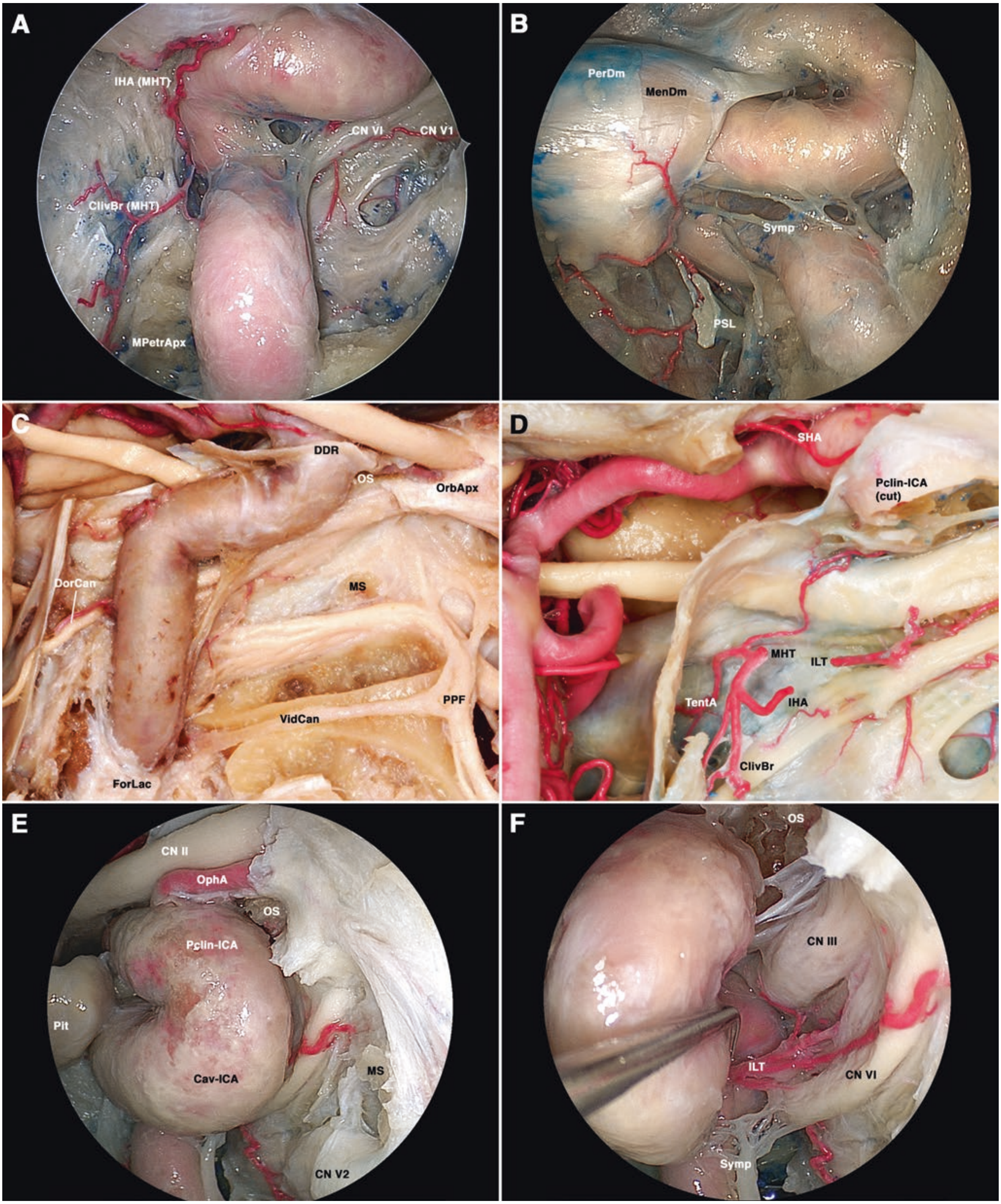

Fig. 7.9 (**a**–**f**) Inferior and lateral compartments of the cavernous sinus, microvascular supply. (**a**, **b**) Inferior compartment, containing the sympathetic plexus of the ICA and distal abducens nerve. (**c**) Microscopic hemisection, illustrating the spatial relationships between the Vidian canal inferiorly and horizontal ICA superiorly. Note the optic and maxillary struts and important endoscopic landmarks. (**d**) Microvascular detail, intracavernous branches of the meningohypophyseal and inferolateral trunks. (**e**) Lateral compartment; in (**f**) the anterior genu of the ICA is retracted medially for visualization. (**a**–**f**, formalin-fixed, silicone-injected cadaveric specimen). *IHA (MHT)* inferior hypophyseal artery of the meningohypophyseal trunk, *ClivBr (MHT)* clival branches of the meningohypophyseal trunk, *CN IV* trochlear nerve, *CN V1* ophthalmic division of the trigeminal nerve, *MPetrApx* medial petrous apex, *Symp* sympathetic plexus of the ICA, *PSL* petrosphenoidal ligament, *DorCan* Dorello's canal, *ForLac* foramen lacerum, *VidCan* Vidian canal, *PPF* pterygopalatine fossa, *MS* maxillary strut, *OS* optic strut, *DDR* distal dural ring, *OrbApx* orbital apex, *SHA* superior hypophyseal artery, *Pclin-ICA* paraclinoidal internal carotid artery, *MHT* meningohypophyseal trunk, *ILT* inferolateral trunk, *TentA* tentorial artery (Bernasconi-Cassinari), *OphA* ophthalmic artery, *Pit* pituitary gland, *CN II* optic nerve, *CN V2* maxillary division of the trigeminal nerve, *Cav-ICA* cavernous internal carotid artery, *CN III* oculomotor nerve

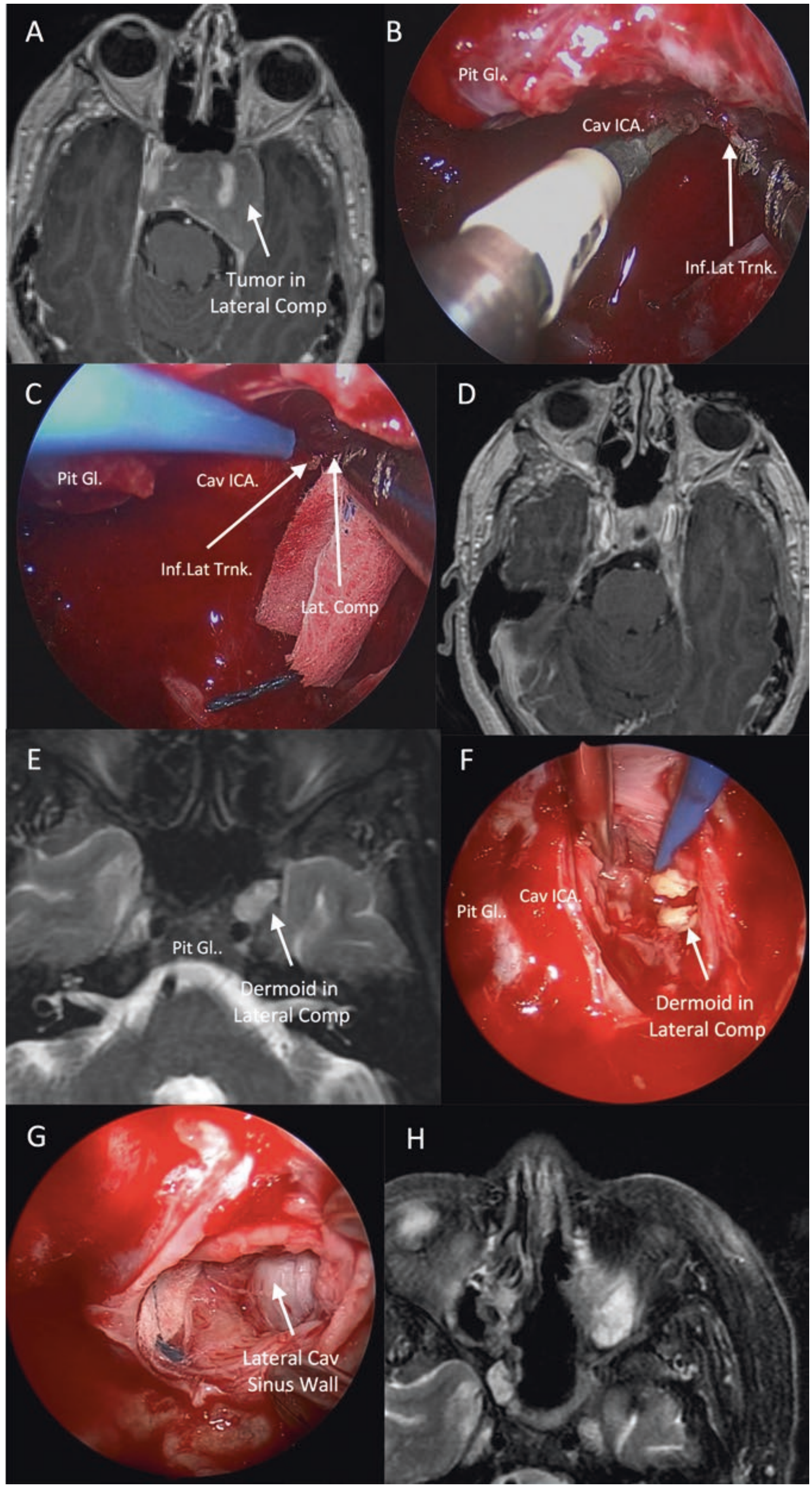

Fig. 7.10 (**a**–**h**) Clinical examples of tumors involving the lateral compartment. (**a**–**d**) Example of a large invasive macroadenoma (**a**). Coagulation and division of the inferolateral trunk allow medial displacement of the carotid artery (**b**). Neurophysiologic confirmation of the location of cranial nerve VI in the lateral compartment is essential (**c**). Postoperative imaging showing gross resection of the tumor. Tumor within the lateral wall was not resected to avoid cranial nerve injury. The patient had no permanent cranial nerve deficits. (**e**–**h**) Example of a dermoid cyst within the lateral compartment (**e**). Intraoperative image showing resection of the tumor with the help of a neurostimulator dissector (**f**). Complete resection of the cyst was confirmed intraoperatively (**g**), as well as on postoperative imaging (**h**)

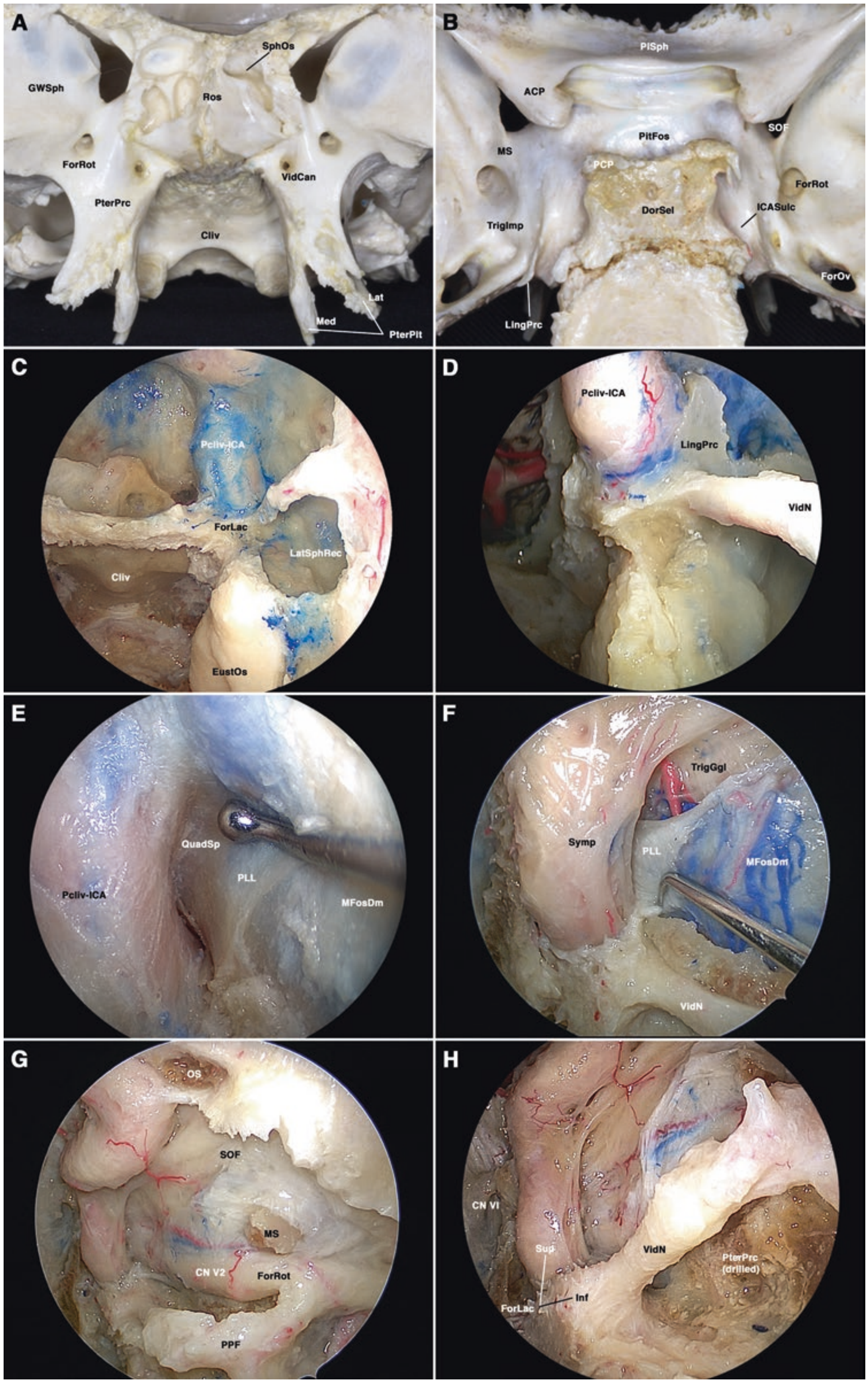

Fig. 7.11 (**a**–**h**) Endoscopic transpterygoid approach to Meckel's cave. (**a**, **b**) Anterior and posterior aspect of the osseous anatomy of the sphenoid, occipital, and pterygoid bones. (**c**–**h**) Stepwise endoscopic anatomical exposure of Meckel's cave. (**c**) Vidian nerve and adjacent areas. (**d**) Lingual process, the door to the (**e**) quadrangular space. (**f**) Petrolingual ligament and the dura of the middle cranial fossa covering the trigeminal ganglion from the endoscopic perspective. (**g**) Superior and (**h**) inferior view of a supravidian transpterygoid approach to Meckel's cave (**a**, **b**, dry, macerated skull specimen; **c**–**f**, formalin-fixed, silicone-injected cadaveric specimen). *GWSph* greater wing of the sphenoid bone, *ForRot* foramen rotundum, *PterPrc* pterygoid process, *Ros* rostrum, *SphOs* sphenoid ostium, *VidCan* Vidian canal, *Cliv* clivus, *Med/Lat PterPlt* medial/lateral pterygoid plate, *ACP* anterior clinoid process, *MS* maxillary strut, *TrigImp* trigeminal impression, *LingPrc* lingual process (sphenoid bone), *PCP* posterior clinoid process, *DorSel* dorsum sellae, *PitFos* pituitary fossa, *PlSph* planum sphenoidale, *SOF* superior orbital fissure, *ICASulc* sulcus of the internal carotid artery, *ForOv* foramen ovale, *Cliv* clivus, *EustOs* ostium of the Eustachian/pharyngotympanic tube, *LatSphRec* lateral recess of the sphenoid sinus, *ForLac* foramen lacerum, *Pcliv-ICA* paraclival internal carotid artery, *VidN* Vidian nerve, *QuadSp* quadrangular space, *PLL* petrolingual ligament, *MFosDm* middle cranial fossa dura mater, *Symp* sympathetic plexus of the ICA, *TrigGgl* trigeminal ganglion, *OS* optic strut, *CN V2* maxillary division of the trigeminal nerve, *PPF* pterygopalatine fossa, *CN VI* abducens nerve, *Sup/Inf ForLac* superior/inferior compartment of the foramen lacerum

enlarged when supplying tumors (e.g., juvenile nasopharyngeal angiofibromas). The petrolingual ligament marks the transition of the lacerum segment of the carotid to the ascending cavernous segment. This transition is important as the dural envelope of the cavernous carotid fuses with the adventitia of the vessel at the lacerum segment.

7.4 Surgical Technique

Cavernous Sinus

Abiding to our cavernous sinus compartment classification, we discuss relevant nuances for each of its compartments separately.

Surgical Nuances Related to the Superior Compartment of the Cavernous Sinus

Exploration of the superior compartment of the cavernous sinus requires removal of the bone covering the paraclinoidal ICA as well as the anterior wall of the cavernous sinus. Although this aggressive bone removal may initially seem unnecessary, it proves critical in allowing lateral displacement of the ICA for direct surgical dissection within this region. Frequently, endoscopic access to this compartment is facilitated by tumor invasion through the medial wall of the cavernous sinus, which creates a natural continuation of a medial-to-lateral surgical corridor from the sella. For cases with true invasion, gentle medial-to-lateral ICA mobilization is performed with the suction shaft while a second surgical instrument is used to remove tumor. Our preference is the use of two 6 and/or 8 Fr teardrop suctions, which allow the surgeon great versatility in the amount of suction applied. The majority of this dissection can be done with zero-degree endoscopes, but the use of angled scopes is helpful to maximize visualization, especially of the dorsal aspect of the anterior genu of the ICA. The interdural segment of the oculomotor nerve is protected as long as the lesion does not invade and extend beyond the roof and lateral wall of the CS. That being said, occasionally the tumor can extend through the oculomotor triangle to the crural and ambient cisterns. Although access to these areas is again facilitated by the corridor created by the tumor, the breach in the posterior cavernous roof may be small, and its surgical expansion requires good knowledge of the anatomic relationships of the oculomotor triangle. The interclinoidal ligament is identified repeatedly during surgery and acts as a landmark for the oculomotor nerve (located lateral to the ligament). Neurophysiology can also help identify cranial nerves III and IV in this area. Our electrophysiological studies have typically shown a positive response for CNs III and IV only at high amperage (2 mAmps) when stimulating posteriorly but a positive response at low amperage (0.5–1 mAmps) when stimulating more anteriorly just behind the genu of the ICA. This is explained by the inner dural layer of the oculomotor triangle becoming thinner as CN III travels anteriorly. The most vulnerable portion of the oculomotor nerve is located just lateral to the anterior genu of the ICA, an area that is not easily accessible from a medial to lateral trajectory. It is also important to note that the superior compartment can be often compressed and not invaded, in which case the medial wall of the CS is displaced laterally but still covers the cavernous ICA and interclinoidal ligament [20].

Surgical Nuances Related to the Posterior Compartment of the Cavernous Sinus

To gain access to this compartment, behind the short ascending segment of the cavernous carotid, extensive bony removal is needed so as to expose the anterior wall of the cavernous sinus as well as the upper paraclival internal carotid which corresponds to the point of entrance of the carotid artery into the cavernous sinus. Although image guidance and Doppler ultrasound are helpful in identifying the paraclival carotids, medial-to-lateral elevation of the dura from the sellar floor to identify the point where the carotid artery dives inferiorly is always a safer strategy before skeletonization of the carotid is pursued. In order to gain access posterior to the cavernous ICA, gentle lateral mobilization of the short vertical subsegment and posterior ICA genu is needed. This may necessitate coagulation and transection of the inferior hypophyseal artery to safely advance toward the posterior compartment. It is extremely important to avoid avulsion of this artery at its takeoff from the carotid, as this may create a defect in the wall of the posterior cavernous carotid genu which may be difficult to control. The short vertical ICA is the best landmark to locate the abducens nerve which will be more or less evident depending on the degree of tumor expansion of this compartment. Electrostimulation will confirm the presence of the abducens nerve at the floor of this compartment just behind the ICA. Venous bleeding from the inferior and superior petrosal sinuses is commonly encountered behind and above the abducens nerve, respectively, and its management requires careful hemostatic technique to prevent nerve damage from excessive packing or unnecessary coagulation.

Surgical Nuances Associated with the Inferior Compartment of the Cavernous Sinus

Access to the inferior compartment requires good exposure of the lateral recess and lateral wall of the sphenoid sinus, which can be achieved with a transpterygoid, Vidian nerve-sparing, and supravidian approach [1]. Aggressive bone removal is essential and includes uncovering the anterior wall of the cavernous sinus extending lateral to the cavernous ICA up to the dural fold marking the transition between the middle cranial fossa dura and the parasellar region dura. Identification of the maxillary nerve entering the foramen rotundum may facilitate the localization of this fold. Such bony removal allows extension of the dural opening lateral and inferior to the anterior genu and horizontal ICA and anterior to the short ascending cavernous ICA segment. Infiltration of this compartment with tumor often leads to its expansion and facilitates this dural opening. Intraoperative use of Doppler ultrasound is very important to confirm the true course of the ICA before the dural opening is pursued. Our preference is to use a right-angled knife with a blunt tip to perform these dural cuts. As described above, the sympathetic nerves are encountered first when following a medial-to-lateral surgical corridor. Electrophysiologic stimulation in this compartment will facilitate identification of the abducens nerve lateral, inferior, and parallel to the horizontal ICA and will enable its distinction from the sympathetic nerve.

Surgical Nuances Associated with the Lateral Compartment of the Cavernous Sinus

This compartment is the least accessible from an endonasal perspective, as the cavernous carotid obscures this area when following a medial-to-lateral surgical corridor. Direct surgery into this area is performed only in select cases to avoid the risk of cranial nerve injury. Full exposure of the anterior genu and paraclinoidal ICA and anterior wall of the cavernous sinus laterally up to the superior orbital fissure is required to access this compartment. The optic and maxillary struts are ideal landmarks for the superior and inferior extent of the exposure, respectively. The dura is opened in front of the anterior genu of the ICA starting from the inferior compartment and advancing superiorly and anteriorly with a blunt tip, right-angled knife. Superiorly, the dura becomes adherent to the ICA as the dural rings meet the anterior clinoid process to form the clinoidal space. The use of a Doppler ultrasound and precise neuronavigation enables accurate mapping of the carotid artery. A surgical corridor needs to be developed between the anterior genu/horizontal ICA and the cranial nerves in the lateral wall of the cavernous sinus. Under normal circumstances, this corridor is very narrow. However, infiltration of this compartment with tumor displaces the cranial nerves laterally and the cavernous carotid medially, widening the surgical access. Although lateral displacement may also be true for the abducens nerve, this nerve is generally an exception, as it does not run in the wall of the sinus and may be embedded within the tumor placing it at a higher risk for injury. The arterial branches of the inferolateral trunk are encountered while developing this lateral corridor, and careful coagulation and division are required. In order to mobilize (from lateral to medial) the cavernous ICA and paraclinoidal ICA, the lateral aspect of the proximal ring (extending between the paraclinoidal ICA and lower aspect of the optic strut) requires partial sectioning. Tumor can be carefully removed between the cavernous ICA and cranial nerves with the assistance of electrostimulation to identify the oculomotor, trochlear, and particularly the abducens nerve. Unfortunately, tumors in the lateral compartment commonly involve the lateral wall of the cavernous sinus, in which case complete resection is not feasible without cranial nerve injury.

Surgical Limitations by Compartment

When reviewing our own experience with 98 pituitary adenomas extending into the cavernous sinus, we found 29 patients to have a single compartment invaded and 69 to have multiple compartments involved. The most commonly invaded compartment was the superior (79 patients), followed by the posterior (64), inferior (45), and lateral (23) compartments. Residual tumor rates by compartment were lateral (79%), posterior (17%), superior (14%), and inferior (11%). This was determined by subsequent postoperative imaging rather than intraoperative assessment. Two patients underwent further resection with subsequent complete tumor resection: one had residual ACTH-secreting tumor in the posterior compartment which grew on imaging, and the other patient had residual nonsecreting tumor in the inferior compartment which was reexplored with complete resection. Twenty-seven of those with residual tumor (74%) were treated with radiosurgery.

Surgical Nuances Related to Endoscopic Approaches to Meckel's Cave

Both transnasal and transmaxillary approaches have been described providing access to Meckel's cave [21]. In general, a transpterygoid approach is required. Coagulation and division of the sphenopalatine artery allows lateralization of the pterygopalatine fossa contents and identification of the Vidian canal and nerve [22]. Superior and lateral to the Vidian canal, the foramen rotundum and V2 are found. Exposure of the bone lateral to the parasellar carotid and inferior to the lateral opticocarotid recess exposes the superior orbital fissure, which provides access to V1. The bone between the superior orbital fissure and the foramen rotundum (a.k.a. maxillary strut) can be removed to provide circumferential access to V2. Removal of the lingual process lateral to the lacerum segment of the carotid artery provides access to Meckel's cave. The surgeon can keep dissecting V2 posteriorly, exploiting the interdural plane all the way back to Meckel's cave. This technique can be used to amputate V2 as proximal to Meckel's cave as possible while avoiding a CSF leak in cases of perineural invasion (such as with adenoid cystic carcinomas). Removal of the bone lateral to the foramen lacerum and the carotid artery, and inferior to V2 (the mandibular strut), provides access to the foramen ovale and V3. The vein of Vesalius or sphenoid emissary vein, which connects the cavernous sinus with the pterygoid plexus, can occasionally be identified here and may be a source of venous bleeding that can be easily controlled with gentle packing. Complete skeletonization of the paraclival carotid is performed when medial displacement will be required to reach the pathology or when proximal and distal control is needed [5].

The Vidian nerve extending posteriorly just lateral to the anterior genu of the petrous carotid artery is usually sacrificed in these approaches. The potential complication for xerophthalmia can be avoided with preservation of the Vidian nerve, although this can limit the exposure caudally and laterally toward V3. To gain access to Meckel's cave, the following landmarks should be readily identified: paraclival ICA medially, V2 laterally, and the horizontal petrous ICA inferiorly. Its superior boundary is formed by the abducens nerve, which runs obliquely and superiorly, medial and inferior to CN V1 within the cavernous sinus on its way to the superior orbital fissure. Staying below the superior margin of V2 is a good way to avoid injury to the abducens nerve, and the dural opening runs parallel to V2 from anterior to posterior [5].

7.5 Operative Setup and Instrumentation

The operating room setup we use for our endoscopic endonasal procedures always follows the team concept, two-surgeon, four-handed technique previously described [23, 24], in which both the neurosurgeon and the ENT surgeon stand on the right-hand side of the patient and the endoscope occupies the right superior nostril, a suction in the right inferior nostril, and other instruments in the left nostril. Our standard head positioning involves slight head extension, right lateral rotation, slight left tilt, and slight translation anteriorly and to the right. For lesions involving the right cavernous sinus or Meckel's cave, the head could be further turned toward the right, whereas for lesions involving the left side, the head can be positioned more or less neutral.

Neurophysiology is an essential component of operating within the cavernous sinus and Meckel's cave. We tend to routinely use somatosensory evoked potentials for all of our procedures. This ensures that head positioning does not result in any significant spinal cord compression or vascular insufficiency (especially in older patients with stiff necks in whom cervical spondylosis is more prevalent). When there is only limited invasion of the medial wall of the cavernous sinus, EMG monitoring of cranial nerve VI may suffice. With gross cavernous sinus invasions, we usually monitor EMGs for cranial nerves III, IV, and VI. Exploration of Meckel's cave mandates EMG of V3 and VI.

Image guidance is a useful adjunct for all endoscopic endonasal procedures. In addition to a contrasted MRI with image guidance sequences, we have found fine-cut CT angiograms to be particularly useful when dealing with cases involving cavernous sinus and Meckel's cave, as they provide a more accurate representation of the bony anatomy relative to the carotid artery.

Another essential tool for approaches involving the cavernous sinus and Meckel's cave is the Doppler ultrasound. Cross-reference between anatomical landmarks, image guidance, and the Doppler adds extra layers of protection when exposing the carotid artery as image guidance can sometimes not be accurate.

For exposures with significant lateral extension, angled endoscopes aid in visualization of areas in question. We typically use a 45-degree endoscope, although occasionally a 70-degree endoscope may be required. Furthermore, although adequate bone removal can allow lateralization of the carotid and the use of straight instruments, curved or malleable instruments (such as suctions) are often very helpful when working around corners.

Furthermore, we routinely use commercially available nasal protection sleeves in all our cases so as to minimize trauma to the nasal cavity, as well as decrease the contamination of the endoscope optics with blood.

7.6 Perioperative Care/Complications

Perioperative Care

Commonly, approaches that involve tumor resection from within either the cavernous sinus or Meckel's cave can be performed without CSF leaks if the tumors do not have true intradural extension. As such, reconstruction is often limited to a free mucosal graft and fibrin glue. For cases with a high-flow CSF leak, a nasoseptal flap is used. Based on the results of a randomized controlled trial performed at our institution [25], we no longer use lumbar drains for small sellar and perisellar defects. However, when these approaches are performed in the setting of large tumors with large clival or anterior cranial fossa defects, then we use a multilayered reconstruction using an inlay collagen matrix, a fascia lata graft, a fat graft, and finally the nasoseptal flap covered with fibrin glue followed by a lumbar drain for approximately 3 days.

Reconstruction in the setting of an exposed carotid artery and anticipated postoperative radiotherapy is controversial. Certainly, a vascularized nasoseptal flap provides the highest degree of protection. However, in the absence of a CSF leak, a free mucosal graft or a layer of fibrin glue usually suffices, and the nasal morbidity associated with a nasoseptal flap is avoided.

When operating within the cavernous sinus or Meckel's cave, 10 mg of dexamethasone is administered preoperatively and re-dosed intraoperatively for long procedures so as to minimize inflammation secondary to manipulation of cranial nerves. However, steroids are only continued postoperatively if the patient is found to have a cranial nerve deficit.

A third-generation cephalosporin (typically ceftriaxone) is used perioperatively, with a second-generation oral cephalosporin (typically cefuroxime) continued for patients with packing, until this is removed.

Complications

Complications related to approaches within the cavernous sinus are mainly related to injuries to cranial nerves III, IV, V1, and VI, while complications in approaches to Meckel's cave may result from injuries of the trigeminal ganglion and any of its branches (V1–3) and cranial nerve VI. The abducens nerve, running within the cavernous sinus, is the most likely to be injured. The use of neurophysiology and a detailed knowledge of anatomy can help decrease that risk. It is also important to remember that approaches to Meckel's cave may compromise the function of the Vidian nerve. The resulting decreased lacrimation may be dangerous in the setting of V1 dysfunction, increasing the risk for corneal keratopathy.

Vascular injuries are also a concern. Injury to the carotid artery is overall rare, but fibrous/invasive tumors increase the risk. A very important point for dealing with tumors within the cavernous sinus is that both the periosteal and meningeal layers of the dura fuse with the adventitia of the carotid artery at the superior extension of the carotid collar, close to the distal dural ring, where the relative protection of the venous channels between dural layers disappears. Therefore, the surgeon has to be extremely cautious when developing a plane between the tumor and the carotid artery in that area as forceful dissection may lead to penetration of the carotid wall. The attachment of both dural layers to the adventitia of the carotid artery inferiorly occurs at the foramen lacerum, something that becomes more relevant for approaches to Meckel's cave. More commonly, when operating within the cavernous sinus, one of the branches of the carotid artery, such as the inferolateral or the meningohypophyseal trunk, can be avulsed from the carotid wall and cause bleeding that is difficult to control; in fact, the origin of the meningohypophyseal trunk is located at the back side of the posterior genu of the cavernous ICA which is difficult to fully visualize.

It is important to remember that once arterial bleeding within the cavernous sinus is suspected, injectable hemostatic agents (e.g., Surgifoam or Surgiflo) should be avoided, as penetration of such agents into the carotid can be the cause of catastrophic strokes. Conversely, inadequate hemostasis can be the cause for postoperative hematomas. As such small cavernous carotid branches should be carefully identified, coagulated, and divided to avoid such events.

Reviewing our recent 5-year experience with 98 pituitary adenomas confirmed intraoperatively to invade the cavernous sinus, we found that no patient developed oculomotor or trochlear nerve palsies. Two patients had transient abducens nerve palsies. Three patients suffered from immediate postoperative hematomas causing symptomatic optic chiasm compression requiring surgical evacuation. The locations of tumor invasion in these patients were inferior/superior, superior/posterior, and in all four compartments. In two of these patients, vision improved significantly immediately after the clot evacuation, and both patients were returned to their preoperative baseline by the first postoperative visit. The third patient suffered from bitemporal hemianopsia up to 6 months after her surgery that was still worse than preoperatively. One patient suffered from internal carotid artery injury during GH-tumor dissection from the cavernous ICA wall; bleeding was controlled with local packing, but intraoperative SSEP (somatosensory evoked potential) recording showed significant drop in potentials suggesting ischemia and high risk of stroke if the vessel were to be sacrificed. Successful vessel preservation was accomplished using covered stents, and the patient did not develop any neurological sequelae postoperatively.

We also reviewed our experience with 17 primary cavernous sinus lesions [26]. The cohort consisted mainly of meningiomas (47%), hemangiomas (11.8%), and V1 schwannomas (11.8%). Tumors were located in the cavernous sinus with extension to the sella (35.3%), Meckel's cave (29.4%), and supraorbital fissure (11.8%). Clinical presentation was mostly a result of intracavernous cranial nerve compression and included diplopia (64.7%), trigeminal dysfunction (29.4%), retro-orbital pain (29.4%), and visual impairment (17.6%). In ten cases, the goal of surgery was biopsy and decompression. Among the other patients, gross total resection was achieved in two, near total (>95%) in two, and intentional subtotal (>50%) in three. Following EES, nine patients (52.9%) showed improvement of symptoms, six (35.3%) remained unchanged, one (5.9%) showed deterioration, and one died 3 days postoperatively because of stroke following intraoperative carotid injury. Other complications included new cranial nerve VI palsy (n = 1) and seizures (n = 1). No patient experienced a CSF leak.

Raza et al. reported their experience with endoscopic endonasal treatment of four patients with trigeminal schwannomas involving Meckel's cave. They reported one patient with posterior fossa disease experiencing sixth cranial nerve palsy in addition to a corneal keratopathy from worsened trigeminal neuropathy. There were no CSF leaks [27]. When reviewing our experience with nine trigeminal schwannomas with involvement of Meckel's cave treated with the EEA, we had four patients experiencing xerophthalmia and three experiencing corneal keratopathy. One experienced a CNVI palsy, and all but two had some degree of sensory or motor trigeminal dysfunction [28].

7.7 Surgical Pearls

- Adequate bony removal allows mobilization of the carotid artery and the cavernous sinus walls, providing access to areas that would otherwise be inaccessible.
- Familiarity with the endoscopic perspective of the anatomy is essential. Understanding the dural layers and ICA segments is critical. The interclinoidal ligament is a very important landmark that is often overlooked.
- Cross-reference between anatomical landmarks, image guidance, neurophysiology, and the Doppler ultrasound is crucial for avoiding neurovascular injuries.
- Endoscopic surgical procedures within the cavernous sinus and Meckel's cave are challenging and should only be undertaken when the team has gained some experience working together in simpler cases.

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Clivus and Upper Cervical Spine

8

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Abbreviations

CCJ	Craniocervical junction
CN	Cranial nerve
CSF	Cerebrospinal fluid
CT	Computed tomography
EEA	Endoscopic endonasal approach
EMG	Electromyographic
GTR	Gross total resection
HD	High definition
ICA	Internal carotid artery
MEP	Motor evoked potential
MRI	Magnetic resonance imaging
NSF	Nasoseptal flap
PPF	Pterygopalatine fossa
PPG	Pterygopalatine ganglion
PSA	Posterior septal artery
SSEP	Somatosensory evoked potential

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8.1 Introduction

Surgical access to the clival and occipitocervical region has long been fraught with numerous complications owing to a deep location, a complex anatomy, and difficulty in obtaining a watertight closure. To gain access to lesions in those deep locations, extensive skull base exposures, including the "far-lateral" suboccipital craniotomy as described by Heros et al. and the transoral approach, refined by Menezes [1], and subfrontal transbasal, transmaxillary, transmandibular, and preauricular infratemporal approaches, were for many years the only available options.

In the past two decades, the refinement and popularization of endoscopic endonasal techniques in neurosurgery have allowed an increasing number of lesions and pathologies located in the clivus and high cervical spine to be treated through a more limited exposure and straighter line of sight and surgical corridor. Jho and Carrau pioneered the endoscopic transsphenoidal surgery for pituitary lesions [2] but are also credited for the description of the first endoscopic endonasal resection of a clival chordoma [3]. In 2002, Frempong-Boaudu first described the use of endoscopic assistance in transoral surgeries [4]. Kassam et al. subsequently described the first case of odontoidectomy performed through an endoscopic endonasal approach in 2005 [5].

A major stride in endoscopic endonasal surgery has been the description of the Hadad-Bassagasteguy flap, or nasoseptal flap (NSF), in 2006 [6]. It has since become the first choice for vascularized mucosal coverage of large skull base defects, both for its ease of harvest and the large surface it can cover, and most importantly the reduction in the cerebrospinal fluid leak rate. Other options for vascularized reconstruction have since been described, and a better understanding has been gained of what is required in terms of closure technique depending on the anatomical location and clinical scenario.

In this chapter, we discuss the more recent advances in endoscopic endonasal, microscopic transcranial, and endoscopic-assisted transcranial approaches as applied to lesions in the lower clival and high cervical region. We will

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focus on the surgical treatment of clival and craniocervical junction chordomas, as these are the archetypal lesions encountered in this location.

8.2 Rationale

Numerous surgical approaches to the clivus and craniocervical junction (CCJ) region have been described, and obviously, no one approach is applicable and most appropriate for all lesions in this location. However, many of the pathologies frequently encountered in the clivus and the CCJ typically affect the ventromedial compartment, making the endonasal corridor the more direct surgical route, thus avoiding extensive manipulation of neurological and vascular structures. This fact may in part explain the generally more favorable results in terms of cranial nerve preservation obtained in endoscopic endonasal series when compared to series reporting on tumor resection performed through a posterolateral corridor [7].

As for high cervical lesions, the rationale behind the development of endoscopic endonasal techniques was the relatively high "cost" of transoral approaches to the ventral CCJ in terms of morbidity. In fact, in many cases, a palatal incision that would have been required in a transoral exposure can be avoided through an endonasal approach. Additionally, the mucosal incision is located higher when an endonasal route is utilized which potentially limits the infectious risk, quickens extubation, and allows earlier reintroduction of oral feeding. There is also reduced tongue swelling in the postoperative period since this approach does not require tongue retraction [8].

However, endonasal exposures of CCJ pathology often require resection of soft tissue and endonasal structures rather than manipulation or transposition as compared to classic transcranial open approaches. It thus involves additional morbidity with at least temporary reduction in quality of life. Additionally, the risk of CSF leak, even though reduced with modern reconstruction techniques, remains a significant issue especially in cases with an expected large dural defect, reoperations, and previously irradiated patients. Hence, the classic transcranial posterolateral approaches, including the far-lateral approach, retain some key advantages when dealing with the CCJ. In addition to the lower risk of CSF leak, posterolateral approaches do not involve resection of parapharyngeal muscles or the Eustachian tube, nor manipulation of the soft palate, allowing for complications such as velopharyngeal insufficiency and chronic serous otitis to be reduced or avoided altogether.

On the other hand, the deep central location of this anatomical region, deep to the soft tissues of the nuchal muscles, makes it difficult to reach through a small incision and keyhole approach similar to the subfrontal, mini-pterional or suboccipital keyhole approaches. However, it does not mean that the keyhole concept together with the endoscope cannot be utilized to reach via the classic far lateral corridor the ventral aspect of the craniocervical junction. Doing so, the "keyhole" is not located on the skin and convexity but is located deeper, at the level of the condylar region (Fig. 8.1a, b). Through a keyhole that is drilled or already made available by the tumor at the level of the condyle, the utilization of the endoscope is paramount to expose and resect parts of the lesion located higher at the levels of the lower- and mid-clivus, the prevertebral space, and infiltrating the odontoid, lateral

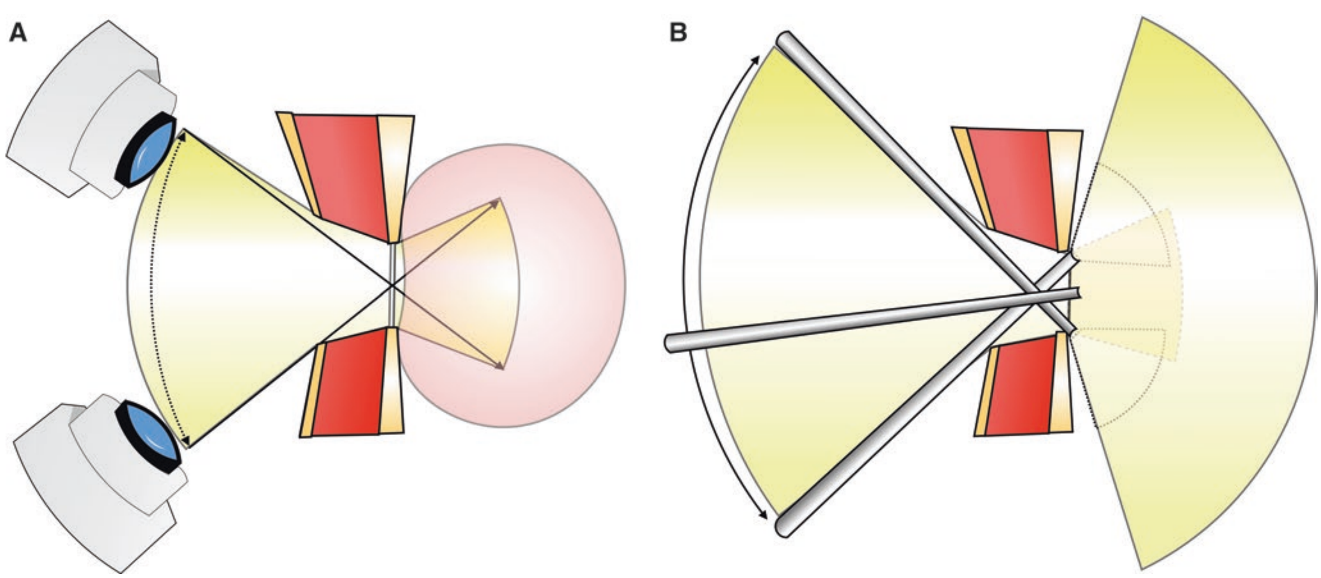

Fig. 8.1 (**a**, **b**). Keyhole concept in posterolateral approaches to the CCJ. (**a**). Exposure afforded by the microscope through a "keyhole" craniotomy. Soft tissues can impose significant limitations to the area visualized with blind spots under the edges of the craniotomy. (**b**). Increased viewing angle and illumination with the use of the endoscope inside the "keyhole," which is at the level of the craniotomy

masses of C1, and body of C2. This allows for equivalent tumor resection or neurological decompression through more limited exposures, which in turn is thought to be associated with less morbidity and shorter hospital stays.

Lastly, in some cases, the best approach is to combine endoscopic endonasal and transcranial techniques so as to benefit from the advantage of both and reduce the overall morbidity. In essence, modern skull base surgeons must utilize multimodal (microscopic and endoscopic) and multiportal (endonasal and transcranial) techniques to optimize clinical outcome.

8.3 Potential Pathologies

The most frequent lesions that neurosurgeons must manage in the lower clival and high cervical region are chordomas and degenerative diseases (rheumatoid arthritis and basilar impression). However, the differential diagnosis is quite varied (Table 8.1), and the clinician must be able to differentiate these preoperatively as the surgical management is often different from one lesion to the other.

Chordomas

Skull base chordomas are lesions arising from notochordal remnants in the clival region. They usually present as well-delineated multilobulated lesions of the midline cranial base, typically hyperintense on T2 and contrast enhancing. Notwithstanding their "benign" histology, these lesions tend to grow on a follow-up, causing osteolysis of the cancellous bone in which they are embedded, and, eventually, break through the cortical bone and invade the surrounding structures, such as the dura and the intradural compartment. They also have the tendency to infiltrate and grow along soft tissue planes, including between dural leaflets which makes gross total resection a true challenge. Clinically, clival chordomas most frequently present with a 6th nerve palsy, while lower cranial nerve dysfunction is also common at presentation at the level of the CCJ [9].

Although the literature has been somewhat conflicting in the past, there is now extensive evidence showing that gross total resection improves overall survival and progression-free survival in skull base chordomas [10, 11]. Another important prognostic factor to take into consideration is the quality of the resection at first attempt. In fact, in most surgical series, GTR appears harder to achieve in residual and recurrent disease than during the first surgical attempt. Every effort should hence be made to maximize resection during the first surgery. Chordomas of the CCJ have a poorer prognosis than those located elsewhere in the skull base or the cervical spine [9]. This finding may be related to the deep location and invasiveness of CCJ chordomas into the surrounding soft tissues and the resulting lower rate of GTR and higher recurrence risk.

Table 8.1 Differential diagnosis of clival and CCJ pathologies

Congenital	
	Malformations of the occipital bone (platybasia, condylar hypoplasia, etc.)
	Basilar invagination
	Irregular segmentation
Inflammatory and degenerative	
	Rheumatoid arthritis[a]
	Basilar impression[a]
	Infectious (osteomyelitis, Grisel's syndrome)
Tumor	
	Chordoma[a]
	Chondrosarcoma
	Meningioma
	Glomus tumor
	Schwannoma of the lower cranial nerves
	Other bone tumors (myeloma, osteoma, etc.)
	Secondary lesions
Vascular	
	Vertebrobasilar aneurysms
Trauma	
	Chronic instability
	Ventral compression

[a]Most frequent lesions

Chondrosarcomas

Chondrosarcomas are rare at the CCJ but it bears mentioning that their surgical treatment is somewhat more complex. In fact, they are often located more laterally and their consistency is more fibrous and they are frequently calcified. Most authors agree that, owing to the indolent nature of this tumor, GTR is not as important as for chordomas and priority must be given to the functional outcome. In some cases of complete resection, close follow-up may be considered. However, upfront adjuvant proton beam therapy is probably safer in case of incomplete resections. These management decisions must also take into account the grade of the chondrosarcoma.

Degenerative Diseases and Malformations of the CCJ

Although a number of degenerative diseases of the CCJ can be treated medically, with cervical traction or standalone posterior decompression, a number of indications for anterior decompression, including odontoidectomy, are still well established. Menezes et al. have published and advocated an algorithm-based management for these lesions in which the first line of treatment is generally reduction with cervical traction [12]. Also, in rheumatoid arthritis with anterior inflammatory pannus, there is usually significant improvement of the anterior compression after a posterior occipito-cervical fusion.

8.4 Patient Selection: Indications/Contraindications

The first and foremost consideration in achieving an optimal resection in lesions affecting the lower clivus and CCJ, especially chordomas, is choosing the optimal surgical corridor. Although chordomas in this location are often midline, they also have the tendency to extend laterally and invade different compartments and anatomical structures around the skull base. More often than other lesions, chordomas at the level of the CCJ present an asymmetric growth pattern and are often comprised of multiple loculations. On occasion, this means that the straighter surgical corridor may not be the one that allows the most complete resection (Fig. 8.2a–c).

A detailed study of the preoperative images should be done by an experienced multidisciplinary skull base team. In tumor cases, we delineate and record all extensions of the lesion to be resected. We give particular attention to extensions causing compression of neurological structures not well addressed by radiation therapy, such as the brainstem and optic chiasm, and those extending into bone or close to metallic reconstruction material.

On the preoperative CT scan, a number of important elements must be considered in order to establish if an endoscopic endonasal route is appropriate and will allow for complete resection and/or sufficient neurological decompression (Fig. 8.3a, b). First, the palatine line must be drawn and the level of the tip of the odontoid marked. In the majority of patients, the tip of the odontoid falls into a ±3 mm interval above or below the palatine line. The nasopalatine line, or Kassam line, between the rhinion and the posterior limit of the hard palate was first described to approximate the lower reach of an endonasal exposure. However, the nasopalatine line does not take into account the soft tissue retraction that is required to gain the down viewing angle and often overestimates the inferior limit of the exposure. We use the nasio-axial line (that connects the posterior limit of the hard palate with the midpoint between the rhinion and the anterior nasal spine) and has a better predictive value of the inferior limit of the dissection possible through an EEA [13].

The relationship of the lesion with the lower cranial nerves, namely, the 9th, 10th, and 11th nerves, at the level of the jugular foramen and the 12th cranial nerve in its hypoglossal canal is another critical element to establish preoperatively. In fact, these landmarks define the anatomical limitations to the coronal expansion of the endonasal corridor. They are also useful when considering a posterolateral approach as lesion compartments located ventromedial to

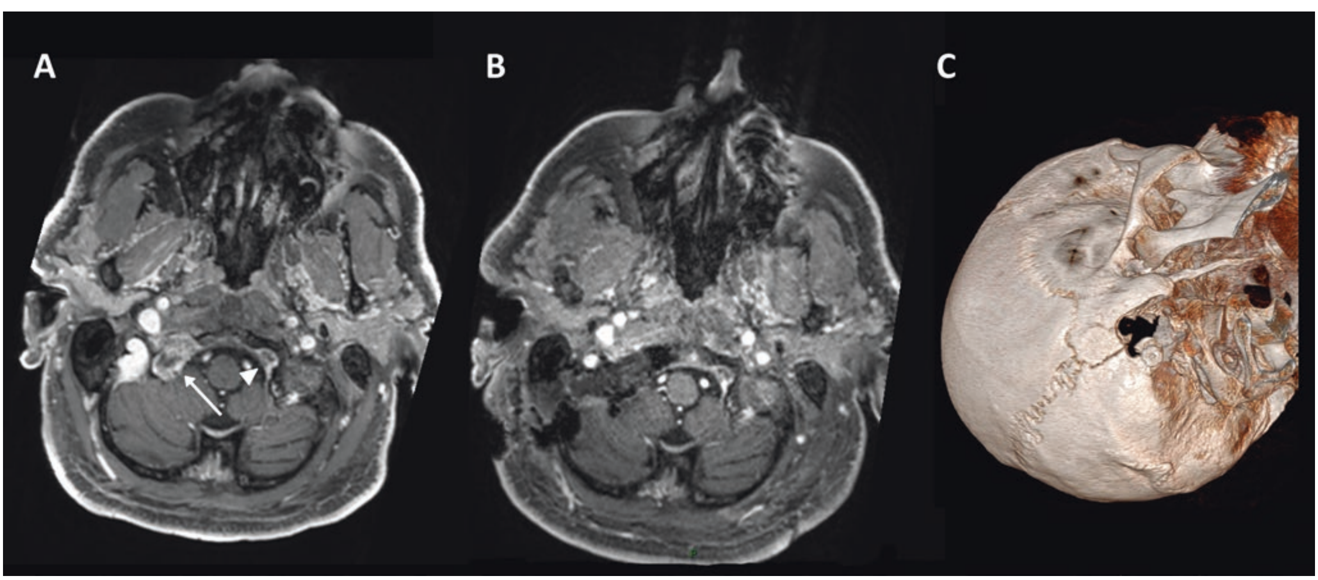

Fig. 8.2 (**a**–**c**). Chordoma presenting unusual extension pattern and resection through keyhole far-lateral craniectomy. (**a**). MRI of a 59-year-old patient presenting with a late recurrence of a right jugular foramen chordoma (*arrow*) with extension to the contralateral hypoglossal canal (*arrowhead*). Clinically, the patient had complete right hypoglossal nerve palsy. (**b**). Subtotal resection was achieved through a right keyhole far lateral approach with endoscopic assistance. Bony resection was done up until the level of the contralateral hypoglossal canal, but tumor was left in that location to avoid functionally unacceptable bilateral hypoglossal nerve palsy. (**c**). 3D reconstruction of the keyhole craniectomy

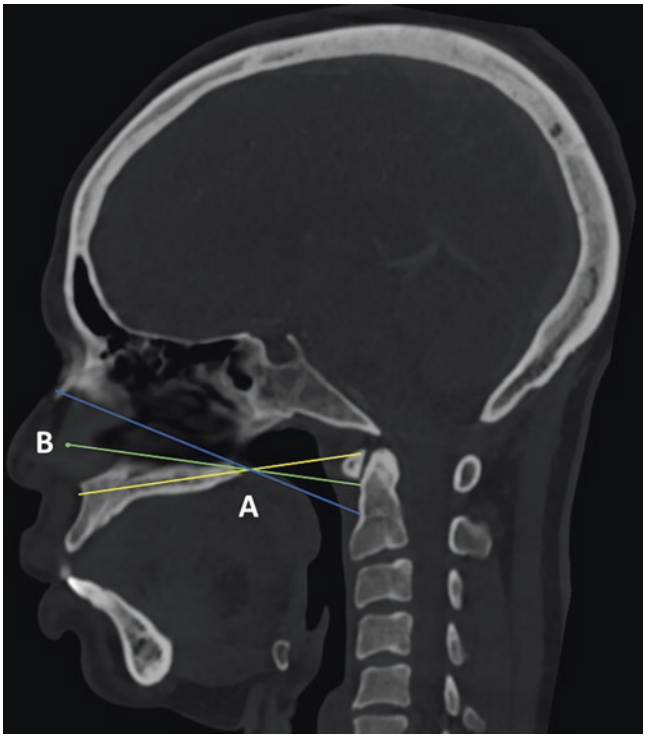

Fig. 8.3 (*AB*). Preoperative assessment of EEA reach. The tip of the odontoid falls into a ±3 mm interval above or below the *palatine line* (*yellow line*) in the majority of patients. The *nasopalatine line* (*blue line*) approximates the lower reach of an endonasal exposure, between the rhinion and the posterior limit of the hard palate (*A*). The *nasio-axial line* (*green line*), connecting the posterior limit of the hard palate (*A*) with the midpoint between the rhinion and the anterior nasal spine (*B*)

these are less well exposed and dissection and manipulation of cranial nerves could be required especially in case of an intradural extension medial to the cranial nerves. Neurological deficits, especially those involving cranial nerves and those that are deemed to have low rehabilitation potential, must be taken into consideration from a functional point of view in order to avoid catastrophic bilateral deficits such as bilateral hypoglossal palsy (i.e., case from Fig. 8.2a–c).

The surgical plan must take into account the patient's particular vascular anatomy and cerebral hemodynamics. In some cases, especially when there is encasement or suspected tumoral invasion of a major vessel, vertebral or carotid occlusion tests may be performed prior to surgical procedures.

Preoperative assessment of instability at the level of the CCJ with dynamic studies is required in all degenerative cases and some tumor cases. Even if the CCJ is deemed stable preoperatively, many cases of EEA and posterolateral approaches for CCJ chordomas that partially infiltrate the condyles mandate a posterior surgical stabilization and fusion surgery because of the additional resection of the condyle and periodontoid ligaments. In chordoma cases, fusion procedures should ideally be discussed and planned with the radiation oncology team as metallic reconstruction material may be an obstacle to proper radiation treatment. In some patients, we have postponed the stabilization as a second stage after completion of proton beam irradiation, with external immobilization of the CCJ using a rigid collar during the proton beam therapy. Another option is to use carbon nonmetallic material which has become available for such indications.

8.5 Surgical Anatomy

The use of technological adjuncts such as surgical navigation, cranial nerve monitoring, and Doppler ultrasonography, while useful and standard of care in such procedures, cannot replace a thorough and in-depth understanding of the anatomy of the anterior skull base and paranasal sinuses.

Osteology

The clival bone is formed by the basilar part of the occipital bone and, at its superior third, by the sphenoid bone. These two parts are joined at the spheno-occipital synchondrosis, just inferior to the dorsum sella. The clivus is separated laterally from the petrous part of the temporal bone by the petro-occipital fissure (or petroclival fissure). Inferolaterally, it is continuous with the condylar parts of the occipital bone that form the anterolateral edges of the foramen magnum along with the clivus (Fig. 8.4a). Classically, the clival bone has been divided into three segments – upper, middle, and lower – although the landmarks delimiting these segments have been subject to some variation from one author to another. From the endonasal perspective, we find it is most useful to divide the clival bone according to lines passing at the level of the sellar and sphenoidal floors (Fig. 8.4b), as this classification correlates well with critical neurovascular relationships of the clival bone with the carotid arteries and intracranial structures and is surgically relevant.

On the exocranial surface of the clivus, the pharyngeal tubercle is found approximately 1 cm above and anterior to the edge of the foramen magnum on the midline. Another reliable osseous landmark is the supracondylar groove, a depression corresponding to the attachment of the rectus capitis anterior, and usually a few millimeters inferior to the level of the pharyngeal tubercle. It is useful in approximating the height of the hypoglossal canal and its external orifice, which are located just posterior and lateral to the supracondylar groove, respectively [14]. The occipital condyles articulate with the lateral masses of the atlas (C1). Their long axes follow an anteromedial trajectory, and their articular surfaces have an oval shape and face anterolaterally.

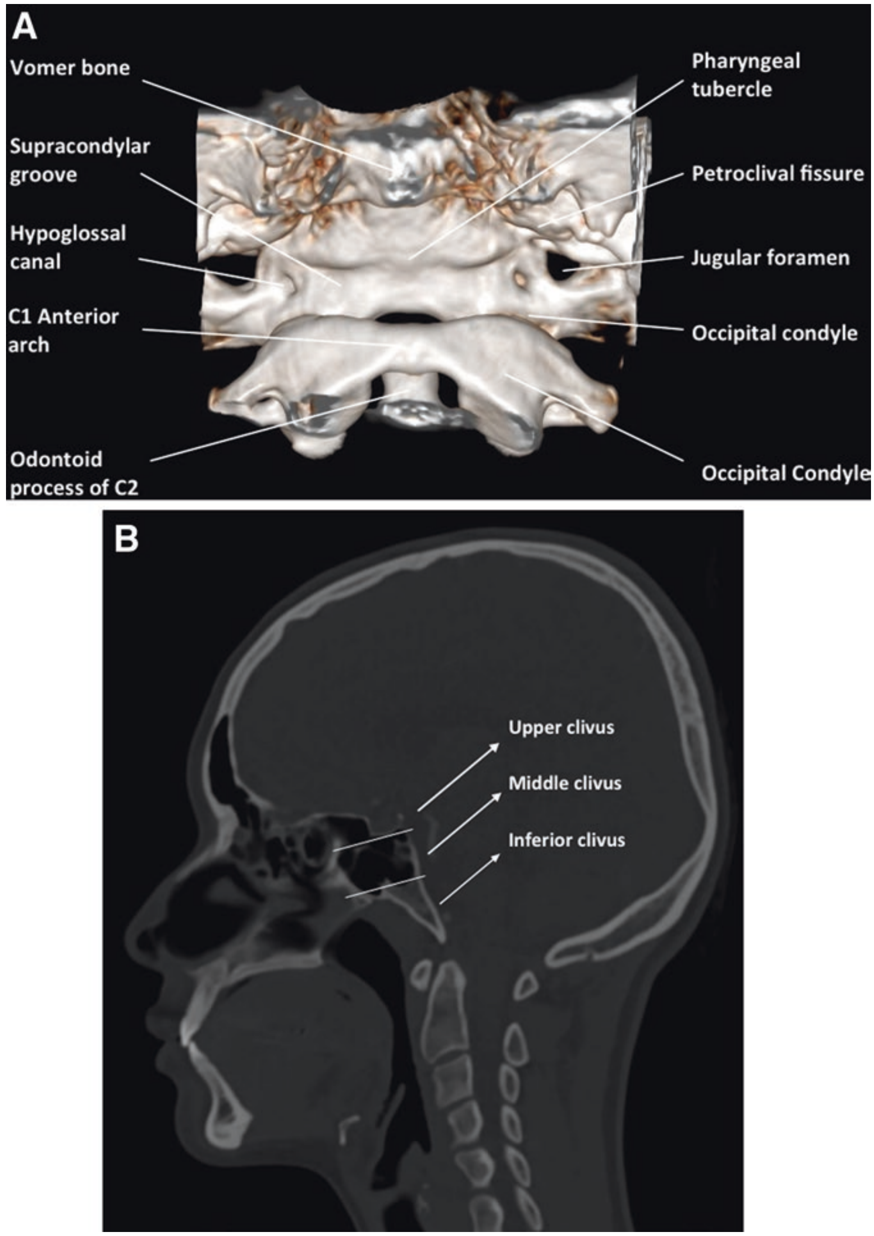

Fig. 8.4 (**a**, **b**). Clival and craniocervical junction osteology

The condyle contains the hypoglossal canal, which encloses the nerve of the same name. The intracranial opening of the hypoglossal canal is located in the middle third of the condyle, and the canal has an anterolateral but also a slightly upward trajectory placing the exocranial exit just above the level of the condyle.

Ligaments located in the CCJ are particularly important in maintaining the dynamic stability of the junction. The anterior longitudinal ligament (anterior surface of C1 and C2 with continuation inferiorly), the anterior atlanto-occipital membrane (between C1 and the foramen magnum), and the apical ligament (between the tip of the dens and foramen magnum) are found from anterior to posterior. Bilaterally, the alar ligaments connect the dens with the alar tubercles of the occipital condyles. The third ligamentous layer is formed by the cruciate ligament, with a vertical part and a horizontal part, also called the transverse ligament, attaching on the lateral masses of C1 on each side.

Endonasal Landmarks

Careful study of the endonasal anatomy on the preoperative CT scan helps the operator in anticipating the patient-specific anatomy and identifying some surgically relevant anatomical variations (e.g., pneumatization of the sphenoid sinus, presence of Onodi cells, or dehiscent carotid canals). During surgery, the first endonasal landmarks to identify are the choanal arches and the middle and inferior turbinates. The uncinate process, the bulla ethmoidalis, and the basal lamella can serve as guides to the superior aspect of the maxillary sinus, the medial wall of the orbit (lamina papyracea), and the posterior ethmoid, respectively.

When following the nasal septum posteriorly, the rostrum of the sphenoid and the vomer bone are palpated. The ostium of the sphenoid sinus is found approximately 1.5 cm above the level of the choana. Just below passes the posterior septal branch of the sphenopalatine artery, which can be traced back to the sphenopalatine foramen. Below the choana, the openings of the Eustachian tubes are located anteriorly and inferiorly to the torus tubarius on the lateral wall of the nasopharynx (Fig. 8.5a). A line drawn between the upper edges of the Eustachian tube approximates the level of the foramen magnum. In the midline, the muscular insertion of the longus capitis muscle (which overly and insert superiorly to the rectus capitis anterior) can be seen as a V-shaped prominence under the mucosal surface. The center of the prominence corresponds to the pharyngeal tubercle [15].

Neurovascular Structures

Knowing the precise localization of the ICA is imperative at all times during endonasal procedures. The ICA is fixed in bony structures at its petrous (C2), lacerum (C3), and clinoidal (C5) segments but is mobile and easily displaced by tumors or other pathological processes in its cervical (C1) and cavernous (C4) segments. Moreover, anatomic variations of the ICA are frequent, including loops and anomalous locations, especially in the cervical segment where it is located deep in the soft tissues of the neck and where it is at risk of injury during expanded endonasal

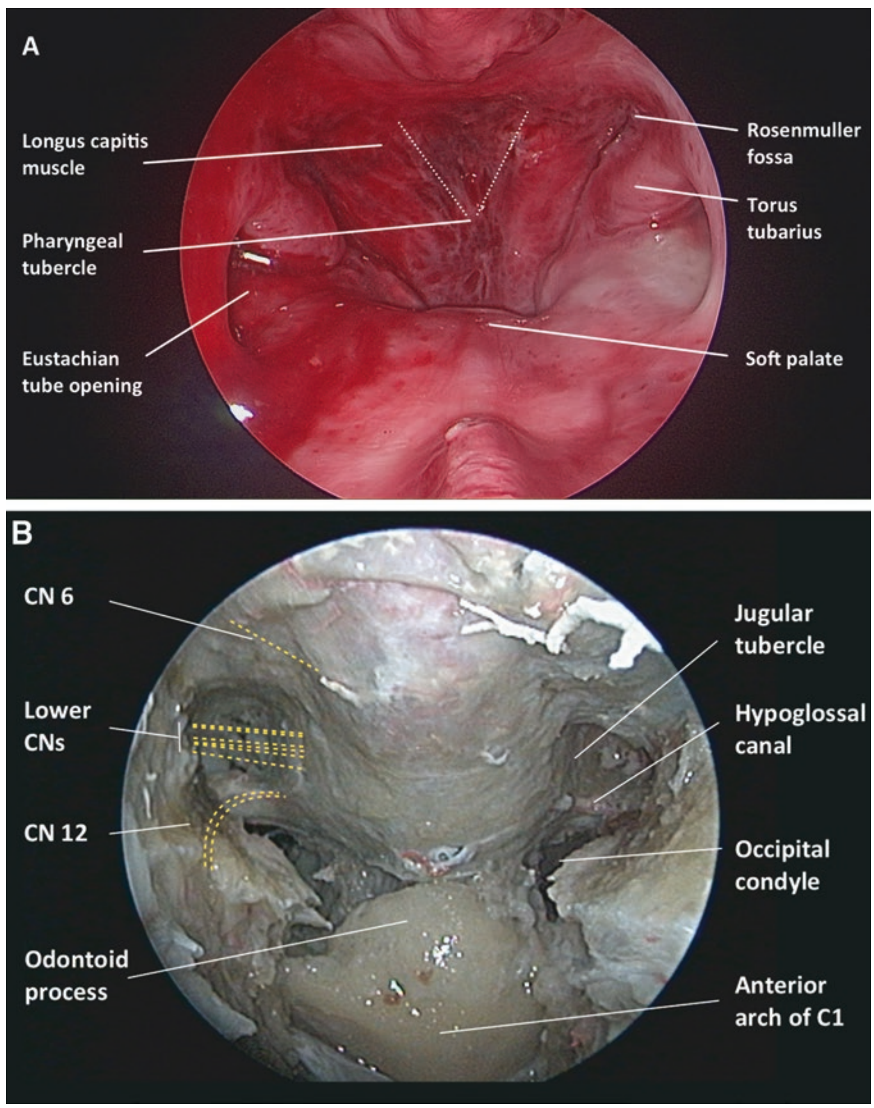

Fig. 8.5 (**a**, **b**). Endoscopic anatomy (cadaveric dissections)

procedures. It is thus critical that anatomical variations be recognized preoperatively on a CTA [16]. The paraclival ICA can be easily identified in many cases where there is a well-defined clival recess inferior to the sella. The vidian nerve can serve to define the depth and location of the ICA (C3 segment) as it terminates at the inferolateral edge of the lacerum segment of the ICA, between the petrous segment (C2) and posterior ascending segment (C4, or paraclival). Before its course in the petrous bone, the parapharyngeal ICA, or distal cervical segment of the ICA, can usually be found posterolateral to the deepest portion of the fossa of Rosenmuller. Although it remains a segment at risk, the parapharyngeal ICA is difficult to locate precisely intraoperatively. It is posterior to the lattermost extent of the cartilaginous part of the Eustachian tube and posterior to the levator veli palatini [17]. The parapharyngeal ICA position can also be approximated by a line going through the anteroposterior axis of the lateral pterygoid plate, which can also be confirmed on the preoperative CTA.

The abducens nerve, or CN VI, the main neurological structure at risk during endoscopic endonasal exposures, emerges on the medial aspect of the pontomedullary sulcus and after a superolateral cisternal course, enters Dorello's canal, an interdural space. The nerve courses under the petrosphenoid ligament (Gruber's ligament) and runs more horizontally lateral to the posterior ascending or paraclival segment of the cavernous carotid to enter the cavernous sinus where it ascends again immediately inferior and lateral to the anterior ascending segment of the ICA. The glossopharyngeal nerve (CN IX) courses in a separate compartment of the jugular foramen than the vagal (CN X) and accessory (CN XI) nerves, the anteromedial *pars nervosa* and the posterolateral *pars vasculosa*, respectively. The hypoglossal nerve (CN XII) arises at the pre-olivary sulcus and has an upward and lateral trajectory toward the hypoglossal canal. After its exit from the exocranial end of the canal, it follows the lateral margin of the rectus capitis anterior muscle (Fig. 8.5b).

8.6 Surgical Technique

Endoscopic Endonasal Approach

As in other complex interventions, a clearly defined surgical strategy is a key factor in gaining proficiency and improving the safety of the procedures performed. Elements to be defined preoperatively can be broken down into three main elements: (1) exposure, (2) resection, and (3) reconstruction [18]. As previously mentioned, the main difficulties in endoscopic endonasal approaches are control of the 6th and 12th cranial nerves and the parapharyngeal ICA. Optimization of the exposure is done by establishing which surgical corridor is best suited for the lesion to be treated (covered in this chapter under "Patient Selection"), but also by planning for extensions of the approach that can improve tumor exposure and neurovascular control. This is part of the "modular" or "building blocks" philosophy that is well described for EEA.

To achieve more complete resections, it is imperative to define all extensions of the tumor according to certain anatomic landmarks (i.e., paraclival carotid artery, hypoglossal canal, odontoid process, etc.). We often prepare a list of targets preoperatively with associated surgical maneuvers or extensions to the approach to perform in order to reach each tumor sub-compartment.

Reconstruction is often the step that leads to the most problematic complications postoperatively, and therefore it must be planned for and executed with careful attention. It is important to consider if previous surgeries were performed and, if so, how reconstruction was achieved and if the mucosal flap can be reutilized. In some cases, a laterally extending approach is required, and a NSF cannot be utilized on that side and must be harvested contralaterally. In situations in which both sphenopalatine arteries are compromised, an alternate vascularized grafting material should be sought for and planned. For example, in some cases with multiple previous surgeries and/or radiation therapy, the best reconstruction material is a temporoparietal fascia flap that mandates a different positioning and preoperative preparation. This particular flap also requires a maxillary antrostomy and transpterygoid dissection, which can also be used to increase the working space inside the nasal cavity. However, in those cases, where the closure may be challenging and requires additional cranial incision and work, a classic transcranial approach should also be considered.

Operative Setup and Patient Positioning

The positioning of the patient should allow for a comfortable and ergonomic position for both surgeons. An ideal setup is comprised of two high-definition (HD) screens, each facing one of the surgeons, who are usually on the right side of the patient (for right-handed surgeons). We also like to have the scrub nurse positioned in front of the surgical team on the other side of the patient to allow for safe and easy handling of the instrumentation. The anesthesia team is usually at the feet of the patient.

The head is positioned in a fixed head holder to avoid surgeon disorientation and loss of optimal positioning. More importantly, the head is flexed, rotated, and tilted toward the side of the surgical team. The rotation and tilt are especially important to center the head in the field of view and action of the surgeons. A practical rule of thumb to estimate the degree of tilt required is to align the patient's head with the ipsilateral nipple. Slight head flexion is also strongly recommended as it grants better exposure of lower clival and CCJ pathologies (unless a temporoparietal fascia flap is needed). To ensure good venous outflow and to reduce venous bleeding, we usually elevate the head and thorax, in addition to a slight reverse Trendelenburg positioning of the surgical table. The abdomen and/or anterolateral thigh are prepared for harvesting a fat and/or fascia lata graft to repair the bony and dural defects.

Instrumentation

In our practice, we have taken up the habit of using almost exclusively 4 mm 30° and inverted 30° scopes (Karl Storz™) in our endoscopic cases. Using the angled scopes from the beginning of the surgery grants the surgeon an almost instinctive understanding of the anatomy under angled visualization, allowing him to make full use of the advantage of endoscopy in terms of panoramic exposure and leaves more room for instrument maneuvering away from the scope. The 45°, 70°, and the EndoCAMeleon® scopes (Karl Storz, Tuttlingen, Germany), with an adjustable viewing angle, are also very helpful in reaching lesions located inferiorly or laterally, typically behind the horizontal segment of the ICA.

Angled and length-appropriate instrumentation are a must in extensions of the endoscopic endonasal approach. While the angled endoscopes provide the visualization, angled instruments, especially malleable rotatable suction-tips, provide the necessary maneuverability to resect laterally reaching compartments of the lesion. Long angled and self-irrigating drill bits are also useful in reaching the lower clival and upper cervical regions.

In all EEA cases, we use surgical navigation with both MRI (T2 weighted or CISS/FIESTA and T1 weighted with gadolinium) and CT imaging fusion. Similarly, the microvascular ultrasound Doppler probe is used systematically to accurately localize the internal carotid and sphenopalatine arteries. The precision of the navigation system should be confirmed regularly during the procedure and must be used carefully and in conjunction with anatomical landmarks.

Another technological adjunct indispensable in EEA is electromyographic (EMG) monitoring of cranial nerves. In chordoma cases, we usually monitor at least both 6th nerves. Additionally, the 5th nerve is usually monitored on the side of a transpterygoid approach, as well as the 12th nerves (bilaterally) and lower cranial nerves (10th and 11th), depending on the tumor extensions and planned surgical exposure. An alternative to EMG monitoring of the nerves innervating the extraocular muscles is electrooculography (EOG) that can be used to detect vertical and horizontal movements of the eye globe.

Technique

In many lower clival cases, a two-surgeon, four-hand technique is employed. The obvious advantage of this setup is the finer dissection that can be accomplished with two instruments. A binostril access and wide septostomy and sphenoidotomy are required to gain adequate maneuverability for both surgeons. However, we now strive to limit as much as possible the endonasal morbidity and have used a uninarial access in many, if not the majority, of our most recent endonasal lower clival cases (detailed below under Minimally Invasive Endonasal Access and Mucosa-Preserving Surgery).

The first step, similar to every endonasal case, is to identify the relevant anatomy and key landmarks, including the choanal arches, the middle and inferior turbinates and sphenoid ostia. A middle turbinectomy can be done on one side, usually on the right, as it provides increased space for the endoscope and suction but it is not always required. The inferior turbinates are preserved in most cases, but it may be useful to down-fracture them to enlarge the endonasal corridor when aiming at lower lesions. The mucosa of the inferior turbinate can also be a valuable vascularized flap option for reconstruction if the NSF is not available [19]. The middle turbinectomy is followed by a maxillary antrostomy, accomplished by identifying and resecting the inferior aspect of the uncinate process. The medial wall of the maxillary sinus is then resected from front to back with a backbiting instrument

A posterior ethmoidectomy facilitates identification of the anatomical landmarks relevant to the harvest of the nasoseptal flap and is helpful when the lesion to be treated is located higher up in the clivus or invades the sphenoid sinus. The bulla ethmoidalis is resected, and the basal lamella of the middle turbinate is opened to gain access to the posterior ethmoid. The lamina papyracea, which is the lateral wall of the bulla, is identified and serves as the lateral limit of the ethmoidectomy. Identifying the sphenoid ostium before resecting the basal lamella is valuable in preserving the posterior septal artery (PSA) and a large mucosal pedicle for the NSF.

Harvesting the NSF is done in a standard fashion, with particular attention given to preservation of the olfactory mucosa. It is also important to try and tailor the flap to the expected dural defect and structures to be covered. Extending the NSF by including nasal floor mucosa and mucosa of the inferior meatus and planning a contralateral NSF flap are options that must be entertained and prepared at this stage of the operation. As discussed previously, when a transpterygoid dissection is planned, the NSF is most often harvested on the contralateral side. Otherwise, it is necessary to free the flap's pedicle along with the sphenopalatine fossa, after sectioning the vidian nerve in order to be able to lateralize the flap sufficiently. The harvested NSF is placed in the maxillary sinus and held in place with the aid of large cottonoids. At this point, a wide posterior septectomy is done, followed by resection of the keel of the sphenoid bone, either with the drill or osteotomes, which have the added benefit of providing bony material for closure. The sphenoid anatomy is explored and the opening is widened to identify the lateral recesses of the sphenoid bilaterally, the paraclival internal carotid arteries (ICA), the sella and optic nerves, the optico-carotid recesses, and the clival recess.

Extensive drilling of the floor of the sphenoid sinus and the clival bone can then be done quickly with a large coarse diamond drill, while keeping track of the exact location of both ICAs and after appreciating the depth of the dural plane. A diamond drill and continuous irrigation should be used when drilling over the ICA and dura mater, preferentially with a large drill bit.

To access the lower third of the clivus and the CCJ, partial/total resection or downward retraction of the muscles attaching to the clivus, namely, the longus capitis and the rectus capitis anterior, is required. In chordoma cases, there are frequently tumor loculations in these muscles, and partial resection of the pre-clival soft tissues is often preferable to achieve gross total resection. In this region, the most reliable landmarks are the Eustachian tubes, which indicate the lateral limit of the surgical exposure and help define the midline. The parapharyngeal ICAs are usually located lateral and posterior to the Eustachian tubes, but this must be verified on preoperative vascular imaging to avoid being surprised during surgery by a medially oriented vascular loop.

Opening and resection of the dura depends on the location and type of pathology. In chordoma surgery, it is often possible to predict if there is dural transgression with T2-weighted images on MRI. During surgery, when the dural transgression is not obvious, indirect signs are the absence of venous bleeding from the venous channels of the clival plexus after completion of clival drilling and splitting of the dura's layers. It is not clear if resection of dural "margins" is required or if it has a survival benefit. When there is no clear dural penetration, coagulation of the attachment base is undertaken while avoiding heat injury to the abducens nerves. In some instances, it is possible to find a dissection plane between the chordoma-infiltrated periosteal layer of the dura mater and an intact meningeal layer, which may allow for a complete resection without dural breach. Radical resection of the dura is reasonable only if complete resection of the intradural extension is achievable, which is not always the case due to brainstem infiltration. In those cases, it is better to keep the dural opening as small as possible to facilitate closure.

Chordoma Resection (Fig. 8.6a–l)

Chordomas are soft, easily aspirated lobulated tumors. The use of an ultrasonic aspirator is often not necessary, although a rotating aspirator tip may prove useful, particularly if used under angled endoscopic visualization. It is not always necessary to perform an extracapsular dissection since there is no "true" tumor capsule in most chordomas. Piecemeal resection often allows safer tumor removal without compromising GTR. The pseudocapsule that is oftentimes identified is usually a conjunctive inflammatory reaction around the tumor that can be aspirated along with the tumor. In the rare occurrences that the chordoma is found to be fibrous, we have found that GTR is hard to achieve safely since these lesions are also more adherent to the ICA and cranial nerves.

During tumor resection, every precaution should be taken to limit tumor seeding in the CSF, but even more so through the surgical corridor. In fact, these tumors are known to have the propensity for metastasizing through both routes, and as a general rule, intratumoral debulking must be maximized before intradural exploration. We also try to limit irrigation when dealing with the intradural compartments of the lesion. As for surgical corridor dissemination, it can probably be minimized by careful inspection and abundant irrigation at the end of the case when the reconstruction has been satisfactorily carried out and the intradural compartment effectively sealed. Likewise, in order to reduce the occurrence of avoidable tumor seeding, biopsy of clivus lesions, when imaging features are highly evocative of chordoma, should be avoided in most cases.

Since chordomas arise from notochordal remnants in the cancellous bone of the clivus, it is important to resect as much of the involved bone as possible. This aspect of the resection is too often neglected. It is reasonable to assume that delayed recurrences after GTR could be due to chordoma cells left in the cancellous bone surrounding the initial "visible" tumor.

Minimally Invasive Endonasal Access and Mucosa-Preserving Surgery

In some cases, it is possible to achieve gross total resection with a uninarial technique and with minimal disruption of the endonasal anatomy (Figs. 8.6a–l and 8.7a, b). While the occurrence of true "empty nose" syndromes is rare, limiting endonasal morbidity clearly improves patient quality of life in the postoperative period. Undoubtedly, the binarial transseptal approach can be "maximally invasive" for the endonasal anatomy, and such extensive dissections are not required in the majority of cases. Nonetheless, going through one nostril and limiting one's approach must not be done at the expense of the quality of the resection.

Uninarial access usually provides sufficient room for only one surgeon, although a third hand to control the suction may be useful in certain stages of the surgery (e.g., drilling and fine dissection). In uninarial access, the hand navigating the endoscope can also be used to hold and control a rotatable suction tip, leaving the dominant hand with the control of another instrument. Although this technique often used by the senior author requires some adaptation and learning curve, it is especially useful in minimally invasive endonasal exposures. Using this technique, the conflict between other instruments and the endoscope is limited, and the endonasal structures that are kept intact serve as a funnel

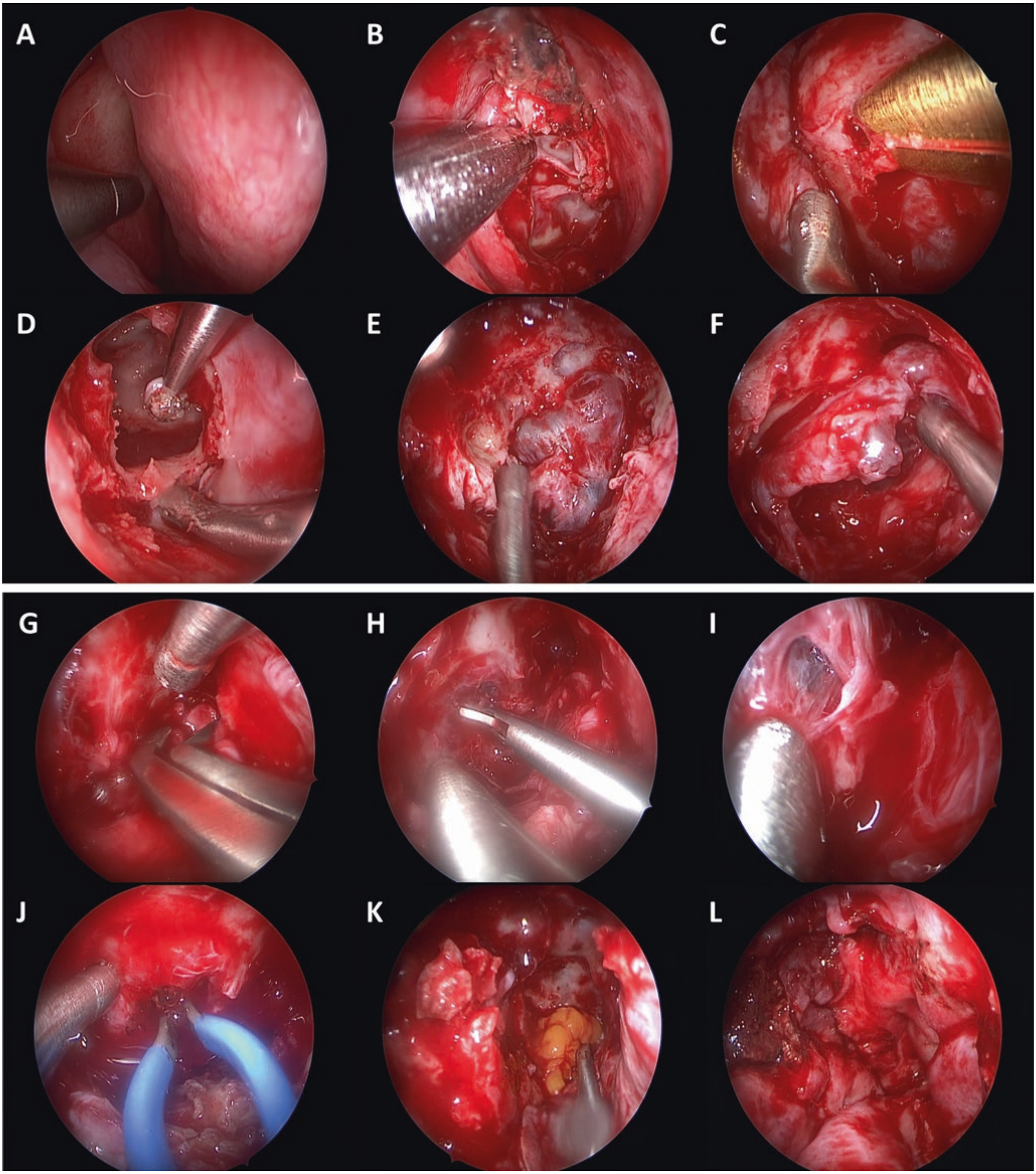

Fig. 8.6 (**a**–**l**). Endoscopic resection of a chordoma of the lower clivus. 56-year-old women with a clival chordoma. Step-by-step description of the endoscopic endonasal exposure and resection of the tumor through a uninarial corridor. (**a**). The middle turbinate is outfractured after identification of the endonasal anatomy. (**b**). After middle turbinectomy, a posterior ethmoidectomy is undertaken with the shaver. (**c**). Posterior septectomy with preservation of the contralateral mucosa. (**d**). A large sphenoidotomy is completed with a large coarse diamond drill and Kerrison rongeurs. (**e**). Extensive drilling of the clivus is done, exposing the extracapsular plane of the tumor. (**f**). The chordoma is debulked, taking care to limit tumor spillage to avoid surgical corridor seeding. (**g**). In lower clivus chordomas, such as in this case, extensions in the longus capitis and rectus capitis anterior muscles must be sought. (**h**). The dura mater is split into its two layers to make certain that all the tumor extensions have been resected. (**i**). A small area of dural defect if exposed with the underlying arachnoid. (**j**). Coagulation of the dural and muscular margins after gross total resection. (**k**). A fat graft is placed to fill the drilled clival bone. (**l**). A NSF is harvested on the contralateral side and is placed over the fat graft and supplemented with fibrin glue

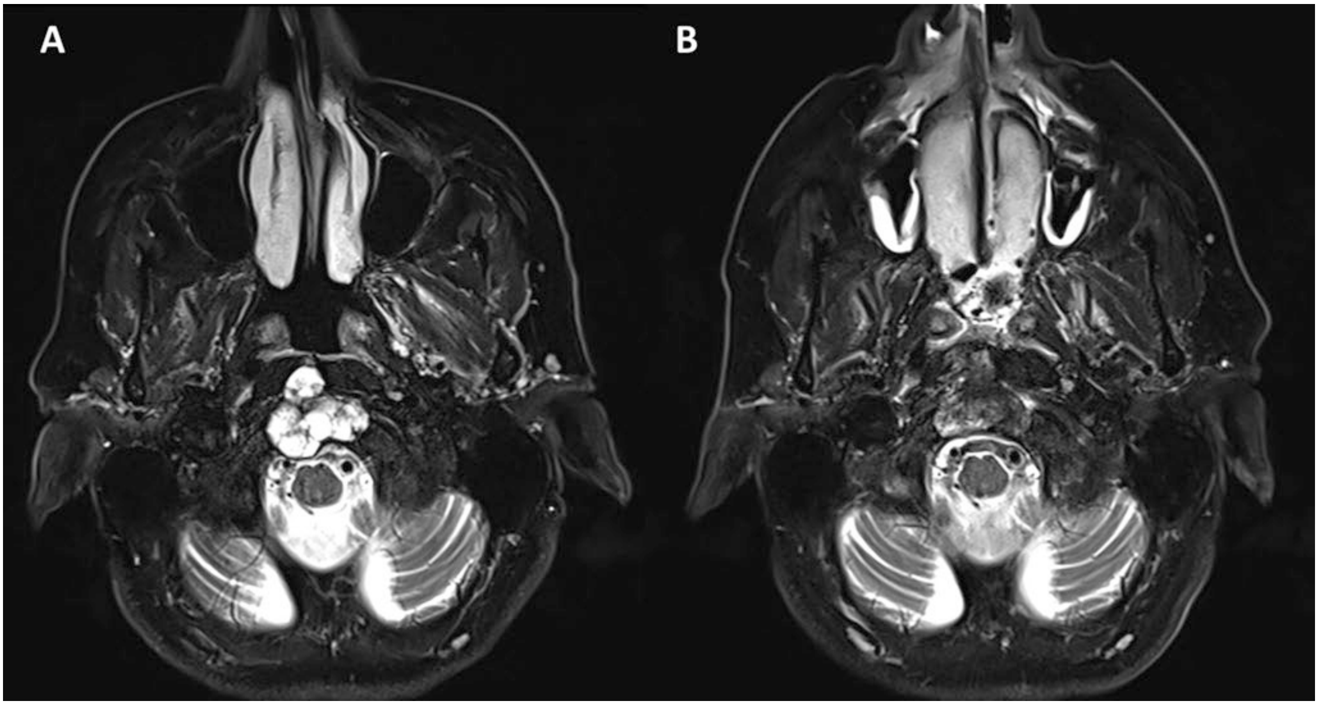

Fig. 8.7 (**a**, **b**). Tailored endoscopic endonasal resection of clival chordoma. (**a**) Preoperative and (**b**) postoperative images of an endoscopic endonasal resection of a chordoma of the lower third of the clivus. Gross total resection was achieved through a left uninarial approach with preservation of the anterior two-thirds of the nasal septal mucosa bilaterally and all turbinates except the left middle turbinate

for the endoscope. The narrower the corridor is, the easier it is to manipulate the instruments. Another advantage is to have the endoscope extremely close to the tip of the instruments.

A contralateral trajectory in lesions extending preferentially on one side may be a valuable option. To gain adequate access to lower clival lesions through one nostril, a middle turbinectomy is usually necessary. The posterior bony septum can be partially resected, but a simple pushover on the other side of the septum along with the contralateral mucosa generally suffices. A wide sphenoidotomy is unavoidable, however, to provide room for both the endoscope and instruments. A posterior ethmoidectomy can also be quite useful to gain additional space to maneuver the instruments without interference from the endoscope. If a NSF is required, it must be harvested on the side of the exposure, which also means that a transpterygoid approach is not feasible if the sphenopalatine artery is to be preserved.

Extending the Surgical Corridor Laterally

Extending the corridor laterally also means that the ICA will be exposed and will have to be managed on a longer segment. A transpterygoid approach helps gain the lateral visualization required to better define the ICA's course. After a wide maxillary antrostomy, the posterior wall of the maxillary sinus is resected. The pterygopalatine ganglion (PPG) and surrounding soft tissues are kept inside the periosteum, which is dissected laterally, and the greater palatine nerve is identified going down. This nerve has a predictable downward trajectory and can be skeletonized out of its bony canal to allow for increased mobilization of the pterygopalatine fossa (PPF). Its sacrifice can result in uncomfortable hypoesthesia in the hard palate and should be avoided if possible, especially if only the superior aspect of the pterygoid has to be removed. The sphenopalatine branch is identified, coagulated and sectioned at its exit from the sphenopalatine foramen after elevating the mucosa anterior to the foramen. After mobilizing the PPF laterally, the vidian canal bundle is progressively identified. Section of the vidian nerve allows for increased lateral mobilization of the PPF. Frequently, before identifying the VN, another bundle is seen, composed of the palatovaginal artery and nerve, which have a more medial direction. It should not be confounded with the vidian canal bundle. The root and medial plate of the pterygoid can then be drilled to increase access inferolaterally in the sphenoid bone and expose the foramen lacerum.

The transpterygoid approach allows better identification of the lacerum segment of the ICA and to gain lateral working space in the middle clivus and petrous apex areas. However, to obtain a more lateral exposure lower in the clivus or at the CCJ, downward mobilization or resection of the Eustachian tube must be done with drilling of the medial plate of the pterygoid. After identification of the foramen lacerum, and clearly defining the limits of the anterior genu of the ICA with a micro-Doppler, a cut in the cartilaginous portion of the Eustachian tube is done below the lacerum segment.

Extending the Surgical Corridor Inferiorly

To increase exposure down to the lower third of the clivus and the upper cervical region, certain simple maneuvers can be done such as outfracturing the inferior turbinates and putting slight retraction on a stitch placed in the soft palate. However, to be able to decompress and resect pathology in the CCJ, some specific landmarks are useful to extend the bony resection to its neurovascular boundaries. One of such landmarks is the supracondylar groove (or inferior clival line), which marks the approximate height of the hypoglossal canal. To better delineate this groove, the muscles are either dissected subperiostally or resected. Drilling the anteromedial aspect of the condyle and thus skeletonizing the hypoglossal canal lead to inferior delineation of the jugular tubercle, which separates the hypoglossal canal from the jugular foramen. While the hypoglossal canal can be exposed through a midline corridor, unroofing the jugular foramen often requires a lateral dissection with a transpterygoid approach and resection of the Eustachian tube. In certain cases, where the condylar bone has been invaded and eroded, this exposure is already done by the tumor, and simple internal debulking is enough to expose both the lower cranial nerves and the 12th nerve in their dural sleeves. Lateral limits to bone drilling and intradural exposure are the inferior petrosal sinus, which courses along the intracranial aspect of the petroclival fissure and the vertebral artery when the drilling involves C1 and C2.

Endoscopic Odontoidectomy

To access the odontoid and perform an endoscopic anterior high cervical decompression, a less invasive endonasal approach is often sufficient. Through a binostril access, we begin by outfracturing the inferior and middle turbinates. In certain cases, partial posterior septectomy may be required to gain adequate exposure posteriorly and under the choanal arches. To increase the inferior angle, a stitch is placed in the soft palate and retracted through the mouth.

The nasopharyngeal mucosa and underlying muscle can either be reclined in a "U"-shaped flap or incised in the midline and dissected laterally. In our opinion, the midline incision is probably the most direct and least traumatic option and often allows for sufficient exposure and satisfactory closure. It can also be easily extended if need be. The pharyngeal tubercle is a reliable landmark to approximate the level of the foramen magnum (1 cm below). Under the longus capitis, the anterior atlanto-occipital membrane is either dissected laterally or resected. At this point, the anterior arch of C1 is visible and can be drilled to expose the dens. After disconnecting the odontoid process from its ligamentous attachments, it is removed with an "eggshell" technique. The apical, alar, and transverse ligaments are then resected, taking particular care in preserving the underlying dura.

After careful hemostasis, and if no CSF leak has been encountered, it is generally sufficient to line the surgical cavity with absorbable hemostatic agents (e.g., Surgicel©, Ethicon, Somerville, NJ, USA). If there is a CSF leak, a NSF is harvested and positioned over the dural defect, after multilayered closure (TachoSil© [Baxter Healthcare Corp, Deerfield, IL, USA] and a fat graft).

Reconstruction

A well thought through reconstruction is an essential part of the preoperative surgical strategy. Whenever there is a dural defect to reconstruct, a multilayer technique is employed. We first put fascia lata as onlay or with a "gasket seal" technique. Fibrin glue and TachoSil© are used as sealants and holding material. In the clival area, a fat graft is placed in virtually every case to fill the void left by the clival drilling. This probably reduces the occurrence of encephalocele over the long term, which can be quite dramatic in this location with anterior displacement of the brainstem or the vertebrobasilar system. Finally, a vascularized flap is placed over the fat graft and serves as the main element contributing to long-term watertight closure.

The mainstay in terms of vascular graft is obviously the NSF. However, there are other options if the nasal septum has been compromised by previous surgery or radiation. In some instances, we have used a "U"-shaped nasopharyngeal mucosa flap, based inferiorly on branches of the ascending pharyngeal and the Eustachian branch of the accessory meningeal artery and rising as high up as the sphenoid ostia and extending laterally to the level of the sphenopalatine foramina. If the mucosa is thick enough, an inferior turbinate flap can be elevated in place of a NSF. As a last resort, we have used the temporoparietal fascia flap, which can be tunneled in the nasal cavity through the pterygopalatine fossa. We do not favor nasal packing in the majority of the patients, although in some cases, we use a silastic patch to hold the vascularized graft in place.

Keyhole Concept in Posterolateral Approaches to the Lower Clivus and High Cervical Region

The keyhole concept can also be applied to posterolateral approaches to the clivus and CCJ, including the far-lateral craniotomy and high lateral cervical approaches. In these skull base exposures, due to the intervening muscular layers to be dissected and the vascular anatomy to be managed, it is not always possible to use limited skin incisions. However, minimal invasiveness is not only about limiting the skin incision. It must be remembered that the keyhole is not necessarily placed at the surface. Following a “hockey stick” incision and exposure of the occipital bone and condyle, a deep keyhole, located in the condylar fossa, for example, can provide access to a large segment of the middle and lower clivus and C1 and C2 vertebrae (Fig. 8.1a, b) with the assistance of the endoscope to browse this area.

We have been recently revisiting our surgical strategy in lower clival lesions, especially in cases where the dural defect is expected to be large and the CSF leak risk deemed high. In some revision cases, reconstructive options may be limited. We also have preferred transcranial approaches in cases where, based on imaging studies, an intracranial component of the lesion appears adherent to the brainstem or neurovascular structures and the microsurgical dissection stage is expected to be complex. The posterolateral transcondylar route is a good option in such cases. In situations where an occipitocervical fusion is necessary, this route also has the advantage of using the same midline skin incision. Conversely, the fusion procedure can also be utilized as an occasion to perform additional tumoral resection from a different trajectory.

Technique

The posterolateral transcondylar approach can be tailored to be both minimally invasive and give maximal exposure of the pathology at hand. We have used both limited skin incisions (“lazy S” and “C shaped”) and the classic “hockey stick” incision for these approaches, depending on the surgical trajectory. We have found the “hockey stick” incision to be helpful in cases where the vertebral artery was involved with the tumor (either encased or displaced) and in tumors with an extension higher up in the clivus. With a well-devised approach, the surgeon can reach tumor extensions located as high as the upper third of the clivus and within the sphenoid sinus. We also use the “hockey stick” incision when an occipitocervical fusion is required.

If a small skin incision is used, the muscular stage of the surgical dissection needs to provide a flat surgical field. In a way, the dissection of the muscles of the retromastoid and condylar fossa is similar to that of the temporalis muscle when performing a pterional or orbito-zygomatic craniotomy. To do so, we first disinsert and recline the sternocleidomastoid muscle from the mastoid, followed by the splenius capitis and longissimus capitis muscles. Underneath, the posterior belly of the digastric muscle and the muscles of the suboccipital triangles can be identified. The superior oblique muscle can be disconnected from the occipital bone to expose the retrocondylar fossa.

At this point, important bony landmarks should be identified, the mastoid tip and digastric groove anteriorly and the occipital condyle and the C0-C1 joint and the foramen magnum medially and posteriorly. The third segment of the vertebral artery is located at this stage. A small inferiorly placed retrosigmoid craniotomy can then be performed, with subsequent drilling of the posterior part of the mastoid to increase exposure of the sigmoid sinus down to the jugular bulb.

In addition to classic skull base techniques such as drilling the jugular tubercle and mobilization of the vertebral artery, the use of endoscopic assistance has also proven to be an invaluable tool to increase the reach of these approaches (Fig. 8.1a, b). The endoscope should be used as a tool to navigate and operate in the depths of the triangles defined by the lower cranial nerves. In chordoma cases, this is safely accomplished in the extradural compartment, in the space left by the drilling or tumoral osteolysis of the condyle and jugular tubercle, superiorly to the 12th nerve and inferomedially to the jugular bulb and inferiorly to the 12th nerve and superiorly to the third segment of the vertebral artery (Fig. 8.8a, b). The endoscope increases both illumination and visualization in these deep locations and can help reach as far as the petrous apex and sphenoid sinus through the condylar keyhole. In soft tumors, the surgeon can easily aspirate and debulk tumors compartments with the endoscope in one hand and a malleable suction tip in the dominant hand.

Role of Neurophysiology

In addition to electromyographic (EMG) monitoring of the 6th, 9th, 10th, and 12th nerves, motor and sensory evoked potentials are utilized in cases of brainstem compression. In some tumor cases, including chordomas, the clival bone is eroded and bony landmarks are lost. In these situations, direct stimulation with an EMG probe can be useful in cranial nerve identification and preservation (especially 6th cranial nerve). Neurophysiological monitoring with somatosensory evoked potentials (SSEPs) and motor evoked potentials (MEPs) also has a pivotal role during the surgical positioning of unstable high cervical lesions.

8.7 Postoperative Care / Complications

In the postoperative period, patients must be monitored for the development of CSF leak, meningitis, and neurological symptoms. We do not routinely leave lumbar drains or continue antibiotics after surgery, although we do occasionally perform evacuative lumbar punctures if patients complain of headaches or in cases of ventricular dilation on postoperative imaging. All patients should have an MRI in the first 48 h following tumor surgery to assess the extent of resection and allow, when indicated, a second surgical stage to be discussed or proton beam therapy to be planned without delay. Specific complications of the EEA approach to the lower clival and the CCJ region include velopharyngeal insufficiency (especially when the soft palate in split), CSF leak, meningitis, and wound dehiscence and infection [20].

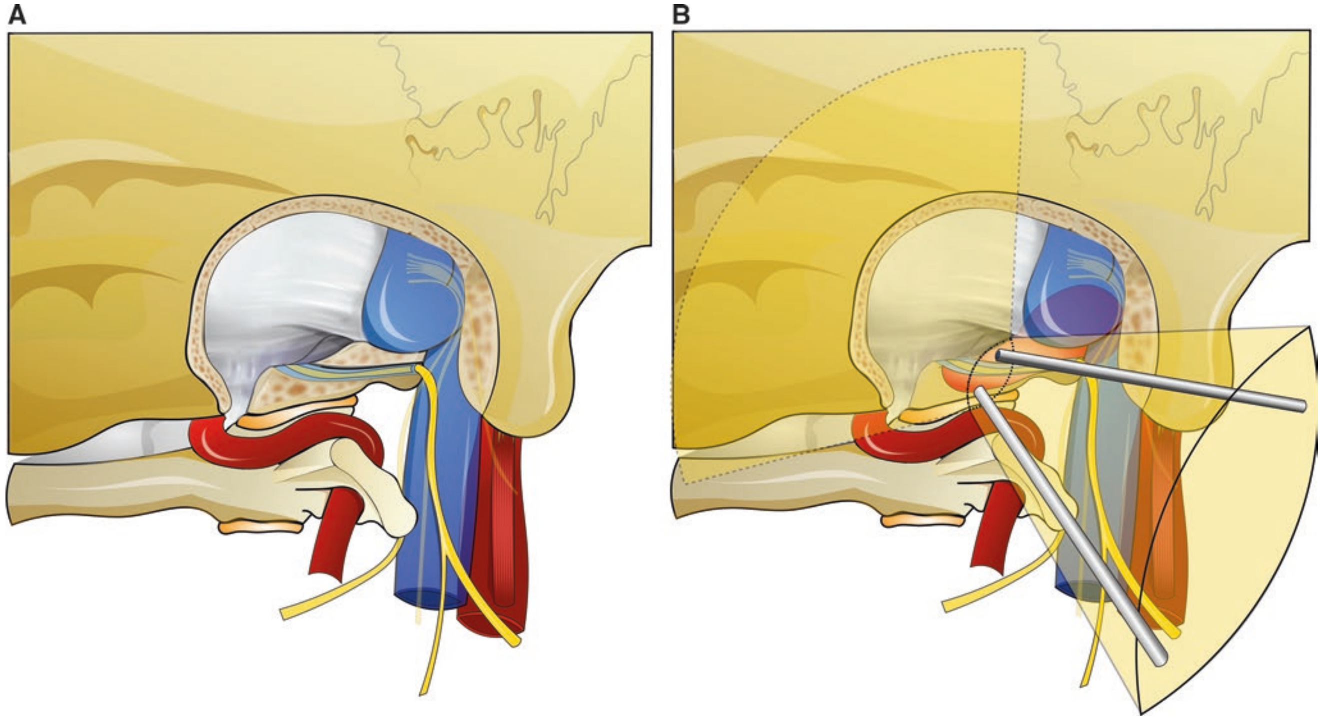

Fig. 8.8 (**a**, **b**). Anatomy and endoscopic windows through extradural triangles in the retrocondylar fossa. (**a**). Exposure of the retrocondylar fossa and identification of the C0-C1 joint, the third segment of the vertebral artery, the sigmoid sinus and jugular bulb, and the suboccipital and foramen magnum dura. The 12th nerve, inside its dural sleeve, has been exposed after drilling of the occipital condyle. (**b**). In the extradural compartment, two windows can be used to access lesions involving the lower and middle thirds of the clivus and the dens and lateral masses of C1. The first space is located superiorly to the 12th nerve, inferomedially to the jugular bulb, and is limited medially by the dura. The second window is inferior to the 12th nerve, superior to the third segment of the vertebral artery, and is also bounded medially by the dura

Endoscopic follow-up for control of mucosal healing, flap intake and vascularization, and removal of crusting must be done during the initial hospitalization and then at regular intervals until satisfactory healing is documented. If a CSF leak occurs in the immediate postoperative period, our first line of treatment is usually bed rest and lumbar drainage, unless the leak is deemed to be "high flow." In such cases, immediate surgical reexploration is undertaken, and graft repositioning or replacement is done. After anterior endoscopic odontoidectomy, the patient is fed through a nasogastric tube placed under direct visualization at the end of the procedure for a period of 5 days to allow for wound healing.

Carotid Injury

Intraoperative internal carotid injury is the most feared complication of EEA in lower clival and CCJ pathologies. A CT angiography should be considered systematically in these cases as it offers a precise evaluation of the main intra- and extracranial vessel trajectories (ICA in all its segments, vertebral and basilar arteries, posterior and anterior communicating arteries) as well as their relationship with the tumor. The need for proximal control of the ICA prior to tumor resection must also be anticipated especially when the tumor is extending laterally and posteriorly to the paraclival ICA. In such cases, an additional transpterygoid approach, following the vidian nerve posteriorly toward the lacerum segment of the ICA, may be necessary before tumor resection. It is important to take into account tumor pathology and invasiveness, as well as prior irradiation, as these factors may be associated with weaker arterial walls and a higher injury risk.

The initial response should be to initiate resuscitation efforts and prepare for massive transfusions on the anesthesia side as well as regain visualization and control of the surgical field, usually with packing. Surgical hemostasis is then obtained, with options including tamponing with crushed muscle, direct suture repair, or clipping. Once hemostasis has been achieved, an immediate angiographic assessment should be sought. Endovascular options in the acute setting are either ICA occlusion or endoluminal reconstruction. Variables to consider when deciding which to choose include the extent and morphology of ICA injury, surgical control of the bleeding, patient factors (age, comorbidities, etc.), angiographic findings (active bleeding, pseudoaneurysm width and morphology, etc.), vascular anatomy and collaterals and, most importantly, expected tolerance of antiplatelet therapy.

Craniocervical Junction Instability

Preoperative assessment of dynamic stability of the CCJ is required in the majority of cases, irrespective of the etiology. When clivectomy and complete condylectomy or odontoidectomy have been undertaken, posterior stabilization is indicated. In cases of partial condylectomy, opinions differ between authors as to the amount of occipital condyle that can be resected before instability is encountered. A recent cadaveric study has suggested that a threshold of 75% anterior condylectomy may be associated with clinical significant hypermobility [21]. Ligamentous resection can also be responsible for instability at the CCJ, particularly when alar ligaments or transverse ligaments are rendered incompetent.

It has been our practice to undertake the posterior stabilization in the early days after the anterior stage in most cases. Some authors advocate doing the posterior fusion in the same surgical sitting, while others even recommend that the fusion precede the anterior decompression. In certain chordoma cases, we even prefer doing the fusion after the proton beam therapy, since the artifacts from the instrumentation have a significant impact on the planning and delivery of the radiation therapy. Neuromonitoring, including lower cranial nerves, MEPs, and SSEPs, is useful during the positioning for the second surgical stage.

8.8 Surgical Pearls

- A detailed study of preoperative imaging is imperative to perform safe endoscopic endonasal procedures. It allows for identification of anatomical variants (including the ICA), inventory of all tumor compartments to be resected, and planning for reconstruction material.
- Precise understanding of ICA anatomy is crucial during endonasal procedures. Key landmarks for its location are the vidian nerve and the Eustachian tube.
- In many endoscopic endonasal approaches for clival lesions, it is possible to achieve gross total resection through a uninarial access and with minimal disruption of the endonasal anatomy.
- The keyhole concept can be applied to posterolateral approaches to the clivus and CCJ, including the far-lateral craniotomy and high lateral cervical approaches with the help of endoscopic assistance.

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Part II

Keyhole Cranial Surgery

Principles of Minimally Invasive Keyhole Surgery

9

Varun R. Kshettry, Tyler J. Kenning, James J. Evans, and Christopher J. Farrell

9.1 Introduction

Axel Perneczky is widely credited with pioneering the concept of keyhole neurosurgery based on the goal of minimizing approach-related surgical morbidity by choosing the optimal trajectory for each specific lesion [1]. The supraorbital keyhole craniotomy as initially described was performed through an eyebrow incision with the location of the keyhole varied anatomically to achieve a more medial subfrontal trajectory or lateral "pterional" trajectory based on the surgical target location along the anterior cranial fossa and parasellar region. Subsequent variations have included an orbital rim osteotomy to attain a more basal approach, alternative incisions to improve cosmesis, and the addition of novel keyhole approaches to access lesions in diverse locations including the pineal, cerebellopontine angle, subtemporal, and subcortical regions (Fig. 9.1a, b). Despite multiple variations, the concept and goals of keyhole neurosurgery remain constant.

Although keyhole surgery is often considered minimally invasive, the primary objective is to achieve maximal surgical efficiency by avoiding unnecessary exposure [2]. Choosing the optimal trajectory to a surgical target allows for less cutaneous and muscular disruption, smaller bony openings, and reduced potential for collateral damage to neural structures that are impertinent to the exposure. With less exposure-related injury, patients can experience more rapid and complete recovery with secondary benefits of potentially fewer infections and improved cosmetic outcome. Although Cheng et al. provided quantitative verification of the keyhole concept, demonstrating similar areas of exposure with the supraorbital keyhole compared to the frontotemporal "pterional" approach, the main disadvantage of the keyhole approach is the limited surgical freedom provided by the restricted corridor [3]. Failure to tailor the opening to achieve the optimal trajectory can lead to a loss of anatomic landmarks, increased need for brain retraction, and decreased vascular control. Because the openings are limited, the ability to correct for suboptimal trajectory and manipulate working angles is more difficult. For these reasons, these approaches are best utilized by experienced neurosurgeons possessing a thorough understanding of surgical anatomy, the ability to anticipate the optimal working angles for specific lesions, and the skills necessary to work efficiently in limited space.

In order to more fully realize the advantages of keyhole neurosurgery and expand its application, surgical illumination and instrumentation have rapidly evolved. Image-guidance systems have shortened the learning curves associated with these procedures, facilitating reduced operative times and avoidance of major complications. The use of single-shaft instruments allows for improved operative manipulation within a limited space while the use of various angle endoscopes allows for increased illumination and multidirectional viewing within the narrow predefined corridor. Additionally, preoperative planning based on three-dimensional imaging of the bony, vascular, and neural structures allows for tailoring and optimization of the approach. As technological advances in surgical simulators continues to progress, their routine incorporation into preoperative planning will allow for enhanced anticipation of the key structures relative to the target pathology. Similarly, advances in surgical robotics will enable improved instrument manipulation within the keyhole corridors and further minimize disruption of non-lesional anatomy. As the adoption of these techniques becomes more widespread, it is incumbent upon

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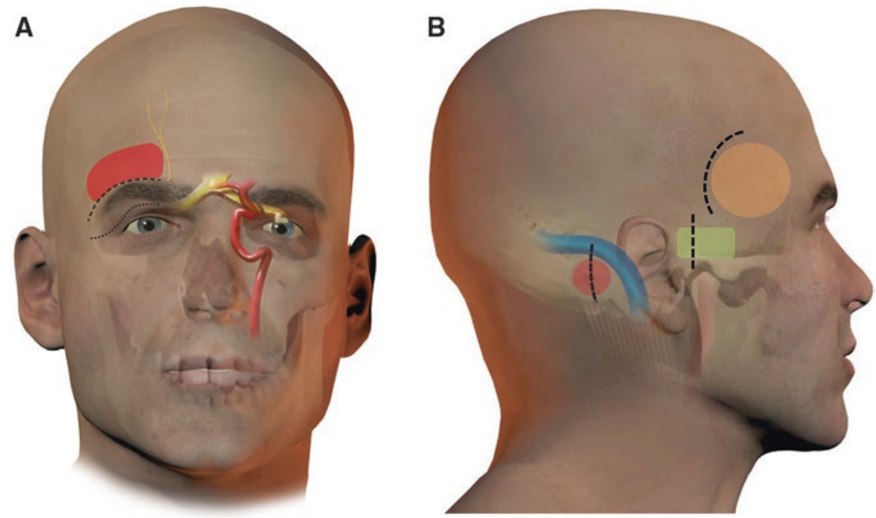

Fig. 9.1 (**a**–**b**) (**a**) Illustration depicting the supraorbital keyhole approach. The *shaded region* represents the size and location of the craniotomy, extending medially to the supraorbital nerve. The approach may be performed via either an eyebrow incision (*wide-hatched line*) or eyelid (*dotted line*) incision. (**b**) Size and location of the mini-pterional (*orange*), subtemporal (*green*), and retrosigmoid (*red*) keyhole approaches

neurosurgeons to more thoroughly define the indications for these procedures through prospective comparison of keyhole and conventional approaches with regard to immediate and delayed complications, neurocognitive outcomes, cosmesis, and functional recovery time.

In this introductory chapter, we provide a brief overview of the more standard keyhole approaches including their indications, advantages, and shortcomings. As with any surgical approach, the keyhole approaches discussed here serve as a foundation that should be tailored on an individual basis to best access the target pathology.

9.2 Supraorbital Keyhole Approach

The supraorbital keyhole approach (Fig. 9.2a–d) serves as an alternative to the traditional pterional and bifrontal craniotomies and is used primarily to access targets along the anterior cranial fossa floor and parasellar regions as well as intraparenchymal lesions involving the anterior frontal lobes. The additional removal of the lesser sphenoid wing can provide a more lateral subfrontal trajectory and is a variation often termed the lateral supraorbital keyhole approach. Each of these approaches is typically performed through either an eyebrow transciliary or supraciliary incision. An alternative transpalpebral "eyelid" incision has also been described which avoids cutting across the frontalis muscle, which may potentially lead to eyebrow asymmetry (Fig. 9.3a–d) [4]. This transpalpebral approach may require the assistance of an oculoplastic surgeon, and, unfortunately, no objective evaluation of cosmetic outcomes between the various incisions has been performed at this time.

The technical steps of the supraorbital keyhole approach are more extensively discussed throughout this book in dedicated chapters. While soft tissue and bony disruption are clearly reduced by the restricted craniotomy and smaller incision, in order to be truly minimally invasive, the approach must not come at the expense of increased brain retraction. Therefore, optimal patient positioning and placement of the craniotomy are critically important. We typically extend the head slightly to allow for gravity retraction of the frontal lobes and rotate the head contralaterally 10–45° depending on the location of the surgical pathology and lateral extension of the frontal sinus, utilizing neuronavigation to help plan the trajectory. The incision extends laterally from the supraorbital notch and may be continued beyond the eyebrow into a skin crease if necessary. A one-piece craniotomy is then turned lateral to the frontal sinus if possible and flush with the anterior cranial fossa floor. The orbital rim may be included in the craniotomy to increase the working corridor; however, we have found that thorough drilling of the excrescences along the orbital roof, progressive opening of the arachnoid, and drainage of cerebrospinal fluid provide sufficient working room in the majority of cases. Meningiomas arising from the olfactory groove, planum sphenoidale, and tuberculum sellae are well visualized via the supraorbital approach. While skull base reconstruction can be performed using a vascularized pericranial graft, the pericranial layer is necessary to help cover the plating system and improve cosmesis. Tumor extension medial to the ipsilateral optic nerve and inferiorly within the olfactory grooves and sella may require endoscopic assistance to expand the field of view and allow for more complete resection. The surgical corridor also allows for inspection and manipulation of the anterior communicating artery complex as well exposure of the A1 segments of the anterior cerebral arteries bilaterally and ipsilateral ICA [5].

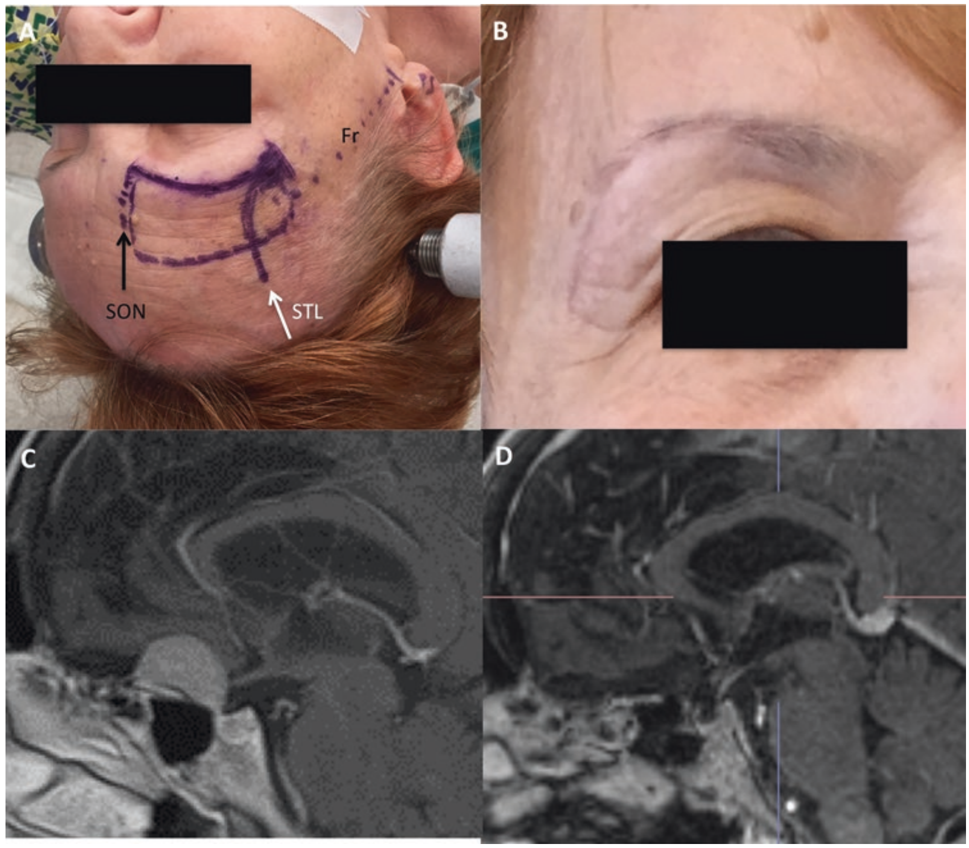

Fig. 9.2 (**a**–**d**) Case illustration of transciliary supraorbital keyhole approach for a planum meningioma. (**a**) Patient positioned with head rotated 30° and vertex extended for frontal lobe gravity retraction. Incision marked along the eyebrow along with markings for the supraorbital nerve (*SON, black arrow*), superior temporal line (*STL, white arrow*), and anticipated course of frontalis branch of the facial nerve (*Fr*) using Pitanguay's line. (**b**) Healing at 6 weeks postoperatively. Preoperative (**c**) and postoperative (**d**) sagittal T1 with contrast demonstrating planum meningioma with gross total resection (Copyright retained by Dr. Varun R. Kshettry. Used with permission)

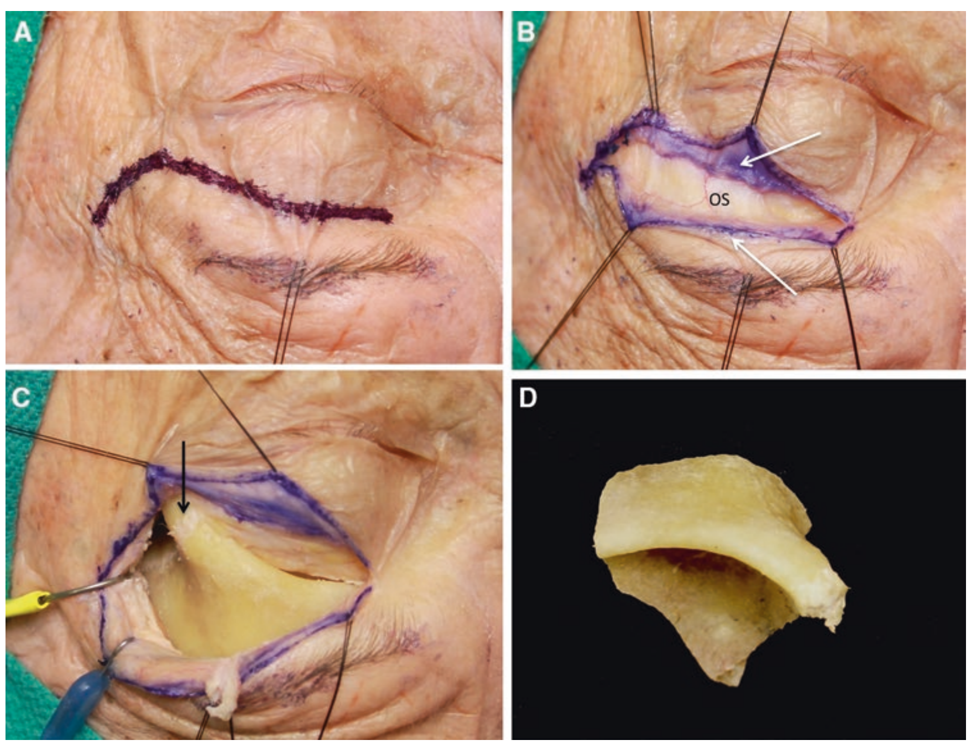

Fig. 9.3 (**a**–**d**) Cadaveric illustration of transpalpebral orbitofrontal craniotomy. (**a**) Incision marked. (**b**) Initial subdermal dissection with spreading of orbicularis oculi fibers (*white arrows*) and visualization of orbital septum (*OS*) with underlying fat that contains the levator aponeurosis and Muller's muscle. (**c**) Exposure of keyhole, frontozygomatic suture (*arrow*), frontal bone, and orbital rim. (**d**) Illustration of craniotomy after performing all osteotomy cuts (Copyright retained by Dr. Varun R. Kshettry. Used with permission)

Approach-related complications may include supraorbital nerve injury resulting in permanent forehead numbness, facial nerve injury resulting in frontalis palsy, and delayed mucocele formation or infection secondary to frontal sinus entry. For tumors extending into the optic canal, early bony optic nerve decompression is extremely difficult through the supraorbital keyhole approach which may potentially lead to worsened visual outcomes, although inferior visual outcomes have not been observed in any of the retrospective studies evaluating the approach.

9.3 Mini-pterional Approach

The mini-pterional keyhole approach is a variation of the standard frontotemporal craniotomy. The primary purpose of the approach is to decrease disruption of the temporalis muscle and reduce total skin incision, while allowing for sufficient access to the anterior and middle cranial fossa, parasellar and cavernous sinus region. The approach is mainly indicated for MCA, supraclinoid ICA aneurysms, and limited lesions around the planum, sphenoid wing, and cavernous sinus. The mini-pterional craniotomy generally resides completely below the superior temporal line, whereas a keyhole variation that shifts the craniotomy more frontally without any temporal exposure is referred to as the lateral supraorbital approach. Because the amount of frontal and temporal craniotomy is reduced, subfrontal and subtemporal access is reduced.

The mini-pterional incision (Fig. 9.4a–d) resides more anteriorly at the hairline and starts about 1 cm superior to the zygoma and is carried just above the superior temporal line. Interfascial or subfascial dissection is performed to elevate the scalp flap anteriorly. The temporalis muscle is detached from the frontozygomatic process and reflected posteriorly and inferiorly. A limited cut in the posterior superior portion of the muscle may be performed to gain wide access. Because the inferior aspect of the temporalis muscle is not cut, there is less chance for neurovascular compromise and subsequent atrophy. An approximately 2.5–3 cm craniotomy is performed centered on the pterion. The craniotomy completely resides below the superior temporal line. Once the opening has been completed, surgery proceeds in standard fashion depending on the target pathology. Because of the limited incision, there is generally inadequate exposure of the orbital rims to perform osteotomies if an orbitozygomatic or orbitocranial approach is needed.

The mini-pterional approach is an ideal keyhole approach for MCA bifurcation aneurysms. However, in ruptured aneurysm cases, if there is increased intracranial pressure, a larger approach is necessary to allow for decompression. The surgeon must also be prepared to utilize an assortment of aneurysm clips and angled clip appliers as the bone edges of the smaller craniotomy can reduce the angulation necessary for straight clip appliers, particularly in a temporally herniating sylvian fissure. The approach provides excellent access to the opticocarotid, carotid-oculomotor, parasellar, and cavernous sinus regions. Small- to medium-sized tumors involving the sphenoid wing, posterior planum sphenoidale,

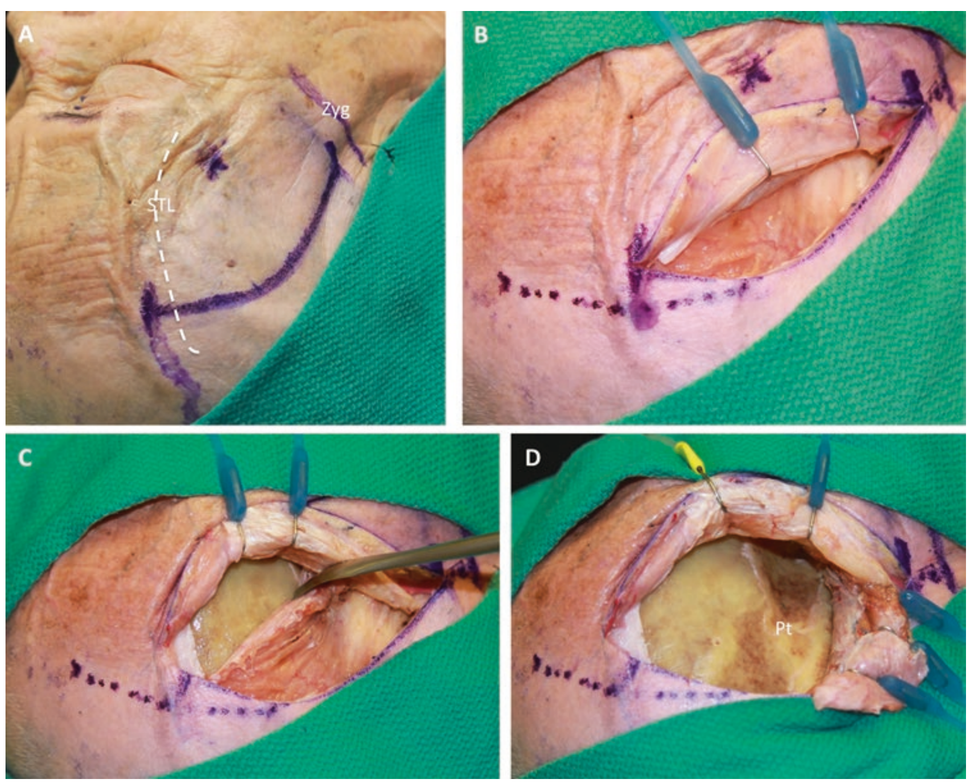

Fig. 9.4 (**a**–**d**) Cadaveric illustration of mini-pterional approach. (**a**) Incision starts 1 cm above the zygoma (*Zyg*) along the hairline and extends just above the superior temporal line (*STL*, *dotted white line*). (**b**) After incision, subfascial dissection is performed here, although interfascial dissection can also be utilized. *Dotted line* above top of incision marks incision course for a typical pterional craniotomy to the mid-pupillary line. (**c**) After subfascial dissection is performed, rather than cutting the temporalis muscle, it can be detached from the frontozygomatic process and mobilized posteriorly. (**d**) Exposure of the pterion (*Pt*). Size and exact position of craniotomy depend on target pathology (Copyright retained by Dr. Varun R. Kshettry. Used with permission)

tuberculum sellae, or cavernous sinus may be treated via this approach. Due to reduced subfrontal access, we generally do not use this approach for tumors located along the anterior planum or cribriform region, but use a variation that includes a more frontally placed craniotomy for better subfrontal access if a lateral approach is desired.

Similar to the supraorbital keyhole approach, while theoretically one might expect a benefit of minimizing soft tissue disruption, there are no studies adequately demonstrating an objective cosmetic advantage of the mini-pterional approach compared to the standard frontotemporal approach.

9.4 Subtemporal Keyhole Approach

The subtemporal approach can certainly be considered a keyhole approach as a small 2.5–3 cm craniotomy can provide panoramic access to the middle and posterior fossa from the anterior clinoid process to the internal auditory canal. The subtemporal approach can be used to treat tumors of the middle and posterior fossae as well as posterior circulation aneurysms.

Multiple variations for skin incision exist including question mark, linear, and horseshoe. The question mark may be useful if more anterior exposure is necessary along the middle fossa. The use of a lumbar drain may be necessary to provide adequate brain relaxation. The craniotomy for the subtemporal keyhole approach can be modified depending on target pathology. A slightly more anterior craniotomy is needed for approaching the anterior cavernous sinus, trigeminal nerves, and petrous apex while a more posterior craniotomy may be made for accessing the internal auditory canal and tegmen region. Generous burr holes can help strip the dura to avoid dural laceration and can allow the surgeon to precisely localize the level of the middle fossa floor to allow the inferior cut to run as low as possible along the floor. For pathology extending superior to the free edge of the tentorium or involving the cavernous sinus, the addition of a zygomatic osteotomy may help to mobilize the temporal muscle more inferiorly and improve the inferior-to-superior trajectory. However, this is frequently unnecessary and may lead to increased pain postoperatively but depends on individual patient anatomy, with the midportion of the zygoma typically lying between 13 mm superior to the middle fossa floor and about 9 mm inferior [6]. A zygomatic osteotomy may be more useful in a “high-riding” zygoma. In both the keyhole and standard subtemporal approaches, the temporal lobe must only be retracted to the extent necessary to expose the pathology as excessive retraction can lead to temporal lobe injury, seizures, and venous infarction. The keyhole approach and zygomatic osteotomy may help to decrease the amount of lateral temporal lobe retraction [7]. Finally, the use of endoscopes in the subtemporal approach may be more limited to the intradural portion of the procedure. During the extradural portion, there is often significant venous ooze that can impair endoscopic visualization, and there is less need to “look around the corner.” Conversely, during the intradural stage, especially when working around the basal cisterns, the endoscope can increase illumination and visualization.

9.5 Retrosigmoid Keyhole Approach

The classic retrosigmoid craniotomy serves as the “workhorse” approach for accessing the cerebellopontine angle (CPA) but requires significant muscular dissection to provide wide exposure, resulting in a relatively high incidence of postoperative headaches, cervical pain, sensory deficit, and cerebrospinal fluid leakage. Progressive reduction of the exposure primarily for microvascular decompression of the trigeminal nerve along with endoscopic assistance has allowed for increased utilization of the retrosigmoid keyhole approach for a variety of indications including microvascular decompression of cranial nerves, resection of neoplastic and nonneoplastic (e.g., epidermoids, arachnoid cysts) lesions, and clip ligation of posterior circulation aneurysms. In Chap. 11, a very thorough overview of endoscopic and keyhole approaches to the CPA demonstrates that through careful tailoring of the retrosigmoid keyhole, excellent access can be obtained to the region extending from the tentorium to the foramen magnum.

In our experience, we have primarily utilized the retrosigmoid keyhole approach for vascular compression syndromes and have found that endoscopic assistance leads to improved visualization of the anterior aspect of the cranial nerves and root entry zone (REZ), while allowing for reduced patient discomfort and accelerated recovery. For microvascular decompression of the trigeminal nerve, neuronavigation may be utilized to help identify the asterion and transverse-sigmoid sinus junction as accurate placement of the incision and craniotomy are critical to perform the procedure in a truly minimally invasive manner. Failure to fully remove the bone over the venous sinus margins will limit the final positioning of the endoscope and require increased cerebellar retraction to provide exposure. Additionally, we utilize a bipolar (SilverGlide® Bipolar Forceps, Stryker, Kalamazoo, MI, USA) with ultrathin tines of variable lengths that enable increased maneuverability when working through small craniotomies as well as bayonetted, rotating micro-instruments (Evans Rotatable Instruments, Mizuho America Inc., Union City, CA, USA) designed for minimally invasive procedures. Although dynamic endoscopy is extremely helpful in establishing depth of field and is the preferred technique for endonasal endoscopy, the restricted anatomy of the CPA lends itself better to fixed endoscopy with an endoscope holder. Additionally, although obscuration of the endoscope lens

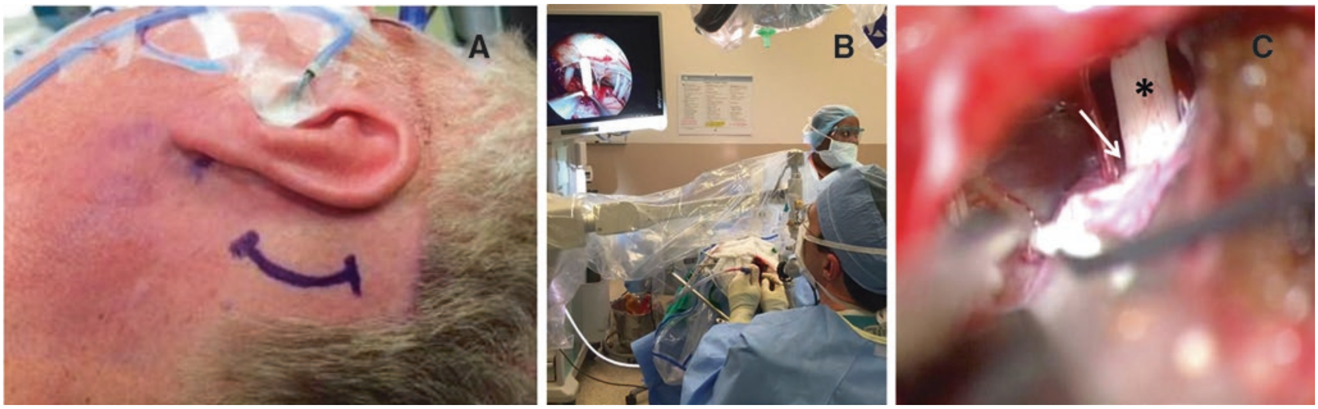

Fig. 9.5 (**a**–**c**) (**a**) Slightly curvilinear 2 cm incision is planned for endoscopic-assisted retrosigmoid keyhole approach microvascular decompression of the trigeminal nerve using anatomic landmarks and neuronavigation at the sigmoid-transverse sinus junction. (**b**) A 2.7 mm endoscope is positioned parallel to the posterior petrous ridge along the tentorium using a pneumatic endoscopic holding arm. (**c**) Endoscopic view of trigeminal nerve (*) and site of vascular compression at nerve root entry zone (*arrow*) (Copyright retained by Dr.Varun R. Kshettry. Used with permission)

may occur, we do not utilize an endoscopic lens cleaner during this procedure as the device increases the circumference of the endoscope and further restricts working area. We typically perform an approximately 2 cm curvilinear retroauricular incision (Fig. 9.5a–c) which enables a 14 mm diameter burr hole-type craniectomy to be performed at the transverse-sigmoid sinus junction. In patients with thicker skin and musculature, as compared to a linear incision, the curvilinear incision allows the soft tissue to be adequately reflected anteriorly using fish hooks such that there is minimal restriction of the endoscope insertion angle.

Once the dura has been opened, access to the cisterna magna is not often possible; however, arachnoid dissection above the facial-vestibulocochlear nerve complex with gentle CSF aspiration facilitates cerebellar retraction. The trigeminal nerve is then inspected circumferentially from the REZ to Meckel's cave for any signs of vascular compression with 0–30° endoscopes. Once the offending vessel has been identified at the REZ and the decompression performed, the dura can typically be repaired primarily as there is no thermal injury or shrinkage as occurs with the microscope.

In addition to the standard complications associated with accessing the CPA, potential complications of the retrosigmoid keyhole approach include cerebellar surface injury due to repetitive "blind" introduction of instruments proximal to the endoscopic field of view. Additionally, the endoscopic view may be impaired by a prominent suprameatal tubercle requiring the use of angled endoscopes or drilling of the tubercle to allow for safe dissection.

9.6 Pineal Region Keyhole Approaches

The pineal region, given its deep location from the cranial surface, is well suited for a keyhole approach. Keyhole approaches in this region are modifications of more traditional open approaches. This includes the midline supracerebellar infratentorial, lateral supracerebellar approach, and occipital transtentorial approach. Both keyhole and more traditional variations can be performed in either sitting, semi-sitting, lateral decubitus, park bench, or prone positioning. The sitting and semi-sitting position have the advantage of gravity retraction of the cerebellum in the infratentorial approaches but at the cost of mild risk of venous air embolism and increased surgeon fatigue. The keyhole variations of these approaches aim to decrease soft tissue disruption via smaller openings. Because much of the surgery is performed under endoscopic control, the need to make a larger opening to allow enough illumination from the microscope is obviated. Particularly for more vascular or fibrous tumors, significant comfort and experience with tumor resection under purely endoscopic control is necessary to tackle such tumors in this delicate area. In addition, keyhole approaches require greater precision with specific patient positioning and sufficient brain relaxation and have less room for error if there is significant bleeding or brain swelling.

9.7 Transcortical Keyhole Approach

The transcortical keyhole approach does not represent a single standard surgical approach, but rather a conscious effort to reduce cortical and white matter injury during the resection of deep-seated intraparenchymal lesions through the use of minimal access corridors. While much of this injury results from the trajectory chosen to access the lesion, the injury may be exacerbated by fixed retraction leading to cerebral edema and vascular compromise resulting in ischemia. Similar to brain retraction for skull base approaches, dynamic retraction is thought to reduce the penumbra of injury but may lead to compromised visualization. More recently, a number of surgeons have reported their experience using tubular retractors for the removal of a diverse array of deep-seated lesions including gliomas, metastases, cavernous malformations, intraventricular tumors, and intracranial hemorrhages (Fig. 9.6a–f). These tubular retractors may be placed using stereotactic guidance and have the advantage of producing equal radial distribution of the retraction pressure which may potentially maintain blood flow to the surrounding tissues and reduce ischemic injury. The use of a tubular retractor for the resection of thalamic gliomas was initially described by Kelly et al. in 1989, and currently there are several commercially available stereotactic tubular retractor systems ranging in diameter from 12 to 28 mm and available in various lengths [8]. Visualization beyond the retractor system is usually augmented with a microscope, endoscope, or exoscope (Fig. 9.7a–e). The retractors may be placed either via a transcortical incision or within a sulcus and are often combined with white matter mapping using diffusion tensor imaging (DTI) in an effort to further minimize neurologic injury. While these retractors certainly enable access to subcortical lesions with less trauma compared to standard fixed retractors, restricted diffusion changes surrounding the retractor corridor have been observed suggesting that there is some element of retraction-related ischemia [9]. In Chap. 14, Pradilla and colleagues provide an excellent description of their experience using the transcortical keyhole approach in the context of white matter fascicular anatomy.

9.8 Conclusion

Although the keyhole concept was popularized in the 1990s, refinement of surgical approaches has been a continuous endeavor, greatly facilitated by an increased understanding of surgical anatomy and advancing technologies including visualization, neuronavigation, and instrumentation. The rationale of keyhole surgery is to reduce the surgical corridor to the smallest extent necessary to achieve the optimal outcome. Critical to performing successful keyhole surgery is the understanding that just because the corridor is reduced,

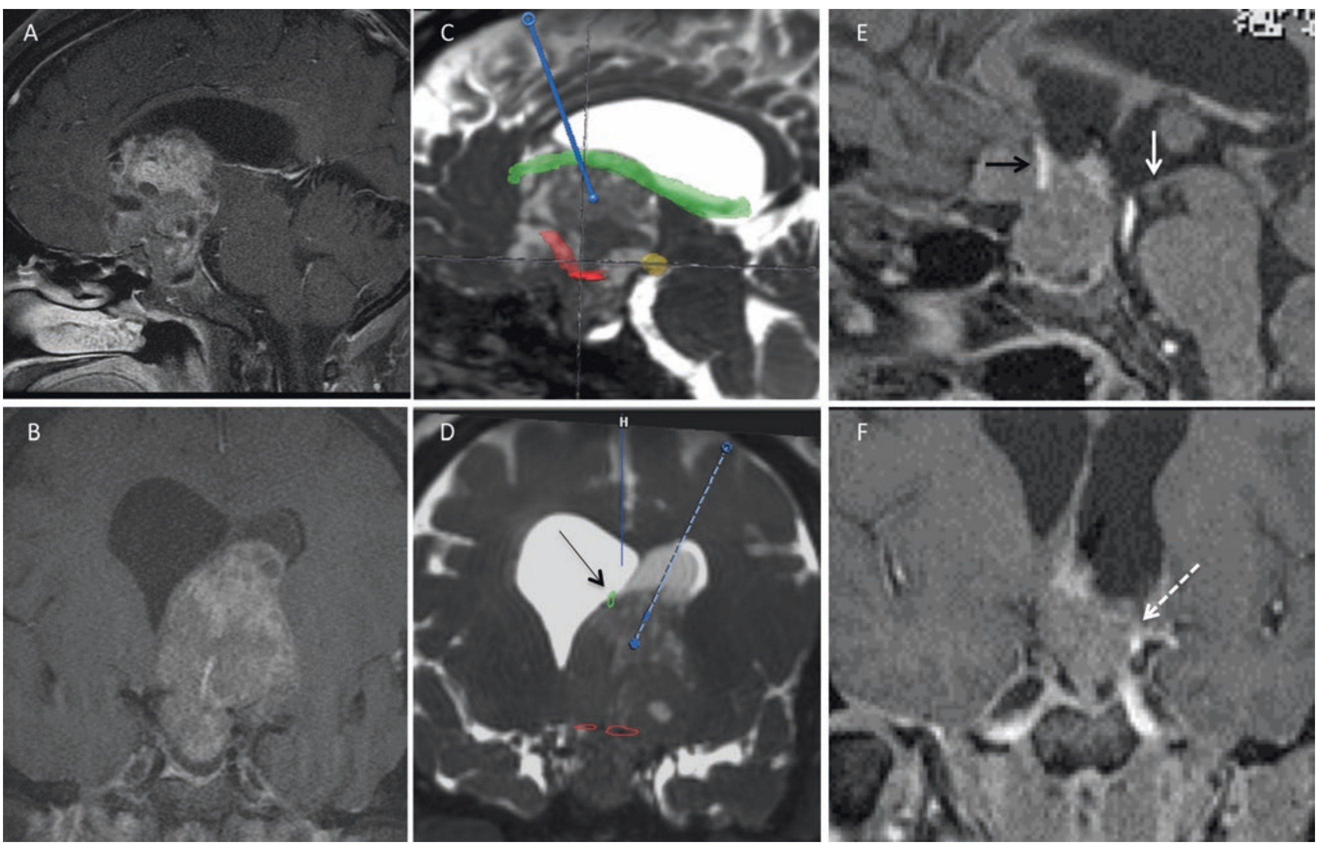

Fig. 9.6 (**a**–**f**) Preoperative sagittal (**a**) and coronal (**b**) T1 MRI demonstrating large recurrent pituitary adenoma encasing anterior cerebral arteries, extending along planum, invading through the frontal lobe, filling the anterior third ventricle, and extending up to the roof of the left lateral ventricle. (**c**–**d**) Preoperative plan demonstrating surgical trajectory (*blue line*) of tubular retractor. Left-side cannulation through smaller ventricle was chosen as this was felt to have less chance of forniceal (*arrow*) injury during tumor resection. *Fornix (green); anterior cerebral arteries (red); mammillary bodies (yellow)*. Postoperative sagittal (**e**) and coronal (**f**) T1 MRI demonstrating resection of ventricular component of tumor down to suprasellar cistern. Depicted are anterior cerebral arteries (*black arrow*), mammillary bodies (*white arrow*), and internal cerebral artery terminus (*dashed white arrow*). The patient later underwent a second-stage endoscopic endonasal resection of residual tumor (Copyright retained by Dr. Varun R. Kshettry. Used with permission)

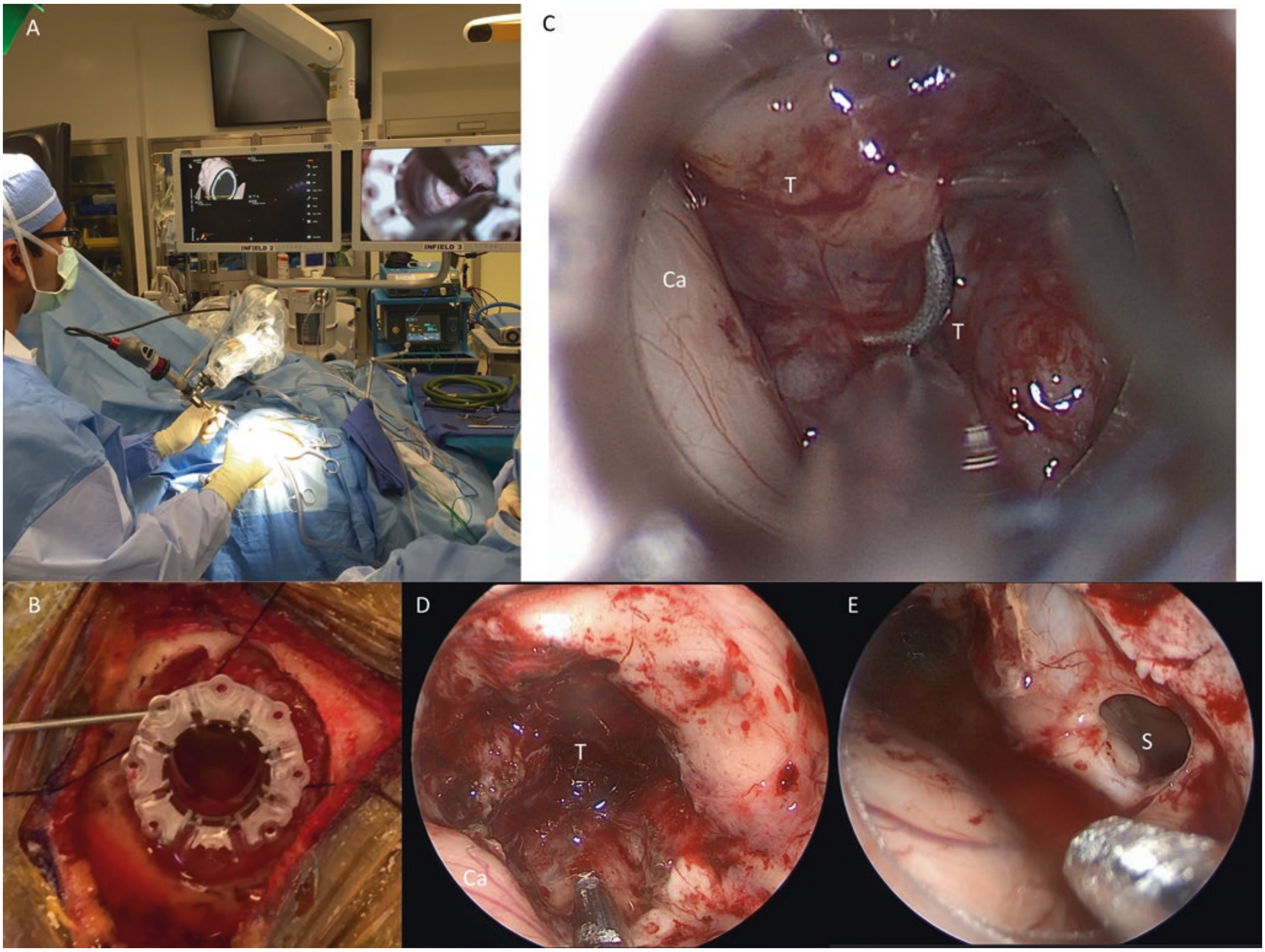

Fig. 9.7 (**a**–**e**) Intraoperative views. (**a**) Depicting OR setup with exoscope held in pneumatic arm. (**b**) Craniotomy and cannulation with 13 mm tubular retractor. (**c**) Exoscope used for initial tumor (*T*) resection. *Ca = caudate head.* (**d**, **e**) Final resection performed with endoscope demonstrating residual tumor (*T*) in suprasellar region encasing ACA as identified by neuronavigation and micro-Doppler, with septostomy (*S*) performed as well

the surgical principles must remain the same as those of more "traditional" neurosurgical approaches including avoidance of excessive retraction and meticulous microdissection under direct visualization.

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10 Keyhole Supraorbital Craniotomy: Eyelid and Eyebrow Approaches

Gordon Mao, Nouman Aldahak, and Khaled Abdel Aziz

10.1 Introduction

Minimally invasive surgery (MIS) approaches to the anterior skull base have continued to evolve in past decades. The anatomy of the parasellar and suprasellar regions lends itself more favorably to an anterior approach. Posterolaterally situated windows to the suprasellar area are bordered by the mesencephalon posteriorly and by the temporal lobes laterally. Approaches from these directions, therefore, require additional brain retraction or a wider splitting of the Sylvian fissure. Conversely, approaches from the anterior direction offer several working windows without the need for significant brain retraction (e.g., subchiasmatic window, carotico-optic window, retrocarotid window) [1].

Historically, the approach to lesions in the midline region of the anterior cranial fossa involves using the bicoronal or "three-quarter Souttar" incision and a frontal or bifrontal craniotomy. Numerous other approaches and skull base variants have also been reported to access this region including frontotemporal, pterional, and orbitozygomatic craniotomies [2]. These approaches were designed to minimize brain retraction and its associated morbidity. However, they often necessitate large incisions that create significant postoperative discomfort and permanent areas of scalp alopecia and carry the risk of disruption of olfaction and facial nerve injury. Another significant complication often associated with the pterional, and even mini-pterional, craniotomy is temporalis muscle atrophy which can cause significant, noticeable cosmetic deformity from temporal wasting in up to 25% of patients [3] along with temporomandibular joint dysfunction or pain.

The introduction of various supraorbital keyhole MIS craniotomy approaches placed a stronger emphasis on cosmesis in addition to surgical approach to the lesion. These techniques carry the added advantage of shorter operation times, decreased hospital stay, and decreased postoperative morbidity [4–9]. A frontotemporal incision behind the hairline generates an excessively large flap for a small craniotomy and may additionally involve the use of subgaleal drains, hair shaving (although hair sparing is an alternative), or incising the forehead in patients with alopecia or a receding hairline [10].

The supraorbital craniotomy through an eyebrow incision was initially described as a "keyhole" craniotomy for pathology ranging from vascular, infectious, and neoplastic etiologies and has grown to become one of the most common MIS approaches to anterior cranial fossa lesions. Anatomical studies have demonstrated that the area of exposure of the parasellar region through the smaller supraorbital keyhole approach is comparable to the larger pterional and supraorbital approaches [11]. Keyhole surgery generally aims to limit the iatrogenic trauma to surrounding structures such as the skin, bone, dura, and brain. The addition of an orbital osteotomy has become an increasingly popular supplement in anterior cranial base surgery with the subfrontal approach that minimizes retraction of normal anatomy. This offers wide access to the anterior skull base and parasellar region by exploiting the subfrontal corridor. Several anatomical and clinical reports document its benefits in terms of increased exposure, decreased brain retraction leading to a lower incidence of frontobasal retraction injury, and obviation of Sylvian fissure dissection, another potential cause of surgery-induced brain injury. The mini-fronto-orbital craniotomy can achieve the same microsurgical exposure as the standard frontotemporal approach.

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There are three primary incisions developed to optimize cosmetic outcomes of the supraorbital keyhole approach: transciliary, supraciliary, and transpalpebral incisions. Transciliary incisions may lead to hair follicle depilation, but this typically does not occur if one avoids the use of cautery [12]. Supraciliary incisions avoid depilating the hair follicles but can leave a visible scar above the eyebrow. Both incisions involve cutting across the frontalis muscle which can result in eyebrow asymmetry, while local hair follicle damage can result in focal alopecia.

In the past decade, incision at the natural skin crease of the upper eyelid via the transpalpebral (aka supratarsal fold) approach has allowed for comparable surgical outcomes while reducing the risk of frontalis palsy or eyebrow alopecia. The transpalpebral approach places the incision through the folds of the eyelid, thus also avoiding depilation of the hair follicles, but typically requires the use of a second specialist with experience performing surgery through the eyelid [6]. The eyelid incision is used in cosmetic blepharoplasty and is well hidden. The standard transpalpebral approach can achieve a one-piece fronto-orbital bone flap that extends from the sphenoid keyhole medially to the lateral edge of the supraorbital notch or foramen with the addition of an orbitotomy dramatically improving visualization and the angle of exposure from the ipsilateral to contralateral optic nerve [6]. All three incisions can become problematic in the setting of infection, but infection risk is low with this approach (2–7%) [13].

10.2 Historical Perspective

Recent advances made in stereotactic image guidance and endoscopic sinus surgery have facilitated the evolution of minimally invasive skull base surgery. As with the open approaches, endoscopic skull base and intracranial surgery evolved from techniques developed to reach sellar lesions through the sphenoid sinus. While the extended endoscopic transsphenoidal approaches to the sella have been used to treat a wide range of anterior intracranial lesions, they may involve indirect working angles and have limited reconstruction options.

The transciliary eyebrow incision approach was first popularized by van Lindert et al. in 1998 for the management of intracranial aneurysms [1]. Czirjak et al. published their experience in 2001 [14] and 2002 [15]. Ramos-Zuniga presented the modification to widen the approach via the so-called trans-supraorbital approach [16].

Application of the endoscope in the keyhole eyebrow supraorbital approach enhances the possibilities of skull base minimally invasive surgery, providing a broader access ranging from the cribriform plate to the mesial temporal lobe, without supplementary brain retraction. This approach represents a valuable alternative to both the classical eyebrow microsurgical and endoscopic endonasal routes in the treatment of anterior and middle fossa pathologies, minimizing the risk of postoperative CSF leaks and avoiding rhino-nasal complications while providing a better control of lateral tumor extensions [17]. H.D. Jho first described an endoscopic, orbital roof craniotomy at UPMC to approach anterior cranial fossa lesions [18] as a modification of a procedure performed by Frazier in 1913 [19].

The more recent introduction of the transpalpebral approach came from its pre-existing role among oculoplastic surgeons to resect orbital tumors and to repair orbital fractures. This upper eyelid incision is also commonly used for blepharoplasty by utilizing the advantage of a natural skinfold [8, 20]. The dissection plane in the eyelid preserves all the functional eyelid structures, thus minimizing postoperative complications. Compared with the suprabrow minicraniotomy approach, the transpalpebral route uses incisions placed in a natural eyelid crease, eliminates the risk of injuring facial nerve branches to the brow elevators, preserves sensory innervation of the forehead, and allows removal of the orbital ridge and frontal bone as a single bone flap. The ability to remove the orbital ridge is particularly desirable as it adds the improved exposure described and quantified by Schwartz et al. in their cadaveric studies [21]. Ohjimi first utilized the transpalpebral incision to repair traumatic orbital roof fractures in 1996 in replacement of the much more extensive bicoronal incisions [22]. Andaluz was the first to document his experience with the eyelid incision to treat anterior circulation aneurysms and suprasellar tumors in 2008 [10]. Aziz released the first significant case series outcome data in 2011 [6]. Recent anatomic research has extended the transpalpebral incision with the extended transorbital minicraniotomy that involves posterior orbital roof and adjacent supraorbital rim, with the advantage of even less frontal lobe retraction than a supraorbital craniotomy [23].

10.3 Indications

The supraorbital craniotomy as an anterolateral approach affords a wide exposure of the skull base, extending from the cribriform plate to the mesial temporal lobe and ventral brainstem. Accessible anatomic regions and structures include the ipsilateral anterior cranial fossa, basal frontal lobe and frontal pole, medial temporal lobe, lateral wall of cavernous sinus, ipsilateral proximal Sylvian fissure, opticocarotid cistern, suprasellar region, superior sella, lamina terminalis, perimesencephalic/interpeduncular cistern, medial or superior aspect of the contralateral optic nerve, chiasm, and internal carotid artery. When supplemented with intracranial endoscopy, lesions of the lateral cavernous sinus, pituitary fossa, contralateral circle of Willis, and ipsilateral retro-orbital space may be addressed.

As for microsurgery, specific indications include neoplastic pathologies within the anterior and middle skull base regions, mainly small- to medium-sized meningiomas (4–5 cm) of the cribriform plate, as well as the planum and suprasellar regions, prechiasmatic craniopharyngiomas, and suprasellar pituitary adenoma remnants as well as vascular pathologies including anterior circle aneurysms, arteriovenous malformations, and cavernous hemangiomas [12].

This approach has been well studied for the management of various aneurysms along the ipsilateral and contralateral arteries of the circle of Willis. In particular, the contralateral ophthalmic artery, medial wall of the internal carotid artery, M1 segment of the middle cerebral artery, A1 segment of the anterior cerebral artery, posterior communicating artery, P1 segment of the posterior cerebral artery, and superior cerebellar artery can be adequately exposed [1] (Fig. 10.1). In addition, for distal aneurysms and lesions in the Sylvian fissure, a more lateral sphenoid ridge keyhole approach has been recently described in detail [24].

The supraorbital approach also possesses key advantages for various tumors of the anterior skull base. Some have also noted that olfactory groove meningiomas with significant lateral extension are ideally suited for approach by the supraorbital route, while managing far anterior olfactory groove meningiomas requires an angled endoscope and instrumentation to directly visualize the attachment point of the tumor in the midline depression of the anterior cranial fossa floor [25]. Significant tumor extension into the nasal sinuses typically calls for an endonasal approach.

Along with the extended endonasal approach (EEA), the supraorbital approach is an effective alternative for the resection of tuberculum sellae meningiomas [26–28]. In cases where no intervening cortical cuff is present between the tumor and cortical vessels, the supraorbital approach offers superior microsurgical control critical vessels during their dissection from the tumor. Additionally, tuberculum sellae meningiomas over 30–35 mm in maximal diameter are generally better approached via a supraorbital or pterional approach rather than an endonasal approach. In cases where the tumor extends lateral to one or both of the optic nerves or lateral to the supraclinoid carotid arteries, the supraorbital approach is also generally preferred [25].

Craniopharyngiomas are frequently approached via either an extended endonasal or supraorbital route. Retrochiasmal craniopharyngiomas are generally best approached via an extended endonasal route that provides direct access below and posterior to the optic chiasm and allows direct access up to the third ventricle. Significant superior midline extension is also a relative contraindication to the supraorbital approach, as the flat trajectory along the floor of the frontal fossa may not provide access to the superior extent of the tumor. Craniopharyngiomas with lateral extension beyond the supraclinoid carotid arteries or large anterior extensions are often best approached via the supraorbital or pterional route [26, 29]. Pituitary adenomas are classically treated via EEA. However, the supraorbital approach can be more beneficial for fibrous adenomas adherent to surrounding neurovascular structures or those with significant superolateral or anterior extension over the planum [30].

Finally, for many intra-axial primary or metastatic tumors of the orbitofrontal region, frontal pole, and medial temporal lobe, the supraorbital approach often provides the most direct surgical trajectory with a shallow corticectomy and little or

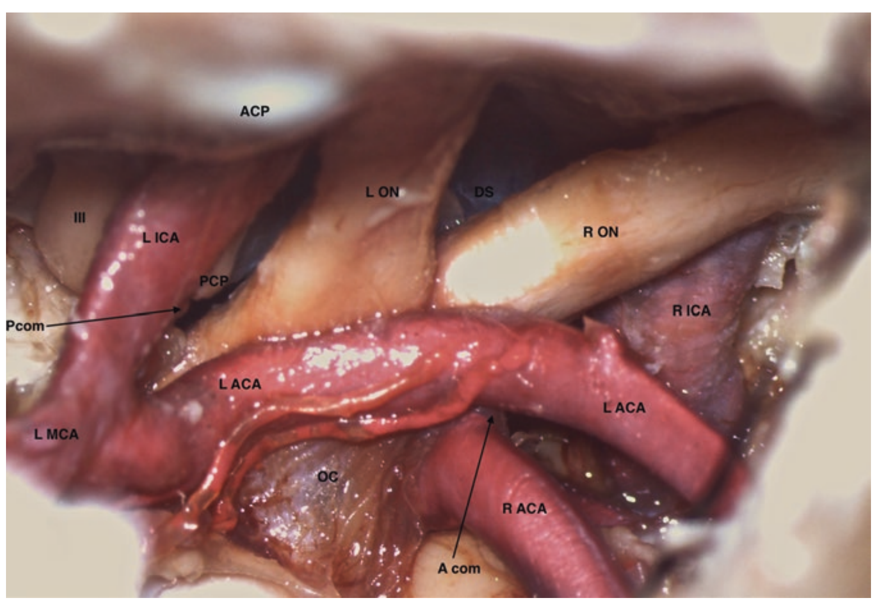

Fig. 10.1 Cadaveric dissection showing exposure of the optic chiasm and anterior portion of circle of Willis. (*ON* olfactory nerve, *III* oculomotor nerve, *ICA* internal carotid artery, *MCA* middle cerebral artery, *ACA* anterior cerebral artery, *Acom* anterior communicating artery, *Pcom* posterior communicating artery, *OC* optic chiasm, *ACP* anterior clinoid process, *PCP* posterior clinoid process)

no need for brain retraction. Navigation and intraoperative ultrasonography are useful adjuncts for these cases [25].

10.4 Facial Anatomy and Cosmetic Considerations

There are several neurovascular and neuromuscular structures to consider during soft tissue dissection. Care must be taken to preserve the supraorbital nerve and its vessels to avoid frontal scalp numbness, which are usually at the junction of the medial one-third and lateral two-thirds of the brow. The medial aspect of the eyebrow projects above the anterosuperior angle of the temporalis muscle attached to the orbitozygomatic process and the superior temporal line.

Because the frontal branch travels forward to enter the frontalis muscle and orbicularis oculi, it is vulnerable to injury during surgery in the periorbital or eyebrow region. The orientation of the frontal branch fibers becomes horizontal as it approaches the lateral canthus and eyebrow. Incisions in this region should therefore be as horizontal as possible. In a study by Schmidt et al., a relative "safety zone" (Fig. 10.2) free of nerve branches was identified that exists up to 2.5 cm lateral to the lateral canthus [31].

The upper eyelid is typically divided by the orbital septum into two lamellae: anterior and posterior (Fig. 10.2b, c). The anterior lamella contains the skin and orbicularis oculi muscle. The posterior lamella consists of the levator aponeurosis, the Muller's muscle, and the palpebral conjunctiva. The septum originates at the superior orbital rim, arising from the arcus marginalis, a thickened band of periosteum that is continuous with the frontal periosteum superiorly and the periorbita. Posterior to the orbital septum is the preaponeurotic fat pad. This fat pad is an important landmark during upper eyelid surgery because the levator aponeurosis lies immediately posterior to it. Staying anterior to this landmark will avoid damage to the levator/Muller complex.

The upper eyelid crease is located 8–11 mm superior to the eyelid margin. It is formed by the attachment of the levator aponeurosis in the subcutaneous tissue inferior to its union with the orbital septum. There is a potential space superior to the upper eyelid crease between the septum and

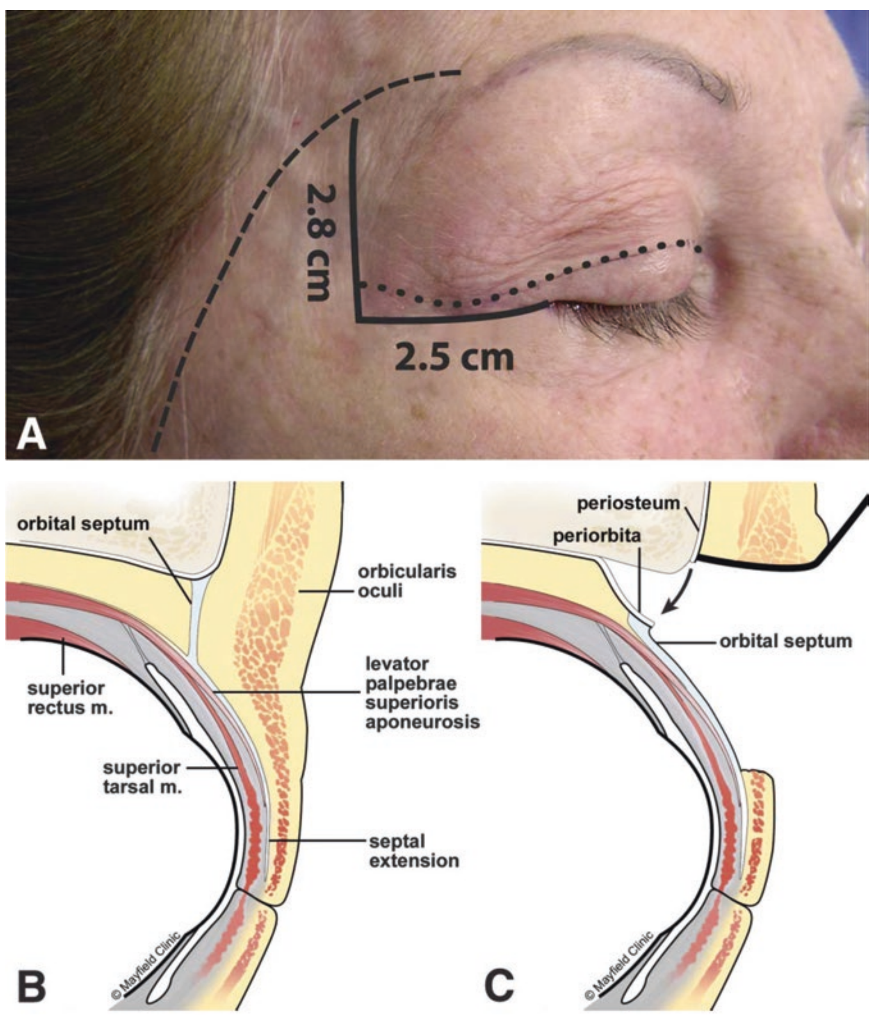

Fig. 10.2 (**a**) Photo showing the "safety zone" as marked on the patient's face. The safety zone is an area 2.5 cm lateral to the lateral canthus, which is free of facial nerve branches. The *solid line* marks the path of the temporal branch of the facial nerve. The *dotted line* marks the extent of the eyelid incision. (**b**) Sagittal cross section of the eyelid demonstrating the layers encountered after an upper eyelid incision. (**c**) Sagittal cross section demonstrating separation of the layers of the eyelid. The periosteum is divided sharply along the midpoint of the orbital rim, just above the orbital septum. Dissection of the periorbita from the roof of the orbit may be extended as far posteriorly as the orbital apex muscle (**b** and **c**: Printed with permission of Mayfield Clinic, Cincinnati, OH, USA)

the skin/orbicularis oculi complex. This space creates a natural, avascular plane that can be used to dissect up to the orbital rim without damage to the posterior lamella.

10.5 Limitations

Numerous shortcomings have been overcome since the introduction of this approach in the 1980s. Probably the biggest limitation was the problem of illumination using the operating microscope down such a narrow corridor to reach a deep-seated lesion. In addition, there is a narrow viewing angle through this approach that may require frequent adjustment of the operating room table and microscope for adequate visualization of a given lesion. Angulated surgical instruments similar to those used in microscopic transsphenoidal surgery have been adapted to access lesions around corners, especially behind the ipsilateral optic nerve.

Endoscopes have dramatically improved visualization of the parasellar or chiasmatic lesions through this approach and allow for safer dissection with better visualization through this smaller incision than can often be achieved with the microscope alone. The supraorbital keyhole approach can never be a standard approach in the sense that the craniotomy is not always placed in the same location and of the same size. When considered for aneurysm treatment, in each individual patient, it should be evaluated whether this approach can allow for proximal vascular control, dissection of the aneurysm, and application of the aneurysm clip. Due to the small craniotomy, there is less opportunity for a change of plan if unexpected findings occur during surgery. Multidirectional viewing is only achievable by the use of intraoperative endoscopy [1].

Lastly, in the setting of vascular pathologies, there may be some difficulty with using two suctions simultaneously in managing prematurely ruptured aneurysms or to obtain proximal control [11, 32, 33]. For this reason, some have even recommended against this approach for vascular lesions [33].

10.6 Techniques

Cosmetic Considerations

The initial incision should be performed through the skin and dermis layers only. Cephalad dissection superficial to the orbicularis oculi, pericranium, and temporalis muscle is important for the development of a separate tissue flap for covering the keyhole craniotomy during closure [11, 30, 32, 34]. Additional considerations for a good cosmetic result include proper replacement of the bone flap. Care must be taken to ensure that the outer cortex of the supraorbital ridge remains intact during the approach. The use of a burr hole cover and square titanium plates prevents the appearance or palpation of the gap between the bone flap and intact native bone following bone flap replacement in the patient.

Positioning

The head is secured in a three-pin Mayfield head holder (OMI, Inc., Cincinnati, OH) in the supine position after general anesthesia and orotracheal intubation. Preoperative imaging to assess the degree of pneumatization of the frontal sinus must be performed in planning the supraorbital approach: in cases of frontal sinus breaching, there is an increased risk of CSF rhinorrhea and infection. The head of the table is angled up to 15–20° to improve venous drainage and ease frontal lobe gravity-related retraction. It is then slightly extended, making the malar eminence the highest point on the face, and rotated to the contralateral side to various degrees depending on the anatomical goal [30, 35–37]. The head is turned to the ipsilateral side 10–45°, depending on the location of the pathology (10–15° for the suprasellar region and mesial temporal lobe surface, 30° for the planum sphenoidale, and 45° for the cribriform plate [12]). It is then slightly extended to the contralateral side to facilitate the frontal lobe to fall away from the anterior cranial floor, but the extension should not exceed 10°; otherwise, there is a risk of hiding part of the skull base. In vascular cases, Berhouma strongly advocates expanded draping for exposure within the surgical field of the classical frontotemporal skin incision for pterional conversion in the event of a vascular injury [12].

Eyelid Approach

Local anesthetic with 1:1 combination of 2% lidocaine with 1:100,000 epinephrine and 0.75% bupivacaine is infiltrated for hemostatic effect. One-half strength Betadine solution diluted with NS is used to prep the upper and lower eyelids to reduce ocular toxicity. The cornea and sclera are protected with a clear shield, while the nose, forehead, and malar eminence are prepped with chlorhexidine.

Care is taken not to incise deep through the orbicularis oculi, which is then elevated from the orbital septum and divided using Westcott scissors. The lateral extension of our eyelid incision beyond the lateral canthus (crow's feet skin crease) carries a minimal risk of injury to the frontal branch of the facial nerve compared with the lateral extension of the eyebrow, which will be in the vicinity of the frontal branch.

Periosteum elevation is then completed beginning along the orbital side with care to preserve the supraorbital neurovascular bundle until it is extended past the frontozygomatic suture. The incision may be extended approximately 1–1.5 cm beyond the lateral canthal angle along the crow's foot if needed. If no crow's foot is present, then the incision line is extended laterally from the lateral canthus along the eyelid crease line.

One relative limitation of the transpalpebral minicraniotomy is the need to traverse the frontal sinus in some patients and the attendant risk of increasing frontal sinusitis and delayed mucocele formation. To minimize this risk, a mucosal sparing technique is important especially in the region of the frontal sinus outflow tract [38].

A spheno-orbital burr hole is initially placed at the junction of the lateral wall of the greater wing and the roof of the orbit at the frontosphenoidal and sphenosquamosal sutures, approximately 1 cm behind the frontozygomatic junction. Drilling should expose frontal and temporal lobe dura as well as periorbita. A one-piece fronto-orbital approach is then established via frontal craniotomy (Fig. 10.3b, c) that extends initially from the frontal portion of the burr hole ending anteriorly at the superior orbital ledge and lateral to the supraorbital notch to avoid frontal sinus violation. The second cut (Fig. 10.3d) extends first cut through the supraorbital ledge. The third cut (Fig. 10.3e) is performed with the footplate extending from the orbital portion of burr hole across the frontal process of the zygomatic bone. The fourth cut (Fig. 10.3f) is made with a custom-designed KA chisel (Axon Medical, Inc.) along the orbital roof, while a malleable retractor protects the periorbita. The final cut is completed with the KA chisel from the orbital portion of the burr hole extending medially to connect to the posterior end of the fourth cut [6].

Residual sphenoid ridge is drilled with a 4 mm rough diamond burr to allow exposure of frontal and temporal dura. The bone flap extends from the sphenoid keyhole opening medially to the lateral edge of the supratrochlear notch or foramen medially; the anteroposterior extension is 2.5 cm from the supraorbital ridge.

During the bony cuts, the skin flap is protected with Desmarres skin retractors. Plastic rubber-covered blunt fishhooks protect the eyelid during the craniotomy and are intermittently released during the case. The dura is opened in a flap fashion on the basis of the periorbita, reflecting the dura anteriorly.

After performance of the intracranial procedure, the dura is closed in watertight manner, sometimes with a synthetic dural graft. Titanium plates with low-profile screws are used to secure the bone flap, avoiding placement directly on the zygomatic or supraorbital processes. Bone cement is applied across the superior orbital ridge cut and zygomatic cut as well as titanium mesh over the sphenoid keyhole. Several tack-up drill holes are placed across the superior temporal line and zygomatic process of the frontal bone to allow for temporalis reapproximation. Frontal periosteum is carefully reapproximated to periorbita with running 4-0 Vicryl to establish proper anatomic alignment, and the skin is closed with 6-0 absorbable running suture. The incision is covered with antibiotic ophthalmic ointment only, and ice packs are placed over the incision for 15 min every hour for the first 3 days postoperatively [6].

Eyebrow Approach

Eyebrow shaving is not necessary to avoid postoperative infection [30]. The incision within the eyebrow should be designed based on the shape and pattern of the brow so that the scar is hardly visible. The incision typically is performed on the right side and extends for 20–25 mm, unless a left-sided approach is needed for a left-sided lesion or to access the medial wall of the right optic canal. The incision is bounded by the supraorbital pedicle medially and the anterior part of the frontozygomatic suture laterally. An incision is made directly in the eyebrow, beveled slightly superiorly to keep parallel to the hair follicles to avoid cutting directly across the hair follicle (Fig. 10.4). The STA and facial nerve branches should not cross the surgical field [30]. Occasionally, the incision must be extended more laterally to obtain a more superior or lateral exposure. If this is necessary, 1 cm of the "laugh line" at the lateral aspect of the brow can be used to hide the incision. The supraorbital nerve is carefully dissected free from the tissues laterally attached to it and is preserved.

The frontal skin flap is dissected subcutaneously to expose the frontal and orbicularis oculi muscles. The frontal muscle is incised with low-intensity monopolar electrocautery parallel to the incision to avoid depilation, while the temporal muscle is detached from the superior temporal line for 1 cm in length and retracted laterally to expose the keyhole position. A careful dissection and precise gentle hemostasis are carried out during this phase in order to minimize the risk of postoperative periorbital swelling [14, 35].

The craniotomy includes the orbital roof, a portion of the frontal bone, and approximately 1 cm of the zygoma (Figs. 10.5 and 10.6). The width of the craniotomy usually measures 2–3.5 cm and the height measures 1.5–2 cm [39]. A small burr hole is made at the superior aspect of the exposed bone, approximately in the middle of the flap. Another hole is made at the keyhole, grooving the underside of the zygomatic arch as well. A small groove is then made just lateral to the supraorbital notch to expose the dura and flatten the orbital rim. A foot-plated, right-angled craniotome is then used to make cuts in the orbital rim medially and laterally, with the lateral cut extending across the zygoma toward the temporal burr hole. The craniotome is then used to connect the burr holes and the craniotomy is reflected inferiorly, fracturing the orbital roof across the gap between the

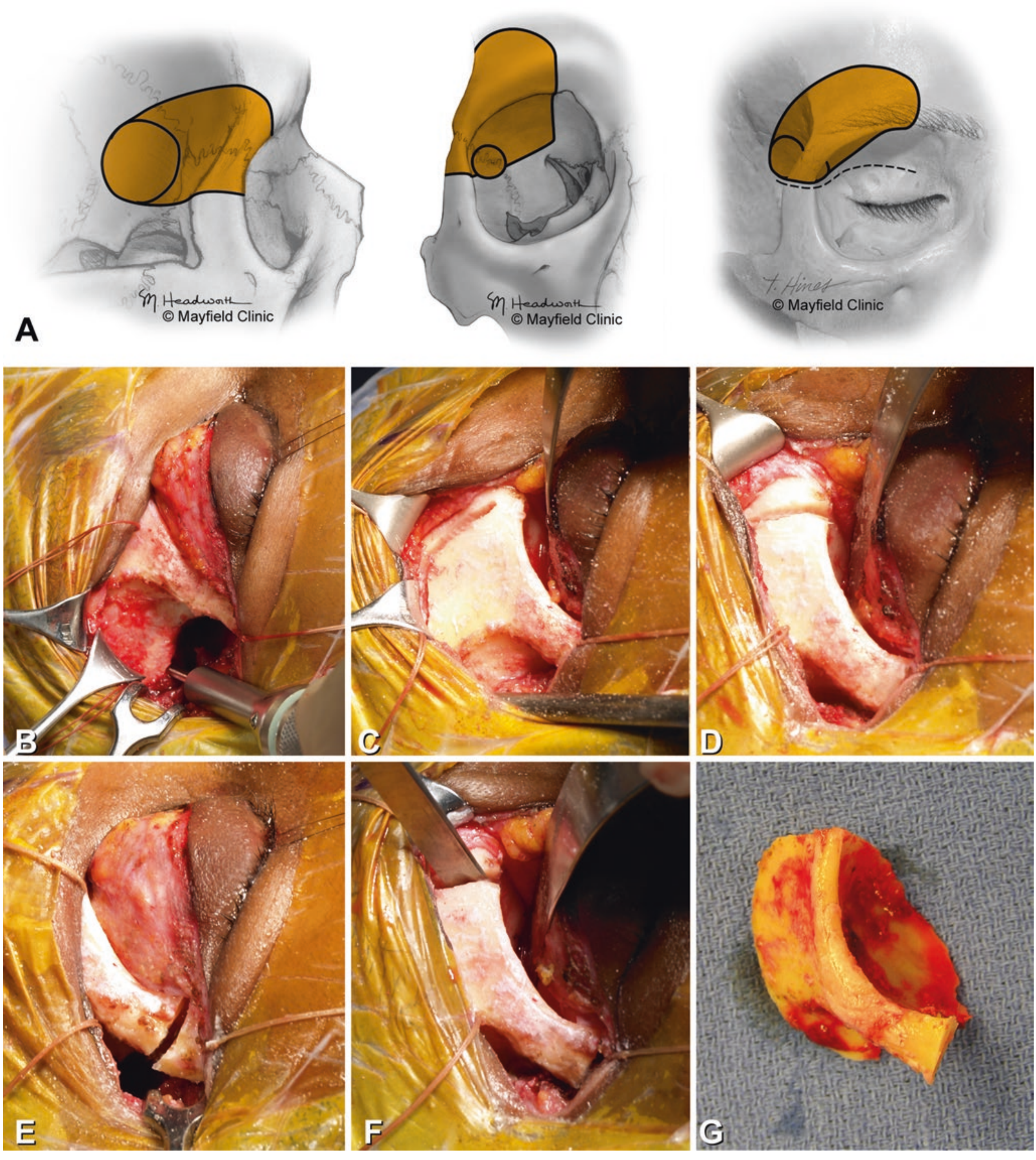

Fig. 10.3 (**a**) Illustrations of the skull with the fronto-orbital bony cuts outlined. Bony exposure, cuts, and craniotomy size are standard in all cases. The bone flap extends from the sphenoid keyhole opening medially to the lateral edge of the supratrochlear notch or foramen medially; anteroposterior extension is 2.5 cm from the supraorbital ridge. The bone flap includes the anterior two-thirds of the roof of the orbit (printed with permission from Mayfield Clinic). Operative still images (**b**–**g**). (**b**, **c**) Frontal bone cut starts from the frontal portion of the sphenoid keyhole and ends at the supraorbital ridge lateral to the supratrochlear notch. (**d**) Supraorbital ridge cut. (**e**) Zygomatic cut. (**f**) Orbital roof cut with the KA chisel with a soft malleable retractor blade protecting the periorbita during this step. (**g**) one-piece fronto-orbital bone flap (**a**: Printed with permission of Mayfield Clinic, Cincinnati, OH, USA)

Fig. 10.4 Intraoperative photograph demonstrating the eyebrow incision outlined

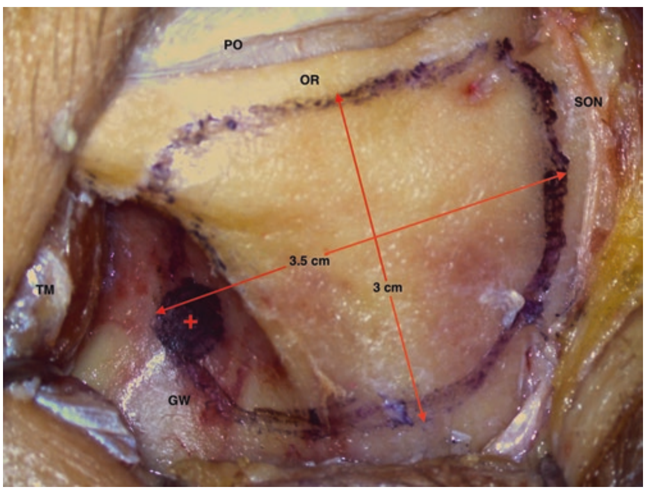

Fig. 10.5 Cadaveric dissection following typical eyebrow incision demonstrating exposure of the frontal bone and orbit and the extent of the supraorbital craniotomy (Abbreviations: *PO* periosteum, *OR* orbital ridge, *SON* supraorbital nerve, *TM* temporalis muscle, *GW* external surface of greater wing of sphenoid bone)

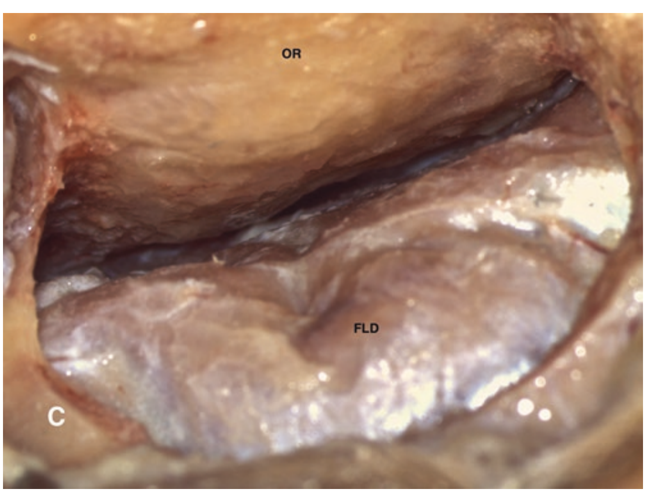

Fig. 10.6 Cadaver dissection demonstrating supraorbital craniotomy with the orbital rim (*OR*) and frontal lobe dura (*FLD*) exposed

two grooves drilled previously. If the frontal sinus is entered, it is exenterated and packed with temporalis muscle.

Paladino et al. recommend performing a craniectomy and reconstruction with a methyl methacrylate cranioplasty after the operation for several reasons [5]. First, the frontal dura is thin and adherent to the bone in elderly patients so that with piece-by-piece rongeuring, uncontrollable dural tears are avoided and waterproof dural closure can be done easily. The craniotomy must be aligned with the anterior fossa; therefore, the bone is usually drilled away to make a straight route over the surface of the anterior cranial fossa in order to reduce brain retraction. The residual groove after the osteotomy results in an unpleasant forehead deformity, so it is preferable to reconstruct it with methyl methacrylate.

Skin closure techniques vary between institutions. While many surgeons advocate temporary nonabsorbable sutures to reduce the risk of scarring following inflammation from absorbable stitches, evidence may suggest no ultimate differences in outcomes [40].

10.7 Results

Clinical validation of the keyhole supraorbital craniotomy began in the 1990s with individual and institutional experience with the eyebrow approach primarily for the surgical treatment of intracerebral aneurysms. Early goals focused primarily on vascular lesions with increasing recognition of the utility of this approach for a broad range of anterior fossa tumors by the early 2000s. The eyelid approach is a more recent introduction to the neurosurgical community and has been gradually gaining attention in the past decade. Its effectiveness has been validated by several authors to be equivalent to traditional open approaches and with the advantage of minimal risk of temporal wasting similar to the eyebrow incision but even lower rates of supraorbital paresthesia or frontalis palsy.

10.8 Approach Complications

Both the transciliary and transpalpebral supraorbital approaches can lead to various neurovascular- and craniotomy-related complications.

During initial soft tissue dissection, the supraorbital nerve is at risk for traction or compression injury leading to frontal scalp numbness. While transient post-op forehead numbness is often noted, the incidence of permanent numbness is low. Perneczky's initial 2004 series noted a 42% incidence of postoperative numbness with 13% permanent supraorbital anesthesia; however, his subsequent larger series noted a reduced 7.5% rate [30]. Gazzeri and Teo more recently noted only a 2% incidence of supraorbital hypesthesia [4]. Czirjak noted only 1.3% incidence among his 155 patients [14], while Fischer reported a 0% rate among his series of 793 patients [41]. A review of the literature up to 2013 shows an average of 2.8% across all case series [13].

The frontotemporal branch of the facial nerve may similarly be injured leading to forehead asymmetry, affecting overall cosmesis. Transient frontalis palsy is not infrequently noted. In addition, weakness of the eyebrow elevation can result from interruption of the insertion fibers of the frontalis muscle to the eyebrow itself or may be due to injury of the frontal branch of the facial nerve, which will be very close to the incision line if extended laterally. Perneczky's initial eyebrow incision case series of 223 patients in 2004 [42] reported 61 (27%) total cases of postoperative palsy, but only 23 (10%) cases of permanent palsy. In his later series of 450 patients, there was a 5.5% incidence of permanent frontalis weakness [30]. Other large series report between 0 and 1% incidence [1, 4, 41] of frontalis weakness with an average of 3% in the literature [13]. Among eyelid outcomes, a similar 0–7% incidence of frontalis weakness has been reported [6, 9].

Frontal sinus violation during the supraorbital craniotomy can be unavoidable in certain circumstances depending on patient anatomy and optimal surgical approach for select patients. Even with the pterional craniotomy, opening of the frontal sinus may not be avoided if the patient has a large sinus. Pondé et al. found that frontal sinuses are larger in males, so the chances of frontal sinus violation in eyebrow keyhole craniotomy are greater than in females [43]. While the frontal sinus is invariably encountered as the approach is more frequently utilized, there has not been any documented sinus infection or mucocele formation in the literature.

Cosmetic results from both the eyebrow and eyelid approach have been uniformly good with high patient satisfaction. In the largest series of eyelid cases, 97.5% of patients were satisfied with the wound appearance with only 1 of 40 patients experiencing significant, visible scarring that required revision. In the Perneczky series, 157 patients (70.4%) reported excellent cosmesis [42].

10.9 Discussion

Within the past two decades, there has been a growing body of literature in support of the numerous cosmetic and direct surgical advantages to a keyhole supraorbital craniotomy over traditional open approaches. With both the transciliary and transpalpebral incision, natural anatomic facial features are used to disguise already small incisions resulting in a large positive impact in overall cosmesis. The shorter incisions reduce wound healing issues with average infection rates between 1% and 2% [13].

In addition, the often present temporal wasting after pterional or bicoronal approaches is completely obviated in the eyelid and eyebrow approaches. Temporalis atrophy frequently causes obvious cosmetic scalp defects as well as persistent temporomandibular discomfort. In the eyebrow approach, incision of the temporal muscle is needed only for 1 cm along the superior temporal line, so there is no need for stripping the temporal muscle from the bone which results in atrophy and deformity. Among the numerous publications of the eyebrow and eyelid approach, no temporalis deformity has ever been documented.

The supraorbital craniotomy also provides the most direct surgical corridor to many anterior fossa and sellar regions. This approach decreases the need for frontal and temporal lobe retraction along with its associated morbidity and reduces the actual working distance to the target lesions. Beretta et al. conducted detailed morphometric cadaveric studies to compare the standard pterional craniotomy against the supraorbital and transorbital craniotomy. Six predefined triangles based on major anatomic landmarks such as the optic foramen and ICA bifurcation were used to numerically analyze the working area. They found that the latter two approaches both exceeded the pterional craniotomy with decreased depth of the surgical window to anatomic targets [44].

Giant olfactory groove meningiomas can be difficult to resect due to a high propensity to hemorrhage intraoperatively and need for cortical retraction during traditional approaches. Their high vascularity often necessitates early surgical control of the feeding ethmoidal vessels to reduce intraoperative bleeding. Normally, this requires a bicoronal flap to reach the medial orbital compartment before the craniotomy begins. With the supraorbital craniotomy, the ethmoidal vessels can easily be cauterized during initial orbital dissection. Further microscopic dissection of the proximal Sylvian fissure permits greater subfrontal dissection with minimal frontal lobe retraction to expose the contralateral aspects of large olfactory groove meningiomas, enabling gross total resection of tumors up to 6 cm in size.

A supraorbital craniotomy through an eyebrow incision can also provide a very efficient and direct approach to pathological conditions. The smaller incision can reduce the time from skin incision to durotomy to approximately 15 min, providing the potential for shorter operating times [1]. Total resection can be safely completed for suprasellar meningiomas, craniopharyngiomas, and Rathke's cleft cysts in less than an average of 2 h [45].

Larger midline lesions that approached via the subfrontal route can also cause disturbances in olfaction during frontal lobe retraction. This complication can be limited with the supraorbital approach. Reisch and Perneczky reported 27 cases of hyposmia in their eyebrow series of 450 patients (6%) with nine cases of bilateral hyposmia (2%) [30]. Other large series by Fischer, Van Lindert, and Czirjak report

0–1.3% incidence of olfactory loss [1, 14, 41]. Aziz's first 71 eyelid approach patients reported no hyposmia [46].

Overall cosmesis and patient satisfaction with both eyebrow and eyelid procedures are excellent. The eyebrow covers the incision well in most cases, while the overlying upper lid provides ideal camouflage of the eyelid incision. The extremely thin lid skin in the crease allows rapid healing with a virtually imperceptible mature scar.

There has been criticism of the supraorbital and transorbital approaches based on the premise that traditional approaches carry the advantage of familiarity, especially for less-experienced neurosurgeons, and because of the risks involved with potential damage to the orbital contents [47]. Surgeons should possess an excellent understanding of the anatomy and the so-called "classic" approaches prior to undertaking the eyelid approach, which may appear simple but become very problematic for less-experienced surgeons [10].

10.10 Surgical Pearls

- Eyelid approach is ideal for various tumors of the anterior skull base up to 5–6 cm in size because it provides for direct access to the lesion.
- The supraorbital craniotomy allows excellent surgical exposure for various unruptured aneurysms of the anterior circulation except for MCA aneurysms which are usually too laterally situated. We do not recommend using this technique for acutely ruptured intracerebral aneurysm due to the degree of brain swelling and therefore difficulty of exposure and retraction.
- Proper wound opening and closure require the expertise of a dedicated oculoplastics surgeon. Gentle tissue handling along with hemostasis using bipolar electrocautery or monopolar electrocautery with a Colorado tip avoids unnecessary skin and soft tissue trauma and improves wound healing.
- A standard five-cut one-piece fronto-orbital bone flap has been used in all cases. The flap extends from the sphenoid keyhole opening to the lateral edge of the supratrochlear notch in the medial-lateral direction and 2.5 cm posteriorly from the supraorbital ridge.
- Excellent wound healing requires reduction of fluid buildup in the soft tissue with the use of sterile suction drainage system as a TLS drain and frequent use of ice packs against the incision every 2–4 h for the first week.

10.11 Conclusion

Supraorbital MIS craniotomy approaches through the eyelid, eyebrow, and suprabrow incisions are all effective alternatives to traditional skull base approaches for lesions of the anterior fossa and parasellar regions. The smaller incisions decrease surgical trauma and postoperative discomfort for the patient and afford superior long-term cosmesis. These approaches, when used for appropriate situations and combined with microsurgical techniques and endoscopic supplementation, are safe and often more efficient at accessing both aneurysms and tumors than pterional and frontal craniotomies.

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11 Mini-Pterional Craniotomy

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Abbreviations

A1 and A2	First and second branch of anterior cerebral artery
ACA	Anterior cerebral artery
ACoA	Anterior communicating artery
CSF	Cerebrospinal fluid
ICA	Internal carotid artery
LSO	Lateral supraorbital approach
M1 and M2	First and second branches of middle cerebral artery
MCA	Middle cerebral artery
MCAbif	Middle cerebral artery bifurcation
PCA-P1	Posterior cerebral artery and its first branch
PCoA	Posterior communicating artery

11.1 Introduction

Craniotomies around the pterion are standard approaches for pathologies involving the sphenoid wings, parasellar region, carotid artery, and proximal anterior circulation. In 1967, Yasargil et al. described the classic pterional approach that became the standard approach to those regions until today [1, 2]. However, during the last two decades, the surgical approaches developed at the founding times of modern microsurgery were further developed to minimize skull openings, operating times, and approach-related soft tissue trauma. Further tailoring of the pterional approach has led to development of the less invasive mini-pterional craniotomy or the lateral supraorbital approach (LSO). The described LSO is executed slightly more frontal to the pterional landmark, which is the junction between the frontal, temporal, sphenoid, and parietal bones. It allows thereby an optimal view to the subfrontal space and a favorable trajectory for the dissection of the parasellar region, ACoA complex, and proximal Sylvian fissure [3]. This approach has found its worldwide spread after its implementation in Helsinki [4, 5].

11.2 Rationale

The lateral supraorbital approach is the consecutive further development of the pterional approach to minimize the approach-related trauma under optimal usage of surgical trajectory through the subarachnoid space.

11.3 Patient Selection

Pterional craniotomies are standard neurosurgical procedures, and therefore there are no specific contraindications, except pathologies that are not accessible by the approach such as distal ACA aneurysms or far distal MCA aneurysms. Like larger pterional craniotomies, the LSO is well suited for the approach of the proximal Sylvian fissure and circle of Willis, as well as lesions in the skull base of the anterior fossa. Moreover, nearly all pathologies around the clinoid are well approachable. Lesions treatable by the LSO approach can be summarized as follows:

1. Intracranial aneurysms located on the ipsilateral MCA (M1, MCAbif, and M2) or the circle of Willis including the ACA (A1, A2), ACoA, ipsilateral and contralateral ICA, basilar bifurcation, and PCA-P1

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2. Meningiomas in the olfactory groove, anterior clinoid, planum sphenoidale, or tuberculum sellae
3. Intracavernous lesions
4. Craniopharyngiomas
5. Pituitary tumors [3–8]

LSO approach may also enable an approach to contralateral MCA aneurysms, [6–8] but during the last decades, we have learned that contralateral MCA aneurysm treatment can lead to significant olfactory nerve injuries [6–8].

11.4 Surgical Anatomy

Extracranial Landmarks

Important extracranial landmarks for the skin incision are the supraorbital rim, the processus zygomaticus of the frontal bone, the superior temporal line, and the zygomatic arch. The pterion itself is the site of fusion of the frontal, parietal, temporal, and sphenoid bones and is a non-palpable landmark situated around 40 mm cranial from the midpoint of zygomatic arch and around 32 mm behind the frontozygomatic suture. The zygomatic process and the superior temporal line are the key landmarks for the initial burr hole and subsequent craniotomy.

11.5 Surgical Technique

Operative Setup

Before beginning surgery, the operating microscope optics and balance are checked. We recommend the use of the mouthpiece to facilitate the focusing and moving of the visual field during surgery. The operative microscope can be positioned on either side of the operating table, usually on the left side while the instrumenting nurse is on the right side.

Positioning

The patient is positioned supine, with the head elevated around 20 cm above the cardiac level to improve venous outflow, reduce venous oozing, and obtain a clean surgical field and a relaxed brain (Fig. 11.1). To obtain this position, the patient's shoulders are elevated around 5 cm from the table using a strong round pillow. The next step is head fixation and we prefer a Sugita (Mizuho, Tokyo, Japan) head holder. The exact positioning of the head is determined by the addressed pathology and the anatomical location to be approached. Usually the head is rotated around 30° to the contralateral side, tilted slightly lateral, and, according to the required

Fig. 11.1 Patient's position for a left lateral supraorbital (LSO) approach. The patient is placed in the supine position. The head is fixed in a four-point Sugita frame, elevated above the cardiac levels, and rotated 15–30° toward the opposite side. Finally, the head is slightly tilted laterally and extended or minimally flexed

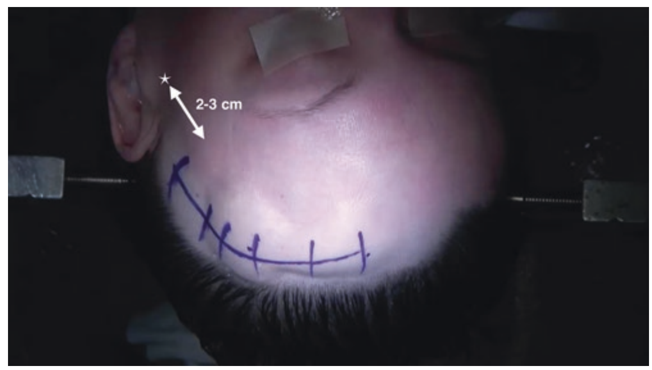

Fig. 11.2 Marking and preparing for an incision. The oblique frontotemporal skin incision is started 2–3 cm above the zygoma (*asterisk*) and continued behind the hairline. The incision is stopped 2–4 cm before the midline depending on localization of the lesion. Then incision line is injected with 20 ml of solution containing local anesthetic with epinephrine

trajectory, a slight retro- or antero-flexion can be used. The exact head position is determined by the pathology and adjusted case by case. Some general principles are:

- The head is rotated only slightly to the contralateral site for visualization of the proximal Sylvian fissure, otherwise the temporal lobe obstructs the trajectory.
- The extension of the head is determined by the height of the pathology from the floor of anterior fossa (for higher pathologies, more extension is used).
- Lateral tilt helps to center the proximal Sylvian fissure.

After positioning of the head, minimal shaving of hair, and skin disinfection, the planned incision is marked with a sterile surgical pen behind the hair line, and an intradermal injection of a vasoconstrictive and local anesthetic solution (lidocaine 5 mg/ml and epinephrine 10 ug/ml) is injected along the drawn incision (Fig. 11.2).

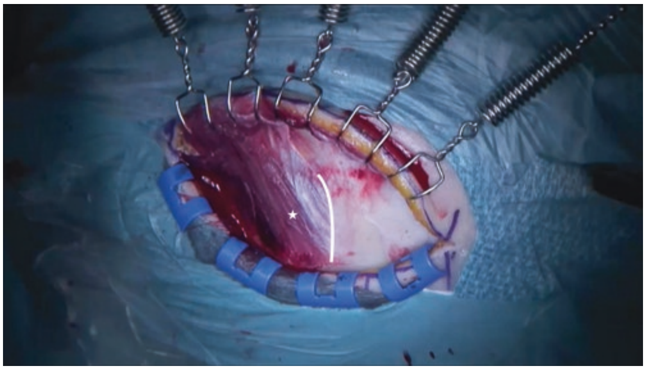

Fig. 11.3 One-layer flap. An oblique frontotemporal skin incision is made in one-layer fashion. The wound is opened with traction of spring hooks fixed to the Sugita frame. Bleeding from the lower margin of the wound is secured with Raney clips. The temporal line (*white line*) and temporal muscle (*asterisk*) are visualized approximately in the center of the wound

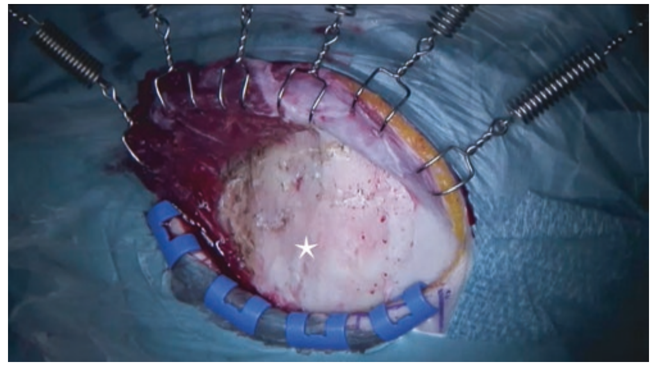

Fig. 11.4 Skeletonized bone. After detaching the temporal muscle with a monopolar knife, the skin flap and muscle are retracted with spring hooks. A burr hole is placed just under the temporal line (*asterisk*), in the point of connection 2/3 and 1/3 of line

Instrumentation

For incision and soft tissue exposure, standard instruments are used (e.g., scalpel, forceps, periosteal elevator, bipolar, monopolar, curved dissector, Raney clips, needle holder, irrigation syringe, suction tip). A burr hole is made by pneumatic drill and the dura is detached. A drill with a cutting blade is used for the craniotomy. Oozing of the bone is managed by "hot drilling" – which is drilling with a diamond blade without using water for cooling the drill.

Technique

A curved frontotemporal skin incision is performed centered to the processus zygomaticus of frontal bone. The incision is made slightly behind the hairline and stopped 2–3 cm above the zygoma (Fig. 11.3). Raney scalp clips are placed at the posterior margin of the incision for hemostasis. Usually the temporal muscle is seen attached to the superior temporal line, where it is cut vertically and by a short incision. The frontal part of the cut temporal muscle is then detached from the cranium with a periosteal elevator and monopolar and retracted frontobasally with the skin flap using a strong spring hook. After muscle retraction, the superior orbital rim, the processus zygomaticus of frontal bone, and a small portion of the temporal bone are visible (Fig. 11.4).

One burr hole is placed under the superior temporal line. The dura is detached from the cranium with a curved dissector. The bone flap is turned with a craniotome in three cuts: (1) from the burr hole toward the frontal base, (2) from the burr hole toward the frontal bone and sphenoid ridge, and (3) over the sphenoid ridge to connect both initial cuts with a

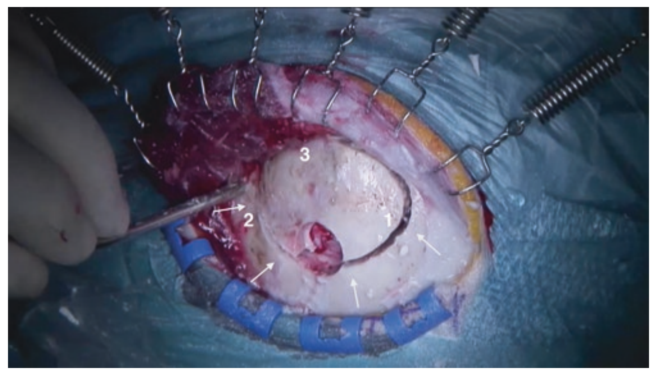

Fig. 11.5 Craniotomy. After placing the burr hole, the side-cutting drill is used to saw right (*1*) and left (*2*) limbs of the craniotomy. Then the craniotome blade without the footplate is used to connect the flap by thinning the bone between the previous two limbs (*3*). Before cracking and lifting up the bone flap, three to five drill holes (*white arrows*) are placed on the perimeter of "mainland" bone for tack-up sutures

blade drill (craniotome drill without the usual cover). Finally, a bone flap of around 3–4 cm is cracked and elevated (Fig. 11.5). The next step is the removal of the sphenoid ridge with a sharp cutting blade drill followed by the use of the diamond drill to obtain more extradural working space before dural opening. General techniques are used to obtain a blood-free surgical field such as "hot drilling" (Fig. 11.6). The dura is then opened in a curved incision. Multiple lift-up sutures are placed to avoid epidural bleeding and to retract the dural flap frontobasally (Fig. 11.7). An orbitozygomatic stitch is performed to achieve more operative space to maneuver [9] (Fig. 11.8).

After opening the dura, the basal frontal lobe is gently retracted using a cottonoid and bipolar forceps, and CSF is released by opening the arachnoid of the exposed Sylvian fissure or the opticocarotid cisterns. This relaxes the brain and produces both visualization of the proximal Sylvian fissure and access to the optic and carotid regions [3].

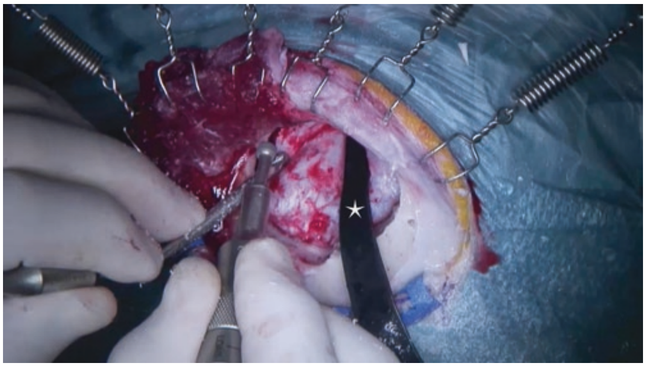

Fig. 11.6 Drilling the lateral sphenoid ridge. To obtain adequate access to the skull base and minimize retraction, drilling of the lateral sphenoid ridge is necessary. To decrease the risk of dural injury, a self-retaining retractor (*asterisk*) can be used. The drilling starts with a cutting drill and continues with a diamond drill, which is also helpful in “hot drilling” – a special technique to stop oozing blood from the bone

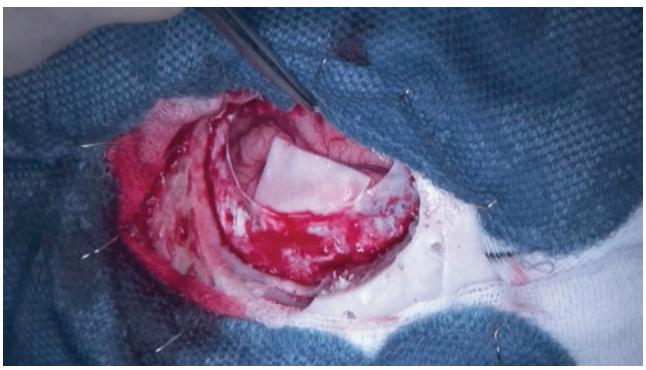

Fig. 11.7 Dura opening. Before opening the dura, the wound is irrigated, hemostasis is performed, and soft tissue is covered with swabs and a green cloth. Then the dura is opened in a curvilinear fashion pointing anterolaterally

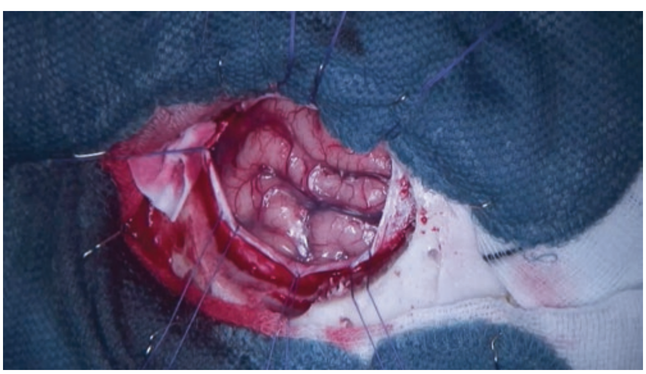

Fig. 11.8 Final stage of approach. Multiple stitches are used to elevate the dural edges. These stitches are fixed onto drapings with hemostats. This helps to prevent epidural oozing during the operation

Anterior Clinoidectomy

If anterior clinoidectomy is needed to access the proximal carotid artery, we prefer an intradural clinoidectomy tailored according to the working space required and the necessary exposure. The dura over the anterior clinoid process is coagulated by bipolar forceps, after which the coagulated dura is opened with a #15 blade scalpel. A micro-dissector is used to detach the anterior clinoid process from its dural cover, following which the cortical bone is drilled down with a 2 or 3 mm diamond drill. Bleeding from the cancellous bone can be stopped by fibrin glue injection or bone wax. The remaining thin bone layers can be mobilized with a dissector and removed with a micro-rongeur.

Aneurysms

General Aspects

Following the LSO craniotomy and opening of the dura, it is important to release CSF by opening the arachnoid of the Sylvian fissure or the carotid and optic cisterns in order to achieve sufficient relaxation of the brain and gain the necessary working space without applying strong retraction to the frontal lobe. After adequate CSF release, no fixed retraction is needed. Gentle minimal retraction applied by cottonoids and the use of bipolar and suction are sufficient to keep the exposure and visibility of the operative field. We prefer to use a 1 ml syringe with an 18G needle (i.e., needle knife) for opening the arachnoid of the proximal Sylvian fissure and the micro-hook or bipolar to dissect the arachnoid of the more basal cisterns.

MCA Aneurysms

Both the MCA bifurcation and the origin of the anterior temporal artery in the M1 segment can be accessed through a small, targeted 1–2 cm opening of the Sylvian fissure placed appropriately in the vicinity of the aneurysm (Fig. 11.9). Length of the M1 segment is used to estimate the proper placement of the keyhole opening on the proximal Sylvian fissure [3, 6, 8].

ACoA Aneurysms

Approach to the ACoA region starts by release of CSF from the carotid and optic cisterns, followed by further dissection of the arachnoid between the optic chiasm and medial frontal lobes. The origin of the ipsilateral A1 is then located and the A1 segment followed to the ACoA region. The contralateral A1 is identified next, and a short segment of both A1s is dissected free for the possible use of temporary clips in case they are needed for proximal control. Great care should be taken to avoid damage to the perforators arising from the A1 segments and the ACoA region, as well as the arteries of Heubner. If necessary to improve visualization of the ACoA region and proximal A2 segments, further brain relaxation can be obtained by release of CSF from the third ventricle by opening the lamina terminalis just posterior to the optic chiasm.

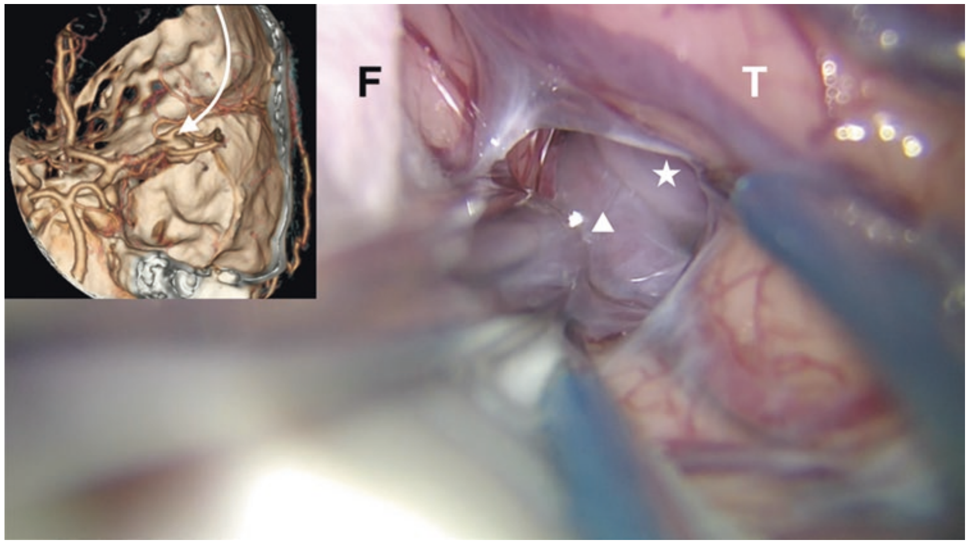

Fig. 11.9 Unruptured middle cerebral artery (MCA) bifurcation aneurysm through right LSO approach. In the left upper corner – 3D reconstruction of computed tomography angiography showing route to the aneurysm (*white arrow*). In the middle – focused opening of the Sylvian fissure. *White arrowhead*, MCA bifurcation; *white asterisk*, aneurysm; *F* frontal lobe, *T* temporal lobe

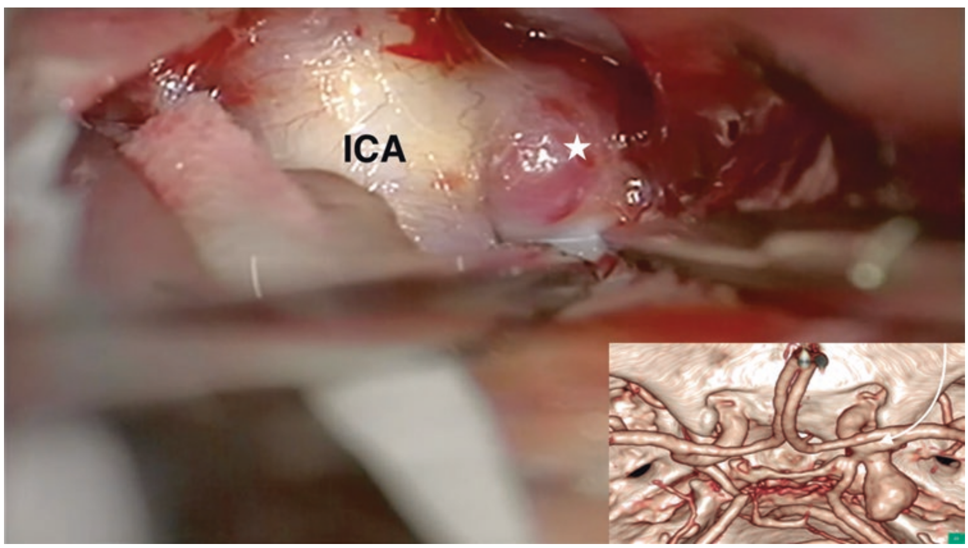

Fig. 11.10 Unruptured posterior communicating artery (PCoA) segment of internal cerebral artery (*ICA*) aneurysm through right LSO approach. In the bottom right corner – 3D reconstruction of computed tomography angiography showing route to the aneurysm (*white arrow*). In the middle – exposed PCoA-ICA aneurysm (*white asterisk*)

ICA Bifurcation and ICA-PCoA Aneurysms

The carotid cistern is opened to release CSF and relax the brain, which allows one to follow the supraclinoid segment of the ICA until the bifurcation. Great care is needed to avoid damage to the perforator arteries arising from the ICA bifurcation and proximal A1 and M1 when dissecting free the neck or the dome of the ICA bifurcation aneurysm. For ICA-PCoA aneurysms (Fig. 11.10), temporal extension of the LSO bone flap is usually helpful. Anterior clinoidectomy may be needed to gain proximal control of the ICA. Vigilance is needed to preserve the anterior choroidal artery when clipping a PCoA aneurysm [7, 10].

Basilar Bifurcation Aneurysms

LSO is a suitable approach for basilar bifurcations that are located at the level of the posterior clinoid process or higher [11]. These basilar bifurcation can be exposed through the carotid-optic and carotid-oculomotor triangles [11] (Fig. 11.11).

Meningiomas

General Aspects

As for the aneurysms, CSF release from basal cisterns is mandatory to achieve sufficient working space without strong retraction of the frontal lobe. Therefore, the first dissection follows the sphenoid ridge toward the optic and carotid cisterns, where CSF can easily be released. The second important step is the devascularization of the tumor from its main feeders by dissection and coagulation of the dural attachments. The third step, tumor debulking, can be required

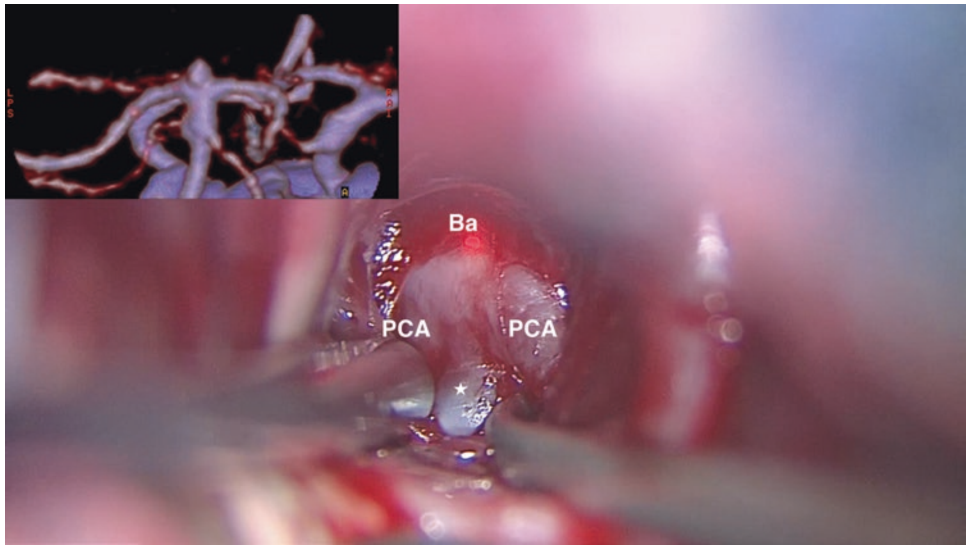

Fig. 11.11 Ruptured basilar tip aneurysm through right LSO approach. In the upper left corner – 3D reconstruction of computed tomography angiography showing basilar tip aneurysm. In the middle – exposed aneurysm (*white asterisk*) through the opticocarotid triangle. *PCA* posterior cerebral artery, *Ba* basilar artery

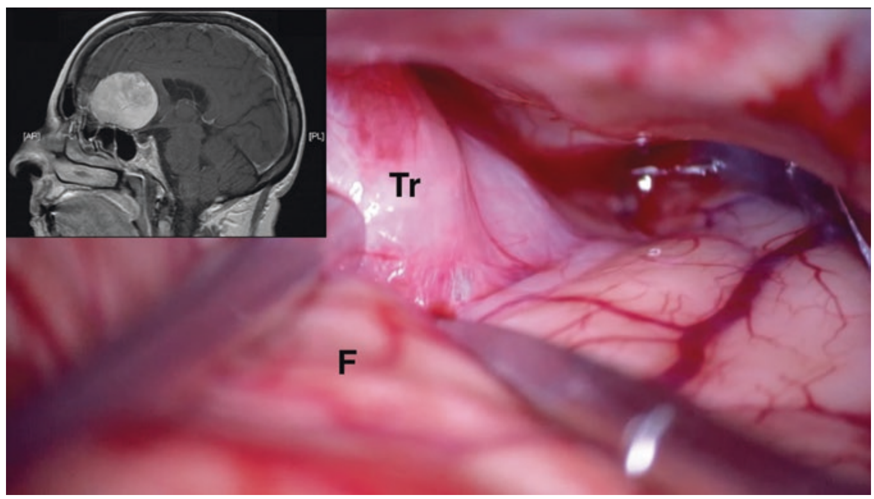

Fig. 11.12 Olfactory groove meningioma through right LSO approach. In the upper left corner – contrast-enhanced sagittal magnetic resonance imaging showing large tumor. In the middle – exposed meningioma. *F* frontal lobe, *Tr* tumor

if the tumor is large and obstructs the view toward significant structures or to obtain sufficient working space. Use of the ultrasonic aspirator (CUSA) can significantly help in tumor debulking. The final step is the dissection between the tumor and brain surface. The dissection is always done toward the tumor surface away from the normal brain tissue. Water dissection facilitates the layer separation, such as within the Sylvian fissure. This dissection always starts at a location where the border zone between the tumor and cortex can be clearly identified. Separation of structures is maintained using small cottonoids.

Anterior Clinoid Meningiomas

In anterior clinoid meningiomas, the ICA, MCA, ACA, optic nerve, and oculomotor nerve can be affected by the tumor formation. The micro-Doppler probe is useful for vessel identification. The main challenge is to detach the optic nerve from the tumor while avoiding new visual deficits. In firm and adherent tumors, it can be necessary to leave tumor in situ for risk minimization. Large tumors require a temporal extension of the previously described craniotomy. In large anterior clinoid meningiomas, it can be necessary to remove parts of the tumor to reach the basal cisterns [12, 13].

Tuberculum Sellae Meningiomas

The dura of the planum sphenoidale and tuberculum sellae is coagulated for devascularization of tumor feeders. In proximity to the optic canal, this must be done carefully. The ICA, MCA, ACA as well as the ACoA complex, optic nerve, and pituitary stalk must be securely identified and preserved [12].

Olfactory Groove Meningiomas

Olfactory groove meningiomas (Fig. 11.12) in general obtain their vascular supply from the ethmoidal arteries that branch from the ophthalmic artery. Coagulation of those feeders is necessary for devascularization of the tumor, but great care has to be taken to preserve the olfactory nerves. To safely perform devascularization, identification of the olfactory nerves is essential, especially the contralateral nerve. It is important to note that in the case of large olfactory groove meningiomas, feeders can also arise from the ACA. Hyperostosis present in the anterior fossa can be drilled down, but if there is opening of the ethmoidal cells, these must be closed carefully to prevent CSF leakage [12, 14].

11.6 Complications

Injuries to the frontal sinus, especially in cases with lateral extension, are possible. Another potential complication during lateral drilling is unintentional opening of the orbital roof. Defects in both of these areas can be closed directly. Epidural hematoma can be avoided by meticulous hemostasis and lift-up sutures. Due to the slight temporal muscle incision and the location of the skin incision, facial nerve injury and temporal muscle atrophy usually do not occur [15, 16].

11.7 Surgical Pearls

- Head positioning should be performed according to 3D visualization of the lesion.
- A short, curved incision is centered on the zygomatic process of the frontal bone.
- The one layer skin muscle flap is retracted with a strong spring hook in a rostral direction.
- A single burr hole is placed at the temporal line.
- Extradural bone removal of the sphenoid ridge is performed with a diamond drill to stop bone bleeding ("hot drilling").
- The LSO provides exposure of the sphenoid wing, MCA, anterior fossa, ICA, ACoA complex, parasellar region, and basilar bifurcation.

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Endoscopic and Keyhole Approaches to the Cerebellopontine Angle

12

Melvin Field and Luke H. Pearson

12.1 Introduction

The retrosigmoid suboccipital approach (RSA) is commonly referred to as the workhorse approach to gain access to the cerebellopontine angle (CPA). The first successful report of approaching the lateral posterior fossa was over 100 years ago by Sir Charles Balance [1]. Over the next 50 years, however, the mortality from this approach remained greater than 50% [2]. During the first half of the twentieth century, the goal of surgery was patient survival. During the 1930s and 1940s, advancements in antiseptic technique and anesthesia allowed Dandy and Olivecrona to demonstrate that this approach to the CPA can be safely performed with an acceptable mortality of less than 20%. Continued improvements in magnification, illumination, and instrumentation occurred in the second half of the twentieth century, and the goals of such surgery now became to cure the pathology involving the CPA. Over the past 25 years, improvements in surgical neurophysiological monitoring and instrumentation and further advancements of visualization refined the expected goals to also include preservation of neurological function. Today, expectations from CPA surgery include curing the pathology of question while at the same time preserving cranial nerve function, cerebellar function, and brainstem function. In addition, patients, hospitals, and insurers have increasing expectations regarding hospital length of stay, recovery, return to work, pain, and disability.

Fully endoscopic surgery has quickly become the standard of care for minimally invasive neurosurgery for pathology of the sella. The endoscope was first used as an adjunct for the microsurgical resection of pituitary lesions. The panoramic view and use of angled scopes provided by the endoscope allowed the neurosurgeon to view portions of the parasellar region that were out of the view using the microscope. For other areas of the brain such as the CPA, the endoscope has not been as eagerly adopted. Endoscope-assisted microscopy, in which the endoscope is used as a complementary device to the microscope, is commonly used for cases requiring access to the CPA. Endoscope-assisted microscopy is a useful start for neurosurgeons aiming to advance to a fully endoscopic approach for microvascular decompressions and the resection of tumors. When the technique is mastered, the fully endoscopic keyhole approach to the CPA leads to better outcomes with fewer complications than microscopy alone.

The RSA has undergone considerable modifications in search of the safest and least invasive approach to pathology of the CPA since being first described. As a result, the retrosigmoid keyhole approach is now the unrivaled route for neurosurgical access to pathological processes of the lateral cerebellar hemisphere, the petroclival region, the foramen magnum, the jugular foramen, and the CPA. This chapter details the retrosigmoid suboccipital keyhole approach to the CPA including a step-by-step guide for employing the technique. The CPA and surrounding area contain important neurovascular structures that can lead to severe neurological deficits when damaged. An intimate understanding of the relationship between the cranial nerves, arteries, brainstem, and cerebellar surfaces within the posterior fossa is necessary before undertaking this approach.

12.2 Rationale

The lateral suboccipital retrosigmoid approach allows for the best visualization of the cranial nerves at their root entry zone and their bony entry points into the skull base. By utilizing the keyhole retrosigmoid approach to the CPA, a safer and more reliable method of access is achieved. The incorpo-

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ration of an endoscope allows the surgeon better illumination through a smaller surgical corridor, a wider field of view than can be accomplished with a microscope, and an improved ability to see medial and lateral structures of the CPA without increasing retraction or drilling of bone intracranially.

The keyhole RSA incision and opening are small and the technique is generally time efficient. The approach provides an unrivaled exposure to the ventral posterior fossa. With the incorporation of an endoscope, most pathology of this region can be accessed and treated. The approach-related morbidity is significantly lower than other more invasive approaches to the ventral posterior fossa.

12.3 Patient Selection

Indications/Contraindications

By utilizing the retrosigmoid keyhole approach, a variety of pathologies involving the CPA can be surgically addressed [3] (Table 12.1). Microvascular decompression (MVD) can be performed for vascular compression syndromes including trigeminal neuralgia, hemifacial spasm, glossopharyngeal neuralgia, geniculate neuralgia, tinnitus, and disabling positional vertigo. MVD is perhaps the easiest procedure to become comfortable with a keyhole craniectomy and the endoscope-assisted or fully endoscopic technique as it provides for relatively normal tissue planes and is performed for a process that does not result in increased intracranial pressure. Small- to medium-sized tumors of the CPA can also be surgically removed via an endoscope-assisted and fully endoscopic keyhole retrosigmoid craniectomy with relative ease. By combining retrosigmoid keyhole craniectomies, large tumors of the CPA can also be removed. Any pathology that can be addressed microscopically via a RSA can also be effectively treated with endoscope assistance or via a fully endoscopic technique.

Table 12.1 List of potential pathology that can be treated via the keyhole RSA

Vestibular schwannoma
Trigeminal schwannoma
Jugular foramen schwannoma
Meningioma
Epidermoid tumor
Glomus jugulare tumor
Trigeminal neuralgia
Glossopharyngeal neuralgia
Hemifacial spasm
Posterior circulation aneurysms
Geniculate neuralgia
Brainstem cavernoma
Chordoma
Arachnoid cysts
Medically intractable vertigo

Relative contraindications to the keyhole RSA either with or without an endoscope include disease extending into the lateral one third of the internal auditory canal (IAC), tumors extending more than 2 cm anterior to the entrance of Meckel's cave, and tumors with significant extension into the posterolateral cranial base. This includes tumors with significant extension into the temporal bone including more than 5 mm extension into the jugular foramen and hypoglossal canal, as well as disease extending into the sigmoid sinus or jugular bulb. Other relative contraindications to the keyhole RSA include disease with extension of more than 10 mm above the tentorial edge or disease with extension beyond the middle third of the ventral skull base. Pathological processes extending beyond these limits can be debulked or biopsied via this approach but not completely excised. In addition, the procedure is relatively contraindicated in patients who do not have a patent contralateral transverse and sigmoid sinus. Procedure-specific systemic contraindications include patients with chronic otitis media or mastoid infections and patients who cannot be temporarily discontinued from antiplatelet or anticoagulant therapies.

12.4 Surgical Anatomy

The surface of the suboccipital region is relatively small, and the triangular configuration of the CPA is perfectly applicable to keyhole or corridor surgery. A large incision and craniotomy/craniectomy do not significantly improve visualization or minimize retraction in this part of the head. As a result, the surgical anatomy of the entire CPA can be visualized and manipulated via three separate or combined 1–1.5 cm corridors (Fig. 12.1a–d). The three keyhole variations are based on the location of the vestibulocochlear complex relative to the keyhole and the main cerebellar artery of the region [4]. A rostral keyhole places the vestibulocochlear complex at the base of the surgeon's corridor, and the superior cerebellar artery is the major vascular complex of the corridor. The middle keyhole centers the vestibulocochlear complex and the anterior inferior cerebellar artery in the surgical corridor, and the caudal keyhole places the vestibulocochlear bundle at the roof of the surgeon's field and centers the posterior inferior cerebellar artery and vertebral artery in the corridor. The rostral keyhole corridor allows for visualization and direct manipulation of the upper half of the CPA. This includes the region from and including the tentorial incisura to just below the vestibulocochlear complex (Fig. 12.2a–c). The middle keyhole corridor allows for visualization and direct manipulation of the middle half of the CPA. This corridor allows the surgeon access from the petrosal venous complex and the trigeminal nerve down to the 9th, 10th, and

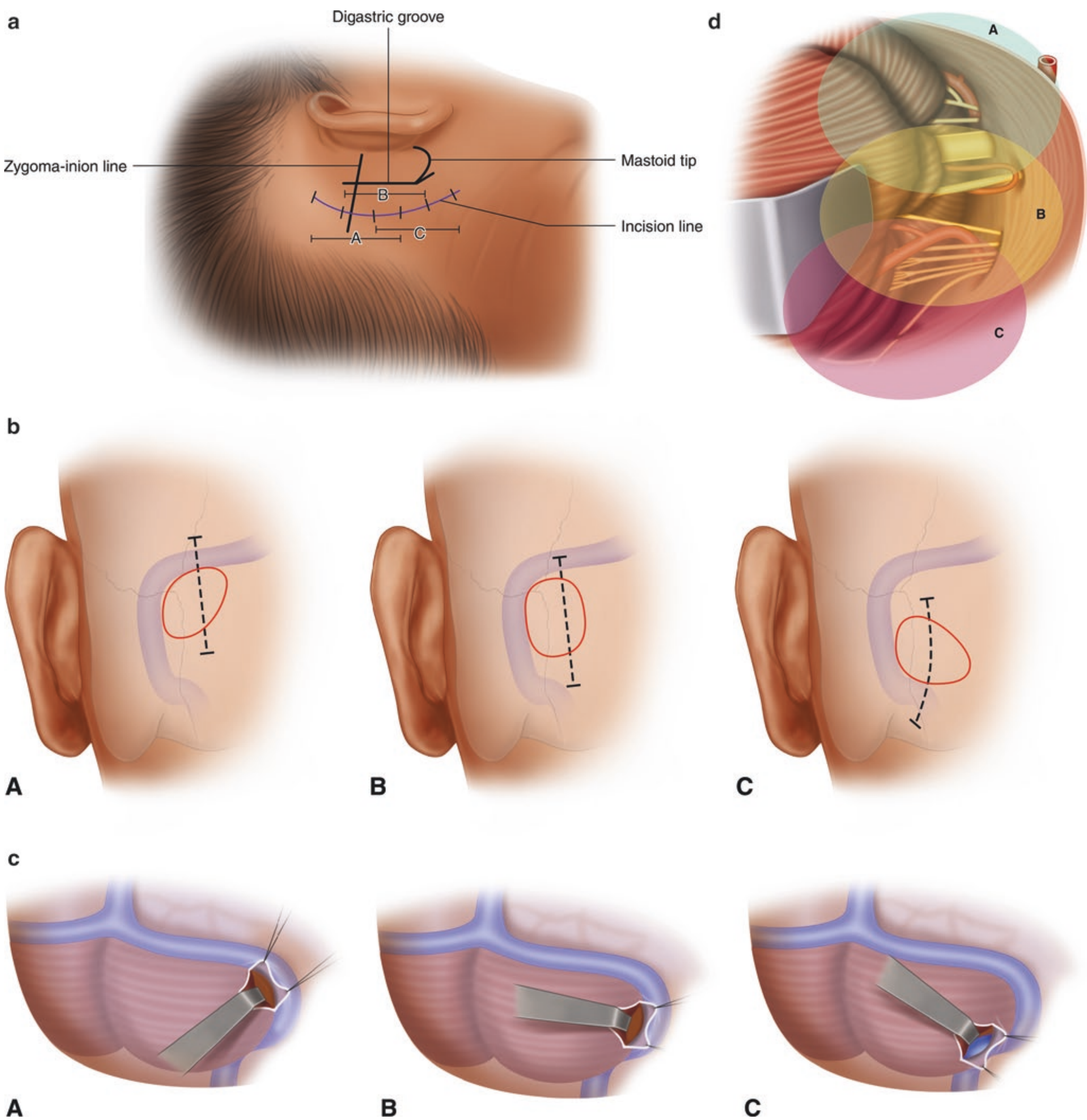

Fig. 12.1 Cutaneous, bony, and dural openings for rostral, middle, and caudal keyhole retrosigmoid approaches to the CPA. *A*. Location of incision relative to zygoma-inion line for each approach; *B*. location of bony opening relative to transverse and sigmoid sinuses for each approach; *C*. dural opening relative to transverse and sigmoid sinuses for each approach; *D*. CPA anatomy easily accessed and visualized for each approach (within *A–D*: *A* rostral approach, *B* middle approach, *C* caudal approach)

11th nerve complex (Fig. 12.3a–g). Finally, the caudal keyhole corridor allows visualization and direct manipulation of the lower half of the CPA. This region essentially includes the vestibulocochlear complex superiorly down to the upper surface of the foramen magnum (Fig. 12.4a, b). The use of angled endoscopes (30–70°) and an advanced comfort level operating with angled lenses can expand the visualization and to some degree the manipulation of structures up or down an adjacent corridor as the surgeon's experience with these corridors grows. For example, a middle keyhole RSA for a CPA meningioma can allow for tumor removal into the tentorial incisura or below the jugular foramen as the central tumor is debulked and angled lenses and instrumentation are utilized. However, the keyhole corridor chosen should be

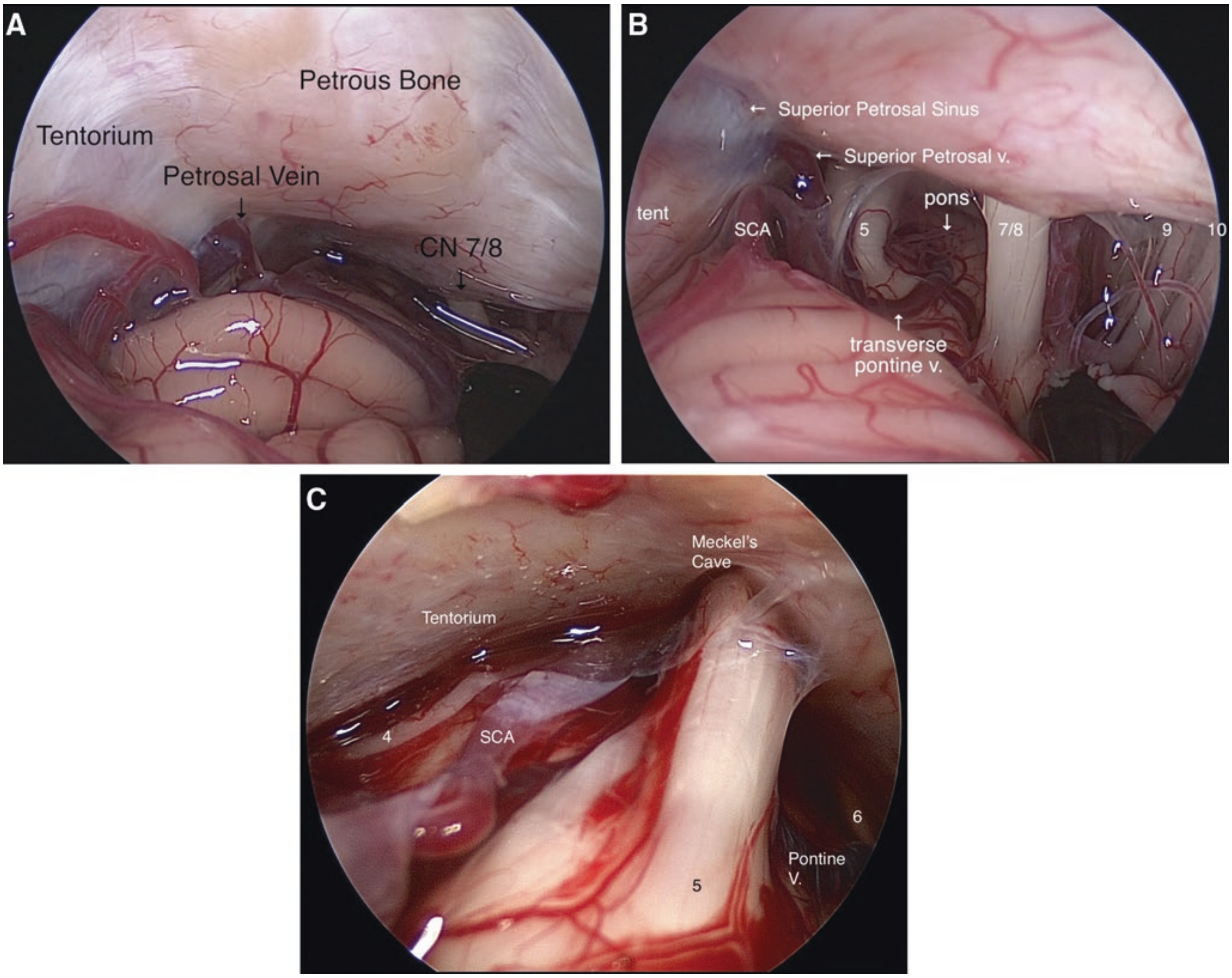

Fig. 12.2 (**a**–**c**) Rostral CPA anatomy. (**a**) Initial endoscopic keyhole exposure of the lateral upper CPA. Superior visualized field includes tentorium above with the petrosal venous complex. The inferior visualized field is the vestibulocochlear complex. (**b**) Rostral CPA anatomy using a 0° endoscope with clear visualization of the tentorium and petrosal sinus down to the superior aspect of the jugular foramen structures. (**c**) Lateral view of the rostral CPA with an excellent view of the distal trigeminal nerve exiting into Meckel's cave as well as the trochlear nerve as it travels above the tentorial incisura. *CN 7/8* facial/vestibulocochlear nerve complex, *tent* tentorium, *SCA* superior cerebellar artery, *5* trigeminal nerve, *4* trochlear nerve

centered over the pathology. For large tumors or lesions extending from the tentorial incisura down to below the jugular foramen, we will often combine our keyhole craniotomy to a size of about 2–2.5 cm in diameter to enhance tumor resection without the need for extreme lens angles or need to more frequently change our scope position.

12.5 Surgical Technique

Operative Setup

Proper patient positioning and operating room setup are vital for successful keyhole surgery of the posterior fossa. Variables affecting the operative setup include (1) neck range of motion, (2) patient weight, (3) pathology laterality, and (4) surgeon handedness.

All operations should be performed under general endotracheal anesthesia. A scopolamine patch is applied behind the contralateral ear to aid in minimizing postoperative nausea. Upon entering the operative suite, patients are given 50 g of mannitol IV, and a Foley urinary catheter is placed once under general anesthesia. In obese patients and patients who take routine diuretics, 20 mg IV furosemide is also administered. We prefer to have mannitol infused and, if needed, furosemide administered 60–90 min prior to dural opening to benefit from its maximal diuretic effect at the time of surgery when the posterior fossa will seem most full (i.e., prior to draining cisternal CSF). Once intubated, the anesthesia provider mildly hyperventilates the patient to achieve an

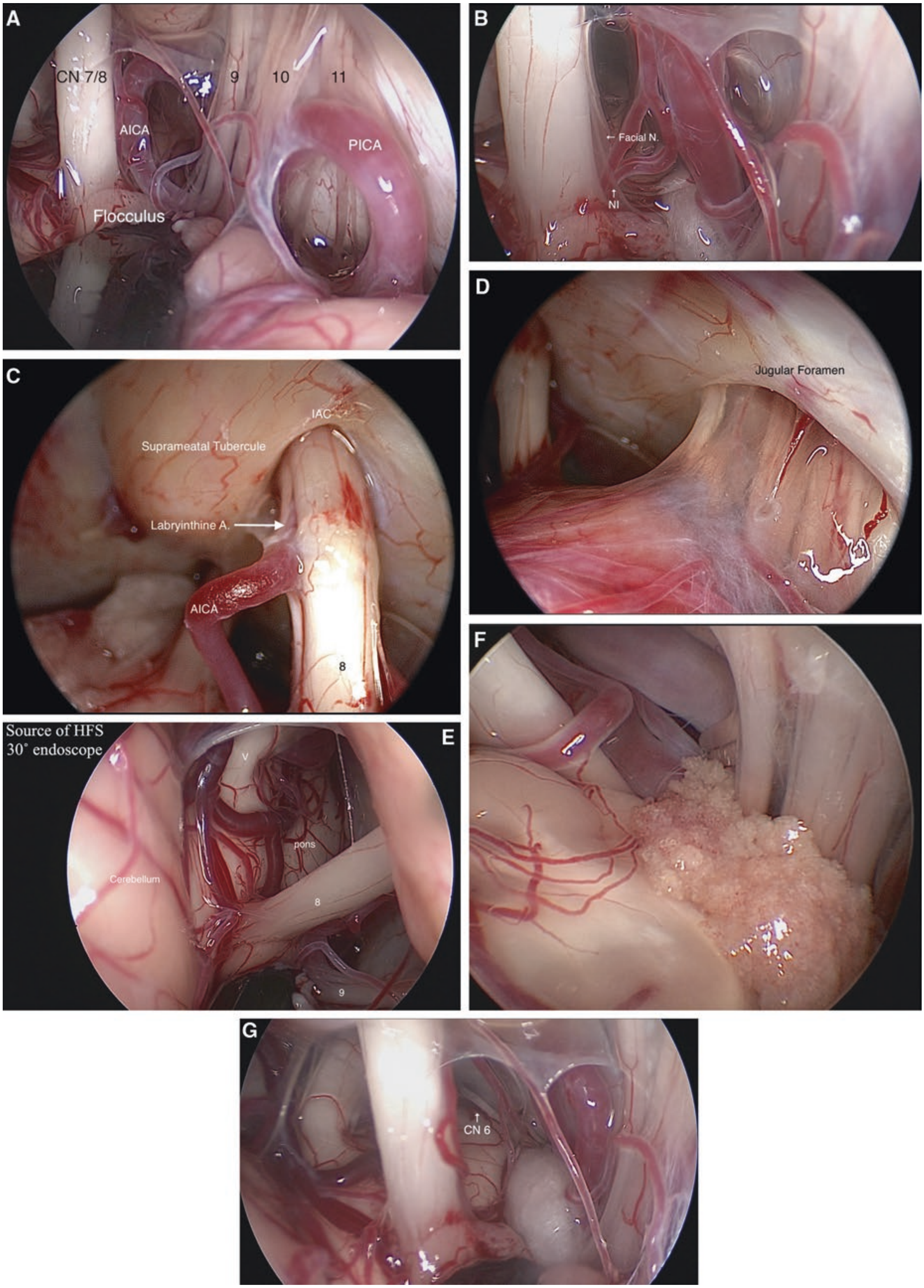

Fig. 12.3 (**a**, **b**) Middle CPA anatomy. A 0° endoscope is used to visualize the structures of the CPA from above the vestibulocochlear complex to just below the jugular foramen. The endoscope magnification and resolution actually allow the surgeon to see capillary flow on the neural elements as seen on the vestibulocochlear nerve in image **b**. (**c**, **d**) Using a 30° endoscope, the vestibulocochlear complex is well visualized beyond the suprameatal tubercle exiting into the petrous temporal bone via the internal auditory canal. The 9th, 10th, and 11th cranial nerves are also clearly viewed exiting the jugular foramen laterally. (**e**–**g**) Using the 30° endoscope to look medially, the root entry zones of the trigeminal nerve, vestibulocochlear nerve, and facial nerve are easily visualized. The posterolateral pons, flocculus, and choroid plexus of the foramen of Luschka are also seen via a middle CPA keyhole approach. *6* abducens nerve, *7* facial nerve, *8* vestibulocochlear nerve, *9* glossopharyngeal nerve, *10* vagus nerve, *11* spinal accessory nerve, *AICA* anterior inferior cerebellar artery, *PICA* posterior inferior cerebellar artery, *NI* nervus intermedius nerve

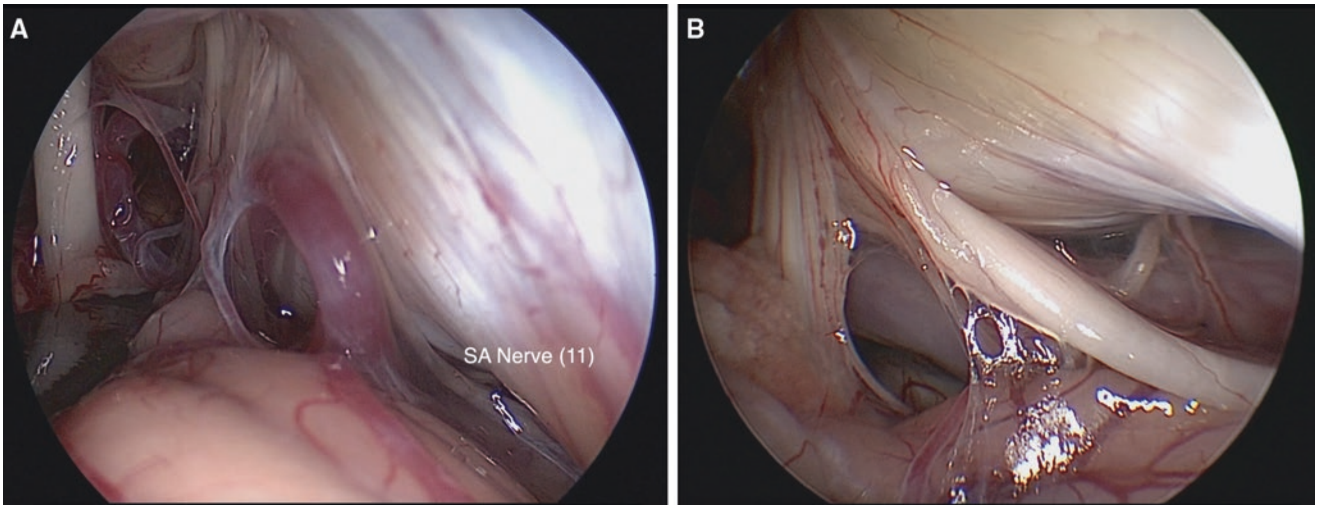

Fig. 12.4 (**a**, **b**) Lower one third of CPA. (**a**) Using a 0° endoscope, the surgeon has a clear view from the vestibulocochlear complex down to just above the foramen magnum. The AICA loop can be seen just below the vestibulocochlear complex and the PICA loop just below the nerves of the jugular foramen. (**b**) By directing the endoscope slightly more caudally, the vertebral artery, hypoglossal nerve, and spinal accessory branch extending down to the foramen magnum can be visualized. *SA/11* spinal accessory nerve

arterial CO_2 of 30 mm Hg. The use of mild temporary hyperventilation and diuresis at the beginning of surgery is performed to maximize brain relaxation at the time of dural opening and minimize the need for cerebellar retraction [5]. Once a posterior fossa arachnoid cistern is opened, cerebrospinal fluid drainage will allow for adequate posterior fossa relaxation, and the arterial CO_2 is then normalized. By employing mild hyperventilation and diuretics prior to exposing the CPA, lumbar drainage or ventricular CSF drainage is almost never necessary. For patients with severe fourth ventricular obstruction and hydrocephalus preoperatively, an external ventricular drain is placed, not to assist in accessing the CPA but rather to manage hydrocephalus in the perioperative period. Prophylactic antibiotics are given between 30 and 60 min of skin incision, redosed every 4 h during surgery, and continued for the first 24 h after surgery.

Operating Room Equipment and Staff Positioning

Whether positioned supine or in the lateral park bench position, anesthesia is situated on the side opposite the surgeon with the patient's head facing the anesthesia team. An arm board is positioned at a right angle to the table toward anesthesia allowing easy access to IV's or arterial lines and is padded to prevent compressive neuropathies. Next to anesthesia, the neurophysiological monitoring team's equipment and staff are positioned to allow easy communication with the anesthesiologist during the procedure and the surgeon. For right-handed surgeons, the scrub nurse is situated to the surgeon's right and to the left for a left-handed surgeon (Fig. 12.5a, b). For microscopic keyhole approaches to the CPA, the authors position the operating microscope base behind the surgeon on the surgeon's nondominant side (i.e., on the side opposite the scrub nurse). For a right-handed surgeon, the microscope's arm and lens apparatus comes over the surgeon's left shoulder to enter the surgical field. For endoscopic and endoscope-assisted approaches, the authors utilize a pneumatic endoscope arm holder. A multitude of different endoscope holder systems exist including bed-mounted mechanical and pneumatic systems, floor-mounted systems, and now robot and robot-assisted systems. The authors use the Mitaka UniARM floor-mounted pneumatic surgical support system with micromanipulator attachment (Mitaka Kohki Co., Ltd., Tokyo, Japan). The system allows for endoscope holder arm balancing, so there is no drift when the pneumatic arm is released or locked and its nitrogen-pressurized pneumatic joint system allows for 6° of motion freedom. A micromanipulator is attached (Karl Storz, Tuttlingen, Germany). We position the UniARM system's base on the side of anesthesia at the junction of the bed with the arm board and bring the pneumatic arm itself over the patient's head so that the endoscope system is positioned at 12 o'clock (superolaterally) in the surgical field away from the surgeon's working field (Fig. 12.6a, b). The endoscope camera and light source cords are taped over the top of the holder system to keep them away from the working field. The endoscope system is connected to a high-definition camera (at least 1080 pixel), and a high-definition monitor is placed at eye level facing the surgeon on the other side of the patient at the head of the bed. The surgeon is situated behind the patient's head and sits during the intradural portion of the surgery (Fig. 12.5b).

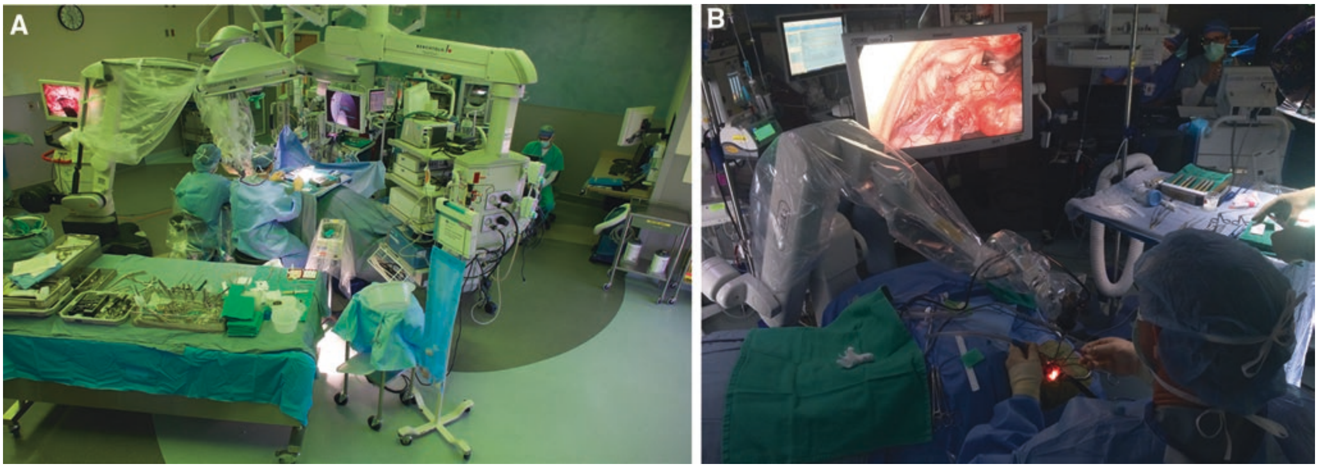

Fig. 12.5 (**a**, **b**) Operating room staff ergonomics. (**a**) Right-sided microscopic keyhole retrosigmoid approach. The patient is in the lateral park bench position facing the anesthesia team. Neurophysiology is positioned next to anesthesia, and the surgical assistant/scrub nurse is to the surgeon's right (for a right-handed surgeon). The microscope is coming over the surgeon's nondominant side. (**b**) Left-sided endoscopic keyhole retrosigmoid approach. The patient is facing the anesthesia team and neurophysiology is next to anesthesia. The surgical assistant is on the surgeon's right (for a right-handed surgeon). Note the relationship of the OR assistant (on surgeon's dominant side), endoscope holder, and monitor relative to the surgeon for an endoscopic retrosigmoid keyhole approaches to the CPA

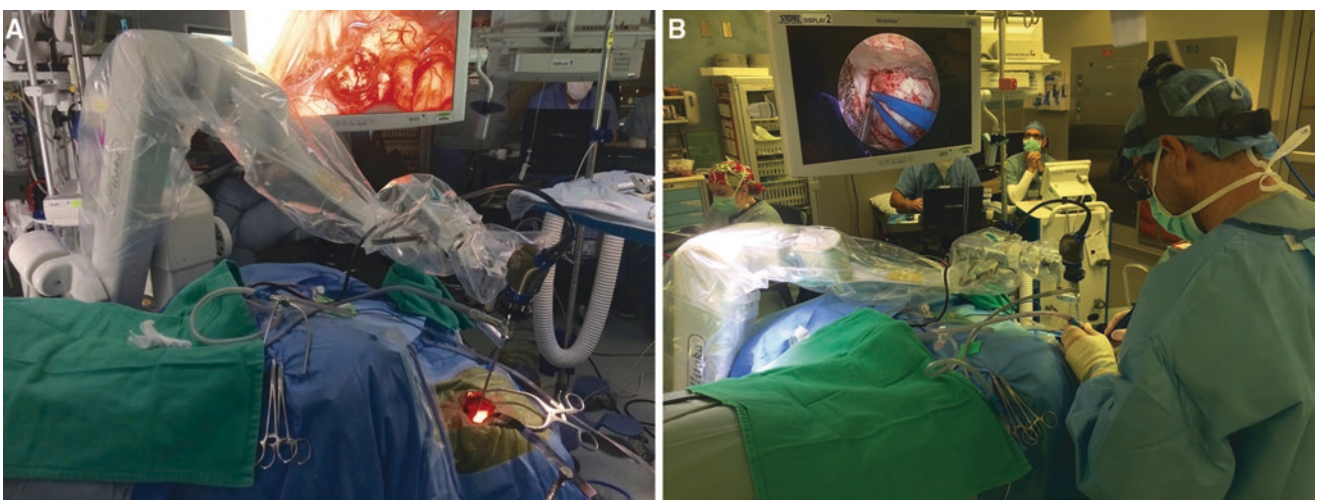

Fig. 12.6 (**a**, **b**) The endoscope holder is attached/positioned just below the patient's arms, at the level of the chest, and extends over the torso into the surgical field at 12 o'clock. The video monitor is positioned at the surgeon's eye level and between the anesthesia team and the patient's face. (**a**) Fully endoscopic left-sided approach with the surgeon sitting. (**b**) Endoscope-assisted left-sided approach with the surgeon standing

Patient Positioning

The goals of patient positioning for keyhole posterior fossa surgery are to (1) minimize tissue obscuration of your target pathology, (2) minimize patient pressure points or discomfort postoperatively as the result of positioning, and (3) maximize surgeon neck, arm, and hand comfort during the procedure.

Positioning of the patient undergoing a RSA to the CPA is one of the most important aspects of the operative setup. Prior to positioning, the patient's head should be secured. We prefer using a Mayfield three-pin head clamp. The patient should be placed in the supine position with the head rotated at least 70–80° toward the contralateral side of the incision. Placing the patient in the supine position ensures that the shoulders will not interfere with the angles necessary for instrumentation through the keyhole (Fig. 12.7a–e). However, this is only appropriate in patients with excellent neck range of motion and is often unachievable in the more elderly population, in patients with degenerative cervical spine disease, and in patients with previous cervical fusions. Some centers also consider significant cervical carotid or vertebral artery disease to be a relative contraindication to

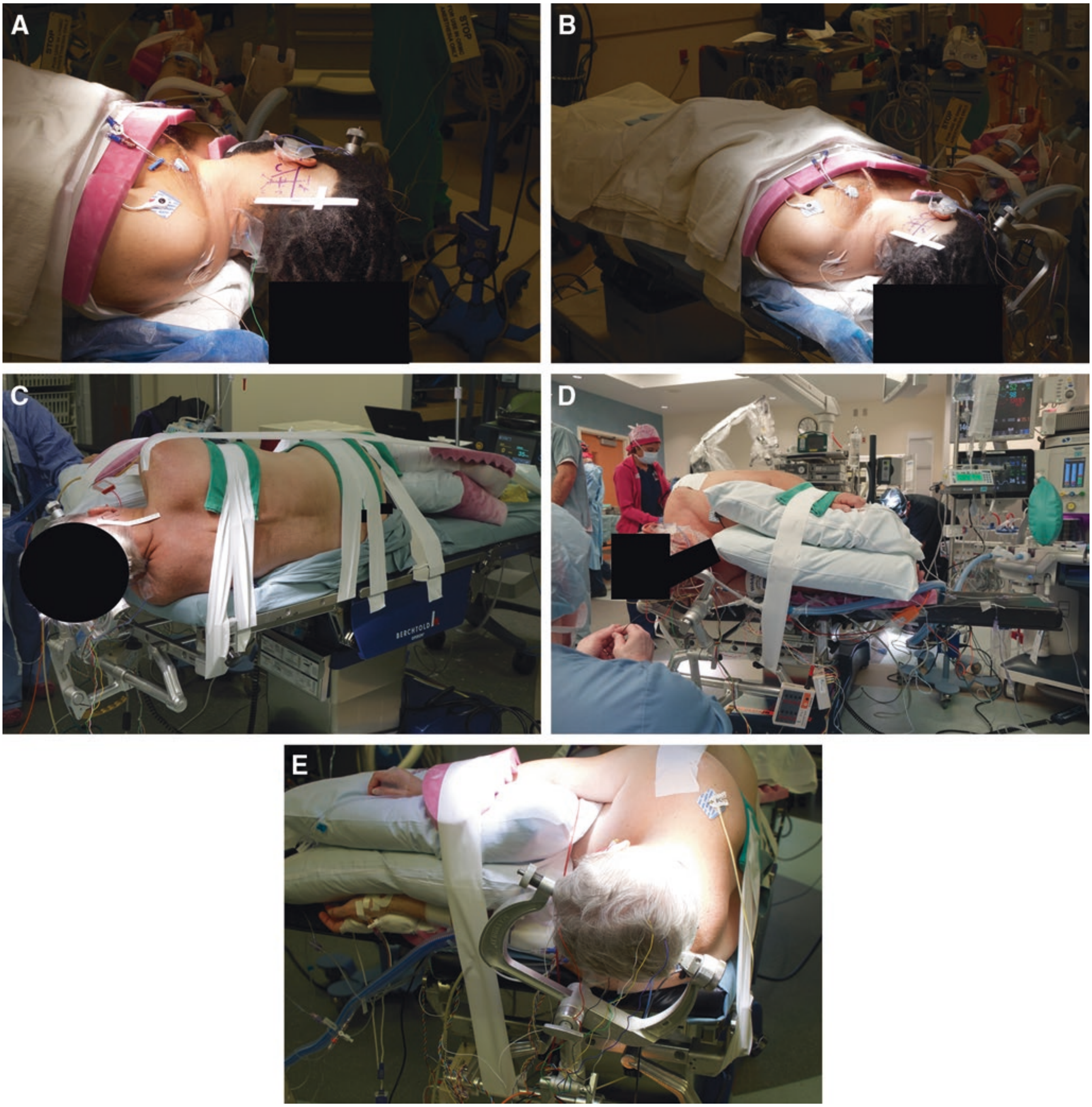

Fig. 12.7 (**a**, **b**) Patient positioning supine with head turn. The patient is placed in a head holder, and the head is turned 70–80° away from the surgeon so that the mastoid eminence is the highest bony landmark on the skull relative to the floor. Padding is placed under the shoulder and torso to minimize strain on the neck, and the patient is secured to prevent shift during table rotation at the time of surgery. (**c**–**e**) Patient positioning in the lateral park bench position with the knees flexed and the upper shoulder taped and mildly retracted away from the head. The patient is in a head holder, and the head is rotated and tilted down very slightly so that the mastoid eminence is the highest point on the skull. An axillary role is placed, and the arms face the anesthesia team hugging pillows to well pad them. This allows access to the IVs and airway by anesthesia during the procedure and minimizes undue pressure along the upper extremities to help decrease the risk of compressive neuropathies. The patient is well padded and secured at multiple locations – torso, hips, and thighs – to minimize movement during table tilting at the time of surgery

positioning in the supine position with head rotation due to the risk of arterial occlusion or ischemia. An alternate position, and the one that we most often utilize in our practice, involves the lateral decubitus position. In the lateral decubitus (aka lateral park bench) position, the patient is positioned laterally on the contralateral side of the incision site, and either a beanbag is used to secure the patient or cloth tape with foam padding is used to secure the patient in position on the operating room table. The legs are bent to flex the patient at the hip and knees. Axillary rolls and pillows are used to help pad the patient (Fig. 12.7a–e). The patient's head should be rotated between 0° and 20° toward the contralateral side of the incision and slightly flexed. The head should not be turned far enough to compromise venous drainage in the neck. The goal of the positioning is to place the mastoid eminence at the highest point on the skull. We also tape the ipsilateral shoulder down to maximize the space between the shoulder and operative field, but we are careful not to retract the shoulder too much to induce damage to the shoulder itself or cause a compressive neuropathy (Fig. 12.7a–e). The area to be shaved should extend approximately 2–3 cm behind the ear. The patient should be secured and padded onto the table to allow approximately 20–35° of bed rotation either toward or away from the surgeon during the procedure without any movement or shift of the patient on the table. Bed rotation aides the surgeon with visualization either more laterally or medially during the procedure. By rotating the patient, less cerebellar retraction is necessary, and less uncomfortable head, neck, and arm positioning by the surgeon is required resulting in less surgical fatigue and increased surgeon comfort. For this reason, we generally prefer tape, pillows, and foam padding over a beanbag during positioning.

Neurophysiological Monitoring

The major roles for the use of neuromonitoring in posterior fossa surgery are to prevent iatrogenic neurological deficits and to map and/or identify neurological structures in the operative site. Table 12.2 lists which modalities are used based on the keyhole approach utilized. To accomplish these goals, we monitor the following modalities: brainstem auditory evoked responses (BAER); free-running EMG for cranial nerves V, VII, VIII, IX, X, XI, and XII; stimulus-triggered EMG for CN VII, CN IX, and CN X; and somatosensory evoked potentials (SSEPs). For BAER, we stimulate both the ipsilateral and contralateral ears. Once baselines are established, no changes are made to filter settings or repetition rate. All mandatory peaks (I, III, and V), when present, are identified. Any increase in wave V latency greater than 1 ms or a decrease greater 50% in amplitude is alerted to the surgeon. Free-running EMG is recorded using bipolar subdermal electrodes placed in ipsilateral muscles: masseter, orbicularis oculi, orbicularis oris, stylopharyngeus, trapezius, and tongue. We also use an EMG endotracheal tube to record EMG activity from CN X. For stimulus-triggered EMG, we use a Medtronic NIM 3.0 to stimulate and record responses from orbicularis oculi, orbicularis oris, stylopharyngeus, and vocal cord muscles. Electrical stimulation is used throughout the dissection of tumor to identify important neural structures involved in the tumor capsule. A rectangular cathodal pulse of 100 us width is delivered through Kartush instruments at 4 Hz. Responses from target muscles are displayed on screen and through a loudspeaker. We start dissection using stimulation intensities of 0.5 mA, and as neural structures are being identified in the field, the stimulus intensity can be reduced to 0.05–0.1 mA. Median nerve and ulnar nerve SSEPs are used to detect neurological deficits that can be caused by compression of the axilla or traction of the shoulder.

Table 12.2 Neurophysiological modalities monitored in keyhole CPA surgery

Approach			
Modality	Rostral keyhole	Middle keyhole	Caudal keyhole
Bilateral BAERS	X	X	X
CN V free-running EMG	X	X	
CN VII free-running EMG	X	X	X
CN IX/X free-running EMG		X	X
CN XI free-running EMG		X	X
CN XII free-running EMG			X
CN VII stimulus-triggered EMG		X	X
CN IX stimulus-triggered EMG		X	X
CN X stimulus-triggered EMG		X	X
Median/ulnar SSEP	X	X	X

Technique

Anatomical landmarks to be palpated prior to skin incision include the mastoid tip, the digastric notch, the zygomatic arch just in front of the tragus, and the inion. A line connecting the root of the zygoma and the inion approximates the location of the transverse sinus relative to the incision, and the digastric groove approximates the posterior margin of the sigmoid sinus (Fig. 12.8). The skin incision generally

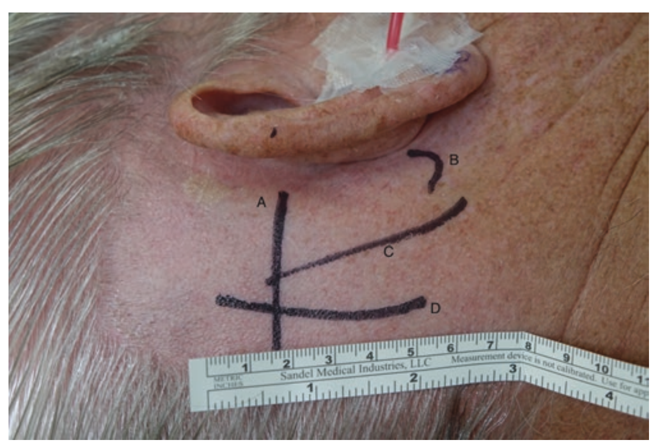

Fig. 12.8 Cutaneous landmarks used for determining incision location with a keyhole RSA to the CPA. The planned incision is 2 cm or 1.5–2 fingerbreadths, behind the ear, and is 4–5 cm in length. A line connecting the zygoma root with the inion approximates the transverse sinus. The digastric groove running just behind the mastoid bone approximates the posterior margin of the sigmoid sinus. *A* zygoma-inion line, *B* mastoid tip, *C* digastric groove, *D* typical planned incision

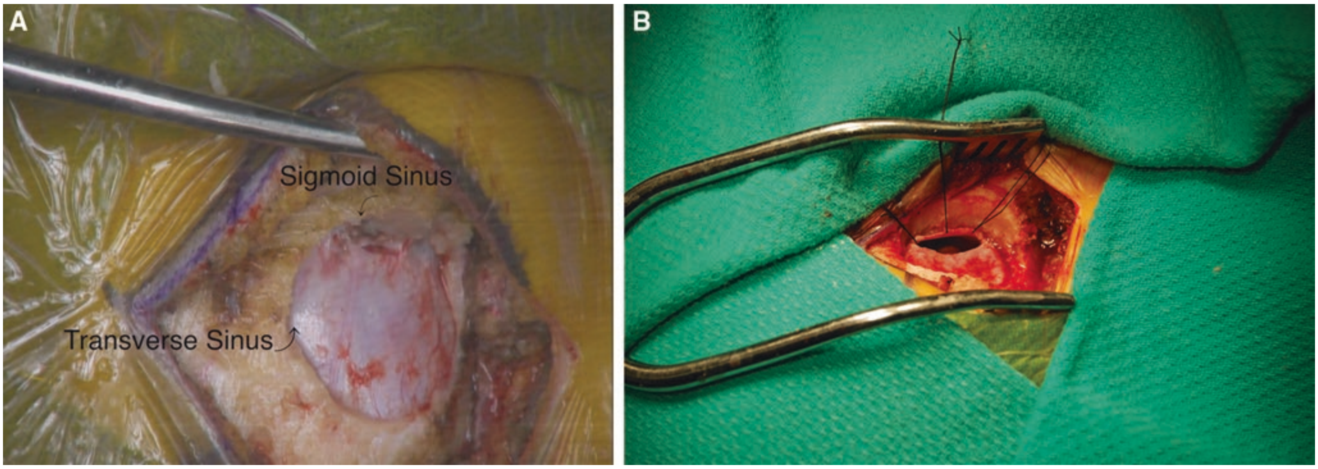

Fig. 12.9 (**a**, **b**) Right retrosigmoid keyhole craniectomy. Standard approach to upper and mid-CPA. (**a**) Exposure includes visualization of inferior margin of the transverse sinus and medial margin of the sigmoid sinus. We recommend skeletonizing these venous dural structures to identify their junction. The junction serves as the upper lateral extent of the dural opening. The initial bony opening is inferior and medial to the sinuses. A Kerrison Rongeur is then used to open more laterally and superiorly until the sinuses are identified. (**b**) Dural opening and retractor placement

is 2 fingerbreadths posterior to the posterior ear crease, 1 cm posterior to the digastric line, and up to, just past, or one third past the zygoma-inion line depending on whether a rostral, middle, or caudal keyhole craniectomy is planned. A single burr hole approximately 1 cm in diameter should be placed at the posterior border of the estimated junction of the sigmoid and transverse sinus. If the posterior margin of the sigmoid sinus is not visualized with your burr hole, then the craniectomy should be extended laterally until it is visualized to confirm that you are lateral enough to gain access to the CPA via a minimal dural opening. If performing a middle CPA keyhole approach, the upper lateral junction of the cranial opening should visualize the junction of the transverse and sigmoid sinus, and if performing a rostral CPA keyhole approach, the opening should visualize the inferior margin of the transverse sinus and the transverse-sigmoid junction (Fig. 12.9a, b). Once the dura is adequately exposed and relevant dural sinuses visualized, the bone edges and any exposed mastoid air cells are sealed with bone wax to prevent postoperative CSF leakage resulting in rhinorrhea, and the wound is copiously irrigated with antibiotic saline to remove bone dust and any loose tissue from the wound. This prevents foreign material from becoming intradural during the surgery and thus will decrease the risk of postoperative chronic headaches. Our soft tissue retractors are then adjusted to minimize interference with introduction of instrumentation into the CPA. Usually "fishhook" scalp retractors or a single cerebellar retractor works well. An approximately 8–10 mm "C"-shaped dural incision is made with the pedicle of the dural opening being the sigmoid sinus. The dura is then tacked up laterally and secured with a stitch reflecting the dura toward the sigmoid sinus. The dural opening should be as far lateral as possible, up to the edge of the sinus, in order to decrease the need for or amount of cerebellar retraction to gain access to the CPA (Fig. 12.9a, b).

The first step in accessing the CPA endoscopically is the positioning of the endoscope itself. The authors use the Karl Storz Hopkins endoscope collection (Karl Storz, Tuttlingen, Germany), although several different endoscope systems are commercially available and are easily adaptable for endoscopic CPA keyhole surgery. A 0°, 2.7 mm outer diameter offset endoscope is generally used instead of a 4 mm sinus endoscope due to the demand for available working space through the keyhole. The 4 mm scope may be of use in cases of large tumors involving the CPA, in which the burr hole and dural opening must be larger. For tumors requiring combined keyhole openings, the 4 mm scope is often used as it does provide better resolution than the 2.7 mm scope. An irrigation sheath is not necessary in the posterior fossa to prevent obscuration of the neurovascular structures by blood or CSF as there is usually very little to no "rundown" during these approaches. In addition, the irrigation sheath decreases the size of the surgeon's working channel making tissue manipulation more restricted.

Once the dura is opened, the operating table is lowered as low as it will go allowing the surgeon to look over the wound in a comfortable sitting position. The bed is then rotated slightly away from the surgeon to allow exposure along the petrous face toward the deep CPA cisterns. A nonstick cottonoid patty is placed in an anterior inferior direction to prevent trauma to the cerebellum and allow for gentle retraction of the cerebellum. The arachnoid is then dissected and CSF is released to slowly relax the cerebellum. As CSF drains and the cerebellum becomes more relaxed, the patty is advanced either under endoscopic or microscopic visualization to access the cerebellopontine cistern arachnoid planes. The arachnoid plane is then sharply opened with an arachnoid knife, and the entire posterior fossa relaxes with minimal to no retraction. Although not necessary, the authors often now place a low-profile malleable cerebellar retractor along the trajectory of approach, not to retract the tissues but rather to protect the cerebellum. Nonstick cottonoid patties could alternatively be used. The patient is now rotated either further away from the surgeon to look further laterally at the IAC, jugular foramen, petrous apex, or midline ventral skull base or rotated toward the surgeon to look more medially at the brainstem and root entry zones. When using the microscope, we now set the light intensity at 90–100% to maximize light into the wound. When using an endoscope, we set the light intensity to 50%, which provides adequate illumination of the CPA while minimizing heat at the lens tip.

A clear difference between an endoscopic approach and a microscopic approach is instrument introduction. With a microscope, the surgeon has a clear view of instrument insertion from the cranial opening down to the focal point of dissection within the CPA, and the entire corridor can be used for instrumentation insertion and removal. The field of view, however, at the level of the CPA is narrowed compared to that of an endoscope requiring more frequent scope position changes, patient table position changes, and often more tissue retraction to see more medially, laterally, superiorly, or inferiorly from the center of your opening. Alternatively, with an endoscope, the surgeon blindly advances instruments under the endoscope lens shaft from the dural opening to just distal to the camera tip. As a result, proximal tissue (i.e., cerebellar) trauma during insertion and removal of instrumentation from the dural opening to the endoscope tip can occur if the tissue is not protected or if the surgeon is not cognizant of this possibility during instrument introduction and removal [6]. For this reason, the authors will often use a thin retractor to protect along this trajectory from the dural opening to the tip of the endoscope camera. Beyond the endoscope lens tip, however, the visualization of the CPA is superior to that of a microscope. The field of view is larger, and lens resolution and magnification are good enough to actually see capillary flow against neural elements. The larger field of view results in fewer lens manipulations, fewer changes in table turning, and less retraction to see medially, laterally, superiorly, and inferiorly. The endoscope also allows for visualization beyond the commonly encountered bony protuberances of the lateral skull base obscuring clear visualization of the internal auditory canal or Meckel's cave (Figs. 12.2a–c and 12.3a–g). As a result, it is less common to require intradural drilling to access disease entering the IAC or intradural lateral skull base. This decreases the risk of CSF leak, cranial nerve injury, and chronic headache. When intradural drilling is needed, the use of an angled endoscope allows for better visualization of the air cells to ensure adequate sealing with bone wax prior to closure. For both microscopic and endoscopic keyhole retrosigmoid approaches to the CPA, the authors use Rhoton (V Mueller™, Becton, Dickenson, Co, Franklin Lakes, NJ, USA), Jannetta (KLS Martin, Jacksonville, FL, USA), Kartush (Medtronic, Minneapolis, MN, USA), and Storz Sepehrnia (Karl Storz, Tuttlingen, Germany) micro-instruments. The specific technique of tumor resection or MVD is the same for keyhole surgery as it is for a traditional suboccipital craniotomy approach. Internal tumor debulking and sharp capsular dissection are employed. For CPA tumor capsular dissection adjacent to or adherent to cranial nerves, stimulus-triggered EMG with Kartush microdissectors is used to minimize the risk of fascicular neural injury.

Endoscope-Specific Surgical Technique Considerations

An endoscope holder should be used to stabilize the endoscope and thus prevent injury to the surrounding neurovascular structures. Securing the endoscope also allows the surgeon to operate with two instruments as opposed to one. Microsurgery in the posterior fossa mandates bimanual dexterity, as should endosurgery of the same area. In general, the endoscope should be kept at the apex of the visualization triangle (12 o'clock). Typically, a 4- to 7-French suction tip is held in the nondominant hand and a microdissector, microscissor, or bipolar is held in the dominant hand. The positioning of the endoscope and instruments is crucial in avoiding the technical challenges faced by crowding within the CPA space. The instruments are passed underneath the endoscope and held at roughly 5 and 7 o'clock positions within the keyhole and at the distal visualization triangle. By maintaining the endoscope at 12 o'clock and keeping the left surgical instrument at 7 o' clock and the right at 5 o'clock, the surgeon can easily pass instruments down the keyhole corridor to the CPA and minimize instrument clashing and confusion [7–9] (Fig. 12.10a–c).

Initial exposure of the CPA and tissue dissection should be performed with a 0° endoscope. For most tumor resections and endoscopic MVD procedures, a 0° endoscope alone is adequate for performing the surgery. If dissection requires visualization more medial, lateral, superior, or inferior to that obtained with a 0° endoscope, a 30° or 45° endoscope can be used and rotated to visualize the region of interest. This is uncommon but can allow for dissection and visualization of pertinent anatomy and pathology outside the intended keyhole corridor approach. For example, when performing a rostral keyhole retrosigmoid craniectomy to access a trigeminal cisternal meningioma, an angled endoscope can be utilized to visualize and remove any disease extending into the mid-CPA such as the IAC or between the IAC and jugular foramen.

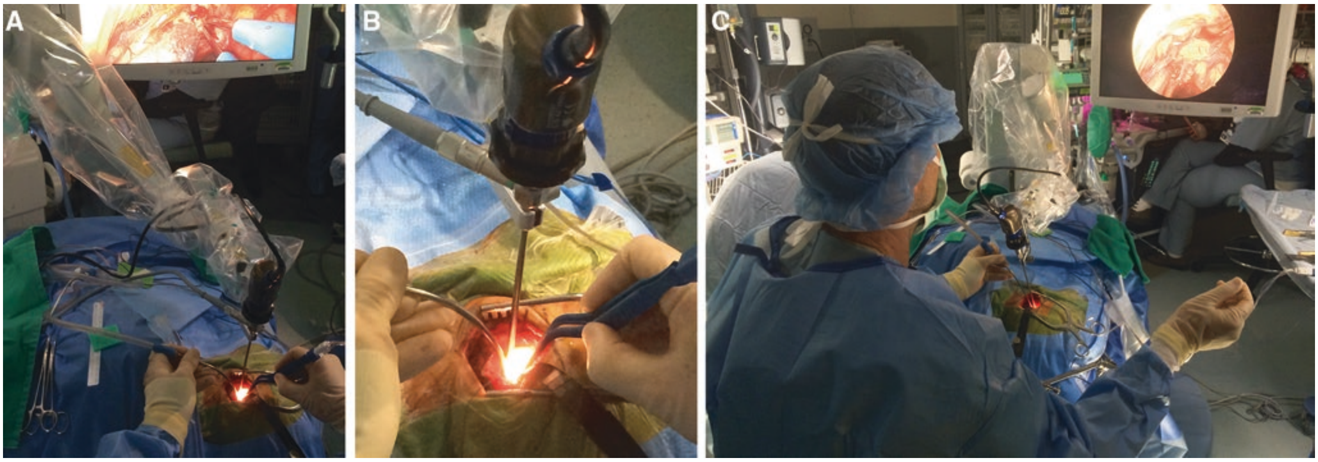

Fig. 12.10 (**a**–**c**) Triangulation of instrumentation relative to endoscope to optimize instrument movement under the endoscope. Endoscope positioned at 12 o'clock, left-handed instrument at 7 o'clock, right-handed instrument at 5 o'clock. (**a**) The endoscope camera cord and light source are secured away from the field so that they do not become tangled with the surgeon's removal and introduction of instruments below the endoscope into the keyhole corridor. (**b**) Positioning of instrumentation within the wound relative to the endoscope. Surgeon's left instrument is at 7 o'clock and the right instrument is at 5 o'clock (**c**) Location of surgeon's hands relative to endoscope. The endoscope comes from the opposite side of the patient and is positioned at 12 o'clock.

Closure

Once the goals of surgery are accomplished, any exposed intradural air cells are waxed under direct visualization. Then, the subdural CPA space is irrigated with warm saline to remove debris, minimize postoperative pneumocephalus, and observe for any sources of bleeding. Caution is used to make sure that direct irrigation over the cranial nerves is avoided to prevent injury. The dura is then closed in a watertight fashion. The craniectomy is closed with a piece of titanium mesh or bone cement to prevent muscular adherence to the dura resulting in post-craniotomy headaches. The fascial and subcutaneous layers are closed with absorbable sutures, and the skin is closed with a running 3-0 nylon suture. No drains are used. A sterile Telfa gauze bandage is placed.

12.6 Postoperative Care/Complications

After surgery, patients are observed in the neuro ICU or neuro-step-down unit overnight. Patients are continued on antibiotics for the first 24 h, and a scopolamine patch is kept in place until the patient is tolerating a regular diet. For tumor patients, an MRI of the brain with and without gadolinium is performed that night to evaluate for completeness of resection, bleeding, stroke, and hydrocephalus. For MVD patients, a CT head is performed. Systolic blood pressure is kept less than 160 mm Hg while in the hospital, and dexamethasone is given postoperatively only in patients with new postoperative neurological deficits, severe headache, or persistent nausea. Diet is advanced to regular as tolerated once the patient is awake, and ambulation is encouraged as soon as the patient arrives to their room. Patients are transferred to the neurosurgical floor on postoperative day 1. Most patients are discharged home on postoperative day 2 or 3. Bandages are removed and incisions are allowed to get wet with a shower 3 days after surgery. Sutures are removed 7–10 days after surgery, and patients are allowed to resume regular activities 2–3 weeks after surgery.

Retrosigmoid Keyhole CPA Surgery-Specific Complications

- Poorly positioned bony opening preventing access to pathology of interest
- Inadequate patient positioning and padding resulting in peripheral compressive neuropathy
- Cerebral stroke due to arterial compression in the neck or decreased venous return in the neck
- Sigmoid or transverse sinus injury resulting in stroke or hemorrhage
- Cerebrospinal fluid leak resulting in rhinorrhea or pseudomeningocele formation
- Cranial nerve injury
- Intradural arterial or venous injury resulting in posterior circulation stroke or hemorrhage
- Chronic headaches

- Cosmetic deformities
- Cerebellar injury with ataxia
- Hydrocephalus

12.7 Surgical Pearls

- Positioning for a keyhole endoscopic or microscopic approach to the CPA should ensure that the mastoid eminence is at the highest point of the skull. This will minimize bed turning and allow for the most comfortable hand positioning during surgery.
- The bony craniectomy of the keyhole approach to the CPA should extend laterally enough to visualize the sigmoid sinus edge, and the dural opening should extend to the sinus edge. This will minimize the need to retract the cerebellum and provide a less obscured view of the CPA.
- The most important initial step after opening the dura in a keyhole approach to the CPA is to identify and fenestrate an arachnoidal cistern to drain CSF. This relaxes the cerebellum and eliminates the need for traumatic tissue retraction.
- Endoscope and instrument positioning for keyhole CPA surgery should form a triangle. The endoscope should be positioned at 12 o'clock, the left-handed instrument at 7 o'clock, and the right-handed instrument at 5 o'clock.

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13 Neuroendoscopic and Keyhole Approaches to the Pineal Region

Hasan A. Zaidi and Peter Nakaji

Abbreviations

CSF Cerebrospinal fluid
CT Computed tomography
ETV Endoscopic third ventriculostomy
FLAIR Fluid-attenuated inversion recovery
hCG Human chorionic gonadotropin
MRI Magnetic resonance imaging
SCIT Supracerebellar-infratentorial

13.1 Introduction

The pineal region is located deep within the brain and cranial vault, and it is surrounded by a variety of important neurovascular structures. It is bordered by the splenium of the corpus callosum superiorly, the quadrigeminal plate of the rostral brainstem anteriorly and inferiorly, and the pulvinar on either side. The pineal gland is a small neuroendocrine gland and, along with the pituitary stalk, is one of the only midline intracranial structures that is unpaired. It is comprised of pinealocytes, whose primary function is to secrete melatonin directly into the systemic blood circulation [1]. Retinal neurons send information regarding ambient light to the suprachiasmatic nucleus of the hypothalamus; this information is relayed to the superior cervical ganglion of the sympathetic plexus and ultimately to the pineal gland, which regulates day–night cycles and moderates hormonal secretion. Primary disorders of pineal gland endocrine function are extremely rare. However, the region is host to a wide variety of tumors of various pathologies, which brings it to the attention of the neurosurgeon.

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13.2 Rationale

Various approaches to lesions in the pineal region have been extensively described [2–4], each with its own set of advantages and limitations. Historically, the pineal region was most often addressed either with stereotactic biopsy or via open posterior approaches. Most open approaches are achieved with the aid of microscopic visualization. During the past two decades, the endoscope has taken a preeminent role in many areas in neurosurgery, especially in skull base surgery. The endoscope improves illumination and visualization within a deep and narrow surgical corridor, which decreases approach-related morbidity. In the past few years, several groups have described [3, 5] endoscopic posterior approaches to the pineal region. With increased surgeon experience with this novel mode of visualization, the use of endoscopic approaches to the pineal region has transitioned from simple cyst fenestrations and tumor biopsies to large tumor resections and removal of vascular lesions. Because lesions in this area are located deep in the brain, a keyhole approach can offer an exposure that is as good as most traditional approaches. In this chapter, we describe the anatomy of the pineal region, the common pathologies encountered within this space, and the traditional open surgical approaches. We then contrast open approaches with our experience with endoscopic keyhole approaches to the pineal region.

13.3 Patient Selection

Any mass in the pineal region that causes symptoms should be considered for resection. Even a germinoma, which tends to respond well to radiation, can be considered for resection if morbidity can be kept low. Any lesion that has been demonstrated to grow should also be considered for resection, regardless of symptoms. Any pineal region lesion that can be removed through a traditional open suboccipital transtentorial or supracerebellar-infratentorial (SCIT) craniotomy can be also removed through an endoscopic keyhole approach.

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For the SCIT approach, a sitting-slouch position is preferred. However, this position is contraindicated in a patient with a patent foramen ovale in the heart because this condition carries a risk that air emboli will transit the septal defect and embolize to the brain.

Pineal Region Pathology

Among all intracranial tumors, pineal region tumors account for 0.1–1% of lesions in the adult population and for 9% in the pediatric population [6]. Tumors arising from the pineal gland tissue are known as pinealomas, with numerous subtypes depending on the cell of origin. Tumors arising from pinealocytes can give rise to pineocytomas or pineoblastomas. Those arising from glial cells can result in astrocytomas, oligodendrogliomas, or glioblastomas. More commonly, those arising from sequestered embryonic germ cells can give rise to germ cell tumors, including germinomas, embryonal carcinomas, lipomas, choriocarcinomas, teratomas, and yolk sac carcinomas. In addition to primary tumors of pineal origin, other lesions can be found in this location and must be included in the differential diagnosis. Other lesions include metastatic disease, cavernomas, pineal cysts, aneurysms, and meningiomas. Nearly 40% of all asymptomatic adults develop idiopathic calcification of the pineal gland, which can be misidentified as a tumor [6]. Pineal pathologies that can be accessed using an endoscopic keyhole approach include the following:

- Primary pineal tumors (pineocytoma, pineal tumor of intermediate differentiation, pineoblastoma)
- Germ cell tumors (germinomas, embryonal carcinomas, lipomas, choriocarcinomas, teratomas, or yolk sac carcinomas)
- Glial tumors (astrocytomas, oligodendrogliomas, or glioblastomas)
- Lipomas
- Meningiomas
- Pineal cysts

The most commonly encountered pineal region pathology in adults is the pineal region cyst, with some reports indicating an incidence as high as 23% [7]. Often, patients with these lesions are asymptomatic, and the lesion appears stable on repeat imaging. Although it can be tempting to treat these lesions, most patients should be followed with serial imaging. In some cases, a large cyst can result in a mass effect on surrounding pineal region structures, such as the superior colliculi or cerebral aqueduct. At our institution, surgical intervention is offered only when neurological findings, such as obstructive hydrocephalus or Parinaud syndrome, are attributable to the pineal region cyst.

The most commonly encountered pineal region tumors are germ cell tumors, which account for 50–75% of all cases [6]. These lesions occur more frequently in males and are often diagnosed during puberty. Among germ cell tumor subtypes, germinomas are the most common, accounting for 50% of germ cell tumors. Germinomas are often exquisitely sensitive to chemotherapy and radiotherapy. Prior to the availability of safe surgical options, many pineal region tumors were treated with chemoradiation with a presumptive diagnosis of germinoma without tissue confirmation, but this practice is not common today. Patients with nongerminomatous germ cell tumors have less favorable outcomes because these lesions often metastasize early in the course of their growth [8].

Pineocytomas account for 11–28% of primary pineal tumors [9] and have varying degrees of aggressive behavior. Grade 2 pineocytomas are often less aggressive and well circumscribed, with minimal invasion into normal surrounding brain tissue. Conversely, grade 4 pineoblastomas often shed tumor cells within the cerebrospinal fluid (CSF) pathway, and patients may present with distant metastases in the central nervous system. For this reason, patients with pineoblastomas require craniospinal radiation treatment, with close radiological follow-up for assessment of possible distant metastases.

Clinical Presentation and Diagnosis

Clinical features associated with pineal region lesions can vary depending on the endocrine functional status of the tumor, as well as its vascularity, size, and configuration. Large tumors or cysts often present secondary to mass effect on surrounding neurovascular structures. Progressive compression of the rostral interstitial nucleus of the medial longitudinal fasciculus of the midbrain can result in Parinaud syndrome, which is characterized by impaired vertical gaze, eyelid retraction, and pseudo-Argyll Robertson pupils. Cerebral aqueduct stenosis can result in obstructive hydrocephalus, with concomitant gait instability, confusion, and urinary incontinence. Patients with acute hydrocephalus can present with more ominous findings, such as brain herniation, obtundation, and acute loss of consciousness, and they may require emergent lateral ventricular CSF drainage. Patients with friable and vascular lesions can present with secondary extralesional hemorrhage that causes damage to surrounding neurological structures, or they can present with intraventricular hemorrhage and subsequent hydrocephalus. Germ cell tumors can produce human chorionic gonadotropin (hCG), which manifests as precocious puberty in young boys and secondary amenorrhea in young girls.

Radiographic imaging is instrumental not only in constructing a differential diagnosis of pineal region masses but also in planning surgical approaches to the lesion. Basic computed tomography (CT) scans are often the first-line diagnostic study used by general practitioners, and CT scans can provide valuable information on the calcification of pineal region masses, peritumoral hemorrhage, and early obstructive hydrocephalus. Germinomas are often hyperdense to the brain and engulf the native pineal calcification; conversely, pineoblastomas are hyperdense lesions that have peripheral or "exploded" calcifications. Mixed-density lesions are often indicative of teratomas, and gliomas of the tectal region are often hypodense or isodense without calcifications [10].

Magnetic resonance imaging (MRI) is the preferred imaging modality in the identification of pineal region masses, and it is useful in assessing the relationship of the mass to surrounding neurovascular structures. MRI provides poor information for assessing the calcification of the lesion but can provide detailed information to differentiate between primary pineal tumors and para-pineal masses [11]. The pineal gland is a circumventricular organ, and it thus takes up gadolinium avidly; similarly, lesions that arise from pineal gland parenchyma also often demonstrate avid gadolinium contrast enhancement on MRI. Germ cell tumors are often hypointense on T1-weighted imaging and hyperintense on T2-weighted imaging. Teratomas are heterogenous lesions characterized by marked hyperintensity on T1-weighted imaging and several calcified nodules. Pineoblastomas often demonstrate a greater degree of surrounding brain invasion, with irregular or indistinguishable borders as compared to pineocytomas, which are often well demarcated from the surrounding brain tissue [11]. Pineal region cysts are characterized on MRI as round masses with smooth surfaces that are isointense to CSF on T1- and T2-weighted images [12] with minimal edema to surrounding brain tissue on fluid-attenuated inversion recovery (FLAIR) sequences. Vascular lesions can be distinguished from other pineal region masses by using dedicated vascular imaging, with particular attention paid to the venous phase to assess for vein of Galen malformations in young children.

Serum and CSF tumor markers are a valuable component of preoperative evaluation of patients with suspected pineal region tumors. Similar to radiographic studies, these markers can only be suggestive of certain tumor subtypes, but occasionally they provide the clinician with diagnostic information. Germ cell tumors often retain expression of their primordial lineage, and they can express hCG and alpha fetoprotein in both CSF and serum. Although not diagnostic, elevated hCG and alpha fetoprotein can be indicative of an aggressive germ cell tumor, with an elevated CSF-to-serum gradient suggestive of an intracranial lesion. These markers may be most helpful in determining the anticipated response to surgery or radiotherapy or chemotherapy; however, active debate remains in the medical literature about the utility of these diagnostic studies. Ultimately, histopathology remains the gold-standard modality in the diagnosis of pineal region tumors.

13.4 Surgical Anatomy

The pineal region lies at the back of the third ventricle; it is bounded above by the junction of the velum interpositum, with the paired internal cerebral arteries in the roof of the third ventricle, immediately laterally by the posterior and habenular commissures, and further laterally by the pulvinar of the thalamus. The pineal gland rests on the tectum, immediately above the superior colliculi. Dorsally, the vein of Galen and, more posteriorly, the culmen of the vermis of the cerebellum bound the space. For anterior transventricular approaches, the relevant anatomy that must be considered is the foramen of Monro. For the anterior interhemispheric approach, the anatomy of the interhemispheric fissure and of the choroid plexus in the lateral ventricle must be considered. A subchoroidal approach will open into the posterior ventricle. The dominant surgical anatomy for the purpose of an endoscopic keyhole approach as emphasized in this chapter is the posterior paramedian SCIT region. The line between the inion and the root of the zygoma describes the approximate level of the transverse sinus. Under the transverse sinus, the intradural space is relatively open, except for occasional bridging veins between the superior cerebellar hemispheric surface and the tentorium. Beyond this, the cerebellum slopes up to the pineal region. In the midline, the culmen rides high and the precentral cerebellar vein tethers the cerebellum (Fig. 13.1). However, there is rarely any obstruction to a paramedian trajectory (Fig. 13.2a, b).

13.5 Surgical Technique

Anterior Endoscopic Approach

Histopathological analysis is imperative in the diagnosis and planning of adjuvant therapy for patients with pineal region disease. In patients with lesions that are large enough to result in obstructive hydrocephalus, anterior endoscopic keyhole approaches to the lateral ventricles can allow for both tissue diagnosis and CSF diversion. These approaches minimize approach-related morbidity compared to that with open microsurgical approaches, and they are often used by surgeons as a first-line treatment [13]. Endoscopic third ventriculostomy (ETV) is one such minimally invasive approach. It consists of guiding an endoscope transcortically into the lateral ventricles and fenestrating the tuber cinereum, which bypasses a block of CSF from lesions in the posterior third ventricle and aqueduct. The alternative route for CSF to flow

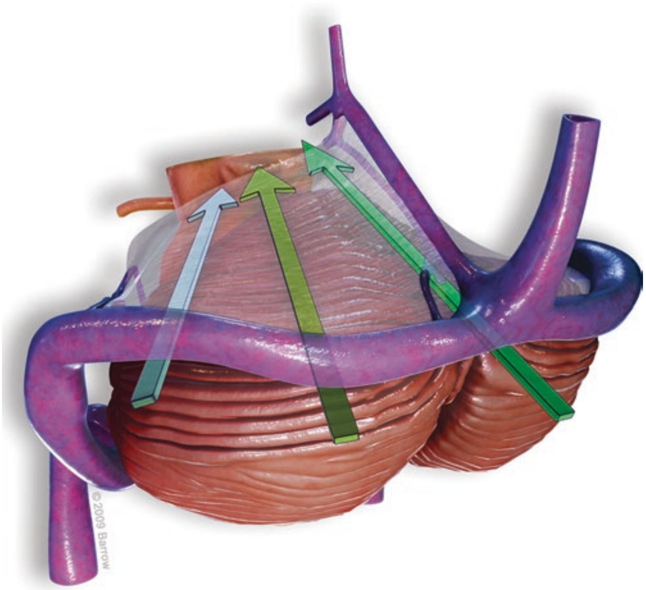

Fig. 13.1 The entire corridor from the retrosigmoid to the midline supracerebellar is available between the tentorium, which lies above the target area, and the superior surface of the cerebellum, which lies below the target area. The paramedian approach (*center arrow*) is the most advantageous for accessing the pineal region (Used with permission from Barrow Neurological Institute, Phoenix, AZ, USA)

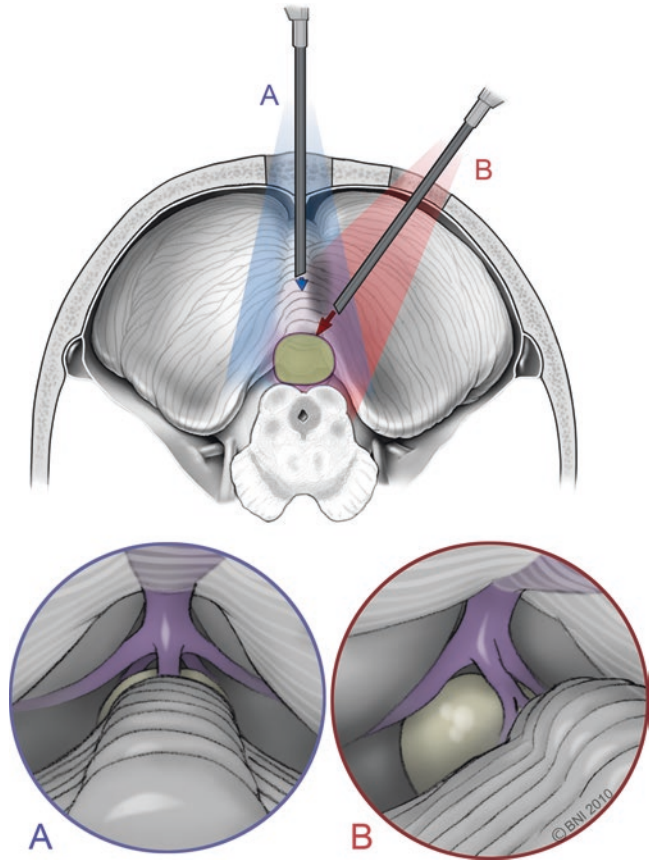

Fig. 13.2 Midline (**a**) and paramedian (**b**) approaches to the pineal region. The paramedian approach avoids obstruction by both the vermis and the bridging veins, and yet it still allows an excellent approach to both sides (Used with permission from Barrow Neurological Institute, Phoenix, AZ, USA)

into the basal subarachnoid space relieves hydrocephalus, such that a shunt is seldom required. This procedure is ideal for patients who have relatively recent-onset hydrocephalus from a pineal region mass because the natural CSF resorption pathways via the arachnoid granulations are intact. For patients with long-term obstruction, success in relieving hydrocephalus can often be variable [14].

The endoscopic unit used in an ETV consists of the endoscope with either a single port or dual ports to allow for placement of various tools, including a probe to make the initial ostomy and balloon catheters to expand the fenestration. The endoscope video output is projected onto a video monitor that is placed at eye level of the primary surgeon for comfort. An assistant also monitors and controls an irrigating pump that allows for clearing of debris and blood products within the lateral ventricles, which may otherwise obstruct the surgeon's view [15]. The procedure is performed by first planning a 1.5–2 cm incision located approximately 13–15 cm posterior to the nasion and approximately 2.5 cm lateral to the midline (Fig. 13.3). Image guidance is highly recommended because the trajectory varies substantially among individual patients. Special attention is placed on the location of this entry point with respect to the coronal suture in order to avoid injury to the motor cortex that would be caused by placing the entry too far behind the coronal suture. Stereotactic neuronavigation should be used to tailor the exact entry point as follows: the trajectory is derived by planning backward from the targeted site in the tuber cinereum, backward through the foramen of Monro, and projecting up to the cortex. Avoiding impact on the fornices is the most important consideration. A burr hole is then created after incising the skin, and a peel-away sheath is inserted into the ipsilateral lateral ventricle. After the lateral ventricles have been successfully catheterized by confirming the egress of CSF, the endoscope is advanced through the cannula, and the ventricular anatomy is identified including the foramen of Monro, column of the fornix, thalamostriate and septal veins, and thalamus. A detailed understanding of the ventricular anatomy is imperative to successfully and safely perform this operation because it is easy to become disoriented, especially if anatomy is distorted in patients with long-standing hydrocephalus. Once the surgeon is oriented, the endoscope is carefully advanced into the foramen of Monro to visualize the third ventricular floor. It is imperative to avoid sweeping motions when in the ventricle in order to avoid shear injury and to prevent vascular injury or excessive traction of the fornix, which is particularly sensitive to manipulation. Anatomical understanding of the third ventricle is also important when performing the next few steps. The floor of

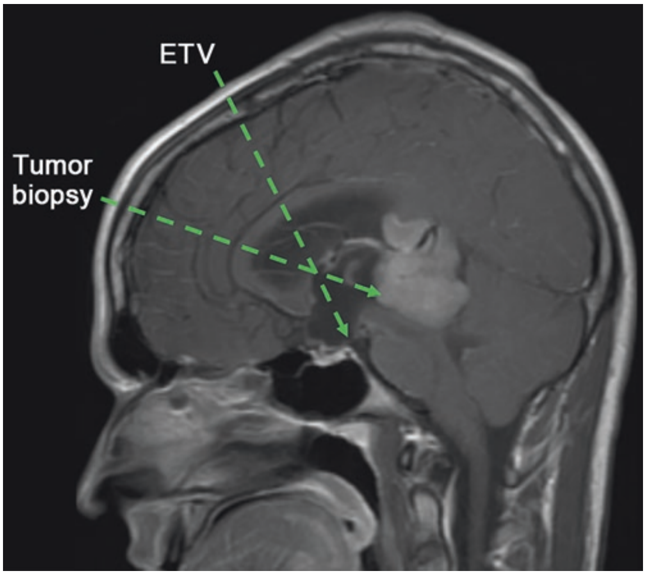

Fig. 13.3 In many patients, the ideal trajectory for access to the pineal region via the transventricular-transforaminal approach is very anterior compared to that preferred for an ETV. Often, separate burr holes are required for each approach (Used with permission from Barrow Neurological Institute, Phoenix, AZ, USA)

the third ventricle contains the cerebral aqueduct, which connects the third ventricle to the fourth ventricle and can be obstructed by pineal region lesions. When the endoscope is within the third ventricle, the mammillary bodies and infundibular recess, anterior commissure, dorsum sella, and membrane of Liliequist are identified. A region approximately two-thirds the distance between these two points is identified, and ports within the endoscope are commonly used as an ideal point of fenestration of the membrane of Liliequist. Often, the membrane of Liliequist is thin and translucent, with the basilar artery visible in the basal cisterns. At our institution, we often place the point of fenestration as far away from the basilar artery as possible to avoid iatrogenic injury to this structure, typically identifying the dorsum sella and working immediately posterior to the dorsum sella to perform the fenestration.

Endoscopic Pineal Region Biopsy

An ETV is often performed in combination with an endoscopic pineal region biopsy. Used in conjunction, these two procedures allow for the relief of hydrocephalus and provide tissue for histopathological diagnosis of pineal region tumors. However, a full resection of most lesions cannot be safely performed using this approach. In this procedure, the entry point required to access the posterior third ventricle necessitates an incision approximately 2 inches anterior to that performed for an ETV (Fig. 13.3). A more anterior entry point allows for more "in-line" access to the pineal region while minimizing traction on the fornix with a rigid endoscope. Often, two separate incisions and burr holes are used to perform a biopsy and ETV using rigid endoscopes. Alternatively, the same incision and burr hole can be used to perform both procedures when using a flexible, steerable endoscope because this tool does not require the lesion and surgical trajectory to be in-line. However, flexible endoscopes can often be difficult for inexperienced surgeons to use because the projected images can be disorienting and resolution is not as high as that of rigid endoscopes. Nevertheless, the flexible endoscope allows for greater access to more challenging lesions [16].

Posterior Endoscopic Keyhole Approaches

Several posterior approaches to the pineal region have been well described in the surgical literature [2–4], and they are used for a variety of lesions. These approaches include (1) the SCIT approach, (2) the supratentorial-transtentorial approach, and (3) the posterior interhemispheric approach. The use of any approach largely depends on the configuration of the tumor and its relationship to the deep venous structures. For lesions that push the vein of Galen posteriorly, more anterior approaches (e.g., the anterior interhemispheric approach or the posterior interhemispheric approach) are ideal. For lesions that push the vein of Galen anteriorly, the SCIT approach or the supratentorial-transtentorial approach is ideal. Each approach is associated with its own set of advantages and disadvantages.

Walter Dandy first described the posterior interhemispheric approach, which necessitates exposure of the superior sagittal sinus and interhemispheric space adjacent to the central sulcus [17]. A confluence of major interhemispheric draining veins can severely limit this approach. More posterior craniotomies adjacent to the occipital lobes, such as that used with the supratentorial-transtentorial approach, can place the primary visual cortex at risk for injury. The posterior interhemispheric approaches for pineal region tumors necessitate either fixed or intermittent retraction of the occipital lobes; thus, they are used less commonly at our institution. When posterior interhemispheric approaches are used, surgeons often rely on endoscopic assistance, with placement of endoscopes in the resection cavity after initially using microscopic visualization; [18] thus, this is not a purely endoscopic keyhole approach.

The use of endoscopy for pineal region tumors has been well described for anterior transventricular approaches, which are used mainly for tissue biopsy and hydrocephalus management [13–15]. As the use of the endoscope by skull base surgeons has increased for a range of anterior skull base pathologies, many of the endoscopic principles learned have been slowly used for posterior approaches to the pineal region. During the past decade, several different groups have described fully endoscopic keyhole SCIT approaches for pineal region pathology [19, 20]. These relatively novel approaches have steadily gained popularity, and their indications have expanded drastically from simple pineal cyst fenestration to complete resection of germinal or parenchymal tumors. The endoscopic keyhole approach allows for improved panoramic visualization of pineal region pathology and increased illumination within a narrow and deep surgical corridor, all the while decreasing approach-related morbidity with smaller incisions and craniotomies.

The SCIT route is a powerful method to access lesions of the pineal region, but the long and deep surgical corridor necessitates large exposures and significant cerebellar traction on the cerebellum when microscopic visualization is used. Microscopic surgeons often advocate exposure of the transverse sinus and/or the torcula to provide upward mobilization of the sinuses and tentorium cerebelli [21]. Because the light source is located outside of the cranial vault, increasing the upward mobilization of the tentorium allows for greater light penetration deep into the surgical cavity. However, this exposure increases the potential for iatrogenic injury to the sinuses, either during the craniotomy or with thrombosis of the vessels during prolonged exposure to the light microscope. Furthermore, microscopic surgeons often use fixed retraction of the cerebellum to increase the surgical working corridor and to decrease instrument obstruction of the surgeon's microscopic view. This increases the risk for retraction-related injury to the normal cerebellum. Lastly, the surgeon's microscopic view of the pineal region is limited to what is available within the line of sight. Manipulation of critical neurovascular structures outside the line of sight necessitates "blind" manipulation, further increasing the risk of inadvertent iatrogenic injury. Conversely, when endoscopic visualization is used, both the camera and light source are inserted into the cranial vault which decreases the need for large exposures to allow for light penetration and magnification of the surgical corridor. Furthermore, angled endoscopes and instruments permit the direct visualization of structures outside the surgical line of sight. The endoscopic SCIT approach may be safer than open microsurgical approaches because of the advantages afforded by the endoscope.

Operative Setup, Instrumentation, and Technique

When using an endoscopic keyhole SCIT approach to the pineal region, we prefer the sitting-slouch position for the patient unless the patient has a patent foramen ovale, in which case the patient should be positioned laterally (Fig. 13.4).

Fig. 13.4 The patient is placed in rigid fixation in the sitting-slouch position, with the back of the bed at the level of the patient's mid-shoulder blade or lower to keep the bed from obstructing the surgeon's hands (Used with permission from Barrow Neurological Institute, Phoenix, AZ, USA)

Several technical considerations must be taken into account when performing this procedure. The operating room setup is different than that used during microscopic procedures. The video monitor is placed directly in front of the primary surgeon who will be performing the bimodal intracranial dissection. A slave projector is placed adjacent to the primary screen to allow the assistant who is holding the endoscope to directly visualize a monitor without having to tilt his or her head; this improves surgeon comfort and decreases surgeon fatigue. We do not routinely employ irrigating sheaths, which are commonly used for endonasal procedures. The endoscope should be trialed and tested before the surgical pause to ensure its usability; replacement endoscopes and a replacement microscope should be available in the operating room should there be a technical issue with the primary equipment. The remaining surgical dissection tools are identical to those used in open microsurgical approaches, and no additional tools are necessary.

The surgical working corridor is optimized for the endoscope by expanding the infratentorial working space using various methods that obviate the need for fixed retraction. These methods include a generous dissection of arachnoid adhesions to permit CSF drainage early in the course of the procedure, the intravenous administration of mannitol, and occasionally the use of lumbar drainage. Furthermore, all patients undergoing endoscopic SCIT approaches at our institution are placed in the sitting-slouch position, unless contraindicated by a patent foramen ovale [3, 22]. This position has several advantages. First, it permits the use of gravity retraction of the cerebellar hemispheres, which further increases the size of the working corridor without the need for fixed retractors. Second, the sitting-slouch position is ergonomically advantageous for the endoscopic surgeon and the assistant, as the surgeon's hands are placed in a comfortable mid-body position for the duration of the procedure. This setup is in stark contrast to that of open microsurgical approaches for which the patient is in a sitting position; with these approaches, the surgeon's hands are elevated in midair for several hours which increases the risk for surgeon fatigue. Third, the sitting-slouch position allows for greater venous outflow and lower venous pressures, which decreases bleeding from the resection cavity during the procedure, and it allows for any blood that is spilled to freely drain away from the surgical field without obscuring the view. The sitting-slouch position also has several disadvantages of which the surgeon should be aware. First and foremost, it increases the risk of venous air embolism. Thus, all patients must undergo preoperative echocardiographic bubble studies to ensure that a patent foramen ovale is not present and to prevent the incidence of paradoxical air emboli. During the course of the procedure, all patients require precordial Doppler monitoring, as well as a central line in the right atrium to detect and treat venous air embolism. Furthermore, should there be a need for emergent conversion to open craniotomy during the course of an endoscopic approach, the sitting-slouch position can become quite uncomfortable for the microsurgeon.

At our institution, we use off-midline SCIT approaches. Midline approaches are avoided because of the larger number of bridging veins from the tentorium to the cerebellum located in this region, as well as obstruction of the pineal region by the high-riding culmen of the cerebellum in the midline [23]. All patients undergo a preoperative MRI with 1-mm cuts that are loaded into a navigation system and registered to the patient using surface contours. Stereotactic navigation techniques allow us to mark a 4-cm incision approximately 12 mm off midline, starting at the level of the transverse sinus and pointing down (Fig. 13.5) [3]. A 25-mm wide and 18-mm high keyhole craniotomy is then performed, exposing the inferior edge of the transverse sinus and dura overlying the upper cerebellum, lateral to the torcula. For lesions that are largely midline, we prefer a left-sided craniotomy when the primary surgeon is right handed to improve ergonomics [24]. Unlike in open microsurgical approaches, the relatively small craniotomy used in endoscopic approaches does not compromise the degree of visualization of the surgical corridor because the light source and camera are located within the cranial vault. However, the smaller the craniotomy, the greater the risk of instrument conflict [25, 26]. Hence, a craniotomy smaller than 20 mm wide may impede progress, and 25 mm is recommended. An inverted U-shaped incision in the dura is then performed up to the level of the transverse sinus, with or without the use of the endoscope, and the dura is retracted superiorly.

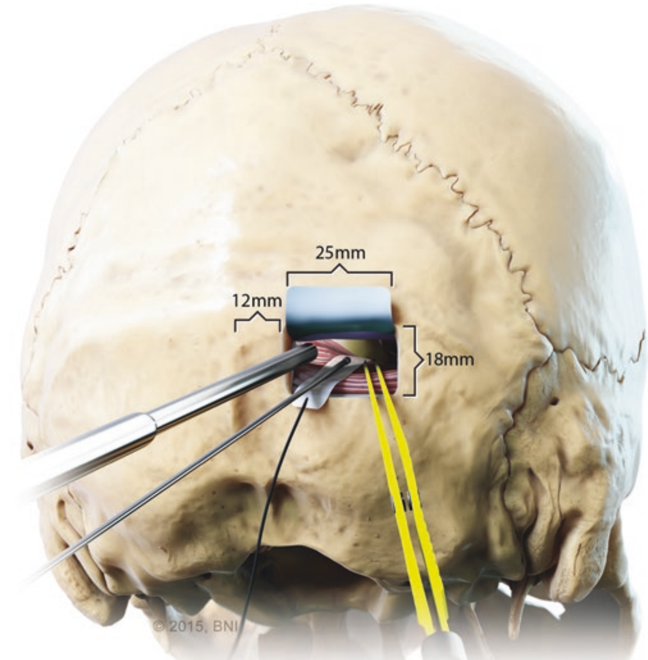

Fig. 13.5 The usual location of the paramedian opening is 12 mm from the midline at the level of the underside of the transverse sinus, and the craniotomy is 25 mm wide and 18 mm high. The endoscope is parked in one of the two upper corners of the craniotomy (Used with permission from Barrow Neurological Institute, Phoenix, AZ, USA)

The endoscope is advanced and wide; generous arachnoid dissection is performed progressively to increase the potential working space and to increase CSF drainage until the pineal region is visualized. Usually, veins can be avoided; in all cases, every attempt is made to preserve veins. When the trajectory is maintained parallel to the tentorium, the vein of Galen can be directly viewed; a slightly inferior trajectory can allow visualization of the pineal region (Fig. 13.6a–e). Larger tumors will have created their own space and the tumor will usually be obvious. If the tumor is not identifiable, it may be necessary to open the thick white arachnoid that often drapes this region. Surgeon knowledge of anatomy is key to navigating the depths of the supracerebellar corridor, especially when normal structures are distorted by tumors in this region. Adapters can be connected to the endoscope that allow for precise triangulation of the endoscopic tip using the neuronavigation platform [24]. During the intracranial segment of the procedure, general endoscopic principles are used for safe and effective surgery. These principles include constant visualization of the tip of instruments when they are introduced into the cranial vault. Such visualization helps to avoid iatrogenic injury to structures outside the line of sight. The endoscope should never be swept side to side because doing so can lead to catastrophic neurovascular injury outside the line of sight. The assistant holding the endoscope should be an active participant in the surgical procedure, frequently advancing and retracting the endoscope

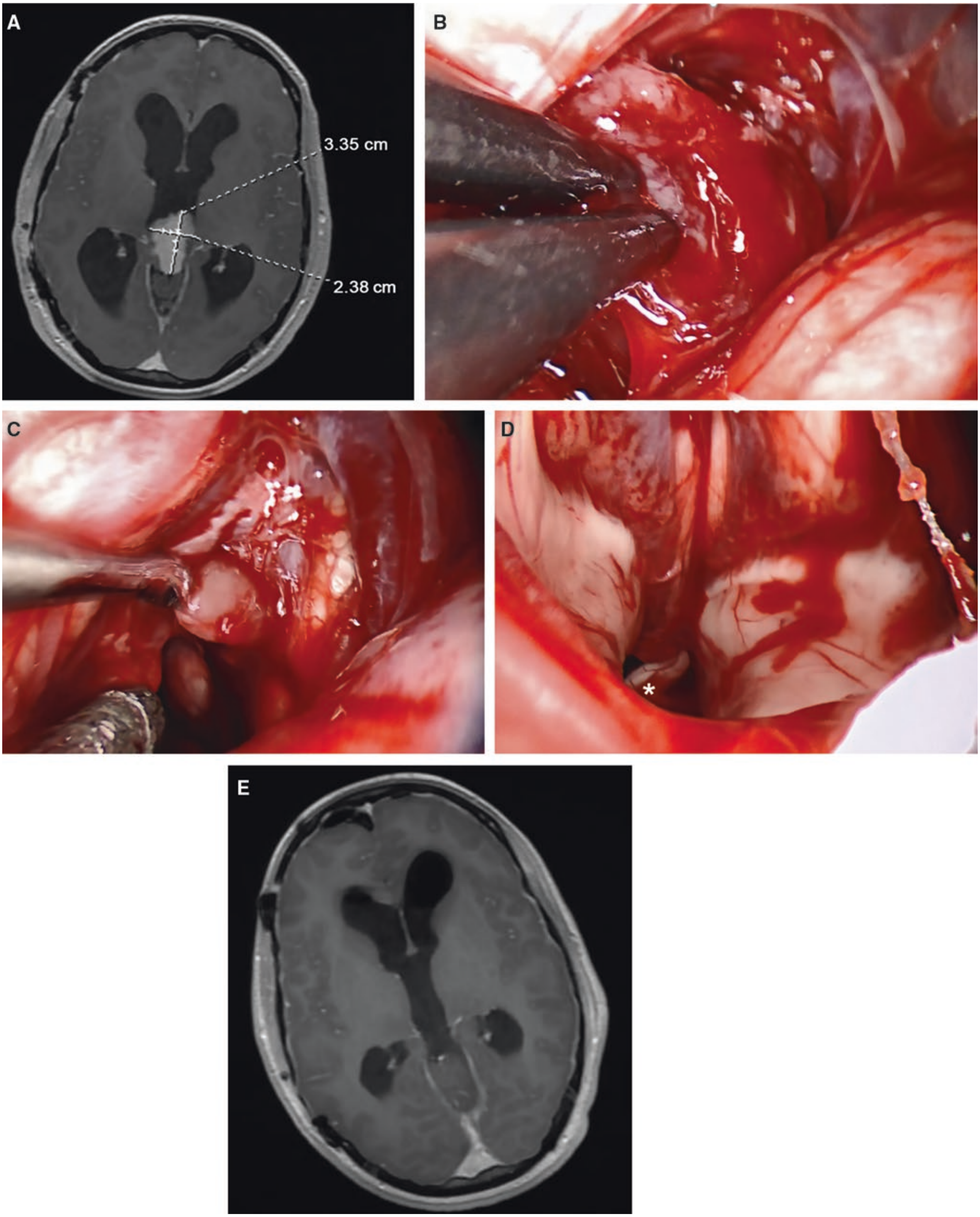

Fig. 13.6 (**a**) Preoperative axial gadolinium-enhanced magnetic resonance image (MRI) demonstrates a medium-sized enhancing pineal tumor. (**b**) Intraoperative view through a three-dimensional endoscope showing the tentorium to the upper left, the vermis to the lower right, and the instruments working on the tumor in the center. (**c**) The main bulk of the tumor has been removed, and a small nodule of residual tumor underneath the internal cerebral veins is removed with a dissector. (**d**) After tumor removal, the roof and anterior superior third ventricle are seen, along with both foramina of Monro (*asterisk*) and the choroid plexus in the roof of the third ventricle. (**e**) Postoperative axial gadolinium-enhanced MRI confirms gross-total removal of the enhancing mass (Used with permission from Barrow Neurological Institute, Phoenix, AZ, USA)

as the primary surgeon is moving instruments in and out of the intracranial vault. Excellent and precise sharp and blunt two-handed microneurosurgical technique should be used throughout. After the surgical resection of the tumor is completed, it is imperative that a watertight dural closure be performed to prevent a pseudomeningocele or CSF leaks.

13.6 Postoperative Care and Complications

Postoperatively, neurological assessment is of paramount importance. Any patient with loss of neurological function or slow emergence from sedation is taken immediately for a CT scan to assess for postoperative surgical bed hematoma or hydrocephalus. The presence of these pathologies should be treated accordingly, either with return to the operating room or placement of a bedside external ventricular drain. At our institution, negative imaging in the presence of neurological dysfunction prompts an MRI in addition to vascular imaging (CT angiography or MR angiography) to assess for a cerebrovascular accident. Patients with stable neurological function postoperatively will undergo a delayed gadolinium contrast-enhanced MRI to assess for residual disease and future treatment planning. All patients who undergo such a surgical procedure are placed in the neurological intensive care unit with continuous cardiac and neurological monitoring. We typically place patients with the head of the bed elevated for 2–3 days to reduce the probability of a pseudomeningocele. We rarely place a preoperative lumbar drain. When additional CSF diversion procedures are performed, they are typically discontinued on postoperative day 1 or 2, depending on the quality of dural closure as assessed intraoperatively. In the absence of known coagulopathy disorders, non-ambulatory patients are also prophylactically treated with sequential compression devices as well as pharmacological prophylactic-dose low-molecular-weight heparin.

13.7 Surgical Pearls

- Virtually all pineal region masses are advantageously approached from a supracerebellar-infratentorial approach.
- A sitting-slouch position uses gravity to create working space between the cerebellum and tentorium. A lumbar drain can be a useful adjunct.
- A preoperative echocardiogram with bubble study to assess for a patent foramen ovale and intraoperative Doppler studies for air embolism are essential.
- A combination of straight and angled endoscopes can be used with two-handed sharp microsurgical techniques to effect removal of a pineal region lesion.

13.8 Conclusion

Neuroendoscopic keyhole approaches to the pineal region are effective and safe methods to address a variety of pathologies. Anterior approaches are primarily used for tissue biopsy and for CSF diversion for lesions obstructing the cerebral aqueduct or third ventricle. Should the anatomical configuration of the lesion permit, posterior SCIT approaches can be used to resect pineal region tumors or cysts. These approaches provide better illumination and magnification of the pineal region than traditional approaches. Endoscopic keyhole approaches are associated with a steep learning curve and require a team-based approach as well as an intimate understanding of the important neurovascular structures of the pineal region.

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14 Transcortical Corridors

Anil Kumar Roy, Nefize Turan, and Gustavo Pradilla

14.1 Introduction

Surgical treatment of deep subcortical and intraventricular lesions presents unique technical challenges related to the depth of the targeted lesion including the ability to deliver proper illumination and magnification to the surgical field, maintenance of an unobstructed surgical corridor without excessive retraction of the surrounding brain, execution of bimanual technique and precise microneurosurgery, ergonomic positioning of the surgeon and visualization tools, and adequate tissue differentiation and sufficient collection of high-quality pathological specimens. Our ability to overcome such technical barriers illustrates multiple areas of progress and development in neurosurgery; however, in order to achieve better functional outcomes, recent efforts have focused on minimizing approach-related morbidity through precise understanding of white matter anatomy to select the least disruptive route and maximize functional preservation.

Several transcortical corridors based on anatomical white matter studies in cadaveric specimens have been described in the neurosurgical literature to access subcortical and intraventricular lesions [1, 2]. Some of these include the superior parietal lobule approach, the transtemporal approach, and the transfrontal approach [3, 4]. Although white matter corridors in these traditional approaches were considered to involve "non-eloquent" areas, increased understanding of white matter anatomy and physiology gained through the human connectome project and similar efforts with high-field functional MRI and diffusion tensor imaging (DTI) and evolving knowledge gained from intraoperative neurophysiological testing during neurosurgical procedures and advanced neuropsychological postoperative testing have together called into question the "non-eloquent" characterization of these areas and increased awareness of the importance of accurate preoperative DTI analysis and judicious respect for white matter fibers intraoperatively [5]. In the presence of sizable subcortical pathology, white matter fibers can be extensively and unpredictably deformed leading to potentially significant tract injuries and functional impairment. Accurately mapping possible subcortical trajectories that avoid damaging healthy neural tissue is imperative if functional outcomes are to be improved.

While treatment of some intraventricular lesions has been greatly improved by advances in neuro-ventriculoscopy, treatment of deep parenchymal lesions has until recently remained stagnant. In this chapter we will discuss basic principles of DTI, review important white matter fibers relevant to common transcortical surgical trajectories, discuss novel techniques for whole brain automated DTI, propose parafascicular fiber-sparing corridors for common deep-seated lesions, review available port-based approaches for deep targets, and highlight relevant technical aspects of these techniques.

14.2 Diffusion Tensor Imaging (DTI)

DTI is an MRI technique based on three-dimensional diffusion of water as a function of spatial location [6]. DTI has been extensively studied in healthy brain to elucidate brain connectivity and architecture as well as in neuropathological states. As water diffusion is very sensitive to microstructural changes in the central nervous system (CNS), several neurological diseases including ischemic stroke, neurodegenerative diseases such as multiple sclerosis and Alzheimer's, intracranial tumors, neuropsychiatric diseases such as schizophrenia, movement disorders such as Parkinson's and Huntington's, and pathological processes such as inflammation, aging, and edema have been evaluated to determine the clinical value of

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diffusion-weighted imaging (DWI) and DTI [6, 7]. In neurosurgery, DTI offers an invaluable insight by providing information about white matter tracts that are critical to preoperative planning and intraoperative guidance [8–10].

As the concept behind DTI is the thermal (Brownian) motion of water molecules, it is crucial to understand different types of diffusion. In principle, isotropic diffusion occurs equally in all directions while anisotropic diffusion is asymmetric resulting from water molecules moving more easily parallel to axonal fibers instead of perpendicular to these fibers. Therefore, anisotropy is high in white matter fibers where movement of water is restricted in one direction but low in gray matter and CSF where diffusion is in all directions and therefore more isotropic. The data generated from DTI analysis thus generate several measures such as mean diffusivity (MD), fractional anisotropy (FA), radial diffusivity (Dr), and axial diffusivity (Da). The FA value, ranging from 0 (completely isotropic) to 1 (movement completely restricted to one axis), is determined for each voxel depending on the differences of anisotropy and is then used to produce a gray scale 2D map. Color codes are then added to each voxel depending on the overall direction of diffusion [11]. Red–green–blue (RGB) color-coded schemes in which each color represents a different direction are frequently used. In this scheme, red represents the fibers crossing from left to right, green visualizes fibers traveling anteriorly–posteriorly, and blue depicts the fibers crossing inferiorly–superiorly [12]. With the information about 3D orientation of white matter bundles, white matter tracts such as the pyramidal tract, corpus callosum, and superior longitudinal fasciculus (SLF) are then reconstructed.

Some of the most common artifacts related to DWI acquisition include eddy current distortions and head motion, which can lead to inaccurate registration and errors in diffusion maps [7]. After preprocessing to correct for these artifacts, data processing and visualization for DTI are commonly performed by a neuroradiologist via a technique called "manual seeding" that involves identifying a region of interest (ROI) by hand to generate images of specific white matter tracts, which are then overlaid onto standard T1 or T2 sequences. Some of the disadvantages of this method include significant processing time, inter-rater variability of interpretation between different neuroradiologists, and limited information gained from evaluating a subset of tracts at a time, which can lead to over- and underestimation of other important structures [13–15]. To overcome some of these challenges, automated seeding and whole brain tractography (WBT) have recently been developed. These techniques allow for data extraction from all voxels and visualization of a three-dimensional map. While various automated DTI post-processing software packages are available, which can lead to variability in output across vendors and centers, tract-based analyses generally outperform ROI analyses [15, 16].

Interpretation of DTI data, however, also has certain limitations as data processing is based on the assumption that fibers within the same voxel are unidirectional, thereby not taking into account the crossing, diverging, or kissing fibers leading to incorrect estimations [7]. Another limitation of DTI analysis especially in the setting of preoperative planning of tumor resection includes perilesional edema. Edema generally leads to reduced FA as the fiber tracts per pixel are reduced. Tumors can cause both destruction and infiltration of white matter tracts with neoplastic cells and induce perilesional edema. In such cases, it can be difficult to establish if the reduced FA is due to edema or to a reduced number of tracts from tumor infiltration [12]. Moreover, highly variable inward and outward intraoperative shifts ranging from 8 to 15 mm have been documented during neurosurgical procedures using intraoperative DTI suggesting that preoperative DTI may not always be reliable during surgery [17, 18]. Therefore, intraoperative DTI is a valuable tool for preoperative assessment, planning, and intraoperative neuronavigation to minimize white matter injury provided the surgeon understands its limitations as described above.

14.3 White Matter Tract Overview

A brief overview of white matter anatomy is required before we can discuss appropriate pathology that can be addressed via a transcortical parafascicular approach. White matter tracts in the cerebrum can be divided into three broad categories: projection fibers, commissural fibers, and association fibers. Projection fibers extend vertically and transmit information from the cerebrum to the rest of the body. Major projection fibers include the internal capsule and optic radiations. Commissural fibers can be visualized as horizontal fibers that carry information across hemispheres with the corpus callosum being the largest of them. Other commissural fibers include the anterior and posterior commissure. Association fibers connect different regions within the same hemisphere and can include longer fibers providing lobar connections and shorter U-fibers providing gyral connections. Major association fibers include the arcuate fasciculus (AF), superior longitudinal fasciculus (SLF), inferior longitudinal fasciculus (ILF), uncinate fasciculus (UF), inferior fronto-occipital fasciculus (IFO), and cingulate fasciculus (CF).

The internal capsule fans out along the lateral aspect of the lateral ventricle with the corticobulbar and corticospinal tracts running along the mid part of the body of the lateral ventricle. The optic radiations can be visualized as running along the roof and lateral edge of the temporal horn. Only the anterior component of the optic radiations has an anterolateral course referred to as Meyer's loop before coursing posteriorly toward the calcarine sulcus.

Although we commonly think of the internal capsule and optic radiations when visualizing white matter anatomy, it is critical to understand their displacement under pathological conditions and the position of important association fibers involved. Deficits that arise from association fiber injury may not be readily apparent on a routine bedside neurological exam but can have significant long-term neuropsychological sequelae. The SLF running just deep to the U-fibers connects portions of the frontal lobe with occipital and temporal areas. Ideomotor apraxia and spatial neglect have been associated with SLF injury [19]. The UF is a hook-shaped bundle that links the ventral portion of the temporal lobe with the inferior frontal gyrus and the lower surfaces of the frontal lobe. Disruption of the UF can cause language deficits, affect, and behavioral changes [20]. The ILF connects the temporal and occipital lobes and runs at the level of the optic radiations from temporal to occipital areas. Deficits associated with its injury include object visual agnosia and prosopagnosia [21, 22]. The CF runs from the cingulate gyrus to the entorhinal cortex and functions in motivational and emotional aspects of behavior and working memory. The IFO as the name suggests is an association fiber from the frontal to the occipital lobe. It is part of the ventral semantic pathway, and its dysfunction leads to conductive aphasia [20, 22]. The AF is a white matter tract between the inferior frontal regions and posterior temporal regions. It plays a critical role in language, and damage to the AF leads to significant language difficulties including problems with repetition [20]. Although language function has been primarily associated with the dominant AF, right hemisphere remodeling of the AF has also been seen in patients undergoing speech therapy with left hemisphere lesions [23].

14.4 Visualization of Deep Surgical Targets

Traditional subcortical surgery has been carried out under microscopic visualization. The operating microscope is designed to deliver both light and magnification to a specific focal point (FP) within the surgical field, adjustable between 200 and 400 mm. One limitation of surgical microscopes is the inability to simultaneously and uniformly deliver light as well as focus throughout the entire working field, requiring moving and refocusing the microscope every time the tissue is beyond the original FP. This discrepancy in light and magnification between the FP and the surrounding fields can result in “pseudo-differentiation,” which can result in iatrogenic injury to normal tissue. Furthermore, the convergent nature in which the light is delivered requires a wide proximal corridor in order to funnel light onto a distal point. Expanding the proximal corridor to accommodate the delivery of better illumination and magnification is not always possible, particularly when the surrounding tissue is eloquent. When utilizing a surgical microscope for deep subcortical or intraventricular lesions, these limitations become more apparent and can affect the safety and efficacy of the procedure.

Improvements in surgical endoscopy have expanded the role of minimally invasive surgery (MIS) in multiple fields. Although several studies have evaluated the use of endoscopy for subcortical parenchymal lesions, several limitations exist. Current endoscopes for cranial applications were designed to perform inside the ventricular system within a liquid medium (CSF). Parenchymal lesions require the endoscope to perform under an air medium for which its rod lens design has not been designed to perform. This results in decreased light and magnification making visualization and dissection challenging. MIS ports for subcortical or intraventricular lesions have limited radial working space. Placement of an endoscope inside a MIS port significantly compromises the available working space and in most cases prevents bimanual technique. Instrumentation in ventriculoscopes is introduced and manipulated through long working channels, which offer limited maneuverability and preclude the concomitant use of bipolar cautery forceps and suction for adequate hemostasis.

A proposed solution to these challenges involves the use of high-definition exoscopic systems. These systems stay outside the working corridor and are able to deliver uniform light and magnification to the surgical field, thus creating a uniform focal field or cubic volume of view (VOV). This VOV can then be viewed on a high-definition (HD) screen. With this system, the size of the HD monitor determines the magnification, rather than the components of the microscope, and thus has no impact on the width of the surgical corridor. In addition, the “flashlight effect” of the telescope, in contrast to the “cone effect” of the microscope, enables the proximal end of the corridor (visualization end) to remain in proportion to the distal end of the corridor (working end), a particularly appealing aspect for the neurosurgeon. An additional benefit of exoscopes when used for minimally invasive neurosurgical procedures is its relatively long working distance. Finally, the issue of lens fogging and contamination with blood is not encountered with the exoscope. The focal length of the exoscope is 15–31 mm (with camera zoom and focus), and the working distance (the range of distances from the object) ranges from 20 to 65 cm.

The exoscope can be mounted on manual, pneumatic, or robotic scope holders. Pneumatic holders use a gas-powered zero-gravity mechanism with a push button for mobilization (i.e., Point Setter®, Mitaka Kohki Co., Tokyo, Japan). Robotic holders can stereotactically position themselves based on the position of the access port (Fig. 14.1). Enhanced visualization tools (i.e., digital imaging filters such as KARL STORZ [Ettlingen, Germany] SPIES Clara

Fig. 14.1 Robotic scope holder with automatic stereotactic positioning based on the position of the access port (Courtesy of Synaptive Medical, Toronto, ON, Canada)

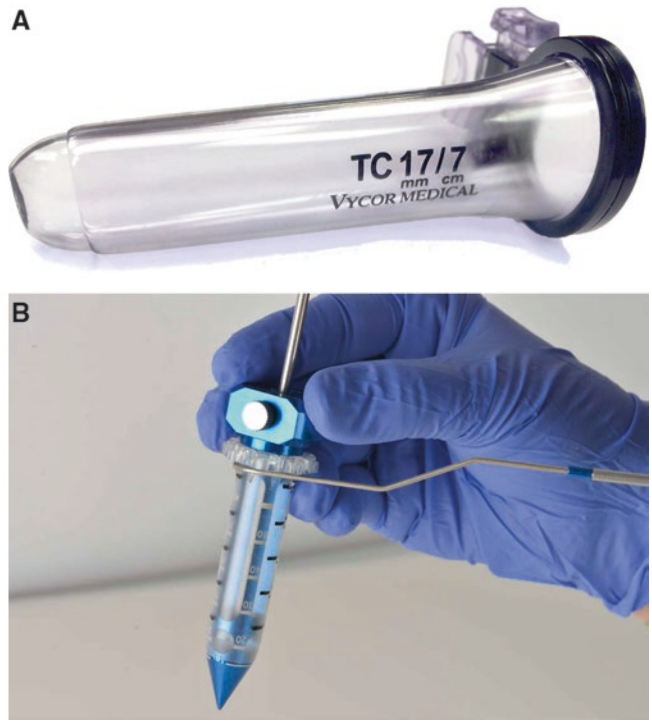

Fig. 14.2 (**a**) Vycor retractor system: ViewSite™ Brain Access System (Courtesy of Vycor Medical, Boca Raton, FL, USA). (**b**) BrainPath® retractor system (Courtesy of NICO Corporation, Indianapolis, IN, USA)

and SPIES Chroma) that can prevent excessive light absorption from hemorrhagic tissue and improve accuracy are recommended.

Currently available exoscopes include the Vitom® HD (Optronics, Goleta, CA, USA), Visionsense (Visionsense, Philadelphia, PA, USA), and BrightMatter™ Vision (Synaptive Medical, Toronto, Canada). The first iteration of exoscopes did not provide stereoscopic visualization, and this was definitely a drawback; however, all currently available systems have a 3D version available or in development to circumvent this issue. Most endonasal endoscopy is performed using 2D visualization in a videoscopic interface, and surgeons trained in these techniques have a shorter learning curve with exoscopic systems.

14.5 Port-Based Approaches

Subcortical lesions are especially suited toward port-based access [24]. To minimize retraction force on the surrounding white matter and prevent "tissue creep" in the surgical field, seminal work by Kelly and colleagues in the 1980s proposed the use of tubular retractors [25]. These retractors can evenly distribute force along their circumference, minimizing trauma to the surrounding tissues. The first generation of commercially available retractors for deep subcortical access was the Vycor retractor (Vycor Medical Inc., Boca Raton, FL, USA) which provided a much needed solution, and its use has been reported in several studies for treatment of multiple conditions [26]. This system was introduced for microsurgical resection with a surgical microscope, and challenges include the difficulty to stereotactically navigate the port insertion and the need for large cortical or sulcal access for insertion. Once introduced the working space of the port can also be a limiting factor. The UPMC Neuroendoport® (UPMC, Pittsburgh, PA, USA) was developed to add stereotactic capabilities and provides an alternative system; the reported experience in the literature, however, is limited to a few studies [27].

Our institutional experience has focused primarily on the BrainPath® system (NICO Corporation, Indianapolis, IN, USA) (Fig. 14.2a, b). The BrainPath system is comprised of a hollow obturator that accommodates a stereotactic pointer for neuronavigation and a clear cylindrical sheath that remains in place after the target is reached. The obturator has a blunt tip with a 2 mm diameter tip that dilates to a 13.5 mm diameter over a 15 mm length. The system was designed for transulcal access to minimize injury to cortical structures and comes in different lengths. In addition to stereotactic deployment, the sheath is secured to a retractor system with a retaining hook, and the surface is frosted to minimize reflection back to the operator. In conjunction with this system, we have utilized an automated whole brain tractography system (Synaptive Medical, Toronto, Canada) for image interpretation and trajectory planning that simulates the surgical corridor and displays fiber tracts at risk (Fig. 14.3a, b). During resection we utilize the same system for intraoperative navigation coupled with a robotic-mounted exoscope that positions the optical system automatically to maintain coaxial alignment. The working diameter in the port is 13.5 mm, and by positioning the exoscope outside of the field, the surgeon is afforded bimanual technique with conventional instrumentation (suction, bipolar cautery, etc.)

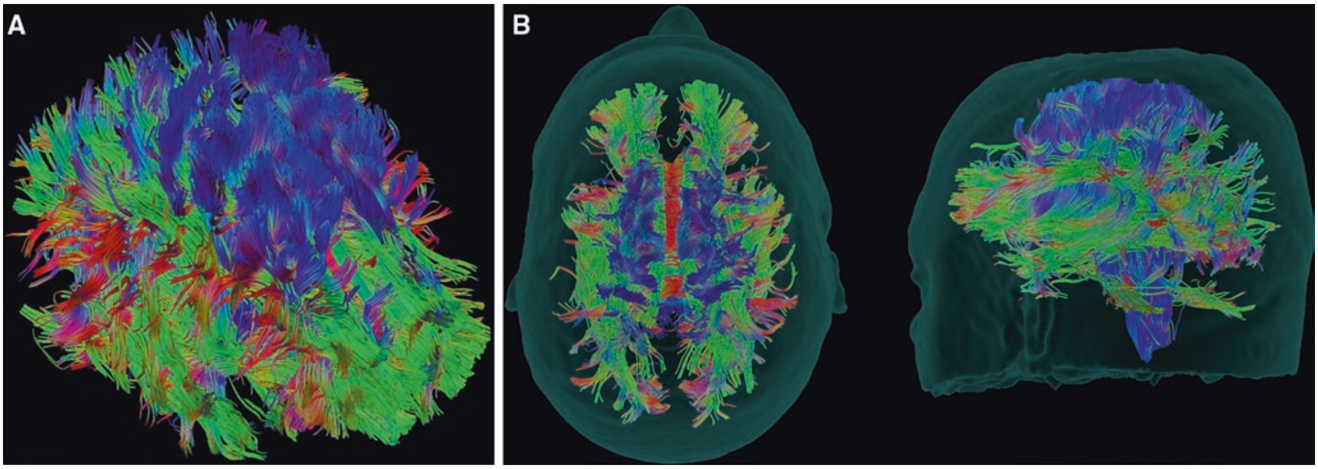

Fig. 14.3 (**a**, **b**) Whole brain tractography generated using automated seeding allows for extremely efficient image generation with consistent quality without interpretation bias

and several surgical adjuncts such as an automated resection device (NICO Myriad, NICO, Indianapolis, IN, USA), port-specific ultrasound probe, and surgical instruments that maximize the safety and efficacy of the operator.

14.6 Patient Selection

Intracerebral Hemorrhage

Intracerebral hemorrhage (ICH) accounts for 10–15% of all strokes with early mortality rates ranging from 35% to 52%, and among survivors only 10–25% return to functional independence [28–32]. This is associated with extremely high productivity losses [33]. Randomized clinical trials have currently not established a role for supratentorial ICH evacuation [34]. A systematic meta-analysis comparing traditional invasive craniotomy for ICH evacuation to conservative treatment with medical care alone found an odds ratio of 0.74 in favor of ICH patients treated with standard craniotomy [35]. The review included 15 randomized controlled trials. Evidence from individual studies such as STICH I and II have not been supportive of surgical intervention [36, 37]. A thorough discussion of these studies is beyond the scope of the current chapter, but it should be noted that subgroup analyses with STITCH I showed that deep ICH had a worse prognosis in both groups and early conventional invasive craniotomy (within 8–24 h) did not result in better outcomes.

Minimally invasive approaches to minimize iatrogenic injury in the evacuation of deep ICHs have been hypothesized to potentially swing the balance in favor of surgical evacuation. The first randomized controlled trial of endoscopic evacuation of ICH was published by Auer et al. in 1989 [38]. The study randomized 100 patients with altered mental status and spontaneous supratentorial hemorrhage >10 mL to endoscopic evacuation within 48 h versus medical management. Endoscopic evacuation resulted in significantly decreased mortality at 6 months compared to medical management but only in patients with favorable preoperative mental status and superficial hematomas. Subsequent trials such as MISTIE II also advocated a minimally invasive placement of the catheter into the clot and delivery of thrombolytics and showed a statistically significant improvement in modified Rankin score (mRS) at 6 months (mRS 0–3, 35% in the surgical groups vs 24% in the medical group) [39].

We have been employing a port-based minimally invasive parafascicular approach to deep ICHs in a selected group of patients with volumes >30 cc with a preadmission modified Rankin score of 0–1 and admission GCS $\geq$ 5. The surgical technique is described in detail below. Our initial experience with this approach for ICH is described in a multicenter study by Labib et al. [24], and these results were replicated by a single center series from the Cleveland Clinic [40]. The Labib et al. study included 39 patients from 11 centers and achieved statistically significant improvements in postoperative GCS with a clot evacuation of ≥90% in 72% of patients. The Cleveland Clinic series included 18 patients with a mean clot evacuation of 95.7% and a median GCS improvement from 10 preoperatively to 14 postoperatively. From these experiences the multicenter randomized controlled trial ENRICH (Early MiNimally-invasive Removal of IntraCerebral Hemorrhage) has been designed. For additional details on selection criteria, please visit www.enrichtrial.com.

Neoplasms

Deep-seated neoplasms are particularly suited for minimally invasive transcortical approaches as they allow biopsy and resection in the same setting, and several centers have reported promising results with port-based access to these lesions [41–44]. In general deep subcortical lesions that are ≤5 cm with moderate to low vascularity are well suited for these techniques (Fig. 14.4a–c). Determining tumor consistency and density utilizing DWI and other MRI sequences can help select lesions more amenable for this approach. When utilizing the BrainPath, port cannulation can be performed through the lesion in less dense tumors allowing for an inside-out and deep to superficial resection or just to the surface of the lesion if MRI suggests a more firm or fibrous consistency. By considering white matter tracts and selecting a trajectory along the long axis of the white matter fibers at risk, as opposed to the shortest distance from the cortical surface, some of the approach-related morbidity can be theoretically minimized. We recommend that practitioners attempt port-based removal of subcortical lesions only after an initial experience with ICH evacuation to develop sufficient experience with the videoscopic 2D workflow and with OR setup and efficiency in general. All the traditional microsurgical techniques still apply to port-based removal of lesions although we have found that non-ablative resection techniques as discussed later work best in these narrow corridors and keep surrounding eloquent tissue out of harm's way.

14.7 Surgical Considerations and Technique

Surgical Corridors

Following DTI reconstruction of white matter fibers, the surgeon must carefully study the displacement of the previously mentioned association, projection, and commissural fibers. Any potential approach needs to minimize transgression of fibers and ideally should run parallel to the major association fibers.

There are three basic corridors depending on the location of the target: anterior, posterior, and lateral (Fig. 14.5a–c). A standardized approach can be followed for ICH patients as multiple DTI reconstructions have revealed a predictable pattern of white matter displacement whereas for neoplasms the approach often needs significant customization. Although approaches to ICH will routinely engage the long axis of the clot, it is critical that the surgeon reviews the tractography data to ensure an atraumatic corridor.

The anterior corridor is the mainstay for all basal ganglia hemorrhages. These typically displace the CF medially and the SLF laterally. If a traditional transfrontal corridor is used, the SLF may be transected. A safe access anterior and inferior frontal corridor that engages the space between SLF and cingulate along the long axis of both fascicles is our most commonly used approach for anterior basal ganglia hemorrhages. For these patients, we do not routinely wait for DTI reconstruction as time to evacuation could play a role in patient outcomes. The posterior corridor engages the parieto-occipital sulcus and provides safe access to subcortical pathology in the peri-atrial region without traversing the optic radiations, the arcuate fasciculus, or the vertical rami of the SLF. It is critical that this corridor be constructed posterior to the primary sensory fibers after DTI reconstruction is completed. The lateral approach in our view is not ideal as it often leads to association fiber violation. Dominant trans-temporal approaches can cause significant violation of language regions, and a longer posterior approach in such cases may actually decrease transgression of white matter fibers. The lateral approach tends to work best with lobar hemorrhages that tend to be superficial, and cortical preservation requires atraumatic access to minimize functional injuries.

Operative Setup

The surgical setup for transcortical access requires the following basic workflow and equipment discussed in further detail below (Fig. 14.6a, b):

1. Preoperative planning, tractography, target selection, and surgical corridor selection
2. Operative equipment:
   (a) Navigation platform.
   (b) Port-based entry system.
   (c) Visualization (microscope, endoscope, or exoscope).
   (d) Large high-definition monitor.
   (e) Standard brain tumor craniotomy tray.
   (f) Resection platform can include any ultrasonic aspirator or non-ablative resection devices (e.g., NICO Myriad, NICO, Indianapolis, IN, USA).
   (g) Long bayonetted bipolars.

Preoperative Imaging

The basic premise of transcortical corridors is atraumatic access to the target with good visualization for a safe and efficient resection. The first step in this process includes obtaining a preoperative volumetric MRI that is merged with preoperative whole brain tractography. This is followed by clearly defining the target and selecting the least traumatic surgical corridor based on the principles discussed earlier. Although we use the Synaptive platform for our initial reconstruction, any navigation platform can be used in this workflow.

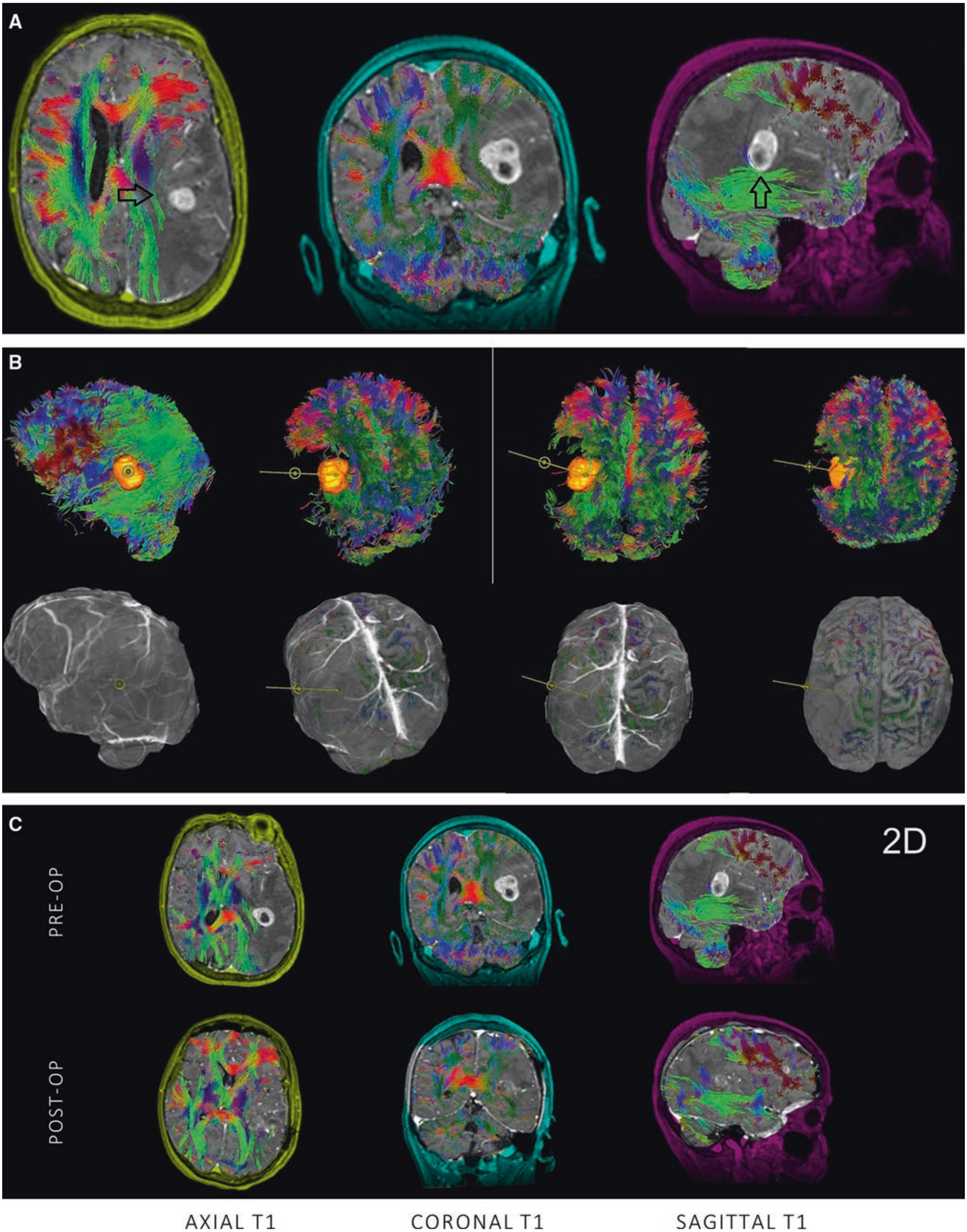

Fig. 14.4 (**a**) Preoperative images reveal a left temporal mass with contrast enhancement with overlayed tractography. Images reveal medial displacement of the posterior limb of the internal capsule (*right-pointing arrow*) and inferior displacement of the optic radiations (*upward-pointing arrow*). (**b**) Planning phase allows construction of a surgical trajectory that clearly avoids critical white matter fibers. (**c**) Postoperative images show a gross total resection of the mass with preservation of all key white matter tracts

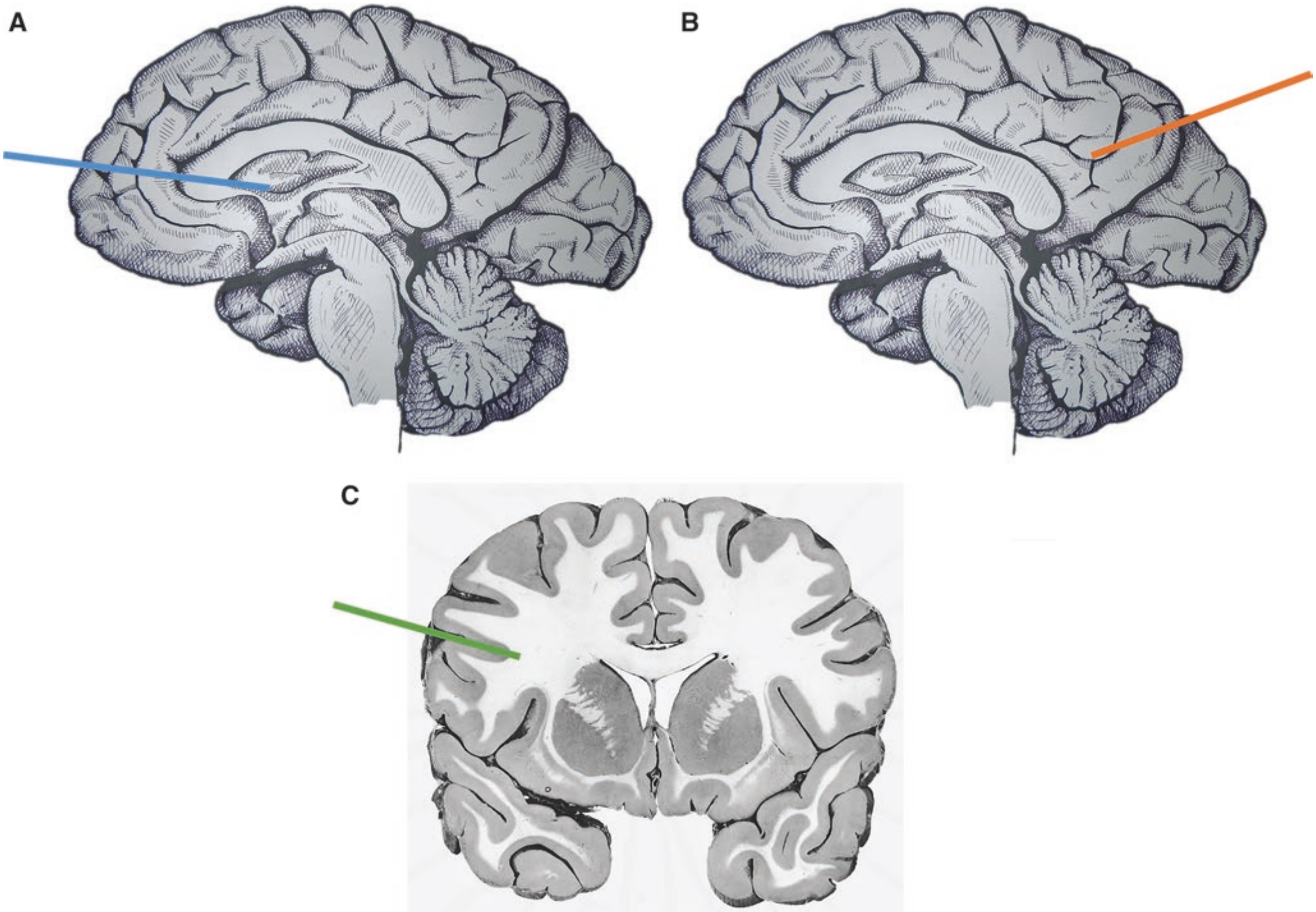

Fig. 14.5 (**a**–**c**) Anterior, posterior, and lateral corridors of entry

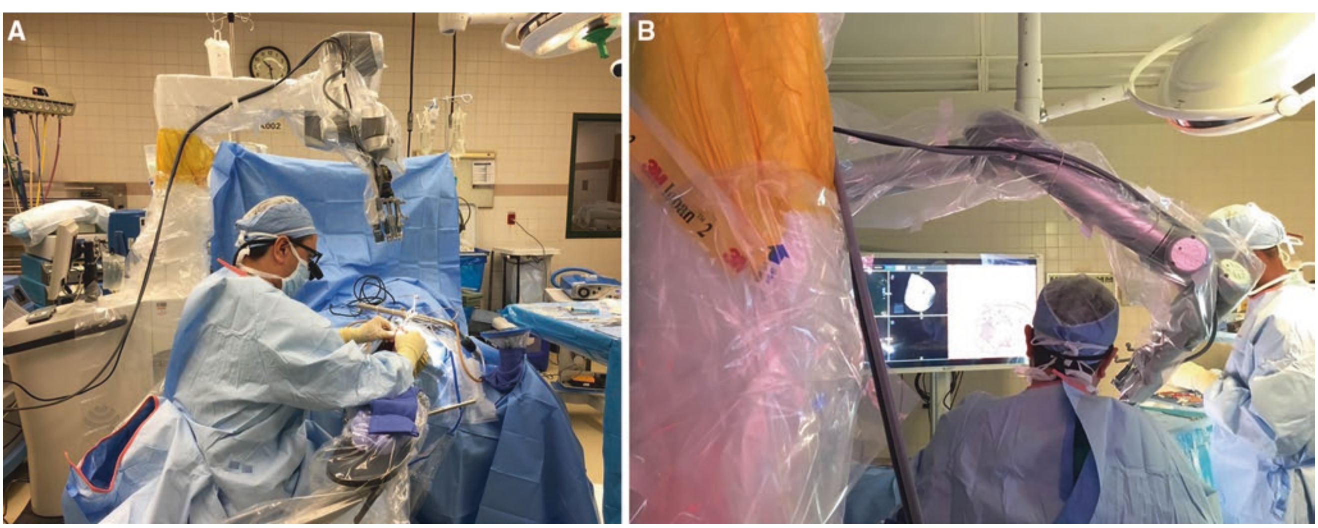

Fig. 14.6 (**a**, **b**) Operative setup should allow adequate mobilization of the visualization port while still maintaining bimanual surgical technique and a direct line of sight to the monitor (**a** and **b**: Courtesy of Synaptive Medical, Toronto, ON, Canada)

Patient Positioning, Craniotomy, and Port Positioning

The patient is appropriately positioned with skull clamps dictated by the site of entry. For most anterior corridors, keeping the head in a supine and slightly extended position works well. For lateral and posterior corridors, either a shoulder bump or a three-quarter position works well. Forehead crease and eyebrow incisions confer quick access for anterior approaches and commonly coincide with the selected entry point in patients with receding hairlines or baldness. For lobar hemorrhages, the involved lobe dictates the position. When possible, the long axis of the existing white matter fibers in the anatomical region will be selected as the loading vector for the planned trajectory. The craniotomy can be planned over the entry site as previously planned in the preoperative phase. The diameter of the port we use is 13.5 mm and comes in lengths of 50, 60, and 75 mm. The appropriate port to use can be determined in the planning phase.

A small incision (usually 4–5 cm) is made, and a craniotomy that affords adequate range of motion of the port along the flight plan trajectory is made. A cruciate dural opening is recommended to facilitate containment of the subjacent edematous parenchyma. The dural opening should be equal to or slightly less than the diameter of the chosen port-based entry system to promote a tight seal around the port sheath (Fig. 14.7). A small opening (2 mm approximately) in the arachnoid is made under exoscopic or loupe visualization with every effort made to preserve surface veins. If a large vascular structure needs displacement during cannulation, the arachnoid along this vessel should be opened sufficiently to facilitate mobilization of the vessel without unnecessary tension.

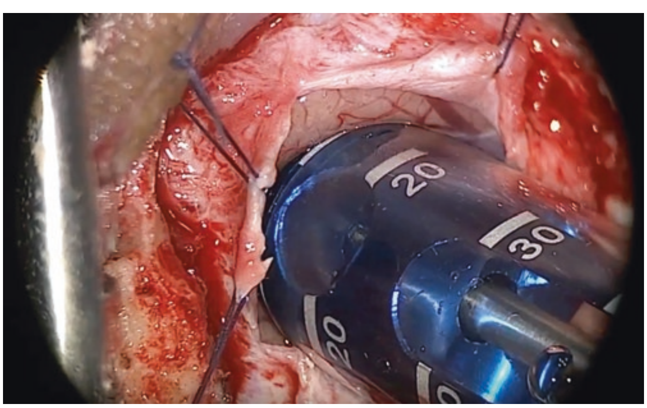

Fig. 14.7 Cannulation of port after opening the arachnoid. The dural opening should be sized to allow adequate mobility of the port but also keep a seal around it (Courtesy of NICO Corporation, Indianapolis, IN, USA)

Anesthetic Plans

Elevated intracranial pressure is advantageous during cannulation to provide adequate resistance to the port and deliver the hematoma or the lesion into the lumen of the port. Mannitol, corticosteroids, hypertonic saline solutions, and other maneuvers aimed to decrease ICP are therefore discouraged during cannulation but may be instituted after clot evacuation has been achieved as deemed necessary by the neurosurgeon. Blood pressure control with a systolic blood pressure goal <160 mm Hg or <180 mm Hh with longstanding hypertension is recommended. In patients with favorable preinduction parameters, immediate postoperative extubation is desirable.

Resection Techniques, Intraoperative Adjuncts, and Hemostasis

Although resection can be attained using either manual or automated surgical instruments, using energy-based ablative instrumentation very close to healthy neural tissue may result in unintended damage, as it is difficult to predict the way in which energy dissipates at the point of application within the lesion. As such we have moved away from using ultrasonic aspirators within the narrow confines of the surgical corridor. Toward this end, we use an automated, non-ablative resection device (NICO Myriad, NICO Corp, Indianapolis, IN, USA). It is a motor-driven suction-aspiration device and well suited when coupled with a tubular retractor. Long bayonetted bipolar cautery forceps and standard surgical hemostatic agents such as Surgicel® (Ethicon, Somerville, NJ, USA), fibrillar, Gelfoam® (Pfizer, New York, NY, USA), FloSeal® (Baxter Healthcare, Deerfield, IL, USA), or SurgiFlo® (Ethicon, Somerville, NJ, USA) are recommended. Proper long tip applicators are useful to maintain visibility and increase accuracy. Once the deepest component of the clot/lesion is evacuated, decannulation proceeds at 1 cm intervals with meticulous hemostasis until the sheath is removed.

14.8 Postoperative Care

All patients that undergo ICH evacuation obtain a postoperative CT of the head, and patients that undergo tumor resection obtain an MRI of the brain with and without contrast. We aim for a systolic blood pressure <180 mm Hg for all patients that undergo MIS surgery while keeping mean arterial pressure ≥80 mm Hg or ≥60 mm Hg when ICP monitoring is available. We routinely add ICP monitoring with CSF diversion in the presence of significant intraventricular hemorrhage or if the patient's mental status remains poor in the postoperative period (GCS ≤ 8).

Deep venous thrombosis/pulmonary embolism prophylaxis is initiated in all patients at 48 h after surgery unless interim imaging reveals a rebleed. Seizure prophylaxis with levetiracetam is also initiated in all patients and continued for 7 days postoperatively unless the patient presented with seizures at which time we obtain consultation with neurology for long-term therapy.

14.9 Surgical Pearls

- All minimally invasive intracranial approaches require a careful evaluation of preoperative whole brain tractography. With time, preoperative tractography is not always needed for ICH evacuations to minimize time to surgery.
- Practitioners should initially start their port-based approaches with ICH evacuations before moving to resection of neoplasms.
- It is critical to preserve surface veins and achieve an atraumatic transulcal opening for best patient outcomes.

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15 Intraventricular Approaches

Rafael Uribe-Cardenas and Mark M. Souweidane

15.1 Introduction

When neurosurgery was still a nascent field, ventricular surgery involved large exposures that were prone to high rates of morbidity and mortality. The refinement of technology and microsurgical techniques allowed open approaches to make their way into mainstream neurosurgery [1–3]. However, as the field progressed, the desire to perform procedures that were less invasive, minimized tissue disruption and maximized successful outcomes increased. Neurosurgeons were slow to pick up the new and evolving field of endoscopic surgery that other fields were already pioneering. Victor Darwin Lespinasse (1878–1946), a urologist from Chicago, is credited with performing the first successful endoscopic ventricular surgery in 1910 to coagulate the choroid plexus of two infants with hydrocephalus with the use of a cystoscope. The first one succumbed, but the second survived the procedure [4–6]. However, performing surgery in the deepest recesses of the brain was still marred by significant morbidity and mortality. One of the greatest limitations for success was the lack of adequate surgical technology. With the development of modern lens and lighting methods came the idea that complex surgeries could be achieved through small working channels. Newer technical developments have paved the way for modern intraventricular surgery. It is now possible to achieve most of what was envisioned by the early pioneers of these techniques.

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15.2 Rationale

Surgery in or around the ventricular system can be extremely challenging and complex. Injuries to critical brainstem, diencephalic, and subcortical structures that lie in direct continuity with the ependymal surface of the ventricles can have devastating clinical consequences. It is therefore of paramount importance for the neurosurgeon to understand the technical nuances of these approaches to obtain successful surgical outcomes. Endoscopic intraventricular surgery can serve as a mean to obtain diagnostic tissue for pathological analysis, for resection of purely intraventricular lesions, for CSF diversion in various forms of hydrocephalus, or for a combination of these pathologies. Although modern endoscopic techniques are very versatile, it is crucial to know their limitations and potential pitfalls.

15.3 Patient Selection

Virtually any patient with intraventricular pathology is a potential candidate for an endoscopic approach. The endoscope can be used to treat a number of conditions from hydrocephalus in neonates with intraventricular hemorrhage to older adults with mass lesions. The selection of a patient for an endoscopic approach will depend ultimately on a combination of factors including the type of pathology that is being treated, the level of comfort of the surgeon with endoscopic equipment, and the overall goal of the surgical procedure itself.

15.4 Indications/Contraindications

Intraventricular surgery can address lesions that fall into three main categories (Table 15.1): space-occupying lesions [7–14], hydrocephalus [15–18], and congenital malformations [19, 20]. These conditions might coexist, and a single procedure might address more than one of them (e.g., ETV can be performed during the same procedure in which a biopsy of a lesion is planned) [21].

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Table 15.1 Intraventricular lesions than can be addressed through an endoscopic approach

Tumors
Tectal/midbrain gliomas
Thalamic gliomas
Pineal region tumors
Choroid plexus tumors
Colloid cysts
Optic pathway gliomas
Craniopharyngioma
SEGA
Langerhans cell histiocytosis (LCH)
Hydrocephalus
Aqueductal stenosis
Loculated hydrocephalus (posthemorrhagic or postinfectious)
Fourth ventricular outlet syndrome
Foraminal atresia
Congenital lesions
Arachnoid cysts
Hypothalamic hamartoma

Endoscopic approaches might also precede the definitive treatment of certain lesions as is the case of choroid plexus tumors. If there is a suspicion that the lesion might correspond to a higher-grade tumor, an endoscopic biopsy might help elucidate the diagnosis and plan for the need of neoadjuvant chemotherapy prior to an attempted open, gross-total resection. Endoscopy can also be an adjunct to other forms of treatment. In the case of complex hydrocephalus, some patients with implanted shunts might not be able to drain both sides of the ventricular system through one catheter. Endoscopy can thus be used to communicate the different ventricular cavities as well as to aid with the adequate placement of the proximal shunt catheter.

Although there are no absolute contraindications to neuroendoscopic procedures, care must be taken to choose the correct patients who might benefit from them. Small ventricles are not necessarily a contraindication; they can be technically challenging to navigate, and therefore the room for error is reduced. The slow instillation of small aliquots of sterile saline solution at the beginning of the case can help improve ventricular size and provide more room for the intended procedure. Patients with severely distorted ventricular anatomy such as in postinfectious or posthemorrhagic ventriculitis can be difficult to navigate, and there is always a risk of injury to critical neurovascular structures that may be far displaced from their typical location.

15.5 Surgical Anatomy

The ventricular system is a complex formation deep within the cerebral white matter. For the purposes of this chapter, we will focus our attention on the anatomy of the lateral and third ventricles which are the sites where most endoscopic procedures are performed.

The lateral ventricles are composed of the frontal horn, body, atrium, occipital, and temporal horns [22]. They communicate with the third ventricle through the foramen of Monro. The lateral ventricles wrap around the thalamus and the caudate nucleus. The frontal horn is bounded superiorly and anteriorly by the fibers of the genu of the corpus callosum. The posterior aspect of the frontal horn is limited by the column of the fornix as it descends in front of the foramen of Monro. The floor of the frontal horn is made by fibers of the rostrum of the corpus callosum. The medial wall is bounded by the septum pellucidum, and the lateral wall is made by the head of the caudate nucleus. The body of the lateral ventricle is limited superiorly by fibers of the body of the corpus callosum. It extends posteriorly to the point where the septum pellucidum disappears and the corpus callosum contacts the fornix. The body of the caudate nucleus forms the lateral wall, and the floor is made up by the thalamus. The thalamus and the caudate nucleus are separated by the caudothalamic groove, a space where the group of fibers that make up the stria terminalis traverse. These fibers connect the hypothalamus with the amygdala. Along the caudothalamic groove lies the thalamostriate vein, an important anatomical landmark that must be identified once inside the ventricle (Fig. 15.1). The atrium and occipital horn take a pyramidal shape. The anterior wall of the atrium is formed by the posterior aspect of the thalamus, the pulvinar. The anterior wall is bordered medially by the crus of the fornix and laterally by the thalamus. The atrium opens up superiorly into the body of the lateral ventricle over the thalamus and inferiorly under the thalamus into the temporal horn. The medial wall of the atrium is formed by two prominences: the superior one is made by fibers from the splenium of the corpus callosum and the inferior (called *calcar avis*) which represents the invagination that corresponds externally with the calcarine sulcus. The roof of the atrium is made of fibers of the body, splenium, and tapetum. The lateral wall of the atrium is formed by the caudate nucleus and fibers of the tapetum. The floor is created by the collateral trigone, an elevation formed by the posterior aspect of the collateral sulcus. The occipital horn is the posterior extension of the atrium and shares the same anatomical boundaries. The temporal horn extends inferiorly beneath the pulvinar into the temporal lobe. The anterior wall of the temporal horn is bounded by the amygdala. The floor is delimited medially by the hippocampus and laterally by the collateral prominence which overlies the collateral sulcus. The roof of the temporal horn is made up medially by the thalamus and the tail of the caudate nucleus. The lateral part of the roof is made up by fibers of the tapetum that extend laterally and anteriorly. The medial wall of the temporal horn is small and is limited by the choroidal fissure, a cleft that separates the thalamus from the fornix. The choroidal fissure extends in a C-shaped configuration from the

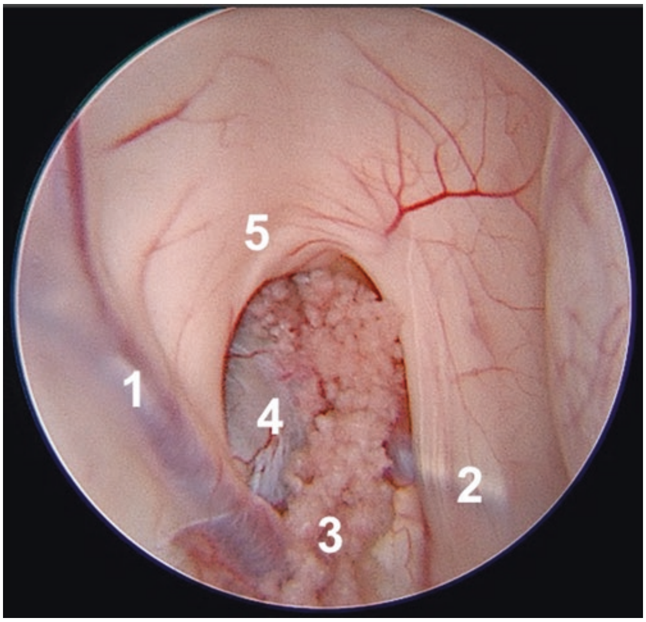

Fig. 15.1 Endoscopic view of the right lateral ventricle. *1* Septal vein. *2* Thalamostriate vein. *3* Choroid plexus. *4* Colloid cyst. *5* Fornix

posterior edge of the foramen of Monro down to the tip of the temporal horn where it terminates in what is known as the inferior choroidal point, the point where the anterior choroidal artery enters the ventricle [23]. The choroid plexus lies along the choroidal fissure, in between the thalamus and fornix. These two structures have ridged edges called the tenia thalami and tenia fornix, respectively. At the temporal horn tip on the side of the fornix, it is called tenia fimbriae. Connecting these ridged edges is the tela choroidea which forms the attachment of the choroid plexus. The choroid plexus thus extends from the medial wall of the temporal horn and extends anteriorly and superiorly until it reaches the level of the foramen of Monro; there, it enters the third ventricle to become two strands of choroid plexus which lie on the roof of the third ventricle. At the level of the atrium, the choroid plexus enlarges to form a tuft called the glomus.

The third ventricle sits at the center of the ventricular system and is the site where most intraventricular endoscopic procedures are performed. It is limited by very important neurovascular structures, and therefore knowledge of its anatomy is crucial for the neuroendoscopist. To understand the third ventricle, one can envision it as a box with six walls. It communicates with the lateral ventricles through the foramen of Monro and posteriorly with the fourth ventricle via the cerebral aqueduct. The roof of the ventricle is made by the body of the fornix anteriorly and the crura and hippocampal commissure posteriorly. It has three additional layers: two layers of tela choroidea and a vascular layer in between them where the internal cerebral veins, the medial posterior choroidal arteries, and their tributaries traverse. The roof extends from the foramen of Monro to the suprapineal recess. The sides of the two layers of tela choroidea run along the medial edge of the choroidal fissure. The two strands of choroid plexus that run along the roof of the third ventricle are attached to the inferior layer of tela choroidea. The floor extends from the optic chiasm anteriorly to the opening of the Sylvian aqueduct posteriorly. Behind the optic chiasm are the infundibulum of the hypothalamus, the tuber cinereum, the mammillary bodies, the posterior perforated substance (visible from the undersurface of the brain), and the midbrain tegmentum. The superior edge of the optic chiasm marks the inferior aspect of the anterior wall and the inferior surface the anterior limit of the floor. Behind it lies the funnel-shaped infundibulum whose axons descend toward the sella where they connect with the neurohypophysis. Behind the infundibulum is the tuber cinereum, a relatively avascular section of gray matter from the hypothalamus. This is the site that is perforated to create a third ventriculostomy. The mammillary bodies are located posterior to the tuber cinereum. They are paired nuclei that connect with the hippocampus via the fornix. The anterior wall extends from the foramen of Monro to the optic chiasm. It is formed by a thin layer of gray matter that connects the optic chiasm with the rostrum of the corpus callosum called the lamina terminalis. Underneath the foramen of Monro lie the columns of the fornix. The anterior commissure (the bundle of white matter fibers that connect the frontal lobes) is caudal to the rostrum of the corpus callosum and is connected via the lamina terminalis with the optic chiasm. The lamina terminalis attaches on the midportion of the optic chiasm creating a small space called the optic recess.

The posterior wall of the third ventricle is constituted by the suprapineal recess, a space between the inferior layer of tela choroidea and the pineal gland. The pineal gland protrudes posteriorly into the quadrigeminal cistern. The pineal stalk has two leafs, a superior and an inferior. The habenular commissure, a white matter bundle of fibers that connects the habenular nuclei, attaches to the superior leaf of the pineal stalk. Finally, the lateral walls are marked by the thalamus and hypothalamus which are divided by the hypothalamic sulcus. The massa intermedia, a bundle of white matter fibers, extends between the thalami.

15.6 Surgical Technique

Operative Setup

For most intraventricular procedures, patients are positioned supine. Pin fixation is usual; the degree of neck flexion or extension has to be adjusted based on the pathology that is being addressed. It is important to make sure that the level of the head is above the heart to ensure adequate venous outflow and to decrease the amount of CSF egress after ventricular cannulation. The bed must be lowered as much as possible to avoid having instruments too close to the surgeon's face. The long axis of the head is kept neutral for most cases; all pressure points must be adequately padded and the patient securely fastened to the operating table. Operative films must be available for review. An indwelling Foley urinary catheter is placed only if the duration of the procedure is expected to be long or if there is an anticipated potential risk of blood loss that might require aggressive fluid resuscitation. An arterial line is not mandatory unless the complexity of the procedure demands close patient monitoring. Broad-spectrum intravenous antibiotics must be administered within an hour of making skin incision to ensure appropriate tissue concentration. Navigational guidance systems are placed at the foot of the bed with the reference array placed on the contralateral side of the surgical approach in order keep the surgeon's space clear (Fig. 15.2). The endoscope tower is also placed at the foot of the bed and must be placed at an angle that will provide the surgeon with a direct, unobstructed view of the monitor (Fig. 15.3). The monitor is more appropriately positioned at the head of the bed for occipital or suboccipital access routes. The anesthesia team will be positioned on one side of the patient, and opposite to them, the scrub nurse will position the instrument Table. A Mayo stand is also placed between the surgeon and scrub nurse to facilitate the transfer of instruments. Prepping and draping are completed in a standard fashion. A craniotomy drape can be used, but it has been our practice for most endoscopic procedures to use Ioban™ (3 M, St. Paul, MN, USA) and a smaller thyroid drape on top of which we place strips of Ioban on the edges. We also place a collection bag making sure that the edges of the Ioban overlap with the bag so that any rundown will go to the bag and not under the drapes. The collection bag is connected to a drainage system to avoid excessive accumulation of fluid which can sometimes put tension on the drapes or the skin incision. An absorbent pad can be placed at the head of the bed to keep the site clean and dry. Before starting the procedure all of the equipment must be checked to ensure adequate functioning. The endoscope has to be adjusted to correct white balance and image definition. It is common to have a multitude of cables and tubes during these procedures: endoscope lighting, camera, irrigation tubing, suction tubing, monopolar Bovie, and bipolar forceps. Care must be taken to ensure that there is enough slack to move the different pieces of equipment freely but without redundant cables that will become tangled during the operation. It is a good practice to inject local anesthetic with epinephrine a few minutes before the start of the case. This can be done immediately following surgical time-out, while the surgical assistant is preparing and organizing the equipment; this will provide better skin hemostasis.

It is important to test all of the endoscope's lenses before the start of the case even if the surgeon anticipates only using

Fig. 15.2 Right frontal endoscopic approach. Notice the placement of the reference array on the opposite side of the surgeon

Fig. 15.3 The video tower and navigation system are placed at the foot of the bed providing the surgeon and assistant an unobstructed view

one as is the case for most endoscopic third ventriculostomies. It is always a safe practice to prepare for distorted anatomy that might warrant the use of a lens with a different viewing angle during part of the procedure.

Instrumentation

There are different commercially available endoscopes suitable for intraventricular procedures. Each one has pros and cons that are beyond the scope of this chapter to discuss. Endoscopes can be flexible or rigid, but for the purposes of this chapter, we will only consider the latter. Most endoscopes consist of a lens connected to a fiber-optic camera and light source and a sheath with a variable number of working portals (depending on make and model). In addition to the endoscope, a basic neuroendoscopy tray will contain a monopolar and a bipolar cautery, scissors, and grasping forceps. The instruments can have rotating tips to help accommodate the sometimes odd working angles. This also allows the surgeon and his assistant to work without having to continuously move their hands during the procedure (Fig. 15.4); it is important to remember that any movement will be transmitted to the shaft of the endoscope thereby creating distortion of the image and potentially injuring deep neurovascular structures. During any endoscopic procedure, an irrigation tube is connected to one of the working channels. Continuous irrigation clears the field of view and helps achieve hemostasis for small bleeding vessels.

Depending on the procedure, the surgeon will want to have available ancillary instruments which include a 3 or 4 Fr. embolectomy catheter with an inflatable balloon tip attached to a 1 cc Luer Lock syringe and a 6 Fr. endotracheal suction catheter. Peel-away sheaths are useful adjuncts to create a permanent working path for the endoscope for the duration of the procedure. There is a caveat to this: the sudden, large-volume egress of CSF from the ventricle might distort the anatomy which in turn will require larger amounts of irrigation.

Nuances of Burr Hole Placement

As detailed in the operative setup section, once the patient has been adequately positioned, all pressure points have been padded, and the navigation system has been registered, the surgeon must proceed to plan the most appropriate location for the burr hole. A standard ETV is planned using the standard frontal approach typically used for the insertion of ventricular catheters. In reviewing the preoperative films, however, the surgeon must anticipate the trajectory of the endoscope that will maximize the effectiveness of the procedure while minimizing the risk to anatomical structures that can occur by moving the endoscope inside the ventricle. If the goal of the procedure is to fenestrate the floor of the third ventricle, the ideal route is one that follows a line that is parallel to the rostral (sphenoidal) clivus. In some circumstances, this will place the burr hole more anteriorly than in a standard frontal ventriculostomy. This will minimize the degree of AP translation of the endoscope which can potentially injure structures like the fornix once inside the third ventricle. It is important then that if the planned entry site is slightly anterior to the traditional approach, the head of the patient be placed in slight extension at the beginning of the case. This same principle of planning for burr hole placement applies to other pathologies and can imply using even less conventional approaches like placing a burr hole on the forehead and using a skin crease to plan the incision (Fig. 15.5). This approach is especially useful for lesions at the posterior or superior third ventricle that would otherwise be difficult to visualize and manipulate. If the surgeon is planning on performing more than one procedure (e.g., ETV + biopsy), it might be necessary to plan for more than one burr hole depending on whether or not both problems can be addressed through the same surgical route.

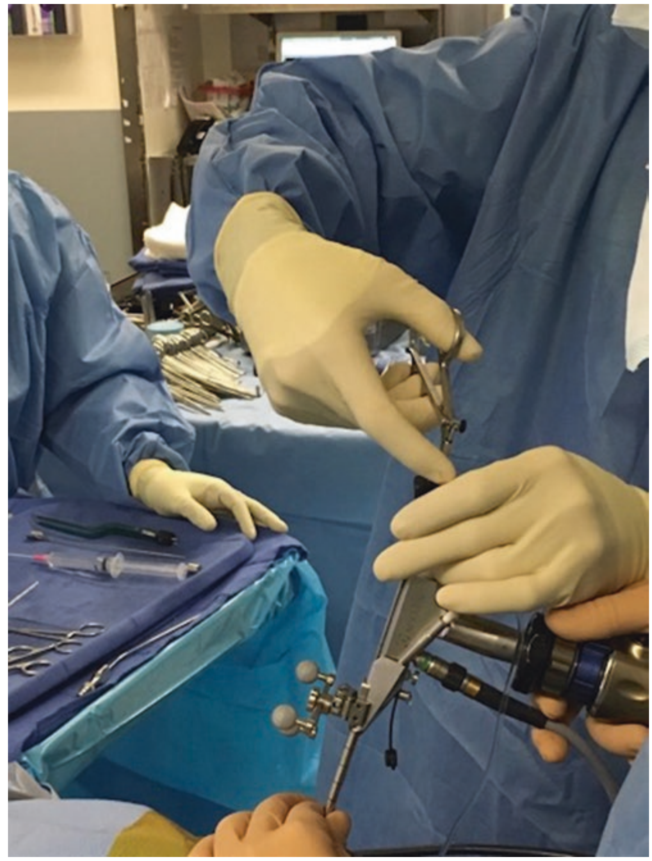

Fig. 15.4 The use of instruments with rotating tips allows the surgeon to adapt to the ventricular morphology without losing an ergonomic posture or transmitting excessive motion to the shaft of the endoscope

Fig. 15.5 T2-weighted sagittal image shows a colloid cyst of the third ventricle (*small arrow*). Planning a trajectory that would be used for an ETV (*dotted arrow*) might limit visualization and force the operator to use unnecessary torque with the endoscope. A more anterior trajectory (*long solid arrow*) will provide a direct view. Preoperative burr hole planning is fundamental for the success of the procedure

Occasionally one might be able to perform an ETV and a biopsy using the same surgical approach as can be the case with a large pineal region tumor. This might not be possible if the lesion is small and does not present anterior to the massa intermedia. In surgical planning, one always needs to confirm that the trajectory does not violate any sulcal margin either on the cerebral convexity or interhemispheric region. In the event that a sulcus is recognized at the time of durotomy, it is prudent to enlarge the burr hole rather than pass the endoscopic sheath into this pial interface.

Avoidance and Management of Injury to Intraventricular Structures

Once the burr hole placement has been decided, the appropriate skin incision is planned. One must take into account the possible need to convert an endoscopic procedure to an open one. This might be the case during tumor resections where the surgeon might encounter brisk bleeding or a tumor with a consistency that does not lend itself to removal through the endoscope's working path. If so, the incision site has to be prepared and the possible flap discussed in order to avoid confusion if and when the procedure changes to an open approach. The head shave that is required for most endoscopic procedures is minimal. Nonetheless, it is also important to take this into account when planning a possible conversion to an open surgery or in the event that an external ventricular drain (EVD) needs to be placed.

Once the incision has been planned and the operative site is marked, sterile prepping and draping of the surgical site are completed in standard fashion. After local anesthetic is injected, incision is performed with a skin knife and carried down to the periosteum using monopolar Bovie cautery with a protected tip. A self-retaining retractor is placed and the burr hole created with a pneumatic drill. A self-stopping perforator drill bit can be used, but it is the senior author's (MMS) preference to use a 4 mm cutting or acorn drill bit. It is important to make sure that the burr hole is large enough to fit the endoscope sheath but also to allow freedom of movement once the endoscope is inserted. Once the dura is exposed, it is coagulated with the bipolar and sharply incised using a number 11 blade. The edges of the dura are coagulated to prevent any subdural bleeding. Then the pia mater is coagulated and cut using a sharp blade. If the ventricles are small, at this time, a ventricular catheter is inserted under navigational guidance. Small aliquots of approximately 5 cc of sterile saline are slowly instilled through the catheter (Fig. 15.6); This can be repeated two or three times and must be done slowly, paying attention to the patient's heart rate to ensure that bradycardia does not develop secondary to intracranial hypertension. Before pulling the catheter out, the surgeon can use it to guide the advancement of the endoscope (Fig. 15.7). If the procedure requires navigation of the endoscope, the equipment must be registered before the start of the case. An important note to take into account is that the reference array must be attached at the most proximal end of the endoscope to ensure that it will not obstruct the path of the endoscope thereby limiting the depth at which the sheath can be introduced. It is important to remember that all of the working channels must have an obturator placed inside. The endoscope is then advanced slowly into the brain parenchyma while simultaneously paying attention to the navigation system and the video monitor. Once the tip of the endoscope traverses the ependymal surface and the ventricle is entered, the irrigation is connected and opened. The continuous flow of lactated Ringer's solution clears the view and helps maintain an aqueous environment inside the ventricle at all times. Before proceeding to complete the intended procedure, it is important for the surgeon to inspect and identify the critical anatomical landmarks inside the ventricle and any variations that could potentially affect the surgical approach. Every time the endoscope is advanced inside the ventricle, the surgeon must not only be aware of what lies in front but also what has been traversed thus far. Structures which might not be visible can be stretched and torn when moving the endoscope. This "blind rear view" is governed by memory of the endoscopic path.

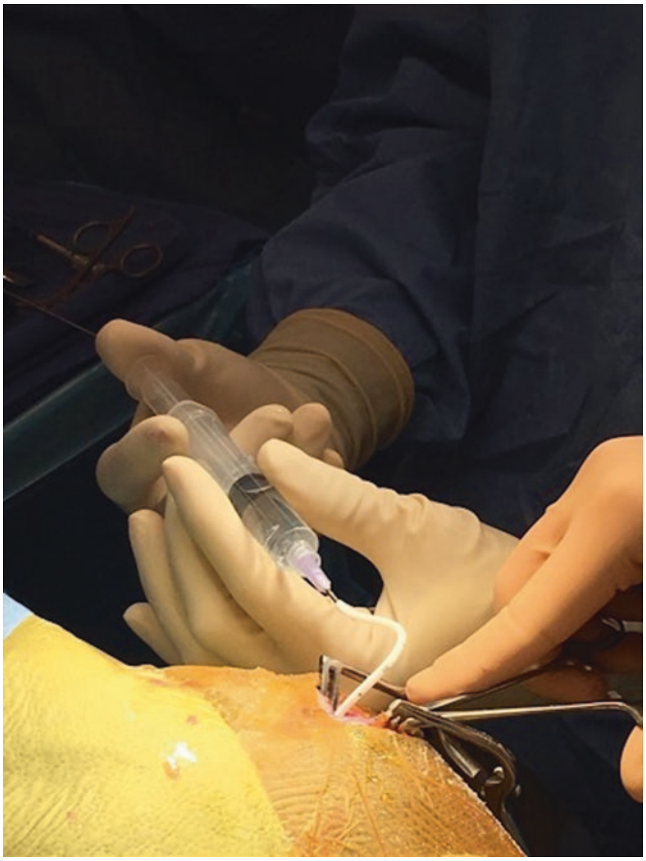

Fig. 15.6 After an intraventricular catheter is placed, small aliquots of sterile saline are slowly injected in order to expand the ventricle. This technique is useful in patients with small ventricles

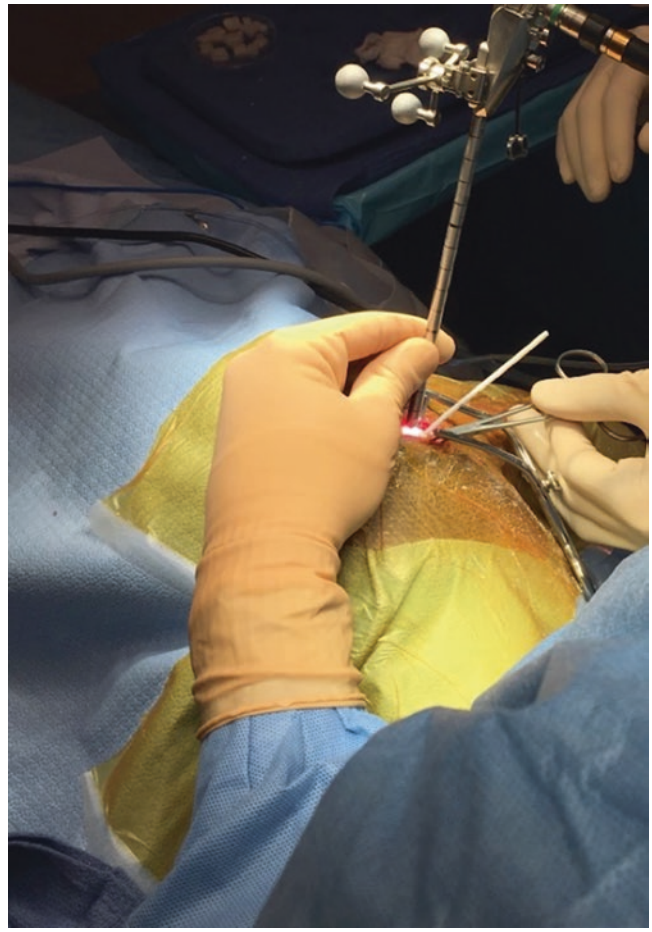

Fig. 15.7 After inserting a ventricular catheter under navigational guidance to infuse sterile saline solution into the ventricle, the endoscope is inserted. The same tract used with the catheter can be followed under direct vision with simultaneous navigation of the endoscope

15.7 Postoperative Care/Complications

For most simple endoscopic procedures, it is possible to discharge the patients after one night of observation. We routinely obtain an MRI on post-op day 1. At our institution, we have developed a specific imaging protocol for endoscopic third ventriculostomies which includes a fine-cut, sagittal, T2-weighted image in order to assess for patency of the stoma. We reserve the use of external ventricular drains to instances where the case is complicated by significant hemorrhage. It is not uncommon to encounter some degree of bleeding during most simple procedures. This, however, can be controlled by irrigation and watchful waiting. If the bleeding proves to be stubborn, the Fogarty embolectomy catheter can be insufflated and used to hold pressure against the bleeding site. If the source is visible and judged to be difficult to control with pressure, bipolar cauterization can be used cautiously. It is important to remember that bipolar diathermy can have the risk of heat transmission to important neural structures adjacent to the site of bleeding. In most cases, the bleeding will be self-limited. Modest venous bleeding is sometimes precisely identified by approaching the site with continuous irrigation. If a small venous tributary is recognized, then direct bipolar diathermy is justifiable and frequently successful. Balloon tamponade is useful if the source of hemorrhage lies within small chambers (i.e., third ventricle) or parenchymal sources (tumor tissue). For significant hemorrhage that clouds visualization, we will remove the endoscope and insert a ventricular catheter through which we will irrigate copiously. Once the bleeding starts to clear, we will take a second look with the endoscope. In some instances, additional irrigation will be required before being able to regain the ability to navigate the ventricles safely. It is important to inspect key areas like the foramen of Monro for abandoned clots. If the clots are overlying the area of suspected bleeding, it is better to leave them untouched. However, if they are distant from the suspect site of hemorrhage and could potentially lead to loculated hydrocephalus, it is better to try to remove them. Retrieval of clot overlying the aqueduct is nearly a routine at the completion of colloid cyst removal. Our approach is to use the 6 Fr. endotracheal suction catheter. Intraventricular clots can be extremely adherent to the underlying tissue, so extreme caution must be exercised when deciding to extract them. As a general rule, hematoma that is adherent to the choroid plexus or at the choroidal interface is best left in situ. A useful trick is to spin the suction catheter at the same time that the endoscope is retracted. If this is insufficient, an aspirator with a side-cutting tip can be used to remove the clot. This can be a two-edged sword because the potency of this device can actually induce additional bleeding.

The use of an EVD must be weighed on a case-by-case basis taking into account multiple factors. If there is bleeding that is judged to be significant enough to potentially cause acute hydrocephalus in the postoperative setting, it is always better to leave a drain. In those circumstances, we will leave the drain open continuously with a low pop-off setting (0–5 mmHg). This will allow continuous drainage of hemorrhagic CSF and monitoring of ICP. If the pressures remain stable and the fluid looks progressively clearer, we start to challenge the patient by elevating the pop-off setting gradually until we can remove the ventriculostomy.

Another reason to leave an EVD can be in the setting of an attempted ETV in a patient who presents with hydrocephalus secondary to a shunt malfunction. Since those patients can have an unpredictable course and are at risk of becoming acutely symptomatic from high intracranial pressure, one can place a drain and leave it clamped. If the patient becomes symptomatic or the pressure becomes persistently high, then a VP shunt is placed.

15.8 Nuances of Intraventricular Approaches in Small Ventricles

It is not uncommon to find in the endoscopy literature that one indication for these types of approaches is the finding of concomitant hydrocephalus associated with the primary pathology to be treated. This contention, however, has been challenged by reports of successful endoscopic treatment of intraventricular lesions in patients without ventriculomegaly (Fig. 15.8). The senior author (MMS) published his series of intraventricular lesions without hydrocephalus; he compared 15 cases with a reference group of patients who had ventricular enlargement and did not find a significant difference in morbidity or the rate of success of the procedure based on the original intended purpose of the surgery [24]. Yamamoto et al. suggested the use of flexible endoscopes for patients with small ventricles [25]; this, however, comes at the price of reduced optical quality which is not justified from our perspective.

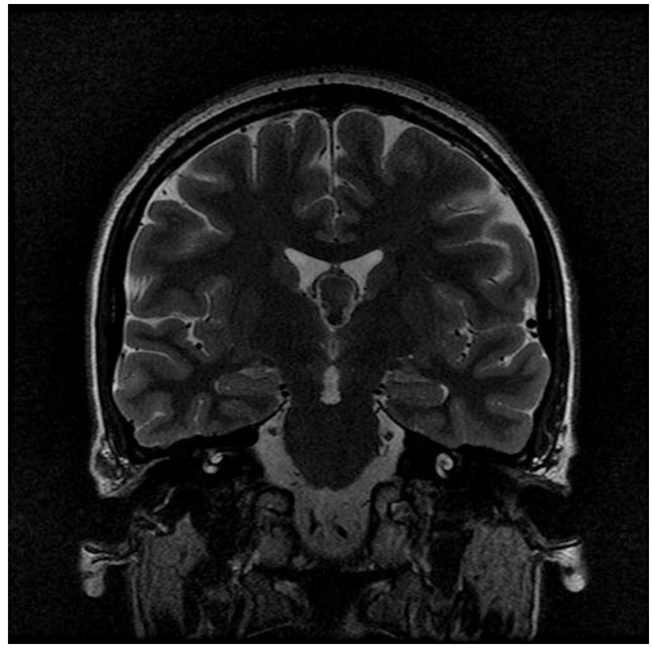

Fig. 15.8 T2-weighted coronal MRI shows a colloid cyst of the third ventricle in a patient with small ventricles. Although technically challenging, these lesions can be safely resected through a purely endoscopic approach

15.9 Surgical Pearls

- Patient positioning and adequate trajectory planning are critical to avoid potential injury to periventricular structures.
- The use of instruments with rotating tips facilitates manipulation and decreases torque of the endoscope's sheath.
- The instillation of small aliquots of saline solution helps to navigate small ventricles.
- The neuroendoscopic surgeon must be prepared to convert to open microsurgical approaches and must plan the incisions accordingly.

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Part III

Target-Based Comparison of Approaches

16 Olfactory Groove Meningiomas

Michael W. McDermott*, Henry W. S. Schroeder*, and Verena Gellner*

Contents

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16.1 Olfactory Groove Meningiomas: Editors' Introduction

Meningiomas are the most common benign intracranial tumors, accounting for up to 20% of all primary brain tumors. Olfactory groove meningiomas (OGMs) comprise up to 10% of all meningiomas [1, 2]. They arise from the midline ventral skull base dura around the cribriform plate and planum sphenoidale. The origin of OGMs is the arachnoid cap cells, with the vascular supply mainly arising from the anterior and posterior ethmoidal arteries. As these tumors enlarge, they can involve the olfactory nerves, compress the optic nerves and frontal lobes, and displace or encase the anterior circulation. As such, the common presenting symptoms are anosmia, changes in mental status, headaches, and visual abnormalities. There is also a propensity to erode into the ethmoid sinus, complicating both the resection and skull base reconstruction.

The traditional resection of olfactory groove meningiomas has been via a bilateral or unilateral subfrontal approach; however, more recently, less invasive techniques have been developed. Two such techniques are the keyhole craniotomy via an eyebrow approach and the expanded endonasal endoscopic approach. In this chapter, the authors will discuss the merits of each technique within the context of the case example presented below (Fig. 16.1a–f).

16.2 Olfactory Groove Meningiomas: Open Transcranial Approaches

Ramin A. Morshed, Stephen T. Magill, Michael Safaee, Jacob S. Young, and Michael W. McDermott

Brief Introduction

Olfactory groove meningiomas (OGMs) arise just behind the crista galli at a point corresponding to the suture line separating

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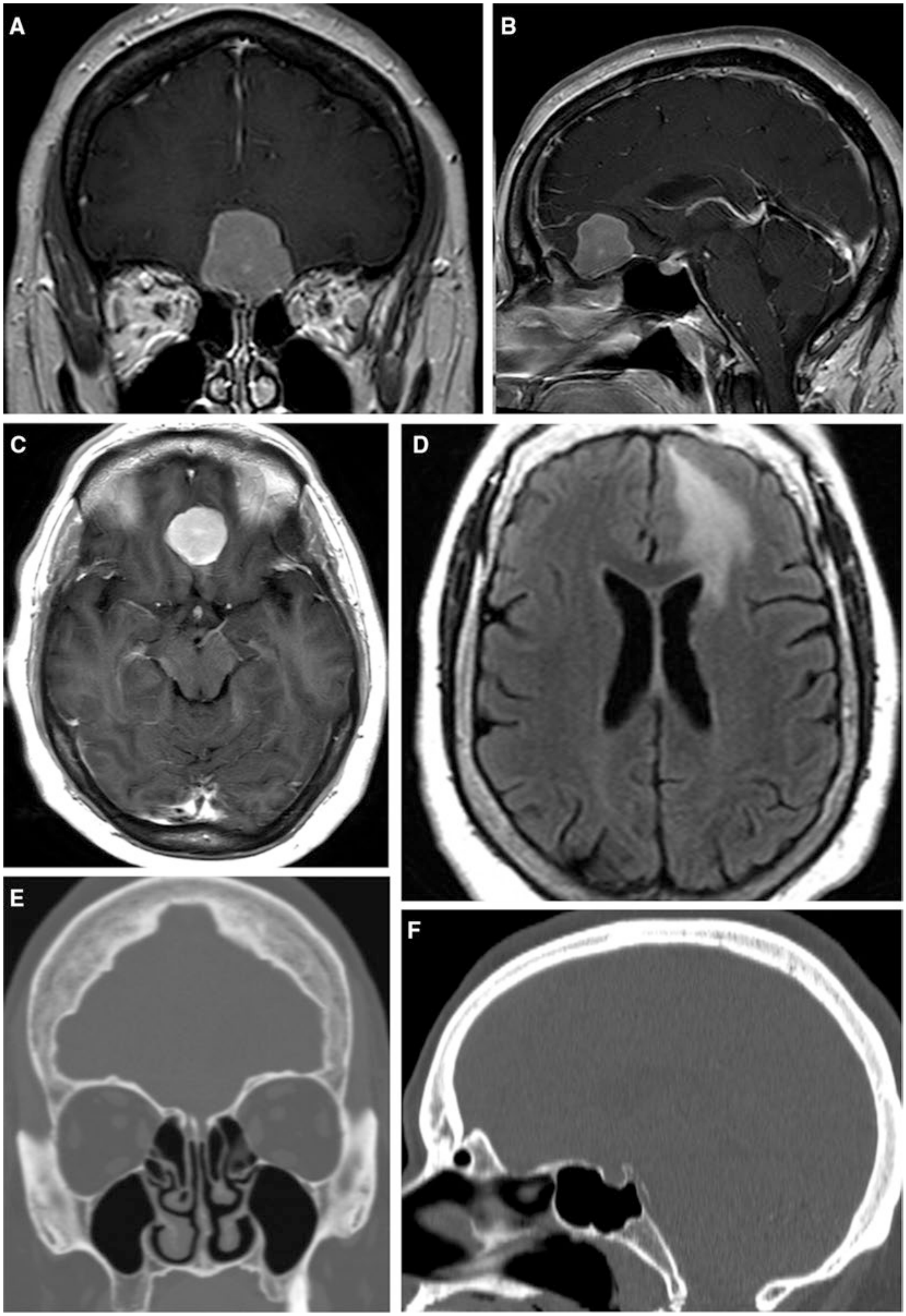

Fig. 16.1 (**a**–**f**) **CASE EXAMPLE**: Olfactory groove meningioma. A 54-year-old woman presents with primary complaints of 6 months of anosmia. Examination: anosmia, otherwise neurologically intact

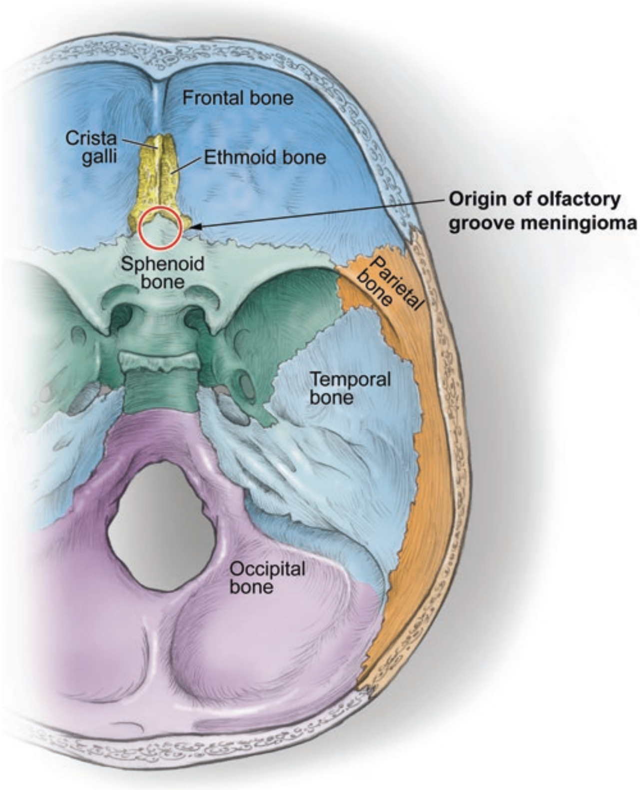

Fig. 16.2 Schematic of internal view of anterior fossa demonstrating origin of olfactory groove meningiomas (OGMs). Note the relationship to the crista galli anteriorly and the optic canals, anterior clinoids, and tuberculum sella posteriorly

the planum sphenoidale and the cribriform plate (Fig. 16.2) [3]. These tumors can grow symmetrically around the crista galli or predominantly involve one side. As these tumors first slowly compress the inferior aspect of the frontal lobes (Fig. 16.3a–d), there is often a lack of focal neurological deficits early in the clinical course. Although patients may experience changes in personality and cognition as a result of the frontal lobe compression, family and even physicians often fail to identify these symptoms as focal neurological findings, which results in delayed evaluation and large tumor sizes at time of diagnosis.

Large OGMs can cause seizures from frontal lobe irritation or visual disturbances secondary to downward pressure on the optic nerves or compression of the optic chiasm. Large unilateral tumors can cause a classic Foster-Kennedy syndrome with anosmia, ipsilateral optic atrophy, and contralateral papilledema. In the original series of 29 patients reported by Cushing, the most common primary symptoms were anosmia (48%), headache (27%), impaired vision (24%), and personality change (21%), demonstrating the rather nonspecific presentation with olfactory groove meningiomas [3].

Classically, open craniotomies have been the workhorse for addressing these lesions. Here, we summarize common surgical approaches and review outcomes from open surgeries targeting these lesions. Finally, we present example cases demonstrating the thought process behind developing a surgical plan that maximizes tumor resection while preventing common postoperative complications.

Considerations for Surgery

The ideal surgical approach should provide exposure of the meningioma and its dural attachments, vascular supply, and any involved bone. Small asymptomatic OGMs can typically be observed until documented growth and/or the development of symptoms attributable to the tumor site. For patients who are asymptomatic from a stable mass in this area, serial imaging every 6–12 months may be warranted and should be presented as an option to patients. Our group has previously found that OGMs/planum meningiomas greater than 42 cm^3 were 8 times more likely to cause visual symptoms compared to those smaller than 42 cm^3 [4]. Earlier intervention on a smaller tumor can be an option prior to development of visual impairment, which may not be salvageable. When considering treatment, age is also an important factor. For younger patients, the goal may be to achieve a Simpson Grade I resection, whereas for an older patient, a Simpson Grade II resection may be acceptable in order to preserve functional status. Additional options for smaller tumors include radiosurgery and endoscopic resection as discussed elsewhere in this book. The focus of this chapter will be on open microsurgical approaches to OGMs that require surgical resection.

The imaging characteristics of the lesion guide approach selection. In addition to standard magnetic resonance imaging (MRI), preoperative magnetic resonance angiography (MRA) is helpful for larger tumors to identify the ascending A2 branches and their relationship to the posterior margin of the tumor. Blood supply to these lesions typically originates from the anterior and posterior ethmoidal arteries, anterior branches of the middle meningeal artery, and the meningeal branches of the ophthalmic artery, although additional blood supply from branches of the anterior cerebral artery (ACA) is not uncommon. Figure 16.4 displays an angiogram of a large OGM supplied by multiple ethmoidal branches of the ophthalmic arteries bilaterally. Given their predominant supply from the ophthalmic artery, they are not typically amenable to preoperative embolization due to the risk of vision loss.

In preparation for surgery, a lumbar drain can be placed preoperatively to improve brain relaxation during the case so that less brain retraction is needed to access the tumor. However, for some approaches, the Sylvian and basal cisterns may be entered to achieve CSF drainage with a similar outcome, negating the need for a lumbar drain. Additionally, preoperative dexamethasone may be considered for 7–10 days if imaging demonstrates significant frontal lobe edema.

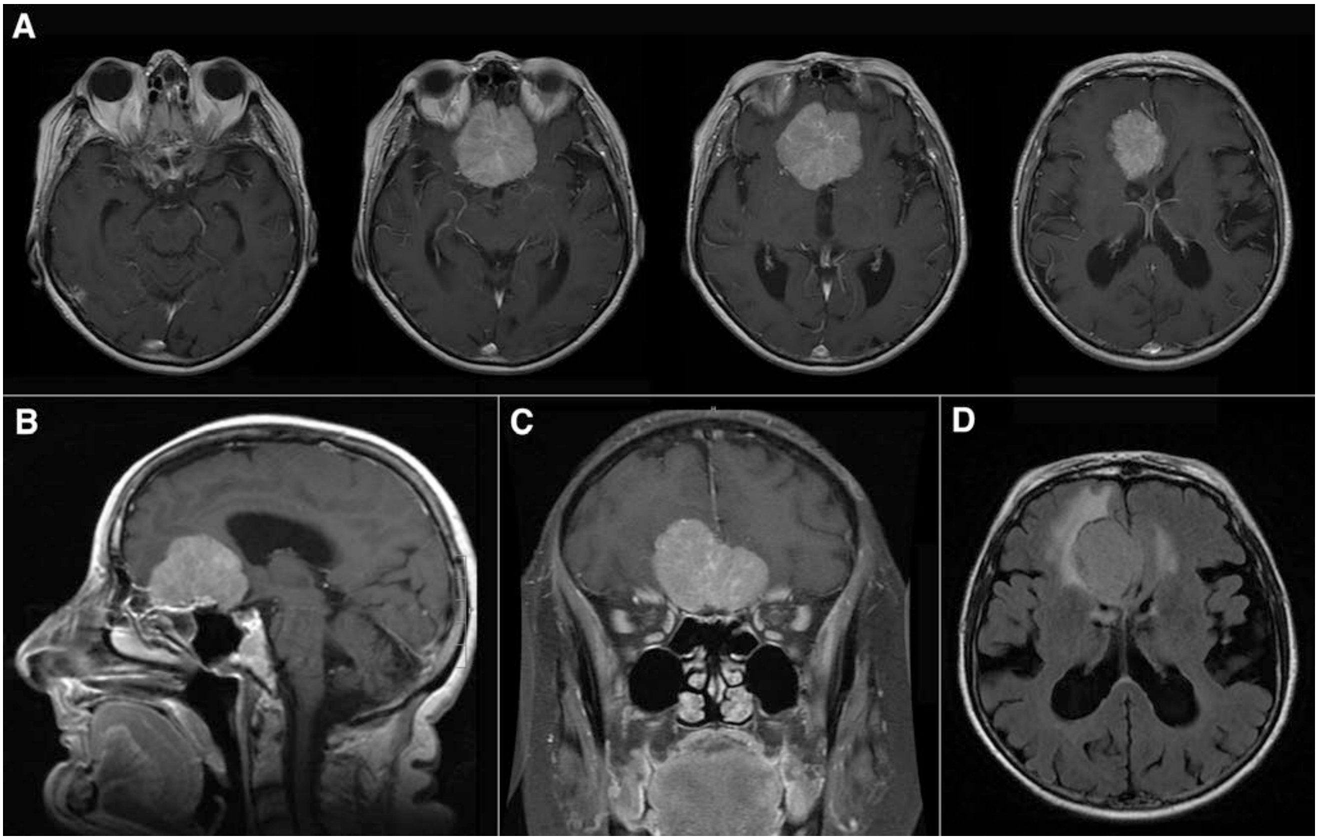

Fig. 16.3 (**a**–**d**) Example of an OGM. Images are from a 70-year-old female who initially presented with worsening headaches, decreased sense of smell, and short-term memory problems. (**a**) Serial axial post-gadolinium T1-weighted images demonstrate a 4.6 × 4.5 × 3.1 cm enhancing mass around the olfactory groove and planum sphenoidale region. Sagittal (**b**) and coronal (**c**) images are also provided and demonstrate extension into the suprasellar region and compression of the right side of the optic chiasm. Draping of the anterior communicating artery over the meningioma was also noted. No extension into the paranasal sinuses was seen. This mass was associated with frontal lobe edema, more prominent on the right side (**d**)

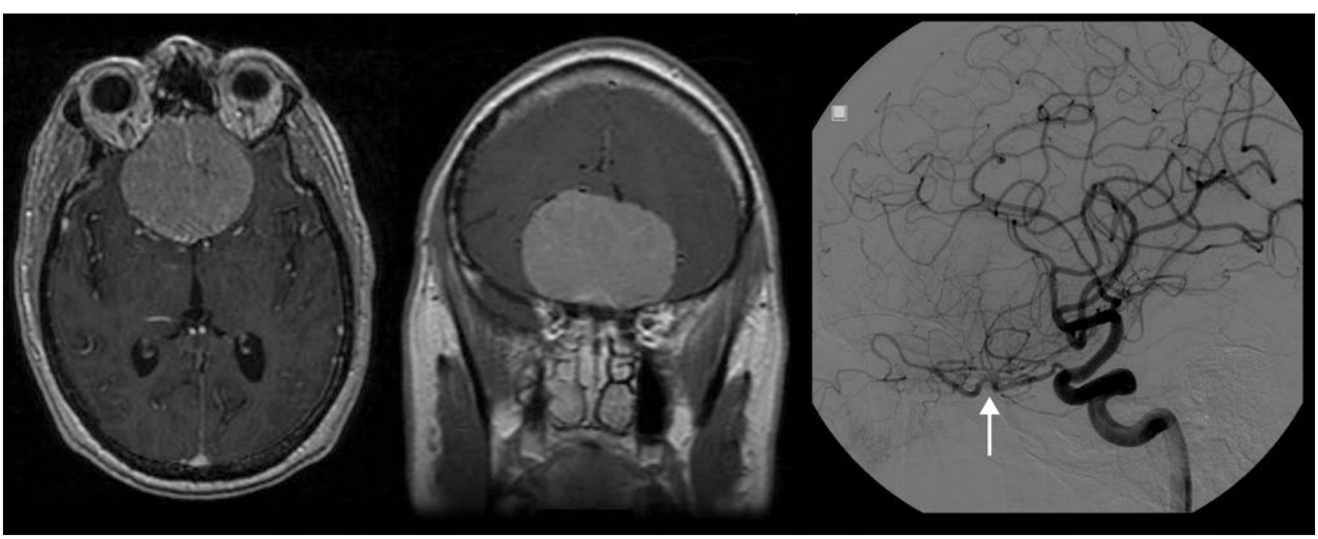

Fig. 16.4 Images from a case of a 60-year-old male who presented with several months of decreased concentration, abulia, and loss of smell who presented with a seizure. Axial (*left*) and coronal (*middle*) post-contrast T1-weighted images demonstrate a 5.9 × 4.5 × 5.4 cm enhancing mass within the olfactory groove displacing the ACAs posteriorly. Angiogram (*right*) demonstrating predominant arterial supply from multiple ethmoidal arterial branches of the ophthalmic arteries bilaterally, precluding embolization

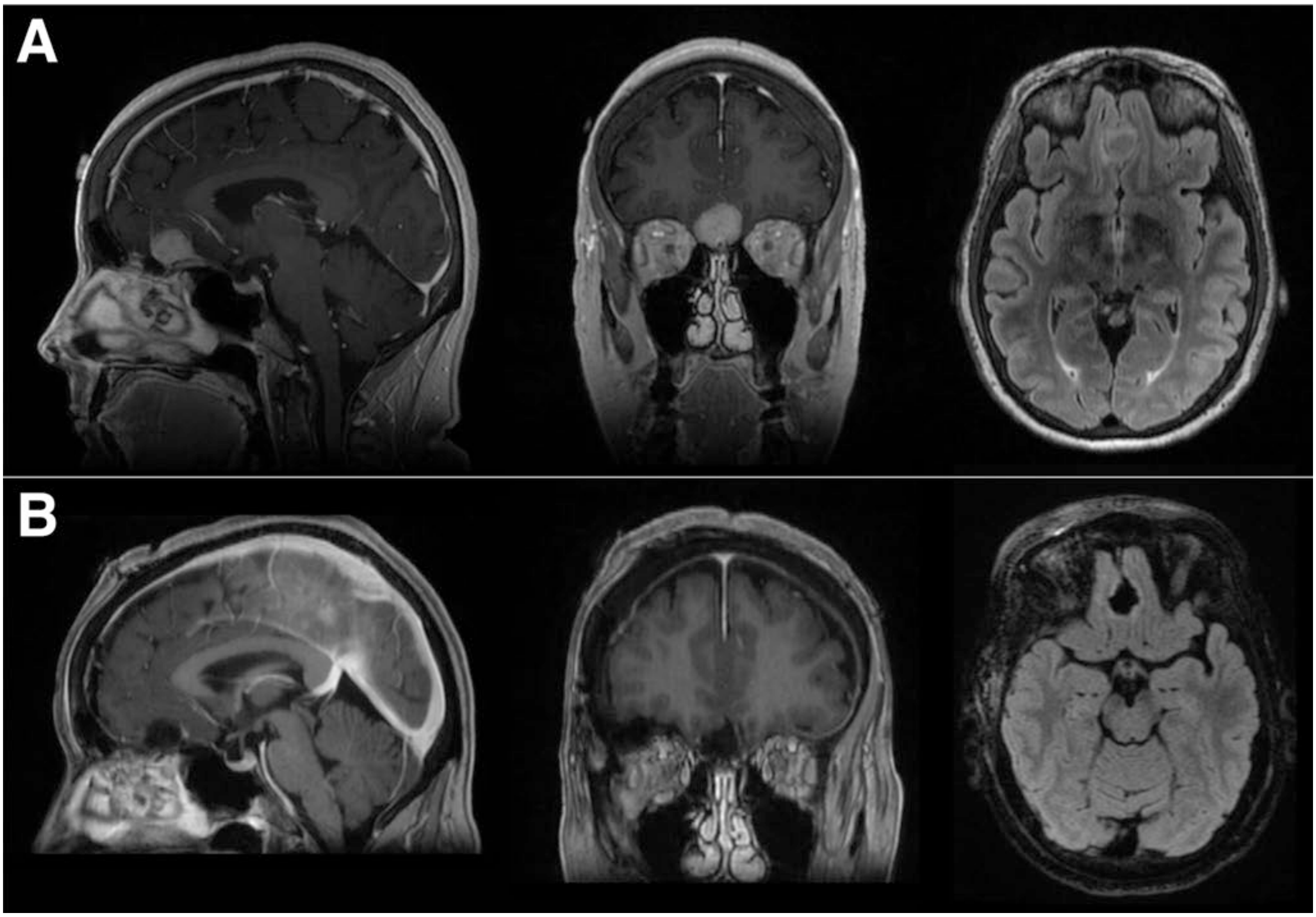

Fig. 16.5 (**a**, **b**) Case 1: management of a tumor less than 3 cm in diameter using a unilateral cranio-orbital approach. Images from a 58-year-old female who presented with headaches with subsequent syncope in addition to olfactory and vision changes. (**a**) Preoperative imaging demonstrated a dural-based enhancing mass (*left and middle images*) originating from the right olfactory groove measuring 2.5 × 2 × 2 cm without appreciable surround edema on FLAIR imaging (*right image*). (**b**) The patient underwent a right frontotemporal cranio-orbital approach with removal of the roof of the orbit. Gross total resection was observed on post-contrast T1-weighted imaging (*left and middle image*) without significant edema on FLAIR (*right image*). The patient had preservation of olfaction

Surgical Technique

We typically use two approaches to address OGMs: (1) a unilateral cranio-orbital approach for tumors less than 3 cm (Fig. 16.5a, b) and (2) a two-part bifrontal craniotomy with tailored nasofrontal osteotomy for tumors greater than 3 cm (Fig. 16.6a–c). Here, we describe these approaches in detail.

Management of Tumors <3 cm: The Unilateral Cranio-orbital Approach

For tumors less than 3 cm in size (Fig. 16.5a, b), a right frontolateral approach is preferred unless the tumor is eccentric to one side. Smaller tumors in this region do not displace the frontal lobe up and away from the skull base and lie medial to the orbital roof, which hides the origin of dural attachments (Fig. 16.7a, b). A curvilinear or coronal incision is used depending on the level of the hairline. We use a frontotemporal craniotomy, staying above the sphenoid wing, and a separate frontal orbitozygomatic osteotomy that extends just below the frontozygomatic suture. After this, the medial roof of the orbit is removed with small rongeurs so that the plane of view is flattened toward the plane of the cranial base behind the crista galli. This allows better visualization of the floor of the anterior fossa up toward the cribriform plate. This is critical as incomplete access to this region may reduce the chance for complete tumor removal and makes drilling of the posterior aspect of the crista galli very difficult. Furthermore, it reduces the need for brain retraction.

With the bony work complete, the dura is opened in a curvilinear manner based anteriorly, and sutures are placed in the dural fold over the orbit to provide additional downward retraction of orbital contents. With the operating microscope, the lateral Sylvian fissure is opened to release cerebrospinal fluid (CSF) and to reduce tension across the fissure with gentle upward retraction of the frontal lobe. By aiming toward the apex of the fissure, the olfactory tract and

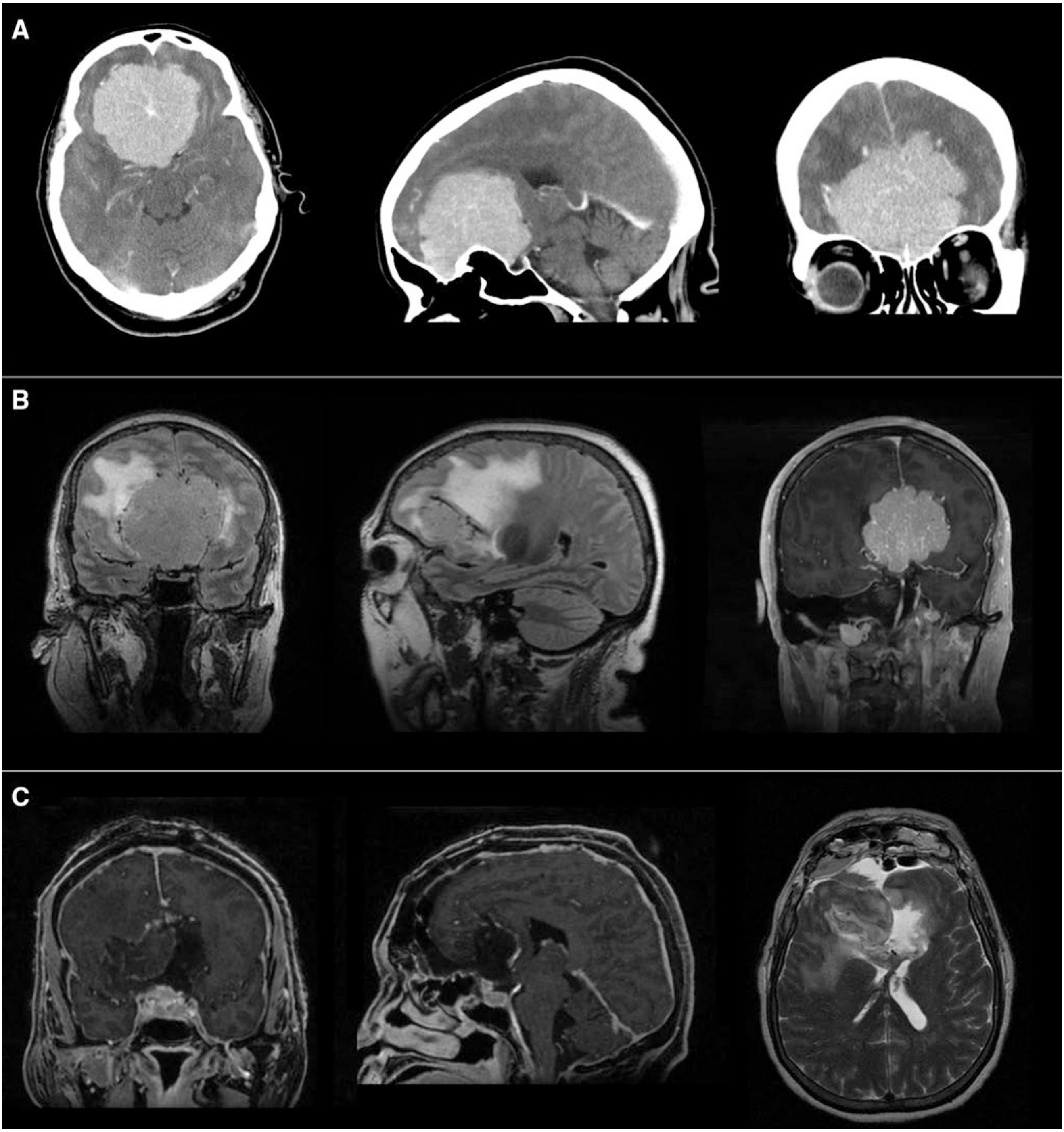

Fig. 16.6 (**a**–**c**) Case 2: management of a large OGM greater than 3 cm in diameter using a two-part bifrontal craniotomy with tailored naso-frontal osteotomies. Images of a 64-year-old female who presented with falls, anosmia, blurry vision, headache, and nausea. (**a**) Preoperative CT scan with contrast (axial, sagittal, and coronal sections) demonstrated a 7 × 6.5 × 5 cm anterior fossa mass associated with focal hyperostosis of the planum sphenoidale. (**b**) On MRI, the mass was noted to compress the optic chiasm inferiorly and either displace or encase branches of the ACA. Additionally, the mass compressed the third ventricle posteriorly and splayed the lateral ventricles bilaterally. On FLAIR imaging (*left and middle images*), significant bifrontal edema was noted. Post-contrast T1-weighted imaging (*right*) demonstrated close association between the posterior aspect of the tumor and branches of the ACA. (**c**) The patient underwent an extended bifrontal craniotomy with orbital osteotomies. Postoperatively, post-contrast T1-weighted imaging (*left and middle images*) demonstrated removal of the tumor. The patient had an excellent recovery and was neurologically intact except for persistent anosmia

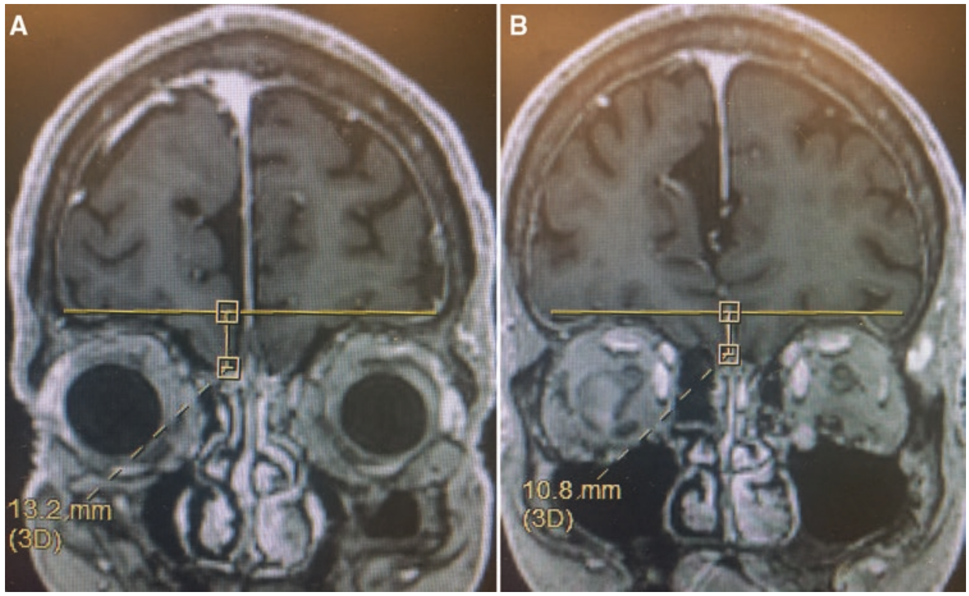

Fig. 16.7 (**a**, **b**) Coronal MRI images illustrating the distance between the plane of the orbital roof and (**a**) the point of origin for tumors just behind the crista galli and the olfactory groove and (**b**) the planum sphenoidale

optic nerve can be identified. By following the olfactory tract on the gyrus rectus proximally, one will see the optic nerve medial to the tract. We then move forward along the tract and coagulate vessels within the dura of the planum sphenoidale while working toward the tumor within the olfactory groove. Usually, it is more efficient to divide the ipsilateral olfactory tract at the posterior margin of the tumor. By doing so, the basal attachments can be coagulated and divided until the back of the falx and crista galli are reached. Bleeding from branches of the ethmoid artery coming through the cranial base can be stopped with bone wax and/or the diamond drill.

The falx is followed until the superior edge of the tumor is reached. We then incise the edge of the falx in an anterior and inferior direction until the crista galli is reached. A diamond drill can be used to remove the crista galli down to its base. Usually the anterior portion of the tumor on the contralateral side can then be seen in addition to the olfactory tract/bulb swinging toward the olfactory groove.

The tumor is debulked centrally followed by peripheral dissection. Small branches of the medial orbitofrontal artery supplying the lesion can be isolated and divided. In most cases of small tumors, the opposite olfactory tract can be dissected free in order to preserve olfaction without compromising the extent of resection. Once the tumor is removed, further removal of the basal dura can be performed along with drilling of hyperostotic bone. However, for small tumors from a unilateral approach, we do not enter the nasal cavity. If tumor is seen within the ethmoid sinuses on preoperative scans, we prefer a bifrontal craniotomy in combination with an endoscopic transnasal transbasal approach.

Management of Tumors >3 cm: Two-Part Bifrontal Craniotomy with Tailored Nasofrontal Osteotomy

For OGMs greater than 3 cm in diameter (Fig. 16.6a–c), we prefer a bifrontal approach. With the patient in the supine position, the neck is flexed on the chest and the head extended so that the forehead is parallel with the floor. A coronal scalp incision is made 2 cm behind the hairline. The scalp is reflected forward in the subgaleal plane up to 1 cm posterior to the supraorbital margin and retracted over a single rolled gauze with scalp hooks. Stopping subgaleal dissection 1 cm posterior to the supraorbital margin ensures that the vascular supply to the pericranium is preserved. The pericranium is then dissected from the superior temporal line bilaterally, cut behind the coronal suture, and reflected forward to the supraorbital margin until the rim of the orbits, the supraorbital notch, and nasofrontal suture are identified. The pericranium is folded over the skin flap and covered with moist gauze to prevent dehydration.

Next, using image guidance, we mark out the midline and plan out a two-piece bone flap with planned cuts just slightly above the rim of both orbits (Fig. 16.8). The two-part craniotomy allows dissection of the superior sagittal sinus from the skull under direct visualization, maximizing safety. To complete the bone flap, a right-sided posterior parasagittal burr hole 1.5 cm lateral to the midline is made in line with the superior aspect of the tumor. The temporalis muscle is detached from the superior temporal line on both sides leaving a muscle cuff. A frontal burr hole is made just behind the base of the frontozygomatic process in the frontal bone

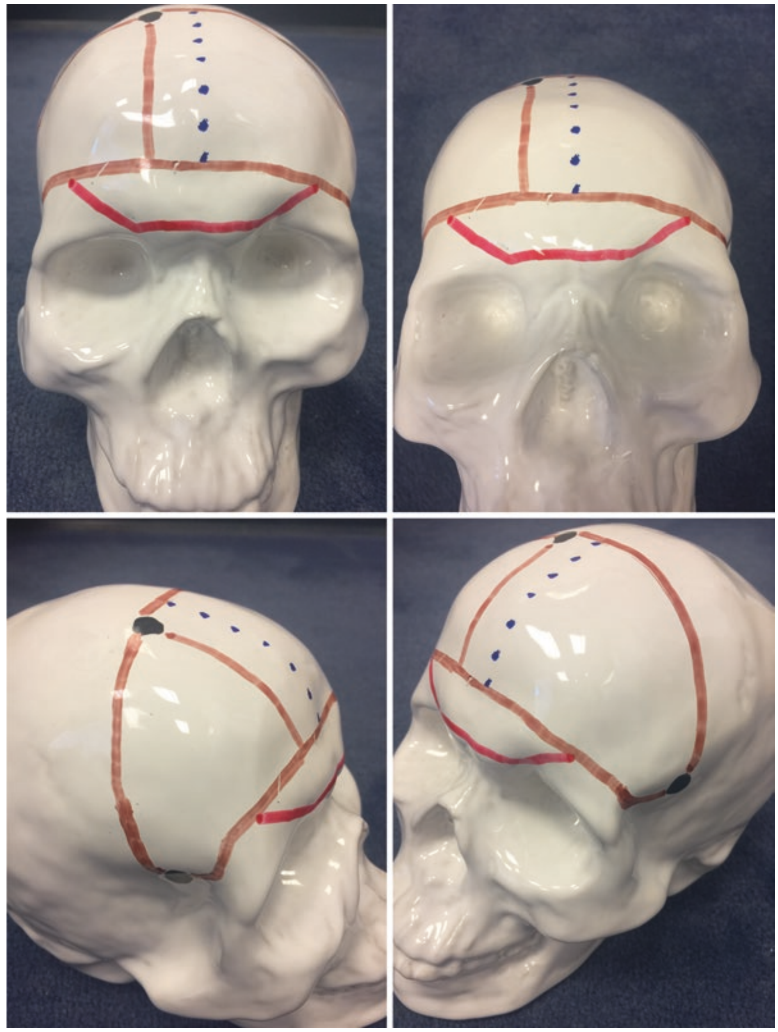

Fig. 16.8 Model skull demonstrating the two-part bifrontal craniotomy with tailored nasofrontal osteotomy. *Black circles* mark out the three burr holes made for the craniotomy. Midline is demarcated with a dotted *blue line*. *Brown lines* mark out the two-part bifrontal bone flap, and the *red line* marks the cut for the nasofrontal osteotomy

above the sphenoid wing on both sides. A small incision is made in the muscle cuff posteriorly on the planned back edge of the craniotomy to allow passage of the cutting blade. After dissecting the dura free, the footplate is used to make a cut below the muscle cuff from the frontal burr hole backward then up over the convexity to join up with the parasagittal burr hole. The next cut is from the frontal burr hole above the rim of the orbit stopping 1.5 cm from midline. The last cut is made from the parasagittal burr hole to the medial aspect of the planned nasofrontal osteotomy cut, and the bone flap is elevated. Under direct vision the dura over the superior sagittal sinus is dissected across midline. Bleeding from the sinus or venous lakes can be controlled with Gelfoam and small cottonoids. Next, the drill is used to cut from the left frontal burr hole posteriorly below the muscle cuff crossing it up over the convexity and across the dissected midline posteriorly. The last cut begins at the left frontal burr hole and crosses the supraorbital margin across midline to the right to complete the craniotomy. The left-sided bone flap can then be elevated. The dura is tacked up on the lateral and posterior margins of the craniotomy. Once these steps are complete, the dura can be reflected from the roof of the orbits, a short distance posteriorly on both sides, and the insertion of the falx into foramen cecum is cut.

We then draw out the template for the nasofrontal (also known as supraorbital) osteotomy (Fig. 16.8). The planned cut runs slightly above the nasofrontal suture and just medial to the rim of the orbit (without entering the orbit) until the supraorbital notch is reached. The cut is completed in an oblique fashion laterally through the remaining bone of the frontal bone. The opening in the frontal sinus is covered with Gelfoam, and any mucosa within the nasofrontal osteotomy is removed to prevent formation of a mucocele. The medial periorbital is dissected along the frontal-lacrimal suture approximately 2.4 cm to identify, isolate, coagulate, and cut the anterior ethmoid arteries to devascularize the tumor, which is supplied by the ethmoidal arteries [5].

The dura is opened a finger's breadth above the orbital rim on both sides, and the superior sagittal sinus is ligated and divided. The tumor is detached from the base in the midline until either its posterior edge or the limbus sphenoidale. Then the lateral attachments are taken down from lateral to medial taking care with the largest tumors to follow the medial sphenoid wing to the clinoid and identify the optic nerves, which will be displaced posteriorly and inferiorly. Tumor removal proceeds in a standard fashion.

When closing, the pericranial flap is laid down over the frontal sinus opening first, and then the supraorbital bone piece is secured in position. Beyond the frontal dural closure line, excess pericranium is removed to prevent congestion and the development of a mass lesion postoperatively. The two-part bifrontal bone flap is connected together with three plates and screws on the inner table side, and the “dog-bone” plates above the superior temporal line are recessed in the outer table to reduce their prominence and provide a better cosmetic result. Gaps in the bone above the muscle cuff can be filled with hydroxyapatite to prevent adherence of the galea to the fibrous union between the bone pieces and also improve the cosmetic result.

Discussion of Surgical Approaches

While we have detailed our recommended approaches for OGMs based on size, there are other surgical options to approach OGMs including a unilateral subfrontal, interhemispheric, and pterional craniotomy, each having their own advantages and disadvantages. Here, we review the merits and drawbacks of these other approaches in addition to postoperative management considerations.

Frontal approaches include both the subfrontal (also known as a transbasal) and interhemispheric approach. There are numerous terms that have been used previously for a transbasal approach, but according to Feiz-Erfan et al., a standard transbasal approach (in which osteotomies of the orbital rim, nasion, or nasal bone are not performed) involves accessing the tumor in an extradural manner and entirely resecting an olfactory groove meningioma intradurally [6]. Of note, if there is a significant intranasal component on preoperative imaging, this approach is often used in combination with endoscopic endonasal assistance, which requires reconstruction of the anterior cranial base.

A modification to the transbasal approach is to include osteotomies of the orbital bar, nasion, or nasal bone in order to approach the tumor from an extradural and slightly more inferior angle. When performing osteotomies of the orbital bar, the transbasal approach requires meticulous attention to the dissection of the periorbital, the supraorbital, and supratrochlear nerves and the insertion of the trochlea for the superior oblique muscle. Manipulation of these structures may lead to transient diplopia especially when the superior oblique muscle tendon insertion to the bone is interrupted. As detailed in our recommended approach, a modified supraorbital craniotomy can preserve structures within the orbit and still allow for adequate operative exposure.

It is often mentioned that one disadvantage with the transbasal approach is retraction-induced injury of the frontal lobes leading to frontal lobe syndromes. However, in our evaluation of patients undergoing an extended bifrontal craniotomy involving orbital osteotomies for midline anterior fossa meningiomas, we found that 91% of patients with no or minimal cerebral edema before surgery remained stable after surgery. Only 4 patients out of 42 included in the analysis experienced worsening edema, all of which had an initial tumor size of greater than 4 cm [7].

Another frontal approach for OGMs is the interhemispheric approach, first reported by Mayfrank et al. [8]. Like other frontal approaches, the patient is positioned supine with the head fixed in a Mayfield headrest. A unilateral craniotomy is performed (typically on the right side unless the tumor is eccentric to the left). A bone flap about 5 × 5 cm or less is made between the upper limit of the frontal sinus and the coronal suture, which is more posterior in location compared to the bifrontal craniotomy performed in a transbasal approach. A U-shaped dural incision is made and reflected medially. Dissection is performed along the falx until the superior pole of the tumor is encountered. Dural feeders are encountered during tumor removal toward the location of the dural attachment with large tumors or by following the anterior tumor surface toward midline. As described by Mayfrank and colleagues, at this point the meningioma should be split into two halves divided by a tumor-free, horseshoe-shaped area that includes the dura around the crista galli, above the cribriform plate, and anterior ethmoid. Next the bulk of the tumor is removed, leaving the capsule behind. As with other frontal approaches, the critical vascular structures and optic nerves and chiasm are encountered at the end of the resection. The surgical viewpoint is perpendicular to the floor of the anterior skull base making it relatively easy to visualize tumor invading into bone and the neurovascular structures at the posterior aspect of the tumor after debulking. Advantages of this approach are that the frontal sinuses are typically not entered, the superior sagittal sinus is preserved, and the risk of postoperative CSF rhinorrhea is reduced. Disadvantages include the need for frontal lobe retraction laterally as well as the possible need to sacrifice bridging veins while dissecting down the interhemispheric fissure, which may predispose the patient to venous infarction or postoperative edema.

A frontolateral approach, our preferred method for accessing smaller OGMs as previously described, offers a unilateral approach to these lesions. The dural attachments may be approached unilaterally with quick removal of dural feeders to the tumor, and the Sylvian fissure may be opened to drain CSF allowing spontaneous relaxation of the frontal lobe. Another benefit is preservation of olfactory sensation, which is more likely compared to bifrontal approaches [9]. A modification to the frontolateral approach includes the addition of an orbital osteotomy as we described above, which allows access to the base of OGMs, reducing the need for retraction. This versatile craniotomy allows access to lesions of the sphenoid wing, anterior and middle fossa floor, orbital apex, parasellar region, and basilar apex [10, 11]. The internal carotid and anterior cerebral arteries are typically accessible along with the optic nerves, allowing safe microdissection of the tumor away from these structures. The orbitozygomatic craniotomy is an expansion of the pterional craniotomy, which provides a wider viewing angle and greater working space over the superior aspect of the tumor with less brain retraction, which is why it is our preferred method for resection of smaller OGMs. Disadvantages include orbital swelling, which is common among patients undergoing this approach and may require temporary tarsorrhaphy to protect the cornea, which is done during positioning of the patient. If the meningioma is too large and cannot be debulked significantly, unilateral frontal lobe retraction may be required to access the most superior aspect of the tumor.

Another widely used surgical approach to OGMs is the pterional craniotomy which allows early visualization of neurovascular structures and control of the posterior aspect of a tumor in its relationship to the optic nerves, internal carotid and anterior cerebral arteries, and pituitary gland [12]. Again, a right-sided approach is chosen unless the tumor is eccentric to the left or involves critical structures on the left. Dissection of the tumor away from critical structures begins at the posterolateral and posterior aspects of the tumor, and much of the relevant vascular supply to the tumor can be coagulated from this unilateral angle. While drilling of tumor invading the floor of the anterior skull base is easier via a frontal approach, this can still be performed through a pterional approach. Typically, the most superior part of the tumor is encountered at the last stage of tumor removal, which may require extensive frontal lobe retraction. The pterional craniotomy spares the frontal sinus, reducing the risk of CSF rhinorrhea and postoperative meningitis, and only involves unilateral brain retraction. It also allows early access to the basal cistern for CSF drainage and brain relaxation, which provides additional working room between the brain and tumor without the need for a lumbar drain.

Many of these tumors involve the bone of the anterior cranial fossa, which requires reconstruction. An autologous pericranial flap, temporalis fascia, and fibrin glue can be used for reconstruction of the dural defect to prevent CSF leak. If larger defects are present, sometimes a split calvarial bone graft can be used in the repair. Typically, when the bone of the cribriform plate is removed during a bifrontal approach, we use a vascularized pericranial graft for repair. However, we do not remove the bone in this region during a unilateral approach.

Cosmesis should be considered when performing craniotomies around the forehead and above the eyes. Standard burr hole covers and dog-bone bridges are often used to hold the bone flap in place but are often visible through the skin, especially in patients with thin skin. We prefer to drill a recess for dog-bone bridges and cover the ridges of the bone flap with methyl methacrylate, which smooths the bone edges, improving cosmesis. Another important cosmetic consideration is to ensure that all scalp incisions are behind the hairline so that as time passes, a patient's hair may cover the scar.

Postoperative Management

Postoperatively, several aspects of patient care should be addressed. Frontal lobe syndrome may develop related to either edema or ischemia from prolonged brain retraction. This may require further treatment with steroids but, if severe, may require hypertonic saline or mannitol administration. Our standard postoperative care also includes seizure prophylaxis for 7 days, unless seizures occurred preoperatively in which case a more extended course of antiepileptics may be used [13]. We also keep patients well hydrated with intravenous fluids for 48 h postoperatively to prevent venous stasis and venous infarction. We also give 24 h of prophylactic antibiotics and select antibiotics that cover sinus organisms such as ceftriaxone. Finally, all patients are given pharmacological DVT prophylaxis at 48 h postoperatively. Additionally, some surgeons elect to leave in a lumbar drain for CSF diversion prior to any evidence of a CSF leak in order to avoid a CSF leak or pseudomeningocele formation; however, this is not our common practice. If a CSF leak occurs, a lumbar drain can be placed for 5–7 days and removed after a clamp trial and no evidence of persistent CSF leak.

Surgical Outcomes

Morbidity

The most common surgical complications reported after resection of OGMs are anosmia, seizures, CSF leak, hydrocephalus, and infection [14–16], in addition to medical complications. Pallini et al. examined outcomes in 99 patients undergoing bilateral subfrontal, frontotemporal, or bilateral subfrontal approach with bilateral orbital osteotomies. The most common complications were retraction-related brain swelling (14.2%), deep vein thrombosis/pulmonary embolism (8.0%), pneumonia (7.1%), and hydrocephalus (5.3%) [15]. Pires de Aguiar et al. reported outcomes in 21 patients who had undergone a bifrontal, pterional, or fronto-orbital approach. The most common complications observed were CSF leak (19%), hydrocephalus (9.5%, all required VPS placement), ventriculitis (4.8%), and postoperative hematoma formation (4.8%) [17]. Similar frequencies of postoperative complications have been observed by others [12, 18, 19]. Our group found that tumors displaying or encasing the ACA, or those with nasal sinus invasion, were much more likely to be associated with postoperative surgical complications. Interestingly, tumor volume was not associated with postoperative surgical complications [4].

Complication frequency varies by surgical approach. Pallini et al. found that the bilateral subfrontal approach with bilateral orbital osteotomies was associated with significantly less brain swelling but greater probability of CSF leakage requiring reoperation compared to bilateral subfrontal or pterional approaches (22.7% versus 15.7% and 9.2%, respectively) [15]. Nakamura et al. found that the most frequent complications for a frontolateral approach were postoperative subdural hygroma (17.6%) and seizures (11.8%), with less frequent hydrocephalus (5.8%) and hemorrhage (2.9%). The bifrontal approach was associated with more frequent hemorrhage (10.9%) and hydrocephalus (8.7%) and less frequent hygromas (2.2%) and seizures (4.3%) [18]. In contrast to these reports, we did not find any difference in complication rate between a more extensive extended bifrontal approach and a less invasive unilateral approach in a series of 44 patients with OGM/planum meningiomas [4]. However, no prospective studies have compared complication rates between these approaches.

Another frequent complication with olfactory groove meningiomas is anosmia. Loss of smell can greatly impact quality of life, especially taste. Preservation of olfactory function is important and a major concern for patients postoperatively. Jang et al. examined 40 patients with OGMs who underwent either a bifrontal or frontolateral approach. Anatomical and functional preservation of olfactory structures was achieved in 65% and 55% of cases, respectively. Olfactory sensation was more difficult to preserve in patients with tumors >4 cm and in patients with preoperative olfactory dysfunction. The authors found that the frontolateral approach was associated with improved preservation of olfactory function compared to the bifrontal approach (71.4% vs. 36.8%) [9]. Nanda et al. found that olfactory preservation was significantly better with lateral approaches (63.4% for lateral vs. 31.3% for bifrontal) [19].

Mortality

OGMs can cause extensive bilateral frontal lobe edema, especially when the tumor is large. This edema can be exacerbated by frontal lobe retraction during surgery, potentially leading to severe cerebral edema and death. Cushing and Eisenhardt's classic report on outcomes in olfactory groove meningiomas had a 22.7% operative mortality [3]. Others had similar outcomes in the decades to follow. Bakay et al. reported a 12% mortality rate in their series of bilateral or unilateral frontal approaches for OGMs between 1950 and 1970 [14]. Similarly, Solero et al. (1983) had a 17.3% mortality rate in 98 patients with olfactory groove meningiomas treated by a frontal craniotomy. The mortality was attributed to cerebral edema, clipping of a major artery, postoperative hematoma formation, meningitis, and other medical causes [2].

More recent patient cohorts demonstrate marked improvement in mortality, now ranging from 0% to 4.9% of patients. Nakamura et al. reported on 82 patients who underwent either a bifrontal or frontolateral approach. Perioperative mortality was reported for four patients (4.9%) of which three were related to postoperative cerebral edema after a bifrontal approach [18]. Aguiar et al. examined surgical outcomes in 21 patients treated with a bifrontal, pterional, or fronto-orbital approach. Perioperative mortality was 4.8% with one mortality secondary to postoperative cerebral edema worsened by hyponatremia [17]. Nanda et al. reported 0% mortality in 57 patients undergoing frontal or anterolateral approaches [19]. Overall, mortality with open approaches has improved markedly in the last several decades.

Mortality rates may differ between approaches. Nakamura et al. reported 8.7% mortality for a bifrontal approach versus 0% for the frontolateral approach [18]. Likewise, Pallini et al. found that the mortality rate was higher for bifrontal (5.7%) compared to lateral (0%) and fronto-orbito-basal (0%) approaches [15]. Bitter et al. had a 1.6% mortality rate with 1 death from a pulmonary embolism in 61 patients undergoing a pterional craniotomy for olfactory groove meningiomas [12]. Similarly, Tomasello et al. reported no deaths with a pterional approach for giant (³6 cm) OGMs [20].

Recurrence

Recurrence of olfactory groove meningiomas generally occurs at the cranial base and into the paranasal sinuses. Recurrence rates range from 0% to 41% depending on the length of follow-up and extent of initial tumor resection [21, 22]. Simpson Grade I or II resections tend to have fewer recurrences compared to higher Simpson Grade resections. It is unclear if the surgical approach influences the ability to achieve more extensive resection. Nakamura et al. reported a Simpson Grade I or II resection in 91.2% of patients undergoing a frontolateral approach and 93.5% of patients undergoing a bifrontal approach [18]. Others have failed to show a difference in extent of resection based on surgical approach [17]. In contrast, Pallini et al. reported more Simpson Grade I–II resections with a fronto-orbito-basal approach compared to the bifrontal and pterional approaches (100% versus 80.0% and 81.0%, respectively) [15]. The ability to achieve a Simpson Grade I or II resection depends on a combination of patient factors, the surgeon's comfort with the approach, as well as the actual tumor characteristics and suitability of the approach for a given tumor.

PEARLS
Brain relaxation can either be achieved via placement of a lumbar drain or by entering the basal cisterns in a lateral approach in order to drain CSF
Postoperative mortality is often due to the development of frontal lobe edema, so any efforts made to minimize retraction during the case can prevent this disastrous complication.
We recommend a unilateral cranio-orbital approach for lesions <3 cm and a two-part bifrontal craniotomy with tailored nasofrontal osteotomies for lesions >3 cm.
Lateral approaches may be associated with lower mortality rates, but these approaches have not been adequately compared to bifrontal approaches incorporating orbital osteotomies.
Lateral approaches may be associated with lower rates of anosmia.

Commentary on Case Presentation in Section 16.1

Case Presentation

A 54-year-old woman presents with primary complaints of 6 months of anosmia. The clinical examination reveals anosmia and no further neurologically deficits. (See Fig. 16.1a–f.)

Discussion

This tumor lesion appears to be less than 3 cm in size and is not significantly eccentric to either side. Additionally, there does not appear to be tumor within the ethmoid sinuses on these preoperative scans. We therefore recommend a right frontolateral cranio-orbital approach.

The tumor does not appear to be intimately related to critical neurovascular structures posteriorly and should be amenable to a Simpson Grade I resection. Of note, there does appear to be some degree of left frontal lobe edema, so the surgeon will be prepared for adherence between the surface of the tumor and the inferior medial left frontal lobe.

As detailed in the chapter, a right frontolateral craniotomy with a cranio-orbital osteotomy would be performed. We would use a coronal skin incision and elevate the pericranium separately to assist with coverage of any opening into the frontal sinus. The frontal craniotomy need not be higher than the superior aspect of the tumor given the thickness and irregularity of the inner table of the skull. During the approach, the medial roof of the orbit is removed as a separate piece so that visualization of the floor of the anterior fossa up toward the cribriform plate is improved. This is essential to allow drilling of the posterior aspect of the crista galli as well as to minimize brain retraction. This bone piece can be reconnected to the supraorbital osteotomy bone for reconstruction of normal orbital roof contours at closure.

After dural opening, the Sylvian fissure is opened to release CSF, and the right frontal lobe should be gently retracted. Vessels within the dura of the planum sphenoidale are coagulated when approaching the tumor. Once the superior edge of the tumor is reached by following the falx superiorly, an incision is made along the edge of the falx in an anterior and inferior direction until the crista galli is reached. A diamond drill is then used to remove the crista galli down to its base. Tumor debulking is performed centrally first followed by peripheral dissection. After the tumor is removed, further removal of the basal dura can be performed.

16.3 Olfactory Groove Meningiomas: Keyhole Craniotomy

Sascha Marx and Henry W. S. Schroeder

Introduction

Increasing technical capabilities and further understanding of surgical anatomy have facilitated the surgical treatment of various pathologies through smaller approaches. Larger craniotomies can be replaced by keyhole approaches without reducing visualization of the surgical target. The supraorbital approach utilizes the keyhole concept and was advocated by Axel Perneczky and colleagues [23–25]. Limitation with the angle of view can easily be compensated by endoscope assistance during the microsurgical procedure [24, 26].

There is no golden rule about the best surgical approach to olfactory groove meningiomas (OGM). Rather an individualized concept, tailored to each patient, should be done to achieve the best oncological and functional outcome. The present chapter discusses pros and cons of the supraorbital approach to OGM.

Surgical Technique

Preoperative Evaluation

Choosing the best approach to an OGM is a multifactorial matter. Anamnesis and clinical examination should be performed before reaching a decision regarding the proper approach. Significantly reduced mobility in the cervical spine (e.g., ankylosing spondylitis) is a contraindication for the supraorbital approach since proper positioning of the head is mandatory to achieve optimal surgical results. Smell should not only be evaluated by history but also by an objective olfactometric test. Since some large OGM may affect the optic nerve and chiasm, vision and visual acuity should be assessed by an ophthalmological examination.

Cosmesis is not the most important point in approach selection, but is increasingly important to patients. Many patients are well informed by the media and Internet and request small skin incisions and keyhole approaches. It is always an individual decision. The eyebrow incision is ideal for men with bald head and bushy eyebrows. On the other hand, if an Arabic woman is wearing a veil, the eye area can be the only part of her body visible in public places. It should be discussed very carefully whether or not the eyebrow skin incision is the best option in this particular situation. However, in our practice, most women choose the eyebrow incision to avoid shaving of hair (although it is very minor in standard approaches) and because of the smaller incision.

Of course, accurate evaluation of imaging is required before decision-making regarding surgery. MR imaging is the imaging modality of choice. We usually perform T1- and T2-weighted images with contrast in all three planes. The degree of contrast enhancement can give an idea about the vascularity of the tumor. Usually, perifocal edema indicates brain invasion by the tumor. Furthermore, MR imaging is necessary to identify intranasal extensions of the tumor and to locate the anterior cerebral artery complex in relation to the tumor (cave vessel encasement). Additionally, size and extension of the frontal sinus is evaluated. CT scans are not performed routinely before surgery, but the day after surgery to rule out complications.

Patient Positioning and OR Setup

One of the most important steps in performing a supraorbital approach is proper patient positioning (Fig. 16.9a–c). The body is elevated 10–15° in a supine patient position to reduce the venous cranial pressure. The head is fixed in a Mayfield clamp with rotation to the contralateral side and hyperextension of the neck in a way that the zygoma is the highest point.

Fig. 16.9 (**a**–**c**) Patient positioning: The patient is in supine position and the body 10–15° elevated (**a**). Sharp fixation of the head with rotation to the contralateral side and hyperextension of the neck (zygoma highest point) (**b**, **c**) (Copyright retained by Dr. Henry W. S. Schroeder. Used with permission)

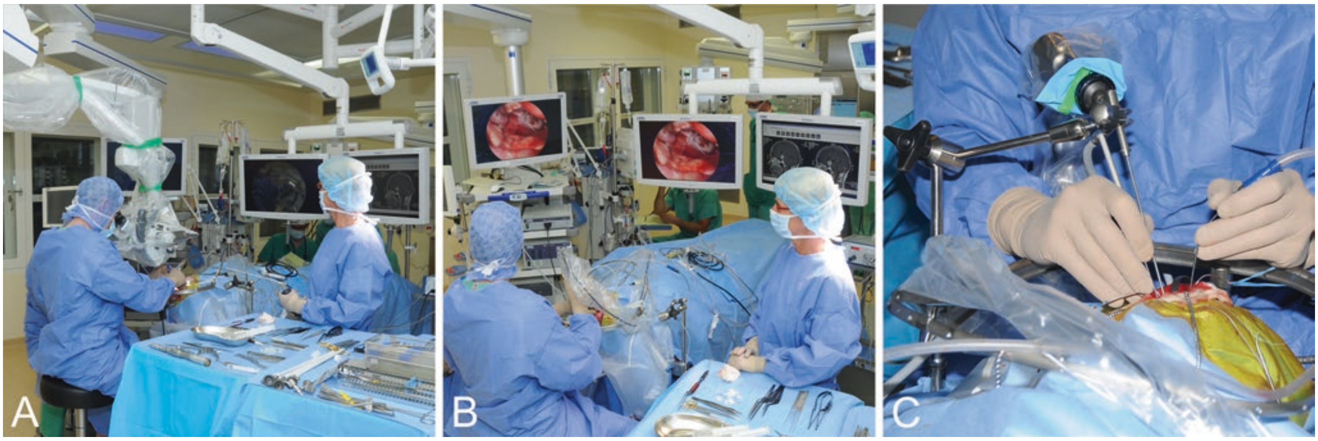

Fig. 16.10 OR setup for endoscope-assisted microsurgery (**a**, **b**). The endoscope is fixed with a mechanical holding arm during bimanual endoscopic dissection (**c**) (Copyright retained by Dr. Henry W. S. Schroeder. Used with permission)

Only minor additional retraction needs to be used in this position, since the frontal lobe falls back by gravity during surgery. We do not use perioperative lumbar drains. The degree of head rotation depends on the target zone but usually is 45° for OGM. After disinfection, the patient is draped as are the microscope and endoscope. The microscope stands behind the surgeon in order to bring it easily out of the surgical field when the endoscopes are used for visualization (Fig. 16.10a, b). Usually the endoscopes are used freehand for inspection but can be fixed to a mechanical holding arm when bimanual dissection and tumor removal are required (Fig. 16.10c).

Supraorbital Approach

"Supraorbital approach" describes the site of craniotomy but can be performed with different skin incisions such as incision in the eyebrow, the eyelid, or behind the hairline [27, 28]. We usually prefer the incision in the eyebrow for a supraorbital craniotomy and use a skin incision behind the hairline for traditional frontolateral or pterional craniotomies. An eyelid skin incision has the advantage of an almost invisible scar, but in our experience the cosmetic result of the eyebrow incision is excellent as well. Complications such as ptosis, diplopia, orbital trauma, and vision loss are described after eyelid skin incision [29]. In cases where an orbital osteotomy needs to be done, an eyelid skin incision would be beneficial [30]; however, this is a very rare situation in our experience.

To avoid injury to the supraorbital branch of the frontal nerve, the skin incision in the eyebrow starts laterally to the supraorbital fissure (Fig. 16.11a). The eyebrow is followed to its lateral end and sometimes a little bit longer (approximately 4 cm). Afterward the orbicularis oculi muscle and the fat tissue overlying the periosteum are incised. The supraorbital nerve can be identified at the medial end of the incision (Fig. 16.11b). The skin is retracted upward away from the orbit. The superior temporal line is identified which is the attachment of the temporal fascia. A periosteal flap based to the orbit is circumcised and elevated (Fig. 16.11c). The lateral periosteal incision follows the superior temporal line. The temporal fascia and the temporalis muscle are detached from the temporal line and adjacent bone and are retracted laterally with fish hooks (Fig. 16.11d). Care must be taken in this step that the tension on the hooks is not too high, otherwise the frontotemporal branch of the facial nerve can be violated. A small burr hole is placed behind the temporal line for cosmetic reasons (Fig. 16.11e). It should be exactly made just over the frontal skull base. If it is too far caudally, the orbit will be entered. Then, a 2.5 × 2 cm craniotomy is made with the aid of a craniotome as close to the skull base as feasible, sparing the frontal sinus whenever possible (Fig. 16.11f). The craniotomy should be approximately 2 cm in height (Fig. 16.11g). If it is too small, the microscopic visualization of the skull base may be compromised, and the margins of the craniotomy may interfere with the bimanual dissection as reported by other groups, too [31]. In select cases, the working space can be increased by the removal of the orbital rim [27] but is rarely necessary in our experience. After elevation of the bone flap, the dura is detached from the frontal skull base. The inner edge of the craniotomy and prominent protuberances of the orbital roof are drilled away to provide more space for visualization and dissection (Fig. 16.11h). To complete the approach, a curved dural incision based to the orbit is finally performed.

The frontobasal cortex is protected with a cotton patty, and the frontal lobe is carefully retracted from the skull base with a suction and anatomical forceps. After the olfactory tract and optic nerve have been identified, the optic cistern is opened and CSF is released abundantly to relax the brain (Fig. 16.12). Alternatively, the Sylvian fissure can be opened to drain CSF. If the head is positioned correctly, the frontal lobe falls back by gravity and provides space for microsurgical

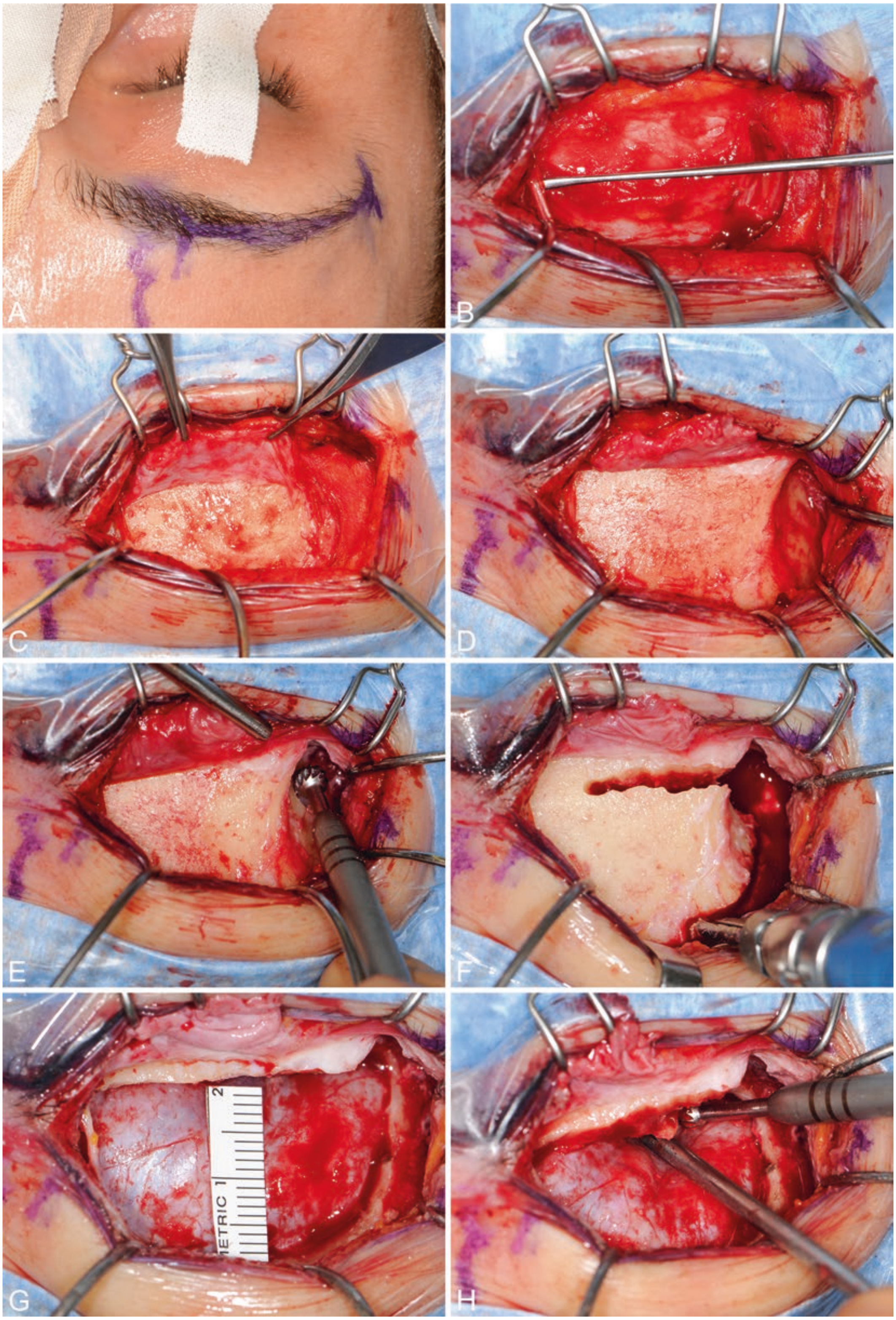

Fig. 16.11 (**a**–**h**) Supraorbital craniotomy via eyebrow skin incision: The skin incision including supraorbital notch are marked (**a**). The periosteum is exposed after skin incision and the supraorbital nerve is identified (**b**). A periosteal flap based to the orbit is exposed (**c**). Temporal muscle and temporal fascia are detached from the superior temporal line and adjacent bone and are retracted by fish hooks (**d**). Burr hole behind the temporal line (**e**). Craniotomy with the craniotome (**f**) and height of the craniotomy (**g**). The inner edge of the craniotomy and prominent protuberances of the orbital roof are drilled (**h**) (Copyright retained by Dr. Henry W. S. Schroeder. Used with permission)

manipulations (Fig. 16.13). Use of retractors is minimized whenever possible. Only to fix the patty which protects the frontal lobe, we temporarily use a retractor with mild retraction force. Excessive retraction of the frontal lobe has to be avoided because it can avulse the olfactory fibers at the cribriform plate.

Tumor resection itself follows microsurgical principles. At first, the anatomy around the tumor is identified, especially the relation of the tumor to the olfactory tract is explored (Fig. 16.14). Then, the tumor base is coagulated to interrupt the blood supply to the tumor. If the patient has preserved olfaction, the olfactory tracts are spared, at least on one side. After having the tumor devascularized, internal debulking is performed. When the tumor is soft, ultrasonic aspiration is used. When the tumor is very fibrous, microscissors are applied. Thereafter, we dissect the brain-tumor interface with two forceps using the traction-countertraction technique. Because of the limited height of the craniotomy, there is usually a problem to visualize the olfactory groove with the microscope, especially when the groove is very steep. Therefore, 30° or 45° endoscopes are mandatory in OGM surgery through a supraorbital craniotomy to inspect the olfactory groove and achieve gross total resection under full visualization (Fig. 16.15a–c). Angulated instruments are used to resect the tumor remnants under endoscopic view. If required,

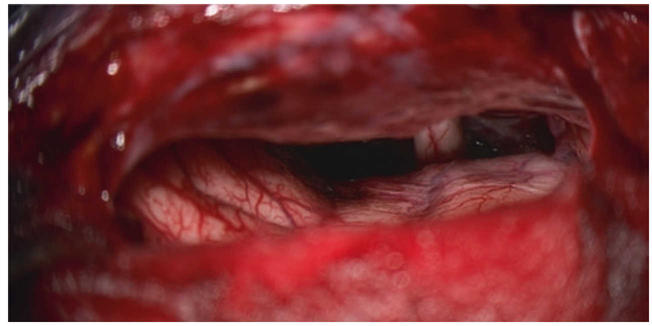

Fig. 16.13 Spontaneous "gravity" brain retraction by releasing CSF through opening of the basal cisterns and proper head positioning (Copyright retained by Dr. Henry W. S. Schroeder. Used with permission)

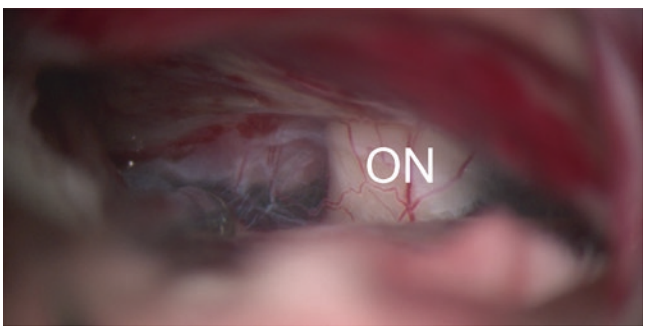

Fig. 16.12 Enlarging the working space by opening the basal cisterns. The frontal lobe is covered by a cotton patty. ON, optic nerve (Copyright retained by Dr. Henry W. S. Schroeder. Used with permission)

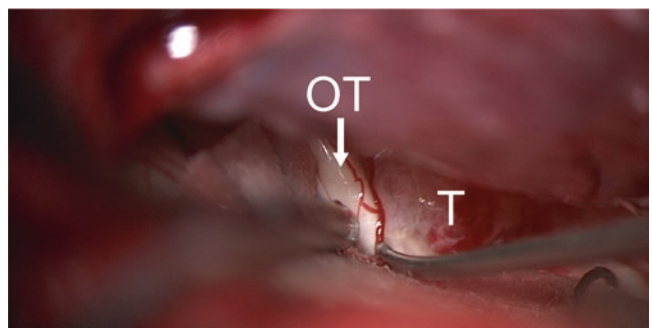

Fig. 16.14 Identification of the anatomy: Olfactory tract (OT) overlying the tumor (T) (Copyright retained by Dr. Henry W. S. Schroeder. Used with permission)

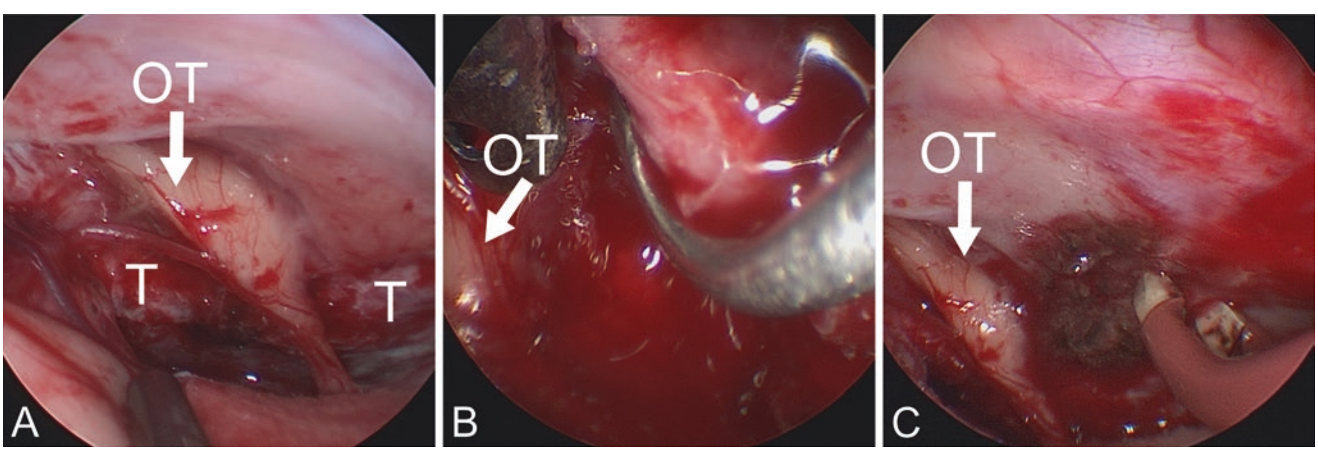

Fig. 16.15 (**a**–**c**) Endoscope-assisted technique. Due to the limited height of the supraorbital craniotomy, the olfactory groove with the olfactory tract (OT) can only be visualized with angled endoscopes (**a**). Resection of the tumor (T) is performed with angled instruments under endoscopic visualization with a 30° endoscope (**b**). Coagulation of the tumor base is performed (**c**) (Copyright retained by Dr. Henry W. S. Schroeder. Used with permission)

the infiltrated dura at the tumor base is resected and the underlying infiltrated skull base drilled. Even tumor in the ethmoidal cells can be resected from this approach. Skull base reconstruction is usually done with TachoSil® (Nycomed Austria GmbH, Linz, Austria) or DuraGen (Integra, Plainsboro, USA) and fibrin glue (Baxter, Vienna, Austria). Pericranium should not be harvested from the surgical field for cosmetic reasons. If it is required, an additional skin incision behind the hairline can be done, or autologous tissue from other sides (e.g., fascia lata) can be harvested.

After skull base reconstruction, the dura is closed with a running suture (Fig. 16.16a). To achieve watertight closure, TachoSil® can be placed on the suture line. This is especially helpful when the frontal sinus was opened during the craniotomy to avoid rhinorrhea. TachoSil® is additionally used to seal the opened frontal sinus. The bone flap is fixed with mini-plates after a piece of Gelfoam is placed in the craniotomy defect (Fig. 16.16b). For cosmetic reasons, the bone flap has to be in close contact to the upper margin of the craniotomy to avoid a visible indentation in the forehead later on. When the bony defect caused by the craniotome is at the lower aspect of the craniotomy hidden under the eyebrow, it usually remains invisible. The initial burr hole and the lower craniotomy defect should be filled with bone cement to avoid a visible indentation after scar formation, if the temporal muscle and scalp are very thin. The wound is closed in layers – periost, muscle, subcutis, and skin. The periosteal suture is important to cover the bony defect caused by the craniotome (Fig. 16.6c). The periosteum is sutured additionally to the temporal fascia. For skin closure, we use running resorbable or non-resorbable sutures (Fig. 16.16d).

Postoperative Care

The patient is observed overnight in the intensive care or intermediate care unit. The day after surgery, a CT scan is performed to rule out any complication and to check the

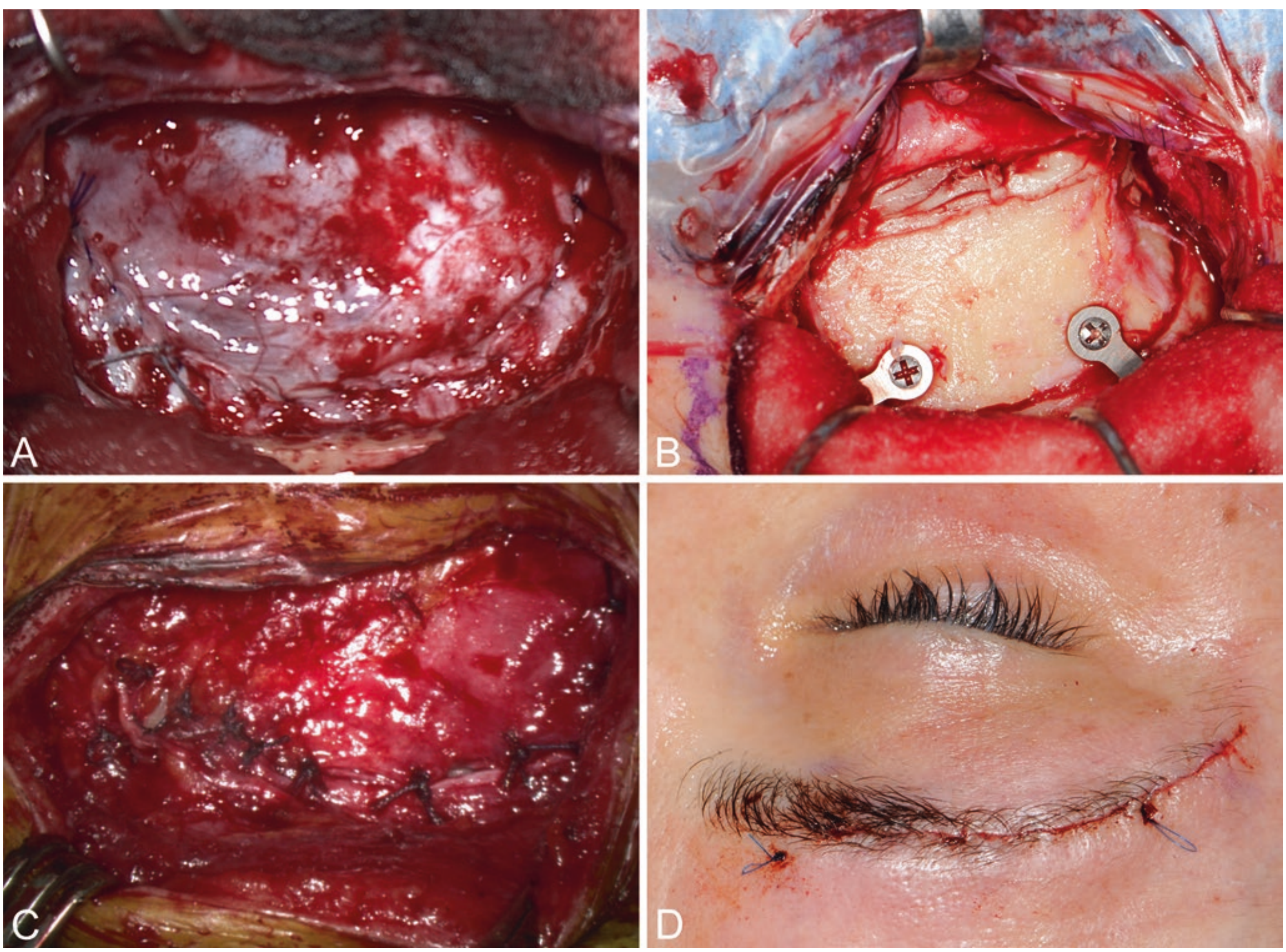

Fig. 16.16 (**a**-**d**) Finalizing the supraorbital approach: A watertight running dura suture is performed (**a**). The bone flap is re-fixated with mini-plates and is attached to the upper craniotomy limit (**b**). The periost is sutured separately (**c**). Skin closure intracutaneously (**d**) (Copyright retained by Dr. Henry W. S. Schroeder. Used with permission)

position of the bone flap (Fig. 16.17). A swollen eye might require ice for several days. Further special postoperative care is not necessary after a supraorbital approach which is a clear advantage compared to extended endonasal approaches. Three months after surgery, we perform an MRI with contrast-enhanced sequences to evaluate the degree of tumor resection.

Management and Avoidance of Complications

The overall complication rate of the supraorbital craniotomy itself is low. CSF leaks did occur in 2.6% in the largest series and were related to occult opening of the frontal sinus or paranasal sinuses [25]. Thus, closure of skull base defects is mandatory. Harvesting of autologous pericranium from the side of craniotomy is not recommended in supraorbital approaches for cosmetic reasons. Since the defect usually is small, we take collagen matrix or TachoSil® as overlay. If we face a larger defect, we do not hesitate to collect autologous tissue from other sites (e.g., fascia lata). Wound healing problems are reported in around 1% [25] and do not play a major role in supraorbital approaches in our experience (Fig. 16.18a–c).

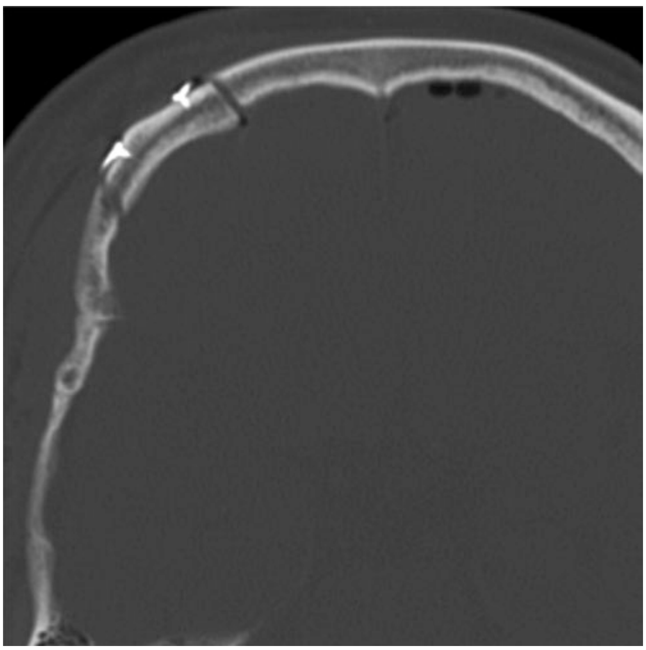

Fig. 16.17 Postoperative CT scan showing proper position of the re-fixated bone flap (Copyright retained by Dr. Henry W. S. Schroeder. Used with permission)

The risk of violating the supraorbital branch of the frontal nerve with resultant scalp hypesthesia is reported to be 7.5% in the largest series [25]. Interestingly, this complication is frequently emphasized when arguing against the supraorbital approach, whereas violation of the auriculotemporal branch of the mandibular nerve in a skin incision behind the hairline is frequently performed without being mentioned as major problem. In older descriptions of the supraorbital approach via eyebrow skin incision, sacrificing of the supraorbital branch was considered as standard, and end-to-end anastomosis was seen as an option for reconstruction [32]. In our experience, proper preoperative planning of the skin incision leads usually to preservation of the nerve. However, in case of an abnormal lateral course of the nerve, cutting of the nerve is required to gain access to the craniotomy site.

Violation of the frontotemporal branch of the facial nerve is described to happen in 5.5% in the largest series [25] but happened only once in our personal experience. Most violations are not sharp, but occur by firm traction of the lateral muscles by fish hooks. Thus, most palsies are transient with a recovery within several months. Nevertheless, the palsy in our experience was permanent. With a well-dosed retraction of the lateral muscles, facial nerve damage can mostly be avoided.

Patient Selection

The most important factor in approach selection is tumor size and location. Due to the nonspecific clinical complaints, OGM are frequently diagnosed with a larger size in comparison to planum and tuberculum sellae meningiomas [33].

Fig. 16.18 (**a–c**) Favorable cosmetic result 1 year after surgery (Copyright retained by Dr. Henry W. S. Schroeder. Used with permission)

For OGM smaller than 4 cm, we usually prefer the eyebrow approach, except if there is significant intranasal tumor extension. In tumors larger than 4 cm, we use a standard frontolateral approach via frontotemporal skin incision behind the hairline. The access to the tumor is straightforward, tumor resection is faster, and the drilling of the anterior skull base is easier with a higher craniotomy. Especially in larger tumors with extension to the tuberculum sellae and significant anterior cerebral artery complex encasement, the standard frontolateral approach gives plenty of space to deal with any difficulty. Additionally, the frontotemporal incision allows the harvesting of sufficient periosteum to reconstruct any large skull base defect. Although giant OGM can be resected via a supraorbital craniotomy [31], we usually do not approach these tumors via this route. This is in agreement with other publications [33, 34].

The lamina cribrosa is the weakest bony point in the anterior skull base and the origin of OGM at the same time. Thus, OGM frequently grow through the lamina cribrosa into the nasal cavity or paranasal sinuses [33]. That is the most common reason for failure of a gross total resection via a transcranial approach [34]. Under these circumstances, either a purely endonasal approach or a combined endonasal and transcranial approach is recommended [36].

When approaching OGM via the eyebrow supraorbital craniotomy, the olfactory groove needs to be evaluated carefully. The depth of the olfactory groove varies remarkably in anatomical studies [33]. Mostly, the olfactory groove cannot be inspected with the operating microscope because of the low height of the craniotomy. Therefore, endoscopes with angulated view are required. We use 30° or 45° endoscopes for inspection and tumor removal [37].

Another important factor in choosing the optimal approach in OGM surgery is the preoperative status of olfaction. When the sense of smell is still good, our intention is to preserve the olfaction, even when there is an infiltration of the cribriform plate. At least one olfactory tract should be preserved. If olfactory fibers are involved on both sides, we leave a small tumor remnant behind. However, smaller tumors can frequently be removed totally with bilateral preservation of the olfactory tracts and nerves. In larger tumors with bilateral infiltration of the cribriform plate, patients have mostly no useful olfaction, and the olfactory tracts and nerves can be sacrificed.

Surgical Outcomes

The literature is lacking larger series of OGM approached via a supraorbital craniotomy. Rather, OGM are included in series of the supraorbital approach for various pathologies [25, 31, 33, 34, 38]. One recent study compared the outcome of OGM resected via a supraorbital craniotomy compared to both endonasal and combined (supraorbital and endonasal) approaches [36]. The supraorbital approach, indeed, is not standard in OGM surgery. In the largest series of OGM, reporting on 99 patients, not a single tumor was approached via this route [15]. In a series reported by Romani et al., 66 OGM have been treated with a modification of the supraorbital approach with a frontotemporal skin incision [35].

Extent of Resection, Recurrence, and Complications

Independent of the approach, the goal of OGM surgery must be gross total resection, and in parallel to this, the recurrence rate must be low [31]. Only in the above drafted scenario, if olfaction needs to be preserved, subtotal resection is acceptable. The literature shows a gross total resection (Simpson grade I or II) in 90–100% [31, 34, 35, 38]. Interestingly, Banu et al. have shown superior extent of resection in supraorbital approaches compared to endonasal approaches (100% versus 87.3%) [36]. The overall complication rate after supraorbital approaches is low and reflects the above discussed issues. Banu et al. have shown a lower complication rate in the supraorbital approach compared to the endonasal approach to OGM, even when loss of olfaction has been included, which is inevitably lost with an endonasal approach [36]. The supraorbital approach is associated with excellent cosmetic results, and the scar is usually hardly visible after 3 months [31, 38]. Of course, CSF leakage rates in extended endonasal approaches exceed rates from transcranial approaches. However, repair techniques still improve in endonasal surgery, and with it the leakage rates will decrease [27].

Preservation of Olfaction

Loss of olfaction is often an insidious process and barely noticed by the patient. Only 14.3% of patients complained of loss of olfaction in a series of Bassiouni et al., but olfactory testing revealed anosmia in 71.7% of the same patients at the same time point [33]. The largest series of OGM showed approximately 60% of patients had preoperative anosmia [15]. Older series rarely refer to functional outcome measures such as pre- and postoperative olfactory function [38]. Since most of the series are of mixed pathologies, the percentage of pre- and postoperative loss of olfaction is not reported separately for OGM [31]. A clear advantage of all transcranial approaches over the endonasal approach is the possibility to preserve olfaction if it is intact preoperatively [36, 39]. Rare reports show an improved olfaction after supraorbital approaches even in patients who have been anosmic prior to surgery [34]. However, if patients are anosmic preoperatively, they usually remain in this state after surgery, even if the olfactory nerve is kept intact [36].

PEARLS
Gross total resection of small- to medium-sized olfactory groove meningiomas is possible via a supraorbital approach with endoscope assistance.
The supraorbital approach via an eyebrow skin incision provides a low rate of complications and favorable cosmesis.
Preservation of olfaction is possible with a supraorbital approach but sometimes needs to be compromised with a waiver of gross total resection.

Commentary on Case Presentation in Section 16.1

Case Presentation

A 54-year-old woman presents with primary complaints of 6 months of anosmia. The clinical examination reveals anosmia and no further neurological deficits. (See Fig. 16.1a–f.)

Discussion

The present case is an ideal candidate for a supraorbital eyebrow approach from the right side. The tumor is midline and small. The olfactory groove is not deep. There are no tumor extensions into the nose. The frontal sinus is small. Since olfaction is already lost, the basal dura and the olfactory bulb can be resected and the skull base drilled. With endoscope assistance a gross total resection is possible. The right side should be chosen if it is the non-eloquent side and if the surgeon is right handed.

16.4 Olfactory Groove Meningiomas: Endoscopic Endonasal Approach

Verena Gellner and Peter Valentin Tomazic

Introduction

There are three main parts of extended endoscopic skull base approaches for olfactory groove meningiomas: the transethmoidal, transcribriform, and transplanum. The approach can be optimally tailored to the tumor size and surgical situation, with the opening as large as necessary but as minimally invasive as possible.

The choice of an endoscopic transnasal or a transcranial approach has been discussed extensively in recent years. Each has its advantages and disadvantages, with optimal results depending on careful patient selection [40–43].

A successful result will owe a great deal to detailed planning and a dedicated surgical team, under the assumption that the hospital has all the necessary infrastructure and resources at its disposal.

Preoperative Considerations/Measures

Preoperative radiological studies should include MRI (T1 and T2 with and without contrast), CT scans if there is bone involvement, and MRA and/or CTA if vascular encasement is present. The ACA (anterior cerebral arteries) and the A2 segment and its frontopolar branches are most likely to be involved. The main vessels supplying the tumor are usually the ethmoidal arteries, which are well accessible via EEA.

Operative Setup and Technique

The tumor should be devascularized early to minimize blood loss. Intraoperative navigation is mandatory, ideally with CT and MR fusion technique. Intraoperative ultrasound aids orientation by detecting vascular structures.

Surgical Technique

Generally, the surgical goal of OGM surgery should be a Simpson Grade 1 gross total resection [44, 45]. The EEA provides a perfect approach for total resection including infiltrated dura and bone. An extensive tumor, involvement of vital structures, patient's age, and/or poor general condition might call for a staged procedure or require other treatment strategies like surgical volume reduction followed by radiation therapy. The endoscopic transethmoidal, transcribriform, and transplanum approaches for larger tumors have been described in detail in the literature [46–55]. Whether nasal structures will be preserved or sacrificed will depend on intranasal tumor infiltration. For strictly intracranial tumors, a standardized sphenoethmoidectomy is performed to identify important landmarks (internal carotid artery, optic nerve) and to gain space. This can be done without resecting the turbinates. After thorough topical decongestion (adrenaline 1:1000), the inferior turbinate is not an obstacle. The middle turbinates can be deflected laterally unless it has to be resected due to tumor infiltration, particularly if the ethmoid is opened. In the postoperative course in our large series of extended skull base cases, all middle turbinates were in orthograde position and stable since their attachment and their basal lamellas were not resected. The maxillary sinus ostium should be identified and opened, but a medial maxillectomy is generally not required. For optimum exposure of the anterior skull base and olfactory cleft, a Draf III procedure is most suitable. Here the frontal sinus floor is drilled away after the frontal sinus outflow tract has been identified. Posterior bone is removed up to the first olfactory fiber (or the anterior superior nasal artery), and the frontal sinus openings are then connected, forming an anterior septectomy. The septum can be resected posteriorly as far as the tumor extends. Once the tumor perimeters can be reached, the skull base can be opened, and the dura devascularized and incised. The tumor removal should be performed strictly following the general microneurosurgical principles of meningioma surgery [56]. Depending on tumor consistency, additional instruments like CUSA® (Cavitron Ultrasonic Surgical Aspirator, Valleylab, Boulder, CO, USA) and/or Sonopet (Sonopet® Ultrasonic Aspirator, Stryker® Instruments, USA) may be very helpful to reduce tumor volume. If there is intranasal infiltration, the turbinates will have to be sacrificed to provide free margins.

Depending on tumor extension and bone infiltration, the skull base defect must be adequate for optimal tumor removal. If additional lateral space is needed, the lamina papyracea can be resected for better access to the tumor's lateral surface. Provision must be made for enough space for microsurgical dissection of the tumor capsule, as well as for handling any vascular complications such as intraoperative bleeding.

Nuances of Skull Base Reconstruction

The most common surgical complication of the EEA is postoperative CSF (cerebrospinal fluid) leakage [57]. Secure closure of the dural defect should be planned preoperatively. Dural and/or osseous defects extending anteriorly to the posterior wall of the frontal sinus are especially challenging. Angled instruments can solve the problem of the working angle. The application of the well-established nasoseptal flaps [58–60] might not be possible or require adaptation because of the long distance between the vascular flap stalk and the anterior border of the defect.

The nasal septum has to be partially resected to access the anterior skull base. Youssef [61] and Rosen [62] describe a technique to transpose the septum, which remains attached to the mucoperichondrium. The septum has to be deflected laterally when the skull base is opened for manipulations but later can be repositioned into the midline. For more anterior tumors where a septal transposition would not be possible, a Draf III opening would best expose the skull base. The same is true if the tumor has infiltrated the septum.

There is no definite consensus about the optimal grafting material, but there seems to be a tendency toward autologous material, with a preference for fascia lata [63]. Its advantages are the abundance of material available for harvest, good cosmetic outcomes, and biological features similar to the dura, as well as avoidance of immunological reactions with allograft material. For smaller defects (<3 cm in diameter) turbinate grafts can be an option. The middle turbinate must be sacrificed, which is understandably controversial with respect to postoperative quality of life (QoL) and nasal function [64]. If a concha bullosa is present, this shortcoming can be avoided as the lateral part of the turbinate can be resected without impairing turbinate function. Sometimes it is also possible to denude the mucosa on the septal side of the concha and create a flap from the medial portion of the concha bullosa, allowing the flap to be swung over the defect with the advantage of maintaining its blood supply. The grafts can be fixed with resorbable material (TachoSil® [Baxter HealthCare, Deerfield, IL, USA] or Oxicel® [Betatech Medical, Istanbul, Turkey]).

For large defects, however, nasoseptal flaps as originally described by Hadad [58, 65], and now with many variations, are the primary workhorse. The complete septal mucosa (and perichondrium/periosteum) can be harvested, whereby the stalk is supplied by the sphenopalatine artery. Since the whole septum is denuded, nasal function and QoL after surgery may be negatively impacted for 2–3 months [66, 67]; further, if the tumor has infiltrated the septum, a Hadad flap is not an option. In these cases a more extensive fascia graft has to be harvested with enough area of overlay in the skull base over the resection margins.

Postoperative packing in olfactory groove meningiomas has to be handled very carefully to avoid prolapse of packing material intracranially and to only gently support the grafting material and keep it in the desired position. Contrary to selected sphenoid closures where only an overlay is possible and hence the packing serves also as a abutment, in OGM the underlay graft serves as an abutment and is stabilized against the skull base by the intracranial pressure. If intranasal packing would be applied too forcefully in this location, the underlay would be lifted off the bone and consequently washed away by CSF resulting in a leak.

Lumbar Drainage (LD)/Fluorescein

We have found no clear evidence or advice in literature about the application of a lumbar drain (LD) pre-, intra-, or postoperatively. Some surgeons place a LD before the procedure to lower intracranial pressure (ICP) and control the flow of CSF during closure. It is possible to leave the drain in place for some days postoperatively to facilitate the healing process and stabilize the closure. Careful perioperative handling of the drain is essential to avoid serious complications like overdrainage or infection. We believe that fluorescein is not necessarily required intraoperatively in EEA of OGM because the location of CSF leakage is clear, and it is not essential in every single case for reconstruction and confirmation of watertight closure. The leak can be identified easily, especially in OGM surgery, due to the good exposure of the operative site. Fluorescein may be helpful for postoperative leaks, especially small ones. In their review, Psaltis et al. [68] report 28 studies that used intrathecal fluorescein intraoperatively to facilitate the identification of the CSF leak site. The dosages ranged from 0.1 mL of 10% to 0.5 mL of 5% fluorescein, usually diluted in 10 mL of CSF or sterile water. As a general rule, 5% sodium fluorescein should be used at a dose of 0.01 ml/KG body weight but never more than 1 mL. Only two studies reported complications: one case of generalized seizures and one of premature ventricular ectopic heart beats.

Postoperative Care

All patients are given very strict recommendations of postoperative activity. Any kind of intracranial pressure elevation such as sneezing, coughing, or pressing should be avoided by all means. Bed rest is recommended for 3–5 days, and stool softeners may be provided if needed. The patient's understanding of the process and full cooperation are important factors for successful recovery especially after this kind of surgery.

Patient Selection

The EEA can provide the perfect approach for olfactory groove meningiomas as the anatomic location and tumor configuration allow early devascularization and complete removal of the tumor's origin. The ideal anatomical constellation for an EEA would be a tumor that does not extend laterally beyond the midline of the orbit and dorsally beyond the orbital apex. There should not be frontal sinus involvement, but an extensive nasal tumor could be an additional indication for this kind of approach. An important factor in patient selection is the lateral and anterior dural involvement and accessibility of the entire tumor. Nonetheless, tumor size alone is not necessarily a factor influencing the decision for or against the EEA. Especially in huge meningiomas with perifocal brain edema, the endonasal approach without any brain retraction might be advantageous. Even vascular encasement is not an absolute contraindication for an EEA, depending on the surgeon's experience and skills. In any case, the surgeon must be thoroughly familiar with the anatomical landmarks and the position of vital structures in the area of interest.

As to the clinical outcome, preoperative nasal impairment and loss of smell seem to be perfect indications for an EEA. If the patient has normal olfactory function and the lesion is unilateral, there is a higher risk of postoperative deterioration, depending on tumor size, tissue consistency, and the surgeon's experience regardless of approaches.

Since the procedure entails opening and then securely closing the dura, nonetheless with the risk of postoperative CSF leakage, only patients in good general condition who are capable of compliance can be seen as candidates for an EEA; cognitive impairment and/or inability to follow the postoperative instructions should be mentioned as relative contraindications for EEA. In these cases a transcranial approach (e.g., eyebrow approach) might be the better option because tight closure of the dura and a fixed reinsertion of the bone flap can be performed.

Surgical Limitations

With better instrumentation and general experience in skull base surgery, surgeons have extended the limits of the EEA over the last decade, e.g., with angled instruments to reach tumor remnants at the anterior and/or lateral borders. Tumor extension laterally beyond the orbital apex and/or the midline of orbit and/or extensive frontal sinus involvement should be considered as a limitation for the EEA.

Contraindications for EEA

Acute nasal, oropharyngeal, or sinus infections are clear contraindications for EEA; antibiotics should be administered and the procedure scheduled after full mucosal recovery. Elderly patients, children, and substance abusers may not be optimal candidates for EEA, as it is of utmost importance that the patient understands and follows the doctor's instructions to avoid elevation of intracranial pressure in the immediate postoperative stage. Postoperative sedation and intubation are not recommended due to the potential side effects and risks involved.

Surgical Outcomes

Generally, it has to be mentioned that the published series are hard to compare because of their lack of uniformity, particularly as to group size and surgeon experience. Even the most experienced skull base centers report a significant learning curve over the past decade. Outcomes as related to the extent of resection, postoperative rate of CSF leak, and morbidity are barely comparable and have to be interpreted with caution.

With regard to the extent of tumor resection, results of as much as 91.7% gross total resection have been reported recently, as most experienced surgeons divide their results into an early and a later learning phase [40, 69].

As to functional outcome, postoperative anosmia is a crucial issue for patients with normal preoperative olfaction. If the tumor infiltrates the cribriform plate and complete resection of the mucosa, bone, and dura is necessary to remove the entire tumor, the procedure inevitably costs patients their sense of smell. Accordingly, for tumors with unilateral extension and preoperative intact olfaction, the supraorbital eyebrow approach might be associated with less morbidity in this regard. The majority of studies addressing EEA do not identify significant postoperative anosmia, but they are mostly related to pituitary surgery. In OGM, a completely different region is affected, and usually the olfactory fibers must be sacrificed during surgery. No study to date has investigated nasal breathing after EEA; Gellner et al. [70] did not find a significant deterioration of nasal breathing after pituitary surgery. The shortcoming of this study is that it only investigated standard endoscopic pituitary surgery in a small case series using VAS (visual analog scale) and no objective measures. Since the approach to OGM differs from transsphenoidal approaches, studies analyzing postoperative nasal patency in that particular approach would be desirable. As regards mucosal function, Alobid et al. published the only study to date [71] describing a decreased saccharin transit time in expanded skull base approaches.

As mentioned above, large dural defects especially showed a relatively high postoperative rate of CSF leakage. The literature shows that this rate has been remarkably improved and reduced to <5% since the establishment of the mucoperiosteal flaps [40, 69].

PEARLS
The EEA for OGM is a skull base approach option only for experienced endoscopic skull base surgeons with appropriate institutional infrastructure.
The endoscopic transnasal technique is a midline approach that provides optimal visualization of OGM, allowing early devascularization and targeted tumor removal.
Careful patient selection based on tumor extension, anatomic conditions, and functional needs is the key for successful EEA for OGM.
Dural closure has to be carefully planned and tailored to the size of the defect.
Since the defect at anterior skull base is flat, grafts can be positioned more efficiently as opposed to more posteriorly located tuberculum sellae meningiomas because you do not have to work around corners.

Commentary on Case Presentation in Section 16.1

Case Presentation

A 54-year-old woman presents with primary complaints of 6 months of anosmia. The clinical examination reveals anosmia and no further neurologically deficits (see Fig. 16.1a–f).

Discussion

Due to the lesion's rather dorsal midline location (no frontal sinus involvement), with moderate lateral tumor extension, absence of vascular encasement or detectable brain edema, and given that the patient suffers from anosmia, the endoscopic endonasal approach (EEA) should be chosen.

Since the nose appears to be quite narrow with a straight septum, the first step would be a sphenoethmoidectomy followed by maximum lateralization of the middle turbinate if possible. A septum tunnel should be created on both sides to prepare to harvest a Hadad flap on one side, which can be parked in the nasopharynx. Mobilization and transpositioning of the nasal septum should be attempted. If this does not work, the median/posterior superior part should be resected, exposing the skull base to the planum sphenoidale. The bone, especially at the edges, should be smoothened with a diamond drill to facilitate later positioning of fascia lata and the nasoseptal flap. The goal of tumor resection should be complete tumor removal (Simpson Grade 1) including the infiltrated dura, with microsurgical dissection technique to a good neurosurgical standard.

The objective should be meticulous skull base reconstruction, e.g., with fascia underlay and Hadad flap or any other technique as mentioned.

16.5 Olfactory Groove Meningiomas: Editors' Commentary

The case provided in Sect. 16.1 (Fig. 16.1a–f) describes a fairly typical situation of a middle-aged patient presenting with anosmia with imaging evidence of an olfactory groove meningioma. A multitude of surgical approaches for the management of these types of tumors have been described ranging from traditional transcranial approaches with or without skull base extensions, keyhole approaches through either a supraorbital or mini-pterional corridor, and extended endonasal approaches. Each of these approaches carries its own advantages and disadvantages, and many of the purported advantages have not been clearly demonstrated in an evidence-based manner. The goals of any surgical approach are to maximize the extent of tumor resection in order to prevent recurrence and to minimize the likelihood of complications. In order to achieve these goals, the surgical approach should be tailored to the individual patient based on a variety of factors including the patient's clinical presentation, individual anatomy, and the intrinsic characteristics of the tumor. In this patient, anosmia is present and the goals of surgery do not include preservation of the olfactory nerves, making a bifrontal craniotomy or endoscopic endonasal approach potentially appropriate. Similarly, the patient's age suggests that they would well tolerate a transcranial approach. In our personal experience, elderly patients do poorly with traditional craniotomy and any frontal lobe manipulation, while relatively better tolerating the extended endonasal approach. Important anatomic factors to consider include the size and extension of the tumor both laterally and anteriorly. In this case, the tumor does not extend significantly beyond the medial aspects of the orbits which would represent a contraindication to the endonasal approach, but the tumor does extend far anteriorly up to the posterior aspect of the frontal sinus. While this anterior extension does not preclude successful resection of the tumor, reconstruction of the bony and dural defects is more challenging with the endoscopic approach where the nasoseptal flap will need to be maximally extended. Additionally, the size and lateral extension of the frontal sinus should be considered. In this case, the frontal sinus is small and would not be expected to be breached via a supraorbital keyhole approach, potentially decreasing the likelihood of CSF rhinorrhea, infection, and delayed mucocele formation. Factors intrinsic to the tumor to be considered include the presence of extensive anterior cranial fossa hyperostosis and intranasal tumor extension, heavy calcifications within the tumor, significant frontal lobe edema, and vascular encasement by the tumor. On sagittal imaging, this tumor does not appear to encase the anterior cerebral arteries nor extend inferiorly into the ethmoid or sphenoid sinuses. There are no heavy calcifications and the associated cerebral edema is minimal.

Based on the presenting symptoms and imaging characteristics for this particular patient, successful resection of this olfactory groove meningioma can be performed via a number of surgical approaches. The primary advantage offered by the endonasal endoscopic approach is avoidance of brain manipulation, potentially resulting in decreased frontal lobe injury. The most dreaded complication of surgical treatment of olfactory groove meningiomas is development of malignant cerebral edema postoperatively, likely related to frontal lobe manipulation and/or venous compromise. Even mild manipulation may result in worsened neurocognition. Direct comparison of endonasal and transcranial approaches in terms of neurocognitive outcome has not been well studied. Using changes in FLAIR volume on MRI as a surrogate for cerebral edema and frontal lobe injury, however, de Almeida et al. suggested the endoscopic approach may result in less frontal lobe injury after controlling for selection bias in terms of tumor size [72]. However, this study also demonstrated that the likelihood of gross total tumor resection and incidence of complications, including cerebrospinal fluid leakage, were higher in the endoscopic compared to the transcranial cohort, although neither of these findings reached statistical significance. A meta-analysis by Komotar et al. comparing endoscopic endonasal to transcranial approaches, however, reached similar findings in terms of extent of resection and complications with the endoscopic approach performing inferiorly in both regards [73].

Transcranial approaches for olfactory groove meningiomas range from the bifrontal craniotomy with or without orbital osteotomies, the interhemispheric approach, and the unilateral approaches performed through either a subfrontal or pterional corridor. The bifrontal craniotomy is often proposed as the optimal treatment for large olfactory groove meningiomas as it provides bilateral access to the frontal lobes and wide exposure of the anterior skull base for vascular control. However, this approach may result in bilateral frontal lobe retraction and injury as well as a high likelihood of bilateral disruption of the olfactory nerves, an important consideration in patients presenting with preserved olfaction. Additionally, interruption of the anterior aspect of the sagittal sinus carries a low but non-negligible risk of venous infarction and may contribute to the incidence of malignant postoperative cerebral edema associated with the bifrontal approach. In order to achieve a more inferior trajectory and avoid frontal lobe manipulation, cranialization of the frontal sinus and the addition of orbital osteotomies are often performed. While this low trajectory allows for excellent access to the tumor, reconstruction of the frontal sinus must be performed with a pericranial vascularized graft and carries the risks of infection and mucocele formation. A gradual evolution away from the bifrontal toward unilateral approaches has occurred over the last several decades, even for accessing large olfactory groove meningiomas. The unilateral approach provides quick and easy access to the tumor and obviates the need for frontal sinus disruption and reconstruction. These approaches have the disadvantage of making radical resection of tumor invading the paranasal sinuses more difficult. The keyhole supraorbital and mini-pterional approaches are the natural evolution of this trend toward unilateral access. The supraorbital approach can be performed through either an eyebrow or eyelid incision. Orbital osteotomy can be added if a more superior trajectory is necessary for larger tumors, although a well-placed craniotomy along the orbital roof with drilling of the bony excrescence of the roof makes this extension unnecessary in the majority of cases without significant superior extension. The most challenging area to access via the keyhole approach is the depressed site of tumor origin within the midline olfactory groove, as demonstrated on the coronal MRI and CT images for the case in discussion. The surgical line of sight along the orbital roof and restricted working corridor created by the small craniotomy makes tumor resection under direct visualization challenging. However, as this represents a relatively safe area for resection free of important vasculature and neural structures, the use of angled endoscopes and instrumentation enables successful removal of tumor from this location relatively easily. The unilateral approach also presents the opportunity for preservation of at least one olfactory nerve, an important consideration in patients with preserved olfactory function as disruption of taste can be associated with significant impairment in quality of life. Importantly, while the keyhole approaches have the potential for less disruption of soft tissues, potentially resulting in improved cosmesis and quicker recovery, these benefits have not been clearly established in the literature and remain to be further explored.

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Planum/Tuberculum Sella Meningiomas

17

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Abbreviations

ACA	Anterior cerebral artery
CSF	Cerebrospinal fluid
CT	Computed tomography
EOR	Extent of resection
ICA	Internal carotid artery
MRI	Magnetic resonance imaging
PS	Planum sphenoidale
TS	Tuberculum sella

17.1 Planum/Tuberculum Sella Meningiomas: Editors' Introduction

Meningiomas are common extra-axial brain tumors that arise from the arachnoid cap cells. Suprasellar meningiomas account for about 10% of all meningiomas [1]. The two most common suprasellar locations are the tuberculum sella and planum sphenoidale, and they may extend to the diaphragma sella. These tumors can grow to displace the optic nerves, chiasm, and pituitary infundibulum. They may also partially or fully encase the internal carotid arteries. Invasion into the optic canals is quite frequent and can complicate the complete removal of these tumors.

Patients that present with visual complaints typically require surgical intervention for decompression of the optic apparatus. Various surgical approaches have been described, ranging from a bifrontal or unilateral craniotomy to more recently keyhole supraorbital craniotomy and endonasal endoscopic techniques. Indications for each approach remain highly variable. Early optic canal decompression is important for improvement of visual outcomes and is often necessary to achieve a gross total resection. In this chapter, we discuss the traditional "open" approaches as well as the minimally invasive surgical approaches within the context of the case below (Fig. 17.1a–d).

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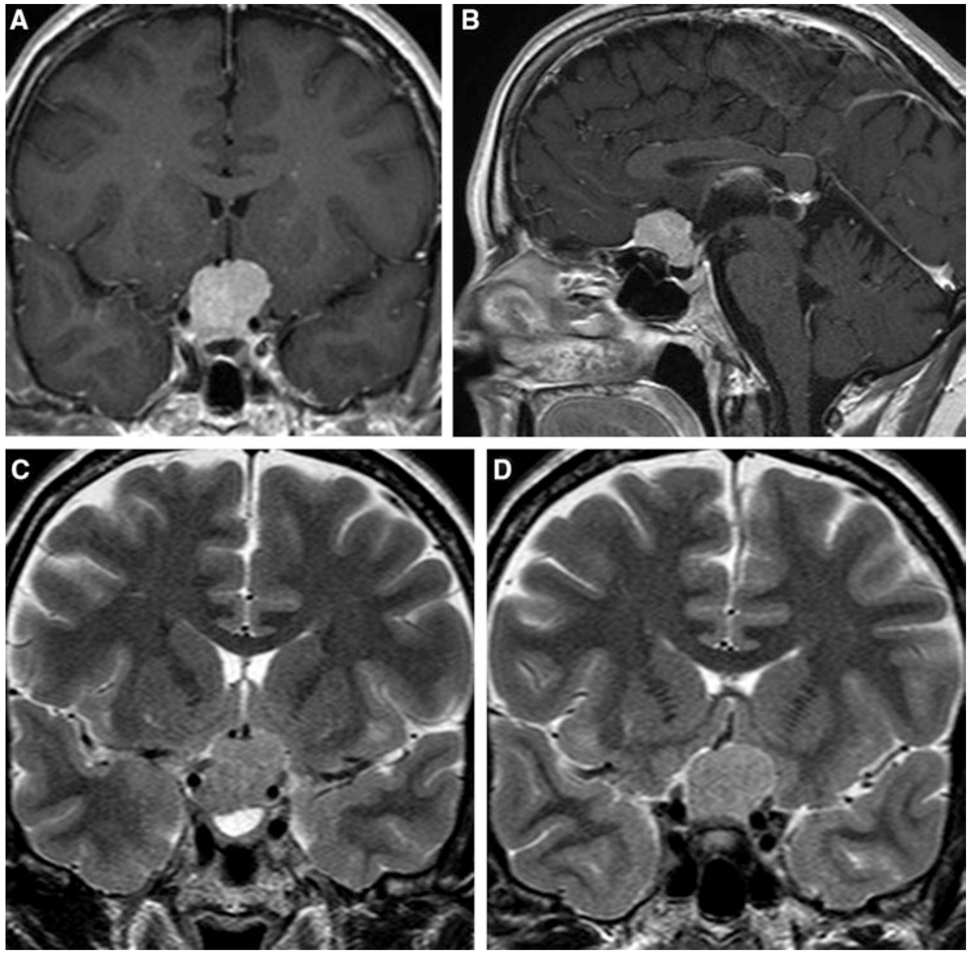

Fig. 17.1 **(a–d) CASE EXAMPLE**: planum/tuberculum sella meningioma. A 52-year-old lady presents with several months of left eye blurriness. Examination: OS, 20/30 (corrected), relative APD, 3/11 color plates; Humphrey VF, superotemporal arcuate field cut; OD, normal. The rest of neurological exam is normal

17.2 Planum/Tuberculum Sella Meningiomas: Open Approaches

Anoop P. Patel, Rakshith Shetty, and Laligam N. Sekhar

Introduction

The first open approach for resection of a meningioma in this region was performed by Harvey Cushing in 1916. He and Eisenhardt subsequently coined the term suprasellar meningiomas in their classification Scheme [2]. The umbrella category of "suprasellar meningiomas" represents approximately 5–10% of all meningiomas [1] and constitutes a broad class of lesions that include meningiomas of the olfactory groove, planum sphenoidale, tuberculum sella, anterior and posterior clinoid processes, optic foramen, medial sphenoid wing, diaphragma sella, cavernous sinus, petrous apex, and upper clivus. Since that time, the term suprasellar meningioma has been refined significantly and broken down into subclasses of lesions that are defined based on their typical presentation and anatomic origination. Lesions that have similar characteristics and therefore common surgical approaches are often grouped together. A brief discussion of the terminology and anatomic definitions of the relevant regions follows.

Anatomic Considerations and Terminology

Olfactory groove (*OG*) *meningiomas*: The olfactory bulb and tract lie in the groove formed by the cribriform plate. Arising from the frontosphenoid suture, meningiomas of this region can involve any area from the crista galli anteriorly to the planum sphenoidale posteriorly. These lesions are considered distinct from PS/TS lesions [1] and are not considered here.

Planum sphenoidale (*PS*) *meningiomas*: The anatomic region of the planum sphenoidale is defined by the plane surface of the sphenoid bone, located superior to the sphenoid sinus and anterior to the sella turcica. The plate of the bone connects the lesser wings of the sphenoid bones and is bounded posteriorly by the anterior border of the grooved chiasmatic sulcus, which leads laterally to the optic canals. As a result of their anatomic proximity, meningiomas of the

PS have sometimes either been considered along with olfactory groove meningiomas [3] or with tuberculum sella meningiomas [4]. Due to their common presentation, we consider them along with the tuberculum sella meningioma group.

Tuberculum sella (*TS*) *meningiomas*: The anatomic definition of the TS is the midline elevation anterior to the dorsum sella but behind the chiasmatic sulcus. This term has been used broadly to describe tumor originating from the TS, chiasmatic sulcus, limbus sphenoidale, and diaphragma sella. Kinjo and Al-Mefty [5] distinguished the supra- and intrasellar diaphragma sella meningiomas from those of the TS and proposed a distinct anatomical classification.

Anterior clinoid (*AC*) *meningiomas*: The AC processes are posterior projections of the bone that form the medial most aspects of the lesser sphenoid wings. They provide a bony surface for attachment of the free margin of the tentorium cerebelli and are located laterally to the optic canal and laterally to the tuberculum sella. Meningiomas arising from AC are usually considered along with medial or inner sphenoid wing meningiomas [6] and are not discussed here.

Diaphragma sella (*DS*) *meningiomas*: The circular-shaped dural fold that forms the roof of the sella turcica is referred to the diaphragma sella. This structure attaches to the TS anteriorly and dorsum sella posteriorly and is bordered laterally by the dural folds of the cavernous sinuses. With the exception of Kinjo and Al-Mefty [5], meningiomas arising from DS are still largely considered along with TS/PS meningiomas. In the following section, meningiomas will be discussed with specific preference to the PS and TS. Due to their large clinical and surgical similarity, DS meningiomas will be discussed as part of PS and TS meningiomas.

Classification and Patient Selection

Lesions in this region can readily be operated on using open or endoscopic endonasal techniques. Patient selection for the appropriate approach is one of the most important factors in ensuring a safe and successful operation that maximizes resection and minimizes patient morbidity. Surgeon comfort with one approach over the other is an important consideration, as inexperience with either endoscopic or open approaches can result in suboptimal results. In terms of anatomic factors that dictate open vs. endoscopic approach, Mortazavi et al. [4] proposed a score-based classification system to predict perioperative morbidity and relative complexity of the case. This scheme assigns relative points to six parameters that are based on preoperative imaging and prior treatment. These parameters include tumor maximal diameter (size), optic canal invasion, internal carotid and anterior cerebral artery encasement, brain invasion, previous surgery, and previous radiation (Table 17.1). As such, tumors are graded on a scale of 0–11 and then subdivided further into classes that generally correlate with complexity of the case; class I, 0–3 points; class II, 4–7 points; and class III, 8–11 points (Table 17.2). An illustration of these classes is seen in Figs. 17.2a–c, 17.3a–c, and 17.4a–c.

Table 17.1 Sekhar-Mortazavi classification system for planum sphenoidale and tuberculum sellae meningiomas

Parameter	Points			
	0	1	2	4
Size	<2 cm	2–4 cm	>4 cm	–
Optic canal invasion[a]	<5 mm	>5 mm	Complete	–
Vascular invasion (ICA, ACA)	No	<180°	>180°	The maximum score for any combination more than four
Brain invasion on MRI	No or mild FLAIR signal	Significant FLAIR signal	–	–
Previous surgery	No	Yes	–	–
Previous radiation	No	Yes	–	–

[a]A maximum of 2 points is given for any combination of optic canal invasion

Table 17.2 Proposed Sekhar-Mortazavi classification system

Score	Class
0–3	I
4–7	II
8–11	III

The goal of this classification system was to use objective parameters to systematically categorize tumors which would allow for comparison of different clinical series, direct the choice of transcranial versus endoscopic approach, and possibly predict morbidity and outcome. The authors generally concluded that a class I tumor is favorable for an endoscopic approach, whereas a class III tumor should be operated on using an open approach. Class II tumors are of intermediate difficulty and should be considered on a case-by-case basis depending on surgeon preference and relative comfort. While this study requires independent validation, it provides a systematic framework for surgeons to consider when making decisions about management of meningiomas in this region.

Surgical Approaches

Choice of Operative Approach

The two standard open surgical approaches for meningiomas of this region are the frontotemporal or pterional craniotomy with orbital osteotomy (FTOO) for small- and medium-sized tumors and a bifrontal craniotomy with naso-orbital osteotomy (BFNO) for large- and giant-sized tumors.

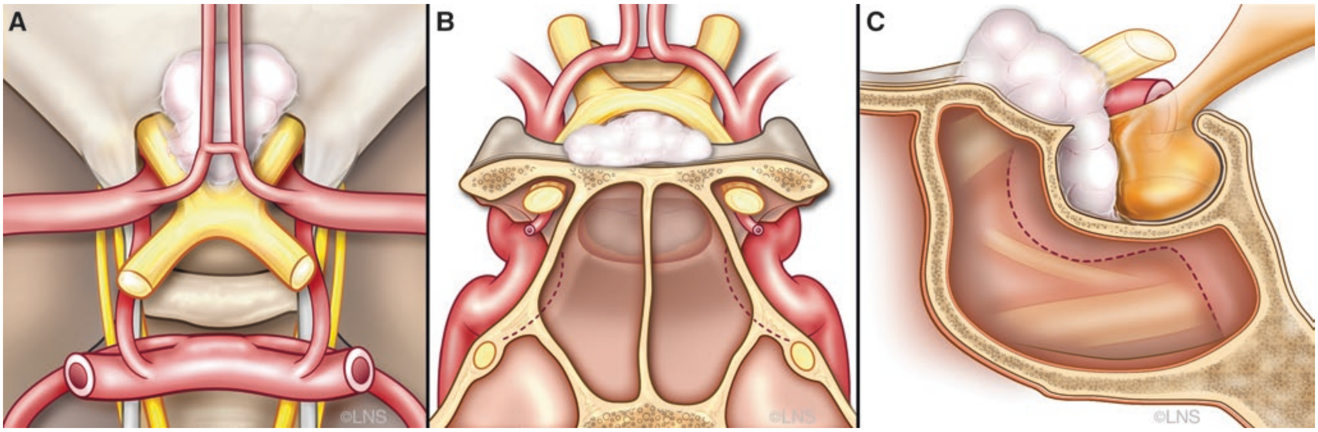

Fig. 17.2 (**a**–**c**) Illustration of the class I tumors based on classification by Sekhar-Mortazavi (Copyright retained by Dr. Laligam N. Sekhar. Used with permission)

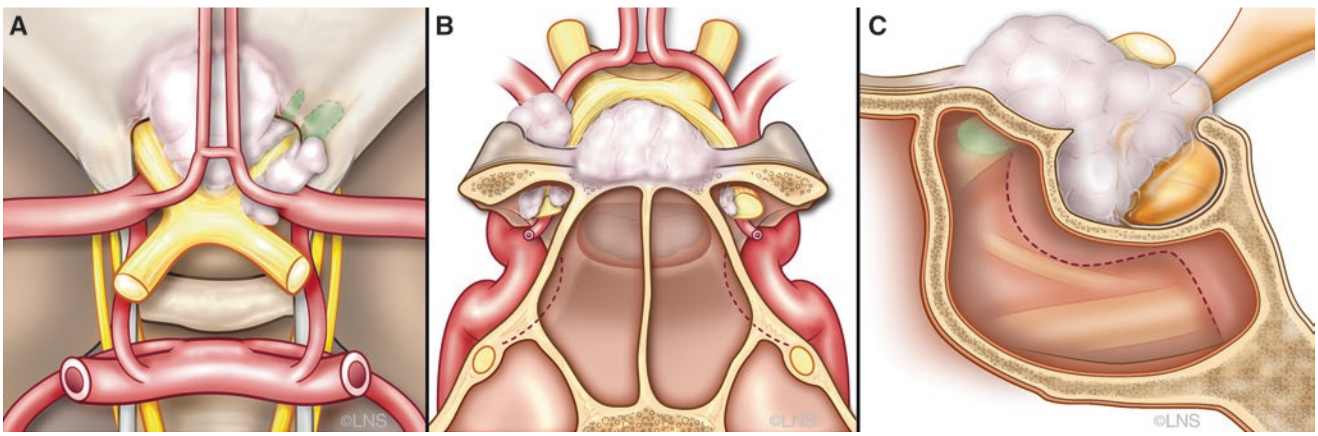

Fig. 17.3 (**a**–**c**) Illustration of the class II tumors based on classification by Sekhar-Mortazavi (Copyright retained by Dr. Laligam N. Sekhar. Used with permission)

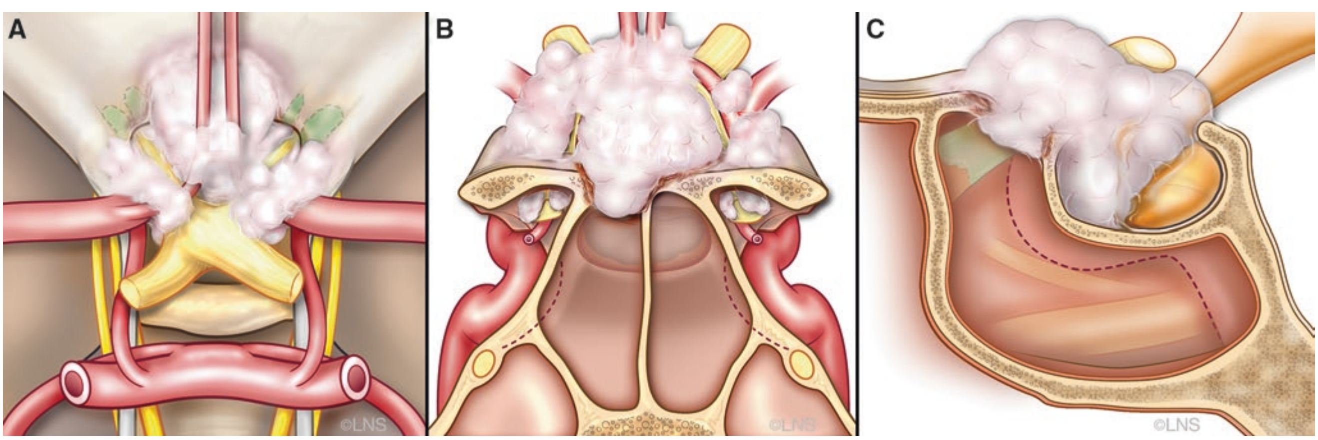

Fig. 17.4 (**a**–**c**) Illustration of the class III tumors based on classification by Sekhar-Mortazavi (Copyright retained by Dr. Laligam N. Sekhar. Used with permission)

Before orbitotomy approaches were popularized, these operations would typically be performed through a simple pterional craniotomy. Our experience has been this often results in the need for lumbar drain or ventriculostomy to obtain a sufficient working corridor and requires greater amounts of frontal lobe retraction. As such, we prefer to perform an orbital osteotomy along with a frontotemporal craniotomy to expand the working corridor and mitigate the need for retraction.

Advantages of the FTOO approach include early visualization of the optic apparatus and anterior cerebral arteries (ACA) allowing for safer dissection of the posterior aspect of the tumor and less retraction of the frontal lobe, and the opportunity avoids the frontal sinus in many cases. However, this approach allows for very limited view of the contralateral side of the tumor, in addition to a narrow working space. Therefore, it is more suitable with unilaterally located or midline small (0–2 cm) and medium-sized tumors (2–4 cm).

The BFNO, on the other hand, has the advantage of wide working space, direct access to the skull base for tumor devascularization and resection, and easier repair of the cranial base using pericranial flap. This approach is also indicated for tumors that involve both optic canals, as it allows for bilateral optic canal drilling. However, the major disadvantage of this approach is the late visualization of the optic nerves and the ACA and the opening of the frontal sinus. In addition, damage to the olfactory nerves is more common. Again, while this approach can be done without naso-orbital osteotomy, we have found that the removal of the naso-orbital bar precludes the need for retraction and CSF drainage. The BFNO approach is recommended for large (4–6 cm) and giant (>6 cm) meningiomas that encase bilateral ACAs and optic canals. Unlike olfactory groove meningiomas, PS/TS meningiomas of this size are far rarer as proximity to the optic nerves typically results in visual disturbance and earlier patient presentation.

Perioperative Considerations

The patient is intubated and maintained under general anesthesia using a balanced anesthetic technique. Routine perioperative antibiotics can be administered, and standard preoperative steroids are typically given. If there is any evidence of preoperative pituitary dysfunction or hypoadrenalism, stress dose steroids should be given to prevent intraoperative or postoperative hypotension. Patients are given anticonvulsants preoperatively and for 1–6 weeks postoperatively as manipulation of the frontal lobes can predispose to seizures. Brain relaxation is typically accomplished with intravenous mannitol and mild hyperventilation. In cases of a young patient or significant frontal lobe edema, lumbar drain or ventriculostomy can be considered. This is not our routine practice, as the combination of a skull base approach, intravenous mannitol, and wide opening of the CSF cisterns typically provides enough working room to remove the lesion without requiring significant retraction. Motor evoked potentials (MEP) and somatosensory evoked potentials (SSEP) are continuously monitored throughout the entire surgery. Electroencephalogram is also monitored to allow burst suppression, if needed.

We do not routinely perform angiography for the purposes of preoperative embolization for PS/TS meningiomas. The vascular supply of these lesions is mainly derived from small perforating branches from the posterior ethmoidal, ophthalmic, superior hypophyseal, and A1/A2 arteries. These tumor feeders are usually difficult or impossible to selectively access [7], and their occlusion may also pose a risk of visual disturbances [8]. However, in cases where the carotid artery (CA) or ACAs are heavily encased by tumor, preoperative angiography may be useful to determine if the vessels are invaded or occluded, though this is rare with PS/TS meningiomas.

Operative Technique for Planum Sphenoidale/Tuberculum Sella Meningiomas: Frontotemporal Craniotomy with Orbital Osteotomy

We typically prefer a right-sided approach to avoid left-sided frontal lobe injury when all considerations are equal. However, factors that would affect the side of the approach include vascular encasement and/or involvement of the optic canal, which would favor an ipsilateral approach to whichever side is involved. A frontotemporal craniotomy is performed (Fig. 17.5), followed by a full orbital osteotomy on the right side (Fig. 17.6a). Alternatively, a posterolateral orbitotomy (Fig. 17.6b) can be performed for smaller tumors or older patients where the brain is less full. The optic canal on the right and the superior orbital fissure are unroofed extradurally (Fig. 17.7), with the aid of a rough diamond drill and copious irrigation to ensure there is no thermal conduction to neural

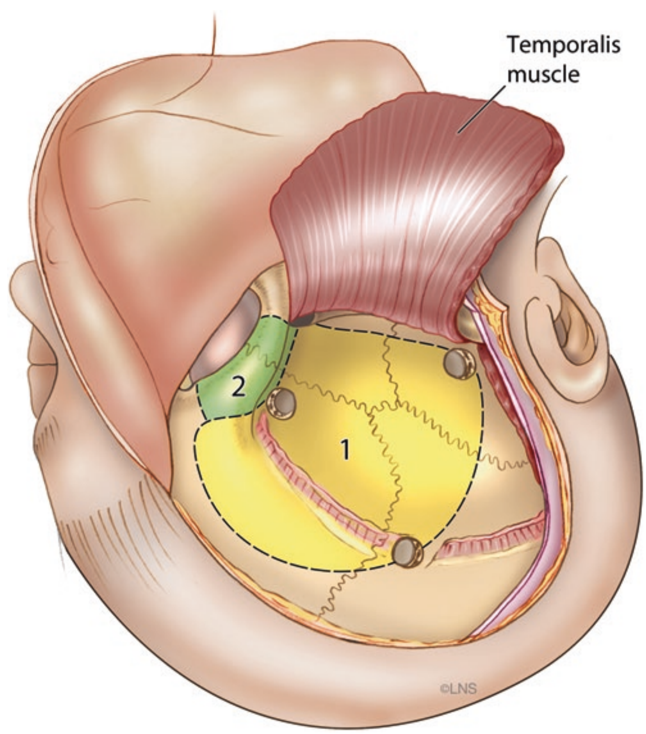

Fig. 17.5 Frontotemporal craniotomy: technical aspects. Note the position of the burr holes and the extension of frontal bone flap limited medially to the notch (Copyright retained by Dr. Laligam N. Sekhar. Used with permission)

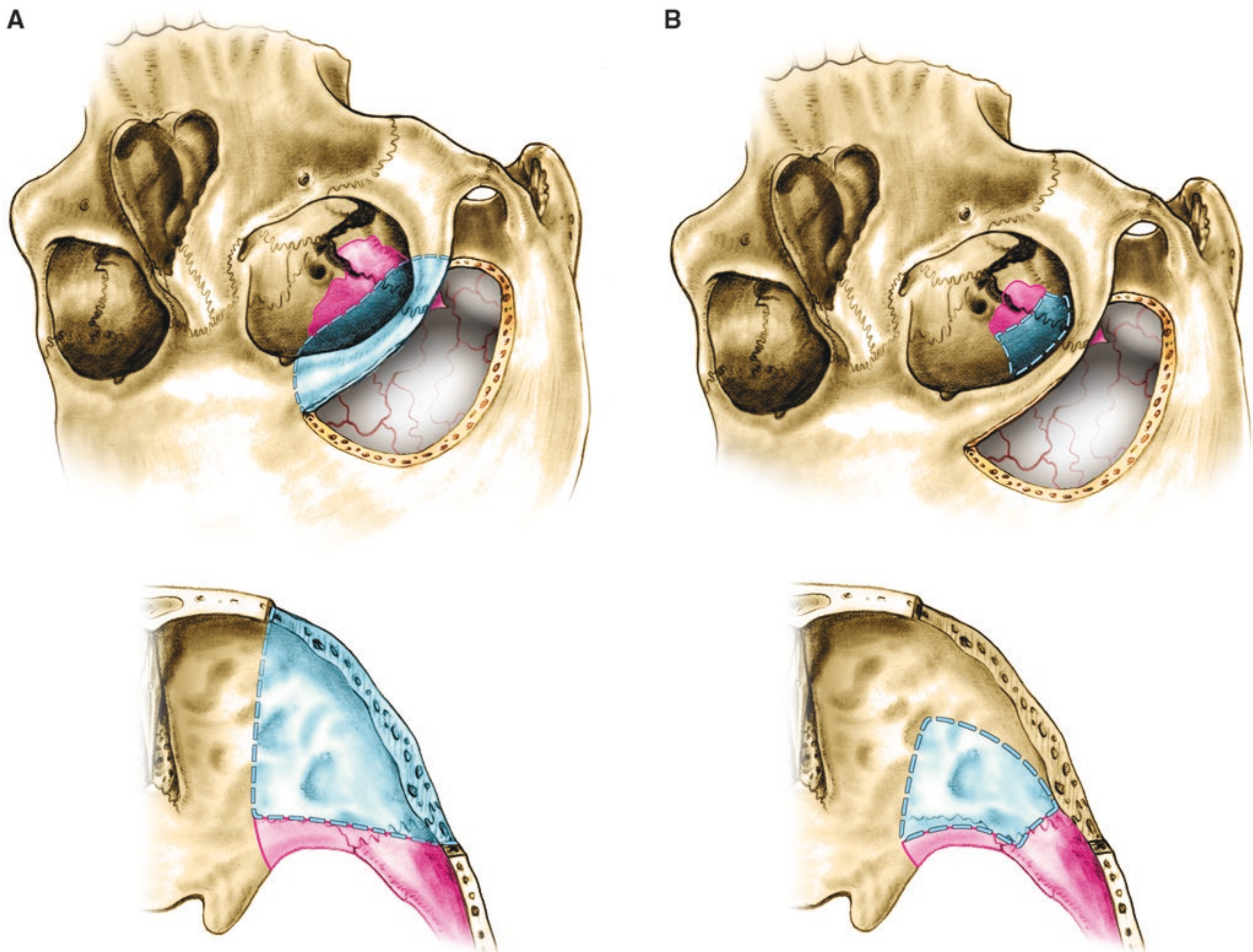

Fig. 17.6 (**a**) Full orbitotomy when brain is full. (**b**) Posterolateral orbitotomy when brain is relatively slack. Both alternatives include decompression of the superior orbital fissure, the optic canal and subtotal to total extradural resection of the anterior clinoid process (**a**, **b**: used with permission from Sekhar and Fessler [20])

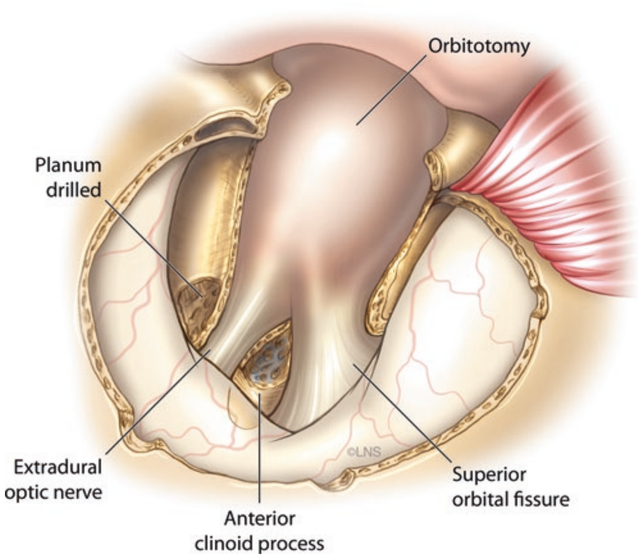

Fig. 17.7 Unroofing of superior orbital fissure and extradural decompression of the ipsilateral optic nerve (Copyright retained by Dr. Laligam N. Sekhar. Used with permission)

structures. This can also be accomplished using a SONOPET bone curette (Stryker, Kalamazoo, MI) [9].

Optic canal decompression is an important adjunct to this operative approach. In any case where the patient has preoperative visual disturbance or obvious involvement of the optic canal, we typically perform a complete extradural optic canal decompression and anterior clinoidectomy. In addition, we typically perform optic canal decompression on the side of the operation regardless of whether there is direct involvement of the tumor or not, as this prevents damage to the optic nerve during manipulation and removal of the tumor. It is important to perform this early on in the operation, prior to any tumor manipulation as there is evidence that visual outcomes are better with early decompression [10, 11]. If the tumor is large and access to the optic canal and clinoid is limited, or the clinoid process is too long to be safely removed extradurally, the decompression can be performed intradurally.

The decision to resect the anterior clinoid is taken on a case-by-case basis. If the clinoid is large and obstructs access to the tumor or is directly involved by tumor, we remove it. The anterior clinoid process may be resected extradurally if it is short or extra and intradurally if long. Careful attention should be paid on preoperative imaging to note whether the clinoid is pneumatized. Removal of a pneumatized clinoid without adequate measures to plug the defect into the nasal sinuses increases the risk of CSF leak significantly.

After the dura mater is opened, the Sylvian fissure on the right is opened widely. Before any subfrontal retraction is done, the dural sheath (e.g., falciform ligament) around the right optic nerve is widely opened to release the nerve intradurally and to release the pressure on the nerve (Fig. 17.8).

Internal tumor debulking is performed toward the base of the lesion, medial to the optic nerve. Sufficient debulking is critical, as it allows the edges of the tumor to be folded inward and away from any vessels or nerves without causing injury. Great care is taken to respect the arachnoid plane around the right ICA as encasing tumor is removed. The tumor is then disconnected from the planum and tuberculum dura and carefully dissected away from the right and left optic nerves and optic chiasm. Once the majority of the tumor has been removed, the contralateral optic nerve is visualized. At this point, any hyperostotic PS is removed, and the contralateral optic canal is also decompressed if necessary with a diamond drill. The dural sheath (e.g., falciform ligament) of the left (opposite) optic nerve is opened, and any tumor inside that optic canal is removed as well (Fig. 17.9). Tumor in the sella turcica is removed, paying attention to preserve the superior hypophyseal arteries (SHA) (Fig. 17.10).

PS/TS will typically respect an arachnoid plane around major vascular structures. As such, tumor encasement can be managed with careful microdissection. In the event of a vascular injury to the CA or ACA main trunks, primary repair

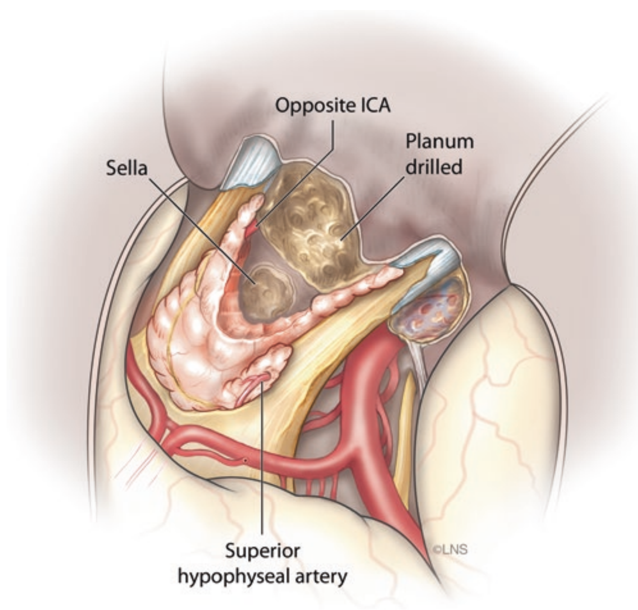

Fig. 17.9 Intradural decompression of the contralateral optic nerve through sectioning of the falciform ligament which allows inspection and resection of any tumor within the contralateral optic canal (Copyright retained by Dr. Laligam N. Sekhar. Used with permission)

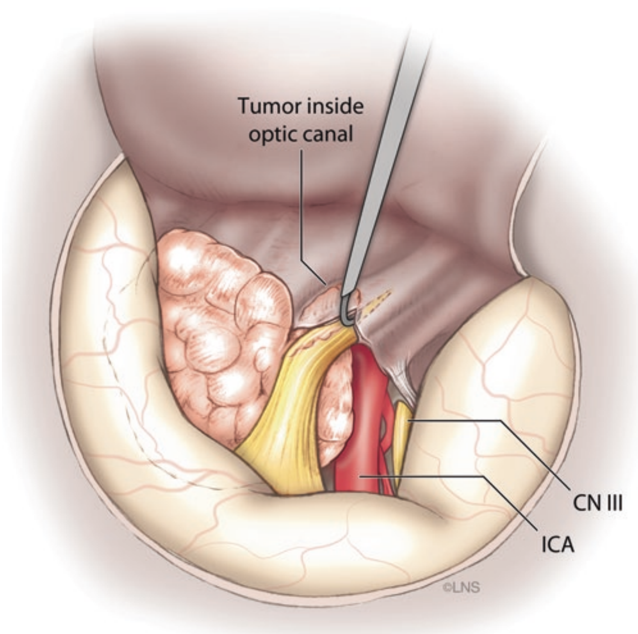

Fig. 17.8 Intradural decompression of the ipsilateral optic nerve by sectioning the falciform ligament superior to the optic nerve (Copyright retained by Dr. Laligam N. Sekhar. Used with permission)

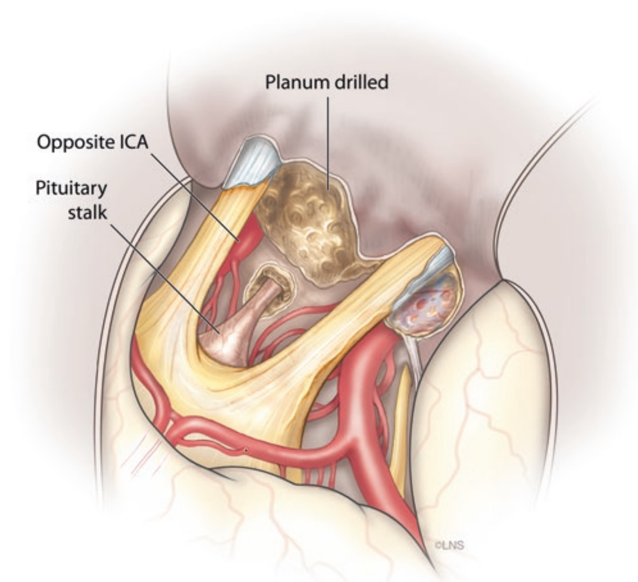

Fig. 17.10 Resection of sellar tumor extension (Copyright retained by Dr. Laligam N. Sekhar. Used with permission)

should be attempted. In the case of the SHA, injury typically requires coagulation and sacrifice. While unilateral SHA sacrifice is typically well tolerated, there have been reports of visual impairment from chiasm ischemia and endocrine dysfunction from pituitary stalk ischemia [12]. If tumor is adherent to or invading vascular structures and cannot be easily dissected, it should be debulked as much as safely possible and left behind as the vast majority of these tumors demonstrate a benign pattern of growth.

Operative Technique for Large- and Giant-Sized Meningiomas: Bifrontal Craniotomy with Naso-orbital Osteotomy

A bicoronal incision behind the hairline is used and typically extends from zygoma to zygoma to allow for complete exposure. The skin flap is dissected down to the naso-frontal suture, and bilateral supraorbital nerves and arteries are liberated from their notches if needed. The periorbital is dissected on one side from the bony orbital roof. In case of a giant tumor, this may be done on both sides.

A bifrontal craniotomy is typically performed in two pieces. The ipsilateral side is taken as far laterally as the keyhole, and the extent of contralateral craniotomy is tailored to the size of the tumor. In many cases, limiting the contralateral craniotomy to the midpupillary line (a one and a half bifrontal craniotomy) is enough to achieve adequate working room (Fig. 17.11). Following this, the posterior wall of the frontal sinus is removed, and the mucosal contents are fully exenterated. The frontonasal ducts are packed with oxidized cellulose. The dura mater of the frontal lobe is separated from the orbital roofs, and a naso-orbital osteotomy is performed. Again, complete bilateral removal is often not necessary and the extent of naso-orbital bone removal can be tailored to the size of the tumor. Care is taken to spare the midline bony structures, including the ethmoidal bone and crista galli (Fig. 17.12). The dura should never be elevated from the skull base floor past the crista galli if olfaction is to be preserved. A linear dural opening is performed bilaterally near the skull base and the superior sagittal sinus ligated just above the nasion (Fig. 17.13). Microdissection is advanced carefully along the skull base until the tumor is identified. Care is taken to try to preserve the olfactory nerves by dissecting them free from the frontal lobe to maintain their apposition to the skull

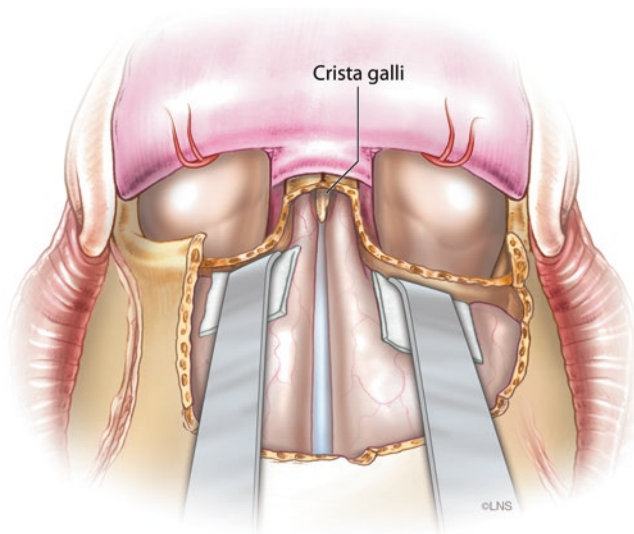

Fig. 17.12 Extradural dissection in preparation for orbito-fronto-nasal osteotomy (Copyright retained by Dr. Laligam N. Sekhar. Used with permission)

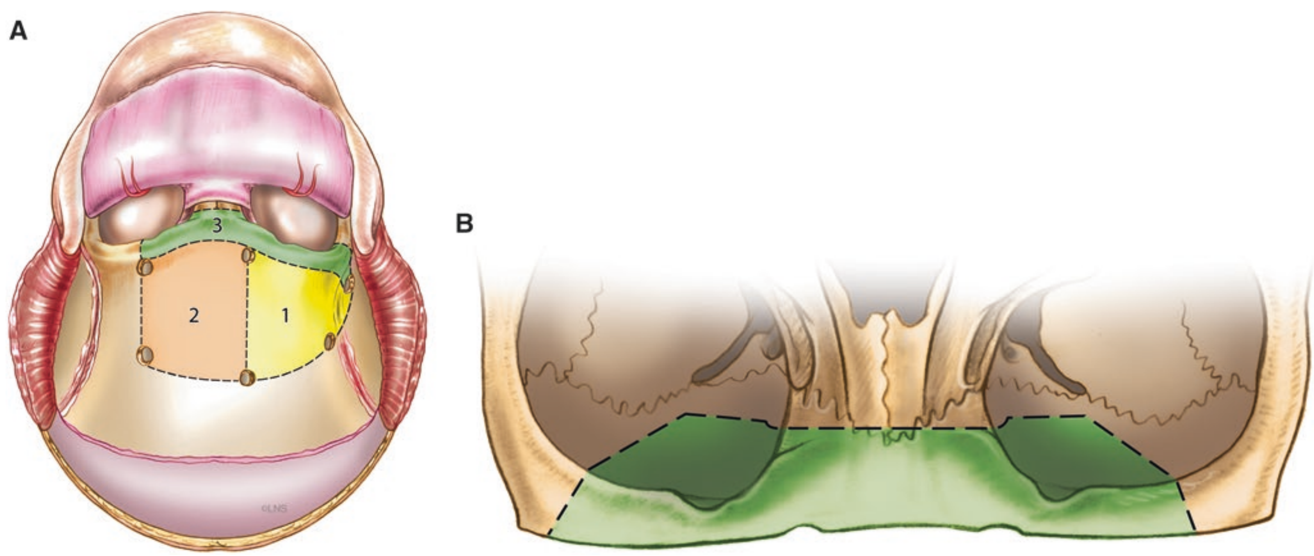

Fig. 17.11 (**a**) One and a half frontal craniotomy. Observe that the cranial flap is taken in two pieces for the safety of the superior sagittal sinus (**a**: Copyright retained by Dr. Laligam N. Sekhar. Used with permission). (**b**) Illustration of the naso-orbital osteotomy (**b**: used with permission from Sekhar and Fessler [20])

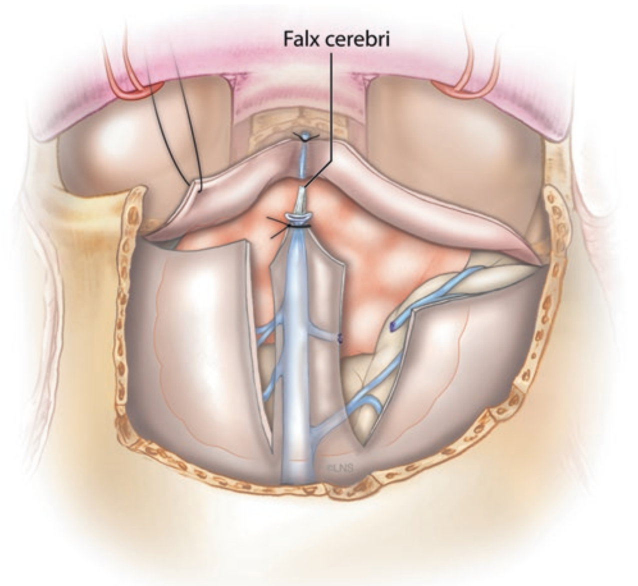

Fig. 17.13 Sectioning of falx cerebri and anterior superior sagittal sinus, in order to gain access to the contralateral side. If necessary, anterior bridging veins can be sectioned (Copyright retained by Dr. Laligam N. Sekhar. Used with permission)

base. If the tumor is biased to one side and/or already involves the olfactory nerve on that side, it is often sacrificed to allow a better working corridor. We always try to preserve at least one olfactory nerve if possible. If the optic canals are easily visible, they can be decompressed bilaterally at this time. In cases of giant tumors, they should be internally debulked to allow access to the optic canals without retraction on the frontal lobes or tumor manipulation that would damage the optic nerves. The remainder of the dissection should proceed as described above, taking care to identify the bilateral optic nerves, optic chiasm, CA, and ACAs bilaterally.

The dura is closed primarily if possible or using a patch if needed. It is often difficult to obtain a completely watertight closure, mandating that all connections to the nasal sinuses are excluded.

Skull Base Reconstruction

At any point during the drilling of the skull base, entry into the nasal sinuses is possible. This can occur during optic canal decompression, clinoid removal, or drilling of hyperostotic bone in the PS or TS. If this is not managed appropriately, the risk of CSF leak is increased significantly. Small defects can be obliterated with bone wax, but larger holes should be packed with muscle or fascia and augmented with fibrin glue.

During either approach, the frontal sinus may be entered. As described above, complete exenteration of the mucosal contents is required to prevent future mucocoele formation. We pack the naso-frontal ducts with oxidized cellulose and fill the dead space in the sinus with bone cement.

In cases where we are sure that frontal sinus exenteration will be required, we typically dissect and preserve a large, vascularized pericranial flap at the beginning of the operation. This can then be laid down along the skull base and carefully sutured to the frontal dura to hold it in place. We typically augment the repair with fibrin glue as well. Once the vascularized flap has been laid down, the naso-orbital or fronto-orbital bone can be fixed in place using a standard titanium plating system. Care must be taken to remove any mucosa from the bone pieces, as residual mucosal tissue can result in mucocoele formation. The remaining bone is then plated back into place as in any standard craniotomy, and routine closure is performed.

Postoperative Considerations

Patients are typically extubated unless there are extenuating circumstances requiring prolong intubation. They are monitored in the intensive care unit until stable, not typically more than 1–2 days depending on the extent of the operation. Complications following microsurgical resection of PS/TS meningiomas include CSF leak, meningitis (often due to CSF leak), seizure, motor deficit (from retraction or frontal lobe edema), vascular injury, pituitary dysfunction, and infection.

Seizures are more likely in patients with a bifrontal approach, significant brain invasion, and higher-grade (WHO II/III) tumors. In general, we maintain patients on anticonvulsants for up to 6 weeks. Steroids are given for a minimum of 4 days but are tailored to the patient's condition and degree of intraoperative and postoperative swelling. If there is evidence of pituitary dysfunction, stress dose steroids should be given and continued postoperatively until they can be safely tapered. While any form of endocrine dysfunction is possible, the most common complication is diabetes insipidus (DI). Urine volume, serum sodium, and patient thirst should be monitored closely.

CSF leak can be prevented by a robust skull base repair as described above. Postoperatively, the patient's head of bed should be maintained in an elevated position to decrease intracranial CSF pressure. Lumbar drainage for 3–5 days to divert CSF and allow scarring to close the defect is typically effective if there is an adequate reconstruction in place. Refractory cases of leak mandate a repair strategy, which can be performed using a vascularized naso-septal flap via endoscopic technique or on rare occasions requires reexploration of the skull base reconstruction via transcranial approach.

Outcomes for Open Approaches

Several reported case series have discussed outcomes for patients treated with open surgical approaches to PS/TS meningiomas [1, 4, 13–16]. A recent study performed a systematic review of the literature on transcranial approaches that included 31 articles for which the patient series was larger than 5 and extent of resection, visual outcomes, and complications were reported [17]. This review included 983 patients with a mean age of 54 years, 75% of whom were female. Mean follow-up was 43.9 months, and mean tumor diameter was 27.8 mm. They reported aggregated gross total resection (GTR) rates of 84% and near-total resection (NTR) rates of 14%. Vision improved, worsened, and remained unchanged in 65.6%, 10.4%, and 24.7%, respectively. They reported a CSF leak rate of 3.4% and transient or permanent pituitary dysfunction in 6.9% of patients (most commonly transient diabetes insipidus). Of note, risk of CSF leak (15.3%) and pituitary dysfunction (9.4%) [18] is both higher with endoscopic approaches as opposed to transcranial approaches, whereas seizure risk is generally considered to be higher in open surgery [19]. Vascular injury was reported as 5.1%, the majority of which were symptomatic in the form of postoperative neurologic deficits. Overall recurrence rate was 3.8% and mortality was 1.1%. While these studies represent the largest meta-analyses of patients with PS/TS meningiomas, individual case series have shown wide variation in reported outcome measures. This is a reflection of the fact that PS/TS meningiomas are a complex entity with many different variables that affect surgical decision-making and outcomes.

Vision-Related Outcomes

Visual symptoms are the most common reason for surgical intervention for PS/TS meningiomas. As such, visual outcomes are intimately tied to preoperative visual status. Patients with intact vision preoperatively have a very high likelihood of having intact vision postoperatively with careful microsurgical dissection and optic canal decompression. Nakamura et al. [16] reported that tumors smaller than 3 cm had lower rate of visual deterioration compared to those larger than 3 cm. In most studies, especially those that reported postoperative visual deterioration, intradural optic canal decompression was performed selectively at a late stage of surgery, not preceding tumor resection, and in some cases, the optic canal was not decompressed at all [14–16]. It was therefore suggested that early extradural optic canal decompression may be associated with better visual outcome by reducing tension on the optic nerve during tumor resection [10, 11]. Early unroofing of the optic canal was found to be an independent factor that increases the chance of better visual outcome. In our series, early extradural optic canal decompression was done in all patients. None of our patients experienced visual deterioration postoperatively (Table 17.3). Besides improving visual outcomes, optic canal decompression reduces the risk of tumor residual in the canals and subsequent tumor recurrence. Among 15 patients with preoperative visual impairment, 14 (93.3%) had postoperative improvement and 1 (6.7%) had no visual change. As such, optic canal decompression, particularly if done early, seems to be an important part of ensuring a good visual outcome in patients with PS/TS meningiomas.

Our Results

Twenty patients (15 females and 5 males) with small- or medium-sized PS and TS meningiomas (<4 cm) were included in our series (Table 17.3) [4]. The most common presenting symptom was visual disturbance, found in 15 (75%) patients, ranging from slight decrease of visual acuity to visual loss to visual field deficit. The tumor size ranged from 15 to 53 mm. Tumor extension into optic canal was found in 14 (70%) patients, and all of these patients had presented with visual disturbance. ICA and ACA partial or complete encasement was found in 17 (85%) patients and 10 (50%) patients, respectively. Significant FLAIR signal was found in two (10%) patients, who had larger tumor size.

Table 17.3 Outcomes of different surgical craniotomies for tuberculum sellae meningiomas

	Frontolateral[a]	Frontotemporal[a]	Bifrontal[a]	Our series[b]
Number of cases	30	21	21	25
Tumor size (mean)	1–5 cm (2.5 cm)	1–5 cm (2.66 cm)	1.5–5 cm (3.49 cm)	1.5–5.9 cm (2.7 cm)
Total resection	28 out of 30 (93.3%)	19 out of 21 (90.5%)	19 out of 21 (90.5%)	23 out of 25 (90%)
Visual improvement	77.8%	68.8%	46.2%	90.4%
Visual preservation	92.6%	81.3%	84.6%	100%
Olfactory nerve sacrificed (side)	2 patients [2], 1 patient [1]	1 patient [2], 1 patient [1]	3 patients [2], 5 patients [1]	–
Subdural hygroma	3 (10%)	–	4 (19%)	–
Hemorrhage	3 (10%)	1 (4.8%)	3 (14.3%)	–
Brain edema	–	–	4 (19%)	–
Brain infarction	–	–	1 (4.8%)	–
CSF fistula	1 (3.3%)	–	2 (9.5%)	2 (8%)
Diabetes insipidus	–	–	–	1 (4%)
Facial paresis	–	–	1 (4.8%)	–
Wound infection	–	–	3 (14.3%)	–
Death	–	–	2 (9.5%)	–

[a]Data from Nakamura et al. [16]
[b]Mortazavi et al. [4]

All of the 20 patients underwent our standard FTOO with extra- and intradural optic canal decompression. GTR was achieved in 18 (90%) patients. Among the two cases with near-total resection (NTR), residual was left due to severe adherence to the optic nerve and ICA, respectively. Two (10%) patients had WHO grade II tumors and received adjuvant postoperative Gamma Knife (GK) radiation. The mean length of follow-up was 25 months. During the follow-up, 14 (93.3%) patients showed visual improvement and 1 (6.7%) patient had no visual change. No patient had visual deterioration. Partial oculomotor paresis was found in one (6.7%) patient. Two patients had postoperative CSF leak and were successfully repaired using endoscopic transsphenoidal approach. No other permanent complication and no mortality were reported, and there was no tumor recurrence.

Another four patients (three females and one male) had large or giant meningiomas (>4 cm) that extended forward past the PS and into the olfactory groove. The most common presenting symptom was visual disturbance (75%). Anosmia was only found in one (25%) patient. The tumor size ranged from 41 to 69 mm (mean 58 mm). Tumor extension into the optic canals was found in two (50%) patients, and both these patients had presented with visual disturbance. ICA and ACA encasement was found in three (75%) patients and two (50%) patients, respectively. Significant FLAIR signal was found in three (75%) patients.

Three (75%) of these patients underwent modified BFNO approach, and one (25%) patient had bifrontal craniotomy without osteotomy. GTR was achieved in all patients. Two (50%) patients had WHO II tumors. One of them received adjuvant postoperative additional proton beam radiation, due to aggressiveness of the tumor. The second underwent GK treatment. During a mean follow-up of 18.8 months, all patients with preoperative visual decline showed improvement. No other permanent complications or mortality were reported. There was no tumor recurrence.

PEARLS
Tumors that are >4 cm, encase major vessels, demonstrate significant lateral extension to one or both sides, have extensive involvement of one or both optic canals, and have been previously operated on or irradiated should be considered for open approaches.
Skull base approaches, including orbital or naso-orbital osteotomy, are important for decreasing morbidity due to brain retraction.
Early optic canal decompression is crucial to ensuring optimum visual outcomes regardless of degree of invasion of the optic canal.
With good patient selection and microsurgical technique, including early optic canal decompression, open approaches can be performed with excellent neurologic outcome and long-term control rates.

Commentary on Case Presentation in Section 17.1

Case Presentation

A 52-year-old woman presents with several months of left eye blurriness. Examinations: OS, 20/30 (corrected), relative APD, 3/11 color plates; Humphrey VF, superotemporal arcuate field cut; OD, normal. Rest of neurological exam is normal (see Fig. 17.1a–d).

Discussion

The case presented here is an example of a class II tumor in the Sekhar-Mortazavi classification. Due to the size of the tumor, the extension lateral to the carotid on the left side, involvement of the left optic nerve, and the partial encasement of the ICAs and ACAs, we would elect for an open approach in this case. Given that the patient has symptomatic involvement of the left optic canal, we would perform a left frontotemporal craniotomy with orbital osteotomy for resection of the lesion. The critical portion of the case with regard to vision preservation is early, up-front decompression of the left optic canal and liberation of the left optic nerve. The frontotemporal approach will also allow for early visualization of the left ICA and left ACA, which can be dissected free after the tumor has been debulked. The lack of FLAIR signal in the brain suggests that arachnoid planes should be intact, facilitating removal of the tumor without damage to the vessels, pituitary gland, or optic apparatus.

17.3 Planum/Tuberculum Sella Meningiomas: Supraorbital Keyhole Approach

Georgios P. Skandalakis, Travis R. Ladner, Christopher A. Sarkiss, and Costas G. Hadjipanayis

Introduction

Over the past two decades, the use of the endoscope in neurosurgery has led to the increased popularity of keyhole transcranial approaches for the treatment of anterior cranial fossa lessons in combination with the operative microscope [21–23]. The eyebrow supraorbital keyhole approach is a minimally invasive approach selected for patients with clinoidal, tuberculum sella, and planum sphenoidale meningiomas [24–26]. This approach offers direct access to anterior skull base lesions through a small eyebrow skin incision, keyhole craniotomy, and minimal brain retraction with reduced complications, morbidity, and hospitalization [27–29].

Patient Selection

The supraorbital eyebrow keyhole approach is a minimally invasive approach tailored to treat patients harboring suprasellar tumors such as tuberculum sella, clinoidal, and planum sphenoidale meningiomas [23–26] for which a frontotemporal, pterional, or bifrontal craniotomy is traditionally employed [4]. Preoperative evaluation of the patient's anatomy, tumor location, and relationship to neurovascular structures is mandatory.

A key characteristic that should always be assessed in the preoperative imaging studies (CT, MRI) when considering the supraorbital approach is the anatomy of the frontal sinus. An extended lateral projection of the frontal sinus may preclude the use of the supraorbital eyebrow approach as it may limit the craniotomy medially and thus compromise the surgical corridor.

Surgical Technique

Skin Incision, Supraorbital Craniotomy, and Dural Opening

The patient, following endotracheal intubation, is positioned supine. The head is secured by a three-point skull fixation clamp. A single pin is positioned on the forehead at the hairline, contralateral to the side of the lesion, with caution not to invade the frontal sinus, while the two-pin rocker arm is positioned on the ipsilateral suboccipital skull region. The neck is slightly extended for frontal lobe relaxation, and the head is rotated approximately 20–25° contralateral to the approach. Image-guided neuronavigation is commonly used to localize and avoid the lateral extent of the frontal sinus and to tailor the supraorbital craniotomy. The skin incision is marked along the course of the eyebrow extending lateral to the supraorbital notch, which is palpated on the medial aspect of the supraorbital rim (Fig. 17.14a). The incision area is prepped in standard fashion after placement of adhesive protective covering over the eyelid to avoid any corneal exposure. Suture placement for closure of the eyelids can also be performed (see Pearls section toward the end of this chapter).

The skin incision is made through the hair follicles of the eyebrow. Some have advocated placement of the scalpel at an angle to the skin to minimize damage to hair follicles. Sharp dissection is performed through the dermal and subcutaneous layers, while monopolar cautery is avoided also to prevent hair follicle and nerve (supraorbital nerve and frontalis branch of facial nerve) damage. Bipolar cautery can be used for hemostasis. No scalp clips should be used for the approach. Blunt dissection is performed above the frontalis muscle and pericranium in a cephalad direction. A curvilinear C-shaped incision of the frontalis muscle and pericranium 3 cm superior to the incision is performed, and this layer is reflected inferiorly toward the eye with skin hooks or a suture. The frontalis/pericranium layer is quite important for reconstruction and covering of the craniotomy site to minimize any cosmetic deformity. Care is taken to protect the supraorbital nerve medially and the frontotemporal branches of the facial nerve laterally to avoid forehead numbness

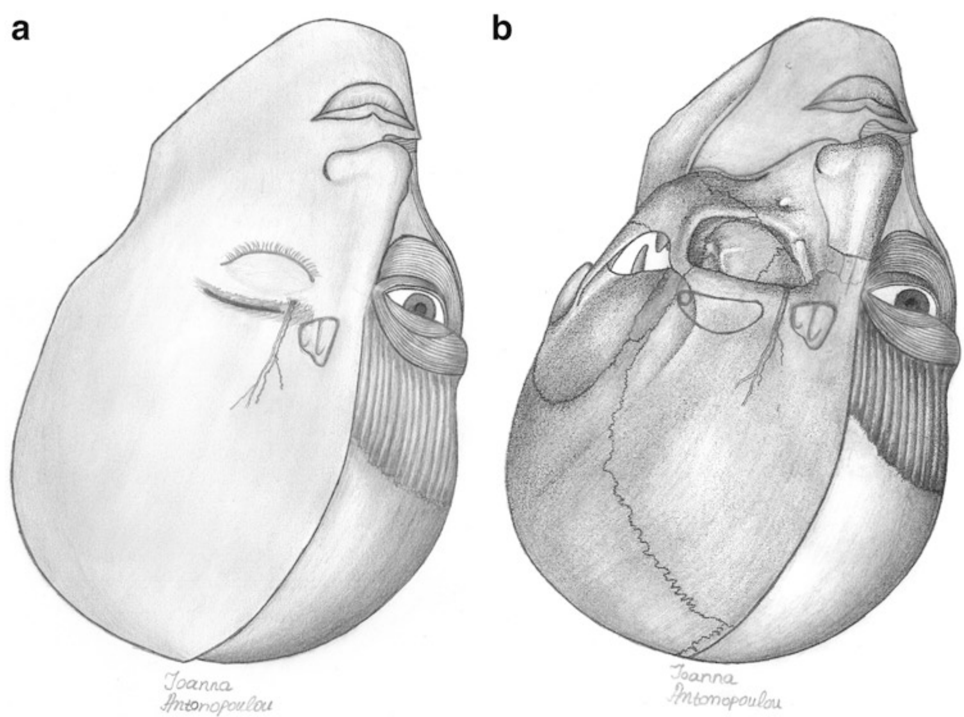

Fig. 17.14 (**a**, **b**) Schematic illustration of the supraorbital approach through the eyebrow. Supraorbital notch, nerve, and artery along with frontal sinus are also being illustrated. (**a**) Skin incision, (**b**) craniotomy (**a**, **b**: Courtesy of Ioanna Antonopoulou)

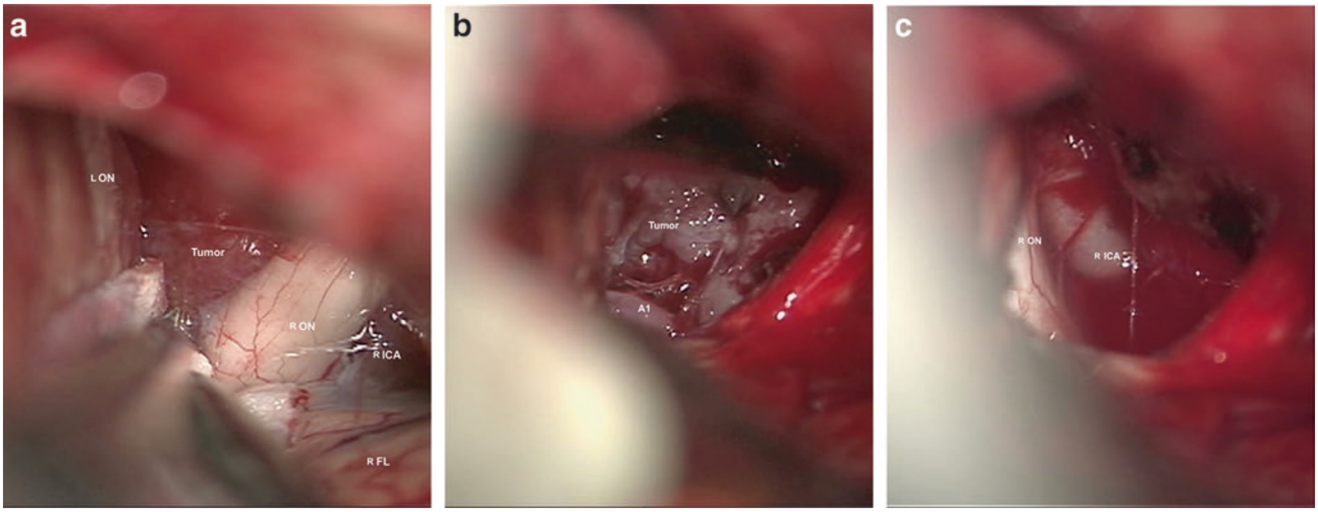

Fig. 17.15 (**a–c**) Intraoperative microscopic views of a right supraorbital approach through the eyebrow. *A1* A1 segment of the anterior cerebral artery, *L ON* left optic nerve, *R FL* right frontal lobe, *R ICA* right internal carotid artery, *R ON* right optic nerve

and a frontalis muscle palsy, respectively [25, 27, 30, 31]. The exposed temporalis fascia and muscle are bluntly dissected off the temporal bone laterally for the placement of the single keyhole burr hole. A 5 mm drill tip is used for the burr hole placement. Temporalis muscle manipulation is carried out cautiously to prevent postoperative atrophy, and its bony detachment should be minimized for placement of a 5 mm burr hole under the muscle.

A 5 mm burr hole is then drilled at the keyhole, inferior to the superior temporal line, with caution not to tear the dura. After freeing the dura off the bone, a tailored "D"-shaped (15–25 mm width and 20–30 mm height) craniotomy is performed lateral to the supraorbital notch (Fig. 17.14b). The dura is detached from the supraorbital craniotomy. If the lateral margin of the frontal sinus is exposed, then this should be sealed off with bone wax, or the frontal sinus opening should be covered with Betadine-soaked gel foam. The inner table of the inferior margin of the craniotomy site along with the bony prominences of the orbital roof is drilled flush using a high-speed drill, in order to maximize visualization and increase surgical maneuverability. The outer table of the supraorbital bar must be spared for cosmetic reasons. During drilling, the dura should be protected at all times, and care is taken not to expose the periorbital. The dura is opened in a "C"-shaped fashion with its base reflected inferiorly toward the orbit with a stitch. The dura is covered with a wet sponge to keep it moist for later reapproximation.

Microsurgical Approach, CSF Relaxation, and Endoscope-Assisted Tumor Removal

The operative microscope is initially utilized for the supraorbital approach. Wet cottonoids are used to cover and protect the inferior surface of the frontal lobe.

After gentle retraction of the frontal lobe, opening of the arachnoid over the optico-carotid cistern is performed for cerebrospinal fluid (CSF) drainage and relaxation of the frontal lobe. CSF drainage is very important for optimal exposure of the suprasellar region and direct visualization of tumor pathology involving the clinoid, tuberculum sella, or planum sphenoidale regions (Fig. 17.15a–c). Lumbar drain placement prior to the initiation of surgery may be helpful for relaxation of the brain but is not required. Microsurgical resection of the tumor can be performed in standard fashion while minimizing the manipulation of the optic nerves and supraclinoid internal carotid artery. Once the tumor resection is felt complete, the falciform ligament is cut, and the optic canal roof is removed using a diamond drilling burr for exploration of tumor extension into the canal. Removal of this residual tumor can be performed to complete the tumor resection.

At this point, a rigid rod lens endoscope can be used to further resect tumors and visualize residual tumor inferior to the optic nerves and extending into the sella. With a standard microscopic view, tumor inferior to the optic nerve may be difficult to visualize. The endoscope permits better visualization inferior to the optic nerve and in the canal to detect residual tumor. Panoramic visualization of the resection site will permit visualization of the dural origin of the tumor for complete resection of the dural attachment (Fig. 17.16a–c). The endoscope is stabilized with one hand, while the other hand is used for tumor dissection and removal. Following microsurgical and endoscope-assisted tumor removal, meticulous hemostasis is performed with bipolar cautery.

Closure and Reconstruction

The dura is now closed in a watertight fashion using a running and locking 4–0 Nurolon® (Ethicon, Somerville, NJ, USA) suture. For dural defects, a duraplasty can be performed to

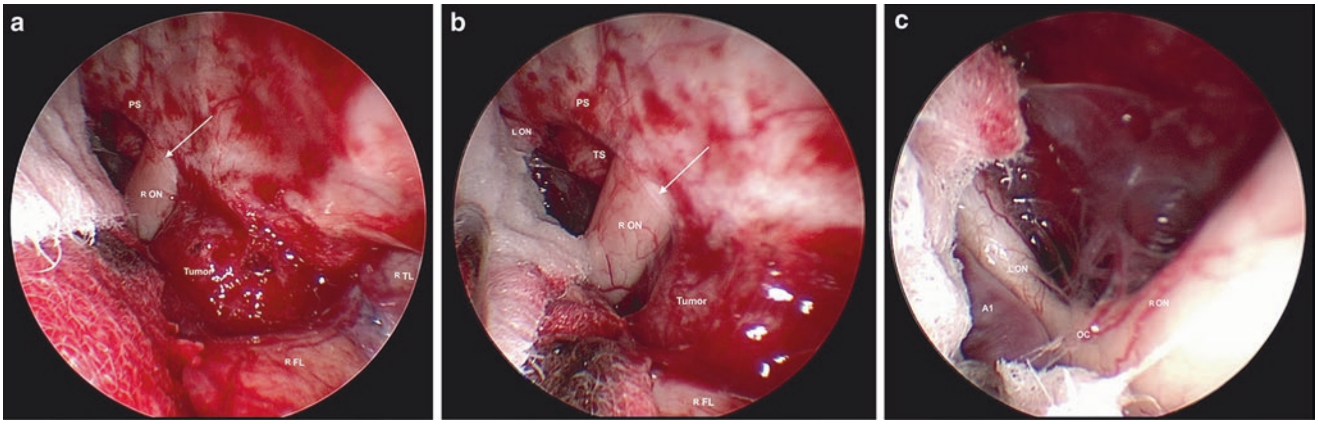

Fig. 17.16 (**a**–**c**) Intraoperative endoscopic views of a right supraorbital approach through the eyebrow. *A1* A1 segment of the anterior cerebral artery, *L ON* left optic nerve, *OC* optic chiasm, *PS* planum sphenoidale, *R FL* right frontal lobe, *R ICA* right internal carotid artery, *R ON* right optic nerve, *R TL* right temporal lobe, *TS* tuberculum sellae, *white arrow* falciform ligament

minimize the risk of CSF leakage. Collagen or a dural substitute can be placed above the dural suture line in combination with fibrin glue. A single titanium burr hole cover is used over the burr hole, and two titanium square plates are used to secure the bone flap aiming at minimizing bone gaps for better cosmesis [22, 27]. The bone flap is placed flush with the superior margin of the craniotomy site. Others have recommended the use of bone cement to minimize any bony defects [26, 32].

Closure of the incision is begun by reapproximation of the frontalis/pericranium layer with interrupted sutures. The subcutaneous and dermal layers are reapproximated with 3–0 Vicryl® (Ethicon, Somerville, NJ, USA) sutures. The skin closure is performed with a 5–0 Prolene sewn in a subcuticular fashion. An initial knot is tied, and then the suture is used to reapproximate the incision. An air knot is placed so the suture can be removed in 10–14 days after surgery. Pressure can be applied above the bone flap with a head wrap overnight to minimize periorbital edema.

Surgical Outcomes

The supraorbital keyhole approach can provide safe and effective access to anterior skull base lesions through a small craniotomy with minimal soft tissue dissection and brain retraction [4, 25, 33–35]. Lower morbidity [4, 25, 26, 28] and shorter hospital stays [23, 25, 27, 29] have been reported with excellent complete tumor removal rates and cosmetic results [4, 25–28, 36]. The approach and working corridor permit early identification of the supraclinoid internal carotid artery and its branches in addition to both optic nerves bilaterally permitting safe tumor resection [28]. The supraorbital corridor can also be applied to lesions extending lateral to the optic nerve and supraclinoid carotid artery as well as lesions extending to the middle fossa [28, 36].

In the largest case series published by Reisch and Perneczky [25], the authors report their 10-year experience with the eyebrow supraorbital subfrontal approach in a total of 450 patients harboring a variety of pathologies including meningiomas, pituitary adenomas, craniopharyngiomas, and anterior and posterior circulation aneurysms. The endoscope was used in 135 cases. Of the 93 anterior skull base meningiomas included in the study, 16 were located in the tuberculum sella and 12 in planum sphenoidale, while the remaining 65 were olfactory groove, anterior clinoid, cavernous sinus, and sphenoid wing meningiomas. In the extent of resection (EOR) that has a gross total in 89.2% (83/93), subtotal in 8.2% (8/93), and only in 2.1% (2/93) of patients, a partial debulking was achieved. With regard to visual outcomes, in the 33 patients that presented with visual deficits, 19 of them improved and nine remained unchanged, while the remaining five experienced deterioration in vision following surgery. According to the study, the most common approach-related complications encountered in the entire cohort of 450 patients were permanent partial supraorbital hypoesthesia in 7.5%; permanent palsy of the frontal muscle in 5.5%; mastication issues without atrophy of the temporalis muscle in 0.6%; permanent unilateral and bilateral hyposmia in 6.0% and 2.0%, respectively; poor wound healing in 1.3%; presence of a cerebrospinal (CSF) pseudomeningocele in 4.4%; CSF leak in 2.6%; postoperative intracerebral hematoma resulting in poor neurological outcome in 0.8% of patients, including one death; and subdural hygroma in 1.6% of cases.

In another study by Paira-Neto and Tella Jr. [37], 24 patients harboring skull base meningiomas, nine of which were located in tuberculum sella, were treated through the keyhole supraorbital approach without the aid of endoscopy. In seven out of the nine cases of tuberculum sella meningiomas, a gross total resection was achieved; while in the remaining two, the removal was near total. Postoperatively, two patients developed transient diabetes insipidus, and one had CSF rhinorrhea that was successfully treated with a lumbar drain.

Kabil and Shahinian treated five patients with suprasellar meningiomas through the supraorbital approach with the use of the endoscope [24]. Although suprasellar meningiomas predominantly originate from the planum sphenoidale or tuberculum sella, the authors do not specifically state their anatomical origin and have broadly categorized them as suprasellar lesions. Of these five cases, two were recurrent tumors. Gross total tumor resection was achieved in four cases. The authors did not reveal any postoperative complications, and all patients were discharged within 48 h and excellent cosmetic results.

In another series by McLaughlin et al. [36], the supraorbital keyhole approach was performed in seven patients with residual or recurrent tuberculum sella meningiomas previously treated through a pterional craniotomy, transsphenoidal approach, or both. The endoscope was utilized in three cases and a subtotal resection was achieved in six patients, whereas in one case, the removal was near total. In the patients presenting with visual deterioration, postoperative improvement of visual acuity and/or visual fields was documented, whereas no case of visual deterioration was reported. One patient suffered an asymptomatic ICA injury, and in another one, a bilateral caudate head infarct resulting in temporary confusion was recorded.

The lesion's specific anatomical characteristics and relationship with surrounding neurovascular structures should be also carefully assessed. In larger tumors that may be fibrous, calcified, or encase, the internal carotid artery may be ideally treated through more traditional approaches [26].

Complications and Their Avoidance

Apart from the common adverse events associated with craniotomies in general, the supraorbital keyhole corridor has some specific approach-related complications. Injury to the supraorbital nerve and frontotemporal branches of the facial nerve results in forehead numbness and difficulty in raising the eyebrow and forehead. Stretching those nerves during soft tissue dissection may also cause similar effects, which are usually transient [25, 26]. To avoid these shortcomings, proper placement of the incision lateral to the supraorbital notch and careful soft tissue dissection should be performed (see surgical technique section) [25, 38].

Inadvertent entry into to the frontal sinus during the supraorbital craniotomy can increase the risk of CSF leak and related infections such as meningitis, brain abscess, subdural and epidural empyema, and subcutaneous and subperiosteal abscess [39, 40]. For small, confined sinus openings, the use of bone wax is adequate, whereas for larger frontal sinus breaches, Betadine-soaked gel foam supplemented with a dura sealant is recommended [22]. The use of harvested, vascularized pericranial flaps has also been suggested with excellent results [41]. To prevent entry into the frontal sinus, imaging studies (CT and MRI) should be reviewed, and neuronavigation should be utilized during surgery to map the lateral extent of the frontal sinus.

Suboptimal cosmetic results have been reported and are related to poor closure technique. The bone flap should be secured as medially and superiorly as possible, aiming to reduce prominent forehead gaps [26, 42]. Closure of the wound should be performed in a meticulous layer-by-layer fashion. Care is taken to reapproximate the frontalis/periosteal layer over the craniotomy. Patients with a known history of impaired healing and a tendency to form keloids or hypertrophic scars should be considered for incisions behind the hairline with traditional craniotomy approaches [32].

Finally, in order to avoid the rare event of postoperative pseudomeningocele formation [43], a watertight dural closure along with application of pressure over the bone flap with a head wrap has been used with good results [28].

Eyelid Approach

A few studies describing an alternative anterior cranial fossa approach through an eyelid incision have also been published recently [44–46]. The "eyelid approach" entails a spheno-orbital keyhole burr hole along with a fronto-orbital craniotomy that incorporates the anterior two thirds of the orbital roof [45]. The potential benefits of this approach are better cosmetic results, lower risk of injury to the frontal and temporal branches of the facial nerve, and preservation of the frontal muscle. Nonetheless, the published cases of tuberculum sella and planum sphenoidale meningiomas treated through this approach are less than ten patients in total. Furthermore, there are no studies directly comparing the supraorbital keyhole to the eyelid approach.

Clinoidectomy and Optic Nerve Decompression

A partial anterior clinoidectomy and unroofing of the optic canal can be performed to aid in total tumor resection by "untethering" the optic nerve from the clinoid segment of the internal carotid and thus providing surgical access to this confined space in cases of TS and PS meningiomas that encase the internal carotid artery and/or the optic nerve [47]. Unroofing of the optic canal can be done with both the microscope and endoscope during the supraorbital approach. Visual function is already compromised in most of these cases [11]. Although there are no published clinical data evaluating the utility of the anterior clinoidectomy or optic nerve decompression during the supraorbital approach, both of them have been extensively performed through other approaches (pterional, lateral supraorbital, frontotemporal, subfrontal with or without supraorbital osteotomy, fronto-lateral, and fronto-orbital) in order to achieve maximal resection of TS and PS meningiomas [10, 11, 48–52]. Clinical studies evaluating the feasibility and utility of anterior clinoidectomy and optic nerve decompression are scarce. Additionally, these studies do not provide a technical description of this specific modification nor report on outcome data [28, 36].

Vascular Encasement

TS and PS meningiomas may commonly encase major vessels such as the ICA, ACA, and their branches [4, 21]. In this instance, tumor dissection should respect the arachnoid plane so as to reduce inadvertent intraoperative injury to surrounding neurovascular structures [21]. It is important to debulk the tumor initially prior to dissection along the arachnoidal plane. The absence of this arachnoid plane significantly increases the risk of vascular injury and complication rate [4].

Postoperative Care

A Band-Aid can be applied to the skin incision after closure. A tight head wrap is applied for 48 h after surgery to prevent pseudomeningocele formation. Care must be taken not to compress the orbits with the wrap. Patients can be mobilized soon after surgery. Routine labs can be obtained that include a basic metabolic panel. If large urine output occurs after surgery, then monitoring for diabetes insipidus is performed. This would be likely transient with these tumors since they are suprasellar and do not typically involve the pituitary stalk. Most patients are discharged home within several days after surgery. Postoperatively, patients should be monitored for rhinorrhea or pseudomeningocele formation. Removal of the head wrap can be done within 48 h after surgery. The Prolene® (Ethicon, Somerville, NJ, USA) stitch removal should occur approximately 10 days after surgery. Postoperative visual function should be assessed within 1–2 months after surgery.

PEARLS
Preoperative imaging to assess frontal sinus anatomy, head positioned 20–25 contralateral to approach with slight extension of neck
Skin incision placed within eyebrow lateral to the supraorbital notch for preservation of supraorbital nerve/artery
Dissection of frontalis/pericranium as separate layer from the skin for reconstruction and optimal cosmesis
Single burr hole placed laterally just below superior temporal line and D-shaped craniotomy performed
Inner table of frontal bone and superior orbital roof drilled to optimize supraorbital approach and maximize visualization
Relaxation of ipsilateral frontal lobe after drainage of CSF from optico-carotid cistern opening
The use of endoscope assistance after microsurgical approach/resection to inspect for residual tumor inferior/medial to the ipsilateral optic nerve and in the sella
Watertight closure of dura
Positioning of bone flap flush with superior portion of craniotomy with the use of titanium plates and screws
Reapproximation of frontalis/pericranium, subcutaneous, and skin layers

Commentary on Case Presentation in Section 17.1

Case Presentation

This 52-year-old lady presents with left eye blurriness for several months. On examination, left eye visual acuity is 20/30 (corrected). She has relative afferent papillary defect and 3/11 color plates, and superotemporal arcuate field cut is seen on Humphrey visual field examination. Right eye visual acuity and the rest of the neurological exam are normal (see Fig. 17.1a–d).

Discussion

MRI studies reveal a midline suprasellar lesion of the planum/tuberculum sella with the radiologic characteristics of a meningioma (Fig. 17.1b). Lesion abuts the internal carotid arteries bilaterally and the anterior cerebral arteries superiorly. There is slight extension of the tumor to the left of midline. There appears to be a small frontal sinus present. This patient may be a good candidate for the supraorbital approach as this tumor can be very well visualized and resected through this approach. Endoscope utilization will offer optimal visualization to ensure removal of any residual tumor that cannot be visualized through the microscope. Bony unroofing of the optic canal can be performed to ensure removal of tumor growth in the canal and inferior to the optic nerves. An endoscopic endonasal extended approach could also be used for removal of this tumor. Visualization of tumor inferior to the optic nerves would be possible with this approach. With the endonasal approach, the tuberculum and the posterior planum sphenoidale would need to be drilled off to access this suprasellar mass. A nasoseptal reconstruction would be needed for covering of the tuberculum/planum bony defect and prevention of CSF leak. A standard craniotomy for a subfrontal or pterional approach could also be used to resect the tumor. Special care would be taken to avoid vessel injury with any approach. The tumor capsule would need to dissect off the anterior circulation vessels, specifically the anterior cerebral arteries superiorly and the internal carotid arteries laterally. The supraclinoid internal carotid artery on the left would be at greater risk for injury since the tumor favors the left of midline. Compared to the other approaches equally suitable for this lesion, the supraorbital approach could minimize the risk of CSF leak and also provide a minimally invasive treatment option with an acceptable cosmetic result.

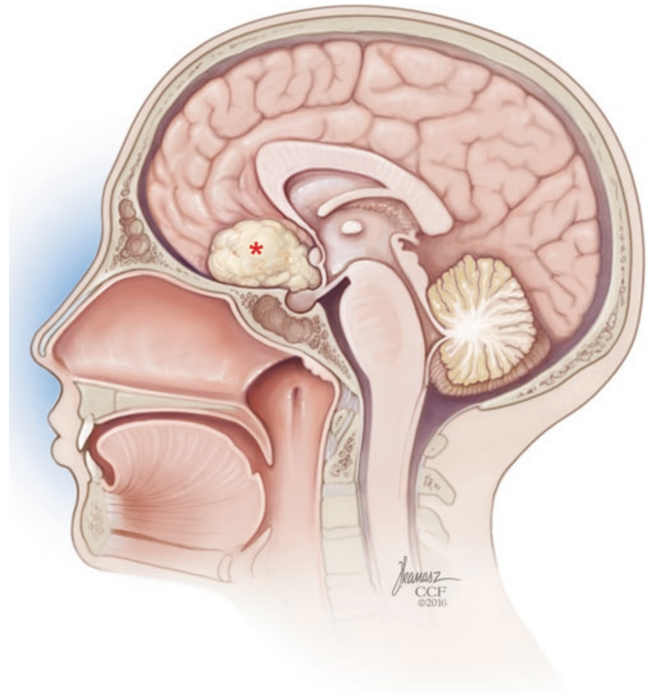

Fig. 17.17 Artist rendition showing a sagittal midline view of the cranial base with a planum sphenoidale meningioma. These meningiomas (*) arise from the dural lining between the tuberculum sellae and cribriform plate and typically become symptomatic after achieving large size due to compression of neurovascular structures posteriorly or from adjacent mass effect on the frontal lobes (Reprinted with permission, Cleveland Clinic Center for Medical Art & Photography © 2016. All Rights Reserved)

17.4 Planum/Tuberculum Sella Meningiomas: Endoscopic Endonasal Approaches

Danilo Silva, Rupa Gopalan Juthani, Brian C. Lobo, Troy D. Woodard, Raj Sindwani, Varun R. Kshettry, and Pablo F. Recinos

Introduction

Cranial base meningiomas represent approximately 40% [53] of all meningiomas with planum sphenoidale (PS)/ tuberculum sella (TS) meningiomas comprising 5–10% of these tumors [53–55]. Although usually grouped together as suprasellar meningiomas, the clinical presentation of PS and TS meningiomas may be slightly different, the former presenting with visual deficits late in the course of its natural history and the latter presenting with visual symptoms early [56–61]. PS meningiomas arise from the dural lining between the TS and the olfactory groove (Fig. 17.17). As they enlarge, they push the optic apparatus posteriorly. Headache is the most common presenting symptom (63%), followed by visual loss (14%) [61]. TS meningiomas arise from the dura of the chiasmatic sulcus between both optic foramina (Fig. 17.18). As they grow, they displace the optic chiasm either superiorly (prefixed chiasm) or posteriorly (postfixed chiasm) [56] (Fig. 17.19). The most common presenting symptom of TS meningiomas is visual deficits (84–89%) [62].Additionally, endocrinopathies can be present in approximately 25% of cases, due to compression of the pituitary gland and infundibulum [62].

Available treatment options include observation, radiation therapy/radiosurgery, and surgical resection. Observation is generally indicated for asymptomatic patients, regardless of age, or non-elderly patients with asymptomatic small tumors. Radiation options are generally reserved as a primary option for high-risk surgical candidates or as a secondary option for growing residual tumor [63, 64]. Notably, radiosurgery is frequently not an option due to the proximity of the tumor to the optic nerves, making fractionated stereotactic radiotherapy the best option in these cases. Surgical resection is the mainstay treatment for symptomatic patients and those with growing tumors. Although transcranial approaches have traditionally been the standard surgical option for PS/TS meningiomas, the expanded endonasal approach (EEA) [65] has risen as an alternative. In this chapter, we describe nuances of the EEA for PS/TS meningiomas, together with our patient selection process and a review of the available data in the literature regarding clinical outcome.

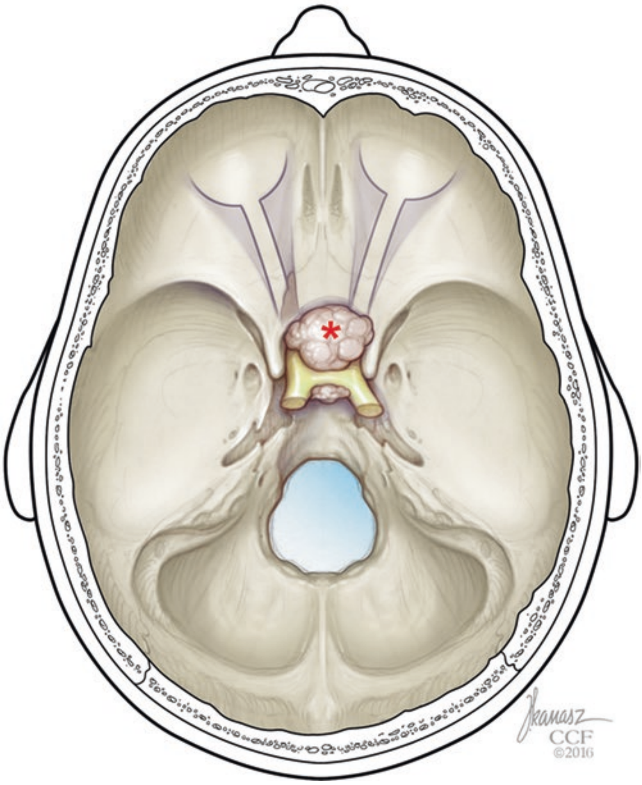

Fig. 17.18 Artist rendition showing an axial view of the cranial base with a tuberculum sellae meningioma. These meningiomas (*) arise from the dura of the chiasmatic sulcus between both optic foramina and become symptomatic relatively earlier in the course compared to other anterior cranial base locations due to invasion of the optic canals, most commonly in its medial compartment as shown above (Reprinted with permission, Cleveland Clinic Center for Medical Art & Photography © 2016. All Rights Reserved)

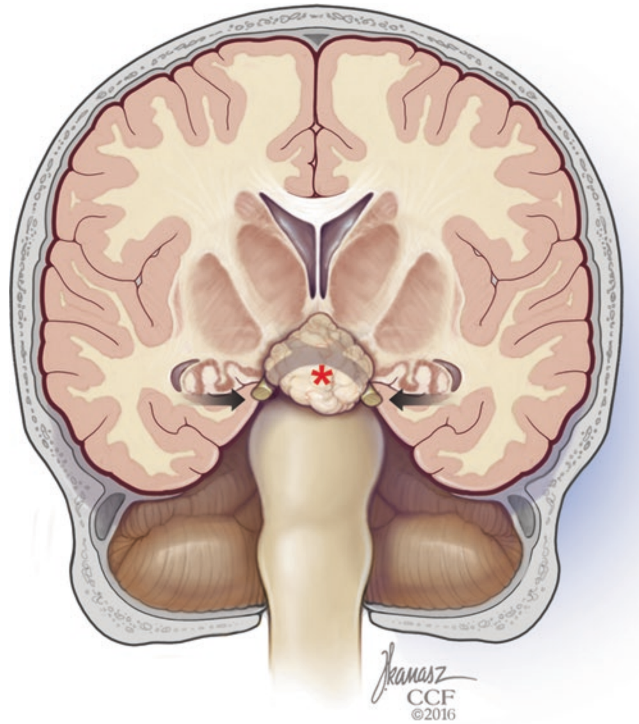

Fig. 17.19 Artist rendition showing a coronal view illustrating the lateral displacement of the optic nerves (*arrows*) and posterosuperior displacement of the optic chiasm (*shadow image*) by a tuberculum sella meningioma (*) (Reprinted with permission, Cleveland Clinic Center for Medical Art & Photography © 2016. All Rights Reserved)

Surgical Technique

Preoperative Planning and Patient Selection

In our experience, the ideal patients with PS/TS meningiomas who should be considered candidates for EEA are those with tumors located medial to the optic nerves and the carotid arteries [56]. This anatomical limit is more important than the size of the lesion. Specifically, the superior and posterior extension of the tumor can be approached via the EEA now that contemporary instrumentation includes angled endoscopes as well as angled and malleable instruments. The goal of surgery for PS/TS meningiomas is maximal safe resection, with preservation or restoration of neurological, visual, and endocrinological function through decompression of the frontal lobes, optic apparatus, and pituitary stalk. For elderly and healthy patients presenting with visual symptoms, either unilateral or bilaterally, a more conservative approach with subtotal tumor resection can be achieved with optic chiasm and medial optic nerve decompression [57] in order to tentatively improve visual outcome. High-risk surgical candidates are usually not managed operatively, and radiosurgery or other forms of radiation therapy should be considered [63, 64]. Embolization is usually not performed for these types of meningiomas, because the blood supply is mainly provided by branches of the posterior ethmoidal, ophthalmic, and superior hypophyseal arteries, with a smaller contribution from branches derived from A1/A2 segments of the anterior cerebral arteries [65].

Our selection process for the recommendation of the EEA for PS/TS meningiomas involves synthesizing clinical, anatomical, and radiological information. All patients with TS meningiomas and those with PS meningiomas with significant posterior extension undergo a neuro-ophthalmological and endocrinological clinical evaluation. Additionally, in cases with significant mass effect on the frontal lobes with associated T2/FLAIR changes on MRI, we also obtain preoperative neuropsychological evaluation to determine baseline status for postoperative follow-up.

Radiological assessment includes a stereotactic sinus CT scan (1 mm cuts) for a detailed understanding of the sinus anatomy, specifically the sphenoid septal anatomy around the sella turcica as well as any important sinus variants impacting the surgical approach. In addition to a stereotactic gadolinium-enhanced brain MRI (0.7 mm cuts), we routinely perform a more detailed imaging study with three-dimensional

CISS (Construction Interference in Steady State) sequences, which has been shown to be of great value in skull base pathologies [66–69]. The CT scan and MRI are fused and utilized intraoperatively with frameless neuronavigation.

Initial Steps

The patient is placed under general anesthesia, with the endotracheal tube to the left side. An arterial line and Foley catheter are placed. Preoperative antibiotics are administered per our institution protocol, cefazolin and/or vancomycin, depending on the result of nasal swab screening for *Staphylococcus aureus* and methicillin-resistant *Staphylococcus aureus*. We routinely place lumbar drains during EEA when addressing intradural pathology and when a high flow CSF leak is expected [70].

The patient is positioned supine on a foam donut with the head slightly rotated to the right side. Nasal mucosa is soaked with epinephrine nasal vasoconstrictor solution. The navigation system is brought into place, and face recognition registration is performed. The abdomen and the lateral surface of the right thigh are prepped in routine sterile fashion in case the abdominal fat or fascia lata is needed. Draping of the face is then performed such that it is kept separate from the sterile-prepped regions. Given the lack of documented benefit and comparable infection rates, it is not our practice to prep the face or the nasal sinuses.

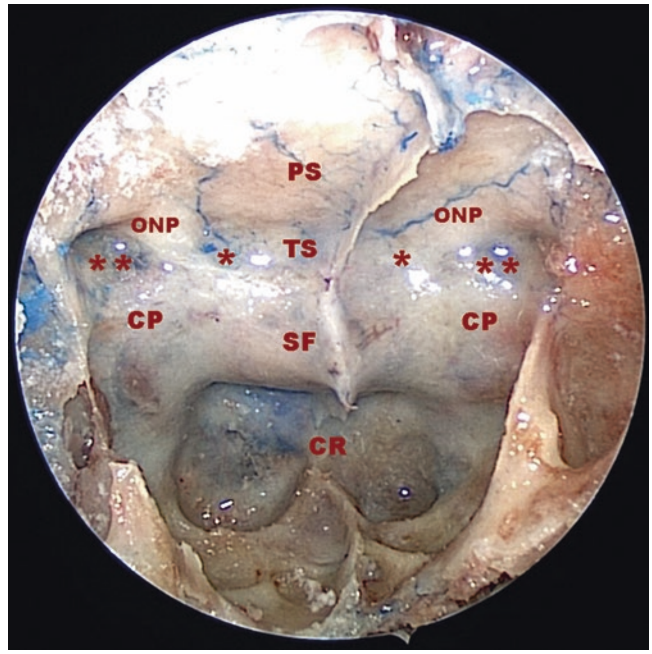

Fig. 17.20 Cadaver dissection image using a zero degree endoscope showing a panoramic view obtained with the endoscopic endonasal transsphenoidal approach. The tuberculum sella (TS) is situated in a deeper plane at a 90° angle between the sellar face (SF) and planum sphenoidale (PS). The carotid protuberances (CP), optic nerve protuberances (ONP), clival recess (CR), medial opticocarotid recesses (*), and lateral opticocarotid recesses (**) are also seen

Endonasal Approach [71, 72]

Using a zero degree endoscope, the middle and inferior turbinates are infiltrated with 1% lidocaine/epinephrine solution. The middle and inferior turbinates are then displaced laterally through the right nostril. The choana, sphenoethmoid recess, and superior turbinate are identified. The sphenoid ostium is located approximately 1 cm superior to the choana and medial to the tail of superior turbinate, which is removed for clear exposure of the ostium. Systematically, the same structures are identified on the left side. We routinely harvest a vascularized, pedicled nasoseptal flap [73] at the beginning of EEAs. Posterior septectomy is performed, followed by exposure of the sphenoid ostia bilaterally. A wide sphenoidotomy and bilateral posterior ethmoidectomies are then executed with subsequent removal of the entire sphenoid sinus mucosa. The sphenoid rostrum and septa are drilled flush to the sellar floor so that the nasoseptal flap can be positioned effectively at the end of the procedure. Identification of the critical neurovascular structures is confirmed with neuronavigation. Doppler ultrasound is utilized to confirm the position of the clinoidal segments of the internal carotid arteries. Exposure should be wide enough so the lateral opticocarotid recesses can be easily identified. The posterior ethmoidal arteries can be identified and coagulated for early tumor devascularization in cases of PS meningiomas.

Intracranial Approach

After full exposure of the sella turcica and the anterior skull base, the carotid and optic nerve protuberances as well as the medial and lateral optico-carotid recesses are typically visualized unless the sphenoid sinus is poorly pneumatized (Fig. 17.20). At this point, we routinely use a 30° angled endoscope looking superiorly for a more detailed view of the anterior cranial base. The TS, which is an elevated bony structure when seen from the transcranial route, is better described as a suprasellar notch when seen from the endoscopic endonasal perspective [66]. This structure lies deeper in the surgical field than the PS and is typically deeper than the sellar floor. Thus, we advocate a systematic approach to drilling and opening of the skull base starting with the sellar face, followed by drilling the PS, and lastly drilling the TS. This technique allows for a safer opening of the TS, avoiding injury of the superior intercavernous sinus, which can complicate the approach with excessive bleeding in early stages of the operation. Depending on how far the tumor extends inside the sella turcica, the opening of the sellar face may be limited to the superior half. In cases where the tumor extends inside the sella, the entire sellar face down to the floor may need to be opened. At the PS level, the opening can be tailored based on the extension of the tumor with assistance of the navigation system. The TS should be opened between the bilateral medial opticocarotid recesses. After the dura of the sella, PS, and TS is exposed, the superior intercavernous sinus is identified and coagulated with a bipolar suction coagulator device (Kirwan®, Marshfield, MA, USA) before opening the dura. Dura over the tumor is coagulated to devascularize the tumor, incised in a linear fashion, and then resected until most of the tumor surface is uncovered.

Tumor Resection

Tumor resection starts with internal debulking. After drilling of the skull base, coagulation and resection of the tumor dural attachment, and cauterization of the posterior ethmoidal arteries (in case of PS meningiomas), early devascularization of the tumor is achieved, which reduces bleeding during tumor debulking. In our experience, it is critical to perform an effective tumor debulking staying inside the tumor capsule in order to avoid injury to the nearby neurovascular structures, such as the optic apparatus and its arterial perforators, as well as ophthalmic and superior hypophyseal arteries. Various adjunct instruments including an ultrasonic aspirator can facilitate this part of the operation, especially in cases of firmer tumors. Following tumor debulking, dissection of the extracapsular arachnoidal plane is performed. Careful bimanual microsurgical technique is used, and the tumor is freed from the arachnoid adhesions and neurovascular structures.

Once complete intradural tumor removal is achieved and confirmed using an angled endoscope (typically using a 30° or 45° scope), we focus our attention on the extension of the tumor into the optic canals. The natural history of TS meningiomas is of early optic canal invasion, even in cases where it is not obviously present on MRI [74, 75]. Thus, opening of the bilateral medial optic canals is performed, with the bony opening during the approach and the dural opening of the orbital apices during the tumor resection. The canals are inspected for tumor remnants despite the findings on preoperative imaging and/or clinical symptom of visual loss. For PS meningiomas, frank invasion into the optic canals is much less common, and opening of the optic canals is only considered when there is frank invasion seen radiographically or intraoperatively. The EEA offers advantages compared to transcranial routes in this setting given that the majority of tumors invade the optic canals in its medial compartment. In order to avoid injury to the ophthalmic artery, which enters the optic canal inferomedially in relation to the optic nerve (Fig. 17.21), we open the optic canal from medial to lateral starting in its most superior aspect, as described by Attia et al. [57].

Reconstruction

As TS/PS meningiomas are intradural tumors, CSF leak is expected during tumor removal. We consistently reconstruct the skull base with a button graft of a suturable dural substitute

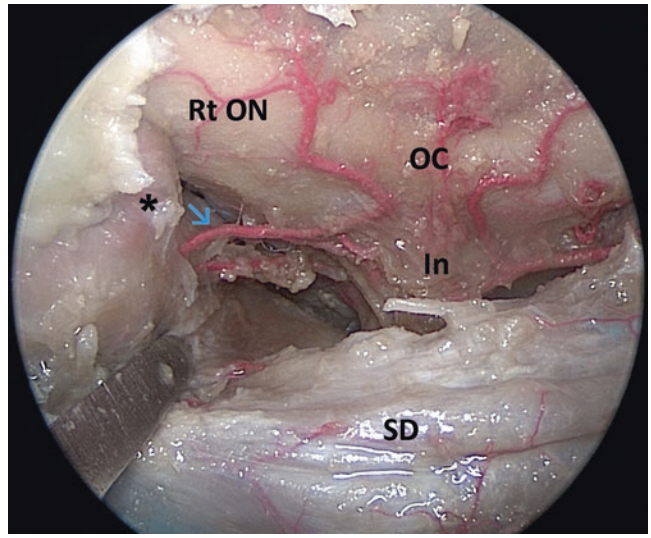

Fig. 17.21 Cadaver dissection image using a zero degree endoscope showing the origin of the right ophthalmic artery (*) and its relationship to the right optic nerve (Rt ON) as it enters the optic canal. The superior hypophyseal artery (*blue arrow*) is also appreciated arising in the medial surface of the supraclinoid internal carotid artery. Optic chiasm (OC), infundibulum (In), and sellar dura (SD) are also seen

placed as a subdural inlay and onlay dual-layered graft followed by the vascularized pedicled nasoseptal flap. The flap has to be in direct contact with the bone surrounding the skull base defect with the mucosa directed toward the nasal cavity. No gaps should exist between the flap and the skull base, hence the importance of drilling sphenoid septa and rostrum flush to the floor of the sella turcica. Following the placement of the flap, a layer of Surgicel Fibrillar® (Ethicon, Somerville, NJ, USA) is applied at the edges of the flap to help create fibrosis in the area, and tissue sealant is spread over the entire skull base. Lastly, absorbable nasal packing Nasopore® (Stryker, Kalamazoo, MI, USA) is positioned in the nasal cavity. A lumbar drain is usually kept for 3 days postoperatively, at a drainage rate of 5–10 ml/h, and removed after being successfully clamped for at least 12 h with no evidence of CSF rhinorrhea. If the patient develops CSF leak, we advocate surgical exploration and repair. Fig. 17.22a–i shows a case example of TS meningioma in which gross total resection was achieved using the EEA.

Anatomical Limitations

Anatomical limitations of the EEA for TS/PS meningiomas can be related to patient anatomy or the tumor itself. In regard to tumor characteristic, the main limitation is extension of tumor lateral to the carotid arteries and optic nerves. When tumors extend lateral to these structures and gross total resection is the goal of the operation, an anterolateral transcranial or supraorbital approach is indicated. This is also true when the tumor invades the optic canal in its lateral

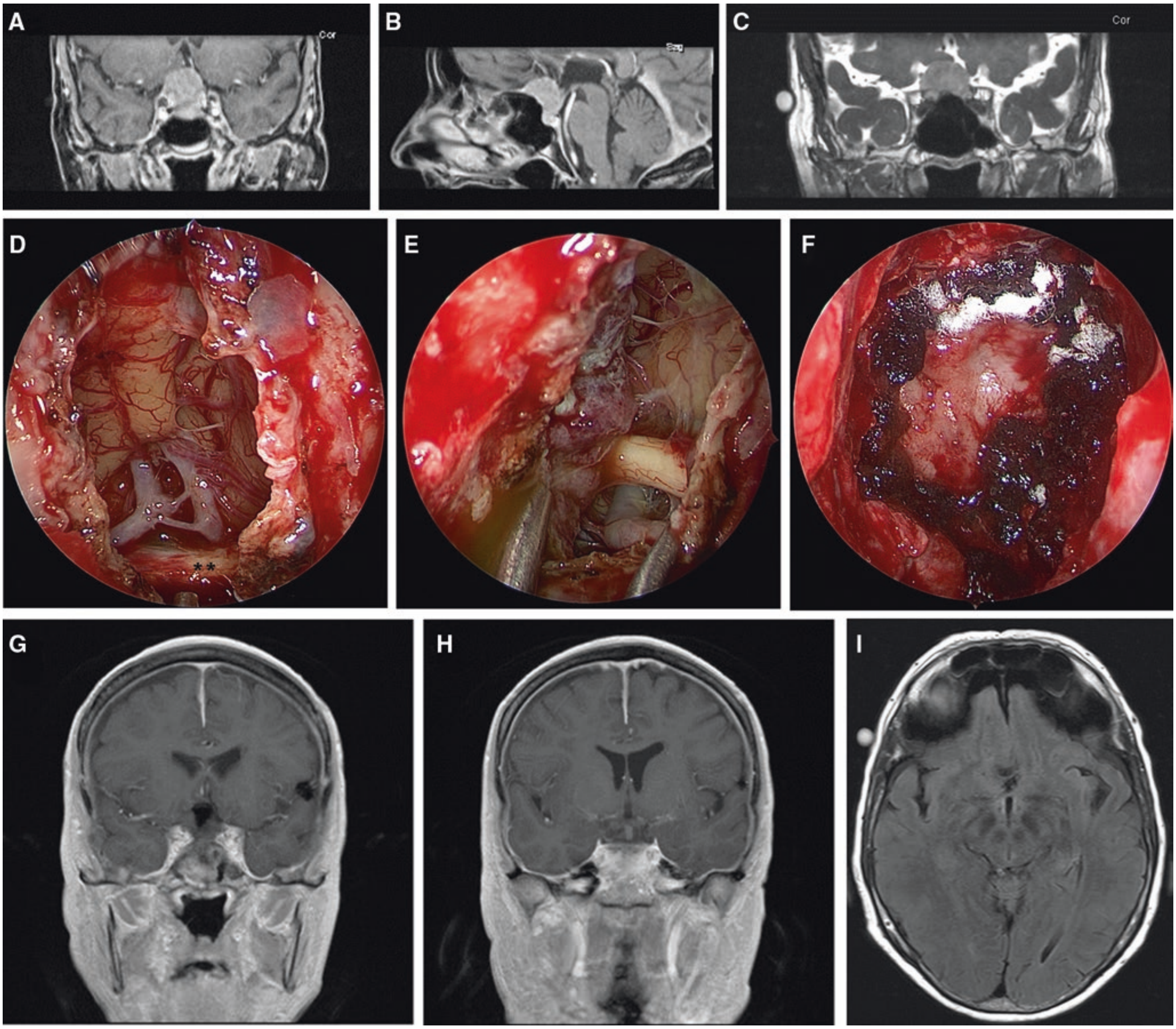

Fig. 17.22 (**a**–**i**) A 77-year-old female with right-side vision loss for 1 year. Coronal (**a**) and sagittal (**b**) T1 post-contrast-enhanced images and CISS coronal (**c**) images showing a tuberculum sellae meningioma extending into the sella turcica above the pituitary gland, displacing the infundibulum posteriorly and invading the optic canals. The tumor lies between the internal carotid arteries making it an ideal case for the EEA. Intraoperative pictures after removal of the tumor and decompression of the neurovascular structures (**d**–**f**). Of note a fenestrated anterior communicating artery complex and decompressed optic chiasm (**) are seen (**d**). Intradural angled view with a 70° endoscope showing tumor invading the optic canal on the right side (**e**). Reconstruction is performed with a vascularized nasal septal flap with Surgicel Fibrillar® (Ethicon, Somerville, NJ, USA) placed around the borders of the flap (**f**). Coronal T1 post-contrast-enhanced images (**g**, **h**) showing gross total tumor resection with preservation of the pituitary gland and infundibulum, decompression of the optic chiasm and axial FLAIR images (**i**) showing no evidence of frontal lobe injury due to retraction

compartment. On the other hand, in cases where a subtotal resection is acceptable and decompression of the optic nerves for improvement in visual function is the goal, the endonasal approach can still be used. Concerning patient anatomy, the pneumatization of the sphenoid sinus can affect the difficulty when performing this approach. A fully pneumatized, sellar-type sphenoid sinus facilitates the exposure. On the contrary, patients with pre-sellar- or conchal-type sphenoid sinus are challenging as the anatomy of critical structures like optic nerves and carotid arteries cannot be reliably identified [76]. In such situations, neuronavigation plays an especially important role during the approach. Another relevant anatomical feature is the distance between the cavernous carotid arteries. When the intercarotid distance is small, the approach and access to the suprasellar region can be difficult due to decreased surgical maneuverability.

Postoperative Care

Patients spend the first postoperative day in a monitored setting. Neurological status is assessed every 1–2 h. For TS meningiomas that are compressing the pituitary gland, fluid status is strictly monitored for diabetes insipidus. Urinary output is recorded hourly, and patients have full access to water at bedside. Sodium levels are assessed in the immediate postoperative period and the next morning after surgery. In cases of urinary output above 250 ml/h for two consecutive hours or over 500 ml/h, stat sodium levels are ordered. As long as the patient can keep up with water intake and the sodium levels are within the normal range, we do not initiate medical therapy for diabetes insipidus. In cases where the patient cannot keep up with water intake or when sodium levels rise above the normal range, we initiate medical therapy. Assessing the integrity of the thirst mechanism is useful in these cases to predict potential need for pharmaceutical intervention. Deep venous thrombosis prophylaxis consists of pneumatic squeezing boots and subcutaneous heparin starting on postoperative day 1. Antibiotics are continued for 24 h postoperatively or as long as a lumbar subarachnoid drain is in place. A brain MRI with and without contrast is obtained on the first postoperative day. Patients are seen in the rhinology clinic for nasal care on the first, third, and eighth week after surgery.

Surgical Outcomes

The EEA for resection of these suprasellar lesions has been increasingly advocated in select patient populations as a safe and effective alternative to open surgical approaches. While long-term data remain limited, several centers have reported positive outcomes using the EEA for TS and PS meningiomas. A selection of results from the larger series is summarized in Table 17.4. In a large retrospective series of 75 patients with TS/PS meningiomas treated at a single institution, Koutourousiou et al. [77] reported a Simpson Grade 1 resection at a rate of 76%, with factors limiting resection being tumor size, morphology, and relationship to vasculature. Notably, optic canal involvement was not reported. Recurrence was reported at 5.3% (mean follow-up 29 months). Patients with recurrence were managed with repeat EEA in one case, radiosurgery in another, and observation alone in two remaining cases. Other authors have reported similar rates of gross total resection (GTR), ranging from 76% to 86% [55, 77, 78], with a recent review of the literature reporting an overall GTR rate of 81% across 29 studies [79]. These results appear comparable to reported outcomes following transcranial surgery. In a meta-analysis of transcranial and transsphenoidal series for TS meningiomas, de Divitiis et al. reported a GTR rate of 87.6% and 93.1%, respectively, with no statistical difference between the two groups [55]. Notably, this study included both microsurgical and endoscopic transsphenoidal approaches.

While some authors have advocated the use of an EEA only for small to midsize tumors [80], others have failed to find a correlation between size and outcome. Khan et al. [78] reported an average TS/PS tumor volume of 11.92–12.02 cm^3 with no influence of tumor size on GTR. As endoscopic techniques and repair methods evolve, the feasibility of treating larger tumors appears to be increasing.

Complications and Visual Outcomes

The most commonly reported complication following EEA for TS/PS meningiomas is postoperative CSF leak, reported in 0–25.3% of patients [55, 57, 77–79]. While this rate may appear high, the routine use of a vascularized nasoseptal flap has been reported in multiple series to dramatically decrease the rate of postoperative leak, with a statistically significant reduction from 69.2% to 16.1% in one large series with use of a nasoseptal flap [77]. Similarly, Khan et al. reported a drop in CSF leak rate following EEA to anterior skull base meningiomas from 14.6% to 0% in recent years, largely owing to improvements in technique and closure [78]. Postoperative meningitis has been reported, which in most cases can be managed with antibiotics alone [77]. Less frequently reported complications include syndrome of inappropriate antidiuretic hormone secretion, seizure, and hemorrhage [77]. A low rate of permanent visual deterioration has been reported, with a range from 0% to 3.6% in the larger series [55, 77–79]. In their meta-analysis of transcranial series for TS meningiomas, de Divitiis et al. reported a 12.9% rate of worsened vision following surgery [55], with rates of deterioration as high as 33.3% in some series [81]. Other meta-analyses comparing EEA to open surgery for TS have shown a significantly higher rate of visual improvement in endoscopic cohorts, although the magnitude of this difference ranges widely from study to study [79].

Table 17.4 Selected outcomes reported in the literature following EEA for TS/PS

Study	Patient size	Mean follow-up time	Rate of GTR (%)	Recurrence rate	Rate of CSF leak (%)
De Divitiis et al. [55]	7	Not reported (3 weeks–20 months)	85.7	Not reported	28.6
Attia et al. [57]	7	20.9 months	85.7	Not reported	0
Koutourousiou et al. [77]	75	29 months	76	5.3%	25.3
Khan et al. [78]	20	51.5 months	85	10%	10

While there remains a paucity of data on EEA compared to transcranial outcomes for TS/PS meningiomas, the literature preliminarily suggests that EEA may offer improved visual outcomes with a trade-off of higher rates of CSF leak, although these rates are decreasing. However, given that larger tumor size is often thought to contribute most to visual deterioration, and that transcranial surgery is often used for larger tumors, further studies are needed to evaluate if these outcomes can be extrapolated regardless of tumor size.

Overall, the reported rates of GTR, visual improvement, and complications suggest that the EEA may be a safe and effective alternative to transcranial surgery. Increasing experience and advancement of endoscopic technique, including repair elements, are likely to further reduce the complication rate as this technology develops.

PEARLS
Exposure is critical. A wide sphenoidotomy with bilateral posterior ethmoidectomies provides clear exposure of both lateral optico-carotid recesses and increases surgical maneuverability.
Ensure that the rostrum of the sphenoid bone and intra-sphenoid septum are flushed to the sellar floor and that all overlying mucosa has been removed for an effective reconstruction with nasoseptal flap to minimize risk of postoperative CSF leak.
When performing the opening, first drill the sellar floor, followed by the planum sphenoidale, and then the tuberculum sellae in order to prevent premature dural violation and avoid injury of the superior intercavernous sinus.
Debulk the tumor first, prior to proceeding with the extracapsular arachnoidal dissection, in order to avoid injury to the surrounding neurovascular structures.

Commentary on Case Presentation in Section 17.1

Case Presentation

The patient is a 52-year-old female with a several-month history of blurry vision in the left eye. Physical examination shows superotemporal visual field cut with relative APD and mild visual acuity deficit (20/30) (see Fig. 17.1a–d).

Discussion

In our opinion, the present case is best approached via an expanded endoscopic endonasal trans-tuberculum/trans-planum approach. The clinical history, radiological features, and physical examination are consistent with a TS meningioma, presenting with asymmetric visual loss on the left side. In this case, the EEA offers a variety of advantages as discussed below:

- *Clinical features*: The natural history of TS meningiomas is to cause vision deficit by invading the optic canal either unilaterally or bilaterally. This is a young patient in her early fifties with a long period of life ahead of her. It is our opinion that the EEA allows for a more effective decompression of the medial aspect of the optic canal. We believe this is an advantage of the EEA, as in the majority of cases TS meningiomas invade the optic canal from the medial side. Of note, in TS meningiomas, we have routinely performed bilateral optic canal exploration, even if the patient does not complain of visual deficit. Thus, in this case, we would also open the optic canal on the right side, in respect of the natural history of this type of meningioma. Following the opening of the optic canal, the tumor is effectively dissected and removed.
- *Radiological and anatomical considerations*: The present case represents an ideal case for the EEA because the tumor is located in the midline and does not extend lateral to the carotid arteries. The anterior cerebral artery complex (A1, Acom, A2) is displaced posterosuperiorly without being encased by the tumor. There is an arachnoidal plane between the tumor and the brain, represented by a thin layer of CSF seen as T2 hyperintensity in the brain tumor interface which allows for a safe extracapsular dissection after effective tumor debulking. The pituitary gland and stalk are displaced posteroinferiorly and would be protected and not seen until the late stages of the operation. Additionally, the optic nerves are displaced laterally and also not encased by the tumor, which permits a safe dissection plane.
- *Technical aspects*: The EEA allows for early devascularization of the tumor by drilling the skull base (TS and PS) as well as coagulating the dural origin. With early devascularization, tumor debulking is facilitated due to decreased bleeding and easier identification of the neurovascular structures. By approaching the tumor endonasally, the working corridor is between the laterally displaced optic nerves without the need to cross the plane of both optic nerves. Finally, effective reconstruction technique with fascia lata, dural substitute, or abdominal fat graft followed by a pedicled nasoseptal flap allows a low rate of CSF leak postoperatively.

Summary

Endoscopic endonasal skull base surgery has evolved over the past years and has now become a safe and effective technique for dealing with skull base tumors. Specifically, PS and TS meningiomas are particularly well approached by this technique due to their intrinsic anatomical location, as midline tumors, and their relationship to the sphenoid sinus. Recent technological advancements in surgical instrumentation and the creation of multidisciplinary skull base teams composed of neurosurgeons and rhinologists have improved clinical outcomes and decreased postoperative complication rates for patient with PS/TS meningiomas treated via EEA. It is yet to be determined the optimal surgical approach to these pathologies, and, certainly, some of these tumors are better approached through a transcranial route as discussed previously. Given that a randomized controlled trial comparing transcranial and the EEA is very unlikely to be developed, the final answer in terms of the ideal approach might never be fully clarified. Lastly, comprehensive skull base centers have demonstrated the safety and efficacy of EEA to PS/TS meningiomas with resection rates, clinical outcome, and postoperative complications comparable to traditional transcranial series.

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17.5 Planum/Tuberculum Sella Meningiomas: Editors' Commentary

The pterional/frontotemporal craniotomies, with possible orbital or zygomatic osteotomy, are very familiar approaches to most neurosurgeons and have been utilized for several decades with good reported success. They most readily allow management of densely calcified tuberculum sella meningiomas and those with vascular encasement that may require repair in the chance of vessel injury. These approaches have set the bar for management of tuberculum sella meningiomas over the past several decades, and it is to these outcomes that other "less invasive" approaches are typically compared.

Keyhole approaches, such as the supraorbital approach, allow a low trajectory to the tumor with limited frontal lobe retraction similar to the traditional transcranial procedures without the need for a larger frontotemporal incision and manipulation of the temporalis muscle. The supraorbital approach can also allow access to superior extension of the tumor with the addition of a superior orbital rim osteotomy or endoscopic assistance. The intradural anatomy is similar to the pterional/frontotemporal approach and familiar to many surgeons. Drawbacks of the supraorbital approach include limited pericranium for cranial base repair and the possible need to transgress the frontal sinus with inherent risk of infection. Although infection with the supraorbital approach is rarely reported in literature, the cosmetic implications may be significant.

Endoscopic endonasal approaches to tuberculum sella meningiomas have become more common over the past decade. These procedures are performed by a neurosurgery and otolaryngology team at most experienced centers. Results in the literature for endonasal removal of well-selected tuberculum sella meningiomas have shown resection rates and visual outcomes comparable or better than the "open" approaches, respectively. However, there is a rather steep learning curve, and the CSF leak rates in some reports remain higher than the transcranial approaches.

The case presented in Sect. 17.1 (Fig. 17.1a–d) is an ideal tuberculum sella meningioma that could potentially be managed by either open, keyhole, or endonasal approaches. Certainly, the surgeon's experience and skill set play a major role. In addition, there are several factors and tumor characteristics that should be considered when selecting the approach: size, consistency and calcification, extension, vascular supply to the tumor, and intracranial vessel encasement. Additionally, the extent of cranial base invasion and the available options for cranial base repair should be considered, especially with reoperations.

Size of the tumor itself is not the main consideration in choice of approach but should be taken into account. Tumors with more superior extension require a lower and more inferior to superior-oriented trajectory for direct visualization of the upper aspect of the tumor. In these cases, the transcranial approaches often require superior orbital or orbitozygomatic osteotomy. The trajectory of the endonasal approach lends itself well to tuberculum sella meningiomas with superior extension. However, larger tumors tend to take more time for resection by the endonasal approach compared to open approaches, even in the hands of surgeons with extensive experience. Dense tumor consistency, particularly calcification, can limit and lengthen endoscopic removal of the tumor. Transcranial approaches may allow en bloc or large piecemeal removal for tuberculum sella meningiomas, whereas endonasal procedures are most effective with initial aggressive internal debulking followed by extracapsular dissection.

Tumor extension plays an important role in selecting the best approach to a tuberculum sella meningiomas. Some tuberculum sella tumors will have lateral extension beyond the optic nerves or encasement of the ICAs that could prohibit complete removal by an endonasal approach. These cases are likely better managed by supraorbital or frontotemporal transcranial approaches that may allow more circumferential access to the ICA and safer management of potential vascular injury. Anterior extension along the planum sphenoidale can be well managed endoscopically with minor modification of the approach and repair. However, if the anterior tail encroaches on the cribriform region, the endonasal approach risks anosmia. Posterior extension into the sella and along the diaphragma sella is readily accessible by an endonasal approach but is often more difficult to directly visualize well by the open or keyhole approaches without endoscopic assistance. Optic canal extension often involves the medial optic canal in the case of tuberculum sella meningiomas. For open and supraorbital keyhole approaches, it can be difficult to adequately access tumor in the medial optic canal, and sometimes this is not addressed until late in the case. Taking a contralateral transcranial approach is sometimes advocated; however, this does not solve the problem of bilateral medial optic canal involvement which is frequently present even when not obvious on preoperative imaging. It can be particularly difficult to access tumor involving both medial optic canals by the unilateral frontotemporal or supraorbital approaches. Alternatively, the endonasal approach readily allows access to both medial optic canals for early decompression of the optic nerves and a more thorough resection of these portions of the tumor.

Vascular considerations include both blood supply to the tumor and intracranial vessel encasement. The tumor blood supply is via the dura and ethmoidal arteries. Although this can be addressed through the transcranial procedures, certainly the endonasal approach has a distinct advantage of providing an early and thorough devascularization of the tumor by the nature of the approach. Intracranial vessel encasement must be recognized and carefully studied on preoperative imaging. Even when vessels seem encased, there is often the presence of a thin cuff of CSF along the vessel on T2-weighted imaging suggesting a preserved arachnoid plane. This will help facilitate removal of the tumor from the vessels which can be done via an open or endoscopic approach. Complete vascular encasement, vessel irregularity, prior radiotherapy, or dense calcification should dissuade the surgeon from selecting an endonasal approach as management of vascular injury and adequate vessel repair remain quite limited endoscopically despite significant technical advances in recent years.

Repair options for the possible cranial base defect must be considered preoperatively when selecting the approach to a tuberculum sella meningioma. Removal of cranial base involvement by the tumor and associated hyperostosis is necessary for complete resection and renders a cranial base defect that must be repaired to prevent CSF leakage and infection. The incision used for the pterional/frontotemporal approaches allows for harvest of a pedicled pericranial graft sufficient for repair of even very large cranial base defects. The endonasal approach can provide a thorough removal of the cranial base involved by the tumor and can effectively utilize a vascularized nasal septal flap repair of the cranial base defect. On the other hand, the supraorbital approach allows for a very limited amount of pericranium that is typically insufficient for repair of a defect at the sella or spheno-ethmoidal region of the cranial base.

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Suprasellar Craniopharyngiomas

18

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Contents

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18.1 Suprasellar Craniopharyngiomas: Editors' Introduction

Craniopharyngiomas are histologically benign World Health Organization (WHO) grade I intracranial tumors that originate from remnants of Rathke's pouch and can involve the sellar, parasellar, and suprasellar areas. Although craniopharyngiomas constitute less than 1% of all primary CNS tumors, they are the most common non-glial tumor in children [1]. The incidence of craniopharyngiomas is estimated at 0.13 per 100,000 person years, with a bimodal age distribution between 5–14 years and 65–74 years [2] and a higher prevalence in childhood. Two pathological types of craniopharyngiomas have been described. The adamantinomatous subtype is more common in children, frequently is cystic, and accounts for 5–10% of pediatric intracranial malignancies. The papillary subtype is more commonly solid with calcifications and occurs almost exclusively in adults.

Craniopharyngiomas are most commonly found in the suprasellar region but can occur along the entire length of the craniopharyngeal duct. Five basic growth patterns have been observed: infradiaphragmatic (sellar), subarachnoid extraventricular, and subpial intraventricular (third ventricle) based on the origin from infrasellar; infradiaphragmatic; transinfundibular; suprasellar; and subarachnoid subpial ventricular locations [3]. The extent of the tumor, coupled with the propensity for local invasion to surrounding critical neurovascular structures such as the hypothalamus, infundibulum, pituitary gland, optic chiasm, and carotid arteries, substantially increases the difficulty of achieving complete surgical resection. Thus, multimodal treatment including radiation and molecular-targeted agents may often be necessary for tumor control. In this chapter, authors will discuss both open and endoscopic approaches to the suprasellar region. In addition, each author will discuss the advantages and disadvantages of their approach to treat the case example below (Fig. 18.1a–c).

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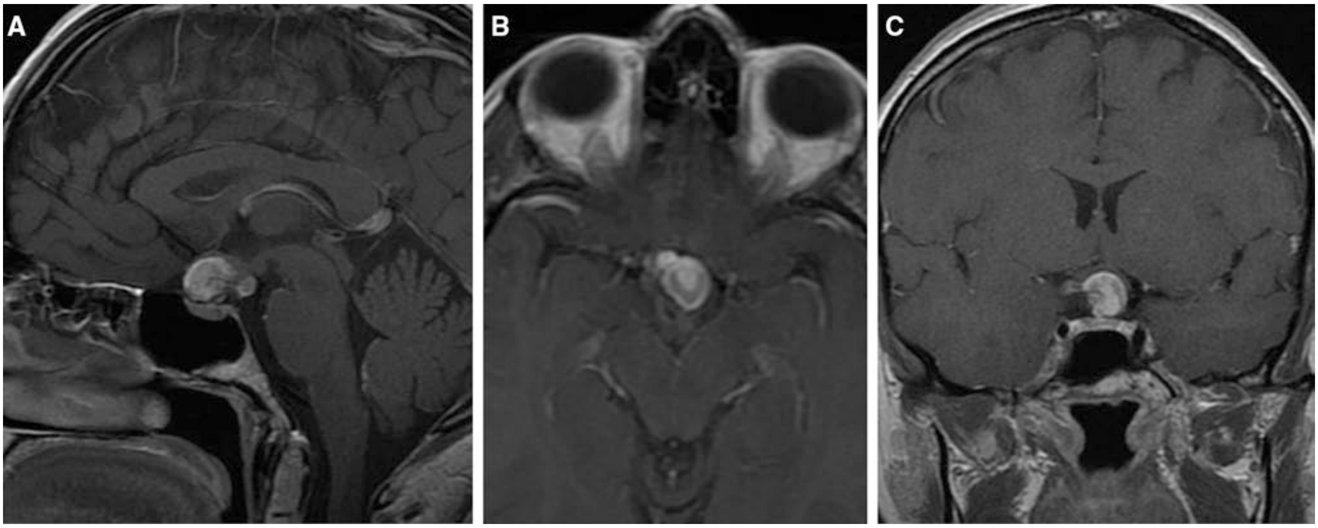

Fig. 18.1 (**a–c**) **CASE EXAMPLE**: suprasellar craniopharyngioma. A 24-year-old lady presents with mild bilateral visual blurriness. Examination: OS, 20/20 (corrected); Humphrey VF, mild arcuate superior and inferior temporal defects. OD, 20/20 (corrected); Humphrey VF, mild arcuate superior and inferior temporal defects. Rest of neurological exam normal. Laboratory workup: normal endocrine function

18.2 Suprasellar Craniopharyngiomas: Open Transcranial Approaches

Amol Raheja and William T. Couldwell

Introduction

Craniopharyngiomas are benign tumors that arise from remnants of Rathke's pouch. Despite their histologically benign nature, their management remains challenging because of the tumor's close proximity to vital neurovascular structures such as the optic apparatus, pituitary stalk, hypothalamus, and vessels of the circle of Willis. They are primarily located in the sellar-suprasellar region, but intraventricular and multicompartmental extensions into the anterior, middle, and posterior cranial fossae are also frequently seen.

The constraints imposed by nearby critical neurovascular structures in the suprasellar region have led to the development of several approaches, including transcranial and transsphenoidal [4–6]. Transcranial approaches are further subdivided into open and minimally invasive keyhole approaches, while transsphenoidal procedures can be either microscopic or endoscopic. This chapter will discuss the current available options for open transcranial approaches to craniopharyngioma tumors and consider their relative merits and limitations, patient selection criteria, perioperative care, complication avoidance, management principles, and surgical outcomes. We will discuss in detail the pros and cons of open surgical technique compared to endonasal technique and discuss the relative merits/demerits of each open technique over another using the illustrative case example.

Open Transcranial Approaches

There are four primary open transcranial approaches to craniopharyngioma tumors: the pterional/orbitozygomatic, the subfrontal/transbasal, the subtemporal approach alone or in combination with a transpetrosal approach, and the interhemispheric transcallosal/transcortical transventricular approaches. The use of extended skull base modifications such as the orbitozygomatic, transbasal, and transpetrous approaches helps to attain a much flatter and inferior-to-superior operative trajectory to more extensive skull base tumors while reducing the amount of brain retraction required. Each of the techniques has its own set of merits and demerits, and the appropriate surgical corridor must be chosen according to the individual tumor and patient characteristics. Many of these approaches require preoperative planning for adequate intraoperative brain relaxation. This goal can be achieved via intraoperative elevation of the head (reverse Trendelenburg position), mannitol or hypertonic saline bolus infusion, lumbar drain placement, ventricular tapping, and early release of cerebrospinal fluid through the sylvian and basal cisterns. One or more of these modalities can be used to achieve adequate brain relaxation depending upon the tumor morphology, patient symptomatology, and ventricular status. The principles of tumor dissection and craniopharyngioma removal include maintaining the arachnoid plane to reduce neurovascular damage, progressive tumor debulking, precise sharp microsurgical extracapsular dissection, and assiduous preservation of neurovascular structures.

Pterional/Orbitozygomatic Approach

The frontotemporal pterional approach, as advocated by Yasargil, uses the parachiasmal spaces (interoptic, opticocarotid, and carotid-oculomotor/carotid-tentorial corridors) and (less commonly) the triangle superior to the carotid bifurcation to access the suprasellar cistern. Performing an additional orbitozygomatic osteotomy can augment the operative access to the interpeduncular, parasellar, and posterior/superior third ventricular regions over and beyond the usual suprasellar access. Drilling of the anterior clinoid process along with optic nerve decompression either extradurally or intradurally helps to alleviate any mass effect on the optic nerve and increase the maneuverability of surgical instruments between the interoptic and opticocarotid corridors for more radical resection of tumor. Although the pterional approach is a versatile skull base approach that provides the shortest distance and most direct transcranial route to the suprasellar region and can be used for a wide spectrum of craniopharyngioma tumors, its best indication is in the resection of prechiasmatic tumors with a secondarily postfixed chiasm. Tumors with large retrochiasmatic and intraventricular extensions may be better managed with other approaches, although many such cases can also be managed by adding a lamina terminalis corridor through the standard pterional craniotomy for better surgical access. The primary disadvantages of this approach include limited access, poor visualization of the contralateral opticocarotid triangle, ipsilateral infrachiasmatic and hypothalamic surfaces and retrocarotid space, and the need for optic nerve/chiasm manipulation to gain access to the suprasellar and interpeduncular regions.

Complications associated with this approach for craniopharyngioma include frontalis palsy if the frontalis facial nerve branches are injured with the craniotomy approach, visual deterioration due to manipulation of the optic apparatus, and vascular injury to the ipsilateral carotid or its branches. Occasionally, especially in children, fusiform dilatation of the carotid may develop after manipulation of the vessel during tumor dissection.

Subfrontal/Transbasal Approach

The subfrontal approach with bifrontal/unilateral craniotomy, as popularized by Tessier et al. and Derome et al., primarily relies on the interoptic, lamina terminalis, and opticocarotid corridors for suprasellar and third ventricular access. As opposed to the pterional/anterolateral approach, the primary advantage for this approach is its midline anterior surgical trajectory, which provides a direct and straight access to the prechiasmatic space and lamina terminalis corridor and offers visualization of the bilateral opticocarotid cisterns. It also provides excellent visualization of both walls of the third ventricle and hypothalamus through the translamina terminalis corridor; however, both the pterional and subfrontal approaches have an inherent limitation of accessing the retrochiasmatic space. If the lamina terminalis approach is chosen to approach tumors with third ventricular extension, a midline transbasal approach offers the optimal trajectory. The presence of a primary or secondary prefixed chiasm becomes a relative contraindication for subfrontal approach if the lamina terminalis approach is not added. The transbasal approach involves the addition of a bilateral orbitotomy and drilling of the crista galli to the bifrontal craniotomy so as to provide a much more inferior surgical trajectory to access craniopharyngiomas with significant third ventricular extension. If the prechiasmatic space is still narrow despite adequate transbasal exposure and the tumor extends inferiorly into the sella turcica, the working channel can be further expanded by drilling the anterior wall of the sella turcica and tuberculum and planum sphenoidale to allow tumor resection. This transsphenoidal transsellar variant of the transbasal approach is associated with higher incidence of cerebrospinal fluid (CSF) rhinorrhea and requires meticulous skull base reconstruction. These approaches can utilize both subfrontal and basal interhemispheric access routes to the suprasellar region. The disadvantage of the subfrontal route as compared with the basal interhemispheric approach is the risk of retraction injury to the bilateral frontal lobes and olfactory tracts; however, if the olfactory tracts are carefully and meticulously dissected from the frontal lobes, a wide operative exposure can be achieved safely, and lateral extensions of tumor can be handled. On the contrary, the basal interhemispheric route provides a relatively narrow operative corridor, requires interhemispheric brain retraction, and has difficulty accessing lateral extensions of the tumor, but it is associated with a lower incidence of olfactory tract injury and risk of CSF rhinorrhea. Complications pertinent to these approaches include anosmia/hyposmia, bifrontal contusions, venous infarctions, and CSF rhinorrhea if the frontal sinus is violated. Additional late complications (mucocele) can be associated with frontal sinus violation that is not addressed properly with the approach.

Subtemporal/Transpetrosal Approach

Given the limitations of the anterior and anterolateral skull base approaches to access retrochiasmatic craniopharyngiomas, Hakuba et al. and Al-Mefty et al. pioneered the subtemporal transtentorial posterior transpetrosal (presigmoid translabyrinthine) approach for such lesions. This posterolateral approach provides a caudocranial operative corridor, as opposed to the craniocaudal trajectory provided by the anterior and anterolateral skull base approaches. The primary advantage is that it allows direct visualization of the infrachiasmatic surface, pituitary stalk, and hypothalamus, thereby allowing for dissection of the upper pole of the tumor from these vital neural structures. It also offers lower risk to anterior circulation perforators, which are more often encountered with anterior approaches; however, it has the limitation of manipulating the vital neurovascular contents of the ambient cistern (oculomotor nerve, trochlear nerve, second segment of posterior cerebral artery, and posterior communicating artery along with their perforating branches). In addition, this approach also carries the risk of temporal contusion due to prolonged retraction of the temporal lobe, seizures, speech disturbances, and vein of Labbé injury-associated venous infarct. With this approach, the posterior communicating artery may be in the corridor; some authors have described division of this artery to enhance the trajectory to the tumor.

Interhemispheric Transcallosal/Transcortical Transventricular Approach

Unlike the other approaches to craniopharyngiomas described earlier, the interhemispheric transcallosal and transcortical transventricular approaches do not employ skull base corridors but rather provide a vertical trajectory from a calvarial aspect. They are essentially reserved for primarily intraventricular lesions with large superior extensions along the foramen of Monro with or without accompanying obstructive hydrocephalus. It is vital to differentiate the primary intraventricular lesions from tumors invading the third ventricle floor secondarily from the suprasellar region, because the former would indicate a transcallosal/transcortical approach while the latter requires a skull base approach. The senior author pays particular attention to the pituitary stalk in this differentiation. Tumors that arise wholly within the third ventricle do not deviate from the pituitary stalk, which may be foreshortened by the mass from above, but is not deviated laterally. Suprasellar tumors will deviate from the pituitary stalk. Therefore, careful preoperative assessment of radiological imaging and appropriate surgical planning is mandatory to preserve the hypothalamus and third ventricular floor. The choice between the transcallosal and transcortical approaches is based primarily on the degree of ventricular dilation, venous drainage anatomy into the superior sagittal sinus, tumor growth pattern, and surgeon's preference. The interhemispheric transcallosal approach offers the advantage of accessing the tumor from the midline and does not require enlargement of the ventricles, whereas the latter has the potential benefits of having lower risk of disconnection syndrome from corpus callosotomy and injury to pericallosal/callosomarginal arteries. The use of intraoperative neuronavigation can be handy for these approaches to better delineate the operative trajectory and accordingly the skin incision and craniotomy. Once the lateral ventricle is entered and the laterality is confirmed using the orientation of the choroid plexus, foramen of Monro, and thalamostriate vein, the third ventricle can be accessed using the transforaminal, interforniceal, suprachoroidal, or subchoroidal corridors. Care has to be taken to avoid iatrogenic forniceal damage and injury to the thalamostriate vein to prevent any memory deficits and postoperative limb weakness, respectively. There is a risk of retraction injury to the frontal lobe and iatrogenic injury to the superior sagittal sinus along with potential venous air embolism with the transcallosal approach. The primary disadvantages of these approaches include long operative distance, putting the hypothalamus and pituitary stalk at risk during tumor dissection, limited access to the anteroinferior aspect of the third ventricular and lateral sellar lesions, and postoperative tendency toward ventricular inflammation and obstructive hydrocephalus.

Surgical Outcomes

Gross Total Versus Subtotal Resection and Adjuvant Radiotherapy

The conventional strategy for craniopharyngioma surgery aims at gross total resection (GTR) because the benign histopathology of these lesions potentially offers the lowest chance of tumor recurrence with complete removal. The factors governing the aggressive resectability of a craniopharyngioma include the presence of preoperative hypothalamic disturbance, evidence of hypothalamic involvement by the tumor on preoperative imaging or intraoperative dissection, intraoperative adherence of tumor to the floor of the third ventricle, vascular encasement, and presence of dense peripheral calcifications stuck to vital neurovascular structures. Radical resection is associated with increased morbidity associated with hypothalamic dysfunction, visual deterioration, and major vessel injury, which may also translate into increased mortality and reduced overall survival. Recent studies have validated that long-term outcomes (progression-free and overall survival) in patients with GTR are similar to those receiving partial resection with adjuvant radiation. Because of these potential sequelae of radical tumor resection, modern surgical strategy takes into account the functional outcome of the surgical resection and emphasizes subtotal resection of the lesion/maximally safe resection and using adjuvant modalities such as radiotherapy to control the residual tumor progression and maintain a good quality of life. It is important to understand that treatment strategy needs to be tailored according to individual case-to-case basis, realistically matching the patient's expectations and optimal surgical outcome. For example, the risk of radiation-associated neurocognitive disturbance, necrosis, arteritis, and secondary malignancy is of paramount concern for pediatric patients.

Pituitary Stalk Preservation Versus Sacrifice

Craniopharyngiomas are often intimately related to the pituitary stalk because of its origin from remnants of Rathke's pouch. Tumor adherence to the pituitary stalk often precludes radical resection without sectioning the pituitary stalk. There are two schools of thought about whether to preserve the pituitary stalk during craniopharyngioma resection. The first presumes that the pituitary stalk is essential to maintaining optimal posterior pituitary function, so near-total resection of tumor is the goal, and tumor adherent to the stalk is left behind. Subsequently, the residual tumor can be either monitored with close radiological surveillance or given up-front adjuvant radiotherapy. Various studies have demonstrated no correlation of stalk preservation with recurrence-free survival rates, validating this surgical strategy. Jung et al. [7] highlighted another important rationale for stalk preservation: its preservation also increases the likelihood of maintaining intact anterior pituitary function. Honegger et al. noted that the attempt to preserve the stalk is a time-consuming and sometimes demanding effort, but it is rewarded with improved endocrinological results. The school of thought regarding stalk resection argues that sacrificing the pituitary stalk during craniopharyngioma resection is a small price to pay for achieving GTR of a benign tumor. Adherents cite strong evidence from the literature supporting the fact that high rates of diabetes insipidus are seen with radical resection of craniopharyngioma, irrespective of the pituitary stalk integrity. Therefore, attempted stalk preservation should not preclude GTR. In these cases, care has to be taken to section the stalk as distal to the hypothalamus as possible without compromising the negative margins, so as to preserve as much antidiuretic hormone production as possible, despite permanent impairment of anterior pituitary function. It is also important to emphasize that preservation of the pituitary stalk does not imply preservation of pituitary function.

Transcranial Approach Outcomes

In a meta-analysis of surgical management of craniopharyngiomas in children, Elliot et al. [8] included 2955 patients operated via transcranial approaches. In these patients, the average GTR rate was 60.9%, and tumor recurrence rate after GTR was 17.6%. The operative mortality was 2.6% and iatrogenic neurological morbidity was 9.4%. Overall, the incidence of postoperative diabetes insipidus (DI), vision improvement, visual deterioration, obesity/hyperphagia, and overall survival were 69.1%, 47.7%, 13%, 32.2%, and 90.3%, respectively.

Transcranial Versus Endoscopic Endonasal Transsphenoidal Approaches

Many studies have attempted to compare the conventional and the newer minimally invasive techniques based on the extent of resection, recurrence rates, and complication profiles. The primary caveats to this comparison include heterogeneity of tumor characteristics (size, location, extent, and neurovascular adhesions), progression of clinical symptomatology, surgical experience, aggressiveness of surgical resection, sample size, and duration of follow-up to assess recurrence. There is increasing experience with using the endoscopic endonasal approach. It has some inherent advantages, largely with the trajectory of approach. It enables direct visualization of tumors growing in a superior direction, with extension directly into the third ventricle. A recent meta-analysis by Elliott et al. [8] compared the transcranial and transsphenoidal approaches for surgical management of craniopharyngiomas in children. A total of 2955 patients operated via transcranial and 373 patients operated via transsphenoidal routes were included. The authors concluded that directly comparing outcomes after the two approaches for pediatric craniopharyngiomas does not appear valid. Baseline differences in patients who underwent each approach create selection bias that may explain the improved rates of disease control and lower morbidity of transsphenoidal resection. It is also pertinent to understand that transsphenoidal approaches are being increasingly used primarily for smaller intrasellar tumors as compared with transcranial approaches, which are often used for much larger tumors with significant suprasellar and parasellar components, those with significant peripheral calcification, and those that engulf vascular structures. In addition, the data for such minimally invasive approaches are limited and have shorter follow-up duration as compared with traditional transcranial approaches.

PEARLS
Anterior (subfrontal) and anterolateral (pterional) skull base approaches are suitable for prechiasmatic craniopharyngioma tumors with limited intraventricular and retrochiasmatic extension.
Augmentation of standard anterior/anterolateral skull base approaches with orbitozygomatic and transbasal extensions may facilitate removal of even larger tumors with significant intraventricular and retrochiasmatic extensions.
Posterolateral (presigmoid translabyrinthine) approach provides a viable alternative for extensive retrochiasmatic lesions and residual tumors along posterior aspect of optic apparatus, which are otherwise not accessible by conventional anterior and anterolateral approaches.
Interhemispheric transcallosal/transcortical transventricular approaches are primarily reserved for purely intraventricular craniopharyngiomas where the bulk of tumor is above the floor of third ventricle.

Commentary on Case Presentation in Section 18.1

Case Presentation

A 24-year-old woman presents with mild bilateral visual blurriness, and the examination reveals OS, 20/20 (corrected); Humphrey VF, mild arcuate superior and inferior temporal defects, and OD, 20/20 (corrected); Humphrey VF, mild arcuate superior and inferior temporal defects. The rest of her neurological examination is within normal limits. The patient's endocrine function is within normal limits. Preoperative gadolinium-enhanced MRI demonstrates a heterogeneously enhancing suprasellar region in close proximity to the pituitary stalk and floor of the third ventricle (see Fig. 18.1). The lesion is splaying and compressing the optic chiasm (left > right side). The primary radiological differential is suprasellar subchiasmatic craniopharyngioma. Ideally, further imaging in the form of computed tomography and coronal T2/FLAIR MR imaging should be evaluated preoperatively to assess the status of peripheral calcification and better delineate for the presence of any radiological hypothalamic involvement, respectively, which might have a bearing on our surgical strategy.

Discussion

Based on the clinical, endocrinological, and radiological information, we infer that the lesion is presumably a moderate-sized suprasellar subchiasmatic craniopharyngioma (Yasargil type C, Puget grade 0, Kassam infundibular type). The lesion does not seem to have any sellar extension as evident by the normal-sized sella turcica. Therefore, endonasal endoscopic TS approach seems inappropriate for the surgical access to this tumor, as it will require pituitary transposition, putting anterior pituitary function at risk. Also, because the sella is not dilated, the surgical corridor for tumor access via a transsphenoidal approach will be quite narrow. A lesser concern is the higher CSF leak associated with the transsphenoidal approach, which may translate into poor patient outcome. The chiasm appears to be just anterosuperior to the lesion and seems to be slightly prefixed, due to the bulk of tumor pushing and splaying it, but there is a reasonable interoptic corridor to this moderate-sized tumor for it to be accessed through both anterior and anterolateral skull base approaches. Because the lesion is located slightly eccentrically toward the left side, performing a unilateral (right-sided) pterional approach would be a reasonable first-line strategy. It will provide us with the benefit of both transsylvian and unilateral subfrontal working corridors for safe access to tumor. In addition to the use of the interoptic corridor primarily, the opticocarotid corridor may also be instrumental for complete resection of tumor safely. We would access this tumor through the right side and not the left side because the oblique field of view from the right side would help us to clearly define the tumor located at the undersurface of left optic apparatus. The vision is affected to the same extent bilaterally so the side of surgical access is not governed by vision in this patient. Had the vision been affected more on the left side, using a left pterional approach would have given us the option of performing ipsilateral optic canal deroofing for better maneuverability of surgical instruments around the optic apparatus and safe resection of the tumor.

Bifrontal craniotomy using anterior subfrontal/interhemispheric approach is another reasonable option and can also be performed in this case depending upon surgeon's preference; however, retraction of the bilateral frontal lobes has a potential for damage to the bilateral olfactory tracts. Had there been significant midline intraventricular extension, this approach would have been the first choice, with a lamina terminalis approach. A posterolateral skull base approach via subtemporal transtentorial posterior transpetrosal (presigmoid translabyrinthine) approach would be our second-line strategy for resection of any tumor left behind from first surgery. Because this approach provides a caudocranial access route and excellent view of the retrochiasmatic region, it can provide the advantage of removal of any residual lesion under direct vision, ensuring safe resection and optimal outcome. This approach has more morbidity than a frontotemporal approach. Lastly, because there does not seem to be any apparent tumor within the third ventricle, interhemispheric transcallosal and transcortical transventricular approaches are not approaches of choice for this case. Another important aspect of this case is the radicality of resection and sacrifice of infundibulum if deemed necessary for radical resection of craniopharyngioma. Considering the normal endocrinological status of the patient and infundibular type of the tumor, the pros and cons of stalk preservation must be discussed in detail with the patient before proceeding with our final surgical planning. Intraoperative adherence of tumor tissue to the infundibulum will also govern our resection strategy and needs to be tailored according to patient's expectations.

Acknowledgments We thank Kristin Kraus, MSc, our medical editor, for her contribution to manuscript editing.

18.3 Suprasellar Craniopharyngiomas: "Eyebrow" Supraorbital Craniotomy Approach

Garni Barkhoudarian and Daniel F. Kelly

Introduction

Craniopharyngiomas have been approached surgically via a variety of surgical corridors. Parameters evaluated for surgical approach selection include tumor location relative to adjacent neurovascular structures, prior treatments, patient's neurological and endocrinological status, and surgeon's experience with each approach. The supraorbital transciliary (eyebrow) or transpalpebral (eyelid) craniotomy is a versatile approach to the parasellar region and is a preferred transcranial approach utilized at our institution for a minority of select craniopharyngiomas.

Patient Selection

The primary factor in surgical approach selection for craniopharyngiomas is the anatomical relationship with the optic chiasm. Most de novo craniopharyngiomas are retrochiasmal in location with their long axis along the sinonasal-sellar-hypothalamic corridor and are best approached via an endonasal endoscopic route along the undersurface of the chiasm. For such retrochiasmal craniopharyngiomas, most anterior transcranial approaches will require significant manipulation of the optic chiasm, nerves, and tracts, thereby increasing the risks of vision loss and vascular injury particularly to the superior hypophyseal arteries (Fig. 18.2), as well as limiting the ability to achieve a safe gross total resection. However, a minority of craniopharyngiomas are suprachiasmatic, either extending directly above the chiasm, anterior to the chasm, or lateral to the chiasm, while some are completely intraventricular and may thus be more safely approached via a transcranial route.

Other important considerations in determining the surgical approach include tumor consistency, prior surgery or radiation, vision, and endocrine function. For example, in many cystic recurrent craniopharyngiomas that were originally approached from the endonasal route, a transcranial route may provide excellent safe access for cyst drainage without the potential additional morbidity of reopening a prior skull base reconstruction and nasal-septal flap via the endonasal route. Thus, the supraorbital approach is ideal for supraoptic and preoptic tumors, as the optic apparatus acts as a barrier to this region from an endonasal or transpetrous approach and is particularly appropriate for recurrent cystic craniopharyngiomas with accessible cystic components in which the main goal is cyst drainage and fenestration [9].

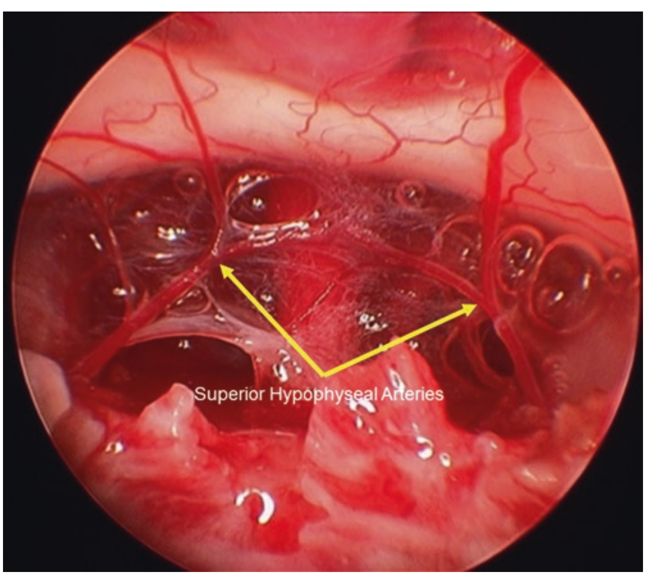

Fig. 18.2 Superior hypophyseal arteries visible via an endoscopic endonasal approach. Note the branches to both the pituitary infundibulum and the optic nerves

Index Case

Commentary on Case Presentation in Section 18.1

Case Presentation

A 24-year-old lady presents with mild bilateral visual blurriness. Examination: OS, 20/20 (corrected), Humphrey VF, mild arcuate superior and inferior temporal defects; and OD, 20/20 (corrected), Humphrey VF, mild arcuate superior and inferior temporal defects. The rest of neurological exam is normal. Laboratory work-up revealed: normal endocrine function (see Fig. 18.1).

Discussion

The MRI of the index craniopharyngioma patient demonstrates a suprasellar, supraglandular tumor. This lesion is heterogeneous in nature and is consistent with a craniopharyngioma. Though there are numerous surgical approaches to this tumor for resection, specific aspects of this lesion make it ideal for the supraorbital eyebrow approach.

- The primary determinant to differentiate between endonasal and supraorbital approaches is the location of the optic chiasm. Retrochiasmal tumors are better accessed from an endonasal approach as the surgical trajectory is ideal, and the optic chiasm would be obstructing the supraorbital surgical corridor. However, this tumor is subchiasmal in location, providing an adequate window beneath the optic chiasm for tumor resection (Fig. 18.3). In many cases, the optic chiasm is not well visualized, particularly in the sagittal plane. However, the anterior communicating artery (AComm) is a good surrogate marker of the optic chiasm as this relationship is not distorted unless the pathology is within the optic apparatus itself (e.g., optic nerve glioma). Additionally, this tumor is not in the third ventricle, hence a trans-lamina terminalis approach would not be feasible here.
- A secondary determinant is the relation of the tumor and the tuberculum sella. Most craniopharyngiomas reside in both the sellar and suprasellar regions (though typically

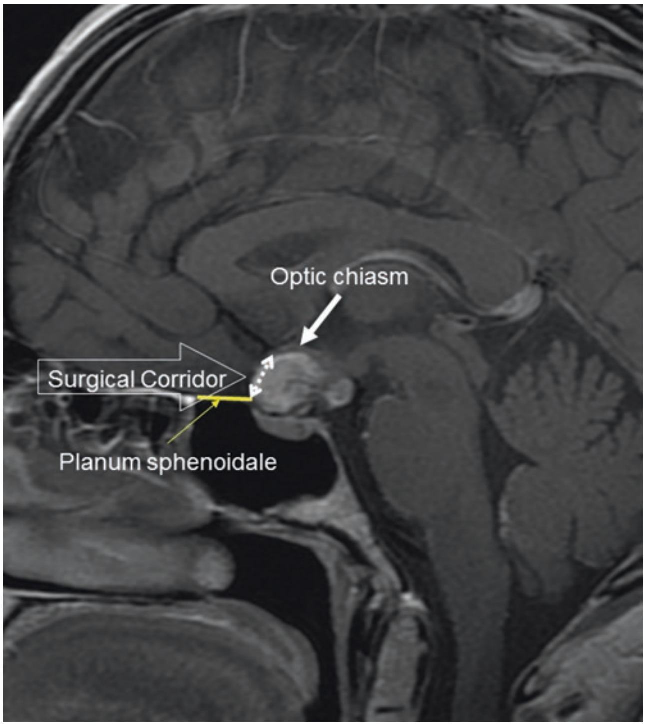

Fig. 18.3 The surgical corridor of the supraorbital approach runs along the planum sphenoidale. The optic chiasm is visible and abutted by the AComm artery. The optic apparatus is displaced superior, resulting in a working corridor (*dotted line*) between the tuberculum sella and optic chiasm

supradiaphragmatic). A high-riding tuberculum sella will result in a blind spot of line of sight in the sellar fossa. This lack of visualization can compromise the dissection of the tumor off the superior hypophyseal arteries and the carotid arteries. However, this can be overcome in some patients by the use of neuroendoscopy and bimanual microsurgical dissection. This tumor is situated just below the plane of the planum sphenoidale/tuberculum sella, allowing for adequate visualization and instrument access with direct line of sight with the microscope alone.

- Relative features that can benefit the supraorbital approach but are not critical include the relationship with the infundibulum as well as the dorsum sella. This tumor is predominantly anterior to the infundibulum (type I), though it does appear to be wrapping around it as well [10]. Hence, there is the possibility to preserve pituitary structure, which may need to be incised via an endonasal approach. Conversely, the technique of incising the pituitary gland has been shown to be safe and does not cause increased pituitary dysfunction [11].

This tumor sits atop the dorsum sella with a subtle extension beyond it. The supraorbital approach offers the ability to directly view this region. For tumors with more significant retrodorsal extension, the use of neuroendoscopy can help visualize the relationship of the tumor and the mesencephalic/pontine structures. Nevertheless, the endonasal approach can access this region via gland manipulation/translocation and drilling of the dorsum sella.

- Potential additional features that are not explicit in the imaging provided include the patient's pituitary hormonal function, visual function, frontal sinus anatomy, age, and comorbidities. Often, patients with craniopharyngiomas have some level of hypopituitarism (although diabetes insipidus is not a common preoperative finding). In the setting of panhypopituitarism, the infundibulum may need to be sacrificed to help facilitate dissection. This is avoided when possible in the attempt to preserve pituitary function. A contemporary series noted 20% of patients had improved postoperative pituitary function after tumor resection [12].
- A typical presentation with large suprasellar craniopharyngiomas is bitemporal hemianopsia. If vision loss is significant, an approach that would minimize optic apparatus manipulation (such as the endonasal approach) may be beneficial. Nevertheless, the small case series of the supraorbital approach demonstrate vision preservation in the majority of patients.
- Often, the craniotomy exposed with the supraorbital eyebrow approach is lateral to the frontal sinus. However, some patients have enlarged and over-pneumatized frontal sinuses. This can be a conduit of postoperative cerebrospinal fluid (CSF) rhinorrhea. This can be prevented with fat graft occlusion of the sinus. However, a very large frontal sinus may be a relative contraindication to this approach if alternative approaches can provide comparable tumor resection.
- Ultimately, patient age and comorbidities can contribute to the approach selection. The approach and closure of the supraorbital eyebrow craniotomy takes about 30–45 min in our experience. However, the endonasal approach can add additional time (often two- to threefold longer) under anesthesia, particularly with the elevation of a nasoseptal flap, securing the reverse flap and gasket seal buttress closure. Additionally, patients with obesity, sleep apnea, or chronic obstructive pulmonary disease (COPD) may have a higher risk of CSF rhinorrhea with the endonasal approach [13–15].

Surgical Technique

When deciding which approach to utilize for a craniopharyngioma in the setting of recurrent or postradiation tumors, a thorough understanding of the vasculature is necessary. Hence, a preoperative CT angiogram (CTA) is helpful to study the relationship of the carotid artery and its branches

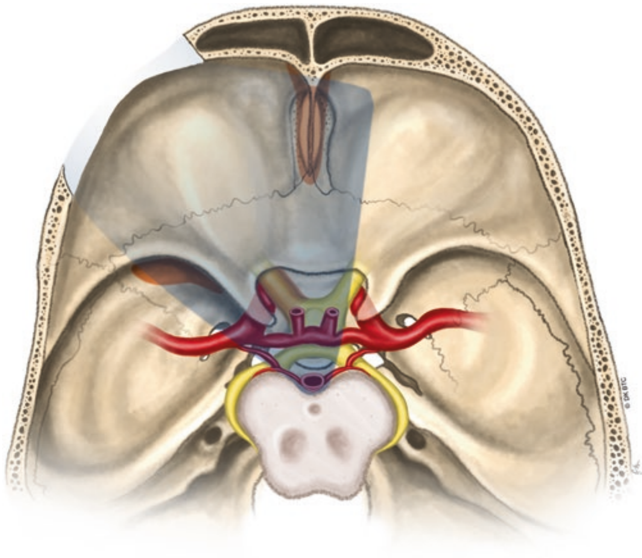

Fig. 18.4 The working area of the supraorbital craniotomy is denoted with the *blue shade*. Relative "blind spots" are denoted with orange shades and include the anterior medial middle fossa, olfactory grooves, sellar fossa, and inferior aspect of the ipsilateral optic nerve (Courtesy of D.F. Kelly Neurological, Inc.)

with the tumor. This can be fused with the preoperative MRI and used for operative neuronavigation. The frontal sinuses are assessed, and, if sinus entry is anticipated, the patient is advised that an abdominal fat graft harvest would be necessary to augment the surgical closure.

The side of the approach is determined by the anatomy of the tumor and its relation with the optic apparatus. A "blind spot" of the supraorbital approach is the region inferior to the ipsilateral optic nerve (Fig. 18.4). Hence, the side with the least volume of tumor beneath the ipsilateral optic nerve is chosen for the approach. If this is not a consideration, then the side with the smaller frontal sinus is chosen. Typical medical and cardiac assessments are conducted to ensure that the patient is optimized for general anesthesia.

Adjuncts used in the operating suite are planned prospectively. These include neuronavigation, neuromonitoring, micro-Doppler probe, and surgical endoscopes. Neuromonitoring typically includes somatosensory evoked potentials (SSEPs) to detect early vascular ischemia. In cases with cavernous sinus or brainstem involvement, monitoring of cranial nerves III and VI is also performed with direct intraoperative nerve stimulation and EMG. The micro-Doppler probe is particularly helpful in identifying and dissecting the involved branches of the carotid artery. The endoscopes are used as adjuncts to the microscope and are helpful for identification and resection of tumor that is located in the blind spots of this approach.

Ample dialogue and interaction with the anesthesia team is necessary for quality surgical outcomes. Given the limited superficial exposure afforded by the eyebrow supraorbital approach, brain relaxation is of paramount importance. The patient is positioned in the typical position of a pterional craniotomy, with the malar eminence at the highest position and the patient's back angled to 20–30°. The head is rotated slightly (10–15°) for parasellar lesions. A modest dose of mannitol is given (25 g for a typical adult), and the patient is mildly hyperventilated with a goal arterial pCO_2 of 30 mmHg. High-dose dexamethasone is often administered – even in the absence of hypopituitarism – which helps to mitigate cerebral edema. Lumbar drainage/CSF diversion is not routinely utilized. If neuromonitoring is employed with cranial nerve stimulation, then muscle relaxants are avoided, and total intravenous anesthesia (TIVA) is employed. TIVA also allows for decreased postoperative nausea/vomiting and a potentially shorter postoperative hospital course. The abdomen is prepped in all patients, even if sinus entry appears unlikely on imaging.

The incision is planned through the center of the eyebrow, starting just medial to the superior orbital notch (or foramen) and extending along the orbital rim about 1 cm inferior to the superior orbital line. If the eyebrow is thin or absent laterally, the incision is continued along the orbital rim. The eyebrow is never shaved, as this results in significant cosmetic deficit with inadequate and delayed hair growth. When the incision is made, the angle of the scalpel blade is situated parallel to the direction of the hair follicles, aiming to minimize transection of the eyebrow hairs, which helps afford improved cosmesis.

The orbicularis oculi is incised sharply and the pericranium is exposed. Then, the supraorbital nerve is carefully dissected at the region of the supraorbital notch. This nerve is typically deep to the orbicularis oculi and often has small local branches that arise from the main nerve (Fig. 18.5a). Efforts are made to preserve each branch, though some branches are very low along the orbital rim and are sacrificed with little long-term clinical significance. A subgaleal pocket is then created to the extent of the planned craniotomy. The pericranium and the temporalis muscle fascia are then incised down to the frontal bone as a single layer and extended to the origin of the zygoma. This layer is elevated and secured with a stay suture. The dissection should avoid entry into the orbit, as this could result in postoperative periorbital ecchymosis. If the orbital rim is to be removed, then the dissection should be carried further, and the periorbital ecchymosis should be reflected off the orbital roof. Fishhooks are used to retract the skin and temporalis muscle superiorly. These are periodically readjusted throughout the operation to prevent laceration or necrosis of the skin edges.

The craniotomy itself has three components: a single burr hole, turning a flap, and drilling the inner table. The burr hole is made with a small burr drill bit (e.g., "matchstick") beneath the superior temporal line. This location allows for placement of a burr hole cover without major cosmetic deficits. The dura

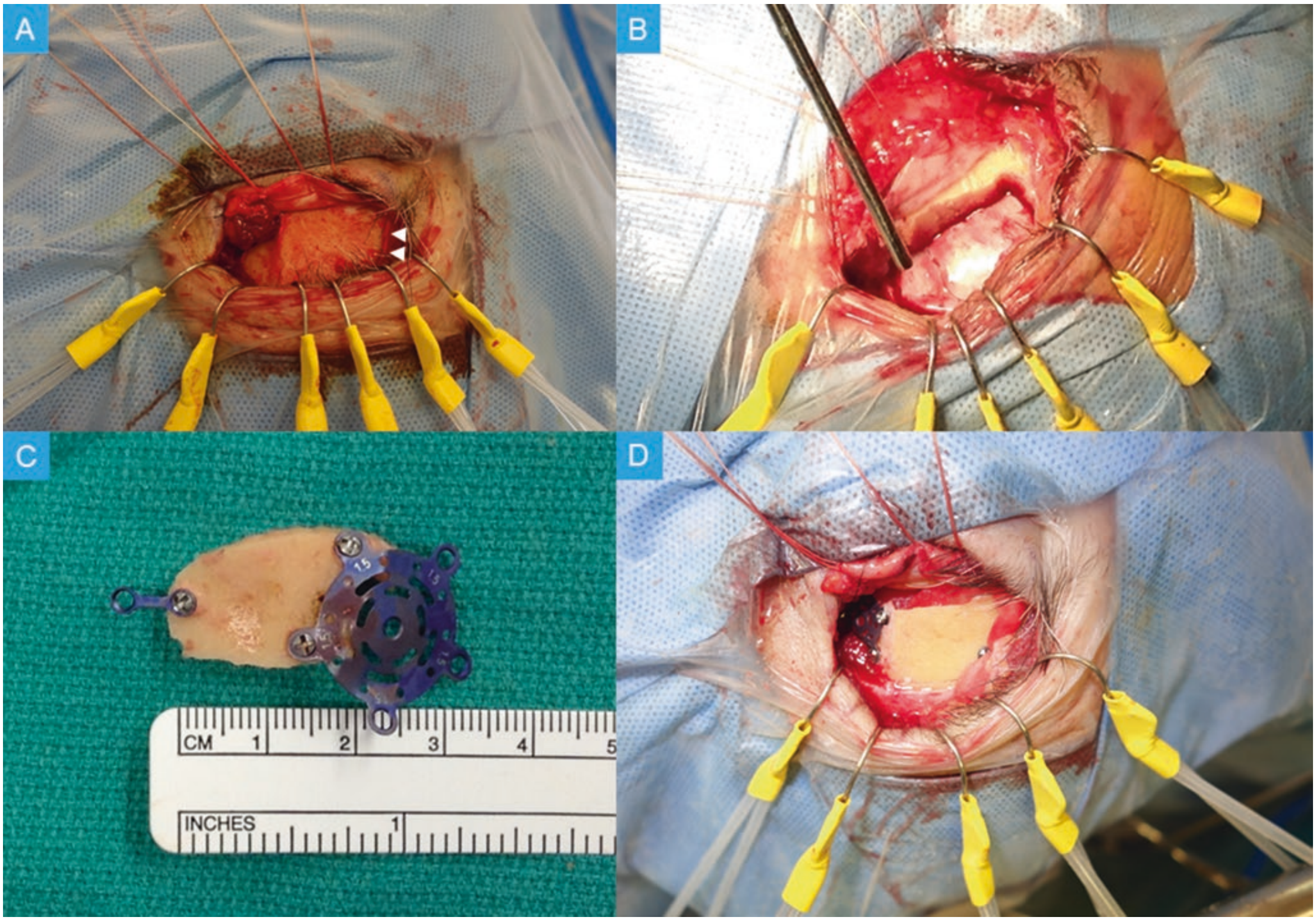

Fig. 18.5 (**a**–**d**) Intraoperative photographs of supraorbital eyebrow craniotomy. (**a**) Pericranial dissection is complete with preservation of the supraorbital nerve (*arrowheads*). (**b**) Craniotomy is completed with burr hole beneath superior temporal line and orbital roof flattened with drill. (**c**) Bone flap with titanium plate locations. (**d**) Positioning of bone flap with superior and medial edges flush with the skull. The gap is left beneath the eyebrow to preserve cosmesis

is carefully dissected, and the orbital roof is palpated. The craniotomy is performed with a craniotome while protecting the supraorbital nerve (the medial limit). The goal is to remain flush with the orbital roof; hence, this cut is made first, and the drill follows the orbital roof. It is key to ensure a symmetric, ovoid craniotomy, avoiding narrowing at either side. This will prevent a "pinching" effect of the keyhole craniotomy, limiting the use of two-tined instruments such as microforceps or bipolars. Minimal dimensions are 1.5 cm (anterior/posterior) and 2.5 cm (lateral). The inner table is then drilled with the "matchstick" drill bit, and the orbital roof is smoothed (Fig. 18.5b). If there is entry into the frontal sinuses, it is either sealed with bone wax (for small or pinpoint breaches) or betadine-soaked sponge for larger defects, to be more definitively addressed during the closure. Though we are not advocates of removing the orbital rim, some authors do this for certain pathologies [16]. There is, as expected, a larger working area and increased degrees of freedom [17]. If the tumor has significant superior extension, requiring a very superior working angle, this may be of some help, though we find the use of angled endoscopes to work just as well.

The dural opening is performed with the intent to achieve ultimately a watertight closure. The initial step is to expose either the optico-carotid or carotid-oculomotor cistern for CSF egress. For craniopharyngioma, this is quite feasible and obviates the need for lumbar drain CSF diversion. It is rare for craniopharyngioma to extend into the optic canal, but if this is noted intraoperatively, the optic canal roof is drilled, the falciform ligament is divided, and the tumor is removed from this location.

The ipsilateral Sylvian fissure is split, and the anterior cerebral artery is dissected, identifying the anterior communicating artery, the contralateral A1 and both A2 arteries. The tumor is dissected from the dorsal optic nerve and chiasm. Tumor extending into the third ventricle via the lamina terminalis is carefully resected while trying to avoid injuring the hypothalamus. Tumor is dissected off the anterior cerebral artery complex, preserving the perforator arterial branches. Most of the unnamed perforators in this region supply the hypothalamus, and a stroke in this region can result in debilitating hypothalamic dysfunction such as hypothalamic obesity, adipsic diabetes insipidus, and cognitive dysfunction.

If there is tumor extending below the plane of the optic apparatus (a relative contraindication to this approach), the interoptic cistern is dissected under direct visualization utilizing angled endoscopes and instruments. The endoscope of choice is the 30°, 4 mm rigid endoscope with HD camera. This offers a low-profile system while preserving high resolution and magnification. We prefer using both the standard and reverse light-post endoscopes to minimize instrument collision while visualizing regions such as the sellar fossa, perimesencephalic region, and infraoptic region. The endoscope is held by an experienced surgeon and brought into the field with the instruments to minimize inadvertent injury with blind movements. Static or pneumatic endoscope holders are not often utilized.

It is imperative that the superior hypophyseal arteries are visualized and preserved, as these are typically involved by the capsule of craniopharyngiomas. For tumors that are mostly cystic, the cyst is drained, and the capsule is resected where feasible. Often, the cyst wall is difficult to dissect off the brain pia mater and must be left as residual, to be treated with adjuvant therapies.

After hemostasis is achieved and the entire cavity is inspected for all resectable tumor, the vasculature is treated with papaverine if there is concern of the potential for vasospasm. Subsequently, the dura is closed primarily in a watertight fashion if possible. This is most relevant if the lamina terminalis was entered for tumor resection, resulting in a high-flow CSF communication.

If there was a significant breach of the frontal sinus, the mucosa is stripped back, and the cavity is obliterated with a fat graft and collagen sponge. The bone flap is plated with a low-profile titanium burr hole cover (lateral) and a short two-hole bar (medial) (Fig. 18.5c). The gap of the bony defect is situated inferiorly – beneath the eyebrow (Fig. 18.5d). In patients with thin eyebrows, bone cement is used to augment the cosmesis of the bone flap. The pericranium is then reapproximated, and the orbicularis/dermal layer is closed, followed by a subcuticular stitch for the skin (typically 4–0 or 5–0 unbraided dissolvable suture). During the closure, pressure is held on the skin to prevent a hematoma from accumulating. The wound is then covered with a nonstick gauze and gently wrapped with an elastic headwrap. This should be tight enough to prevent a hematoma but loose enough to avoid scalp necrosis or pressure urticaria.

Postoperative Care

Much of the success of any skull base surgery relies on appropriate and meticulous postoperative management. Similar to most skull base craniotomies, the patient is admitted to the intensive care unit for close neurovascular monitoring. Tight blood pressure parameters are applied, keeping the patient normotensive and avoiding blood pressure spikes. The patient's vision, cranial nerve, and cognitive exam are routinely assessed and compared to the preoperative and immediate postoperative baselines. The head-of-bed is maintained at about 30°.

A postoperative CT head without contrast is performed immediately after the patient is discharged from the recovery unit. This assesses the tumor cavity for any potential hemorrhagic complication. An assessment of the degree of pneumocephalus is performed. If there is excessive pneumocephalus, the patient is treated with 100% oxygen for 24 h to help decrease postoperative hypotensive headaches. This CT can be compared with future scans, and if the pneumocephalus has not resolved or has worsened, this could reflect a possible cerebrospinal fluid leak.

A postoperative "pituitary protocol" MRI is performed on either the first or second postoperative day. The primary purpose is to assess for the extent of tumor resection. In the unlikely situation of a large residual tumor, the patient may require an early reoperation. Certain cystic craniopharyngioma can develop early cyst recurrence. Hence, the immediate postoperative MRI is helpful to compare to interval postoperative scans at 1–2 months which can also help for stereotactic radiotherapy planning.

A deviation from our standard cranial postoperative protocol is the endocrine assessment and treatment, which mirrors our endonasal postoperative protocol. As stated, these patients typically are prescribed dexamethasone or stress-dose hydrocortisone perioperatively. These medications are tapered down, and if the pituitary gland could be preserved, they may be discontinued to assess the adrenal axis with adequate morning serum cortisol levels. As many of these patients are at high risk for diabetes insipidus (DI), close monitoring of serum sodium and urine output would prompt administration of desmopressin (ddAVP). These patients are comanaged with our pituitary endocrinologists. In follow-up, the remaining pituitary axes are assessed including the thyrotrope, gonadotrope, and somatotrope axes. If the pituitary stalk was sacrificed, the patient is typically discharged with low-dose levothyroxine replacement to be adjusted at the 6-week follow-up visit. Whether or not the patient has developed diabetes insipidus, a delayed postoperative serum sodium level is performed at 5–7 days after surgery. If the gland is preserved, this would help diagnose a delayed syndrome of inappropriate antidiuretic hormone (SIADH) or DI. If the patient was discharged on ddAVP, the serum sodium helps assess for proper dosage.

Long-Term Follow-Up

Following the initial postoperative visit, the patient is seen at about 2 months after surgery. At this time, a repeat "pituitary protocol" MRI is performed, and the patient's neurological symptoms and signs are assessed. If the patient had vision loss preoperatively, a formal visual field test is performed, primarily to serve as a baseline for future examinations. Given the high incidence of tumor recurrence, even in the setting of gross total resection (GTR), fractionated stereotactic radiation therapy is administered either prophylactically or at the earliest sign of tumor recurrence/progression [18, 19]. There are sufficient data to suggest that patients have superior outcomes with combinatorial therapy compared to surgical resection alone [19, 20].

Long-term endocrinological treatment is of paramount importance. In the setting of pituitary stalk preservation, there is possibility of hormonal recovery, and periodic assessments of the adrenal and thyroid axes in particular may be helpful. In these patients, radiation therapy can also result in hypopituitarism over time [21, 22].

Complication Avoidance

Approach-specific complications have overlap with more extensive craniotomies such as the pterional and orbitozygomatic approaches. These include cosmetic/wound healing issues, CSF rhinorrhea prevention, supraorbital nerve injury, frontotemporal nerve injury, stroke/vascular injury, optic nerve injury, and hypothalamic injury.

The supraorbital approach has the potential for very visible cosmetic deficits. Hence, much effort is taken to prevent wound issues. This includes a precision craniotomy that is re-plated and approximated to the superior edge of the defect to position the gap beneath the eyebrow. Bone cement is used to fill in the gap if the patient has a thin or absent eyebrow. Meticulous hemostasis helps prevent a postoperative hematoma that typically can result in periorbital ecchymosis. During the operation, the "fishhooks" are frequently repositioned to prevent laceration of the skin edges.

CSF rhinorrhea can occur if the frontal sinus is breached. A watertight closure is always attempted but sometimes not possible. Hence, abdominal fat grafting is helpful to seal this structure. A multilayer collagen sponge reinforcement is helpful as well.

The supraorbital nerve dissection is necessary to minimize the incidence of long-term postoperative supraorbital anesthesia. Once the nerve course is traced, the main branches are preserved. The "fishhooks" are positioned to avoid direct traction along this nerve. During closure, the medial plate is often positioned beneath this nerve. This requires gentle retraction during screw placement to prevent damage to the nerve. Similar efforts are made during pericranial flap closure. Overall, transient supraorbital hypesthesia occurs for about 1–2 months in many patients, but is permanent in about 3.4–7.5% [23, 24].

The frontotemporal branch of the facial nerve is not visualized during the operation. However, the majority of patients will develop an immediate postoperative frontalis paresis. Many of these will resolve within 3 months following surgery. Permanent paresis occurs in about 2% of patients [23]. The nerve has variable trajectories along the orbital rim, and its location correlates with postoperative paresis [25]. Frequent repositioning of the "fishhook" retractors during the operation will minimize the tension on this nerve and may prevent permanent injury.

A concern with a smaller exposure compared to pterional or orbitozygomatic approaches is vascular control and the management of vascular injury. A key adjunct to performing safe tumor resection is the micro-Doppler probe [26, 27]. Particularly in the setting of recurrent and/or radiated tumors, the carotid artery and its branches can be difficult to identify or dissect off the tumor. Hence, frequent use of Doppler ultrasound is helpful to avoid vascular injury.

Vessels that are most vulnerable to inadvertent injury are the superior hypophyseal arteries. These vessels are not only hidden by the optic chiasm but often adherent to the capsule of the craniopharyngioma. Other branches that can be involved include the anterior choroidal, posterior communicating, and recurrent Heubner arteries. Hence, blind dissection or excessive traction of the craniopharyngioma should be avoided.

Preserving the optic apparatus and vision function is dependent on vascular preservation (superior hypophyseal and anterior choroidal arteries) as well as careful optic sheath dissection. Often, recurrent/radiated tumors can be adherent to the optic chiasm. Hence, a small residual tumor is allowed to prevent optic nerve injury. Over the past few decades, the trend in craniopharyngioma surgery has shifted from attempting gross total resection to achieving maximal safe resection. A large series of craniopharyngioma patients cared for in the 1980s demonstrates a 90% gross total resection rate, but with 15% worsened vision and 17% mortality [28]. This is in stark contrast to a contemporary single-center, high-volume study that reported 38% gross total resection and 34% near-total resection with no new vision loss or increased mortality [12]. In the era of stereotactic radiation (IMRT) and targeted molecular therapy, permissive tumor residual is acceptable, even in the setting of recurrent tumor.

The hypothalamus borders the floor and the inferior half of the third ventricle lateral walls. Within this thin structure exists the numerous hypothalamic nuclei regulating pituitary hormone release as well as homeostatic functions. Hypothalamic injury can present with fatigue, memory loss, behavioral changes, adipsic diabetes insipidus, and obesity. These can be quite challenging to treat and very debilitating to the patient. Hence, measures to prevent hypothalamic injury are important for maintaining quality of life. Tumor that is densely adherent to the hypothalamus is debulked with deliberate residual left on the ependymal surface [29].

Surgical Outcomes

The supraorbital eyebrow craniotomy approach has been successfully used for a variety of pathologies. These include benign and malignant brain tumors as well as intracranial aneurysms, cavernous malformations, and other nonneoplastic lesions [24, 30–33]. Parasellar lesions are ideal for this approach, given its anatomic exposure previously described. Numerous surgical series have demonstrated the versatility of this approach with comparable outcomes to traditional frontal approaches such as the pterional, orbitofrontal, and orbitozygomatic craniotomies.

There are limited published series utilizing the supraorbital craniotomy for craniopharyngioma, primarily due to the infrequence of anterochiasmal tumors. Reisch et al. reported 39 of 1125 (3.5%) supraorbital "eyebrow" craniotomies over a 10-year period were for craniopharyngiomas [24]. Seventy-four percent of these patients achieved gross total resection, though only 36% were recurrent tumors. Fatemi et al. reported a more contemporary series comparing endonasal and supraorbital approaches for craniopharyngiomas [34]. Only four patients of 22 underwent the supraorbital approach, and 50% were recurrent tumors compared to 33% of the endonasal cohort. Conversely, only 50% gross or near-total resection was accomplished, compared to 67% with the endonasal approach [34]. In a follow-up article, McLaughlin et al. reported four supraorbital operations for recurrent tumors in patients that had previously been treated via craniotomy [9]. Overall outcomes were good, though one patient did experience a CSF leak, which required reoperation.

PEARLS
The supraorbital eyebrow craniotomy is ideal for the craniopharyngioma that is anterior or superior to the optic apparatus.
Meticulous attention to detail during exposure, nerve dissection, and closure is necessary to achieve optimal cosmesis.
Preservation of vascular and hypothalamic anatomy is essential to prevent postoperative morbidity.
Permissive near-total resection is acceptable with adjunct stereotactic radiation therapy and targeted molecular therapy.

18.4 Suprasellar Craniopharyngiomas: Endoscopic Endonasal Approach

Amanda Carpenter, Jean Anderson Eloy, and James K. Liu

Introduction

The endoscopic endonasal approach (EEA) has evolved significantly in the last decade. With more accurate neuronavigation, improved endoscope optics, and the evolution of skull base reconstruction materials and techniques, endoscopic endonasal surgery is safer and more effective and has become the preferred approach for a variety of skull base lesions. Traditional transcranial approaches to craniopharyngiomas often require some degree of brain retraction (and potential cerebral edema) and lack complete direct visualization of critical structures in the retrochiasmatic region. Traditional speculum-based microsurgical transsphenoidal approaches provide a direct endonasal route but with limited field of view and surgical freedom. On the other hand, the extended EEA via the transplanum transtuberculum corridor provides direct midline exposure to intrasellar/subdiaphragmatic, supradiaphragmatic, and retrochiasmatic craniopharyngiomas that extend up to the third ventricle without any brain retraction [35]. The extended EEA offers unmatched visualization of the undersurface of optic nerves and chiasm, pituitary stalk, third ventricle, perforators, and hypothalamus. Craniopharyngiomas that are retrochiasmatic in location should be strongly considered for resection via the EEA.

Patient Selection

When choosing a surgical approach for craniopharyngiomas, the optimal choice should be the shortest and most direct route that will provide maximal exposure and visualization of the tumor's interface with surrounding critical structures. The anatomic location of the tumor and its degree of extension are of paramount importance, particularly its relation to the optic chiasm, the pituitary gland and stalk, the hypothalamus, the carotid artery, the anterior cerebral artery complex, as well as the sella and diaphragm. Retrochiasmatic lesions are particularly well suited for an extended EEA via the transplanum transtuberculum corridor rather than a transcranial route in order to avoid manipulation of the optic nerves and chiasm. It is important to note that tumor extension into the third ventricle can be removed via an EEA, as long as there is communication with suprasellar space [36]. However, pure intraventricular craniopharyngiomas situated within the third or lateral ventricles may be better accessed with a transcranial transventricular approach via a transcortical or transcallosal route [35]. A combined approach of both open and endoscopic techniques may be necessary for extensive lesions that involve multiple anatomic compartments. In cases of pure intrasellar craniopharyngiomas, a transsellar approach is favorable. However, the majority of suprasellar supradiaphragmatic craniopharyngiomas require an extended EEA via the transplanum transtuberculum corridor to gain optimal exposure. Tumors with significant lateral extension (>1 cm lateral to the carotids) may not be amenable to EEA, as are tumors with significant superior extension into the interhemispheric fissure [36]. Another limitation of the EEA is the inability to perform direct vascular repair or bypass in the case of arterial vessel injury.

The age of the patient and medical history must be taken into account when choosing surgical approach. For example, in an older patient with many medical comorbidities, a more conservative approach with a goal mainly to decompress neural structures may be most appropriate. In addition, surgeon's preference, experience, and skill level are also important considerations. The EEA is associated with a significant learning curve, and the surgeon's comfort level performing this approach should be considered. It is also crucial to have a collaborative experience with an otolaryngologist specializing in rhinology and endoscopic skull base surgery, which provides a multidisciplinary team approach to the patient. Some factors that make EEA less favorable include a hypoplastic sphenoid sinus, significant lateral extension of the tumor into the Sylvian fissure, significant superior extension into the interhemispheric fissure, a narrow intercarotid artery distance, and a narrow infrachiasmatic window [36].

Preoperative Evaluation

Preoperatively, in addition to conducting a thorough neurologic exam, it is crucial to obtain neuro-ophthalmologic evaluation to assess visual fields and acuity. It is also routine to have a neuroendocrine evaluation, which includes measurement of pituitary hormone levels and a body mass index measurement, as hypothalamic involvement can affect appetite and weight. Evaluation with an otolaryngologist should be obtained for surgical planning. Recent neuroimaging studies are imperative as well. CT scan shows the bony anatomy of the nasal sinuses and skull base that will be encountered during the approach and reveals calcifications and cystic components of the tumor [36]. MRI demonstrates tumor extension and can also differentiate solid and cystic components, position of the chiasm, and relationship of the tumor to neighboring vascular structures, the pituitary stalk, and the third ventricle [36].

Surgical Technique: Endoscopic Endonasal Transplanum

Transtuberculum Approach

Patient Positioning

The patient is placed under general anesthesia with the endotracheal tube secured to the left side of the patient. We generally place a lumbar drain at the time of surgery to minimize the risk of postoperative CSF leakage. The patient is positioned supine on the operating table with the head in a three-point Mayfield head holder. The bed is arranged to keep the head slightly elevated above the heart to promote venous return. The head is slightly rotated to the right to facilitate easier access for the operating surgeons standing on the right side of the patient. The head is also slightly extended to improve access to anterior skull base. Frameless stereotactic image guidance is used for intraoperative navigation and for anatomic localization. It also helps guide the extent of anterior bone removal from the planum sphenoidale based on the sagittal trajectory to the lesion [37]. The nose and nostrils are prepared with Betadine, and the nasal cavity is packed with Afrin-soaked pledgets. The abdomen and thigh are also prepared for harvest of autologous fat and/or fascia lata for dural repair and reconstruction. Intravenous antibiotics and 10 mg of dexamethasone are administered prior to incision. Mannitol and antiepileptics are usually not used because there is no brain retraction or manipulation during the EEA.

Endonasal Sphenoid Sinus Exposure

In our center, we use a two-surgeon, three- to four-hand binostril technique with a neurosurgeon and otolaryngologist. The initial endonasal exposure to the sphenoid sinus is performed primarily by the otolaryngologist using a high-definition 30°-angled endoscope (Karl Storz, Tuttlingen, Germany). We prefer the 30°-angled endoscope because of the viewing capabilities around corners with simple rotation of the scope. The tail and anterosuperior attachment of the middle turbinates, as well as the nasal septum, are infiltrated with 1% lidocaine with epinephrine (1:100,000 dilution). Both middle and inferior turbinates are mobilized laterally. In some cases, the right middle turbinate can be removed to allow for more room for multiple instruments in the right nostril, if needed. The sphenoid ostium is identified bilaterally about 1–1.5 cm superior to the choanal arch and medial to the superior turbinate. A wide sphenoidotomy and posterior ethmoidectomy are performed with a microdebrider and Kerrison rongeurs. The same maneuvers are performed in the left nostril with an additional posterior septectomy of about 1.5–2 cm in order to create a unified working corridor to the anterior skull base. The posterior septectomy allows triangulation of surgical instruments through both nostrils so that bimanual dissection can be performed. It is important to recognize the presence of an Onodi cell (posterior ethmoid cell that is positioned superolateral to the sphenoid sinus), because the optic nerve and carotid artery may often course through the lateral aspect of that cell.

At this point, a vascularized, pedicled, nasoseptal flap is harvested from the nasal septum and rotated posteroinferiorly into the nasopharynx until later use at the time of reconstruction. Care must be taken to protect the vascular pedicle arising from the posterior septal branch of the sphenopalatine artery from inadvertent injury. At this juncture, the neurosurgeon and otolaryngologist work simultaneously using a binostril technique. The otolaryngologist provides guidance and optimal visualization with the 30° endoscope in the right nostril in the 6 o'clock position looking superiorly. The neurosurgeon uses bimanual surgical technique, with a suction device placed in the 12 o'clock position in the right nostril and the working instrument (drill, dissector, scissors, bipolar device, or tissue aspirator) in the left nostril.

Transplanum Transtuberculum Bony Opening

During the bone drilling, we prefer to use a double-barrel suction-irrigator in the right nostril. The self-irrigating system keeps the surgical field clear of bone dust and also cools the drill tip to protect underlying structures from heat injury. Irrigation is also provided from the self-irrigating high-speed drill and the irrigating endoscope sheath. The sphenoidotomy opening is maximally widened, removing all sphenoid septations and bony ridges that may hinder instrument maneuverability and surgical freedom. It is important to ensure that the line of sight to the transplanum transtuberculum region is unobstructed. A high-speed diamond drill with copious irrigation is used to remove bone over the sella turcica, planum sphenoidale, and tuberculum sellae. It is also important to identify the medial and lateral opticocarotid recesses on both sides. The medial opticocarotid recess is an indentation in bone that is formed at the medial junction of the parasellar carotid canal and the optic canal. This recess represents the lateral aspects of the tuberculum sellae as viewed from the endonasal perspective [10]. The lateral opticocarotid recess represents the optic strut from the endonasal perspective. Once the tuberculum strut and both medial opticocarotid recesses are thinned down to eggshell thickness, an up-angled 5-0 curette is used to remove the remaining remnant of tuberculum strut and medial opticocarotid recesses. By removing the medial opticocarotid recesses, the medial aspect of the optic canals are unroofed, which facilitates exposure of the optic nerves and paraclinoid carotid arteries in the opticocarotid cistern [10]. It is important to avoid using a Kerrison rongeur in the region of the optic canal since this can cause potential injury to the optic nerve.

Next, we prefer to open the dura in a transdiaphragmatic fashion, at the level of the planum and sella. An arachnoid knife or number 11 blade is used to make a cruciate incision over the sellar dura, and a second horizontal incision is made in the dura of the planum sphenoidale above the intercavernous sinus. The superior intercavernous sinus is coagulated with an endoscopic bipolar and divided sharply with scissors. This incision is continued along the diaphragma sella to expose the suprasellar cistern.

We typically use a 30° endoscope which gives the surgeon additional angled views. The endoscope is placed at the 6 o'clock position, with the suction at the 12 o'clock position in the right nostril when using the 30° endoscope to look up into suprasellar cistern or retrochiasmatic space. The neurosurgeon is therefore working "above" the endoscope while maintaining optimal surgical exposure. When using a 0° endoscope, we prefer to do the opposite and place the endoscope at the 12 o'clock and the suction at the 6 o'clock position.

Intradural Tumor Dissection and Removal

The arachnoid is dissected to expose the underlying tumor in the retrochiasmatic space. We recommend using an extra-arachnoidal dissection technique, in other words, dissecting in the plane between tumor capsule and the tumor arachnoid, instead of between tumor arachnoid and cisternal arachnoid. This allows mobilization of the arachnoid layers toward the critical neurovascular structures to provide a buffer of protection. Both carotid arteries are visualized underneath the optic nerves as they exit the distal dural ring. It is important to identify the superior hypophyseal arteries that arise from the carotid arteries and to preserve branches that supply the undersurface of the optic apparatus to avoid postoperative vision deficits. There is typically an arachnoid layer investing these perforators, and by working between the tumor capsule and the arachnoid layer, the perforators can be safely mobilized and preserved during tumor removal.

Initial decompression of the cystic contents is performed to allow collapse of the tumor capsule. This facilitates descent of the superior extent of the tumor and allows for subsequent extracapsular microdissection. Dissection of the tumor capsule from the undersurface of the optic chiasm and hypothalamus is achieved using conventional bimanual microsurgical techniques. The cyst wall is placed under gentle countertraction, while the suction is used as a dissector to sweep the capsule off of the neural structures. In some cases, arachnoid adhesions are lysed with sharp dissection using microscissors. By using bimanual microsurgical dissection techniques, the tumor capsule is identified in relation to the optic chiasm, optic nerves, and pituitary stalk. In retrochiasmatic craniopharyngiomas, the tumor is located underneath and posterior to the optic chiasm. It can be adherent to the undersurface of the optic apparatus and hypothalamus, with extension superiorly into the third ventricle and posteriorly into the interpeduncular fossa and retrosellar space. The anterior communicating artery complex is located superior to the optic chiasm and is therefore protected from the plane of dissection.

For solid craniopharyngiomas, the tumor is internally debulked with a side-cutting tumor aspirator device (NICO Myriad®, Indianapolis, IN, USA) or ultrasonic aspirator. Initial debulking of the solid component and/or aspiration of cystic fluid allows for decompression of the tumor capsule. Once the tumor is adequately debulked and decompressed, extracapsular dissection of the tumor capsule away from optic chiasm and hypothalamus is performed with careful bimanual microdissection. Care is taken not to prematurely amputate the tumor capsule, as it provides a surgical "handle" to provide countertraction for extracapsular dissection. After meticulous microdissection, the most superior extent of the tumor should descend into the retrochiasmatic space.

In most cases, the membrane of Liliequist is intact and can act as a plane of dissection to peel tumor safely from the basilar artery, posterior cerebral arteries, and P1 perforating vessels. To visualize this region, the 30° endoscope is pointed inferiorly and placed in the 12 o'clock position in the right nostril. The inferior aspect of the tumor is elevated from the top of the pituitary gland to identify the base of the pituitary stalk. We attempt to preserve the pituitary stalk, especially if the tumor can be readily dissected off of the stalk. However, if a gross total resection is possible and there is tumor invading or expanding the stalk (type II transinfundibular), we prefer to do a low stalk transection just above the pituitary gland to facilitate gross total removal and place the patient on postoperative hormone replacement therapy. We agree with the opinion of Dr. Oldfield that this strategy may be better to prevent tumor recurrence rather than leaving tumor on an anatomically intact stalk that may not retain normal pituitary function [38].

The tumor is generally most adherent at the level of the hypothalamus where meticulous and careful microdissection is performed. Once the tumor is free from all areas of adherence, the tumor is carefully delivered through the nose. Premature pulling of the tumor without complete dissection can potentially result in catastrophic nerve or vessel injury. If the floor of the third ventricle is open, a 30° and 70° endoscope can be used to look inside the walls to inspect for residual tumor.

Gross total resection (GTR) should be the goal of craniopharyngioma surgery if safely possible. However, in some cases, GTR may not be feasible due to the intimate nature of some of these lesions to critical neurovascular structures. Therefore, it may be necessary to leave some residual tumor that is densely adherent to important nerves or vessels yet maximize the extent of safe resection. In the event of subtotal or near-total resection, radiation therapy is an appropriate adjuvant treatment and has been shown to decrease recurrence rates [39].

Closure and Skull Base Reconstruction

Closure and reconstruction are of utmost importance to prevent a postoperative CSF leak. Although various techniques exist, we prefer a multilayered closure with autologous fascia lata and a vascularized pedicled nasoseptal flap for transplanum defects. An initial piece of Gelfoam is placed underneath the dural opening as an inlay to slow the pulsations of CSF out the dural defect. Next, an autologous fascia lata graft is placed over the dural opening and held in place with a monolayer of Surgicel® (Ethicon, Somerville, NJ, USA). This step is repeated with a second layer of fascia lata to reinforce the initial layer. Another monolayer of Surgicel is placed over the bone defect to hold the fascia graft in place. Finally, the vascularized pedicled nasoseptal flap is then rotated superiorly to cover the dural closure and bony skull base defect. Care is taken to ensure that the edges of the nasoseptal flap are in contact with the bone. The bony surface must be devoid of any sinus mucosa as this will increase risk of flap dehiscence and possibly delayed formation of mucoceles. Cottonoids are used to apply gentle pressure on the flap to ensure a good seal against the skull base without any trapped air pockets. After removing the cottonoids, another monolayer of Surgicel is placed over the edges of the flap against the surrounding bone to prevent flap migration. The flap is then bolstered with several layers of gentamicin-soaked Gelfoam pledgets followed by a Merocel® (Medtronic, Dublin, Ireland) expandable nasal tampon, positioned in the sphenoid sinus posterior to the nasal septum. The packing expands as it is hydrated with gentamicin irrigation. The lumbar drain is opened temporarily during extubation to allow preferential drainage of CSF through the lumbar catheter rather than through the repaired defect and to prevent increases in intracranial pressure [35].

Potential Complications

One of the most feared complications is direct injury to a major vascular structure. This can be devastating and requires prompt hemostasis via coagulation or clip ligation. A carotid injury may require packing with crushed autologous muscle followed by nasal packing to tamponade the bleeding. An emergent angiogram is then performed to rule out a pseudoaneurysm and to perform potential intervention. Postoperative vision worsening secondary to manipulation of the optic nerves and/or chiasm or vascular compromise is also possible. Anterior pituitary insufficiency and diabetes insipidus may also occur due to pituitary stalk manipulation or intentional stalk transection. The most frequent postoperative complications are discussed below.

Postoperative Care

Care is taken to monitor for signs of intracranial hypotension and CSF rhinorrhea postoperatively. The lumbar drain is kept open at 5–10 ml of drainage per hour for about 72 h after surgery. If a persistent CSF leak is suspected, reexploration and revision of the skull base repair in the operating room may be necessary. An MRI of the pituitary region with and without contrast is performed routinely on postoperative day 1 or 2 to evaluate extent of resection. These patients are also closely monitored for development of diabetes insipidus, which is managed with vasopressin. If the pituitary stalk is intentionally sacrificed, the patient is immediately placed on hormone replacement therapy with hydrocortisone, levothyroxine, and vasopressin. Patients are also preemptively kept on high-dose dexamethasone (10 mg every 6 h) with elevated systolic blood pressures to minimize optic nerve swelling and to optimize chiasmal perfusion, respectively. This is weaned off as tolerated over the course of 7 days. Formal visual field and acuity are assessed by the neuro-ophthalmology team. It is recommended to avoid nasal positive pressure ventilation out of concern for development of pneumocephalus. Patients are maintained on broad-spectrum antibiotics until the Merocel packing is removed by the otolaryngologist in the office on postoperative day 10–12.

Surgical Outcomes

Tumor Resection

It has been reported that the EEA achieves greater rates of gross total resection (GTR) than open approaches. In a recent meta-analysis by Komotar et al., EEA had greater rates of GTR (66.9% vs. 48.3%) and decreased rates of tumor recurrence than transcranial approaches [40]. There was also improved postoperative visual outcomes (57%) but a higher rate of postoperative CSF leak (18.6%) with EEA compared to open approaches [40]. However, reported rates of postoperative CSF leak have declined significantly in recent years with the development of more sophisticated reconstructive techniques, particularly when using the nasoseptal flap. With our technique described above using a multilayered reconstruction technique, our CSF leak rate has been 3.2% [41]. Other groups have reported postoperative CSF leak rates after EEA under 5% as well when using the nasoseptal flap [42].

Visual and Endocrinological Outcomes

Improved visual outcomes after EEA compared to open approaches are routinely reported, likely due to less manipulation of the optic apparatus [12, 40]. Komotar et al. reported improved or stable visual outcomes in 66% of EEA patients [40]. Recent reports of permanent diabetes insipidus after EEA range from 27% to 48% and of panhypopituitarism ranging from 38% to 47% [12, 40, 42, 43].

PEARLS
The extended EEA via the transplanum transtuberculum corridor provides direct midline exposure for retrochiasmatic craniopharyngiomas with excellent visualization of the infrachiasmatic and hypothalamic region.
The EEA is limited for craniopharyngiomas that extend laterally into the Sylvian fissure or superiorly into the interhemispheric fissure.
Multilayered dural defect closure with a nasoseptal flap is essential to minimizing risk of CSF leak.
Postoperative complications include diabetes insipidus, CSF leak, vision deficits, panhypopituitarism, and vascular injury.

Case Illustration

This 56-year-old female presented with progressive headaches, confusion, memory loss, and bitemporal visual field loss. MRI demonstrated a solid, retrochiasmatic craniopharyngioma compressing the optic chiasm with an associated giant right frontal cyst causing significant mass effect (Fig. 18.6a–f). Various surgical approaches were considered including a transbasal subfrontal approach, right orbitozygomatic approach, EEA, and a combined microscopic/endoscopic endonasal approach. After careful deliberation, it was felt that the optic chiasm would be best decompressed with an EEA with decompression of the frontal cyst. If the cyst wall could not be completely removed endonasally, a second-stage transcranial procedure was anticipated if the cyst recurred.

At surgery, the solid, retrochiasmatic component of the tumor was readily dissected off of the optic chiasm and hypothalamus (Fig. 18.7a–f). The pituitary stalk was identified at the base of the tumor and was preserved anatomically. Remnants of microscopic calcifications were left adherent to the top of the optic chiasm and the anterior communicating artery complex. The right frontal lobe cyst was decompressed, but the cyst wall was very adherent to the frontal lobe and anterior communicating artery complex and could not be safely removed. The solid component of the tumor was completely removed with excellent decompression of the optic chiasm and preservation of the pituitary stalk. Here, a decision was made to preserve the stalk because it was anatomically intact, and a complete tumor resection was not achievable.

Postoperatively, the patient had restoration of normal vision, preservation of normal pituitary function without requiring hormone replacement therapy, and no CSF leakage. MRI performed at 3 months follow-up showed no evidence

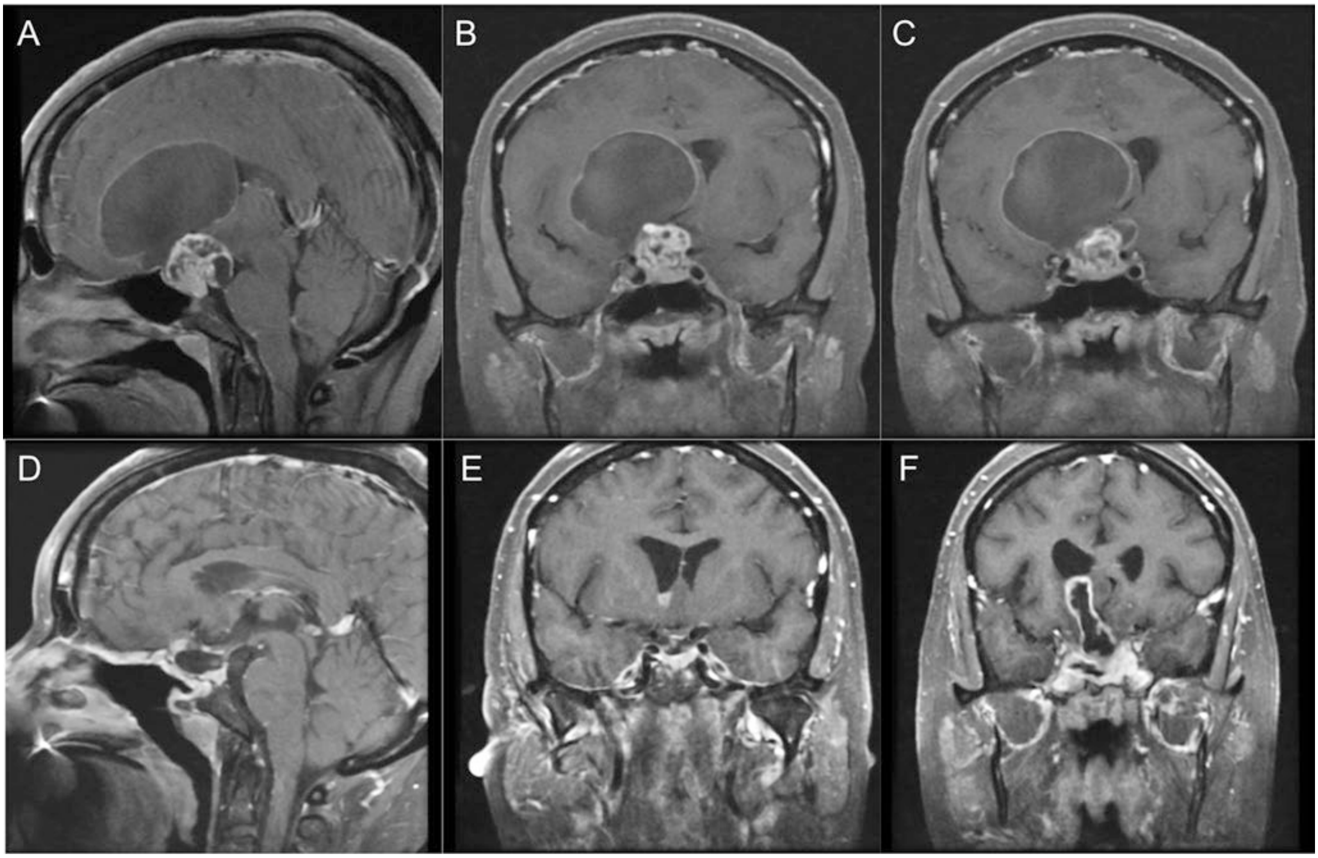

Fig. 18.6 (**a**–**c**) Preoperative MRI of solid retrochiasmatic craniopharyngioma with associated giant right frontal lobe cyst. Removal of the solid retrochiasmatic component was performed with wide fenestration of frontal lobe cyst into suprasellar cistern. (**d**–**f**) Postoperative MRI at 3 months follow-up shows regression and collapse of frontal lobe cyst. Optic chiasm is well decompressed, and the patient had normal pituitary function with stalk preservation (**a**–**f**: Copyright retained by Dr. James K. Liu. Used with permission)

of solid tumor recurrence and regression of the right frontal lobe cyst (Fig. 18.6a–f). It appeared that the frontal lobe cyst was well fenestrated into the suprasellar cistern. The patient was referred for radiation therapy to maintain tumor control. Further imaging follow-up is warranted to determine long-term tumor control. This case illustrates the limitations of complete tumor removal via an EEA when superiorly extending cyst walls are adherent to critical structures. However, we felt it was a reasonable first surgical option since it allowed complete removal of the solid component in the retrochiasmatic space with excellent visual and endocrine outcomes.

Commentary on Case Presentation in Section 18.1

Case Presentation

A 24-year-old lady presents with mild bilateral visual blurriness. Examination: OS, 20/20 (corrected); Humphrey VF, mild arcuate superior and inferior temporal defects; OD, 20/20 (corrected); Humphrey VF, mild arcuate superior and inferior temporal defects. The rest of neurological exam is normal. Laboratory workup: normal endocrine function (see Fig. 18.1).

Discussion

In the case provided by the editors, the patient has a midline suprasellar supradiaphragmatic craniopharyngioma that is primarily retrochiasmatic in location. The lesion is situated below the optic chiasm and does not have any third ventricular or lateral extension. It appears that the majority of the tumor is pre-infundibular and a smaller component is post-infundibular. Nevertheless, one must be prepared for a tumor that is invading or encasing the pituitary stalk. With that in mind, the patient is also a young female of childbearing age which factors into the goals as well as the limits of aggressive surgical resection. In this particular case, the authors would favor an endoscopic endonasal transplanum transtuberculum approach, which provides midline access to the tumor with excellent visualization of the retrochiasmatic region. The challenge here is primarily in preservation of the pituitary stalk and pituitary gland function due to the patient's potential desire for having children. A gross total resection

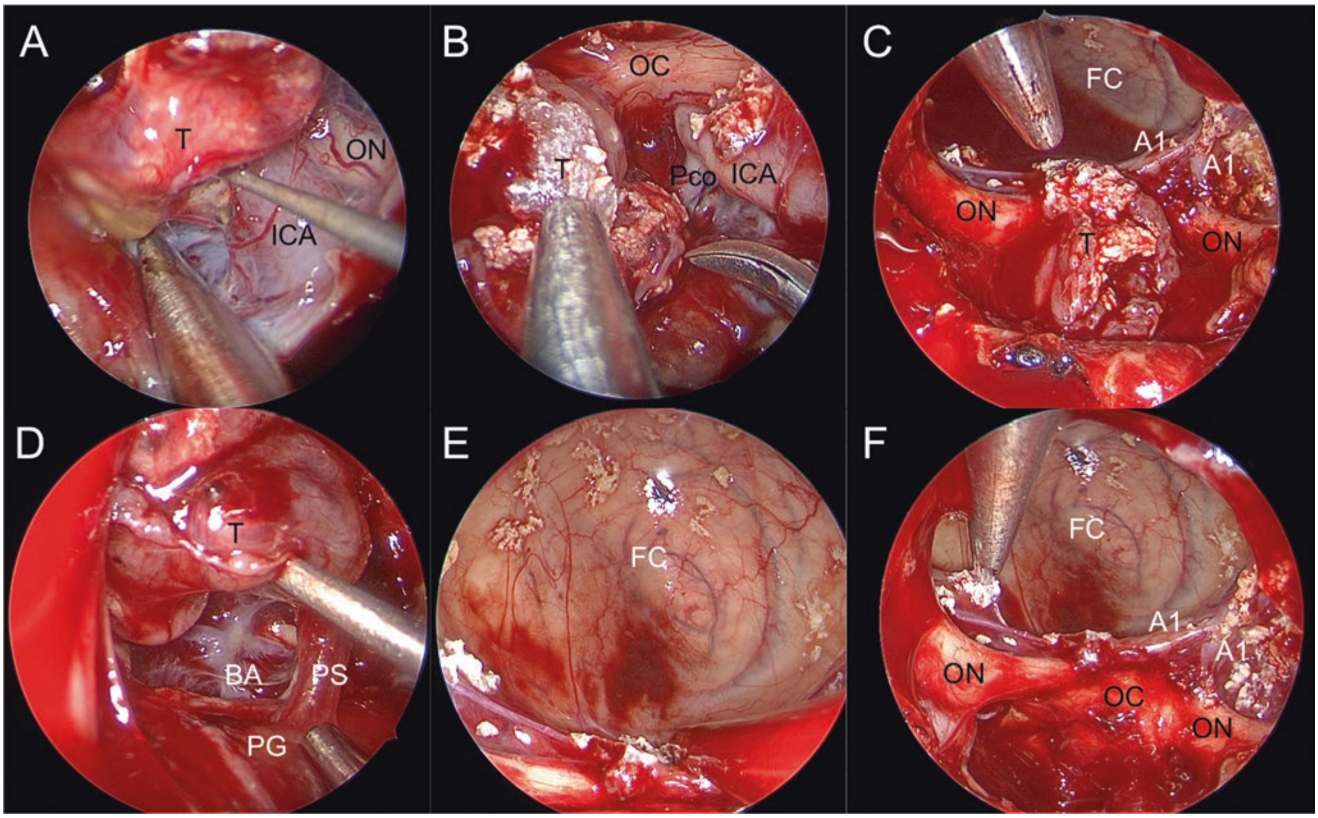

Fig. 18.7 (**a**–**f**) Intraoperative photographs of EEA resection of solid retrochiasmatic craniopharyngioma with associated giant right frontal lobe cyst. (**a**, **b**) Extracapsular dissection of tumor (T) off of the left internal carotid artery (ICA), left posterior communicating artery (Pco), left optic nerve (ON), and optic chiasm (OC). (**c**) Right frontal cyst (FC) is widely fenestrated into suprasellar cistern. The tumor (T) is very adherent to the optic chiasm. (**d**) Elevation of the tumor from the top of the pituitary gland (PG) reveals the pituitary stalk (PS) which is able to be preserved. The basilar artery (BA) complex and left oculomotor nerve are visualized. (**e**, **f**) Final view after near-total resection. The optic chiasm is well decompressed, and there is adherent microscopic tumor to the both A1 vessels. The frontal lobe cyst (FC) was also very adherent to the brain and the A1 vessels (**a**–**d**: Copyright retained by Dr. James K. Liu. Used with permission)

should be attempted if safely possible. However, if there is any adherence or invasion of tumor into the pituitary stalk, one should leave adherent remnants to the stalk if the patient wishes for normal gland function and the ability to have children. However, if the tumor is transinfundibular in nature, it may not be possible to preserve the stalk in order to remove the tumor, and stalk sectioning may be considered if gross total resection can be performed.

18.5 Suprasellar Craniopharyngiomas: Editors' Commentary

Although histologically benign, craniopharyngiomas represent some of the more daunting intracranial tumors due to their propensity for dense adherence to critical neurovascular structures. Management of these tumors typically is aimed at surgical gross-total resection (GTR) in order to prevent recurrence. Attempts at GTR, however, can be accompanied by significant morbidity from endocrinopathies, hypothalamic dysfunction, or neurovascular injury. Therefore, more recently, the emphasis in the treatment of craniopharyngiomas has shifted from the *need* to obtain a GTR to a paradigm that focuses on maximal safe resection followed by radiotherapy as necessary for long-term tumor control.

Surgical treatment of craniopharyngiomas is aimed at pathologic diagnosis, relief of mass effect, restoration of any visual compromise, preservation of endocrinologic function, and prolonged oncologic control. This can be accomplished through multiple surgical approaches, which can be grouped into either transcranial or endonasal techniques. The transcranial approaches can be further subdivided into "minimally invasive" supraorbital craniotomies, traditional cranial base approaches, and interhemispheric transcallosal or transcortical transventricular approaches. Each of these has its advantages and disadvantages. The choice of surgical technique is dependent on a number of factors, including the tumor's radiographic characteristics, the patient's demographics and presenting symptoms, and the surgical team's experience. Often, the primary factor in surgical approach selection is the anatomical relationship of the tumor with the optic chiasm.

Regarding the traditional open cranial base approaches, these present familiar anatomy to the neurosurgeon, and their use over many decades has resulted in ample experience with these techniques. They generally allow for better vascular control and management of any potential large vessel injury compared to endonasal or supraorbital trajectories. Also, with the exception of those approaches that add extensive skull base osteotomies, postoperative cerebrospinal fluid leaks are less common.

In general, anterolateral approaches permit access to any lateral tumor extension beyond the internal carotid arteries and into the sylvian fissures. Subfrontal transbasal craniotomies allow resection of more midline tumors, and opening the lamina terminalis permits removal of tumor located within the third ventricle. These anterior and anterolateral corridors are best indicated for prechiasmatic tumors with a secondarily post-fixed chiasm. Posterolateral approaches through subtemporal transtentorial and posterior transpetrosal craniotomies, however, provide a method for accessing the retro- and infra-chiasmatic space, but traversing significant neurovascular structures and cranial nerves along this surgical corridor may limit its use. These traditional "open" cranial base procedures have some additional disadvantages, namely: potential difficulty in visualizing the interface between the tumor and the hypothalamus with more rostral tumor expansion due to the craniocaudal surgical trajectory, potential release of caustic cyst tumor fluid into CSF spaces due to wider CSF arachnoidal openings, brain retraction that can be minimized by potentially cosmetically disfiguring skull base osteotomies, and the need to work around or mobilize critical neurovascular structures to access the tumor.

To circumvent some of these limitations, the endonasal corridor and supraorbital craniotomies have been utilized with increasing frequency. The eyebrow or eyelid approach can be more cosmetic than the incisions used for open craniotomies, while providing a very low subfrontal approach. With the addition of an orbital osteotomy or the use of angled endoscopes, the degree of brain retraction can be further minimized. Frontal sinus management can be a large concern with the supraorbital craniotomy, and any wound healing issues can be particularly disfiguring. The surgical freedom may be limited, and it is often difficult to adequately visualize the retrochiasmatic space. The endonasal endoscopic technique, conversely, does provide access posterior to the chiasm as well as into the third ventricle. As a direct route along the long axis of the tumor, the endonasal approach affords excellent retrochiasmatic visualization, optimal cosmesis, and perhaps the best method to identify vascular perforators to the chiasm and infundibulum. With no brain retraction or manipulation of the optic apparatus, excellent visual outcomes can be achieved, and a more clear distinction can often be made of the tumor-hypothalamic interface. Despite these advantages, there is a higher CSF leak rate, the potential for sino-nasal complications, limited access to extension of tumor lateral to the internal carotid artery, and possibly a greater risk of postoperative endocrinopathies.

The index case (Fig. 18.1) demonstrates a multiloculated, partially cystic craniopharyngioma with intratumoral calcifications that is largely located in the midline suprasellar cistern. Although the lesion is retrochiasmatic in location and the sella is minimally expanded, there is a fairly sizeable space between the pituitary gland and the optic chiasm for the approach. Additionally, the intervals between the neurovascular structures in the basal cisterns are likely expanded, allowing for tumor resection through the corresponding triangles, and there is minimal rostral extension. These factors make this particular lesion amenable to removal through both transcranial and endonasal approaches, as detailed in the preceding sections. There is not significant intraventricular extension, and therefore only the interhemispheric transcallosal and transventricular approaches would not be considered appropriate for this lesion.

There are some additional characteristics of this particular case to be considered in deciding the optimal surgical approach. The patient's gender and age are critical to note as an emphasis should be placed not only on achieving a maximal resection but also in maintaining normal endocrinologic function in a female of child-bearing age. In such a patient, preservation of the pituitary gland architecture as well as the pituitary infundibulum should be attempted, if possible. Additionally, this patient has only minimal visual compromise on presentation. Therefore, it is especially critical that maximal efforts are made to not sacrifice any vascular perforators to the optic nerves and chiasm and to limit manipulation of these structures in order to ensure an optimal visual outcome.

This illustrative case is fairly representative of most de novo craniopharyngiomas in that it is located largely within the retrochiasmatic suprasellar cistern. As the long axis of the tumor is midline along the sino-nasal-sellar-hypothalamic corridor, we feel that it is best approached via an endoscopic endonasal transplanum transtuberculum approach. Although in experienced hands the anterior transcranial approaches could achieve excellent results, these may require manipulation of the optic chiasm, nerves, and tracts, thereby increasing the risks of vision loss and vascular injury. A multilayered closure of the dural and cranial base opening with a nasoseptal flap is essential to minimize the risk of a postoperative CSF leak.

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19 Petrous Apex Cholesterol Granulomas

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19.1 Petrous Apex Cholesterol Granulomas: Editors' Introduction

Cholesterol granulomas of the petrous apex are slow-growing, expansile, cystic lesions that contain cholesterol crystals within a thick capsule of chronic inflammation. These lesions are possibly caused by the obstruction of the petrous air cells. As a result, cholesterol crystals accumulate and cause chronic inflammation and skull base erosion [1, 2]. This "obstruction-vacuum" theory has been widely accepted but may be oversimplified, leading to the "exposed marrow" hypothesis [2]. Cholesterol granulomas in the petrous apex are distinct entities from the cholesterol granulomas that develop in the tympanomastoid area [2]. Symptoms include deep-seated pain, visual dysfunction, hearing loss, tinnitus, and multiple cranial neuropathies.

Cholesterol granulomas are typically hyperintense on T1 and T2 images without enhancement [3]. Bony erosion is best appreciated on CT imaging, with a specific focus on the clivus, sphenoid sinus, and petrous apex. Grossly, dark fluid and degraded blood products are contained within the cyst. Histologically, the cholesterol granulomas contain giant cells with cholesterol clefts surrounded by inflammatory cells with areas of subacute hemorrhage [4].

While asymptomatic lesions can generally be observed, surgical management remains the primary mode of treatment for symptomatic lesions. The goals of surgical intervention are cyst drainage and marsupialization to relieve mass effect and prevent recurrence, which may occur in up to 35% of patients [5]. Surgical options range from open middle and posterior fossa cranial base procedures to endoscopic approaches. In this chapter, the authors discuss the various approaches relevant to the case example described below (Fig. 19.1a–d).

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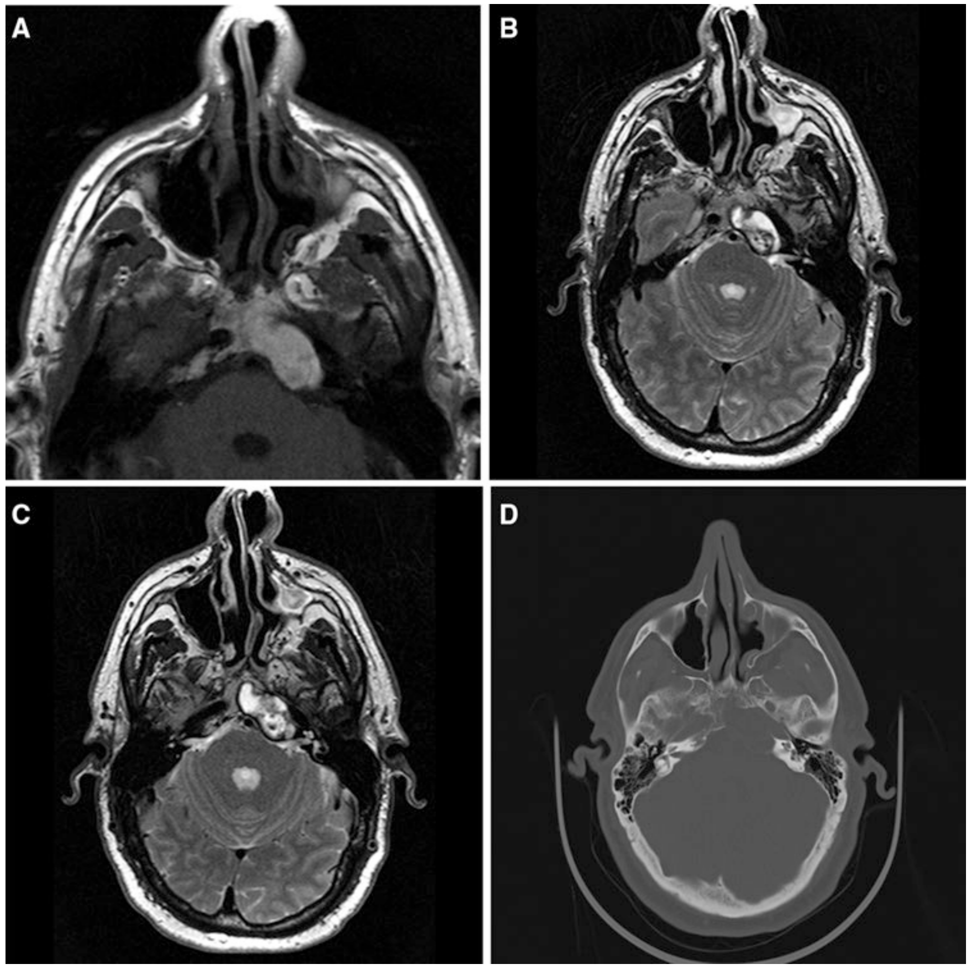

Fig. 19.1 (a–d) **CASE EXAMPLE**: petrous apex cholesterol granuloma. A 52-year-old man presents with diplopia and headaches. Examination: left sixth nerve palsy, otherwise neurologically intact

19.2 Petrous Apex Cholesterol Granulomas: Open Transcranial Approaches

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Introduction

Lesions of the petrous apex remain a challenging entity for all skull base surgeons. Cholesterol granulomas (CG) are no exception. Cholesterol granulomas comprise the most common cystic lesion of the petrous apex and typically affect young to middle-aged patients. The pathophysiology of CG involves a foreign-body, giant-cell reaction to cholesterol deposits, with fibrous and vascular proliferation. There are two prevailing theories of their genesis [2, 6]. One postulates there is occlusion of the air cell system, either from trauma or chronic Eustachian tube dysfunction, and air present in the cells is gradually resorbed, causing negative pressure, leading to hemorrhage within the air cells [7]. A second alternative involves spontaneous hemorrhage from residual rests of marrow within a well-pneumatized petrous apex. While some patients have a history of chronic ear disease, many do not and thus, the search for other possible explanations. Both of these mechanisms are likely to occur and may lead to CG formation. Presentation depends on the exact location they occur: the petrous apex, the middle ear, mastoid, or other pneumatized spaces of the temporal bone. If they occur in the petrous apex, patients are often asymptomatic but can present with headache, cranial nerve dysfunction (CN VI, VII, VIII), tinnitus, and dizziness, among other symptoms. A definitive diagnosis can typically be made with MRI alone, and CT can further clarify the extent of bone erosion. On MRI, CGs are generally hyperintense (bright) on TI, T2, and fluid-attenuated inversion recovery (FLAIR)-weighted sequences and without restriction on diffusion-weighted sequences. There may be peripheral enhancement after administration of gadolinium. CT scans show a nonenhancing cystic structure that involves the bone of the petrous apex causing osseous erosion with smooth peripheral margins and loss of internal bony septations. A subtle finding, often a clue to the diagnosis, is that often the contralateral petrous apex is highly pneumatized [6, 8, 9]. Other typical petrous apex lesions to consider include epidermoid, asymmetric bone marrow, chondrosarcoma, metastatic tumor, mucocele, or retained secretions.

Traditionally, CGs have been treated by drainage and stent placement via an infracochlear route. While immediate postoperative results have been acceptable, recurrence rates have been reported to be as high as 60% [10]. Modern thinking and experience show that many CGs do not require any treatment and are often incidentally discovered. For symptomatic lesions, proper preoperative diagnosis remains of the utmost importance. When treatment is indicated, CGs may be best managed through a more aggressive surgical approach for definitive management, with complete resection of the cyst contents and wall.

There remains controversy on the optimal surgical approach to definitively address these lesions. The surgical management of petrous apex CG is complicated by the proximity of critical neurovascular structures such as the petrous internal carotid artery (ICA), cochlea, internal auditory canal (IAC), superior petrosal sinus, cranial nerves IV–VIII, the greater superficial petrosal nerve (GSPN), and the brainstem. However, when these lesions become symptomatic, treatment is indicated, and the only definitive treatment is surgery. Understanding the natural history of these lesions is essential to set realistic treatment goals, as watchful observation is usually the best strategy when CGs are discovered incidentally.

Surgical Technique

Preoperative Evaluation

Patients who present with symptomatic CGs are evaluated with CT and MR imaging with and without contrast. Preoperative audiometric evaluation provides baseline hearing analysis, and if the patient complains of diplopia, preoperative neuro-ophthalmologic evaluation with Lancaster red-green testing can provide objective measures of abnormalities of extraocular movements. Depending on the surgical approach, intraoperative electromyographic monitoring of cranial nerve V, VI, and VII is essential. Choosing the surgical corridor to use depends on the specific location of the lesion (e.g., superior or inferior petrous apex), patient-specific temporal bone structure (e.g., high jugular bulb, poorly pneumatized mastoid), preexisting cranial nerve deficits, and patient health.

Anterior Petrosectomy (Figs. 19.2 and 19.3)

Indications

The authors' choice for the complete resection of CG of the petrous apex is the extended middle fossa, or anterior petrosectomy, approach. The anatomical borders of the anterior petrosectomy are the ICA with the overlying GSPN laterally, the cochlea and IAC posteriorly, the superior petrosal sinus medially, and the trigeminal ganglion and third division of the trigeminal nerve anteriorly [11]. One of the major advantages of the middle fossa approach, as well as the retrosigmoid approach, is that it provides extended access to the lesion with very little risk of hearing loss. Additionally, the operative corridor can be extended further by drilling of the labyrinth if the patient has profound ipsilateral deafness and vestibulopathy on presentation [11–13]. However, when hearing and balance function have already been lost, a more direct approach may be through the transcanal transcochlear/translabyrinthine approach with closure of the external auditory canal. Surgeon familiarity and comfort can reasonably play a role in this decision.

Technique

With the patient supine on the operating table in mild reverse Trendelenburg position, the head is placed in three-point fixation and turned ~60° to the opposite side. A curvilinear incision is made beginning in front of the tragus of the ipsilateral ear and extending up to the frontoparietal junction. The temporalis muscle is released and turned inferiorly leaving a cuff of muscle for later closure. A middle fossa craniotomy is turned 2/3 anterior and 1/3 posterior to the external auditory canal. Great care is taken to make the inferior bone cut parallel to the floor of middle cranial fossa. Using the operating microscope, the dura is carefully dissected from the floor of the middle cranial fossa working from posterior to anterior. The geniculate ganglion is identified with direct stimulation and protected. The GSPN is sharply dissected away from the dura, and the dura is elevated out to the petrous apex. The groove signifying the superior petrosal sinus is identified, and a self-retaining retractor is placed with the tip of the blade in this groove. After dissecting the foramen spinosum, the middle meningeal artery is coagulated and divided. After finding the foramen ovale, the GSPN is further dissected off the dura up to this margin. The petrous carotid can be exposed by drilling along the course of the GSPN in the confines of the posterolateral triangle. The third branch of the trigeminal nerve comprises the superior margin of this approach. Using a 3-mm diamond burr or an ultrasonic bone aspirator, the bone of the petrous apex is drilled away in a slow, meticulous fashion. The CG is widely opened in this fashion and then extensively debrided. Meticulous hemostasis can be obtained in this way, and the cavity is obliterated with a small autologous abdominal fat graft. The CG has typically eroded the bone to the posterior fossa dura, and the cyst wall can most often be dissected free from the dura, but if not, it is left in place rather than create a large dural defect, CSF leak, and intracranial communication with the cyst contents.

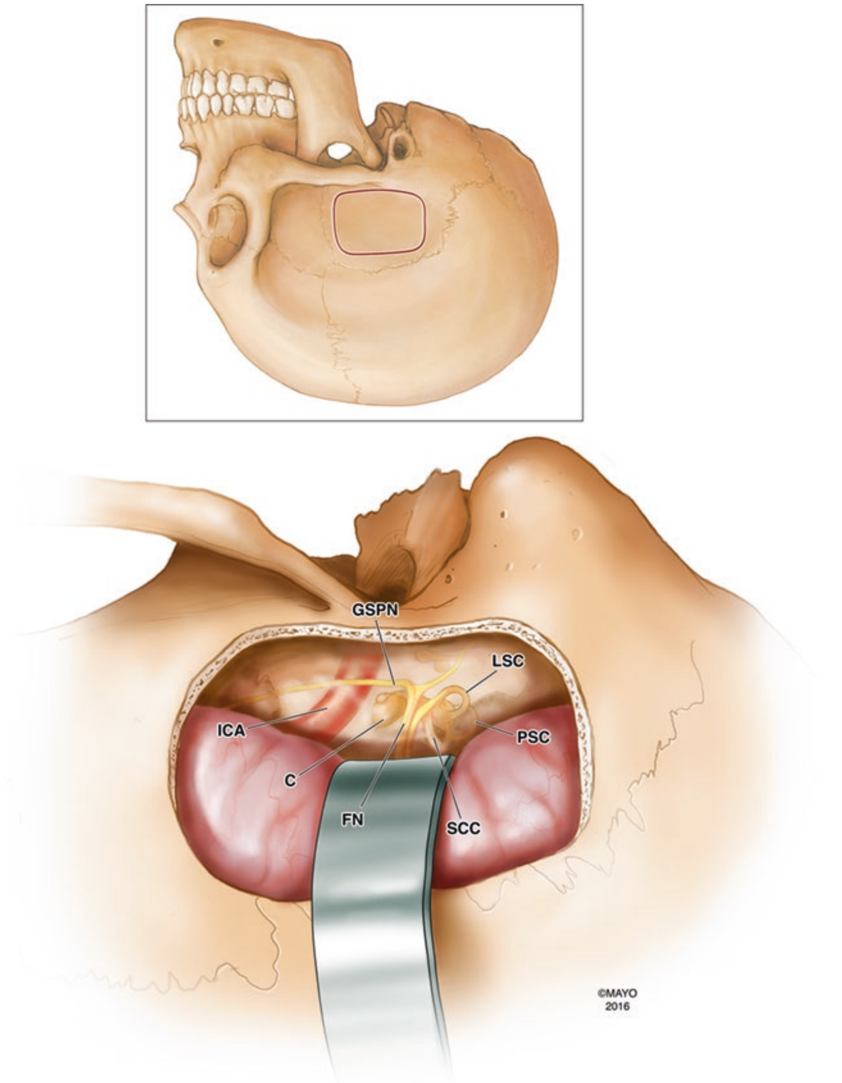

Fig. 19.2 Anterior petrosectomy. The operative anatomy of the anterior petrosectomy, the authors' preferred choice for definitive resection, is illustrated. *LSC* lateral semicircular canal, *PSC* posterior semicircular canal, *SCC* superior semicircular canal, *ICA* internal carotid artery, *GSPN* greater superficial petrosal nerve, *C* cochlear, *F* facial nerve (Used with permission of Mayo Foundation for Medical Education and Research. All Rights Reserved)

Retrosigmoid Inframeatal Approach

Indications

The retrosigmoid (RS) approach is very familiar to neurosurgeons and is a mainstay for microvascular decompressions and for resection of tumors in the CPA. It allows for wide exposure to the posterior fossa. The fourth through the twelfth cranial nerves are well visualized, but it is a moderately longer corridor to reach the petrous apex. As mentioned previously, it is a hearing preservation approach.

Technique

The transverse-sigmoid sinus junction is approximated using the intersection of a line connecting the inion to the root of the zygoma and a line conforming to the medial outer limit of the mastoid. A postauricular C-shaped incision is made approximately three fingerbreadths behind the ear. The soft tissue is reflected anteriorly, and the suboccipital musculature is reflected inferiorly. The bone is exposed from the asterion to just above the foramen magnum, with the arch of C1 palpable through the soft tissue. A burr hole is made just inferior to the transverse-sigmoid sinus junction, and a generous craniotomy is made. Further exposure with a diamond burr and a rongeur can help expose the edges of the transverse and sigmoid sinuses. It is helpful to keep the sinuses almost entirely covered in bone to help prevent inadvertent injury during retraction or by surgical instrument introduction/removal. The dura is opened in a semilunar fashion based on the edges of the transverse and sigmoid sinuses. The cerebellum is gently retracted, and the arachnoid of the lateral cerebellomedullary cistern is sharply opened to allow for decompression of CSF. Drilling of the inframeatal zone starts inferolaterally to the internal auditory canal. Using a diamond burr, the bone of the inframeatal ridge is removed,

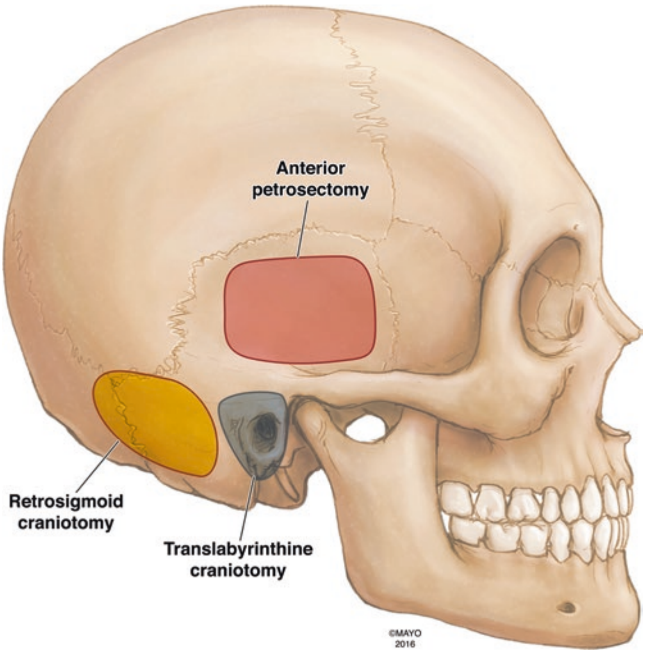

Fig. 19.3 Overview of various surgical approaches to the petrous apex (Used with permission of Mayo Foundation for Medical Education and Research. All Rights Reserved)

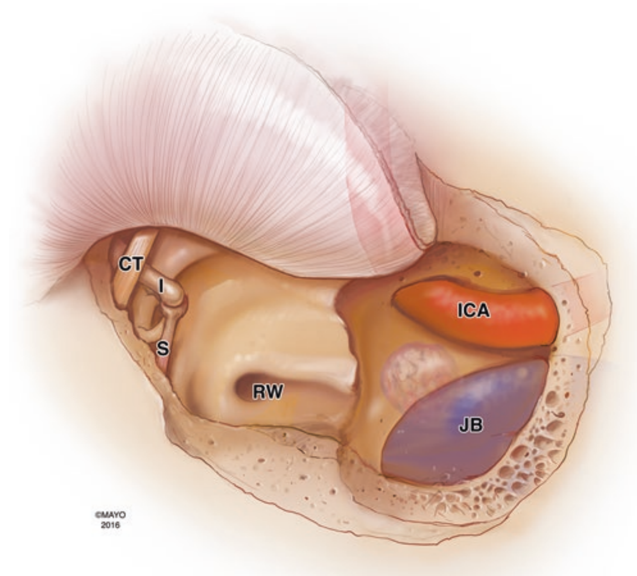

Fig. 19.4 Transcanal Infracochlear. The operative anatomy of the transcanal infracochlear approach is illustrated. *ICA* internal carotid artery, *JB* jugular bulb, *RW* round window, *S* stapedius tendon, *I* incus, *CT* chorda tympani (Used with permission of Mayo Foundation for Medical Education and Research. All Rights Reserved)

exposing the petrous apex. The entry point is located between the CN VII/VIII complex and the lower cranial nerves. A 30° endoscope is often helpful in this scenario to inspect for residual lesion and to remove the most anterior portions of granuloma.

Transmastoid Infralabyrinthine

Indications

This approach allows for access to the petrous apex through the retrofacial air cell tracking inferior to the labyrinth. It is an anatomic area familiar to neurotologists and carries a low risk of hearing loss, vestibular dysfunction, facial nerve weakness, or carotid injury. The jugular bulb and sigmoid sinus are at increased risk compared to the middle fossa approach, and the access to the lesion is limited [11].

Technique

For this approach a wide mastoidectomy is performed. The posterior and horizontal semicircular canals are identified, and the facial nerve is skeletonized along its mastoid segment. A communicating passage can then be made through the bone between the labyrinth superiorly and the jugular bulb inferiorly until the cholesterol granuloma is entered. This is a very narrow working corridor on the order of 2–3 mm. This allows, typically, adequate drainage of a CG but does not provide the ability to work in the cavity to completely exenterate the lesion, etc. Similar to other "minimally invasive approaches," careful examination of preoperation imaging, in particular a thin-cut CT scan, is important to assess for a high-riding jugular bulb [14]. If this is encountered, infralabyrinthine access is limited. Preoperative measurements of the distance between the labyrinth and the jugular bulb are helpful. Studies have shown that a distance of less than 1 cm is inadequate to satisfactorily drain cholesterol granulomas and that another approach, such as the transcanal infracochlear approach, is more appropriate [15]. This approach can be combined with the middle fossa approach to create a pathway for drainage when there is incomplete removal of the cyst wall.

Transcanal Infracochlear (Fig. 19.4)

Indications

This particular approach is best suited for a lesion that occupies the inferior aspect of the petrous apex and for patients who may not tolerate more invasive procedures. In general, this is one of the most common approaches chosen for drainage of CGs of the petrous apex, as it anatomically provides a dependent position for drainage to a well-aerated region proximal to the Eustachian tube and provides for relatively easy re-exploration via a transcanal approach [11, 12]. That said, if the transmastoid infralabyrinthine approach will allow access to the CG, it is increasingly preferred by many. It is efficient, offers quicker recovery, avoids risk to the carotid, obviates the need for a wide canalplasty, and has no risk of conductive hearing loss due to tympanic membrane perforation or poor healing.

Technique

A C-shaped postauricular incision is made, similar to that used for standard mastoid surgery. The external auditory canal is entered just medial to the bone-cartilage junction, and a tympanomeatal flap is elevated to expose the middle ear space. The anterior, inferior, and posterior portions of the ear canal skin are superiorly reflected to the level of the umbo. Utilizing a high-speed drill with continuous irrigation, the ear canal is widened to expose the hypotympanum. Care should be taken to not expose the temporomandibular joint anteriorly. In most cases the chorda tympani nerve can be preserved. Intraoperative facial nerve monitoring is used, but only in very rare cases does the facial nerve limit exposure. It is important to recognize that the facial nerve can be lateral to the annulus, particularly inferiorly. Access to the petrous apex is gained by traversing the space between the carotid artery, jugular bulb, and inferior aspect of the cochlea. This is a narrow space that even in the most favorable conditions can be exceedingly tight. A small diamond burr is used to skeletonize the carotid artery anteriorly, basal turn of the cochlea superiorly, and jugular bulb posteriorly. Special care must be taken during drilling to protect the cochlea, carotid artery, and jugular bulb. The opening is very narrow and precise drilling around these structures is essential. Dissecting medially becomes increasingly blind. Drilling posteriorly too far can result in encountering the posterior fossa dura and CSF leak. Too aggressive dissection anteriorly puts the carotid artery at risk, and superiorly, care needs to be taken to not enter the cochlea. A successful approach will allow entry into the anterior-inferior and posterior-inferior petrous apex. Opening into the cholesterol granuloma will result in drainage of some fluid, but it can be difficult or impossible to remove the more fibrous tissue typically found within these cysts. As mentioned, this approach is most often chosen for drainage of cholesterol granulomas of the inferior petrous apex and in patients who may not tolerate a craniotomy. The exposure obtained is inferior to that achieved through a middle fossa approach or a retrofacial approach in many cases. It is important to carefully study the anatomy on a high-resolution CT to determine which approach may be best suited to the clinical scenario. Preop 3-D modeling based on imaging and intraoperative image guidance can be useful adjuncts.

Lesion Resection

Care must be taken with drilling, as oftentimes much of the bone over the petrous apex has already been eroded by the lesion. Typically, during an anterior petrosal approach, after encountering the cholesterol granuloma, the cyst is opened, and its contents are evacuated via microsuction. On opening the cavity, there is oftentimes a dark thick fluid that drains, which may be under pressure. The cavity should be well irrigated. The authors' preference is to use a combination of sharp-angled curettes and an angled Takahashi or tumor forceps to debulk the granuloma and resect the cyst wall from the carotid artery and middle and posterior fossa dura with care not to cause a CSF leak. Oftentimes, a portion of lining of the cyst is quite adherent along the internal auditory canal and cannot be visualized from a middle fossa corridor. The use of a 30° or 45° endoscope may help to evaluate for any residual lesion. It is important to thoroughly and meticulously irrigate any bone dust from the cavity, as it may accumulate behind surgical corners. When resection is complete, generally a fat graft is placed in the bony defect; this may decrease the risk of re-accumulation of blood products. A thin layer of Tisseel (Baxter Healthcare Corp, Deerfield, IL, USA) fibrin sealant is placed over the covered resection cavity.

Stenting

Due to the high rate of recurrence with marsupialization, if surgical resection is not possible via an anterior petrosectomy, the authors advocate for drainage with the use of the largest possible silicone elastomer tube [Silastic® (Dow Corning, Midland, MI, USA), Silopren]. With the infracochlear approach, to prevent mucosal overgrowth of the catheter tip and stent occlusion, the lateral end of the catheter should extend just beyond the medial wall of the middle ear [16]. When utilizing the infralabyrinthine or transmastoid approaches, the newly created openings should be stented into the mastoid cavity. With the middle fossa approaches, the catheter can be placed along the tegmen into the epitympanum, although with this approach catheter drainage is almost never required because the lesion can be so effectively resected. If draining from the middle fossa is needed, this approach can be combined with the infralabyrinthine approach.

Recurrence

Optimal treatment for cholesterol granulomas of the petrous apex involves complete resection of the lesion and cyst wall through an open approach. This provides the least chance of recurrence. Recurrence rate for drainage procedures ranges between 0% and 62%, compared to recurrence rates for open resection between 0% and 5% (average, 3%) [7]. Residual granuloma and inadequate or blocked drainage or marsupialization by fibrous tissue are also causes of recurrence [10, 16, 17]. In fact, in studies presenting cases of complete resection, no recurrences were reported in the literature [10, 17–19]. Previous studies have also suggested the use of a vascularized galeofascial flap, or a pedicled strip of temporalis muscle, to prevent recurrence [7, 10]. Recurrence should be treated by open re-exploration and complete resection.

Patient Selection

Patients with asymptomatic lesions are managed nonsurgically, with routine imaging and follow-up to evaluate for the new onset of lesion growth or symptoms. As discussed, symptoms comprise a wide range, including hearing loss, loss of balance, tinnitus, headache, facial nerve or trigeminal dysfunction, otorrhea, and diplopia. Surgery is reserved for symptomatic or rapidly expanding lesions. Drainage and marsupialization are the current standard, with the goal to establish a permanent outflow pathway. Surgical approach selection is discussed in detail above, with important considerations that include location of the jugular bulb and internal carotid artery, hearing ability, general health of the patient, facial and/or trigeminal nerve involvement, and anterior extent of the lesion. In general, an infracochlear approach provides for the most dependent drainage, an infralabyrinthine approach is an alternative that is attractive when the jugular bulb is favorable, a middle fossa approach provides a broad, direct operative corridor for complete removal, a retrosigmoid approach provides for a familiar approach, and a translabyrinthine approach provides for the most direct route to the petrous apex when hearing is already lost. The authors suggest complete removal of these lesions including cyst wall resection whenever safely possible. Due to the high rate of recurrence with marsupialization, if surgical resection is not possible, the authors advocate for drainage with the use of a large silicone elastomer stent (Silastic, Silopren). It should be noted that in cases with significant intracranial extension into the posterior fossa, an open resection is recommended, as a drainage procedure would result in open communication with the dura and the risk of meningitis.

Surgical Outcomes (Tables 19.1 and 19.2)

Sweeney et al. presented the clinical and radiographic characteristics of cholesterol granulomas of the petrous apex, as well as the outcomes of operative and conservative management at two independent tertiary academic centers, including the authors' home institution [20]. Both adult and pediatric patients were included. All patients with less than 6 months of follow-up and those with iatrogenic postoperative cholesterol granulomas were excluded. Ninety petrous apex cholesterol granulomas were included (57.8% females, 55.6% right sided), 23 of which eventually underwent surgical intervention. Average age at presentation was 43.1 years (median 42.0, range 8.0–77.0 years). Signs and symptoms in decreasing frequency included headache (56.7%), dizziness (35.6%), facial paresthesia/pain (12.2%), hearing loss (6.7%), facial weakness (2.2%), diplopia (1.1%), and seizure (1.1%). Average lesion size in greatest diameter was 2.1 cm (median 1.7, range 0.7–5.0 cm). Mean follow-up was 46.0 months. No intraoperative complications were noted. In terms of outcomes, 23 patients (25.6%) required surgical treatment, most commonly for intractable headache. Surgical approaches included the transcanal infracochlear, the retrofacial infralabyrinthine, and the endoscopic transsphenoidal or transmaxillary approaches. The transcanal infracochlear and retrofacial infralabyrinthine approaches were the most commonly used lateral surgical approaches for cyst drainage (8 patients), whereas anterior approaches (endoscopic transsphenoidal or transmaxillary) were used at the same rate (8 patients). Only 47.8% of these patients experienced durable symptom improvement by their last follow-up. Among the patients who underwent primary observation, 85.7% reported no symptom progression or significant radiographic growth through an average of 45.5 months of follow-up (median 27.8, range 6.4–221.5 months). The authors concluded that most cholesterol granulomas of the petrous apex remain stable over time and can be safely managed with observation and routine follow-up. Surgery is reserved for lesions that are causing neurologic dysfunction from mass effect or erosion of critical structures such as the otic capsule. While cranial neuropathy associated with cholesterol granuloma is found to improve after surgical intervention, symptoms such as headache and dizziness were less likely to benefit from surgery.

Table 19.1 Patient characteristics at presentation

Variable	Value
Patients	
Total number	90 (100%)
Female	52 (57.8%)
Right sided	50 (55.6%)
Average age	43.1 year (median 42.0 year, range 8–77 year)
History	
Headache	51 (56.7%)
Dizziness	32 (35.6%)
Facial paresthesia/pain	11 (12.2%)
Hearing loss	6 (6.7%)
Facial weakness	2 (2.2%)
Diplopia	1 (1.1%)
Seizure	1 (1.1%)
Migraine history	17 (18.9%)

Used with permission from Sweeney et al. [20]

Table 19.2 Symptom improvement postoperatively in 23 patients who underwent surgical intervention

Symptom	Percent of patients improved
Headache	29.4
Dizziness	36.4
Facial weakness	50
Facial pain/paresthesia	83.3
Diplopia	100
Seizure	100

Used with permission from Sweeney et al. [20]

PEARLS
Cholesterol granulomas of the petrous apex are difficult lesions to access, and surgery should be reserved only for those cases that are symptomatic or show growth.
For complete resection of cholesterol granulomas of the petrous apex, the extended middle fossa approach, or anterior petrosectomy, is recommended to provide definitive care.
Special care must be taken during surgical exposure of the petrous apex, as openings are often narrow and precise drilling is necessary to protect vital structures, such as the cochlea, carotid artery, and jugular bulb.

Commentary on Case Presentation in Section 19.1

Case Presentation

A 52-year-old man presents with diplopia and headaches. Examination: left 6th nerve palsy, otherwise neurologically intact (see Fig. 19.1).

Discussion

The MRI shows a petrous apex lesion that is hyperintense on both T1 and T2 imaging. CT scan (bone window) shows a structure that involves the bone of the petrous apex causing osseous erosion with smooth peripheral margins and loss of internal bony septations. There are no other lesions or abnormalities identified on the imaging. Considering the differential, due to the specific imaging characteristics, we agree with the preliminary diagnosis of cholesterol granuloma.

Our next step would include a detailed history and physical examination. If an asymptomatic lesion that is stable in size is encountered, the authors would advocate for close observation. In light of the patient's 6th nerve palsy, as well as headaches and diplopia, the recommendation would be for surgical intervention for this symptomatic lesion.

On careful examination of the preoperative imaging, the lesion extends into the clivus. The anterior extension of this lesion makes it very amenable to an endoscopic transnasal, transclival approach for drainage. Upon recurrence, it would be recommended to undergo an anterior petrosectomy for resection (procedure outlined above). In this way, the capsule could be widely resected, and the petrous apex could be packed with subcutaneous fat to provide definitive cure.

19.3 Petrous Apex Cholesterol Granulomas: Endoscopic Endonasal Approaches

Daniel M. Prevedello, Ana Belén Melgarejo, Laila Pérez de San Román Mena, and Ricardo L. Carrau

Introduction

Advances in minimally invasive surgery including an improved understanding of endoscopic skull base anatomy, intraoperative image guidance, and the description of surgical approaches to the ventral skull base have led to new endoscopic treatment options for petrous apex cholesterol granulomas (PACGs). Advantages of endonasal approaches include avoidance of a craniotomy, minimal postoperative symptoms, faster recovery, cosmetically acceptable results, shorter hospitalization, and also the possibility for the reestablishment of a natural drainage pathway into the nasopharynx.

Montgomery [21] was the first to describe the transsphenoidal approach to the petrous apex in 1977. Fucci et al. [22] reported the first endoscopic endonasal approach (EEA) for addressing a giant cholesterol cyst of the petrous apex in 1994. Since then, numerous groups have reported treatment of cholesterol granulomas (CGs) via EEA, establishing it as another surgical option for the treatment of this pathology.

The petrous apex (PA) is one of the least accessible regions of the skull base. Vital structures are located in its vicinity, and therefore a thorough understanding of the surgical anatomy is mandatory. As previously described by Scopel et al. [23], the PA can be divided into three zones: superior PA, between the level of Dorello's canal and foramen lacerum; anterior-inferior PA, between the foramen lacerum and the carotid foramen; and posterior-inferior PA, between the carotid foramen and the jugular foramen.

Zanation et al. [24] described three endoscopic approaches to reach the PA: (1) medial transsphenoidal approach, for lesions that extend medially and/or reach the sphenoid sinus; (2) medial approach with internal carotid artery (ICA) lateralization, for lesions with a more posterolateral location, minimal medial expansion, or when the contour of the lesion cannot be appreciated because of poor pneumatization of the sinus; and (3) transpterygoid infrapetrous approach, when the lesion cannot be reached through the sphenoid sinus.

Paluzzi et al. [25] developed an algorithm to guide selection of the best approach for PACGs. They recognized three types of CGs based on their location in relation to the ICA. Type A includes larger CGs that extend medially and can be approached through a transclival approach, with the possibility of lateralizing the paraclival ICA to gain few more degrees if necessary. Type B includes CGs with lateral and inferior extension below the petrous ICA, which therefore requires an infrapetrous approach. And type C refers to very lateral CGs for which one should consider a lateral approach. This algorithm is based on a retrospective review of cases in which the type of approach was chosen depending on radiological criteria, namely, the relationship between the CG and the carotid artery.

Surgical Technique (Fig. 19.5a, b)

As in any routine endoscopic surgery, the patient is positioned supine with the head slightly rotated toward the right side and fixed with a Mayfield head holder. The skull base endoscopic team is located at the patient's right side for right-handed surgeons. Neuronavigation is always used to identify the course of the ICA and PA areas, and neurophysiologic monitoring (SSEPs and cranial nerve VI monitoring) is always utilized due to the potential need for manipulation of the ICA and the proximity of the surgical approach to cranial nerve VI. Topical oxymetazoline (0.05%) is used for vasoconstriction. Although PACGs may be approached by a unilateral sphenoidotomy, performing bilateral sphenoidotomies is thought to be more appropriate when working in this region as it allows four-handed surgery. Removal of sufficient bone at the skull base is also necessary to create a wide surgical corridor that exposes key anatomical landmarks and prevent crisscrossing of instruments. Moreover, it minimizes soiling of the endoscope lens, allows dynamic movement of the scope, helps maintain an unobstructed view of the surgical field, and increases lateral angulation. Furthermore, bimanual dissection is especially important for managing significant bleeding as it allows visualization while controlling the hemorrhage, thus avoiding injury to adjacent structures.

Medial Approach (Transsphenoidal Transclival)

Bilateral sphenoidotomies are performed with wide resection of the sphenoid rostrum and posterior septal attachments. The vascular pedicle for a septal mucosa flap can be preserved on the opposite side of the lesion in case it is needed.

Anatomical landmarks within the sphenoid sinus are identified: planum sphenoidale, sella, clival recess, and optico-carotid recess. Septations within the sphenoid sinus are removed. Also, the paraclival ICA course can be identified. Usually PA lesions are apparent due to medial bony expansion at the level of the clival recess.

If the lesion expands into the sphenoid sinus, the overlying mucosa is stripped, and the bone overlying the cyst is thinned with a 3- or 4-mm coarse diamond burr. Drilling starts on the medial surface of the bony deformity and runs along a vertical plane parallel to the course of the ICA. Once the bone is thin enough to fracture, small pieces of bone are removed with a 1-mm or 2-mm angled Kerrison rongeur, therefore leaving the lesion completely unroofed. The cyst wall is opened and its content is evacuated with suction, curettes, and irrigation. Wide communication with the cystic cavity is achieved after evacuation of the contents.

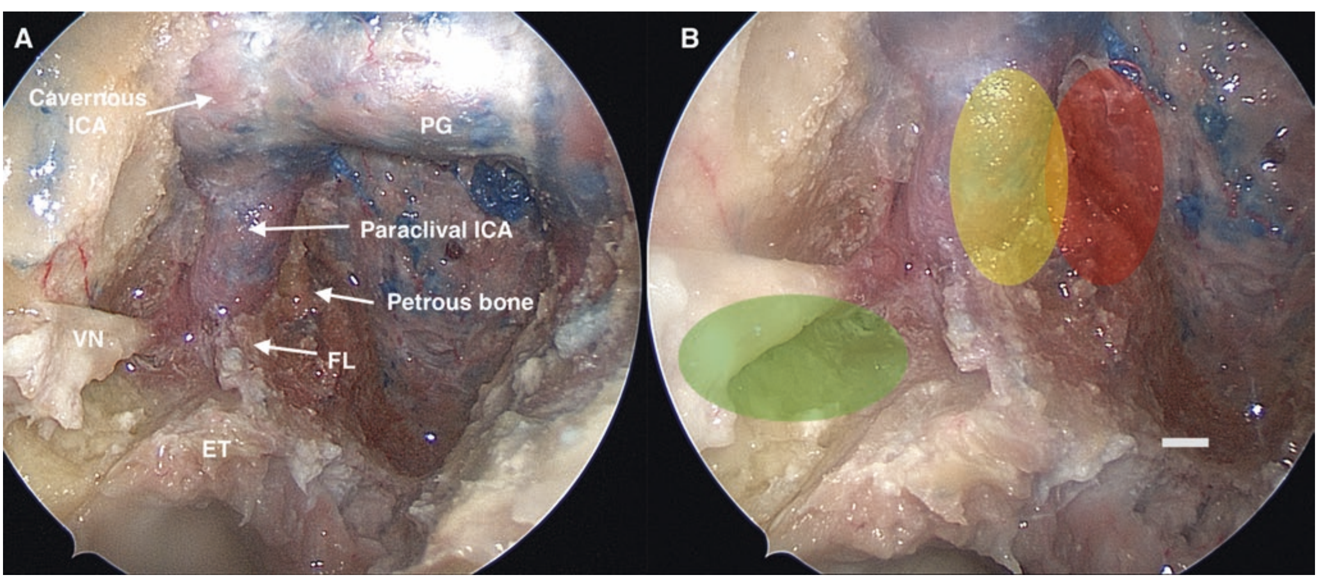

Fig. 19.5 (**a**) Anatomical image under 45° endoscopic endonasal view of the right petrous apex region. (**b**) Illustrative image of different PACG locations and their ideal approaches. *Red circle* represents a CG with medial extension (medial approach). *Yellow circle* represents a posterolateral CG to the ICA (medial approach with ICA lateralization). *Green circle* represents inferolateral CG below the petrous ICA (infrapetrous approach). *PG* pituitary gland, *ICA* internal carotid artery, *VN* vidian nerve, *ET* eustachian tube, *FL* foramen lacerum

Medial Approach with ICA Lateralization

When the contour of the PA lesion cannot be appreciated due to poor pneumatization of the sphenoid sinus, minimal medial expansion of the cyst, or a more posterolateral location of the cyst, additional bone removal is necessary. Decompression of the ICA allows lateral displacement of the artery and creates a larger medial window. Greater access can also be provided by thinning the clivus with the drill until the dura is exposed.

Key landmarks for locating the ICA include the vidian artery and nerve. A pterygoid approach is used to identify the vidian artery and trace its course to the lacerum segment of the internal carotid artery. A middle meatal antrostomy is first performed, and the sphenopalatine artery is located at its exit from the sphenopalatine canal. Overlying palatine bone is removed to expose the medial contents of the pterygopalatine space. The posterior nasal and sphenopalatine arteries are cauterized and transected, and the soft tissues are elevated from the underlying bone to expose the base of the pterygoid. The tissues are dissected laterally until the vidian artery and nerve can be visualized exiting from the pterygoid canal. Bone medial and inferior to the canal is removed with a hybrid diamond drill until the course of the ICA can be delineated at the lacerum foramen posteriorly. The bone overlying the vertical paraclival segment of the ICA can then be thinned and elevated to completely unroof this segment of the carotid canal. This widens the exposure by several millimeters allowing lateral displacement of the paraclival vertical carotid segment. Then the PA is approached via this now expanded medial corridor as described above for the medial approach. Care should be taken to cover the ICA exposed in this approach. A free mucosal graft is enough in the majority of the cases. A nasoseptal flap could also be used if the case in consideration would require this type of reconstruction.

Transpterygoid Infrapetrous Approach

If the PA lesion is not accessible through the sphenoid sinus or if it is best approached inferiorly to the petrous ICA, a transpterygoid infrapetrous approach is required, as well as dissection of the eustachian tube (ET) and foramen lacerum. The vidian artery and nerve are transected, and pterygopalatine soft tissues are elevated from the base of the pterygoids in a medial to lateral direction until the second division of the trigeminal nerve is identified where it exits foramen rotundum. The vidian artery is traced back to the lacerum foramen where the ICA turns from its petrous horizontal segment to the paraclival vertical segment. Additional bone is removed from the base of the pterygoids and the superior aspect of the medial and lateral pterygoid plates. The cartilaginous segment of the ET is resected for approximately 1 cm. Dissection along the posterior edge of the lateral pterygoid plate exposes the third division of the trigeminal nerve. The inferior surface of the PA is reached by drilling the bone between the horizontal segment of the petrous ICA and the ET, medial to the third division of the trigeminal nerve. Once the vertical and horizontal segments of the ICA are visualized, then drilling proceeds inferior to the petrous carotid into the PA. Here the cyst cavity is widely opened and its contents are evacuated with angled suctions and irrigation. It is also possible to use a 30°–45° endoscope to explore the cavity and ensure complete resection.

In order to avoid postoperative stenosis and preserve a drainage pathway, two main techniques have been described—the use of silastic stents and vascular pedicled nasoseptal flaps (NSF). Both techniques can be used concomitantly in cases where the opening is large and satisfactory.

Silastic stents can be placed into the opening to maintain patency during the healing process (Fig. 19.6). There are multiple options for stents including a red rubber catheter, mushroom, gastrostomy tube, and neonatal endotracheal tube. The stent is usually removed in the office, approximately 3–6 months following surgery, unless the presence of adhesions required its removal to be performed under general anesthesia. However, no consensus exists on the role of stenting for skull base CGs and it is mainly at the discretion of the surgeon.

In recent years, the use of vascular pedicled NSF has been described, proving to be helpful in avoiding postoperative scarring and subsequent restenosis while ensuring ventilation and drainage of the cyst. The NSF has to be placed inside the cavity, in contact with denuded bone after the sphenoidal mucosa has been removed in order to allow good integration and avoid mucocele formation. As Karligkiotis et al. [26] described, it may not be necessary to cover the whole surface of the CG cavity with the flap. In all cases, but especially in those with narrow opening, it is enough to cover the edges of the marsupialized cavity for at least 180 degrees of its circumference. This interrupts the circular reepithelialization and, when ipsilateral, also protects the exposed ICA.

Once the cyst cavity has been widely opened, we prefer to combine the use of silastic stent and vascular pedicle NSF, which will be located at the floor of the cavity. Sometimes an absorbable polyglycolic acid sheet and fibrin glue can be applied to prevent flap mobilization or stent migration. In case a wide aperture cannot be performed to nicely accommodate the flap, a silastic stent will be used alone. With this technique there is no need to remove the walls or capsule of the cystic cavity as it is marsupialized into the sphenoid sinus. It is important to avoid the use of free grafts in this case, as it may induce the formation of scarring and/or obstruction.

Preoperative Evaluation and Postoperative Care

Preoperative assessment is based on nasal endoscopy and neuroimaging. All patients must receive radiological assessment by computed tomography (CT) scan and/or magnetic resonance imaging (MRI) to better evaluate the bony boundaries,

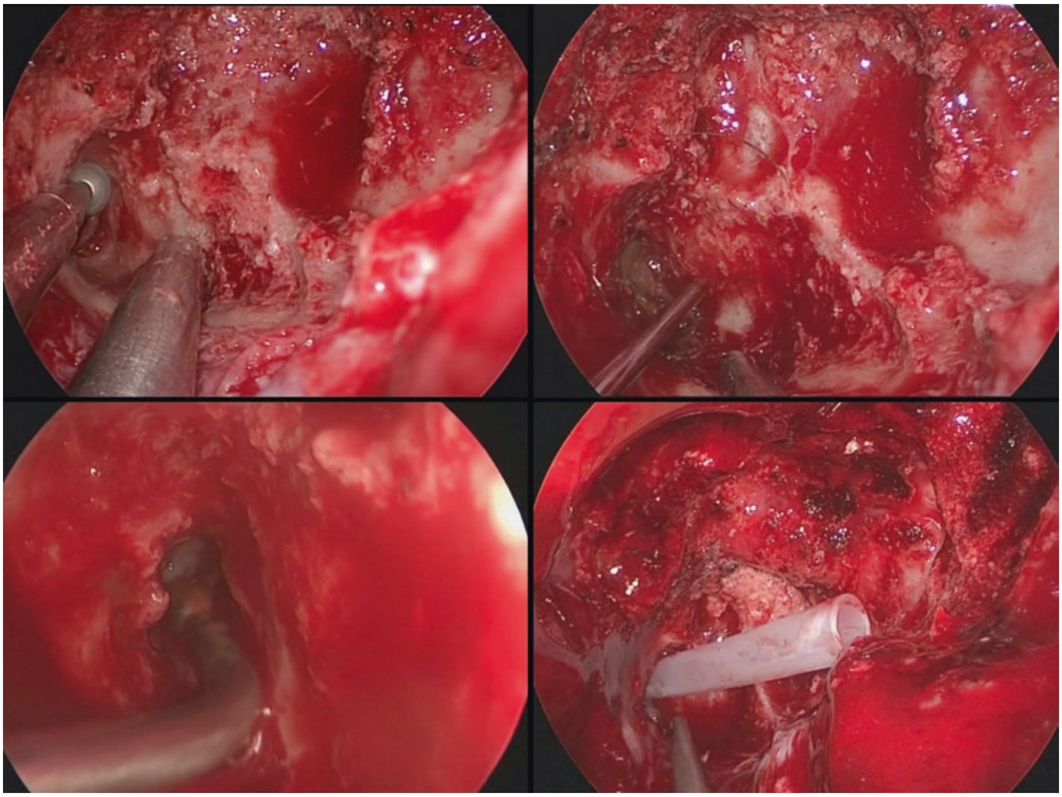

Fig. 19.6 Intraoperative pictures of an infrapetrous approach for the treatment of a right PACG. Curettes were used for denuding the cyst cavity, and finally a silastic tube was placed in order to maintain potency of the new cavity created

the extension of the lesion, and its position relative to adjacent structures. The medial expansion and the relationship to the ICA are highly significant in determining the best surgical corridor. All patients have to be fully informed about the method of treatment and give their written consent.

The postoperative care of patients undergoing this procedure is routine for endoscopic sinus surgery and consists of periodic office visits for removal of crusts until healing is complete and for checking the patency of the drainage tract. Examinations should be performed 1 week, 3 weeks, 2 months, and 4 months following surgery. If necessary, until full healing is observed, examinations every 3 months during the first year will be carried out. A CT will be performed in all patients in the immediate postoperative period in order to rule out complications and check the correct placement of the silastic tube. Later on, MRI will be performed for the follow-up to rule out recurrences.

Complication Avoidance and Management

Complications may be classified as vascular, neurological, and functional.

Among the vascular complications, injury to the ICA would be the main concern, which is increased in the infrapetrous approach. The knowledge of the course of the ICA and its landmarks are of paramount importance [27]. If carotid injury occurs, the surgeon should control the bleeding by compressing the vessel with pledgets immediately followed by exchanging it for a muscle graft that is kept in position for several minutes. Packing should then be used to keep reasonable pressure in the area. After bleeding is controlled, an angiogram should be performed to investigate for the presence of a pseudoaneurysm at the site of the vascular injury and to evaluate the collateral circulation. ICA endovascular stenting should then be considered to facilitate endothelial healing and complement the repair. All endoscopic surgery should be performed at an institution with immediate availability of endovascular neurointerventionalists for immediate angiography if damage to the ICA is suspected. Injury of the internal maxillary artery (IMA) and its branches may also occur when dealing with this pathology, which may result in a significant intraoperative bleeding. If muscle and packing are not sufficient to control the bleeding, then clipping of the vessel trunk may be preferred. Neuronavigation and Doppler are useful supports for early identification of the vessels.

Regarding the neurological complications, when approaching this region we should bear in mind the potential risk of injury to the 6th cranial nerve as it courses around the posterior aspect of the PA as it ascends to Dorello's canal, resulting in diplopia. In addition, injury to the vidian nerve may result in reduction of lacrimation function and loss of emotional tearing ipsilaterally. Vidian nerve function should not be sacrificed in a patient with dry eyes or V1 hypesthesia. There is also a risk of injury to the maxillary and mandibular divisions of the trigeminal nerve resulting most of the times in facial dysesthesia. CISS/FIESTA sequence MRI can help to delineate the course of the cranial nerves in relation to the lesion in order to better plan the approach.

Considering functional complications, we should take into account that the resection of the cartilaginous portions of the ET may result in a serous middle ear effusion that has to be treated with a tympanostomy tube. This occurs in less than half of the patients. Trismus is common after dissection of the pterygoid muscles. Analgesics and stretching exercises should be started early in the postoperative period to avoid permanent and progressive scarring of the muscles that could lead to severe limitation of oral opening. CSF leak is extremely rare when dealing with this kind of pathology and was not found in any of the cases of our series.

All of these complications are diminished when surgery is performed by highly trained skull base teams and also based on the proper selection of the approach. For instance, CGs located very lateral will be better approach through open craniotomy in order to avoid injuring the ICA. This is one of the main anatomical limitations for the EEA.

Patient Selection

Although CGs are benign lesions, they can behave aggressively and depending on the anatomical location and involvement of adjacent structures, clinical manifestations can be present. Therefore, there are two main treatment strategies when dealing with this pathology: observation and surgical treatment.

The first option is recommended for asymptomatic patients who are diagnosed incidentally by imaging, for symptomatic patients who have poor general conditions, or elderly patients. Patients managed conservatively will undergo serial radiologic evaluation with MRI or CT imaging.

The surgical treatment is advised to patients with symptomatic disease, cranial nerve deficits, and extensive lesions and when growth of the lesion is documented.

Surgical Outcomes

The success of the surgical management of CG is dependent on the evaluation criteria. Demonstration of pneumatization of the cyst on follow-up has been considered a desirable result of drainage procedures; however, this is not achieved frequently and is usually not considered a criterion for success. Instead, the absence of further growth and relief or resolution of symptoms is considered signs of a successful procedure despite continued opacification of the cyst cavity [28]. In the series of Brackmann and Toh, 20 of the 21 patients had either decreased or unchanged cyst size but only 5 regained aeration [29].

The EEA for PACGs has a recurrence rate of between 12% and 15.7%. This is significantly lower than that reported following transcranial approaches, which can be as high as 60%. The factors that can contribute to this lower recurrence rate include a wider bony opening and, consequently, a more extensive removal of the cyst's anterior or medial wall afforded by the EEA. The superior visualization of the cavity using angled endoscopes enables management of remaining septations and/or debris, optimizing the drainage.

Silastic stents and drains have been used with varying success. In 2002, a series published by Brackmann and Toh [29] reported no recurrence of CGs when a stent was used, but a recurrence rate of 30% when no stent was inserted. Conversely, Mosnier et al. [30] reported that recurrence of the lesion most commonly occurred secondary to stent occlusion. Georgalas et al. [31] highlighted the importance of the location of the lesion. If the cyst is situated adjacent to the posterior wall of the sphenoid sinus, then marsupialization is likely to be more successful and amenable to endoscopic exteriorization that allows drainage via sinonasal mucociliary clearance. McLaughlin et al. [32] consider that the use of a stent to preserve a drainage pathway throughout the healing process may contribute to reducing recurrences. This is especially important with the infrapetrous approach since the path of dissection is deeper than that performed in the medial transsphenoidal approach and therefore may be at more risk of scarring.

Terranova et al. [33] reported a case using a vascular pedicle NSF, with 3 years of follow-up, without clinical signs of recurrence, and with an MRI that revealed a complete reepithelialization of the sphenoidal mucosa, complete integration of the septal mucosal flap with wide opening of the sphenoidal drainage pathway, and no evidence of recurrence of the disease. Karligkiotis et al. [26] described ten cases of GC in which they used a NSF and reported that nine cases were without evidence of disease after a mean follow-up 35.7 months; one case presented with recurrence due to a technical error in placement of the flap and was retreated endoscopically. Shibao et al. [34] described a surgical technique that they used to treat two patients with PAGCs. They combined a NSF with a silicone tube and reported no recurrence after 12 and 24 months of follow-up.

Table 19.3 presents a summary of 66 reported cases: a transsphenoidal approach was carried out in 26 cases, a transclival approach in 17, and an infrapetrous approach in 23. All of the patients had clinical improvement, except for eight cases where the details were not available. Ten cases developed recurrence and all of them were treated by endoscopic redrainage. There were six cases which developed

Table 19.3 Summary of cases using a transsphenoidal approach (Number of Cases, 26), a transclival approach (Number of cases: 17), and an infrapetrous approach (Number of Cases: 23)

Author	Cases	Preoperative symptoms	Procedure	Clinical outcome	Complication	Recurrence
Fucci et al. (1994) [22]	1	Headache	TS drainage of cyst; wide opening of the cyst cavity; placement of T-shaped stent	Improved	None	None
Griffith and Terrell (1996) [35]	2	1. Dysequilibrium 2. Hearing loss, V3 hypesthesia	1. TS drainage of cyst, wide opening of the cyst cavity 2. First: Subtemporal transzygomatic approach, second: TS drainage of cyst, third: TS drainage of cyst and wide opening of the cyst cavity	Asymptomatic	1. Transient epistaxis 2. None	1. None 2. Recurrence 2 months after first OR recurrence 2 monthsFU after second ORno recurrence after third OR
Michaelson et al. (2001) [36]	1	Headache, sixth nervepalsy	TS drainage of cyst, wide opening of the cyst cavity, marsupialization	Asymptomatic	None	None
DiNardo et al. (2003) [37]	1	Dysequilibrium	TS drainage of cyst, wide opening of the cyst cavity	Asymptomatic	None	None
Presutti et al. (2006) [38]	1	Headache, sixth nerve palsy, increasing vertigo	TS drainage of cyst, wide opening of the cyst cavity, placement of T-shaped stent	Asymptomatic	None	None
Oyama et al. (2007) [39]	1	Hemifacial pain	TS drainage of cyst, wide opening of the cyst cavity	Asymptomatic	None	None
Georgalas et al. (2008) [31]	4	Facial palsy, sensorineural hearing loss, vertigo, headache, sixth nerve palsy, visual loss	TS drainage of cyst, wide opening of the cyst cavity, marsupialization	Asymptomatic	None	None
Samadian et al. (2009) and (2015) [40]	4	Headache, sixth nerve palsy, hearing loss	TS drainage: 3 TS + stent: 1	Asymptomatic	None	None
Zanation et al. (2009) [24]	8	All had headache, two had occasional vertigo	TS: 2 TS with ICA lateralization: 3 IP approach: 3 drainage of cyst and stenting	No detail	None	One recurrence at 2.5 years after first OR
Jaberoo et al. (2010) [41]	2	Diplopia, dizziness, headache	TS and TC drainage of cyst, wide opening of the cyst cavity TP	Improved	None	None
McLaughlin et al. (2012) [32]	1	Headache, sixth nerve palsy	First: TS drainage of cyst, wide opening of the cyst cavity, partial marsupialization second: TS and IP for drainage of cyst, very wide opening of the cyst cavity,marsupialization, placement of a Doyle splint	Asymptomatic	After first OR: delayed epistaxis	Recurrence 4 months after first OR no recurrence yet aftersecond OR
Paluzzi et al. (2012) [25]	17	Headache, imbalance, diplopia, severe vertigo, ear pain, conductive hearing loss, facial pain, dizziness, tinnitus	TC + miniflap: 3 TC + mini flap + stent: 1 TC with ICA lateralization: 3 TC with ICA lateralization + stent: 2 TC-IP approach + stent: 8	All improved	Epistaxis, chronic serous otitis media and eye dryness, transient CN VI deficit	2 recurrence after 11 months and 2.8 years in 2 redrainage + stent (previous TC + ICA lateralization)
Sade et al. (2012) [42]	3	Headache, hearing loss, diplopia, dysequilibrium	TS: 1 TS + stent: 2	Improved	Chronic sphenoid sinusitis	3 recurrence, one patient after 7 and 13 months (TS) second after 5 months
Tomazic et al. (2013) [43]	4	VI palsy	TS and TC	Asymptomatic	None	None
Emanuelli et al. (2013) [44]	4	Headache, vertigo, tinnitus	Tc-IP	Improved	None	None
Shibao et al. (2015) [34]	2	1. Vertigo, hearing disturbance 2. Headache	1. First: TS drainage of cyst second: TS drainage + T-shaped stent + flap 2. TS drainage + flap	Improved	None	1. Recurrence 2 months after first OR
Karligkiotis et al. (2015) [26]	10	Headache, hearing loss, diplopia	TC + flap: 4 TP + flap: 6	Improved	None	One recurrence after 12 months (TP)

TS transsphenoidal, *TC* transclival, *FU* follow-up, *OR* operation, *IP* infrapetrous, *ICA* internal carotid artery complications: three experienced epistaxis, one chronic serous otitis media and eye dryness, one transient VI CN palsy, and one chronic sphenoid sinusitis.

PEARLS
Neuronavigation is always used to identify the course of the ICA and PA areas and neurophysiologic monitoring may be considered when manipulation of the ICA is anticipated.
PACG that extend medially enough, and/or reach the sphenoid sinus, a transsphenoidal approach is required.
PACG that do not extend into the sphenoid sinus may require a medial approach with ICA lateralization, mainly if the lesion is located posterolateral to the ICA or a transpterygoid infrapetrous approach for lesions located inferior to the petrous ICA.
In order to avoid postoperative stenosis and preserve a drainage pathway, two main techniques are recommended: the use of silastic stents and vascular pedicled nasoseptal flaps (NSF).

Case Examples

Case 1 (Provided by Editors in Section 19.1)

A 52-year-old man with diplopia and headaches. Examination: Left 6th nerve palsy, otherwise neurologically intact (see Fig. 19.5).

A medial approach would be the selected endoscopic technique. In this case, the PACG is located medial to the petrous ICA, extending into the clival region. Therefore, the medial extension of the cyst provides an optimal corridor to open the cyst without the need to lateralize the ICA or to perform an infrapetrous approach.

Case 2 (Provided by Authors)

A 49-year-old male with headaches, facial paresthesia, and dizziness. Physical examination was uneventful. MRI was performed showing an expansile lesion within the right PA compatible with a CG. EEA was carried out with an infrapetrous approach. Silastic stent and NSF vascularized on the left posterior nasal artery were used. Postoperative images showed decompression of the lesion and proper positioning of the silastic tube. The patient experienced improvement of his symptoms and the silastic tube was removed 6 months after surgery (Fig. 19.7a–e).

Case 3 (Provided by Authors)

A 65-year-old female who presented with hearing loss. No other neurological deficits were detected during physical examination. MRI showed a cystic lesion located at the left petroclival region. An EEA transclival transpetrous approach was performed. Skull base reconstruction was performed using an ipsilateral NSF and marsupialization with silastic tube inside the cystic cavity. Postoperative CT scan demonstrates adequate cyst decompression and proper positioning of the silastic tube, which was removed 6 months after surgery. No recurrences were found during the follow-up (Fig. 19.8c).

Conclusion

The EEA to PA lesions is safe and effective for appropriately selected patients in the hands of experienced endoscopic skull base surgeons. The approaches can be classified based on their relationship to the petrous ICA. Approach selection depends on the medial expansion of the lesion and the position of the paraclival carotid artery. EEA offers the advantage of avoiding hearing and potential facial nerve damage from transtemporal/transcranial approaches and allows for the possibility of reestablishing a larger and more natural drainage pathway into the sinuses.

Potential advantages of an endonasal approach (especially compared with a transcranial approach) include avoidance of a craniotomy, faster recovery, minimal postoperative symptoms, no external scars, lower recurrence rates, and shorter hospitalization.

19.4 Petrous Apex Cholesterol Granulomas: Editors' Commentary

Historically, the petrous apex has been a difficult region of the cranial base to surgically access. One of the most common lesions to occur in this location is the cholesterol granuloma (CG). These are often asymptomatic and discovered incidentally. In this subset of patients, CGs should be observed radiographically. In patients with referable symptoms, however, treatment is warranted and is typically performed through surgical drainage with resection of the cyst contents and wall. The ultimate goal is to establish a permanent outflow pathway to prevent recurrence.

Traditionally, treatment of CGs had been accomplished through transcranial approaches. Even with drainage and stent placement, recurrence rates have been reported to be as high as 60%. Surgical exposure of the petrous apex often results in relatively narrow openings and precise drilling is necessary to protect vital structures such as the cochlea, carotid artery, and jugular bulb. As described by the authors of Sect. 19.2, the lateral cranial base approaches include:

- Transcanal infracochlear approach – provides for the most dependent drainage, especially in patients who may not tolerate more invasive procedures

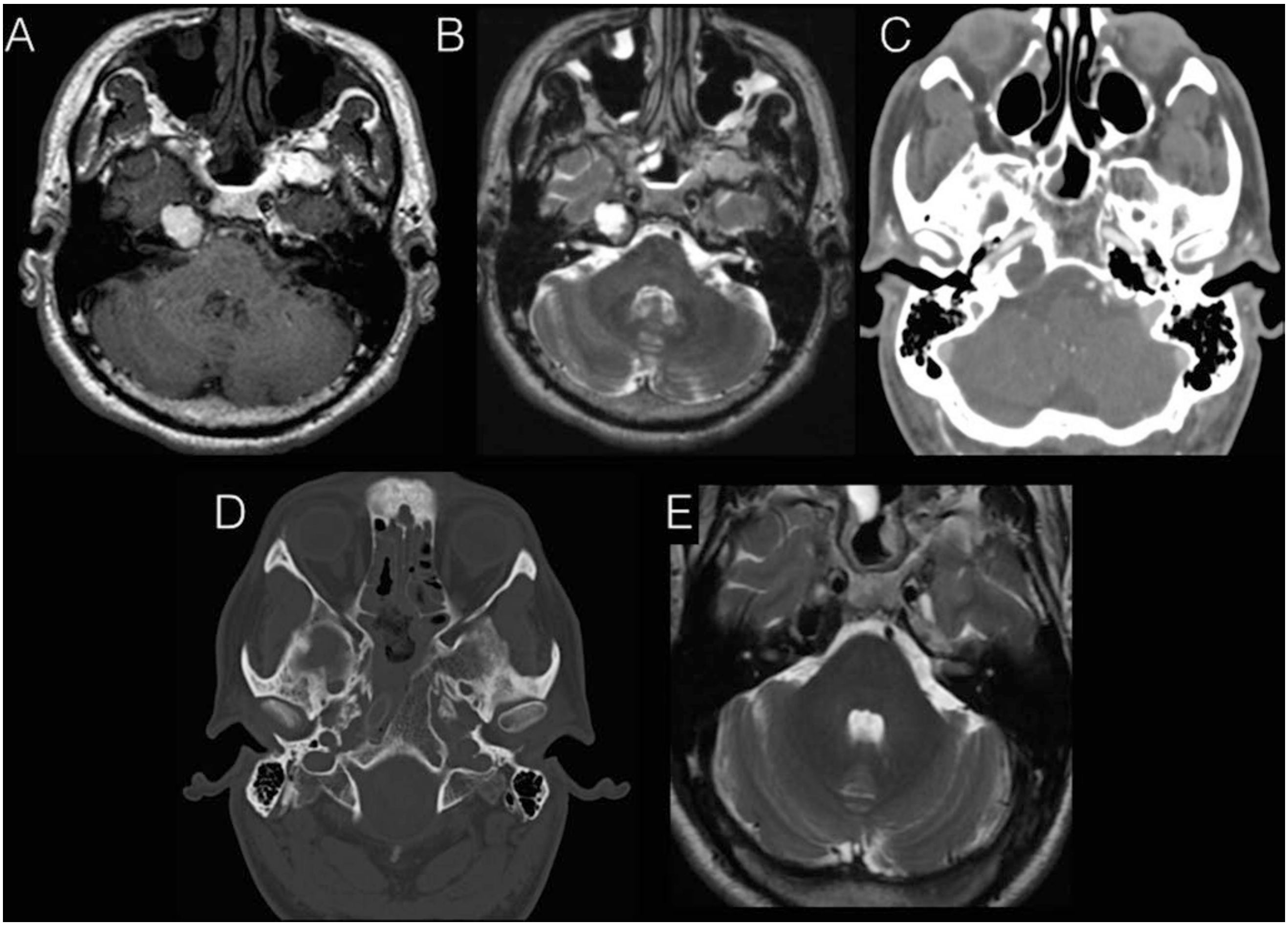

Fig. 19.7 Right PACG preoperative (**a**–**c**) and postoperative (**d**, **e**) images. (**a**, **b**) Axial T1-/T2-weighted MRI showing a hyperintensive signal lesion. (**c**) Axial CT angiogram showing an expansile cyst. (**d**) Postoperative CT scan showing the bone removed through an infrapetrous approach combined with a medial transclival approach (ICA lateralization was not necessary in this case) and the placement of a silastic tube into the cyst cavity. (**e**) Axial T2-weighted postoperative MRI showing decompression of the lesion

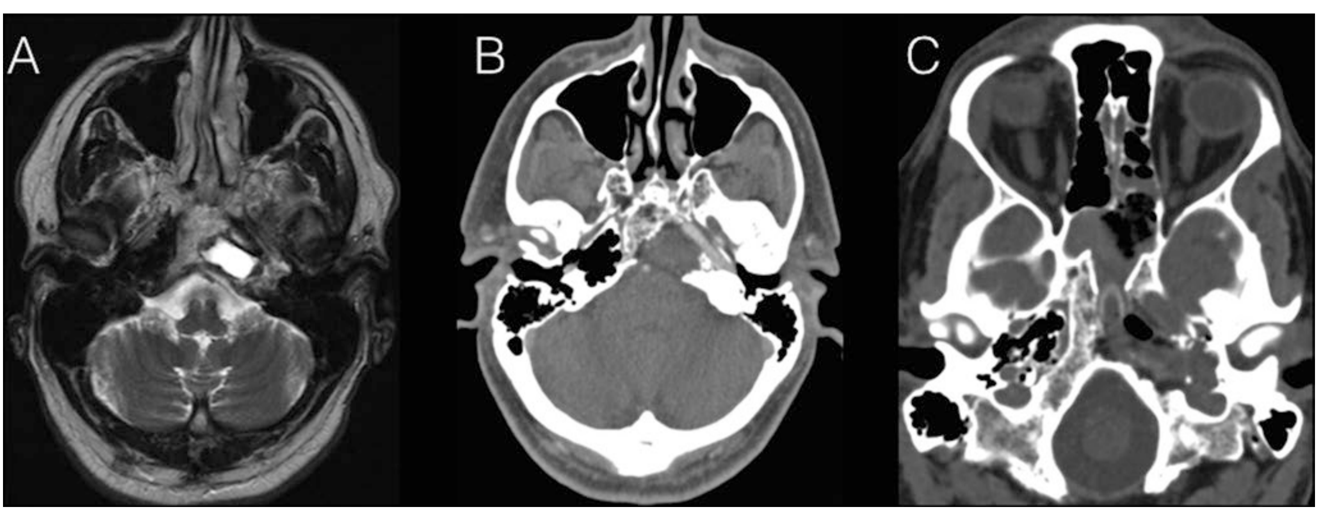

Fig. 19.8 (**a**–**c**) Left PACG. (**a**) Axial T2 preoperative-weighted MRI showing a high signal lesion. (**b**) Axial CT angiogram showing an erosive cystic lesion. (**c**) Postoperative CT scan showing optimal decompression and correct positioning of the silastic stent

- Transmastoid infralabyrinthine approach – when the jugular bulb placement is favorable, provides a surgical corridor familiar to neurotologists and carries a low risk of hearing loss, vestibular dysfunction, facial nerve weakness, or carotid injury, but provides somewhat limited access to the petrous apex
- Extended middle fossa/anterior petrosectomy approach – provides a direct operative corridor for complete removal with hearing preservation
- Retrosigmoid inframeatal approach – familiar approach to neurosurgeons that preserves hearing but is a longer course to the petrous apex
- Translabyrinthine approach – provides direct route to the petrous apex when hearing is already lost

Alternatively, the endoscopic endonasal approach (EEA) to petrous apex cholesterol granulomas can provide for direct access to these lesions with the potential for more reliable long-term drainage, resulting in lower reported recurrence rates (12–15%) as compared to transcranial approaches. Three distinct corridors can be utilized depending on the location of the lesion:

- Medial transsphenoidal approach – for CGs that extend medially and/or reach the sphenoid sinus
- Medial approach with internal carotid artery (ICA) lateralization – for CGs with a more posterolateral location, with minimal medial expansion, or with poor pneumatization of the sphenoid sinus
- Transpterygoid infrapetrous approach – for lesions located inferior to the petrous ICA

Currently, the endonasal approach is preferred for accessible petrous apex CGs due to its significantly lower recurrence rate. For the illustrative case provided, for surgical teams skilled in endoscopic procedures, an EEA would likely be the most appropriate approach due to the CGs medial extension and proximity to the sphenoid sinus, providing for a well-aerated surgical corridor to access the lesion and into which the lesion can be marsupialized and drained. Additionally, endonasal approaches avoid the potential morbidity and cosmetic deformity associated with a craniotomy, minimize postoperative discomfort, provide a faster recovery and, most importantly, allow for the establishment of a natural drainage pathway into the nasopharynx. The use of silastic stents and/or vascularized pedicled mucosal flaps may provide better long-term patency of the outflow tract. Endonasal approaches to the petrous apex, however, may be limited with infrapetrous and more lateral lesions and accessing these CGs endonasally may come with signficantly increased risk and morbidity such that lateral transcranial approaches may be more appropriate for CGs in these locations in order to optimize resection and prevent vascular injury.

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Pineal Region Tumors

20

Jeffrey N. Bruce* and Charles Teo*

Contents

Abbreviations

AFP	α-Fetoprotein
CSF	Cerebrospinal fluid
CT	Computed tomography
ETV	Endoscopic third ventriculostomy
EVD	External ventricular drain
ICP	Intracranial pressure
MRI	Magnetic resonance imaging
OTT	Occipital transtentorial
PPTID	Pineal parenchymal tumor of intermediate differentiation
SCIT	Supracerebellar infratentorial
VPS	Ventriculoperitoneal shunt
βHCG	β-human chorionic gonadoptrin

20.1 Pineal Region Tumors: Editors' Introduction

Tumors occurring in the pineal region are uncommon. The overall incidence of any tumor in the pineal region is 0.10 per 100,000 population [1]. These tumors account for 0.4% of all central nervous system (CNS) tumors overall but are more frequent (2.5%) in the younger population (0–19 years old) [1]. The most common presenting symptom of pineal region tumors is headache which may be related to obstruction of cerebrospinal fluid (CSF) flow. Gait ataxia and Parinaud's syndrome with upgaze palsy may also be present. Differential diagnoses of lesions in the pineal region include neoplasms, benign cysts, vein of Galen malformations, and inflammatory disorders. True neoplasms must be distinguished from benign pineal cysts, which have an incidence of 20–40% in both autopsy and high-resolution MRI studies [2–4]. A wide array of neoplasms can occur in the pineal region including germ cell tumors, pineal parenchymal

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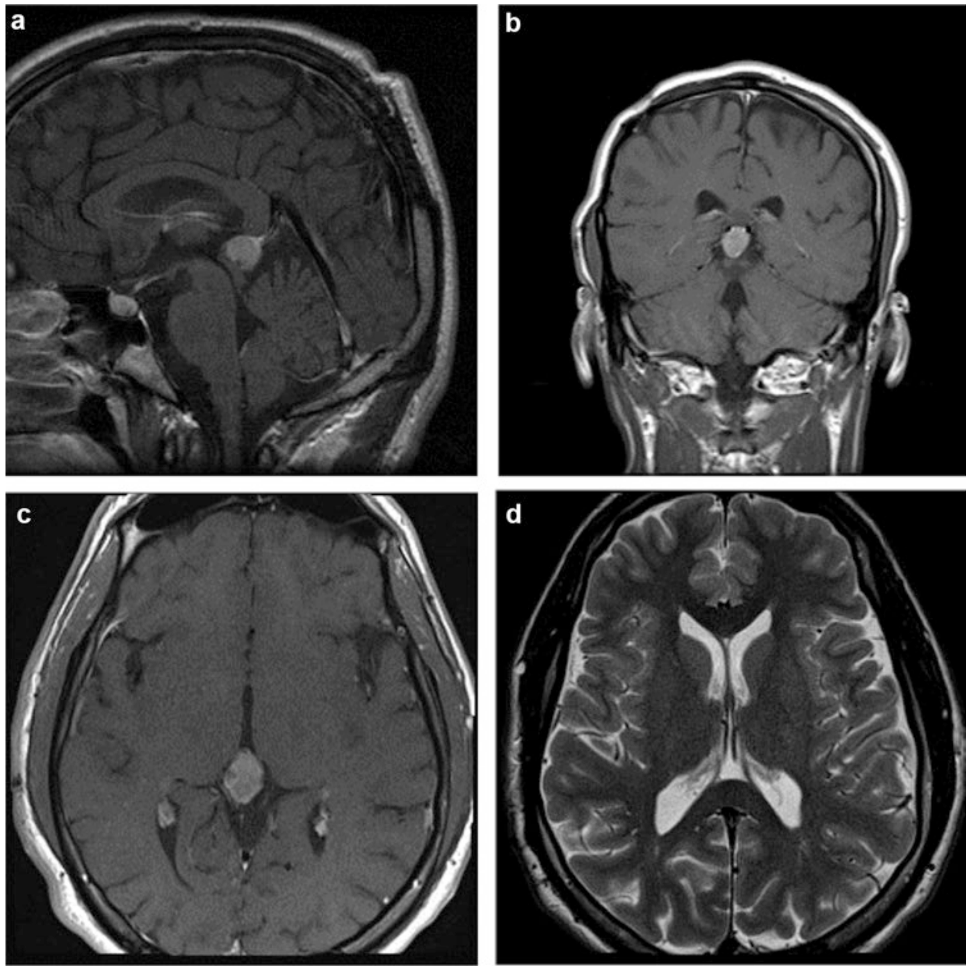

Fig. 20.1 **(a–d)** **CASE EXAMPLE**: pineal region tumor. A 19-year-old presents with an incidental lesion discovered after he sustained a concussion after a motor vehicle accident. He is currently asymptomatic. The lesion was initially followed but demonstrated growth in a 6-week follow-up scan, which was concerning for high-grade neoplasm. Serum markers were negative. No hydrocephalus

tumors, papillary tumors of the pineal region, meningiomas, astrocytomas, ependymomas, and metastases [5]. Germ cell tumors (GCTs) are the most common primary pineal region tumors. GCTs include germinoma, yolk sac tumors, teratomas, embryonal carcinoma, and choriocarcinoma [6]. Pineal parenchymal tumors are divided into three categories in the World Health Organization (WHO) classification: grade I, pineocytoma; grade II/III, pineal parenchymal tumors of intermediate differentiation; and grade IV, pineoblastoma [5, 7]. The diversity of tumors occurring in this region including the possibility of mixed pathology and malignant potential presents a diagnostic challenge for clinicians. Tumors that are symptomatic or show evidence of growth require tissue diagnosis and treatment. Incidentally discovered asymptomatic tumors can be typically observed with short-interval follow-up for growth. CSF may be obtained via lumbar puncture or ventricular sampling (when obstructive hydrocephalus is present) to evaluate cytology and the presence of hormonal markers. Tissue diagnosis can be obtained via an endoscopic transventricular approach, a stereotactic needle biopsy, or an open surgical approach. The most common open approaches include the occipital transtentorial and supracerebellar infratentorial approaches. Keyhole endoscopic variants of these approaches are also increasingly being utilized. In this chapter, the authors will discuss open transcranial, endoscopic, and keyhole approaches to the pineal region. Each author will discuss the relative merits of their surgical approach to the case example below (Fig. 20.1a–d).

20.2 Pineal Region Tumors: Open Transcranial Approaches

Randy D'Amico and Jeffrey N. Bruce

Preoperative Considerations

The pineal region gives rise to a variety of neoplastic and nonneoplastic lesions with individual therapeutic and prognostic implications. Neurosurgical intervention remains vital to the diagnosis and management of pineal region lesions, and surgical interventions range from biopsy to complete resection with variable benefit [8–12]. Choice of intervention is dependent on the relationships between the tumor and surrounding anatomic structures, the pathologic features of the identified lesion, and the comfort and experience of the operating surgeon.

Preoperative evaluation invariably begins with contrast-enhanced, cranio-spinal magnetic resonance imaging (MRI). Dedicated imaging of the spinal column is critical given the proclivity of malignant lesions to seed the spinal canal, as well as to avoid difficulties with interpretation of imaging studies obtained following intradural neurosurgical intervention. While MRI of the pineal region does not reliably differentiate histologic subtypes [13], it is necessary to plan the operative approach, identify relationships between the tumor and anatomic structures, and determine if safe, complete resection is possible.

Hydrocephalus

Obstructive hydrocephalus due to compression of the cerebral aqueduct or posterior third ventricle is present in most patients and should be managed prior to resection. Preoperative cerebrospinal fluid (CSF) diversion also offers an opportunity to send CSF for evaluation of tumor markers.

If urgent, bedside placement of an external ventricular drain (EVD) can be performed until a definitive treatment strategy is determined. Patients with chronic, symptomatic hydrocephalus classically underwent ventriculoperitoneal shunt (VPS) insertion prior to tumor resection. However, VPS insertion has been supplanted with preoperative endoscopic third ventriculostomy (ETV) as an effective treatment strategy. Preoperative ETV is safe and effective and eliminates the risks of shunt infection, shunt malfunction, overshunting, subdural hematoma, and peritoneal seeding of malignant cells, classically associated with VPS insertion [11]. Furthermore, ETV permits endoscopically assisted biopsy of pineal region tumors through the posterior third ventricle during the same procedure when indicated. Contraindications to ETV include the presence of tumor occupying the floor of the third ventricle or an unfavorable relationship between the floor of the third ventricle and the basilar artery.

Of note, in mildly symptomatic patients, complete resection of the pineal mass may circumvent the need for permanent CSF diversion. In these situations, the reduction of mass effect on the cerebral aqueduct, and communication of the third ventricle with the quadrigeminal cistern through resection of the lesion, restores the normal circulation of CSF.

Germ Cell Tumor Markers

The identification and accurate interpretation of germ cell tumor markers are critical components of the preoperative evaluation of pineal lesions (Table 20.1). The presence of germ cell markers is pathognomonic for malignant germ cell elements and may preclude surgical intervention for tumors with sensitivities to chemotherapy and radiation, with resection saved for residual or recurrent tumors only. However, management of hydrocephalus is still usually warranted.

Table 20.1 Serum and CSF germ cell tumor markers

Tumor type	AFP	β-HCG
Yolk sac tumor	++	±
Embryonal cell carcinoma	±	±
Immature teratoma	±	±
Choriocarcinoma	–	++
Germinoma	–	–
Germinoma with syncytiotrophoblastic cells	–	+[a]
Mixed germ cell tumor	Variable	

Data from Refs. [14–16]

[a]Weakly positive, typically found in CSF analyses when present

Serum levels of relevant tumor markers should be drawn during the initial preoperative consultation. Sampling of CSF is more sensitive for germ cell tumor markers and should occur during procedures for CSF diversion or during careful lumbar puncture. Specifically, the presence of α-Fetoprotein (AFP) indicates the presence of fetal yolk sac elements and is associated with endodermal sinus tumors, embryonal cell carcinomas, or immature teratomas. Measurable β-human chorionic gonadotropin (β-HCG), produced by trophoblastic elements, indicates the presence of choriocarcinomas, embryonal cell carcinomas, or germinomas. In addition to their role in diagnosis, tumor markers are also used to establish a postoperative baseline to monitor treatment response or detect recurrence [10, 11, 17].

Of note, while measurable germ cell markers are confirmatory for a malignant germ cell tumor, their absence does not rule out the presence of a germinoma or embryonal cell carcinoma and should be interpreted cautiously. Furthermore, elevation of AFP in the presence of a germinoma suggests that an embryonal cell carcinoma or endodermal sinus tumor is present as part of a mixed tumor.

Biopsy Versus Resection

Given the diversity of histologic findings in pineal masses, the most reliable strategy to ensure accurate tissue diagnosis is with adequate tissue sampling. Strategies for tissue sampling include stereotactic and endoscopic biopsies or open surgical procedures. In general, biopsy may be preferred in patients with known primary systemic tumors, multiple lesions, or other comorbidities that increase surgical risk. In these situations, where open surgical resection is deemed too risky, the options for ensuring accurate tissue diagnosis include either stereotactic or endoscopic biopsy. Unfortunately, tissue sampling is limited in either technique and may result in inaccurate diagnosis due to sampling error. Furthermore, both techniques have been associated with risks of postoperative hemorrhage and obstructive hydrocephalus [18–21].

Alternatively, open surgical resection provides maximal tissue sampling concurrent with a reduction of mass effect due to tumor debulking. In addition, aggressive resection offers the potential to control hydrocephalus without a second procedure and reduces the risk of postoperative hemorrhage into the residual tumor bed [22].

Surgical Approaches

Operative approaches to the pineal region are broadly categorized as supratentorial or infratentorial (Fig. 20.2). In general, surgical approach is interchangeable and relies on careful anatomic evaluation and the experience and comfort of the surgeon with a particular approach. Supratentorial approaches provide greater exposure than infratentorial approaches with the caveat that they require the surgeon to work carefully around the convergence of the deep cerebral veins. Supratentorial approaches are preferred for accessing large tumors that extend supratentorially or laterally to the ventricular atrium.

Alternatively, the midline, infratentorial location of most pineal region tumors offers many advantages to infratentorial approaches. In particular, the deep venous system is dorsal to the tumor, permitting it and the velum interpositum to be more easily dissected off the tumor with an infratentorial approach.

Supratentorial approaches include occipital transtentorial (OTT), interhemispheric transcallosal, and the rarely used transcortical transventricular. The supracerebellar infratentorial (SCIT) approach is the most commonly used infratentorial approach.

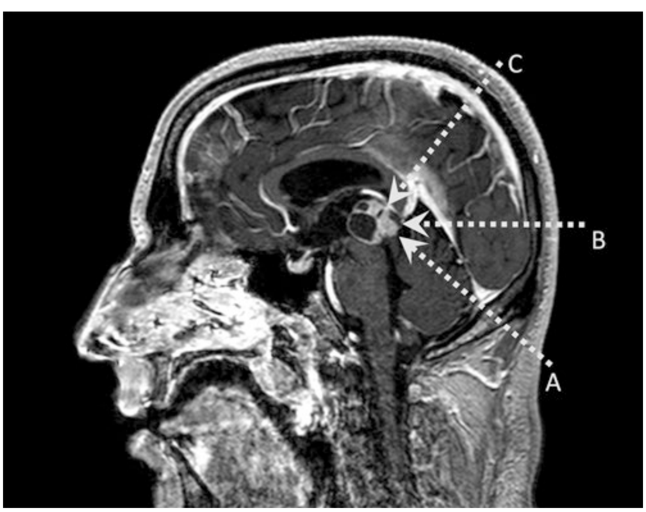

Fig. 20.2 The approximate approach of each of the three major open surgical approaches to the pineal region. Supratentorial approaches include the occipital transtentorial approach (**b**); the interhemispheric transcallosal approach (**c**). The most common infratentorial approach is the supracerebellar infratentorial approach (**a**). In general, surgical approach is interchangeable and relies on careful anatomic evaluation and the experience and comfort of the surgeon with a particular approach

Patient Position

Sitting Position

The sitting position is generally preferred for SCIT approaches. The patient is seated upright in a neutral position. Gravity works in favor of the surgeon to reduce pooling blood and CSF in the operative field, to allow the cerebellum to fall away naturally with minimal retraction, and to facilitate dissection off the deep venous system dorsal to the tumor. The head should be flexed so that the tentorium is approximately parallel to the floor. Care is taken to avoid kinking of the ET tube, obstruction of venous outflow, and cervical spine injury. The legs should be elevated and slightly flexed to facilitate venous return and to minimize sciatic stretch injury. Risks of air embolism, pneumocephalus, or subdural hematoma should be anticipated and proper precautions taken. Precordial Doppler and end-tidal CO_2 monitoring are used to detect air emboli and rarely, a central venous catheter can be used to remove entrapped air.

Lateral and Three-Quarter Prone Position

The lateral decubitus position is another commonly utilized position. The head is raised above the horizontal, in particular for the interhemispheric transcallosal approach. For the OTT approach, the head is positioned with the patient's nose rotated 30° toward the floor to improve visualization. The three-quarter prone position is an extension of the lateral position, with the head turned at an oblique 45° angle. Lateral approaches are thought to reduce surgeon fatigue as the hands are not extended as they are in the sitting position. Again, gravity is exploited to facilitate retraction of the dependent hemisphere.

Prone Position

Prone positioning allows two surgeons to work together through an operative microscope. In a variant known as the Concorde position, the head is rotated 15° away from the craniotomy side. In general, the prone position is simple and safe for supratentorial approaches but can be cumbersome in infratentorial approaches where the benefits of gravity are lost.

Operative Approaches

Supracerebellar Infratentorial

Performed most commonly in the sitting position, the SCIT approach follows a relaxed, natural midline corridor to the pineal region and is best suited for tumors arising below the deep cerebral veins, without extensive lateral or caudal expansion [10, 12, 22]. While the midline SCIT approach offers easy orientation and a wide operative corridor, the angle to the lateral and caudal side of the surgical field is narrow and limited in patients with a steep angle of the tentorium, often necessitating excessive neck flexion which may compromise airway safety or venous return. As a result, preoperative evaluation for a steep angle of the tentorium or a low-lying torcular Herophili is critical and may suggest an alternate approach.

The patient is positioned neutral with rigid fixation as described above. A skin incision is made from just above the torcula to approximately the level of the C4 spinous process. The suboccipital musculature is released and retracted laterally. Typically, craniotomy is preferred over craniectomy as replacement of the bone reduces the incidence of postoperative aseptic meningitis, fluid collections, and discomfort. A wide craniotomy is centered just below the torcula. Slots are drilled over the sagittal sinus above the torcula, over the transverse sinuses bilaterally, and 1–2 cm above the foramen magnum in the midline over the occipital sinus. The slots are connected with a craniotome and the bone flap elevated. Bone edges are waxed and all venous bleeding is controlled to avoid air emboli. Care is taken to expose enough bone above the transverse sinus to clearly view the tentorium and to ensure the craniotomy is extensive enough to provide room for instruments and adequate light from the microscope.

The dura is opened in a curvilinear fashion, reflected superiorly along the transverse sinus, and anchored with slight tension to avoid obstructing the sinuses. The inferior dura remains intact and acts to support the cerebellar hemispheres. A corridor is developed above the cerebellum and beneath the tentorium through cautery and division of arachnoid adhesions and thin midline bridging veins. Bridging veins are cauterized and divided midway to minimize bleeding from the sinuses. Great effort is made to minimize disruption of the extensive collateral circulation by sparing thick bridging veins and hemispheric bridging veins found laterally. Following division of the cerebellar attachments to the tentorium, gravity permits the cerebellum to fall away with minimal retraction. The cerebellum is protected, and deeper adhesions and bridging veins are divided when they become visible near the anterior vermis until the arachnoid covering the pineal region is seen. The precentral cerebellar vein courses from the anterior vermis to the vein of Galen and should be dissected, cauterized, and divided as far as possible from the confluence of the basal veins of Rosenthal to avoid progression of thrombosis and potential venous infarct. Sacrifice of additional veins of the deep venous system is not advisable.

The retractor (if still necessary after CSF removal) and the microscope are adjusted to visualize the inferior portion of the tumor (Fig. 20.3a–c). The arachnoid covering the pineal gland is sharply opened, and the posterior surface of the tumor is exposed. The central portion of the tumor is cauterized and opened, and specimen can be taken and sent for frozen intraoperative pathological consultation. The tumor is subsequently debulked using a variety of instruments including suction, cautery, tumor forceps, or an ultrasonic aspiration device if necessary.

With progressive internal decompression, separation of the tumor from the surrounding thalamus and midbrain becomes possible. Tumor dissection continues until the third

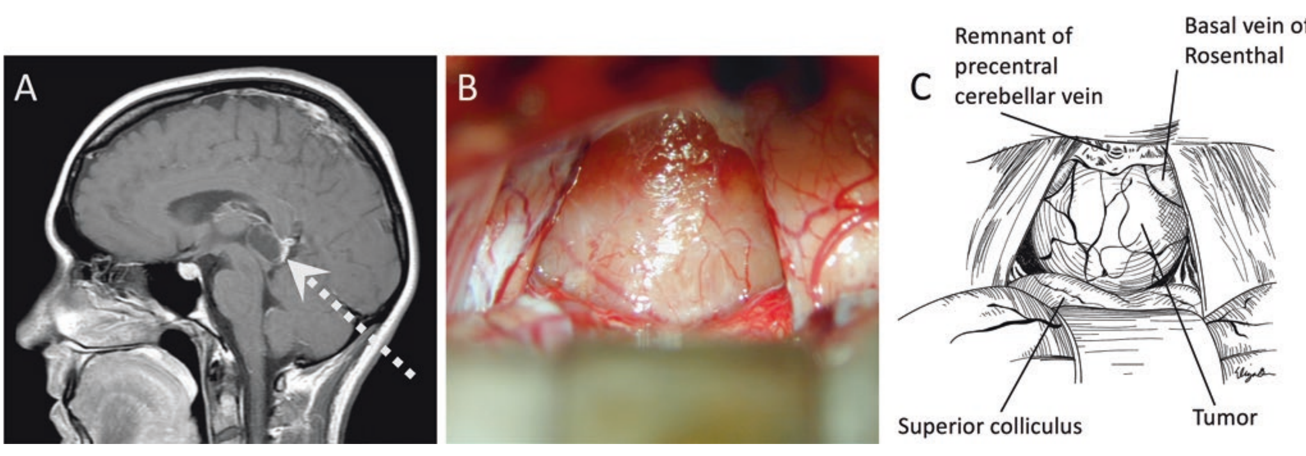

Fig. 20.3 (**a**–**c**) Supracerebellar infratentorial approach. The supracerebellar infratentorial (SCIT) approach follows a relaxed, natural midline corridor to the pineal region and is best suited for tumors arising below the deep cerebral veins (**a**). Surgery is typically performed in the sitting position with the head midline. A wide craniotomy is centered below the torcula, and a curvilinear dural opening permits development of an operative corridor between the cerebellum and the tentorium through separation of arachnoid adhesions and thin midline bridging veins until the arachnoid covering the pineal region is seen. The precentral cerebellar vein is dissected, cauterized, and divided as far as possible from the confluence of the basal veins of Rosenthal, and the tumor is debulked and separated from the surrounding brain (**b**, **c**). (**c**: Used with permission of Eliza Bruce.)

Fig. 20.4 (**a**–**h**) Lateral supracerebellar infratentorial approach. A 24-year-old male with history of an ETV for obstructive hydrocephalus due to non-enhancing pineal lesion was taken for resection of the lesion using the lateral supracerebellar infratentorial approach (**a**). The patient is placed in the sitting position, and the head is placed in rigid fixation, turned slightly and flexed slightly to optimize the operative corridor (**b**). Incision is marked halfway between the inion and the transverse-sigmoid junction, extending above and below the transverse sinus (**c**). The suboccipital musculature is separated and retracted laterally, and a small paramedian craniotomy exposes the transverse sinus (*dotted line*) while leaving the torcula protected (**d**). The dura is opened as a single curved flap and retracted toward the sinus exposing the operative corridor (**e**). The operative corridor is developed with great effort to avoid sacrificing large median veins as well as lateral bridging veins, and the arachnoid covering the pineal gland is sharply opened demonstrating the posterior surface of the tumor (*) (**f**). The tumor is internally debulked initially and subsequently separated from the surrounding brain (**g**). Postoperative MRI within 48 h demonstrates gross total resection (**h**)

ventricle is encountered. Choroidal arterial vessels along the lateral wall need not be preserved. The tumor is lifted superiorly, and blunt dissection at the junction with the brainstem is performed. The superior attachment to the deep venous system is carefully released last, and the lesion is removed. Flexible mirrors are useful to examine the caudal portion of the tumor bed in order to verify the extent of resection and to remove any residual tumor or blood clot obstructing the cerebral aqueduct that may cause hydrocephalus. Careful cautery is preferred for hemostasis at this point as hemostatic agents may float into the ventricle and cause CSF obstruction. The dura is closed in a watertight manner and the bone flap is plated into place.

Risks associated with the SCIT approach mainly reflect the sitting position and include air embolism, pneumocephalus, and subdural hematomas.

Lateral Supracerebellar Infratentorial Approach

The lateral SCIT approach utilizes a slightly longer trajectory over the quadrangular lobule with a less steep, more direct angle to the pineal region (Fig. 20.4a–h). The use of a paramedian corridor to the pineal region avoids the apex of the culmen and the numerous bridging veins of the median supracerebellar region [23, 24].

The lateral SCIT is most frequently performed in the sitting or lateral decubitus position. The head is placed in rigid fixation, turned slightly, and flexed to ensure optimal utilization of the surgical corridor (Fig. 20.4a, b). A spinal drain or occipital EVD is placed to assist with brain relaxation. A 7–8 cm linear incision is made halfway between the inion and the transverse-sigmoid junction extending above and below the transverse sinus (Fig. 20.4c). The suboccipital musculature is separated and retracted, and a small, paramedian craniotomy is performed exposing the transverse sinus while leaving the torcula protected (Fig. 20.4d). The dura is opened as a single curved flap and retracted toward the sinus where tentorial retraction sutures are used to elevate the transverse sinus and expand the operative corridor (Fig. 20.4e). CSF release through lumbar drainage facilitates retraction of the cerebellar hemisphere caudally. A corridor is developed above the cerebellum and beneath the tentorium as in the standard SCIT approach. Again, great effort is made to leave large median and lateral bridging veins intact. Arachnoid dissection and tumor resection are carried out as described above with care to preserve the arachnoid membranes of lateral structures such as the SCA or the trochlear nerve (Fig. 20.4f–h).

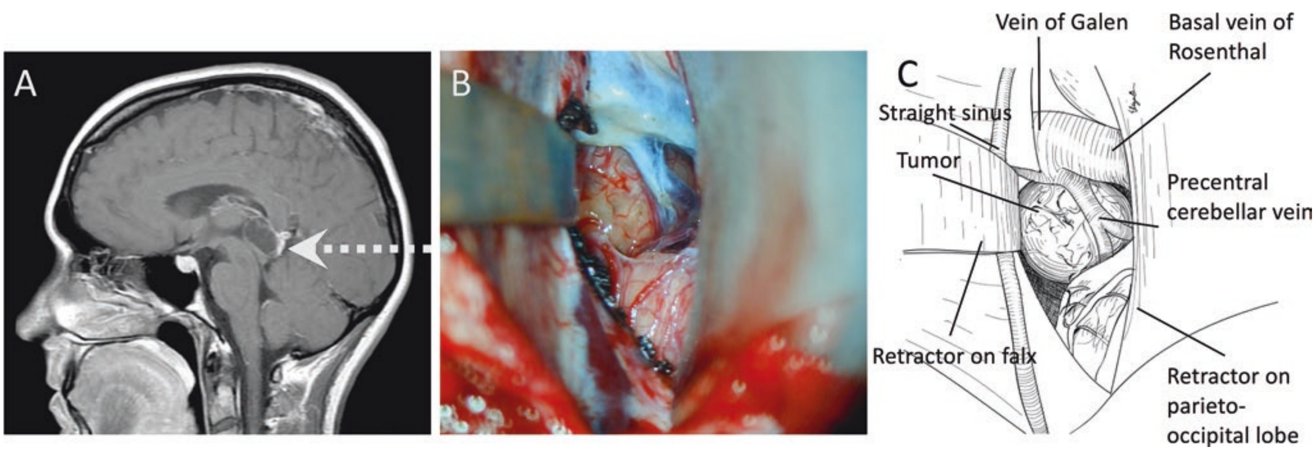

Fig. 20.5 (**a**–**c**) Occipital transtentorial approach. A lateral, three-quarter prone position is preferred for the occipital transtentorial approach (OTT). A paramedian linear incision and generous craniotomy extending to the opposite of midline result in a natural corridor between the falx medially, the tentorium inferiorly, and the medial occipital lobe laterally (**a**). The dura is opened and reflected toward the sinus. Microscopy and Doppler ultrasound are used to map the location and course of the straight sinus, and the tentorium is divided just lateral to the sinus. Splitting the tentorium offers panoramic supra- and infratentorial views of the tumor, the surrounding venous structures, and the collicular plate (**b**, **c**). The arachnoid overlying the tumor and quadrigeminal cisterns are opened, and tumor removal proceeds with great care to avoid injury to the deep venous system. (**c**: Used with permission of Eliza Bruce.)

Occipital Transtentorial (Fig. 20.5a–c)

The OTT approach is the most commonly used supratentorial approach. Utilizing a natural corridor between the falx medially, the tentorium inferiorly, and the medial occipital lobe laterally, the OTT provides a wide operative field with an unobscured view of the tentorial notch, the deep venous system, and the pineal region from above [25]. As the majority of pineal lesions arise inferior to the deep veins, the OTT approach requires the surgeon work past the deep venous structures, damage to which may prove neurologically devastating.

Typically, a lateral, three-quarter prone position is preferred for the OTT approach as it permits gentle, gravity-assisted retraction of the occipital lobe. Mannitol or spinal drainage is used for additional brain relaxation when necessary. A linear, slightly paramedian scalp incision permits placement of a burr hole or slot over the sagittal sinus just above the torcula with a second burr hole 6–10 cm above this. A generous craniotomy is performed extending 1–2 cm to the opposite of the midline with care to avoid injuring the sinus. The dura is opened and reflected toward the sinus. A lack of bridging veins near the occipital pole allows wide retraction of the occipital lobe with gravity alone, minimizing the use of fixed retractors. Microscopy and Doppler ultrasound are used to map the location and course of the straight sinus, and the tentorium is divided just lateral to the sinus. A retractor can be placed to facilitate retraction with care to avoid injuring the infratentorial bridging veins connecting the cerebellum to the tentorium. Splitting the tentorium offers panoramic supra- and infratentorial views of the tumor, the surrounding venous structures, and the collicular plate [26]. If necessary, the falx can be divided to further facilitate retraction.

The arachnoid overlying the tumor and quadrigeminal cisterns are opened, and tumor removal proceeds as described above with great care to avoid injury to the deep venous system. While division of the tentorium provides excellent exposure of the quadrigeminal plate, the OTT provides a limited view of the contralateral quadrigeminal cistern.

The OTT approach may be associated with transient postoperative hemianopsia due to excessive retraction on the occipital lobe, which becomes permanent in 1–3%. Permanent morbidity is reduced by avoiding rigid brain retraction if possible. Potential damage to the deep cerebral veins is also possible as instruments are manipulated between them during tumor removal.

Interhemispheric Transcallosal Approach

The interhemispheric transcallosal approach is an alternative supratentorial approach that utilizes an operative corridor along the parieto-occipital junction, between the falx and the medial cerebral hemisphere, and through the posterior corpus callosum. Patients generally positioned in the prone or sitting position. Mannitol and spinal drainage may be used for additional brain relaxation if needed. Craniotomy location varies depending on the location of the tumor within the third ventricle, and the use of navigation is helpful. Burr holes or slots are made over the sagittal sinus anteriorly and posteriorly, and a generous craniotomy extending 8 cm in length and 1–2 cm to the side opposite of your approach to the tumor is performed. A wide craniotomy is recommended as it provides flexibility in avoiding bridging veins when choosing an operative corridor and is generally centered over the vertex to minimize manipulation of the occipital lobe. The dura is opened in a U-shaped fashion and reflected medially toward the sagittal sinus. An operative corridor is chosen which minimizes the number of bridging veins for sacrifice. If further exposure is required, the inferior falx and tentorium may also be divided.

Using gentle retraction of the parietal lobe, the corpus callosum is identified, and the pericallosal arteries running over the corpus callosum are retracted to one side or to separate sides. A 2 cm opening is made in the corpus callosum over the bulk of the tumor using suction and cautery. If possible, opening through the splenium is avoided to minimize the risk of a disconnection syndrome. Once through the corpus callosum, early identification of the veins of the deep venous system is critical, and the dorsal surface of the tumor is identified. The tumor is then debulked and dissected using the same techniques described above. Leaving a ventricular drain is optional.

Transcortical Transventricular Approach

This approach is rarely used given its limited exposure and the need for cortical incision. When indicated, a standard craniotomy is planned over the desired approach to the lesion using navigation. A cortical entry point is typically chosen so as to pass through non-eloquent cortex. Stereotactic guidance is particularly useful in the setting of a tumor that extends into the lateral ventricle.

Postoperative Care

Postoperatively, all patients are maintained on high-dose corticosteroids, which are tapered off as their condition improves. A short course of antiepileptics is generally recommended for supratentorial approaches. Postoperative lethargy and cognitive impairment are common following surgical resection and manipulation of the third ventricle and in patients with extensive subdural air after surgery in the sitting position. Careful and frequent neurological examination in an intensive care setting is appropriate until clinical improvement is noted. Any evidence of neurologic worsening should always be investigated with appropriate imaging to rule out hydrocephalus, hemorrhage, or pneumocephalus. Patients with a history of VPS insertion and neurological worsening should be evaluated for possible shunt malfunction caused by air, blood, or operative debris.

Early mobilization and ambulation with physical therapy and rehabilitation improve outcomes. Postoperative MRI should be performed within 48 h to determine the extent of resection. In addition, spinal MRI to evaluate for CSF dissemination of disease should be performed if it was not done. Drains placed at surgery should be removed or converted to permanent CSF diversion within 72 h to minimize infection risks. Tumor markers should be serially measured postoperatively to serve as a baseline for detecting recurrence or monitoring response to treatment.

Surgical Outcomes

Outcomes following microsurgical resection have improved dramatically as a result of modern microsurgical techniques, advances in neuroanesthesia, and improved postoperative care. Large, modern series (Table 20.2) evaluating surgical outcomes since the 1990s report mortality rates between 0% and 2% and permanent minor morbidity persisting in 3–20% of patients [25, 27, 28].

The most serious postoperative complication is postoperative hemorrhage, with a higher incidence in the setting of

Table 20.2 Outcomes of large, modern surgical series since the 1990s

Study	Year	No. cases	Approach	Age	GTR (%)	Mortality (%)	Major morbidity (%)	Permanent morbidity (%)
Bruce and Stein [22]	1995	160	SCIT IHTC OTT	All	45	4	3	19
Konovalov and Pitskhelauri [25]	2003	255	SCIT OTT Other[b]	All	58	7.8[a]	15	15
Hernesniemi et al. [27]	2008	119	SCIT OTT	All	88	0	<1	4.2
Bruce [16]	2011	128	SCIT IHTC OTT	All	49	2	1	NA
Qi et al.[c] [28]	2014	143	OTT	All	91.6	0.7	3.5	5.6

SCIT supracerebellar infratentorial, *IHTC* interhemispheric transcallosal, *OTT* occipital transtentorial, *NA* not available, *GTR* gross total resection

[a]Mortality rate of 1.8% in resections after 1990

[b]Other approaches included: subchoroidal, fourth ventricular approaches and combined approaches in 30 patients

[c]Only included nongerminomatous pineal region tumors

malignant, vascular tumors that have been subtotally resected. Additional common postoperative sequelae include dysfunction of extraocular mobility, pupillary abnormalities, and ataxia related to brainstem and cerebellar manipulation. These minor complications are typically transient, resolving over days to months. In addition, the presence of intracranial air often results in transient cognitive impairment as well. Venous infarcts are not uncommon on postoperative MRI. Rarely, these venous infarcts extend into the midbrain and create devastating consequences. Venous infarction is typically unpredictable and likely results from venous insufficiency in a small subset of patients who cannot tolerate the disruption of collateral circulation within the cerebellum due to sacrifice of bridging veins. Permanent, mildly limited upgaze is frequently observed and carries little clinical significance. In general, complications are more common in previously irradiated patients, patients with invasive tumors, and patients who were progressively symptomatic preoperatively.

Long-term clinical outcomes vary according to specific tumor pathologies, and the analysis of the efficacy of tumor debulking is most instructive in the context of specific pineal histologies. Generally, benign tumors are amenable to long-term, progression-free survival and likely cure with complete resection. Outcomes are more variable for malignant tumors, with several reports correlating extent of tumor resection with improved response to adjuvant therapy and increased survival [11, 22, 29]. Clear trends demonstrate the benefits of aggressive surgery for the various pathologies arising within the pineal region.

Benign Pineal Region Tumors

Benign lesions, including meningiomas, epidermoids, teratomas, pineal cysts, and pilocytic astrocytomas, are all associated with excellent prognosis [8, 10, 22]. Gross total resection invariably results in long-term remission and potential cure. In addition, operative complications are rare in this group of patients.

Germinomas

The most common tumor in this region, germinomas, is exquisitely radiosensitive with 80–90% long-term survival with radiation to the tumor and the surrounding ventricles [30, 31]. Typically, stereotactic radiosurgery and chemotherapy are attempted as a first-line therapy, reserving whole brain radiation and its associated risks of cognitive and endocrinological morbidity for non-responders. Successes with adjuvant therapies have decreased the emphasis on debulking in germinoma, and the importance of extent of resection in these patients remains debatable [32–34]. Regardless, tissue-proven diagnosis is recommended.

Nongerminomatous Germ Cell Tumors

Nongerminomatous malignant germ cell tumors, including endodermal sinus tumors, choriocarcinomas, and embryonal carcinomas, carry a poor prognosis. Biopsy is often unnecessary as the majority of these tumors are diagnosed on the basis of elevated tumor markers. These tumors respond best when radiation and chemotherapy are combined, and most analyses report only anecdotal benefits of initial complete resection that do not reach statistical significance [30, 33].

Interestingly, second-look surgery for the presence of residual mass following radiation and chemotherapy increases 5-year survival rates and offers diagnosis of residual malignant elements requiring further chemotherapy [35].

Pineal Parenchymal Tumors

Pineal parenchymal tumors exist along a continuum from the benign pineocytoma to the aggressively malignant pineoblastoma.

Pineocytomas are optimally treated by radical surgery alone, and their prognosis parallels that of benign pineal tumors with long-term remission after resection with or without radiation for residual [22]. Alternatively, pineoblastomas are aggressive and prone to metastasis. Pineoblastomas are treated with radical surgery along with adjuvant radiation and/or chemotherapy. In the pediatric population, treatment strategies for pineoblastomas mirror those of the more common posterior fossa primitive neuroectodermal tumors (PNETs) whose prognosis is tied to the extent of surgical resection [36]. Modern practice advocates gross total resection and focuses on chemotherapy regimens to reduce radiation in children older than 4 years as radical surgery is associated with trends toward better outcomes [37, 38]. In adults, long-term remission is not unusual, and aggressive resection also seems beneficial [39].

There is little agreement on the optimal management strategy to address PPTIDs as they demonstrate unpredictable behavior. However, median survival is significantly longer following radical resection of PPTIDs as compared with pineoblastomas and supports complete resection. In studies of papillary pineal tumors, only gross total resection and younger age were associated with a longer overall survival with radiotherapy and chemotherapy having no significant impact [40].

Pineal Astrocytomas

Astrocytomas arising in the pineal gland are often cystic. Although few reports specifically assess the management of pineal region astrocytomas, complete resection is achievable and likely results in cure [41].

PEARLS
Management of symptomatic hydrocephalus should precede surgical intervention.
Serum/CSF germ cell tumor markers should be evaluated in all patients as their presence may preclude surgical intervention.
Accurate tissue diagnosis is ensured with adequate tissue sampling.
Supratentorial approaches are preferred for accessing large tumors that extend supratentorially, laterally to the ventricular atrium, or have significant caudal extension.
Infratentorial approaches are preferred for midline, infratentorial lesions arising ventral to the deep venous system.

Commentary on Case Presentation in Section 20.1

Case Presentation

A 19-year-old presents with an incidental lesion discovered after a motor vehicle accident and concussion. He is currently asymptomatic. The lesion is initially followed but demonstrated substantial growth on a 6-week follow-up scan, concerning for high-grade neoplasm. Serum markers are negative. There is no associated hydrocephalus (see Fig. 20.1a–d).

Discussion

Despite its incidental nature, this patient has a newly diagnosed and progressively enlarging pineal mass. In the absence of symptomatic and radiographic hydrocephalus, as well as serum germ cell tumor markers, this patient warrants evaluation for surgical intervention via resection, or biopsy, with the primary objective to ensure an accurate diagnosis. As discussed above, adequate tissue sampling is paramount to ensuring accurate diagnosis in pineal lesions, and this is best achieved with open surgical resection in a healthy 19-year-old with no significant medical comorbidities precluding surgery. A treatment algorithm for managing pineal region lesions is shown in Fig. 20.6.

In the absence of hydrocephalus, preoperative ETV is not required, and the risk of hydrocephalus can be addressed intra- or postoperatively. Spinal drainage or an intraoperative EVD, in addition to mannitol administration, may be helpful to assist with brain relaxation.

Given the midline, infratentorial location of the lesion, resection via the SCIT approach in the sitting position would provide adequate visualization and minimize the risk of damage to the deep cerebral veins and potential for postoperative hemianopsia associated with supratentorial approaches. However, given the steep angle of the tentorium and relatively low-lying torcula, the patient would require significant neck flexion to optimize the working angle. In this particular situation, a lateral SCIT approach may offer a less steep working angle and excellent access to this small midline lesion while potentially minimizing the number of midline cerebellar veins sacrificed. Depending on frozen pathology, an attempt at gross total resection or internal debulking is attempted with the goal of minimizing postoperative neurologic deficits.

Postoperatively, the patient would be managed in an intensive care setting with frequent neuro exams. Craniospinal MRI would be performed within 48 h to establish extent of resection and look for evidence of CSF seeding. Any temporary CSF diversion would be considered for permanent placement. Adjuvant chemotherapy and radiation would be considered for malignant lesions and for recurrence or progression of subtotally resected benign lesions.

20.3 Pineal Region Tumors: Keyhole Craniotomy

Charles Teo and Reid Hoshide

Introduction

The pineal region presents unique challenges to the cranial neurosurgeon. The area is deep within the brain, surrounded by vital venous structures and in juxtaposition to eloquent parts of the brain, including the tegmentum and tectum of the midbrain, fornices, thalamus, and occipital lobes [42]. Other authors have covered the general philosophy concerning approaches to this region, and therefore, this chapter will concentrate on the keyhole philosophy as it relates to pineal tumors.

The keyhole approach to any tumor is not about size as much as it about limiting collateral damage, improving outcomes, and reducing complications. The surgeon should consider the *entrance*, the *corridor*, and the *rooms*. When removing tumors that are deep to the surface, the *entrance* does not need to be large. Of course, it needs to be big enough to allow good visualization of the entire tumor, but the deeper the region, the smaller the opening required. The *corridor* needs to offer the most uncluttered trajectory to the most superficial part of the tumor *and* the deepest part of the tumor, i.e., it should be along the long axis of the tumor (the "2-point rule") (Fig. 20.7a–c). Finally, the concept of the *rooms* is simple. If all tumors were perfectly cylindrical or spherical, in other words totally within the *corridor*, the operating microscope would give you adequate visualization

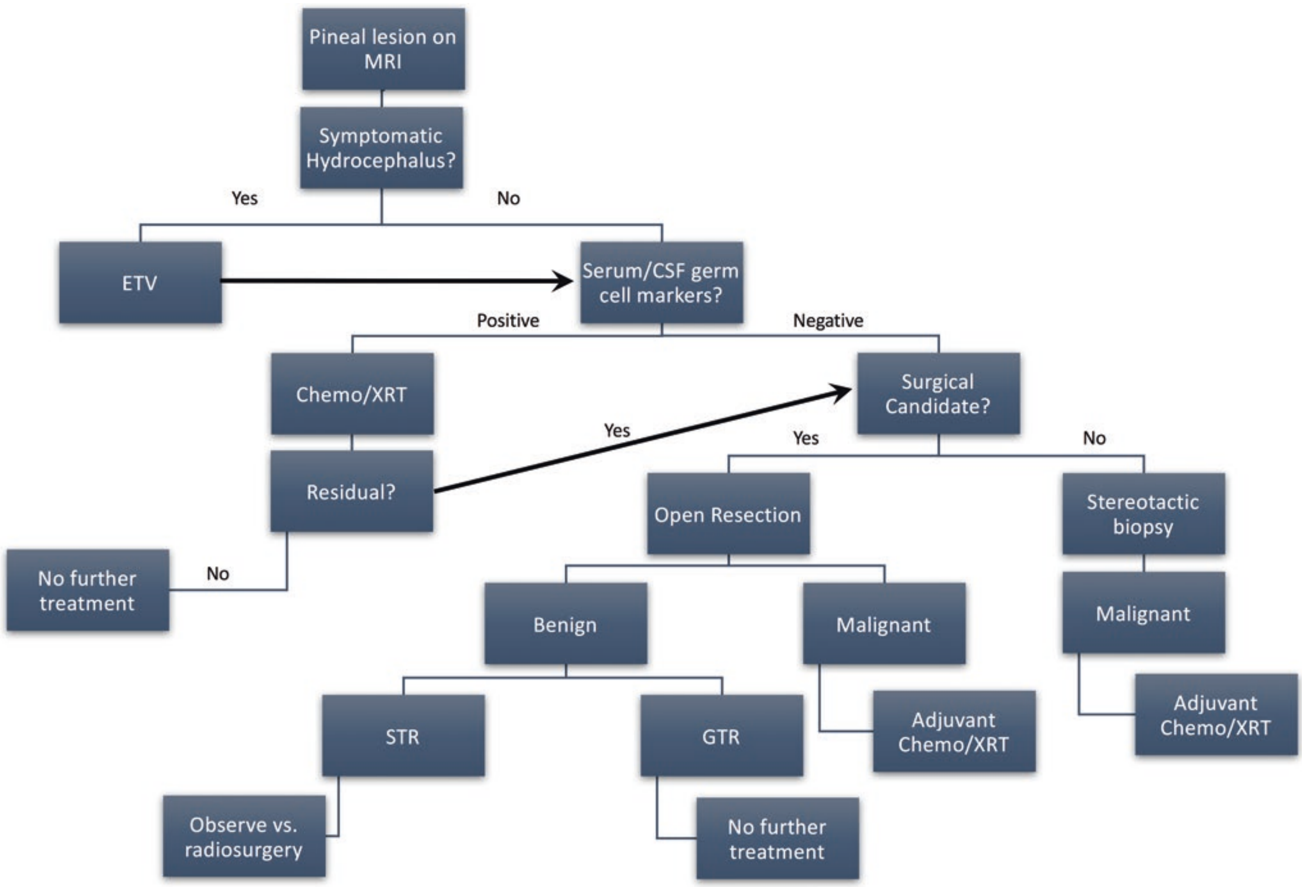

Fig. 20.6 Treatment algorithm for pineal region lesions (Abbreviations: *Chemo/XRT* chemotherapy and radiotherapy, *ETV* endoscopic third ventriculostomy, *MRI* magnetic resonance imaging, *CSF* cerebrospinal fluid, *STR* subtotal resection, *GTR* gross total resection)

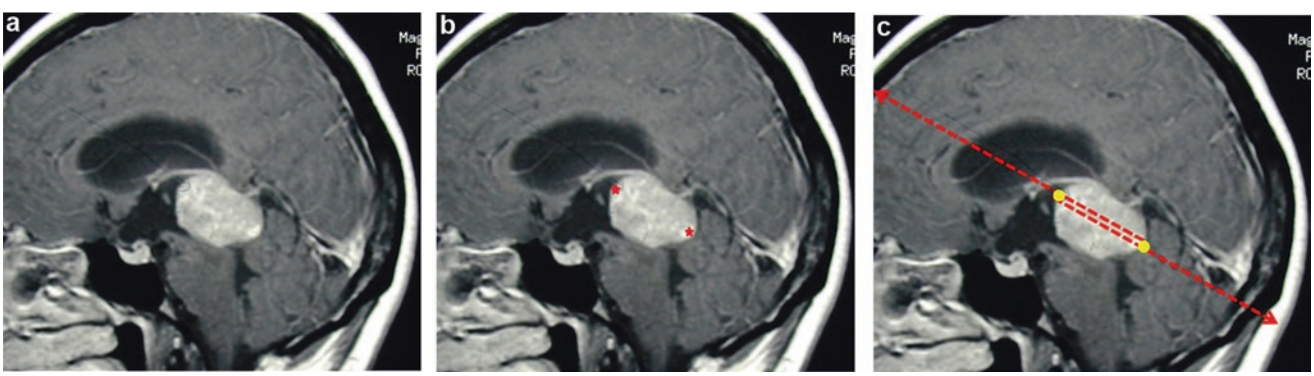

Fig. 20.7 (**a**) This pineal tumor was most extensive in the anteroposterior plane. (**b**) The 2-point rule simply states that the deepest and most superficial points of the tumor need to be identified. (**c**) A line drawn between the two points and carried to the surface will indicate the ideal surgical trajectory. This pineocytoma was completely excised using an infratentorial keyhole approach but could equally have been removed by an anterior, interhemispheric, transcallosal approach

of the entire tumor (Fig. 20.8a–f). However, most tumors are not confined to the corridor and occupy different *rooms* that require you to either knock down walls or look through doors or around corners. Those walls are either functional brain tissue or important neurovascular structures that cannot be "knocked down." This is when the endoscope becomes an invaluable tool (Fig. 20.9a–f).

With these three concepts in mind, the correct approach is that which considers the long axis of the tumor, the "obstacles" on the way to the tumor, and the structures that need to be

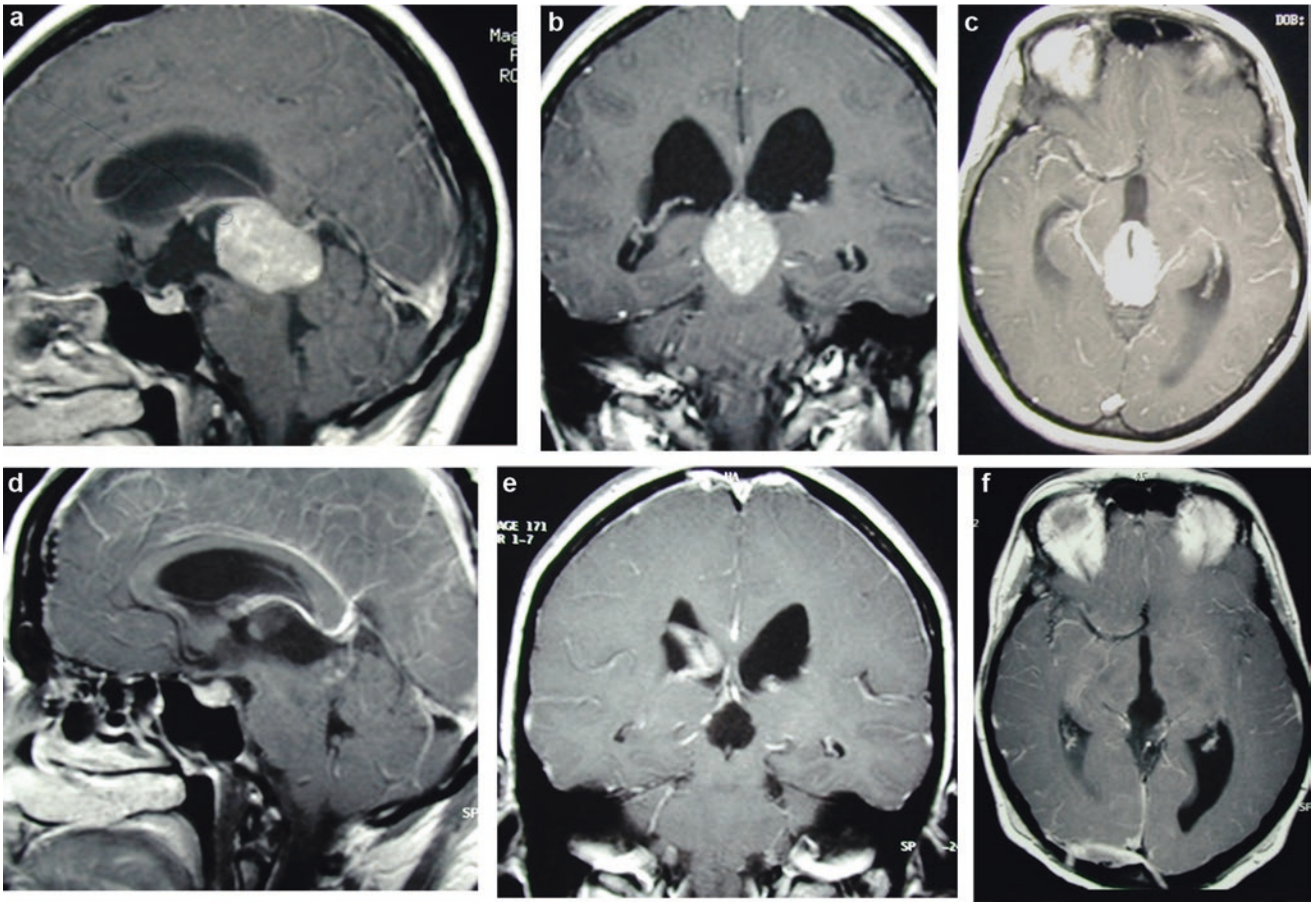

Fig. 20.8 (**a**–**c**) The sagittal and coronal views show clearly that this tumor is almost like a cylinder and therefore can be removed without the need for angled visualization. (**d**–**f**) Although the endoscope was used to inspect the tumor bed, the view from the microscope was sufficient for a complete macroscopic resection

identified and preserved when "looking around corners." Of course, in order to be effective, the angled view is worthless without angled instruments. The essential tools for endoscopic-assisted surgery are angled bipolar forceps (with varying lengths of the angled distal end), angled scissors and grabbing forceps, and angled suckers. Techniques vary with personal preference. We recommend holding the endoscope in your non-dominant hand and instruments in the other. To avoid being a "one-handed" surgeon, many of the medical device companies have developed hybrid instruments that allow the surgeon to both suction and coagulate, curette, irrigate, and "liquefy" tumor with a single instrument. Indeed, available to neurosurgeons now is a disposable and malleable instrument with four functions in one. We are not comfortable using currently available scope-holding devices. First, they still have micro-movement. Second, they can be bumped during the case, and this may result in damage to important normal structures. Third, until we have 3D high-definition cameras, the most effective way to achieve a sense of depth with 2D vision is to move the endoscope in and out. This cannot be done when it is fixed.

Infratentorial/Supracerebellar Keyhole Approach

This inferior approach is more applicable to tumors in which the long axis is more anteroposterior and especially if the anterior part extends into the posterior third ventricle.

The patient is positioned either prone, sitting, or lateral decubitus. All have their advantages and disadvantages, but our preference is the lateral decubitus position if their BMI is >25 (Fig. 20.10a, b) and prone if less (Fig. 20.11a, b). The sitting position requires anesthetic expertise and lengthy preparation compared to the other approaches and increases surgeon's fatigue. The risk of air embolism is greater, and hypoperfusion of the cervical cord has been well documented [43]. The prone position is comfortable for the surgeon, but venous congestion and subsequent brain swelling are inevitable with even slightly obese patients. The lateral decubitus position reduces some of the venous obstruction compared to the prone position yet still provides an anatomical approach when the head is flexed and slightly rotated to face the floor. The approach is through the side that is uppermost, and the

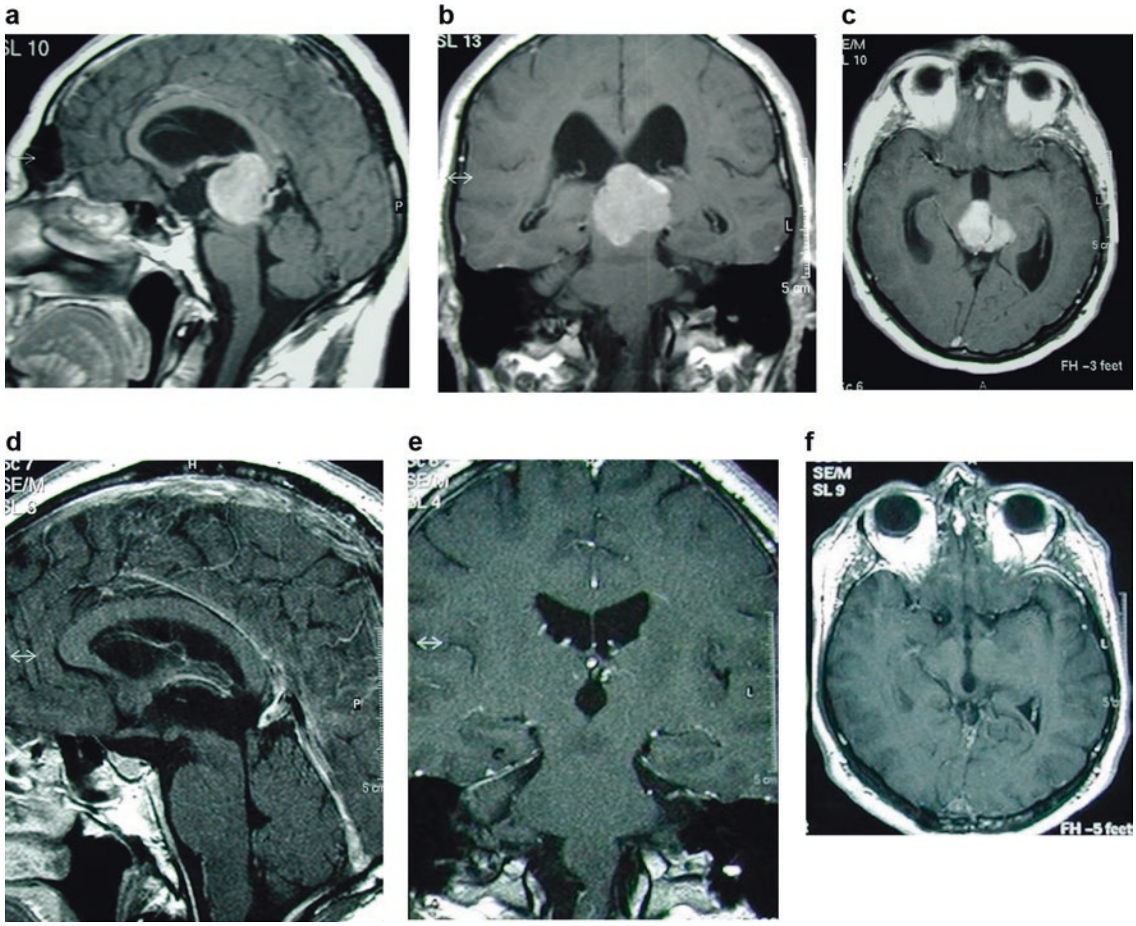

Fig. 20.9 (**a**–**c**) Unlike the tumor in Fig. 20.8a–f, this tumor has a left lateral projection and a superior projection. This illustrates the concept of a tumor having several "rooms" off the main "corridor." (**d**–**f**) This tumor was completely removed using an occipital supratentorial approach. It is also a good example of a steep sloping tent, making the infratentorial approach less attractive. The endoscope was invaluable in this case

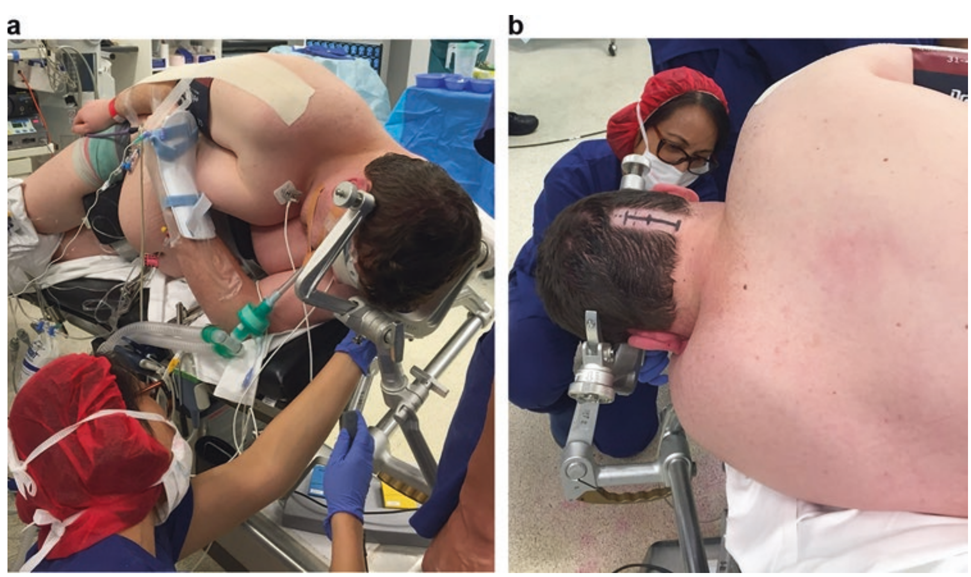

Fig. 20.10 (**a**, **b**) The BMI of this patient was >25. The prone position would have created severe venous congestion and resulted in challenging conditions

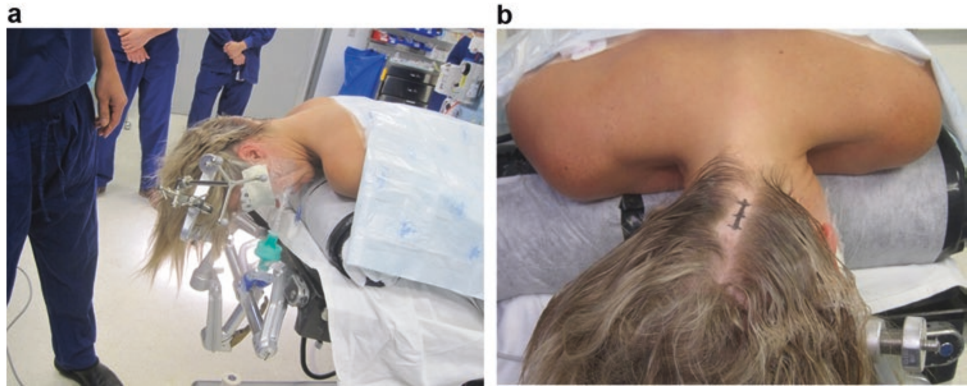

Fig. 20.11 (**a**, **b**) The BMI of this patient was <25, and hence the prone position was used without any compromise to her venous return

preferred side depends on the position of the transverse sinus and the morphology of the tent: the higher the sinus, the better the approach. The sagittal MRI views should be examined in order to ascertain whether the tent is "drooping" downward, thereby creating a slightly more limited corridor. Also, an extremely steep slope to the tent is also a relative contraindication to this approach (Fig. 20.9a, d).

The patient is then placed in a reverse Trendelenburg position to encourage venous return, and the incision should be made approximately 3 cm from the midline and just below the nuchal line. The incision is paramedian (*not* midline) as the approach is slightly off the midline to avoid the high-riding vermis. In other words, the corridor is along the sloping superior surface of the cerebellar hemisphere and not the superior vermis. The inferior edge of the transverse sinus should be seen ("blue-lined") before opening the dura, and the craniotomy should be no more than 2 cm in diameter. A triangular dural flap is made and reflected superiorly against the transverse sinus. Any bridging vein encountered along this path may be sacrificed with impunity (Fig. 20.12a–e). Midline veins, on the other hand, are better preserved, although the more posteriorly placed ones, and even the precentral vein, may be sacrificed without a high risk of venous congestion/infarction. Lysis of all the thick and opaque arachnoid membranes will aid in the early identification and subsequent preservation of the deep complex of veins.

Once the tumor is removed using standard microsurgical technique, the endoscope is brought into the surgical bed. This will serve two purposes. Firstly, it confirms total tumor removal, if the surgeon intends to do, and believes he has done, a complete resection. Secondly, an angled endoscope helps to visualize areas that are hidden from the microscope. Typically, this approach gives excellent views into the posterior third ventricle and the undersurface of the vein of Galen complex. However, if the tumor extends into the fourth ventricle, the angled visualization afforded by the endoscope is invaluable.

The closure is also crucial. The dural closure should be as watertight as possible. The bone is replaced and secured if a craniotomy was performed, and if a craniectomy was performed, then the defect can be filled with either artificial bone or bone dust. Conversely, the layer of the overlying muscle is so thick that if the defect is not filled, there will be no significant cosmetic effect (Fig. 20.13a–d).

Occipital/Supratentorial/Interhemispheric Keyhole Approach

This superior approach is more applicable for tumors in which the long axis is supero-inferior and especially if the caudal end of the tumor extends into the fourth ventricle.

The patient is placed prone or lateral decubitus with the ipsilateral side slightly down, to allow the ipsilateral occipital lobe to fall away from the falx. The head is either slightly flexed or slightly extended, but in most cases the neutral position is good (Fig. 20.14). To determine the final position of the head, another rule of keyhole surgery should be obeyed. It states that self-retaining retractors should be avoided whenever possible. To avoid the walls of the *corridor* from collapsing into the tumor bed and, hence, obstructing the surgeon's view, the long axis should be perpendicular to the floor. This is the rationale for taking time to position the patient correctly. Frameless stereotactic guidance is extremely helpful in this circumstance. Identify the deepest, most difficult part of the tumor. Draw a line from this point to the entrance. Ensure that this line is perpendicular to the floor in all planes by positioning the head before locking it into its final position. Reverse Trendelenburg position is helpful for venous drainage.

The incision is made in the midline and is invariably centered approximately 6 cm from the inion, but ultimately it is determined by the long axis of the tumor. The rationale for a midline incision is that the craniotomy sometimes needs to be bilateral. Choosing the operative side is based on the surgeon's comfort (right-handed surgeons prefer a right-sided approach), the pattern of bridging veins, and the long axis of the tumor. Clearly, if the tumor extends to the right side,

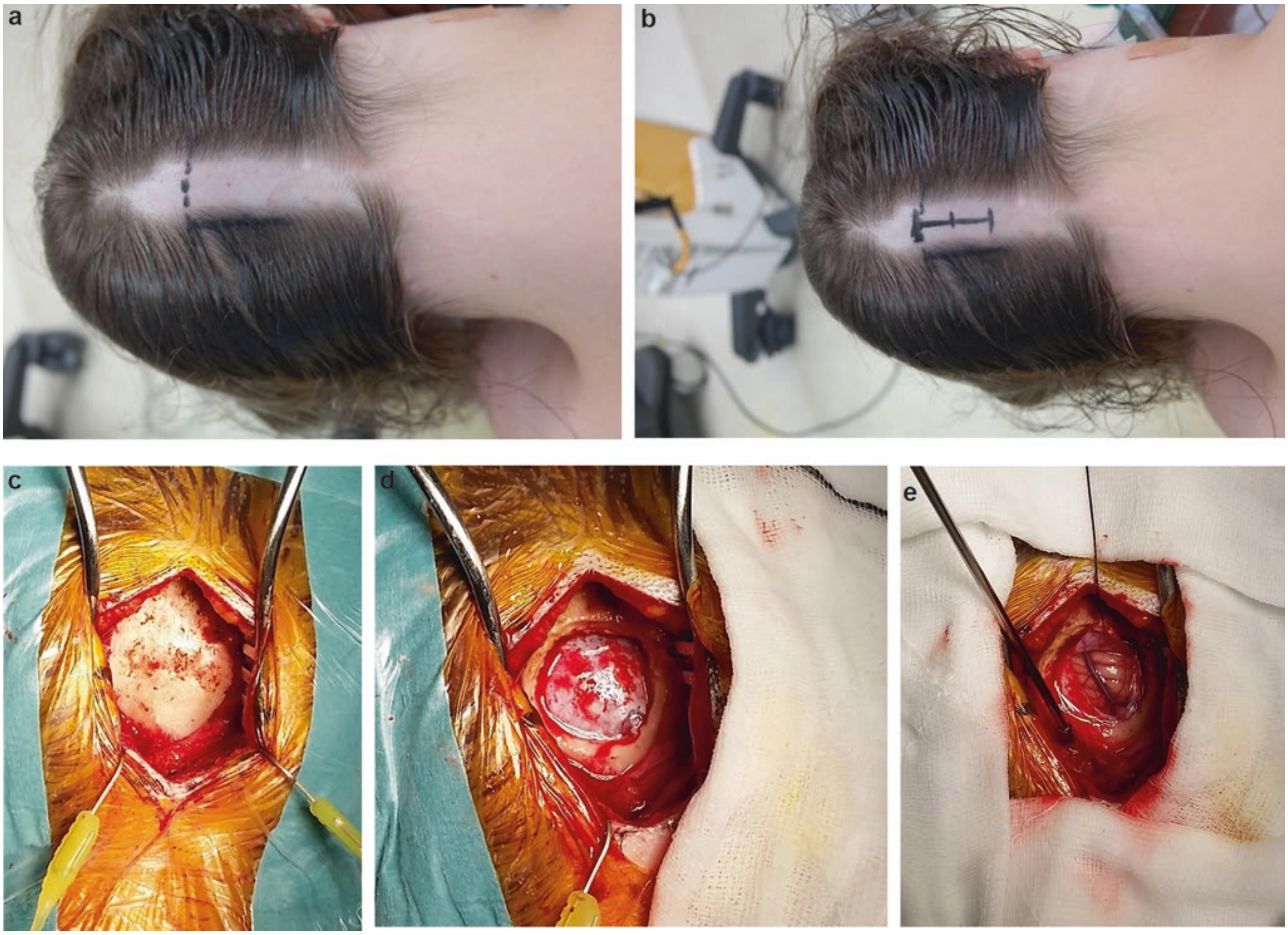

Fig. 20.12 (**a**, **b**) When the patient is placed in the lateral decubitus position, the craniotomy is performed on the upper side, and the incision is approx. 2 cm from the midline. (**c**–**e**) One can gain access either by a craniotomy or a craniectomy. The crucial step is to be sure the opening is directly below the transverse sinus

a left-sided craniotomy is preferable and vice versa. Sometimes, a bridging vein, invisible on preoperative imaging, is encountered on dural opening. If the vein is in the middle of the craniotomy and of a significant size, then the cranial opening needs to be long enough to operate either anterior or posterior to the vein (Fig. 20.15a–c). The occipital lobes are often full and bulging once the dura is opened despite good positioning and a nonobese patient. This is common and does not present an insurmountable problem. The first maneuver is to open some of the sulcal subarachnoid spaces immediately after the dural opening. Although the CSF that escapes from these spaces appears minimal, occasionally it is enough to relax the brain without any other maneuver. The next step is to ask the anesthesiologist what the end-tidal CO_2 is. If it is greater than 28 mmHg, hyperventilation will help. The next maneuver is to position the bed even more head-up. It is crucial not to panic and place unwarranted pressure on the brain in an attempt to rapidly find deeper CSF spaces. The advantage of the small craniotomy is that the brain will not herniate through the cranial opening, thereby giving the surgeon time to achieve a relaxed brain. If all of these maneuvers fail, the occipital horn of the ipsilateral ventricle may be cannulated.

If any bridging veins are encountered, they should be treated with respect and preserved. The microscope is used for the interhemispheric dissection. The splenium of the corpus callosum is the first and most easily identified structure seen. The dense and consistently opaque arachnoid seen behind the splenium contains a complexity of large and important veins. It is very important to be cognizant of this as indiscriminate dissection in this area may lead to disaster. Once CSF is released and the brain is relaxed, the straight sinus is identified and the incision in the tent made parallel and at least 3 mm lateral to it. The edges of the dura are coagulated to further increase the corridor. Be prepared to stem quite copious bleeding from venous lakes in the tentorium. This is a consistent finding. Do *not* use fixed brain retraction. It is not necessary and will cause damage to the underlying brain. Another consistent finding is a bridging vein between the occipital lobe and the vein of Galen or the basal vein of

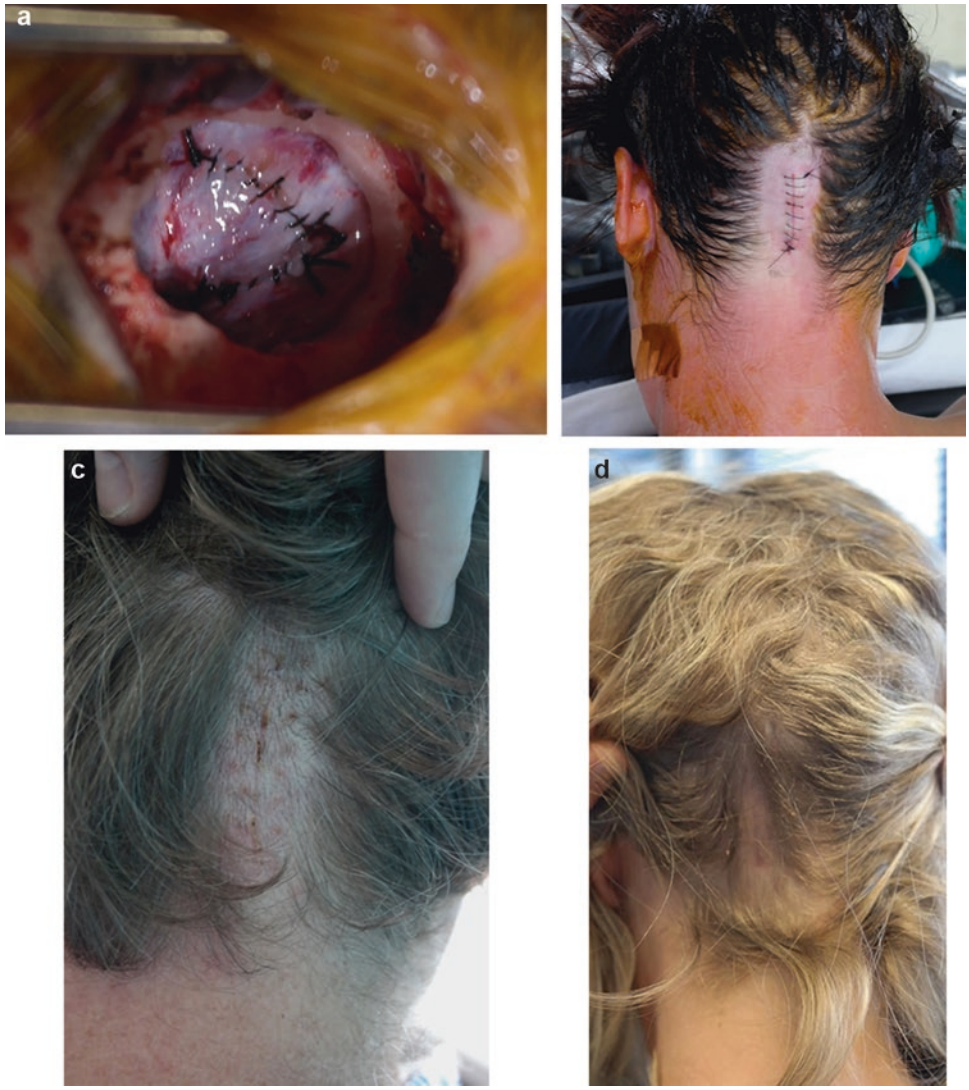

Fig. 20.13 (**a**, **b**) Given the disruption of the cisterns and the direct approach to the third ventricle, a watertight dural closure is crucial. A tight and precise multilayered closure of the muscle, fascia, and skin will also help prevent a CSF fistula. (**c**, **d**) The cosmetic result is excellent, even if the bony defect is not filled

Fig. 20.14 The occipital supratentorial approach does not require flexion or extension of the head. The neutral position is conducive to good venous return

Rosenthal (Fig. 20.16a–c). This can and probably should be sacrificed. Traction on this vein may result in damage to one of the larger and more important receiving veins. Initial tumor resection is performed with the microscope. The endoscope is only brought into the field once the surgeon feels that the microsurgical resection is complete. The endoscope now assists in viewing areas that are hidden from the microscope. The pineal region has many "hidden" *rooms*. Typically, the supratentorial approach does not afford adequate visualization of the apex of the tent. Indeed, the angle required to look superiorly into the apex is often greater than that which is given by a 30° endoscope. A 70° scope improves visualization of this area, but operating at this angle is difficult. Another "blind" spot is the area underneath the confluence of the internal cerebral veins and the vein of Galen. The endoscope is an invaluable addition to the surgeon's armamentarium with tumors that extend into the back of the third ventricle (Fig. 20.17a–e).

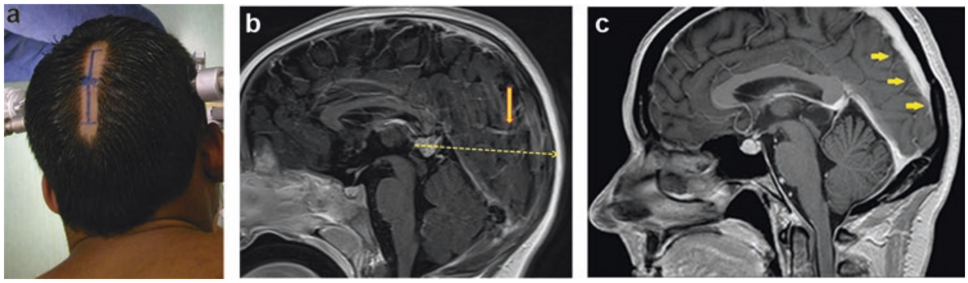

Fig. 20.15 (**a**) The midline incision is long to prepare for the possibility of a major draining vein being encountered in the middle of the craniotomy. This could potentially halve the size of the workable opening. (**b**) The preoperative MRI gives a rudimentary idea of the anatomy of the draining veins but is never 100% reliable. In this patient, the major venous complex appears to be out of the way of the ideal approach. (**c**) In this patient, with a symptomatic pineal cyst, the preoperative imaging showed multiple draining veins, all of which might have limited the occipital/interhemispheric approach. This cyst was removed using the infratentorial approach

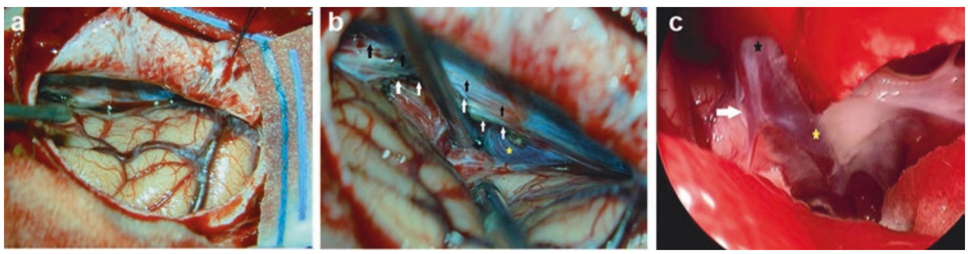

Fig. 20.16 (**a**) The large venous complex is at the anterior part of the dural opening and was preserved without compromising the approach. (**b**) The *black arrows* outline the straight sinus. The *white arrows* outline the tentorial incision. The *yellow asterisk* is the vein of Galen. (**c**) This is an endoscopic view of the occipital bridging vein (*white arrow*) attaching to the junction of the vein of Galen (*black asterisk*) and the basal vein of Rosenthal (*yellow asterisk*)

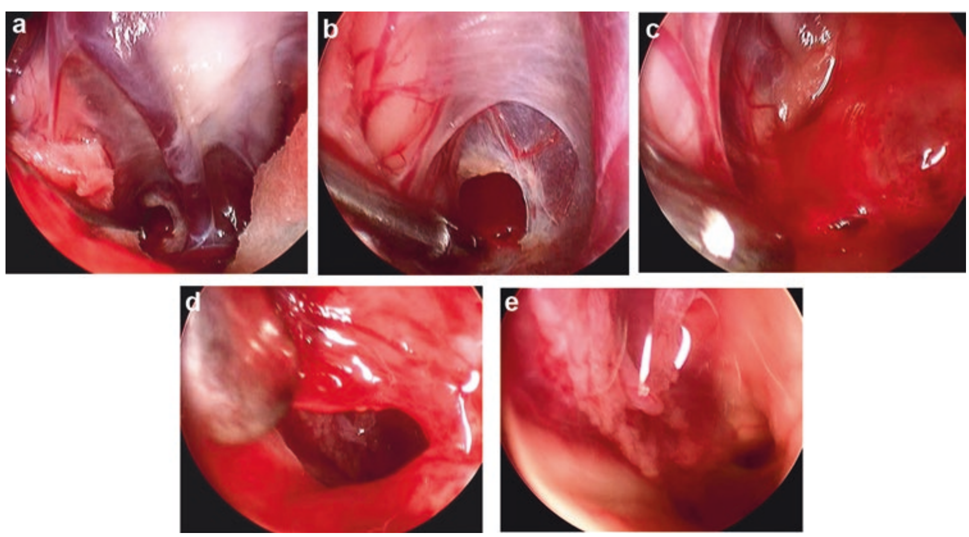

Fig. 20.17 (**a**) Once the microscopic-assisted view showed a "complete" resection of this glioma, the endoscope was brought into the field. The vein in the foreground is the basal vein of Rosenthal. (**b**) The arachnoid separating the pineal space from the ventricle has been opened. (**c**) Residual glioma is readily seen with endoscopic assistance. (**d**, **e**) Once the glioma is removed, the back of the third ventricle is opened, and the normal anatomy of the roof of the third ventricle is seen

Postoperative ipsilateral symptomatic subdural collections can occur after this approach. In my experience, this has not been a problem, and it may be due to diligent postoperative care. Patients are placed supine and with the ipsilateral side down for the first 24 h after surgery. The weight of the brain possibly reduces the potential subdural space.

Anterior/Transventricular Keyhole Approach

This anterior approach is more applicable for third ventricular tumors that extend into the pineal region but is nevertheless a reasonable alternative approach for pineal tumors [44].

Fig. 20.18 Ideal positioning of the patient for an anterior/interhemispheric approach

The patient is positioned supine with the head neutral in the coronal plane and flexed 30° in the sagittal plane. The bed is slightly headed up to encourage venous drainage. The paramedian incision is made approximately 1 cm from the midline, behind the hairline, and usually in front of the coronal suture but ultimately determined by the long axis of the tumor (Fig. 20.18). The size of the craniotomy should never be greater than 2 cm given the depth of the target area. The side of the craniotomy is determined by the long axis of the tumor. If the tumor extends to the right, then the approach is from the left and vice versa. If the dural opening is anterior to the coronal suture, as a general rule, any bridging vein can be taken with relative impunity. The interhemispheric dissection is carried out using standard microsurgical technique. The corpus callosotomy is not made until the trajectory is checked with the frameless stereotactic guidance system, in order to limit the size of the opening. When dissecting through the corpus callosum, be sure to err on the ipsilateral side. It is a common mistake to inadvertently enter the contralateral ventricle. Before extending the opening, the endoscope is useful in determining the location of the foramen of Monro. Many tumors can be removed through a transforaminal approach. If the foramen is too small, then it should be expanded by dissecting open the choroidal fissure. Any of the small bridging veins contained within this arachnoid sleeve can be sacrificed as long as the thalamostriate vein or a similarly large choroidal bridging vein is preserved. Once again, the use of self-retaining brain retractors should be discouraged. It is both unnecessary and dangerous, especially if placed against a sensitive structure such as the fornix.

Once the bulk of the tumor is removed, the endoscope is gently placed within the third ventricle. This is a difficult maneuver given the sensitive nature of all the surrounding structures. The fornices frame the foramen and roof of the third ventricle. The internal cerebral veins are contained within the velum interpositum. The wall of the third ventricle is the thalamus posteriorly and the hypothalamus anteriorly. The floor of the third ventricle is the midbrain! Any injudicious placement of the endoscope could be disastrous. When the endoscope is used in such an eloquent area, it should be positioned accurately using the microscope, and once in position, the surgeon then switches to an endoscopic view. It is not uncommon for the surgeon to maintain the microscopic view, looking at both the shaft of the endoscope and the operating instrument, while the assistant guides the dissection by looking at the endoscopic view and relaying this added information to the primary surgeon. Although this sounds counterintuitive, when an endoscope and an instrument are both within the third ventricle, any minor displacement can destroy an attenuated fornix!

Management of Preoperative Hydrocephalus

Preoperative hydrocephalus secondary to pineal region tumors can be either communicating, noncommunicating, or, rarely, both [25]. Compression of the underlying aqueduct by the tumor itself can result in a noncommunicating hydrocephalus. If so, removing the tumor will likely resolve the preoperative hydrocephalus [25, 45]. However, if the hydrocephalus is a communicating type, then decompression of the tumor will not be as successful in resolving the hydrocephalus. Depending on the tumor etiology and behavior, communicating hydrocephalus can be secondary to leptomeningeal involvement, proteinaceous obstruction of CSF outflow mechanisms, or inflammatory scarring of arachnoid granulations. Of course, if the patient presents in extremis and requires immediate CSF diversion, then an ETV or an EVD may be performed urgently for noncommunicating and communicating hydrocephalus, respectively. If an anterior transventricular approach is chosen as the route of access to the pineal tumor, the surgeon can consider doing an endoscopic third ventriculostomy within the same procedure. Otherwise, the measurement of success in the resolution of hydrocephalus by surgical management of the tumor can be seen in postoperative examinations and imaging. The decision to shunt or perform a third ventriculostomy for CSF diversion should be made in the immediate postoperative period. In our experience with pineal region tumors, patients rarely present acutely unwell, and therefore, we have never been forced into doing a CSF diversionary procedure before definitive treatment of the tumor itself [46]. Similarly, only a small percentage of patients have required CSF diversion post resection (see below).

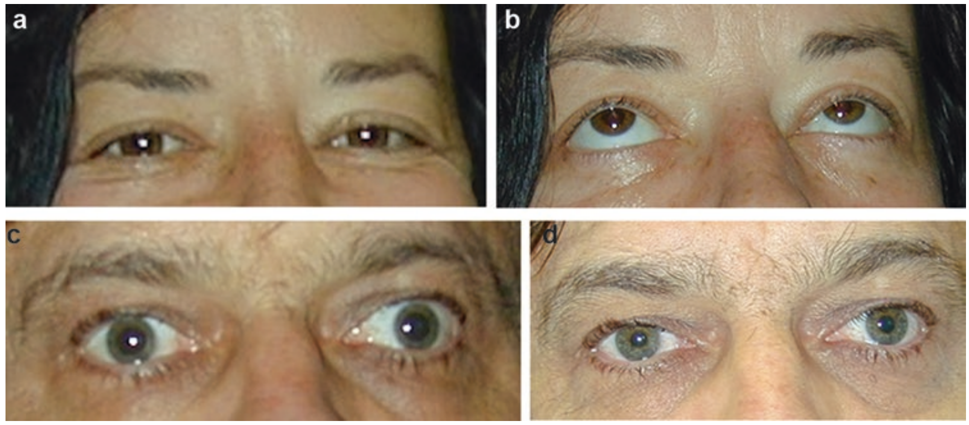

Fig. 20.19 (**a**, **b**) Normal EOM in the immediate post-op period is more commonly seen than dysconjugate EOM. (**c**, **d**) Immediate dysconjugate EOM after removal of a giant pineocytoma with slight improvement after 6 weeks and dramatic, but not complete, improvement after 1 year

Philosophy of Gross Total Resection Versus Subtotal Resection

By and large, the degree of resection of pineal tumors is best determined when the pathological diagnosis is understood. In cases of a germinoma, experts argue that tissue diagnosis is a sufficient surgical objective alone [15]. These tumors are known to be very radiosensitive, and therefore carrying out a larger tumor resection can lead to unnecessarily increased surgical risks. However, the argument for complete resection of germinomas is equally compelling. Once the approach has been made, then removing the tumor completely will likely not increase the risk of surgery any more than the risk of an open biopsy. Also, many presumed pure germinomas are in reality mixed germ cell tumors, even if serum and CSF biomarkers are negative. These tumors may benefit from a more tailored oncological approach. Finally, reestablishment of CSF pathways may require significant decompression, and therefore a complete resection would have the added benefit of obviating the need for an ETV or shunt. Non-germinomatous germ cell tumors are far less radiosensitive, and thus, we aim for gross total resections as best as possible. Surgical resection of pineoblastomas and pineocytomas plays a critical a role in their outcome, and therefore gross total resections are essential [15, 47]. In cases of pineal region gliomas and meningiomas, we advocate for a maximal safe resection to gross total resection as possible to control tumor growth. In cases of pineal region cysts, simple fenestration with generous cyst wall resection has been occasionally successful, although in our experience complete removal of the cyst is desirable [48, 49].

Outcomes

As described above, the pineal region can harbor a wide spectrum of tumor pathologies. The goals of surgery are different for each tumor type, as are the outcomes. In cases of germinomas, survival is generally higher than cases of pineoblastomas. However, it is well known that the degree of tumor resection plays a key role in tumors that are less responsive to chemotherapy or radiotherapy, and thus, a greater, safer resection should be performed with endoscopic assistance [15].

Surgery of the pineal region is plagued with danger. The old dictum of the more experience one accumulates, the lower the complication rate, certainly applies to these techniques. The keyhole techniques combined with improved visualization provided by the endoscope surely lessen the rate of collateral damage and postoperative complications and increase the oncological benefit. The most common complication is temporary and rarely permanent dysconjugate eye movements (Fig. 20.19a–d). The incidence is directly related to the degree of involvement of the tectum although the infratentorial/supracerebellar approach may have a lower incidence than the other approaches.

PEARLS
The selection of the approach will vary on the location of the tumor, the orientation of the long axis of the tumor, and the orientation of the surrounding anatomy.
Keyhole techniques are ideal for deep-seated tumor resections, such as the pineal region.
Mastery of the keyhole techniques can allow for a safer, more efficient surgery to be performed compared to traditional techniques.

Commentary on Case Presentation in Section 20.1

Case Presentation

A 19-year-old presents with incidental lesion discovered after motor vehicle accident and concussion. He is currently asymptomatic. Lesion initially followed but demonstrated substantial growth in a 6-week follow-up scan concerning for high-grade neoplasm. Serum markers are negative. No hydrocephalus. (See Fig. 20.1a–d.)

Discussion

In this case of the 19-year-old with an incidental lesion, we see a well-circumscribed mass centered in the pineal region. (It likely enhances with Gd, although we cannot make a definitive statement about this without a non-contrasted scan.) There does not appear to be ventriculomegaly or overt distortion of the neighboring structures. Its growth over the 6-week follow-up scan is concerning, and, though asymptomatic, we would prefer to surgically address this mass before it becomes symptomatic.

On review of his midsagittal MRI, the slope of the tentorium is quite steep. The trajectory to access this tumor by a supracerebellar, infratentorial approach would be quite difficult and would not be our initial option. The tumor mass is also situated in the pineal region, not in the posterior third ventricle. With respect to the location of this tumor, we would perform an occipital interhemispheric, transtentorial approach for surgical resection. We would attempt a complete resection with identification and preservation of the surrounding veins and underlying tectal plate. We would warn the patient of all the general risks associated with a craniotomy and specifically warn of the risk of dysconjugate eye movements that would probably be temporary but potentially permanent. Although the approach would obey all the rules of a keyhole craniotomy, the endoscope would probably not contribute to the resection of such a well-circumscribed lesion that has no *rooms*.

20.4 Pineal Region Tumors: Editors' Commentary

Regardless of surgical approach, decision-making in the management of pineal tumors remains the same. This includes proper management of hydrocephalus if present, evaluating serum and CSF markers, and establishing surgical goals (e.g., biopsy vs resection). When biopsy is indicated, it is critical to obtain enough tissue sampling to confirm a diagnosis, particularly as some of these tumors may have mixed pathology.

Open and keyhole approaches to the pineal region share many similarities. There may not be a dramatic difference in a keyhole versus open supracerebellar infratentorial surgery, for example. The primary difference is in relation to the area of the opening compared to the area of access to the target. The keyhole concept seeks to minimize the size of the opening but maximize access to the target. Decreasing the approach opening may help to reduce the risk of postoperative CSF leakage, operative time, blood loss, and postoperative pain. In addition, if intraoperative frozen section demonstrates radiosensitive pathology, the surgery may be reasonably terminated having performed a smaller opening. However, keyhole approaches are also less forgiving. They require exquisite understanding of anatomical relationships, careful preoperative study and planning, alternative strategies for brain relaxation, and experience. Attaining maximal target access and safely achieving the surgical goals require mastery of neuroendoscopic tools and techniques.

Larger approaches as performed with standard pineal region access may provide greater surgical freedom to maneuver instruments and perform bimanual microdissection. A reasonable size opening is required to achieve sufficient illumination of the surgical target with the operative microscope. The supracerebellar infratentorial (SCIT) approach may be more amenable to performing a smaller craniotomy due to greater availability of local CSF spaces to achieve adequate brain relaxation, and the sitting or semi-sitting position may also help to maximize the operative corridor without brain retraction. In the occipital transtentorial (OCTT) approach, a larger cranial opening may be necessary to avoid compression of the eloquent cortical surface against the lateral craniotomy edge, even despite efforts to achieve brain relaxation including hyperventilation, hyperosmolar therapy, and ventricular puncture. The lateral supracerebellar approach provides a better view of the ipsilateral interface between pineal tumors and the quadrigeminal plate but creates a blind spot on the contralateral interface. The position of the vein of Galen is also an important consideration. When the tumor has pushed this anteriorly, a posterior approach (SCIT or OCTT) is preferred. If the tumor pushes the vein of Galen posteriorly, a more anterior interhemispheric transcallosal subchoroidal approach, although a long trajectory, may prevent obstruction of the tumor by the vein of Galen.

Regardless of surgical approach, the use of endoscopic-assisted technique or intermittent endoscopic-controlled dissection can be invaluable to increase illumination and improve visualization in the deep working channel as well as to look around surgical corners that may not be visualized with the microscope alone. In the case example provided, the angle of the tentorium is quite steep. This favors either an occipital interhemispheric transtentorial or lateral supracerebellar infratentorial approach.

Frozen section and adherence of the tumor to surrounding neurovascular structures will dictate the extent of resection intraoperatively. The use of an endoscope can help confirm complete resection if that becomes the surgical goal and inspect for any tumor remnants, although given the tumor configuration, the entire tumor should be visualized well with the microscope.

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21 Clival Chordomas

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Contents

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Abbreviations

AICA	Anterior inferior cerebellar artery
CSF	Cerebrospinal fluid
CT	Computed tomography
ELD	External lumbar drainage
GTR	Gross total removal
ICA	Internal carotid artery
LC	Local control
LCR	Local control rate
MRI	Magnetic resonance imaging
OS	Overall survival
PFS	Progression-free survival
SR	Survival rate

21.1 Clival Chordomas: Editors' Introduction

Clival chordomas are histologically benign, but clinically aggressive tumors due largely to their propensity for local invasion and recurrence as well as their proximity to critical neurovascular structures. These tumors are thought to originate from remnants of the notochord. Clival chordomas occur more frequently in men, with a mean age in the mid-40s [1].

Diplopia is the most common complaint in patients with clival chordomas, often from abducens palsy [2]. Other symptoms include headache, visual disturbance, and disequilibrium. Radiographically, chordomas are located in the midline within the clivus and can extend posteriorly toward the brainstem, superiorly to compress the pituitary gland, anteriorly into the sphenoid sinus and retropharynx, and inferiorly into the craniocervical junction and upper cervical spine. Lateral extension into the cavernous sinus can occur in up to 75% of cases [3]. A CT scan will show evidence of cranial base bony destruction and marrow replacement. MRI

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typically shows a T1 hypo- to isointense lesion with heterogeneous enhancement and T2 hyperintensity. Neither of these characteristics is completely distinct to chordomas and may also be seen with chondrosarcomas.

Histologically, chordomas are low-grade neoplasms characterized by polygonal cells with an eosinophilic cytoplasm and eccentric nuclei within the mucoid matrix. The hallmark pathologic feature is identification of vacuolated physaliphorous cells [1]. A chondroid type variant does exist which is differentiated by the presence of hyalinization and may carry a better prognosis.

Surgery remains the primary mode of diagnosis and treatment for this tumor. Aggressive surgical resection to achieve a gross total removal with clear bony margins is ideal; however, radical resection is often limited by the local invasiveness of the tumor and involvement of surrounding neurovascular structures making resection challenging even when multiple surgical approaches are utilized. Progression-free survival can be as high as 79% at 5 years, especially when gross total resection is coupled with radiation therapy [4]. Various surgical procedures have been described ranging from open cranial base approaches to less invasive endoscopic endonasal approaches. In this chapter, the authors describe the advantages and limitations of open and endoscopic techniques as well as discuss their approach to the case illustration below (Fig. 21.1a–e).

21.2 Clival Chordomas: Open Transcranial Approaches

Wenya Linda Bi and Ian F. Dunn

Introduction

Although chordomas are classified as a histologically benign tumor, their propensity for aggressive growth and local invasion and recurrence requires an oncologic approach in their management. Radical surgical treatment has been associated with improved long-term disease-free survival, but also heightened morbidity [4–7]. The clivus in particular – long the target of a wide range of surgical approaches – challenges the choice and execution of surgery. Moreover, chordomas exhibit relative radioresistance, but administration of high-dose radiation appears to demonstrate synergistic disease control when combined with radical resection. Taken together, surgery remains the most effective treatment for both newly diagnosed and recurrent chordomas, with the imperative for the surgeon to minimize the risk profile through safe skull base techniques.

Preoperative Evaluation

A comprehensive history and physical examination are complemented by ophthalmologic, audiologic, endocrinologic, and swallow assessments. Specifically, for lesions involving the upper clivus and sella, endocrine labs may be warranted to assess for hormonal hypersecretion or insufficiency. Ophthalmologic examination establishes a baseline for extraocular movements, especially that of the abducens nerve. Lateral extension at the level of the internal auditory canal (IAC) merits careful assessment of facial function and a formal audiogram. For lesions involving the lower clivus and oropharynx, formal swallow evaluation provides a baseline in the setting of lower cranial nerve involvement or an anticipated approach involving the oropharynx. Should the lesion extend lower and involve C1, craniocervical stability should be assessed.

MRI of the brain classically reveals a T1-hypointense to isointense, T2-hyperintense, and frequently heterogeneous mass of the clivus, with variable extension into the posterior fossa, middle fossa, sphenoid sinus, and suprasellar space. High-resolution CT provides additional value in delineating the extent of osseous involvement, including erosion or expansion, for operative planning. This is frequently obtained in conjunction with a CT angiogram and venogram (CTA/V) to allow depiction of vascular relationships. Dynamic CTA/V additionally offers temporal resolution in providing vascular flow-related data, if available [8], although formal digital subtraction angiography with balloon occlusion test to evaluate for collateral circulation may be indicated in the setting of internal carotid artery ensheathment by the tumor.

Patient Selection and Goals of Surgery

The goals of surgery reflect that of other locally aggressive skull base tumors: relief of mass effect, preservation or restoration of neurologic function, cytoreduction and minimization of tumor recurrence, and procurement of a histopathologic diagnosis. Given the complementary role of adjuvant radiation for chordomas, risks incurred by surgery should be weighed carefully as part of a multimodality treatment plan. When radical removal of tumor is deemed imprudent because of the risk to critical neurovascular structures, an attempt should be made to improve the safety profile between tumor and normal neural tissue for subsequent radiation treatment fields.

Reoperation, following prior surgical treatment or radiotherapy, is associated with higher rates of postoperative complications. Indications for repeat surgery include control of locally progressive disease, alleviation of symptoms from

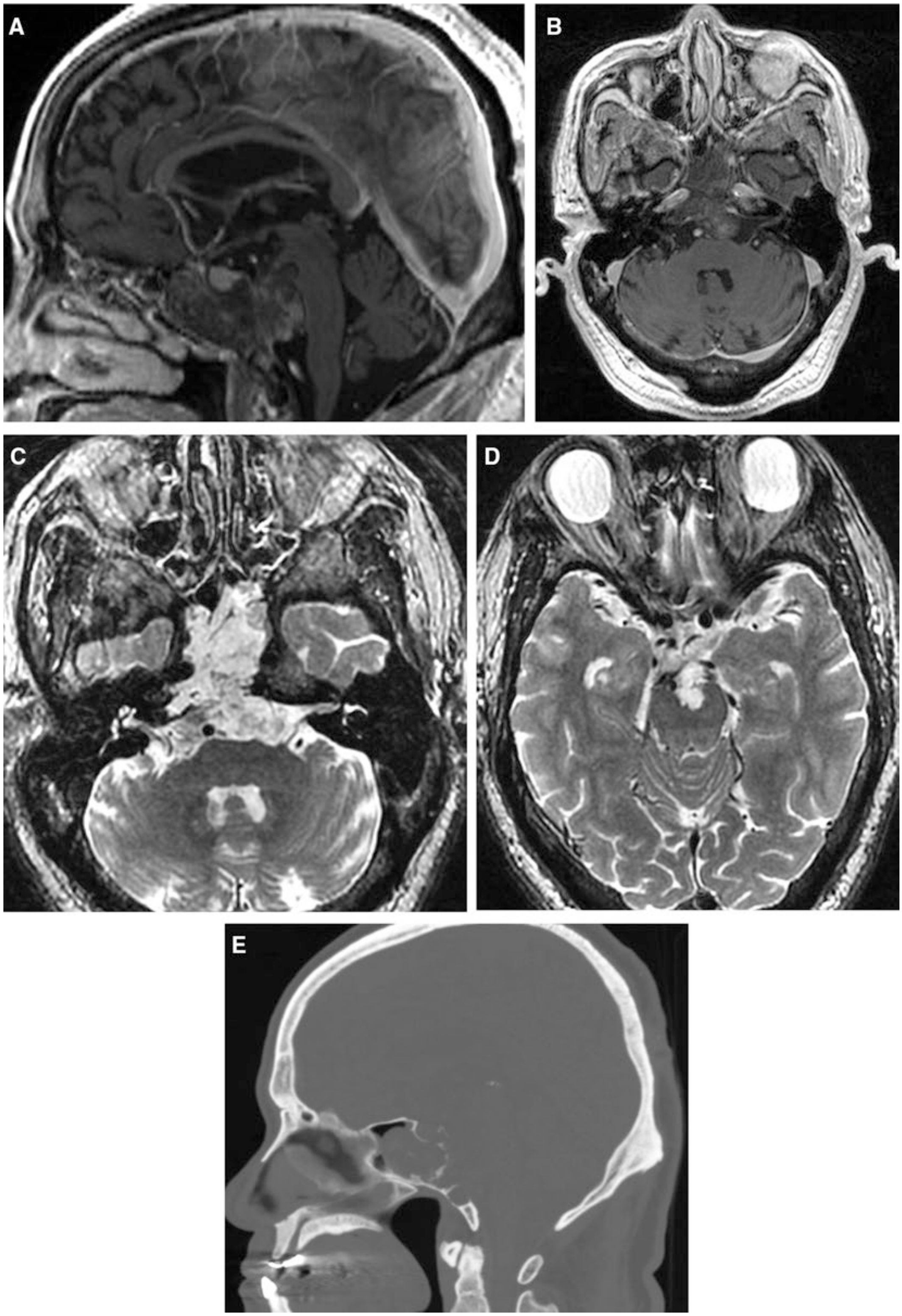

Fig. 21.1 (**a–e**) **CASE EXAMPLE**: clival chordoma. A 68-year-old man presents with 2 months of double vision and headaches. Examination: right sixth nerve palsy, otherwise neurologically intact. Laboratory workup: normal endocrine function

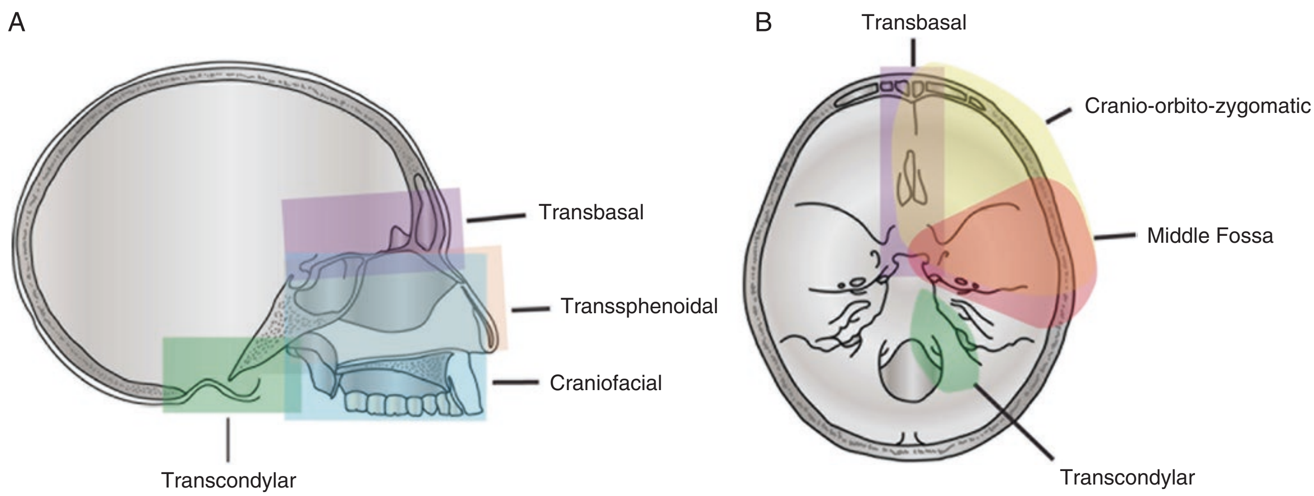

Fig. 21.2 (**a**, **b**) Surgical approaches to the clivus. Illustration of target anatomic regions accessed by the transbasal, transsphenoidal, craniofacial, cranio-orbito-zygomatic, middle fossa, and transcondylar approaches in (**a**) sagittal and (**b**) axial views

neural compression, and providing a safer margin between residual tumor and critical neurovascular structures for subsequent radiation. In patients with recurrent chordoma, re-resection with curative intent to achieve gross total resection is targeted toward patients with isolated disease, long disease-free interval, and good functional status [9].

Surgical Approaches

The choice of surgical approach depends on tumor size, its extent of growth, the patient's clinical status, and the surgeon's operative philosophy and comfort (Fig. 21.2a, b). One surgical classification for clival chordomas based on their patterns of extension defines type I tumors as restricted to a solitary anatomic compartment, type II tumors as extending to two or more contiguous areas, and type III tumors as those with extensive spread requiring two or more skull base procedures to achieve radical resection [10]. Tumor growth often creates a natural surgical corridor by eroding the obstructing bone and expanding or displacing the soft tissue and vascular structures. The endoscope can further abet visualization of obscured corners to seek out isolated nests of tumor during microscopic approaches. If necessary, skull base approaches can be combined to ensure the goals of surgery are maximized.

Open surgical approaches to clival chordomas frequently place a significant demand on meticulous reconstruction to prevent leakage of cerebrospinal fluid (CSF) and to optimize cosmesis. Additionally, planning for the surgical approach should also account for the risk of recurrence from chordoma seeding of the surgical tract, including the fat graft site [11].

Intraoperative Considerations

Given the extensive growth patterns of chordoma and its propensity for multi-compartment invasion, neuronavigation with high-resolution CT and MRI provides a helpful adjunct to localize tumor and assess the extent of resection. Doppler ultrasound may be used to verify the location of the carotid and vertebral artery. Additionally, as resection proceeds, endoscopic inspection frequently reveals nests of tumor which may be obscured by immobile neurovascular structures. These tools should be incorporated into routine use at skull base centers with fluency in the surgical treatment of clival chordomas.

Neurophysiologic monitoring is regularly integrated in the surgical treatment of chordomas. Somatosensory evoked potentials (SSEPs) and brainstem auditory evoked responses (BAERs) are routinely applied for all tumors, especially with posterior fossa involvement. Cranial nerves are monitored as necessary, based on the selected approach and potential involvement by the tumor. Innervation of extraocular muscles and the facial nerve is monitored in anterolateral and lateral approaches, especially the extended middle fossa approaches. Lower cranial nerves are monitored during the transcondylar approach.

Anterior Approaches

Transbasal Approach

Anatomic Target

Chordomas arising from the upper clivus, with the involvement of the sella and sphenoid sinus, may be accessed from a rostrally based anterior approach, capitalizing on the subfrontal corridor. These transbasal approaches allow for craniofacial resection of tumor without the risks entailed by transfacial exposure. Transbasal approaches, with their many variations, allow a rostral-caudal corridor from the anterior cranial fossa and suprasellar space to the arch of C1 [12–15]. The internal carotid artery, optic nerve, globe of the eye, Dorello's canal, and hypoglossal nerve limit the corridor laterally. The posterior clinoid has been considered a "blind spot" in the traditional descriptions of the approach [14].

Surgical Technique

The patient is positioned supine with the head in neutral position. A bicoronal incision behind the hairline allows the exposure for all transbasal approaches, without the potential cosmetic considerations of a periorbital or facial incision. As the scalp is mobilized forward, vascularized pericranium should be preserved and harvested as a pedicled flap for reconstruction at the end of the surgery. Removal of the orbitonasal bar minimizes retraction on the frontal lobe, which can be combined with or without a low bifrontal craniotomy (Fig. 21.3) [15, 16]. Osteotomies through the frontonasal ducts, nasal septum, cribriform plate, or planum sphenoidale can be connected at the lateral boundaries, as defined by the junction of the orbital roof and the lamina papyracea. The lamina papyracea itself can be removed for access to the periorbita if the tumor extends into the orbit. Removal of the lateral orbit permits greater retraction of orbital contents

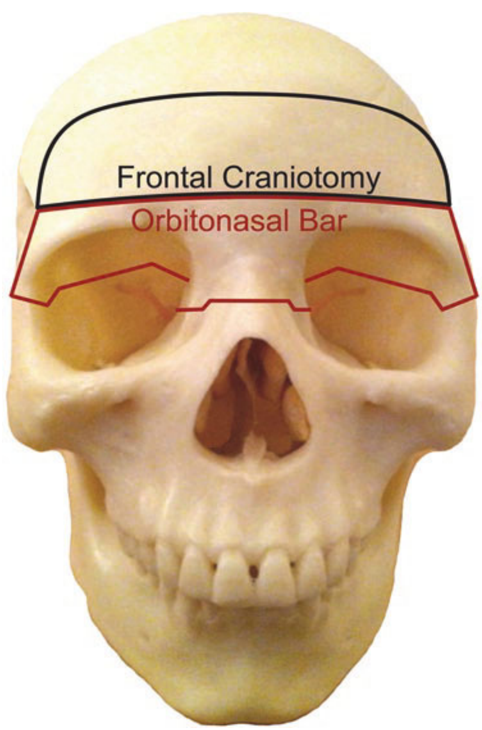

Fig. 21.3 Transbasal approaches incorporate a low bifrontal craniotomy with or without an orbitonasal bar, to allow subfrontal access to the superior and mid-clival area

from medial to lateral. Osteotomy around the cribriform plate may also be performed to preserve olfaction, if uninvolved by the tumor. Exenteration of the ethmoid, sphenoid, and frontal sinuses ensues, as indicated, for tumor resection and reconstruction. The clivus is drilled from the sella floor to the foramen magnum, as indicated by the extent of the tumor. The dura is opened if necessary for tumor extension.

Reconstruction

Closure begins with watertight dural repair, if feasible. The anterior cranial fossa floor defect is most effectively sealed with a vascularized graft, such as the pedicled pericranial or a rotated temporalis flap. To augment this, or in the absence of vascularized grafts, fat, fibrin glue, free flaps, fascia lata, and other adjuncts may also be used. In the face of extensive osseous removal, a bony strut, such as the inner table of the craniotomy, may be laid across the anterior fossa floor, but this is rarely required. The medial and lateral canthal ligaments are reapproximated, if cut. The frontal bone flap and orbitonasal bar, in one or separate pieces, are then secured to the frontal, nasal, and maxillary bones. Augmentation of crevices with hydroxyapatite or bone substitute may improve the cosmetic outcome.

Transoral Approach

Anatomic Target

The transoral approach may be considered for extradural mid- to lower clival chordomas extending from the nasopharynx to the top of C3 [17]. Midline tumors with ventral extension are favored by this approach, while extension lateral to the jugular or hypoglossal foramen or intradural extension may be better handled by a unilateral or bilateral transcondylar approach.

Surgical Technique

The patient is positioned supine with the head slightly extended. A self-retaining oral retractor displaces the tongue inferiorly and the soft palate superiorly, creating a minimal aperture of 2.5 and 3 cm between the incisors. Local anesthetic is infiltrated into the mucosa of the posterior pharyngeal wall. The soft palate and posterior 1 cm of the hard palate may be divided for visualization, although this is unnecessary with endoscopic assistance for visualization. An inferior incision from the nasopharynx to the inferior border of C2 allows for lateral retraction of the mucosa and subsequent separation of the longus capitis and longus colli muscles from the ventral face of the clivus and cervical vertebrae, respectively. The Eustachian tube, lower cranial nerves in the jugular foramen, hypoglossal nerve in its canal, and the vertebral artery confer a natural lateral boundary for the exposure, which may be displaced from their normal positions by the growing chordoma. The clivus, anterior arch of C1, and the axis are drilled as necessary for tumor resection. If tumor is pursued into the intradural compartment, the venous lakes of the ventral dura behind the clivus should be adequately coagulated.

Reconstruction

A primary dural closure may be difficult to achieve in this setting, and intradural or epidural reconstruction with dural substitute or fascia lata can be reinforced with fat and fibrin glue. Reapproximation of the longus colli muscles and posterior pharyngeal mucosa follows.

Craniofacial Approaches

Anatomic Target

Transfacial approaches employ the natural air spaces as well as the modular nature of the nasal, paranasal, and oropharynx cavities for direct access to the clivus. These encompass transethmoid, transmaxillary, transsphenoidal, and facial translocation procedures, all of which can be combined with transbasal approaches. In the endoscopic era, approaches utilizing transfacial incisions are becoming relics in some centers and are reserved most commonly for invasive cancers in which covering tissue may also require resection in addition to the main target lesion. Open access to these midface corridors is increasingly supplemented or replaced by endoscopic techniques, which allow ready access from the anterior skull base to C2 in the rostral to caudal direction, and is discussed separately. The anatomic landmarks and surgical principles for ventral midline approaches remain constant, regardless of the technique.

Lateral anatomic boundaries for midline anterior approaches are established by the orbital globe, optic nerve, cavernous sinus, internal carotid artery, bilateral foramina for the exiting cranial nerves, the Eustachian tube, and the vertebrobasilar complex. The supraclinoid, cavernous, and petrous segments of the carotid artery may be displaced, along with investing soft tissues and remodeled osseous sheaths by the midline-based chordoma. However, tumor extension lateral to neural foramina and cranial nerves may necessitate an alternative or additional lateral approach if the goal of surgery is radical resection.

Surgical Technique

The patient is positioned supine with the head in neutral to slightly extended position, based on the caudal extent of tumor growth. The endotracheal tube should be secured inferiorly, or wired to the mandible, if extensive manipulation of the midface is anticipated. Preoperative placement of a nasogastric feeding tube may also be desired in the setting of anticipated postoperative swallowing dysfunction. Temporary tarsorrhaphies are often performed. The specific incision in the face depends on the area of attack.

Transmaxillary Approach

The transmaxillary approach involves a unilateral maxillotomy with paramedian splitting of the palates or the LeFort I osteotomy, with or without midline split of the hard and soft palates (Fig. 21.4a, b) [18–20]. All variants of this approach

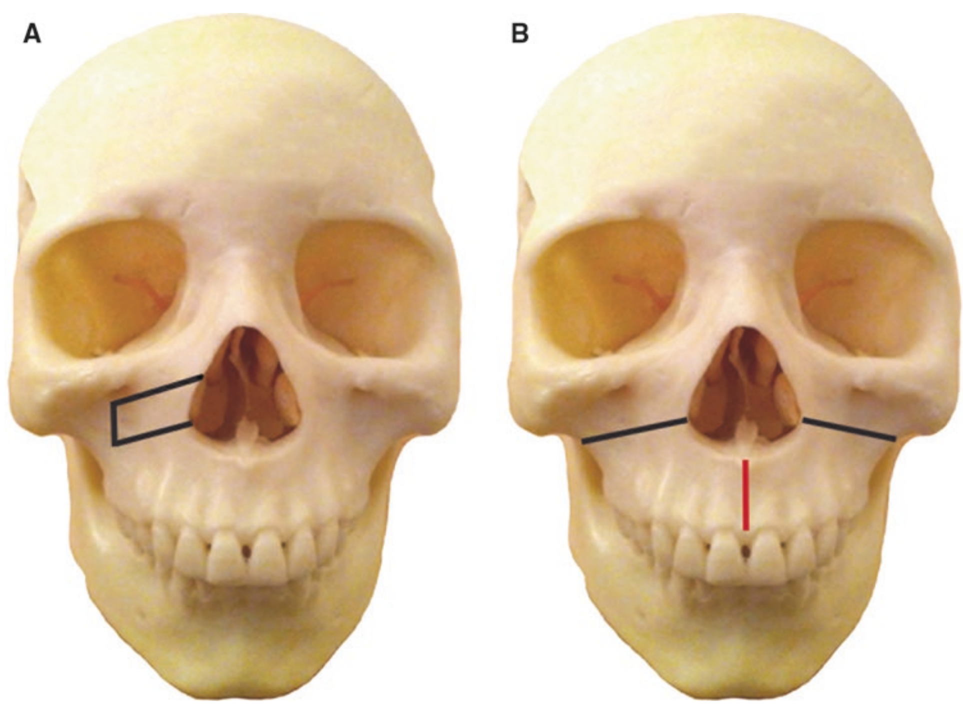

Fig. 21.4 (**a**, **b**) Transmaxillary approaches employ sinus space to create surgical corridors. (**a**) Unilateral maxillotomy osteotomy cuts (*black line*). (**b**) Bilateral osteotomies along the maxillary face (*black line*) with midline split (*red line*)

provide access to midline lesions of the full clivus, from the anterior cranial fossa to the superior aspect of C3. The internal carotid artery, pterygoid plate, jugular foramen, and hypoglossal canal bound the exposure laterally. Local anesthetic is infiltrated into the nasoseptal and gingivobuccal mucosa. A transfixion incision is made anterior to the nasal septum as well as a transverse sublabial gingivobuccal incision extending to both maxillary tuberosities. Perichondral and periosteal dissection of mucosa from the nasal septum and maxillary face continues until the infraorbital foramen and the entire superior surface of the hard palate are exposed. Upon exposure of the maxilla, the osteotomy cuts and splitting of the palates varies based on surgeon preference.

A LeFort I osteotomy cuts along the maxilla face, superior to the piriform aperture, parallel to the dental arch, to allow for downward displacement of the maxilla, thereby increasing the exposure of the bony septum and posterior pharynx. A unilateral transverse cut can be combined with two vertical osteotomies, one midline and one laterally, to create a unilateral maxillotomy if the tumor extends asymmetrically to one side. A median split of the hard palate can be accomplished, with or without sparing of the soft palate and its vascular supply to the maxillary segments. Prior to any osteotomies of the maxilla and midface, the drill holes for the attachment of titanium plates should be made to facilitate subsequent reconstruction. The resection defect is filled with fat, fibrin glue, dural substitute, and possible pedicled nasoseptal flap in the face of a CSF leak. The soft tissues and palate are reapproximated, the maxilla returned to its anatomic position, and the nose packed. One risk of dividing the hard palate is velopharyngeal insufficiency and oronasal fistulae. Furthermore, postoperative respiratory and swallow function should be monitored closely.

Facial Translocation

Facial translocation approaches utilize the concept of modular craniofacial disassembly to provide wide exposure to the ventral skull base through mobilization of the soft tissue and bone but carry concordant risk for significant cosmetic dissatisfaction to some patients [21–24]. The embryologic fusion of tissues at the midline, bilateral neurovascular supply of facial units, and internal hollow spaces provided by paranasal sinuses and the oral and nasal cavities motivate the principles of transfacial approaches. The midfacial translocation variant combines the exposures provided by transethmoid, transsphenoid, and transmaxillary approaches. This can be extended into a standard facial translocation to expose the infratemporal fossa. Further medial, inferior, or bilateral extensions allow for stepwise exposure of bilateral sphenoid, ethmoid, maxillary sinuses, nasopharynx, oropharynx, pterygomaxillary fossa, infratemporal fossa, and paracentral skull base, from the anterior cranial fossa to C3. A preauricular frontotemporal incision and craniotomy can also be incorporated into the exposure.

For the central midfacial translocation, the endotracheal tube is wired to the mandibular dentition, or a tracheostomy is considered upfront. Local anesthetic is infiltrated into the nasal soft tissues and superficial skin incision. A Weber-Ferguson incision, with possible extension across the bridge of the nose, is frequently employed. Extensive soft tissue dissection to expose the facial bones should account for handling of the medial canthal ligaments, lacrimal sac, and ethmoidal arteries. Drill holes are made for subsequent plating of the midface construct before osteotomies of the nasomaxillary, nasofrontal, zygomatic-orbital, and pterygomaxillary bones. The nasal septum and palate are also freed to permit lateral rotation of the soft tissue and osseous flap. Prevertebral soft tissue is incised and dissected for exposure of the clivus and tumor, as discussed above. Meticulous dural closure is achieved with fat, fibrin glue, dural substitute, or pedicled nasoseptal flap, especially if in the setting of CSF leak. Soft tissues are reapproximated in a layered fashion, especially of the soft palate to prevent velopharyngeal fistula; the bones replated using original drill holes; and the facial incision closed with plastic surgery techniques.

Anterolateral Approaches

Cranio-orbito-zygomatic Approach

Anatomic Target

The cranio-orbito-zygomatic (COZ) approach opens a broad anterolateral trajectory that allows exposure along the subfrontal-transbasal, transsylvian, subtemporal, and infratemporal routes [25, 26]. Chordomas arising from the upper clivus, with lateral extension to the parasellar area, cavernous sinus, middle fossa, petrous apex, infratemporal fossa, or intradural, may be effectively addressed by this approach.

Surgical Technique

A skin incision is fashioned starting at the tragus and extending toward the contralateral superior temporal line behind the hairline, with care to preserve the superficial temporal artery. The scalp is flapped forward to allow harvest of a pedicled pericranial graft for subsequent reconstruction. The superficial and deep temporalis fascia is sharply incised posterior to the fat pad containing the frontal branches of the facial nerve and reflected forward in the subfascial plane to allow exposure of the lateral orbit and zygoma. The zygoma is cut and deflected downward to allow maximal inferior mobilization of the temporalis muscle. A one- or two-piece craniotomy is fashioned to remove the lateral and superior orbit, usually to the point of the supraorbital nerve, although medial extension of the orbitotomy can be achieved by freeing the nerve from its notch or foramen, thereby allowing for an extended subfrontal access. Further removal of the sphenoid wing and anterior clinoid may abet exposure. Extradural dissection can be pursued to reveal the cavernous sinus, Meckel's cave, petrous apex, and planum sphenoidale. Mobilization of the dura propria, or lateral wall, of the cavernous sinus and the investing nerve sheath of trigeminal branches from the inner membranous layer allows access to tumor that has invaded the cavernous sinus. Resection of extradural and intradural tumor, as well as removal of any infiltrated dura, proceeds with attention to involved perforating vessels. Overall, upper clival and anterior/middle fossa exposure can be afforded through this approach.

Reconstruction

The dura is closed primarily when possible. Potential openings into the frontal, sphenoid, or ethmoid sinuses and to the Eustachian tube are obliterated with fat or muscle and fibrin glue. Vascularized pericranium can then be positioned under the frontal lobe or over any sinus entry zones in the middle fossa or petrous apex. When pericranium is not readily available, such as in the setting of reoperation, or with larger defects, a pedicled temporalis muscle may be rotated inferiorly for reconstruction. The orbital roof is reconstructed to prevent enophthalmos or pulsatile exophthalmos. The cranio-orbital flap is replated, with calvarial defects, especially along the orbital rim, augmented by bone substitute such as hydroxyapatite. The temporalis muscle is realigned to the superior temporal line, either to a fascial cuff or to securing holes placed in the cranium. The zygoma is returned to anatomic position with titanium plates, and the scalp is closed in a layered fashion.

The cosmetic outcome of the cranio-orbito-zygomatic approach is favored by preservation of the temporalis muscle vasculature and reconstruction of the orbit. The temporalis muscle is supplied by branches of the superficial temporal artery as well as deep temporal arteries medially [27]; preservation of these branches during scalp incision and forward mobilization of the muscle from the bone minimize subsequent risk for temporalis atrophy.

Lateral Approaches

Extended Middle Fossa Approaches

Anatomic Target

For chordomas of the upper or mid-clivus with extension to the cavernous sinus, lateral to the internal carotid artery, middle fossa, petrous bone, infratemporal fossa, and pterygopalatine fossa, an extended middle fossa approach may be indicated. These encompass the zygomatic middle fossa approach, an anterior petrous approach, or a combination of the two [28]; note that both of these may be achieved through the more comprehensive COZ. The extended middle fossa approach provides access from the superior orbital fissure to the internal auditory canal, especially with the removal of the petrous apex.

Surgical Technique

The patient is positioned supine with the ipsilateral shoulder bolstered by a roll, and the head is turned toward the contralateral side such that the middle fossa becomes vertical and the temporal lobe can fall away from the field during extradural dissection. A preauricular question mark incision originates at the level of the superior temporal line behind the hairline, curves over the top of the auricle, and terminates in front of the tragus. This may be extended inferiorly, to the level of the mandible, if needed for infratemporal fossa exposure. The superior temporal artery is preserved to minimize subsequent temporalis wasting. The temporalis fascia is dissected from the lateral orbital rim and superficial surface of the zygoma, and the zygomatic arch is cut at its origin and insertion and deflected downward. The temporalis muscle is then mobilized downward through a subperiosteal dissection in an inferior to superior and posterior to anterior direction, until it is detached from the superior temporal line. A temporal craniotomy is then constructed.

Extradural dissection along a broad front proceeds from a posterior to anterior direction to identify the middle meningeal artery at the foramen spinosum. The foramen ovale and the mandibular branch of the trigeminal nerve lie immediately anteromedial to the spinosum and frequently cover the traversing internal carotid artery, which should be identified before drilling ensues for the anterior petrosectomy. The horizontal segment of the internal carotid artery is often exposed further with a high-speed diamond drill, to allow placement of a temporal clip or a Fogarty balloon catheter, if necessary, for proximal control. Care should be taken to avoid traction on the greater superficial petrosal nerve (GSPN), which may transmit to its origin at the geniculate ganglion, to prevent inadvertent injury to the facial nerve, which should be monitored. Anterior petrosectomy drilling is limited by the internal carotid anteriorly underneath V3, the IAC posteriorly, the cochlea laterally, and the inferior petrosal sinus and jugular bulb inferiorly. If the tumor extends intradurally, a low transverse cut in the temporal dura is made, and a vertical one in the posterior fossa dura is made; the superior petrosal sinus is coagulated and cut. The fourth nerve is identified prior to sectioning of the tentorium to access the posterior fossa, and care is taken not to inadvertently section the nerve. The cavernous sinus can also be exposed by sharply dissecting the dura propria and investing the dural sheath of the trigeminal nerve from its inner membranous layer.

Reconstruction

The dura is closed primarily when able. Potential openings into the frontal, sphenoid, or ethmoid sinuses and to the Eustachian tube are obliterated with fat or muscle and fibrin glue. Vascularized pericranium can then be positioned under the frontal lobe or over any sinus entry zones in the middle fossa or petrous apex. When the pericranium is not readily available, such as in the setting of reoperation, or with larger defects, a pedicled temporalis muscle may be rotated inferiorly for reconstruction. The orbital roof is reconstructed to prevent enophthalmos. The cranio-orbital flap is replated, with calvarial defects, especially along the orbital rim, augmented by bone substitute such as hydroxyapatite. The temporalis muscle is realigned to the superior temporal line, either to a fascial cuff or to securing holes placed in the cranium. The zygoma is returned to anatomic position with titanium plates, and the scalp is closed in a layered fashion.

Transcondylar Approach

Anatomic Target

The transcondylar, or extreme lateral, approach well suits chordomas based within the lower clivus and craniocervical junction, with or without lateral extension [29, 30]. The position of the jugular bulb dictates the superior limit of this approach, although it can be combined with a transtemporal approach superiorly or an infratemporal approach inferiorly for expanded access.

Surgical Technique

The patient is positioned supine with a large ipsilateral shoulder roll, and the head is slightly turned or in a lateral decubitus position. The vertex is tilted slightly toward the contralateral shoulder, and the ipsilateral shoulder is gently displaced inferiorly to optimize the working angle. If craniocervical fusion is anticipated due to disruption of the atlantoaxial joint and condyles, either by tumor or by the approach, the caput should be positioned in a neutral position relative to the neck. A C-shaped incision is made beginning at the level of the pinna, 3 cm behind the mastoid tip, and extends inferior and forward to reach the anterior border of the sternocleidomastoid muscle at the level of the mid-cervical spine. The skin is mobilized forward in a subfascial plane. The transverse process of C1 is identified through dissection along the anterior border of the sternocleidomastoid muscle. The sternocleidomastoid, splenius capitis, and longissimus capitis muscles are then mobilized inferiorly after detachment from the mastoid. The spinal accessory nerve should be identified at its entrance into the sternocleidomastoid muscle. The suboccipital triangle and the horizontal segment of the vertebral artery should be identified, after which the superior and inferior oblique muscles can be mobilized from the transverse process of C1. Preservation of the alveolar tissue around the suboccipital venous plexus ensheathed around the vertebral artery minimizes bleeding during manipulation and displacement of the vertebral artery. If required, the vertebral may be mobilized out of the C1 transverse foramen. The mastoid is drilled to expose the jugular bulb and occipital condyle, which is removed as needed to gain access to the lower clivus and retropharyngeal space, as dictated by the tumor. Endoscopic inspection is frequently helpful to identify hidden islands of tumor.

Reconstruction

Primary dural closure should be attempted if the dura is opened. Fat, dural substitute, and fibrin sealant are applied for reinforcement. Occipital-cervical fusion is performed if eventual craniocervical instability is strongly suspected. The mastoid cortex is reconstructed, with bone substitute for cosmetic repair. Muscle layers are reapproximated and the skin is closed. Pseudomeningoceles may develop if dural closure is incomplete, particularly due to the multilayer dissection. A tight pressure wrap is applied after surgery and kept on for several days to augment fastidious reconstruction.

Complication Avoidance

General Considerations

Skull base surgery for chordoma resection faces approach-specific risks as well as generalized considerations for complication avoidance. CSF leak and injury to cranial nerves and vessels are the most frequent concerns across all surgical approaches.

Potential sites for CSF leak include the facial sinuses, air cells, the Eustachian tube, and incompetent dural closures. CSF leak is most effectively prevented by a layered closure with vascularized tissue, such as pericranium or a pedicled muscle flap, and obliteration of dead space with fat, fibrin glue, or dural substitute. Postoperative CSF leak may be observed in a delayed fashion through egress from drilled anterior clinoid, petrous apex, or mastoid air cells. While lumbar drain placement for CSF diversion may temporize a leak, re-exploration and repair are often indicated in the setting of early or persistent CSF leak.

Risk to cranial nerves and the internal carotid artery or vertebrobasilar complex is dictated by the pattern and extent of tumor growth. Proximal control of the carotid artery should be prepared for and can be obtained at the petrous carotid or at the cervical segment. Previous radiation to the operative bed increases risk of vascular injury. In the event of injury, primary vascular repair is optimal, if feasible; otherwise, tamponade with the muscle or autograft should be achieved, followed by endovascular imaging. Postoperative epistaxis and retrobulbar hematoma are additional bleeding risks from injury to branches of the sphenopalatine artery and ethmoid arteries, respectively.

Care should be taken to avoid seeding of the surgical tract, including of the abdominal fat graft site, with chordoma cells, which display an avid propensity to uptake into host sites.

Transbasal Approach

Complications from transbasal approaches arise from exposure, tumor resection, and inadequate reconstruction. Anosmia may result either from osteotomies involving the cribriform plate or from retraction of the frontal lobes with stretch of the olfactory nerves. A unilateral cribriform osteotomy to preserve the olfactory fibers on one side may permit functional olfaction postoperatively. Ocular complications (including epiphora, diplopia, enophthalmos, and loss of vision) may result from injury to the lacrimal system, extraocular innervation, and optic nerve. Specifically, the lacrimal sac may be injured during removal of the medial orbital wall, with potential for postoperative epiphora. Manipulation of the periorbita and retraction of the orbit may result in enophthalmos as well as risk to the oculomotor and trochlear nerves. The abducens nerve is also vulnerable throughout its course in the clival basal dura and at its insertion into Dorello's canal during drilling of the clivus.

Frontal contusion, pneumocephalus, and epidural hematoma from potential space are further risks of the subfrontal approach. Inadequate reconstruction of the anterior cranial fossa may result in CSF leak, meningocele, encephalocele, or pulsatile exophthalmos. Care in reconstruction of the nasal bone is exerted to prevent a saddle nose deformity.

Craniofacial Approaches

Craniofacial procedures demand meticulous reconstruction for acceptable cosmesis. Optimal alignment of the maxilla, nasal bone, and other midface constructs is abetted by creating drill holes in the plating construct prior to any osteotomies. Low-profile titanium plates and screws should be positioned in areas with maximal overlying soft tissue and fat, if possible. The skin is closed in a layered fashion, with care for flawless epidermal apposition.

In craniofacial approaches, significant devitalization or removal of nasal mucosa may result in sinusitis, crusting, and adhesions, with negative impact on the patient's sinonasal quality of life. Olfaction may be impaired by damage to the olfactory cilia in the superior nasal mucosa. Facial numbness results from sectioning of the infraorbital nerve during the midfacial translocation approach. Aseptic necrosis or collapse of the nasal dorsum and the maxilla may result from excessive devascularization from osteotomies. Incompetent closure of the soft palate risks velopharyngeal insufficiency, with consequent hypernasality, swallowing dysfunction, or breathing difficulty.

Anterolateral Approaches

Specific consideration of the internal carotid artery, the facial nerve, and hearing function should be made during the extended middle fossa approach. The horizontal petrous carotid artery is exposed during initial extradural dissection of the temporal lobe and remains at risk during the petrous apex drilling. One should prepare for proximal control, either at the petrous segment or with cervical carotid exposure.

The facial nerve may be injured at several stages of the approach and tumor resection. First, during initial extradural dissection along the middle fossa, the geniculate ganglion may be dehiscent and inadvertently injured. Traction on GSPN during the dissection may also avulse the facial nerve at the geniculate ganglion. Lastly, resection of posterior fossa tumor extension as reached from the middle fossa may also result in injury of the facial nerve at the internal auditory canal. Continuous facial nerve monitoring is especially helpful during this approach.

Hearing may be jeopardized through injury to the cochlea or arcuate eminence during the anterior petrosectomy or to the cochlear nerve at the internal auditory canal. Furthermore, the abducens nerve is vulnerable at its entry to Dorello's canal.

Transcondylar Approach

In the transcondylar approach, identification of the vertebral artery, from its anatomic landmarks, as well as with use of the micro-Doppler intraoperatively, is critical for avoiding inadvertent injury. The vertebral artery is surrounded by a venous plexus and fat, which may provide cues to its proximity after identification of the transverse process of C1 and the suboccipital triangle.

Condylar integrity may be compromised by tumor erosion or intentional removal during exposure. During condylar drilling, the course of the hypoglossal canal should be noted to preserve the hypoglossal nerve. In the setting of greater than 50% condyle erosion or removal, craniocervical stability may be compromised. Upfront craniocervical fusion can be performed upon completion of tumor resection, or a rigid halo or collar may be applied with subsequent formal testing for craniocervical instability before commitment to occipital-cervical fusion. Removal of greater than 50% of the occipital condyles and disruption of the atlantoaxial ligaments should prompt consideration of craniocervical stabilization at the same setting or, in a staged fashion, after formal evaluation of craniocervical stability.

Postoperative Care

Perioperative antibiotics should cover broad-spectrum organisms, especially for anterior approaches or with violation into sinus cavities. Vigilance for potential CSF leak should remain even with careful reconstruction strategies.

Postoperative sinonasal function, including crusting, adhesions, and impaired breathing, should be monitored in craniofacial approaches involving the nasal cavities. Corneal protection with lubricants and possible gold weight insertion may be indicated for postoperative hypoesthesia and/or facial weakness leading to corneal exposure.

Postoperative impaired swallow function may require nasogastric access for nutritional supplementation. Should there be concern for or known violation of the ventral skull base, nasogastric access should be placed under direct visualization or radiographic guidance to prevent inadvertent intracranial transgression. Alternatively, patients may receive total parenteral nutrition until recovery of their swallow function.

Even after radical surgical resection and adjuvant radiation, clival chordomas may exhibit delayed recurrence and merit long-term follow-up.

Adjuvant Treatment

Proton-beam radiation is frequently considered the standard of care following surgical resection of clival chordomas [31]. High-dose radiation, ranging from 70 to 80 Gy, is necessary for efficacious control of chordoma given their relative radioresistance [32]. The close proximity of neurovascular structures abutting the clivus and tumor bed favors the dose delivery profile of proton-beam over photon radiation due to the Bragg peak effect. However, external beam, intensity-modulated radiation therapy, Gamma Knife, and CyberKnife have also been administered as postoperative radiation therapy adjuvants with reported efficacy at local control, although long-term follow-up of tumor and side effects remain to be studied [6, 7].

The molecular profile of chordomas may also impact the prognosis and potential targeted therapeutic strategies. In vitro models and isolated case reports have suggested a role for inhibition of tyrosine kinases, epidermal growth factor receptor (EGFR), signal transducer and activator of transcription 3 (STAT3), vascular endothelial growth factor (VEGF), mammalian target of rapamycin (mTOR), platelet-derived growth factor receptor (PDGFR), and histone deacetylase (HDAC), although the efficacy of targeted therapies remains to be demonstrated in larger clinical trials of chordoma patients [33–35].

Surgical Outcomes

For clival chordomas, 5-year local control is approximately 47–75%, with 81–86% overall survival, as influenced by the histologic subtype, extent of resection or volume of residual tumor, primary or recurrent status of tumor, administration of radiation, and possibly the molecular signature [7, 31, 36, 37]. Longer-term follow-up data are more sparse, with estimates of approximately 70% overall survival and 30–35% progression-free survival at 10 years [7, 38]. Hyperfractionation of adjuvant high-dose proton-beam radiation has been reported to be associated with improved progression-free and overall survival in limited clinical series [31].

PEARLS
Surgical goals for chordoma include gross total resection of microscopic tumor when anatomically feasible or radical subtotal resection to improve the safety margin between tumor and critical neural structures for postoperative radiation when complete removal risks neurovascular compromise.
The choice of surgical approach is dictated by the anatomic compartments invaded by tumor, preparation for reconstructive needs, and minimization of recurrence risk from seeding of the surgical tract.
Chordomas are extradural in origin. Even with possible intradural growth and invasion, the surgical approach should allow for a primarily extradural route for tumor removal, thereby minimizing contamination of CSF spaces unless mandated by the tumor.
The course of the internal carotid artery and position of exiting cranial nerves establish the lateral boundary for anterior midline approaches

Commentary on Case Presentation in Section 21.1

Case Presentation

A 68-year-old man presented with 2 months of double vision and headaches. Examination was notable for a right sixth nerve palsy, with no other focal neurological deficits. Audiogram revealed symmetric and intact hearing bilaterally. Laboratory workup included normal endocrine function. (See Fig. 21.1a–e.)

Discussion

CT scan demonstrates a lytic mass extending from the sphenoid sinus to the lower clivus, with erosion of the inferior sella and posterior clinoid (Fig. 21.1e). Sagittal and axial T1-weighted contrast-enhanced MRI reveals a heterogeneously enhancing mass centered at the upper and mid-clivus, filling the sphenoid sinus, with dorsal intradural extension into the prepontine cistern extending to the left cerebellopontine angle and compressing the brainstem (Fig. 21.1a, b). The T1-hypointense and T2-hyperintense mass demonstrates minimal lateral extension beyond the internal carotid arteries and internal auditory canal, consistent with a clival chordoma (Fig. 21.1b, c). Both petrous apices appear to be involved.

Given the central location of the tumor, with epicenter in the upper and mid-clival region, the lesion lends itself well to a microscopic or endoscopic endonasal approach with associated modifications. In considering approaches that are not directly transnasal, an extended transbasal approach could be pursued for ventral midline access from the sphenoid down to the clivus. This approach may not allow sufficient exposure of the leftward intradural and extradural extension. We would monitor SSEPs, BAERs, and bilateral abducens nerves and the left oculomotor, trochlear, and facial nerve. A lumbar drain would be placed for CSF egress to minimize brain retraction.

We would begin the approach with a coronal exposure and harvest of the pericranium. A bifrontal craniotomy would be taken to the level of the orbital rim, abetted by a diminutive frontal sinus. Removal of the supraorbital bar would be undertaken as a separate step to increase the floor exposure. While separation of the canthal tendons has been described to take the orbital removal down further, it would not be necessary in this case. The first portion may be pursued through an extradural or intradural approach, and we would make an attempt to spare the olfactory tracts during the exposure. CSF drainage is undertaken at this time. The planum sphenoidale and upper clivus are drilled for resection of the tumor from the sphenoid sinus and followed to the right and left. A 30° endoscope could be introduced to inspect and guide microsurgical resection of the cerebellopontine angle tumor extension (Fig. 21.1c).

Following resection, the reconstruction would begin with an intradural and extradural graft repair, and the anterior cranial fossa floor defect would then be bolstered with an abdominal fat graft augmented by fibrin glue followed by onlay of the pedicled pericranium. The frontal bone flap would be plated and hydroxyapatite applied to the crevices for cosmesis.

An alternative left anterior petrosectomy may also be considered to augment the posterolateral exposure and resection if the first exposure was inadequate. Intraoperative imaging can aid in this assessment. This could be done in a single or staged fashion.

He would undergo proton-beam radiation therapy postoperatively.

21.3 Clival Chordomas: Endoscopic Endonasal Approach

Anne-Laure Bernat, Stefano Maria Priola, and Fred Gentili

Introduction

The current standard for the treatment of chordomas includes safe, aggressive, and maximal surgical removal of the tumor, usually followed by high-dose radiation therapy for residual or recurrent lesions [39–44]. A variety of surgical approaches have been described for the treatment of clival chordomas. Among these, the transsphenoidal approach is considered the less invasive and most direct route to the clivus. The introduction of endonasal endoscopic techniques has further reduced the invasiveness of this approach, allowing more detailed visualization of the tumor and, with the use of angled lenses and instrumentation, the ability to address tumors with lateral extension [45–52].

According to long-term follow-up studies, the prognosis is primarily related to the extent of resection [3, 53–55]. Although gross total removal (GTR) is often stated as the desired goal of surgery, in many cases this is not possible. Nevertheless, the selection of the surgical approach is important in attempting to achieve this goal safely [55]. The overall results of the management of chordomas remain disappointing with local control rates (LCR) in adults of only 40–55% at 10 years [39].

Preoperative Evaluation

Preoperative evaluation first requires a detailed review of the neuroradiological imaging. This enables the surgeon to obtain important information, such as the radiologic appearance, location, extensions, and relationship of the clival tumor with surrounding neurovascular structures [56]. The best surgical approach is chosen on the basis of this information.

The clivus can be approached through multiple surgical routes, mainly classified into anterior and posterolateral approaches. The anterior approaches include bifrontal transbasal craniotomy [14, 57], lateral rhinotomy [20, 58], and the transsphenoidal and extended transsphenoidal routes [59–61].

Also included are the transfacial approach [21, 62, 63], transoral, transmaxillary, and transpalatal [64, 65], with anterior cervical decompression and fusion [66, 67]. The lateral approaches include the pterional, transpetrosal [54, 68–71], infratemporal [54, 72–74], and fronto-orbito-zygomatic approaches [75], as well as the retro-sigmoid, far lateral suboccipital [29, 76], and the combined supra-infratentorial presigmoid approaches [77, 78]. In some cases, because of the size and tumor extensions, more than one surgical route used either simultaneously or sequentially may be necessary to achieve maximal tumor resection [44, 75, 79].

The endoscopic endonasal transsphenoidal approach is currently considered as the most direct and safest route to the clivus (Figs. 21.5 and 21.6), with better visualization of affected anatomical structures, allowing for better preservation of neurovascular structures.

Neuroradiological imaging should include high-resolution computed tomography (CT) scan and MRI. CT scan is mostly useful in the assessment of bony anatomy, focusing on possible bony involvement such as erosion and/or osteolysis. Moreover, based on the relationship of the tumor to vascular structures, CT angiography may be necessary to determine arterial displacement, encasement, or anatomical variations such as a medial course of the internal carotid artery [80, 81]. On occasion, conventional angiography may be useful for certain cases in determining whether sacrifice of an encased artery is feasible by defining the patient's tolerance and collateral flow on balloon test occlusion [49, 81].

MRI is still the gold standard, providing more detailed information about the relationship with brain stem, neurovascular structures, and the pattern of cranial nerve displacement or encasement, especially using fast imaging employing steady-state acquisition (FIESTA) sequences [82]. Most chordomas are hypo-isointense on T1-weighted images. T1 hyperintensity, when present, may be related to hemorrhage or mucinous changes within the tumor [83]. T2-weighted images usually demonstrate hyperintensity of the lesion. After injection of gadolinium, the contrast enhancement is usually heterogeneous with a "honeycomb" appearance [80, 84]. While diffusion-weighted MR imaging may be useful in assessing clival tumors, including differentiating chordoma from chondrosarcoma [85], radiological imaging alone is often unreliable in distinguishing between these two entities.

The various neuroimaging studies can be fused with a neuro-navigation system and used for image guidance intraoperatively during the procedure.

One cannot overemphasize the role of careful preoperative evaluation in choosing the best surgical approach and in understanding the tumor relationship with the dura and surrounding neurovascular structures. While chordomas are generally considered extradural lesions, intradural extension is not uncommon and can occur in up to 30% of tumors (Figs. 21.7 and 21.8a–f). Lesions with intradural extension

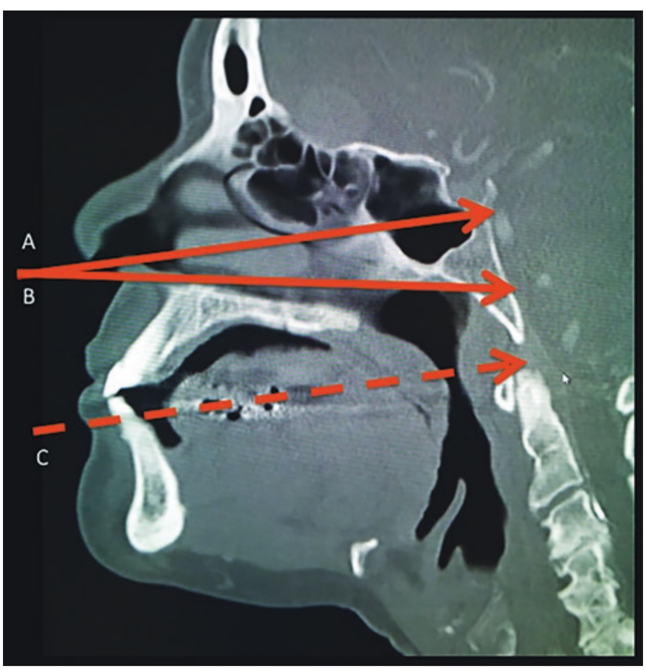

Fig. 21.5 Endoscopic endonasal approach of the clivus. Upper (**a**), middle clivus (**b**), transoral (endoscopic or microscopic) approach that may be combined with an endonasal approach allowing to reach the lower clivus toward the junction C0–C1 (**c**)

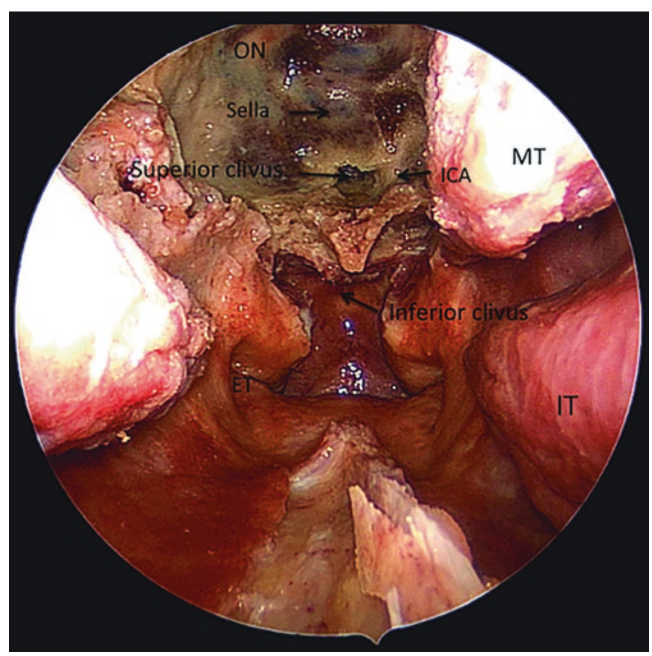

Fig. 21.6 Specimen anatomical view of the clivus with endoscopic endonasal approach. *ON* optic nerve, *ICA* paraclival carotid artery, *MT* middle turbinate (*left*), *IT* inferior turbinate (*left*), *ET* Eustachian tube (*right*)

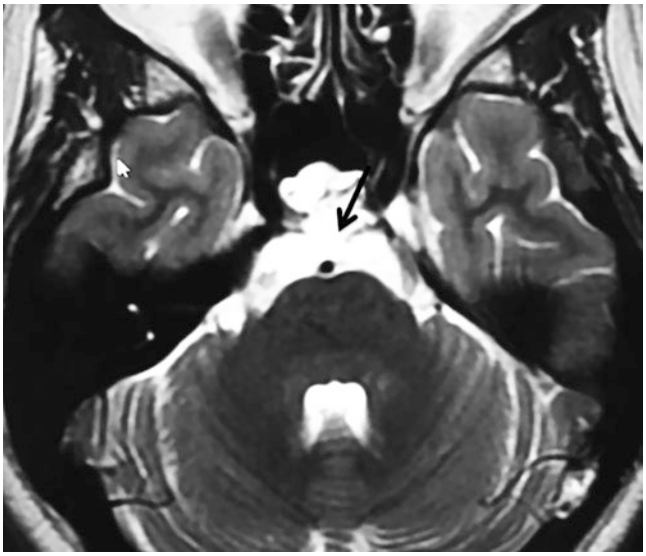

Fig. 21.7 Case of chordoma with intradural extension (*black arrow*) through the upper-middle clivus

are often more difficult to manage, and the surgeon should be aware of any possible intradural extensions before surgery, thus avoiding hazardous traction and manipulation of the tumor during removal.

Surgical Technique of the Endonasal Endoscopic Transclival Approach

Surgery usually involves a team of two surgeons, an otolaryngologist/rhinologist and a neurosurgeon. The otolaryngologist usually performs the initial nasal part of the approach and then drives the endoscope providing a dynamic view of the surgical field during the second stage of the surgery performed by the neurosurgeon.

Different types of endoscopes are used during the procedure, usually starting with a 0° endoscope and if necessary 30°, 45°, and 70° scopes during advanced stages of the procedure for a more lateral view.

Image guidance and neurophysiological monitoring (cranial nerves, brain stem evoked responses, and motor-somatosensory evoked potentials) are used routinely in all cases.

The surgery is performed under normotensive general anesthesia with the patient in the supine position. The head is usually fixed in pins, slightly flexed to improve the view toward the clivus, and the head of bed is elevated 30° to reduce venous bleeding. Lumbar drainage is not used routinely.

Cottonoids soaked in adrenaline 1:1000 are placed in the nasal cavities 10 min before the beginning of the surgery to decongest the nasal mucosa, and the patient is then adequately prepared and draped. With intradural extension of tumor, the thigh is prepped out for possible harvesting of fascia lata and fat used in the reconstruction.

A binostril approach is used, with the endoscope typically positioned in the right superior nasal cavity to allow the operating surgeon to perform a bimanual approach with an instrument in each nostril.

The nasal phase starts with the introduction of the endoscope in the right nostril and the injection of lidocaine 1% with a 1:100,000 dilution of epinephrine at the upper attachment of middle turbinate. This first surgical step includes out-fracture and lateralization of the inferior turbinates. The resection of the right middle turbinate is recommended, but not mandatory, and allows for a wider surgical corridor. Possible alternatives include preservation of one middle turbinate with its lateralization or resection of both middle turbinates [51, 86–88], depending on tumor features and surgeon's preference. These initial surgical steps are completed by mono- or bilateral removal of the uncinate process and ethmoid bulla with the opening of the medial wall of the maxillary sinus, thus increasing the width of the surgical corridor.

The nasal septum is completely exposed, and a pedicled vascularized nasal septal flap is harvested from either the right or left side of the nasal septum based on the anatomy and location of the tumor. The flap consists of the nasal septum mucoperiosteum and is based on the nasoseptal artery, a branch of the posterior septal artery (Hadad-Bassagasteguy flap [HBF]) [89]. A second septal flap may be raised from the contralateral side depending on the need and size of defect. Once harvested, the nasoseptal flap is carefully placed inside the maxillary sinus and not into the pharyngeal space to avoid conflict with the surgical procedure.

At this point, a posterior septostomy is carried out including the maxillary crest, thus obtaining a panoramic view of the sphenoid sinus and nasopharynx mucosa (Fig. 21.9a). This allows a free introduction of instruments from the contralateral nostril improving adequate surgical maneuverability. From this point on, a bimanual binostril technique is routinely utilized.

The sphenoid sinus is opened widely, and the floor is drilled down to the level of the clival recess and nasopharynx in the coronal plane allowing low exposure and bimanual manipulation. The degree of drilling will depend on the pneumatization of the sphenoid sinus. In the presellar or conchal type, significantly more drilling is required (Fig. 21.8b). In that case, image guidance is critical to identify the location of the paraclival carotid arteries. By drilling the lateral recess of the sphenoid laterally to the medial aspect of the paraclival carotids, the surgeon obtains a wide window that includes the floor of the sella, the two paraclival carotid protuberances (Fig. 21.9a, c), and the upper third of the clivus including both medial and lateral optico-carotid recesses, parasellar carotids, and floor of the sella (Fig. 21.9a).

In cases with superior extension of the tumor to the dorsum sella behind the pituitary gland, the bone over the

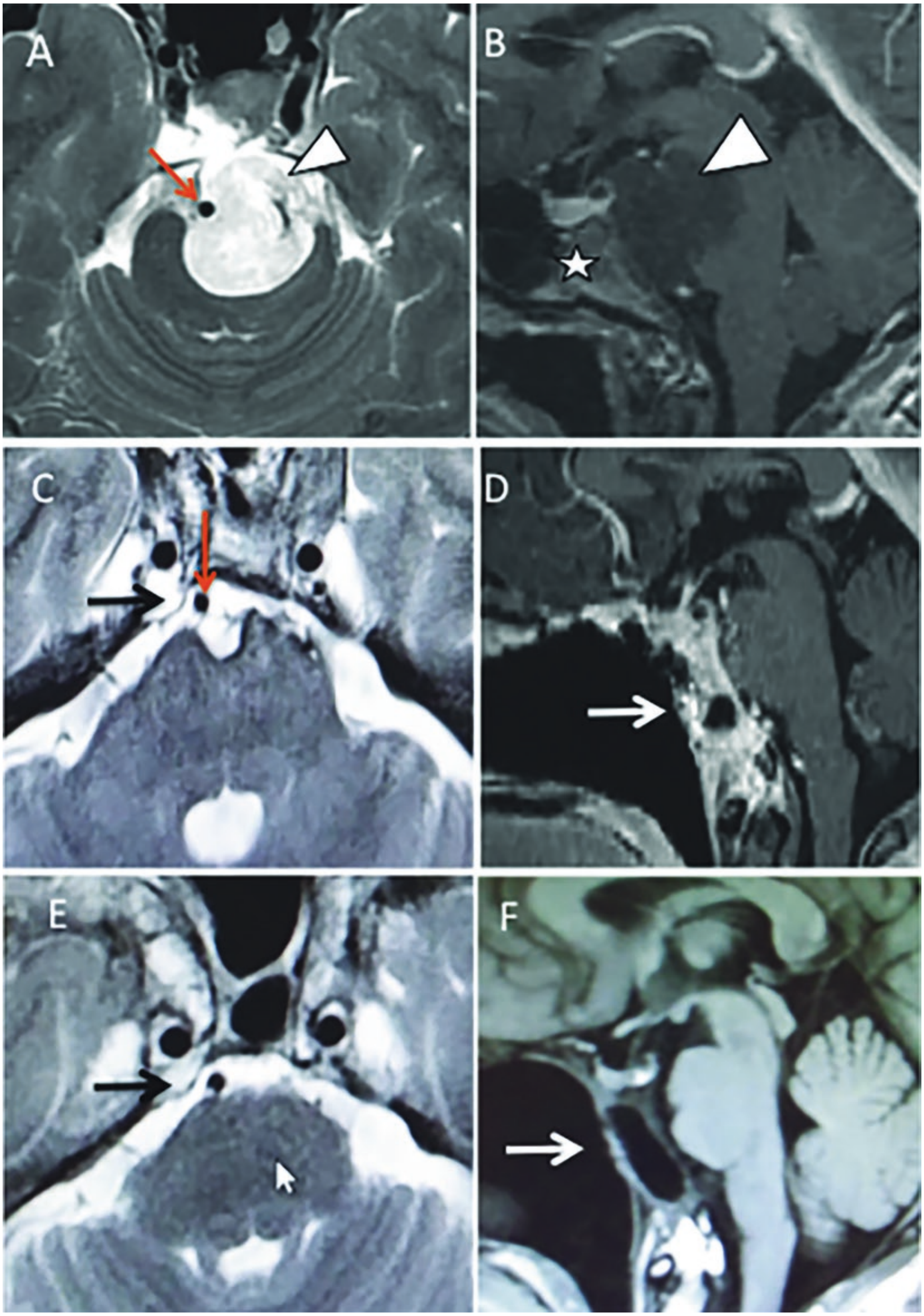

Fig. 21.8 (**a**–**f**) Case of intradural clival chordoma. (**a**) Axial T2. Lesion extended to the right paraclival ICA (*white arrow head*), basilar artery deviated to the right (*red arrow*). (**b**) Sagittal T1 post-contrast. Upper-middle clival chordoma (*white arrow head*), presellar pneumatization sphenoid sinus type (*white star*) requiring an extensive drilling. (**c**) Post-op axial T2. Subtotal resection. Small residual behind the right paraclival ICA (*black arrow*). (**d**) Post-op sagittal T1. Multilayer reconstruction (*white arrow*). (**e**, **f**) Three years after surgery and irradiation. Axial T2: slight shrinkage of the residual (*black arrow*). Sagittal post-contrast: multilayer reconstruction flap after healing (*white arrow*)

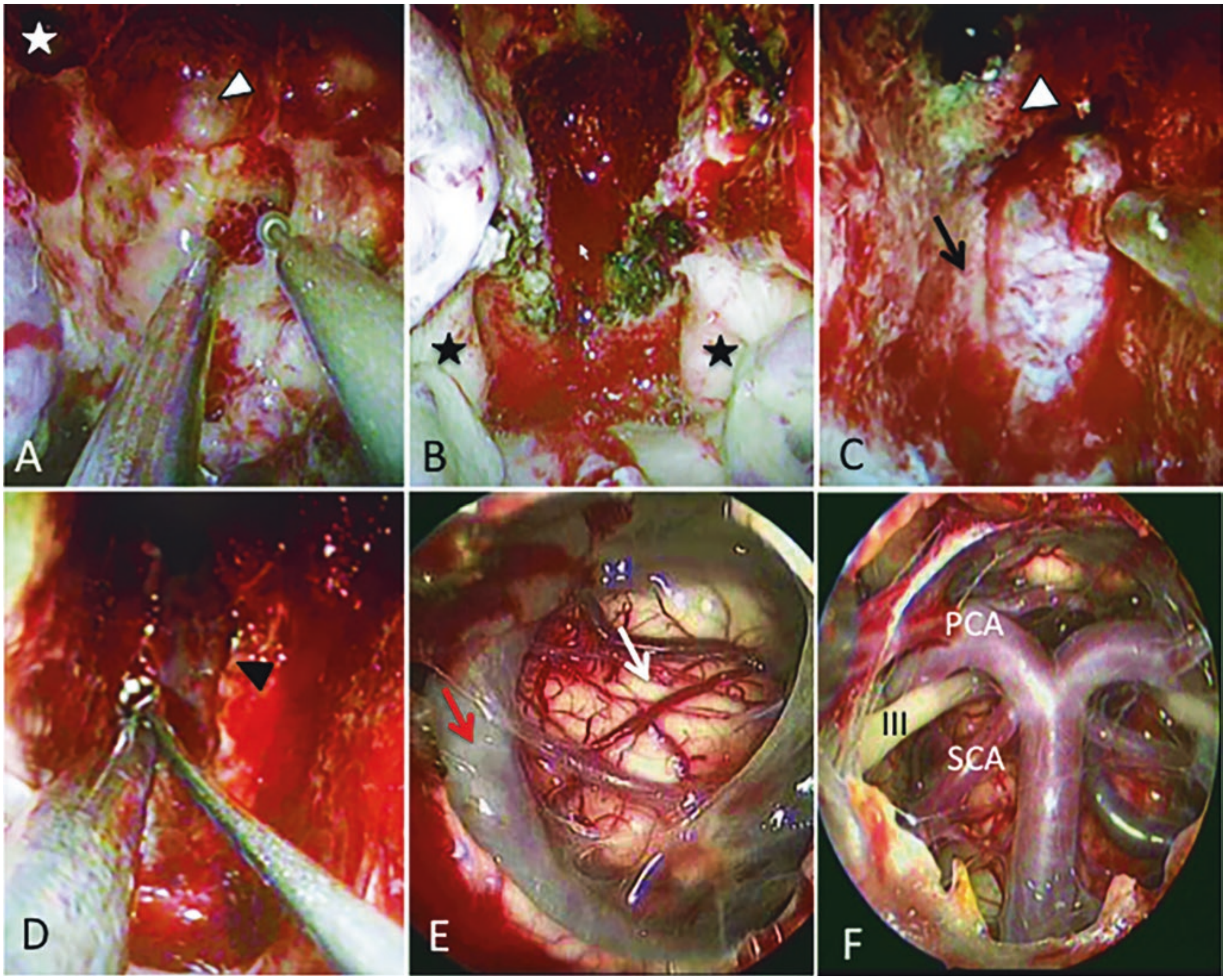

Fig. 21.9 (**a**–**f**) Perioperative pictures from the case reported. (**a**) Extensive drilling of the clival bone. Right optico-carotid recess (*white star*) and floor of the sella (*white arrow head*). (**b**) Exposure of the clivus, sellar floor above, vomer, and nasopharynx below with Eustachian tubes laterally (*black stars*). (**c**) Exposure of the clival dura and right paraclival artery (*black arrow*). (**d**) Debulking of the tumor after opening of the dura (black arrow head). (**e**) After resection, basilar artery view (*red arrow*) and brain stem (*white arrow*). (**f**) Basilar tip with emergence of left and right oculomotor nerves (III) between posterior cerebral and posterior communicating arteries

sella should be removed, and the pituitary gland can be elevated creating a corridor between the pituitary, sellar floor, and cavernous sinus allowing removal of the dorsum sella and posterior clinoid processes [90].

Exposure of the middle and inferior third of the clivus is achieved by a midline incision (with U- or I-shaped fashion) of the nasopharynx mucosa, and the underlying pre-clival fascia and the longus capitis muscles are reflected inferiorly and laterally. The pre-clival soft tissues can be very adherent to the clival bone and can be taken down with cautery.

A key point during this step is to maintain a midline route between the Eustachian tubes (Fig. 21.9b), particularly when using cautery, as the ICAs run just lateral and posterior. Evaluation of the preoperative images allows one to monitor for an excessively medial course of the ICAs [87, 90–92].

The drilling of the clivus is then completed using various-sized high-speed drills and finally with a coarse diamond hybrid burr. Depending on the extent of involvement of the bone by the tumor, this step may be associated with extensive bleeding from the clival venous plexus usually easily controlled by hemostatic agents and gentle pressure. Fortunately, the clival venous plexus is often already thrombosed by compression or invasion by the tumor.

In selected cases, a combined transsphenoidal and transoral approach can be utilized if the tumor extends inferior to the craniocervical junction and the second cervical vertebra. The transoral route may also be indicated for extradural lesions of the inferior clivus that are confined to the midline, protrude into the posterior pharyngeal region, and extend to C1–C2 region. Extensive osteotomy, hard palate resection, or soft palate splitting is not usually required [40, 86, 93].

At this point, the tumor is often visible, especially if the clivus is eroded anteriorly. Rarely, the lesion is not visible, so its location is confirmed with intraoperative navigation.

The tumor is then removed using the classic microsurgical principles, such as intracapsular debulking, followed by extracapsular sharp dissection when a capsule is present (Fig. 21.9d).

As previously noted, it is not unusual for the tumor to have eroded the dura with an intracranial extension. In these cases, the role of image guidance and micro-Doppler becomes crucial in order to locate the course of internal carotid arteries, basilar artery, and vertebrobasilar junction. This is also useful in order to identify the course of sixth cranial nerve before the dural opening, as it usually has a close relationship with the vertebrobasilar junction [94, 95].

The removal of the intradural portion of the tumor should be undertaken with care when using suction or pituitary forceps given the close relationship of the tumor with the cranial nerves, brain stem, and perforating vessels (Fig. 21.9e, f). We do not recommend the use of an ultrasonic aspirator. While the identification and careful dissection of arachnoid planes when present aids a safe resection, chordomas can infiltrate both neural and vascular structures.

Chordomas are usually soft and easily removable by suction. They often do not have a true capsule and extracapsular dissection is not possible. However, some tumors are more fibrous, often the case in some recurrent tumors that have undergone multiple surgeries and/or after radiation. In these cases, surgery may be more challenging because of nerve and/or vascular encasement. Nerves may then be encased in the tumor pseudo-capsule and not easily dissected. In these cases, it is safer to keep working carefully within the tumor until one is able to identify these critical structures. When nerves are encased by the pseudo-capsule, it is rarely possible to dissect them free without serious risk of damage. In these cases, it is better to leave the pseudo-capsule and adherent neurovascular structures.

When a sixth nerve palsy exists prior to surgery, which is not uncommon [96], it may be due to compression at the cisternal segment of the nerve (often a partial palsy) (Fig. 21.10a)

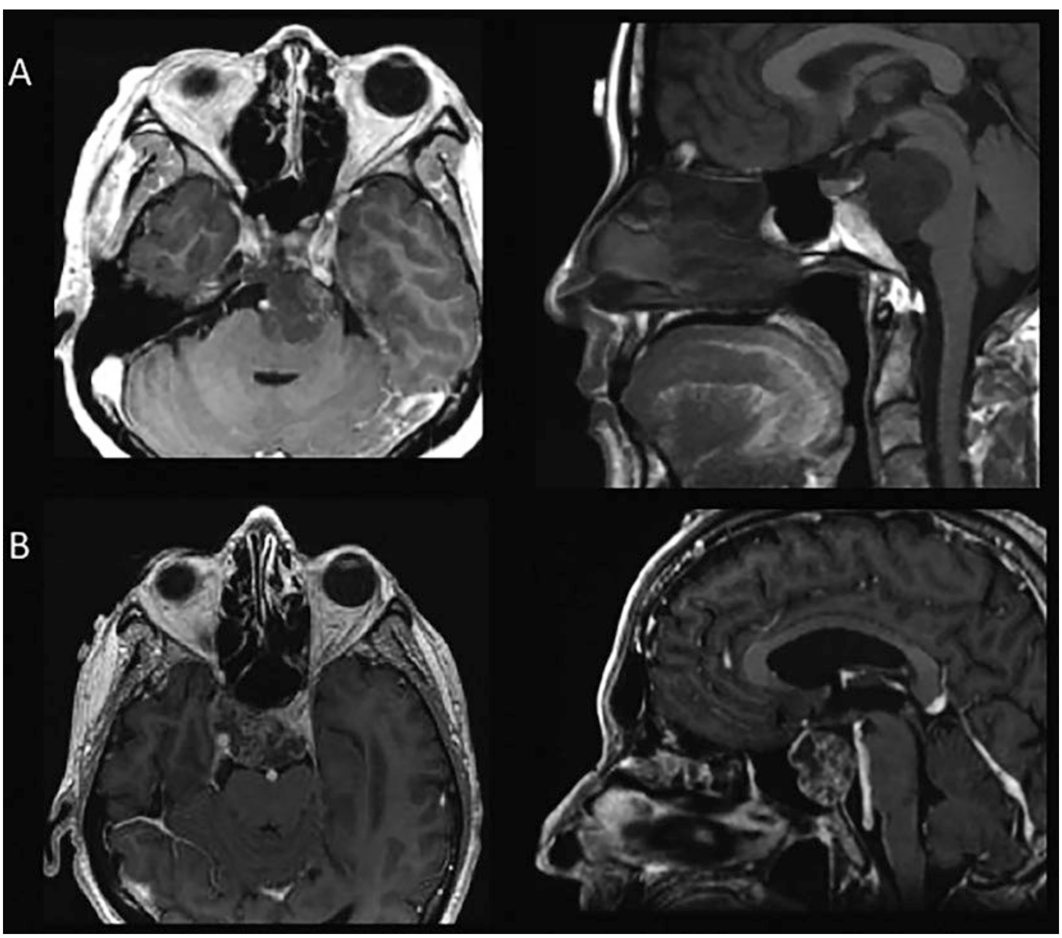

Fig. 21.10 (**a**, **b**) Cases of upper and middle clival chordoma presenting with sixth nerve palsy. (**a**) Compression of left sixth nerve palsy in its cisternal segment, partially improved after resection. (**b**) Encasement of right sixth nerve palsy in its intraosseous and intradural segments with permanent complete palsy

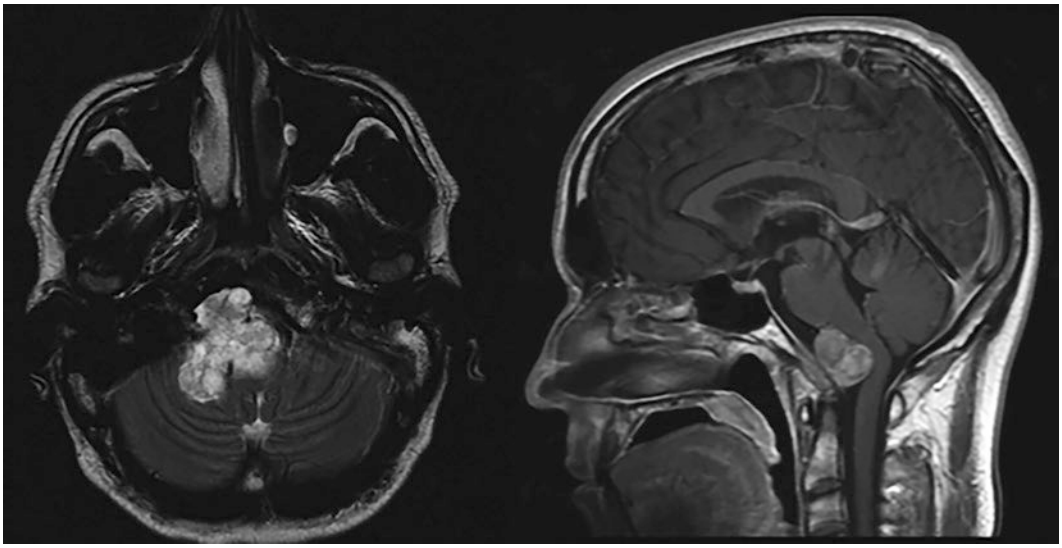

Fig. 21.11 Case of lower clival chordoma presenting with right abducens palsy

or by encasement, especially in case of infiltration of the petrous apex bone and/or Dorello's canal (intradural segment) (Fig. 21.10b). Surgery may result in improvement if the mechanism is compression, but is often not the case when it is related to infiltration of the intraosseous and/or dural segment of the nerve. Temporary sixth nerve palsy or deterioration of pre-existing palsy is a frequent complication of the resection of a clival chordoma because it is the most vulnerable nerve and even gentle manipulation may injure it. Chordomas of the lower clivus may involve multiple lower cranial nerves (Fig. 21.11). On occasion, tracheostomy may be necessary before resection in cases with pre-existing swallowing difficulties.

It is important to avoid a bilateral sixth or lower cranial nerve palsy that may lead to serious clinical consequences for the patient.

When ICA encasement is present, especially in case of recurrent and/or irradiated tumor, it is preferable to perform an occlusion test before surgery in order to know the therapeutic options in case of injury and for subsequent surgical planning.

The surgeon should adapt his/her technique and surgical goal to the clinical situation with the understanding that the prognosis is correlated to the degree of resection.

The removal of tumors with more lateral extensions that spread posterior to the paraclival and petrous portion of ICA is more problematic. In these cases, it is important to skeletonize the ICAs, with a gentle lateralization and careful removal of the tumor using angled endoscopes, angled dissecting tools, and curettes [97]. However, a significant lateral extension is a contraindication for the use of the endonasal transclival approach alone.

Reconstruction Technique

Reconstruction of the skull base defect is an important step in all the endoscopic approaches to the skull base. In extended procedures with a significant dural opening, meticulous closure techniques are crucial to prevent postoperative CSF leakage. A meticulous attempt to recreate the natural resistance offered by the arachnoid and dura mater represents key steps in the repair.

A number of reconstructive techniques have been described in the literature [96, 97]. We use a multilayer reconstruction technique preferably with biological tissue (fascia lata, fat) (Fig. 21.12). The first layer is a piece of DuraGen™ (DuraGen Plus™ Adhesion Barrier Matrix, Integra LifeSciences Corporation, Plainsboro, NJ, USA) to try to reconstitute an arachnoid layer. Next, a piece of fascia lata is placed as an intradural inlay graft slightly larger than the defect. This is followed by a second piece of fascia lata applied as an onlay graft covering the entire bony defect. Next the previously harvested vascularized nasoseptal flap is then placed over the onlay fascia lata graft. Careful attention is paid to make sure the flap is firmly in contact with bone edges around the defect. The flap is then framed with Surgicel® (Ethicon, Inc., Somerville, NJ, USA) to create a matrix, and this is then covered with dural sealant.

The closure is completed by the application of a piece of autologous fat that temporarily wedges the contour of the flap, keeping it in the appropriate position, while the reconstruction is completed by placing gelatin foam pieces (Gelfoam®, Upjohn Co., Kalamazoo, MI, USA) followed by a balloon stent (Foley n°14) which is usually kept in for 3–4 days. An alternative to a Foley catheter is packing with Vaseline gauze.

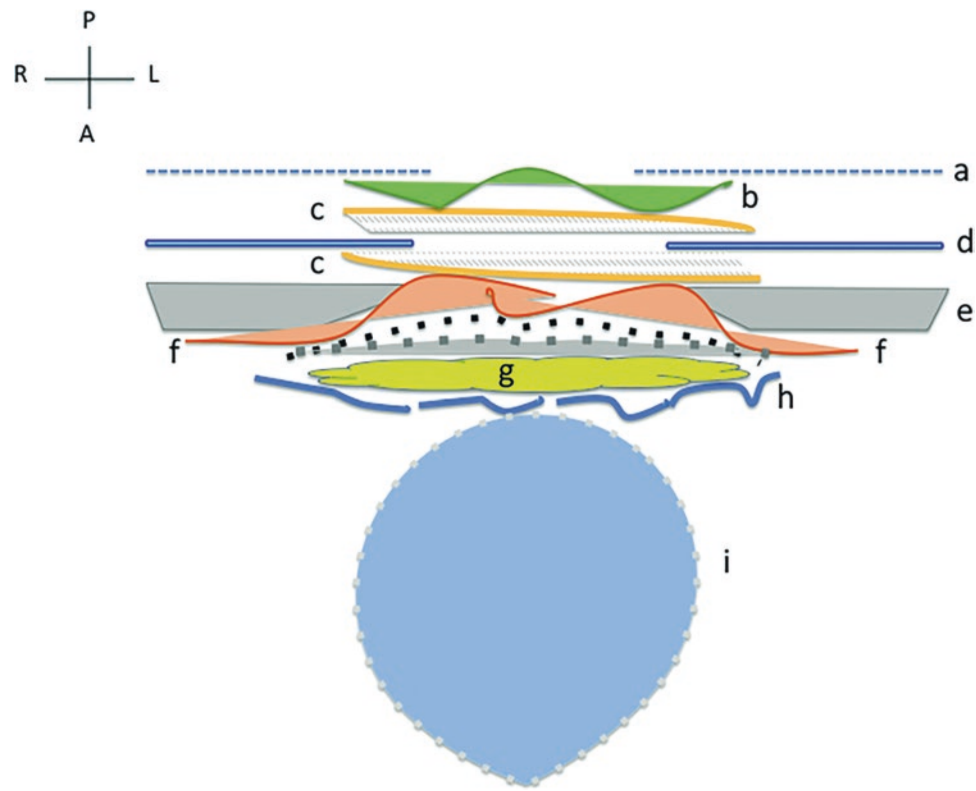

Fig. 21.12 Multilayer reconstruction technique. (**a**) arachnoid; (**b**) DuraGen flap; (**c**) fascia lata (inlay and onlay flaps); (**d**) dura mater; (**e**) clival bone; (**f**) nasoseptal flaps; *black dotted line*: Surgicel® (Ethicon, Inc., Somerville, NJ, USA); *gray dotted line*: dural sealant; (**g**) autologous fat; (**h**) gelatin foam pieces (Gelfoam®, Upjohn Co., Kalamazoo, MI, USA); (**i**) balloon stent (Foley n°14)

A lumbar drain is not routinely used. In cases where the reconstruction is not felt to be entirely satisfactory with a high likelihood of CSF leak, a lumbar drain is placed and typically drained at 5–10 ml/h for 2–3 days.

Other closure techniques have been described including the "Gasket-seal" watertight closure by Leng et al. [98] and the "extradural (overlay) technique" by Cavallo et al. [99]. The former is characterized by the application of a piece of autologous fat on the bone defect, followed by the positioning of an autologous piece of fascia lata harvested and fashioned to be larger than the cranial base defect. Then, a piece of rigid material—such as vomer, titanium plate, or Porex—is fashioned in order to fit with the bone defect and gently countersunk into the bone defect.

In the latter technique, a large piece of lyophilized human pericardium, exceeding the size of the osseous defect, is placed over the defect, is cut slightly larger than the bone defect, and is wedged into the extradural space, holding the dural substitute in the overlay position. A nasogastric tube is lastly inserted allowing a better healing of the nasopharyngeal mucosa.

Postoperative Care

At the end of the procedure, the patient is transferred to a critical care bed with close observation for at least 48 h. Wide-spectrum antibiotics are given 30–60 min before surgery and should be continued postoperatively for 2–3 days or as long as the nasal balloon catheter or packing is in place.

Our protocol for postoperative adjuvant treatment of clival chordomas is conformal intensity-modulated radiation therapy, usually scheduled between 2 and 6 weeks after surgery [100, 101].

Complications

The extended endoscopic endonasal transclival approach requires an experienced and well-trained team [102]. Strict observance of basic neurosurgical principles, detailed knowledge of endonasal anatomy, and meticulous dissection of neurovascular structures are required in order to reduce the incidence of complications.

According to the time of appearance, we classify the complications as immediate or delayed.

Immediate (Occurring During Surgery)

Injury of Major Vessels (ICA, Basilar Artery, Vertebral Artery, Perforators) or Venous Sinuses

There is an increased risk of vascular injury in tumors with significant infiltration of the bone, with subsequent loss of anatomical landmarks [103], or with blind traction during the removal of the intradural portion of the tumor. The use of the image guidance and Doppler probe will reduce the incidence of these complications [104].

Management of a major vascular rupture is immediate compression and the use of hemostatic tissue and muscle if immediately available, followed by an immediate endovascular assessment and treatment if necessary including stenting or occlusion of the vessel [103, 105].

Injury of the Brain Stem or Cranial Nerves (Especially the Sixth Cranial Nerve)

Although large clival chordomas can involve several cranial nerves, specific consideration should be given to the sixth cranial nerve, which is at particular risk during the endoscopic endonasal transsphenoidal transclival approach.

Usually the sixth cranial nerve arises at vertebrobasilar junction and then runs superolaterally in the prepontine cistern. It then enters the petroclival venous confluence under the posterior petroclinoidal dural fold and travels through Dorello's canal under the petrosphenoidal (Gruber's) ligament into the cavernous sinus [94, 106].

It can be damaged as it exits Dorello's canal coursing toward the cavernous sinus especially during the drilling of the clivus or during tumor resection.

Knowledge of its anatomical course and careful review of neuroimaging are crucial points in order to preserve its integrity.

Another important factor is the opening of the dura, which when needed should be done in the midline above the level of the vertebrobasilar junction at the origin of the nerve (in 80% of cases behind the AICA origin) [107].

Delayed

CSF Leak [61, 108–112]

While CSF leaks can occur intraoperatively, the incidence of postoperative CSF leakage after endoscopic resection of clival chordoma ranges between 6% and 25% [46, 87, 113–115]. The incidence is in part related to the extent of resection during surgery, the size of the resultant defect, and the quality of the reconstruction. The introduction of the vascularized pedicled nasoseptal flap has dramatically reduced the incidence of postoperative CSF leaks. The reconstructive steps should be meticulous, and placement of an external lumbar drain (ELD) should be considered at the end of surgical procedure if there is concern regarding the integrity of the closure. Likewise, if the leak occurs immediately in the postoperative period and is minor without significant pneumocephalus, an ELD can also be placed for 3–4 days. Surgical repair should be considered in cases with persistent CSF leakage despite the ELD insertion or in large leaks with pneumocephalus. Biological tissue with abdominal or thigh fat and fascia lata is recommended. If the original nasoseptal flap is not viable, the use of an inferior turbinate flap or the posteriorly based lateral nasal wall flap is the other intranasal options available [115].

In cases of persistent/recurrent CSF leakage, where no other intranasal options are available especially after multiple previous surgeries and irradiation, a tunnelled vascularized temporoparietal fascial or frontal pericranial flap [116, 117], the Oliver-modified palatal flap, and the occipital galeo-pericranial flap can be used [115].

One should keep in mind and allow for the fact that the size of the flap at harvest has the potential for retraction over time (up to 20%) [115].

Bacterial Meningitis [108, 118, 119]

The incidence of bacterial meningitis ranges from 0.5% to 14% in extended transsphenoidal approaches. It is usually related to CSF leak in cases where there is a large communication between the intradural space and the nasal cavities. This complication can be avoided if a watertight closure of the skull base defect is performed. The failure of an adequate reconstruction with CSF leak and meningitis remains a major concern in expanded endoscopic techniques and can compromise surgical outcome.

Craniocervical Instability [120]

This complication is seen in large lesions involving the lower portion of the clivus and often related to extensive drilling of the occipital condyles. It can also be related to bone invasion of these structures by lesions with lateral extension. A second surgical procedure, often a posterior fixation, may be required based on documented instability. This complication can be anticipated and planned prior to surgery.

Hypopituitarism (Temporary or Permanent)

These complications are more frequent in cases with sellar invasion with involvement of the pituitary gland and especially in cases where pituitary transposition is carried out [90].

Nasal Complications

While minor nasal issues including discomfort are common, some uncommon nasal complications include hearing loss (often related to Eustachian tube injury), anosmia, and dysgeusia. Careful postoperative assessment, follow-up, and treatment by the rhinologist are important to manage these issues.

Metastasis

Although rare, local recurrence and seeding along the surgical route (1–7%) [121–124], ventricular space [123], and neural axis [125, 126] have been reported.

Surgical Outcomes

The extent and quality of resection is generally felt to influence the surgical prognosis in patients with chordomas [96, 127]. Bresson et al. [96] reported a mortality of 20.5% and recurrence rate of 28% after GTR compared to 52.5% and 47.5% after subtotal resection.

Some authors have reported better surgical results following the endoscopic approach in terms of the degree of resection with GTR rates of 55–83% vs 40% for open procedures. Likewise, lower rates of complications such as cranial nerve palsy and CNS infection were reported [127–129], when compared to classic transcranial approaches.

The overall survival rate at 5 years was close to 70% and 60% at 10 years [96, 130, 131]. The improved outcomes with the endoscopic approach were attributed to the more direct midline access to the clivus when compared to the antero- or posterolateral transcranial approaches. Nevertheless, the learning curve had a significant impact on GTR, with increasing rates in more recent reports of 89% by Koutourousiou et al. [132].

The most frequent complication, significantly more frequent than with transcranial approaches, was cerebrospinal fluid leakage with up to 20% reported by some authors [79, 132]. With increasing experience and the development of better reconstructive techniques, this complication has decreased to 6–10% in more recent reports [47, 133, 134].

Neurological complications, especially of the sixth or other cranial nerves, including new cranial neuropathies occurred in 6.7%. These were more frequent especially if the neuropathy existed preoperatively and were seen in up to 60% for sixth cranial nerve [79]. Nevertheless, this complication is less frequently reported than with transcranial posterolateral approaches where it reaches 34% [135].

Fortunately, ICA injury is a rare complication reported in less than 2% of cases [92, 136, 137] and seen more often in recurrent lesions which had been radiated or with cavernous sinus and/or petrous ICA invasion. The most frequent site of arterial injury was the upper or middle third of the clivus.

GTR and long-term control remain very difficult to achieve in chordomas regardless of the route or the radiation modality used postoperatively [6]. For lesions with significant lateral or inferior extension, a combined endoscopic and transcranial approach in two or more stages can be done to allow maximal safe resection.

The best surgical outcomes for the endoscopic approach have been observed in patients presenting with a midline tumor where a nasoseptal flap had been included in the reconstruction [79]. Significant intradural invasion is a limiting factor for GTR especially in recurrent lesions.

Likewise, chordomas with an exophytic growth pattern are more difficult to achieve a GTR and tend to recur more often than those with an endophytic growth pattern. Although the endoscopic approach may allow for higher GTR rates and less neurological morbidity than traditional transcranial routes, their impact on long-term survival and disease control remains unknown.

Recurrences within the surgical pathway by seeding were reported in 2.8% in non-endoscopic approaches [138] and in 1.3% for the endoscopic approach [122].

Safe, function-sparing surgery with GTR followed by radiotherapy seems to be the optimal management of these tumors [44, 127, 139]. Although proton beam has been the traditional modality of radiation given after GTR [96], no definitive superiority of proton versus photon or other modalities has been demonstrated [6]. The literature would suggest [39, 140] that radiation therapy improves the efficacy of surgery, especially if associated with maximal total resection. Partial resection is strongly associated with poor progression-free survival (PFS).

Management After Surgical Resection

Postoperative Studies

The routine first follow-up MRI is done between 3 and 6 weeks after surgery unless there are clinical indications for an earlier scan. In very early post-op scans, there may be many postoperative changes that make accurate analysis of any residual tumor difficult. Subsequent MRI assessment can be based on the surgeon's impression, pathological analysis, and postoperative radiation plan [79].

While there is no consensus, in cases of documented residual disease, in a resectable location, a second procedure can be considered in an attempt to achieve a more complete resection. Regardless of the degree of resection, but especially with documented residual disease, postoperative irradiation is recommended.

Posterior Fixation

A posterior fixation might be required in cases of cervicospinal instability from tumor involvement or extensive drilling of the condyles and when the patient is symptomatic. This risk is increased after a combined anterior endoscopic and posterior open approach [120].

Radiation Therapy

A variety of radiation modalities have been used and demonstrated efficacy in chordomas. Among these, proton radiation therapy and combined proton/photon radiation therapy have been the standard therapy recommended in the literature. Whether photon or proton beam therapy or other radiation modalities such as carbon ion are used, it depends on the experience and individual practice of the local radiation oncologist and discussion at a multidisciplinary team conference. Radiation is usually performed within 3 months after surgery.

The recent literature [141, 142] attests to its efficacy, although radiation toxicity remains high given the high dose (70 Gy) administered for this tumor.

The impact of radiation toxicity on surrounding structures in patients with chordoma is important because the organs at risk include the pituitary gland, with the risk of complete or partial pituitary insufficiency; the optic nerves and cavernous sinus, with the possibility of oculomotor or other cranial nerve palsy; and medial temporal lobe toxicity with the potential for seizures.

While a recent review of the literature by Holliday et al. [143] would suggest an advantage of proton therapy compared to other radiation modalities, the latest consensus on the management of chordoma [144] confirms both proton and photon radiation therapy in high doses as standard treatment (recommendation A, level of evidence V). Other studies and trials are still ongoing to test the efficacy and feasibility of carbon ion therapy compared to other forms of radiation [145, 146].

Management of Recurrence

Surgery

Surgery is usually required for recurrent tumor in cases of surgically accessible lesions with acceptable risk and when the condition of the patient warrants intervention. Some cases, with very aggressive tumors with early and frequent recurrences, often require multiple surgical interventions [147]. The cervical location is often associated with more frequent recurrences than other locations, especially those limited to the clivus.

Radiation Therapy

There is no consensus in the literature regarding the role of repeat radiation therapy in recurrent chordomas, although a number of recent papers have discussed the role of a second-line radiation therapy for recurrent chordomas. McDonald et al. [148] reported on repeat proton radiation therapy for recurrent chordomas, with a 2-year estimate chordoma-specific survival at 88% and a 2-year estimate of local control (LC) at 85%, without significant toxicity. This review demonstrates better results, in terms of progression-free survival (PFS) and overall survival (OS), for recurrent chordomas when treated by repeated surgery and second-line radiation therapy than surgery alone [141, 148].

Palliative Management

Unfortunately, no treatment has a proven long-term efficacy in chordomas. The recent reports looking at molecular markers in chordomas are of significant interest and suggest a potential in the future for targeted therapy of this tumor. A multidisciplinary, multimodality management is critical and offers the patient the best chance for prolonged survival along with an acceptable quality of life [149].

PEARLS
Regardless of the approach used for chordomas, initial adequate exposure of the lesion is critical for safe maximal resection of the lesion.
Patients with chordomas can have long-term functional survival such that quality of life issues must be taken into consideration when attempting aggressive resections with major risks to critical neurovascular structures.
Intimate knowledge of the vascular, neural, and bony anatomy involved by the chordoma is important and aids cranial nerve preservation especially the sixth cranial nerve during the drilling of the clivus or during tumor resection.
Expertise in both open approaches and combined open and endoscopic techniques is important in dealing with the varied locations of chordomas.

Commentary on Case Presentation in Section 21.1

Case Presentation

A 68-year-old man presents with 2 months of double vision and headaches. Examination shows right sixth nerve palsy, but otherwise neurologically intact. Laboratory workup: normal endocrine function (see Fig. 21.1a–e).

Discussion

This patient presents with the classical presentation and radiological features of a clival chordoma. The MRI shows the lesion extends both anteriorly into the sphenoid sinus and posteriorly where it appears to have gone intradurally adjacent to the basilar artery with extension along or into the brain stem. As to the optimal operative approach for this lesion, the expanded transclival endoscopic endonasal approach offers a number of significant advantages.

These include the fact that it is the most direct approach to the lesion with the least disruption of normal anatomy. In addition, the endonasal approach would allow access to both the sphenoid sinus component and the clival and intradural component without transgressing any cranial nerves or vascular structures. This approach would also allow for the removal of the component involving the brain stem. While a total removal is unlikely by any approach, the endoscopic approach would allow for the safest maximal removal of this lesion. Careful reconstruction is necessary and would require a multilayered closure with fascia lata, fat, and a vascularized nasoseptal flap.

21.4 Clival Chordomas: Editors' Commentary

Clival chordomas represent a very challenging pathology due to their difficult to access location in the cranial base, frequent involvement of critical neurovascular structures, as well as infiltration of the bone and potentially dura with intradural extension. These characteristics make complete removal of chordomas difficult and contribute to the high recurrence rate despite aggressive surgery with adjuvant radiotherapy. The general premises of surgical resection of chordomas are maximal safe resection, accurate tissue diagnosis, relief of tumor compression of the brainstem and cranial nerves, and to provide a safer distance between the tumor and normal neural tissue for radiation when gross total resection cannot be achieved. Tumor recurrence and repeated surgical interventions can create substantial morbidity and potential mortality. Outcomes, therefore, are clearly related to the degree of tumor resection coupled with postoperative high-dose radiation. The current standard management begins with a maximal safe resection of the chordoma by one or more surgical approaches.

The clivus can be accessed through multiple surgical routes. Traditional open transcranial approaches have been the workhorse of chordoma resection for several decades; however, they may be associated with significant approach-related morbidity, CSF leakage, and tumor seeding along the approach corridor. The open transcranial routes can be divided into anterior, lateral, and posterolateral approaches. Anterior transcranial approaches encompass the extended subfrontal, transbasal, and transfacial approaches (i.e., transmaxillary, transpalatal, and transoral). The lateral approaches include the pterional, orbitozygomatic, subtemporal anterior petrosectomy, and subtemporal infratemporal fossa approaches. Posterolateral approaches encompass the posterior transpetrosal and far lateral approaches with various degrees of occipital condyle removal.

The clear advantage of the anterior approaches is direct access to the tumor origin within the clivus without the need to transgress cranial nerves or neurovascular structures. Although effective, many of the open anterior approaches have more recently been replaced by endoscopic transnasal procedures which may reduce much of the approach-related morbidity and actually provide better visualization for tumor resection. Improved outcomes reported with endonasal approaches to midline chordomas are likely related to the more direct access to the clivus with less disruption of uninvolved anatomy compared to the open anterior, lateral, or posterolateral approaches. A particular advantage of the endonasal approach is in the case of recurrent extradural chordomas in the region of the clivus. The endonasal corridor lends itself very well to repeated surgery without the need for external incisions or disruption of extensive areas of normal anatomy. Limitations include the ability to adequately address tumor that extends lateral to these the cranial nerves and internal carotid arteries, unless the tumor has provided a corridor, and particularly when there is a recurrent chordoma that is mainly located laterally. Despite advances in expanded endoscopic techniques, the lateral and posterolateral open cranial base approaches have not been supplanted. At most centers, patients with mainly laterally located chordomas are managed with the lateral or posterolateral approaches alone or in staged combination with the open or endoscopic anterior approaches when there is a midline tumor component.

In the case example (Fig. 21.1a–e), a 68-year-old male presents with headaches and 2 months of diplopia secondary to a right sixth nerve palsy. The imaging reveals a chordoma with its epicenter in the clivus. Tumor extension anteriorly into the sphenoid sinus with the long axis of the tumor largely in an anterior-posterior orientation lends itself well to an anterior approach. The lack of involvement of any structures anterior to the sphenoid sinus would render the open anterior approaches rather destructive to normal and uninvolved anatomy. Thus, the case would favor an endoscopic endonasal approach. The lateral extension of the tumor dorsal to the petrous carotid artery could represent a limitation of this approach; however, expanded techniques as well as angled endoscopes and instruments may largely overcome this issue. Also noted is evidence of intradural extension of the tumor. The endonasal approach has initially carried a higher risk of postoperative CSF leakage, but improved repair techniques and surgeon experience have diminished the risk of postoperative CSF leak such that rates are comparable to those of the open approaches with the literature demonstrating continuous improvement. Skull base reconstruction in this case would certainly require a synthetic or autologous primary dural repair and a vascularized nasal septal flap to reduce the risk of CSF leakage and provide a robust barrier that can withstand mandatory adjuvant high-dose radiation postoperatively.

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22 Colloid Cysts

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Contents

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22.1 Colloid Cysts: Editors' Introduction

Colloid cysts are benign intraventricular lesions that originate from the roof of the third ventricle. They are thought to be developmental cysts and constitute up to 2% of intracranial neoplasms. The cyst is thought to arise from the endodermal cells within the velum interpositum, with an attachment point dorsal to the foramen of Monro. Symptoms associated with this lesion may occur at any point but most often occur in the fourth decade and are characterized by positional headaches. Sudden death can rarely occur secondary to acute obstruction of cerebrospinal fluid outflow [1]. Colloid cysts are typically T1 hyperintense and T2 hypointense on MRI and may occasionally exhibit rim enhancement. Pathologically, the cystic fluid is described as turbid and viscous. Histological findings include a fibrous wall composed of simple columnar epithelium resembling bronchial epithelium, which stains for keratin and EMA.

Colloid cysts are often found incidentally and can be managed conservatively if the cyst is small, asymptomatic, and without hydrocephalus. Surgical intervention is the mainstay of treatment for symptomatic lesions [2]. Treatment goals range from fenestration and drainage to complete surgical resection, and a variety of surgical approaches have been described for colloid cyst resection. In this chapter, the open and endoscopic approaches for colloid cysts will be discussed by the authors including their management of the case example below (Fig. 22.1a, b).

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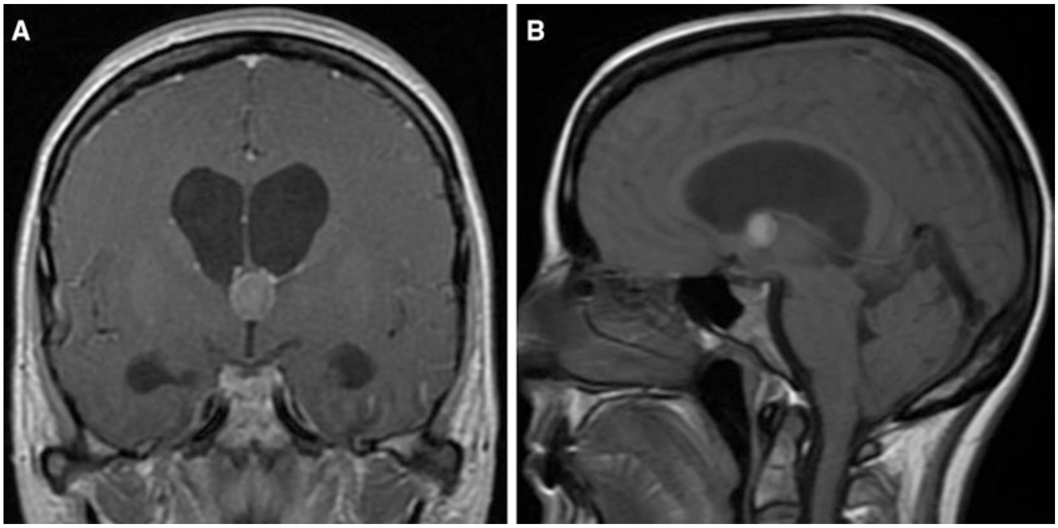


Fig. 22.1 (**a**, **b**) **CASE EXAMPLE**: colloid cyst. A 42-year-old man presents with headaches but is otherwise neurologically intact

22.2 Colloid Cysts: Open Transcranial Approaches

Anil Nanda and Devi Prasad Patra

Introduction

Colloid cysts are benign lesions of the third ventricle yet are sometimes associated with poor outcomes if not treated appropriately. A complete removal of the lesion is imperative to provide symptomatic relief as well as complete cure. Since the description of the transcortical-transventricular approach by Dandy [3], many of these lesions have been successfully removed. Over the years, other surgical corridors including the interhemispheric transcallosal and subfrontal lamina terminalis approaches have been explored with comparable efficacies in dealing with this lesion. Advancement of the endoscopic technique has recently surpassed all the problems of open approaches and is being utilized more frequently for its minimal invasive nature. In spite of that, open approaches are still practiced by many neurosurgeons around the world and are regarded as the gold standard for complete excision of colloid cysts [4–7].

Preoperative Evaluation

Most of the patients with colloid cysts present with nonspecific headaches and episodes of acute rise in intracranial pressure. In such cases neurological examination is mostly normal. A preoperative documentation of the higher cognitive function, including memory and language function, is important to detect any possible postoperative deterioration. Colloid cysts are readily visible in standard radiological studies like computed tomography (CT) or non-contrast MRI. Images in every surgical plane should be evaluated properly to identify the cyst's exact location in the third ventricle and its extension. Degree of ventricular dilatation and the thickness of the cortical mantle should be evaluated if a transcortical approach is being planned. For a transcallosal approach, the most important study to evaluate is the cerebral venogram, which can provide details of the cortical draining veins and their location in relation to the coronal suture. Secondly, the position of the internal cerebral veins should be carefully evaluated to detect any displacement or other anatomical variations. A preoperative assessment of the degree of dilation of the foramen of Monro or any deviation of the forniceal columns is helpful in selecting a suitable approach. The use of neuronavigation is particularly helpful for appropriate placement of the craniotomy and to aid the localization of the cyst inside the ventricle if it is not visible during surgery.

Surgical Technique

Colloid cysts are located in the anterior third ventricle, and surgical approaches for these lesions revolve around the ease of transventricular entry. Almost all cases require initial entry into the lateral ventricle and then into the third ventricle. Any intraventricular entry requires division of neural structures; hence, surgical approaches are weighed upon the relative deficits and complications from this anatomical disruption. For proper understanding, all the surgical approaches are divided into three sequential steps: (a) entry into the lateral ventricle, (b) entry into the third ventricle, and (c) cyst removal.

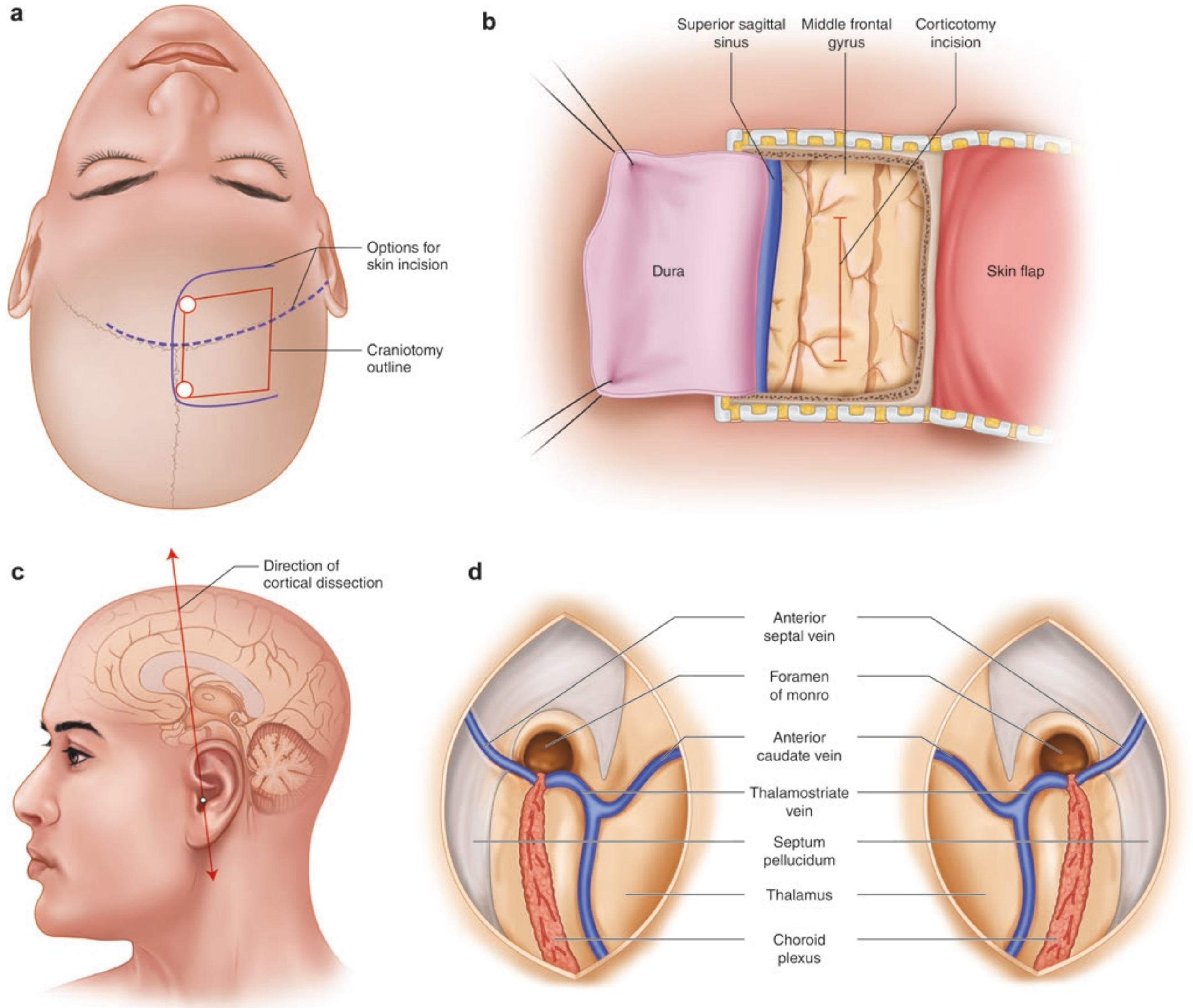


Fig. 22.2 (**a**). Outline for the skin incision (*blue line*) and craniotomy (*red line*) for transcortical approach (**b**). The dura has been cut and reflected medially based on the superior sagittal sinus. The middle frontal gyrus is the usual site of cortisotomy (*red line*) (**c**). The direction of opening of the cortisotomy can be traced along the line connecting the coronal suture to the external acoustic meatus that leads to exposure of the lateral ventricle at the level of the foramen of Monro. Neuronavigation can also guide dissection. (**d**, **e**). Entry can occur in either ventricle; understanding the orientation of the thalamostriate vein to the choroid plexus is helpful in determining the side of the ventricle. In the left ventricle (D) the thalamostriate vein lies on the left, and on the right ventricle (E), it lies to the right of the choroid plexus

Entry into the Lateral Ventricle

Multitudes of approaches have been proposed for entry into the lateral ventricles with specific indications and complications; however, we will mainly discuss the two main approaches adopted by many surgeons, the transcortical-transventricular and interhemispheric transcallosal approaches.

Transcortical-Transventricular Approach

The transcortical-transventricular approach, as the name implies, involves the opening of the cerebral cortex (frontal cortex) to gain access to the ipsilateral lateral ventricle. The nondominant side (usually right) is usually preferred unless dictated by the asymmetric dilatation of the lateral ventricle or significant extension of the tumor into the left lateral ventricle, which would otherwise be more suitable for a left-sided approach.

Operative Steps

The patient is placed supine, and the head is fixed with Mayfield head holder with the head slightly flexed. A pure midline head position is favored for proper orientation of the intraventricular structures during surgery. The scalp incision can be fashioned in multiple ways, but we prefer a lateral-based horseshoe scalp flap that allows a craniotomy of approximately 6 cm × 4 cm (Fig. 22.2a). The exact position of the craniotomy should be properly estimated by the location of the lesion in the third ventricle using neuronavigation; however, the posterior extent should not exceed beyond 2–3 cm behind the coronal suture to avoid the motor strip. The midline extent of the craniotomy is up to the sagittal sinus, which need not be fully exposed. The dura is divided along the craniotomy line and is reflected based on the sagittal sinus. The middle frontal gyrus is the usual site of the corticotomy, but a trans-sulcal approach can be more direct and less invasive (Fig. 22.2b). Cortical veins should always be preserved, although they do not usually pose a significant problem in this approach. Dissection proceeds toward the ipsilateral foramen of Monro. The direction can be predicted by an imaginary line connecting the coronal suture to the ipsilateral external auditory meatus; alternatively, navigation can be used (Fig. 22.2c). The ventricular ependymal lining is appreciated by a grayish hue with ependymal vessels. After the ventricle is opened, the choroid plexus and thalamostriate veins are seen and are traced to identify the foramen of Monro (Fig. 22.2d, e). Although rare, it is possible to enter into the opposite ventricle, especially in patients with asymmetric ventriculomegaly. In these cases the aforementioned structures are used to confirm the side of entry. The choroid plexus is initially identified and traced anteriorly to look for its disappearance in the foramen of Monro. At the foramen, the thalamostriate vein is identified running lateral to the choroid plexus. The position of the thalamostriate vein to the right or left of the choroid plexus confirms the side of entry into the right or left lateral ventricle, respectively.

Complications

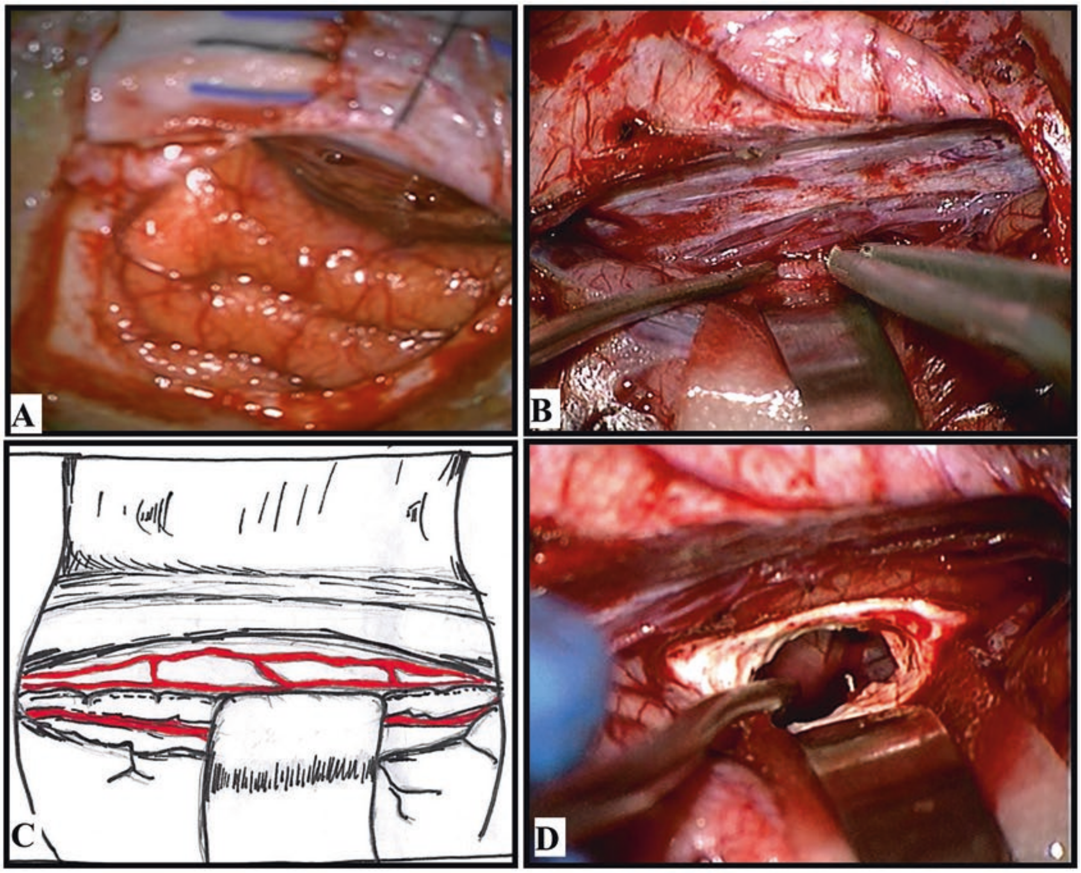
The most commonly discussed complication in this approach is seizure which can occur in as many as 30% of the patients [8, 9]. With the advancement of microsurgical techniques, the incidence of seizure has decreased, and recent series report seizure occurrence rates of 5–10% [10, 11]. The occurrence of seizures is related to the cortical incision which acts as the seizure focus; however, cerebral edema secondary to venous compromise or retraction can also be contributing factors. The other important potential complication is focal motor deficit, most commonly transient hemiparesis due to retraction of the motor cortex. A dense hemiplegia suggests a direct injury to the internal capsule, which lies just lateral to the lateral ventricular wall at the level of the foramen of Monro and can be damaged by errant pathway of cortical dissection. Other less common complications are subdural hematoma due to rapid decompression of hydrocephalus and subdural hygroma due to persistent cerebrospinal fluid leakage through the corticectomy site, meningitis, and ventriculitis.

Interhemispheric Transcallosal Approach

The interhemispheric transcallosal approach involves a pure midline approach to the ventricle between the two cerebral hemispheres without the need of a cortical incision. As compared to the cortical approach, it seems to be less invasive but is more difficult and technically demanding.

Operative Steps

The initial patient positioning and placement of scalp flap are nearly the same as the transcortical approach except that the craniotomy requires a lesser lateral extension and more exposure across the midline. The sagittal sinus should be exposed at least by half of its width to give an unrestricted midline access (Fig. 22.3a). The position of the craniotomy in the anteroposterior direction is dictated not only by the location of the lesion but also by the pattern of cortical draining veins on the preoperative venogram. One of the most important challenges in this approach is to develop a corridor between the cortical veins draining into the sagittal sinus. The course of the coronally oriented draining veins should be studied in the preoperative venograms to identify the widest possible corridor in front of the coronal suture. A forced anterior or posteriorly placed craniotomy based on the venous course does not create difficulty in gaining a comfortable path toward the ventricle, which can be easily accomplished by lowering or elevating the head, respectively. All veins are to be carefully protected at all costs to minimize the chance for complication. In difficult cases the smaller veins can be sacrificed if required; however, larger veins should be mobilized by carefully dissecting and dividing the arachnoidal attachment from the brain surface and the sinus. The medial edge of the hemisphere is carefully retracted over a cottonoid to enter into the interhemispheric fissure (Fig. 22.3b). A few retracting sutures on the dura just lateral to the sinus can effectively increase the angle of exposure. Small bridging veins between the hemispheres are coagulated and divided. Important structures to identify at this point are the cingulate gyrus, pericallosal artery, and callosomarginal artery. The callosomarginal artery runs above the cingulate gyrus, whereas the pericallosal artery runs below it and above the corpus callosum, although variations may exist (Fig. 22.3c). The corpus callosum is identified by its shining white color as compared to the cingulate gyrus, which is a dull white similar to the cortex with overlying pial vessels. This differentiation is critical to avoid inadvertent entry into the cingulate gyrus. Both pericallosal arteries are to be identified running parallel to each other, and a surgical plane between them is developed by dividing the arachnoid adhesions. This midline separation of the arteries is essential to avoid injury to the lateral cortical perforating arteries. About 3–4 cm of the corpus callosum is exposed in the midline, and the site of incision is determined by using navigation. The callosotomy is done by a knife with gradual



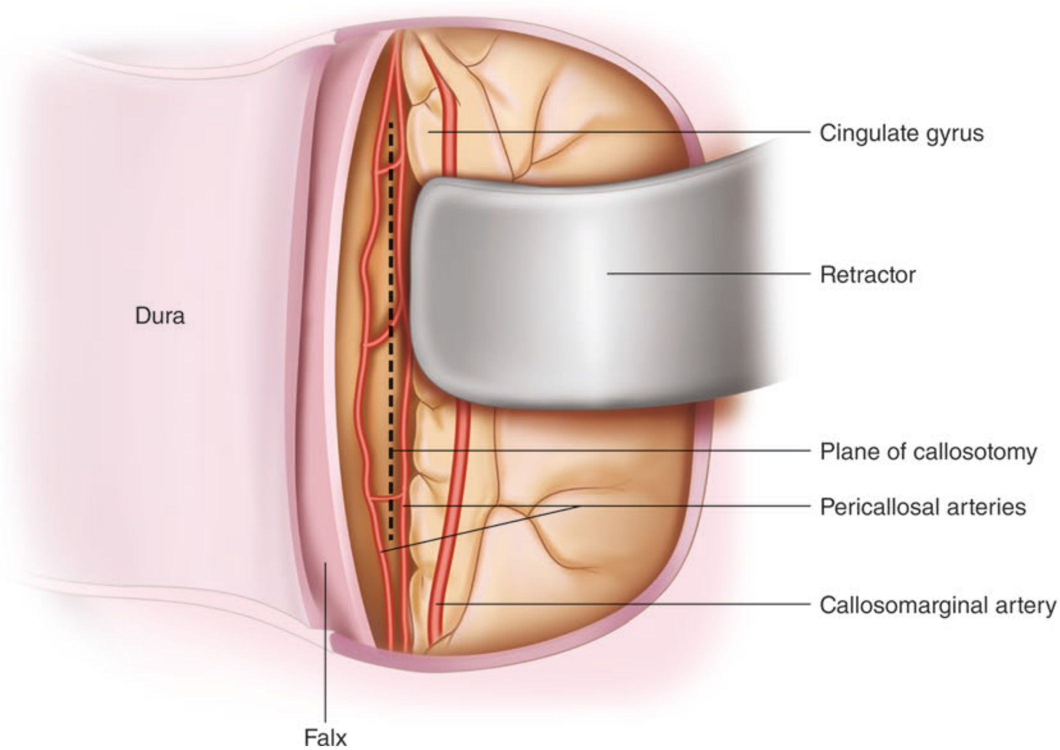


Fig. 22.3 (**a–d**). A. Intraoperative image of the interhemispheric transcallosal approach. The dura has been reflected medially based on the superior sagittal sinus exposing the interhemispheric fissure. (**b**). The medial hemispheric cortex has been retracted to expose the pericallosal vessels below the falx. (**c**). Illustration showing the orientation of the pericallosal vessels. The plane of dissection should be between the two pericallosal arteries to avoid lateral cortical perforators. (**d**). Small callosotomy has exposed the lateral ventricle. Note the bright white color of the corpus callosum

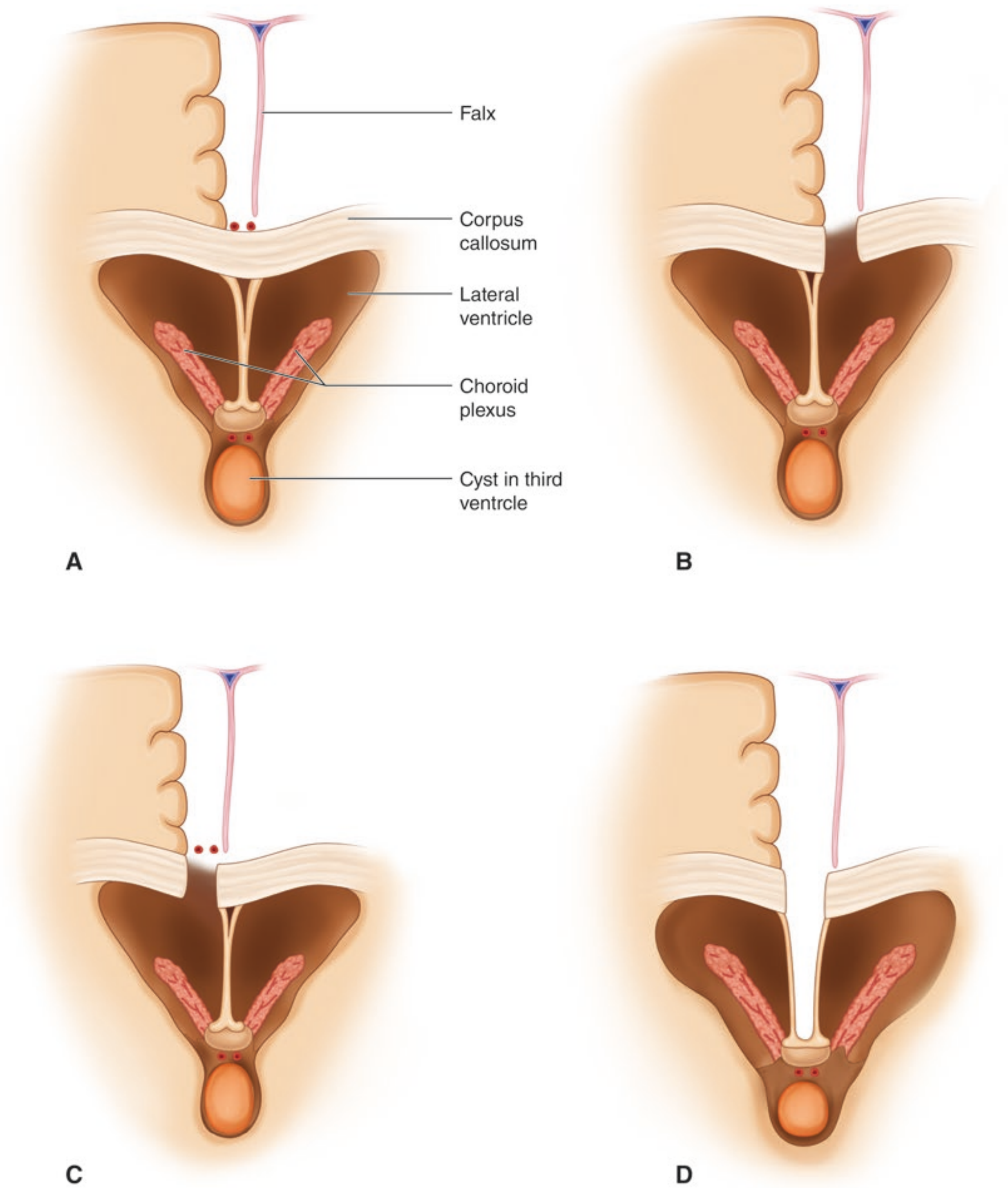


Fig. 22.4 (**a**–**d**). Interhemispheric transcallosal approach (**a**) can open into the right ventricle (**b**), or into the cavum of the septum pellucidum (**c**), or into the left ventricle (**d**)

separation of fibers using bipolar forceps and a suction tip over cottonoids. About 1–2 cm of callosotomy is usually satisfactory to access the colloid cyst (Fig. 22.3d). Depending upon the direction of dissection and orientation of ventricles, either of the lateral ventricle or the cavum of the septum pellucidum may be entered (Fig. 22.4a–d). The side of the ventricle can be estimated based on the orientation of the thalamostriate vein and the choroid plexus. A septum pellucidum entry makes the view blind without any visible structures in the cavity. A simple fenestration on the lateral wall of the septum is all that is needed to enter the ventricular cavity.

Complications

The most feared complication in this approach is damage to a major cortical vein and subsequent venous edema and hemorrhage. Desai et al. have reported this complication in 4 out of 62 patients they have operated through this route [11]. On the other hand, a few series, including our own series, did not have such complications [12, 13]. In the meta-analysis by Sheikh et al., the transcallosal approach was associated with venous infarction in 2.7% of the patients as compared to none in the transcortical approach [10]. Postoperative focal deficits like monoparesis, hemiparesis, amnesia, or aphasia can be secondary to cortical retraction, cortical vein, sagittal sinus thrombosis, or injury to the pericallosal artery. A specific concern about the development of disconnection syndrome due to callosotomy has been widely discussed in the literature. An impairment of the interhemispheric transfer of sensory or motor information is theoretically possible; however, measurable deficits have not been reported frequently. A possible hypothesis is that the callosotomy required in this approach does not involve the splenium and thereby avoids significant impairment of inter-cortical transfer. The callosotomy required for the surgery of colloid cyst is presumed to be small enough to avoid causing any demonstrable disconnection symptoms [14–17]. However, a few series have reported subtle deficits like tactile and auditory transfer deficits [18–20]. Other complications are similar to the transcortical approach which includes subdural hematoma or hygroma, meningitis, and ventriculitis.

Selection of Approach

After the initial description by Dandy, the transcortical approach was probably the most widely method used for third ventricular lesions. Consequences of callosotomy have been overestimated in older literature until the last three decades when many reports suggested minimal to no deficits after a limited anterior callosotomy. Each approach has its advantages and limitations (Table 22.1). Several factors guide the suitable approach, which include:

1. Degree of dilatation of the lateral ventricles: In the presence of hydrocephalus, both approaches serve equally because the stretching of neural structures helps to decrease the working depth required to enter into the ventricle. However, the absence of ventriculomegaly or the presence of a functioning shunt poses problems for the transcortical approach because of the need for a large cortical incision and more retraction. The transcallosal approach is particularly suitable in these instances as it does not require a cortisotomy.
2. Specific location and extent of the colloid cyst in the third ventricle: In the transcallosal approach, a direct midline view is beneficial to visualize both the lateral ventricles with equal ease and so is more versatile to deal with lesions extending bilaterally (Fig. 22.5a, b). Similarly, a midline approach is favorable should an interfornicial approach be required to access the third ventricle. In the transcortical route, the angle of vision makes it easier to access the ipsilateral lateral ventricle and the contralateral wall of the third ventricle (Fig. 22.5a, b). Therefore, a cyst extending predominantly to one side in the setting of hydrocephalus can be easily removed through a transcortical approach with minimal damage.
3. Venous anatomy of the frontal cortex: Lack of a suitable corridor between draining veins on the preoperative venogram makes the transcallosal approach less suitable.
4. Patient presentation and preoperative cognitive function: Acutely decompensated patients or patients with cognitive dysfunction, a transcortical route may be favorable because it is less technically demanding and avoids retraction of the cingulate gyrus or division of the corpus callosum.

Table 22.1 Comparison of approaches to the lateral ventricle

	Transcortical approach	Transcallosal approach
Advantages	Easy and less technically demanding	Direct and shorter length of access
	Good visualization of ipsilateral lateral ventricle and contralateral wall of third ventricle	Good visualization of both lateral ventricles as well as third ventricles
	Less risk of cortical vein injury	Does not require cortisotomy, so less seizure risk
	No risk of disconnection syndrome	Suitable for non-dilated ventricles
Disadvantages	Not suitable for non-dilated ventricles	Risk of cortical vein injury or thrombosis
	Poor visualization of contralateral foramen of Monro	Risk of disconnection syndrome
	Risk of seizure	Risk of damage of pericallosal arteries
	Risk of focal neurological deficits	Technically more difficult

Entry into the Third Ventricle

Once inside the lateral ventricle, the lesion is readily appreciated bulging through the foramen of Monro. In rare instances, the cyst is located posteriorly or buried behind the opposite foramen of Monro, and so it may not be readily visible with a standard lateral ventricular exposure. Most cysts can be easily decompressed without any need to enter inside the third ventricle; however, some form of access to the anterior third ventricle is needed for the final detachment of the cyst from the third ventricular roof. The most important factor guiding the further steps is the amount of dilatation of the foramen of Monro. An adequately dilated foramen as seen in most of the cases allows further dissection and removal of the cyst without any hindrance. However, in patients with a normal-sized foramen or non-visualized cysts, the anterior third ventricle needs to be exposed for safe manipulation of the cyst. The anterior third ventricle can be entered by various routes, and we will discuss the three most important approaches.

Transforaminal Approach

Being cystic in character, most of the colloid cysts can be decompressed by needle aspiration and shrunken to a level that can be easily removed even in a non-dilated ventricle. However, in some cases without proper visualization of the

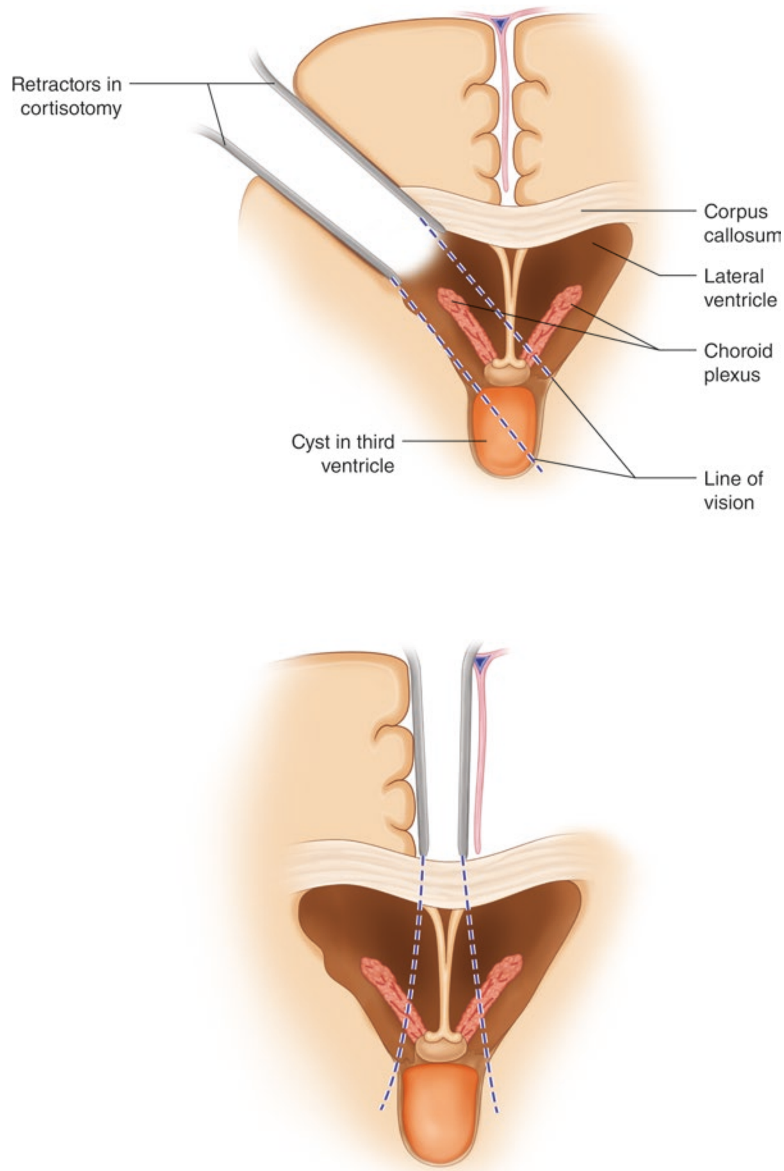


Fig. 22.5 (**a**, **b**). Comparison of the line of exposure in the transcortical approach (**a**) and interhemispheric transcallosal approach (**b**)

extent of the cyst, it is appropriate to enlarge the foramen for a safe manipulation of the cyst. Should the need arise, an initial attempt can be made to enlarge the foramen by separating the closed tips of the forceps in either direction to mobilize the cyst without cutting any neural structure. However, in difficult cases further dilatation can be achieved by cutting the wall of the foramen. Two options have been described in literature in this regard: (1) cutting the inferior aspect of the fornix unilaterally and (2) cutting the posterior inferior aspect of the foramen with partial removal of the anterior thalamus (Fig. 22.6a).

Operative Steps

The column of the fornix can be divided unilaterally in its inferior aspect anterior to the septal vein as it starts separating from the opposite forniceal column anterior to the foramen (Fig. 22.6b). This widening is mostly helpful in lesions that are visible but too large to be retrieved through the small foramen. Removal of the anterior part of the thalamus gives more access to the posterior aspect of the foramen where the cyst is attached to the roof of the third ventricle. The thalamostriate vein is the main hindrance and limits the posterior extent of division. The thalamostriate vein can be separated from the substance of the thalamus by carefully cutting its arachnoid attachments and can be mobilized posteriorly to expose the anterior thalamus, which is subsequently curetted out. In both the situations, the foramen can be reasonably dilated to remove most of the lesions; however, as there is a limit in the allowable division of the neural structures, very large lesions should not be treated via this route.

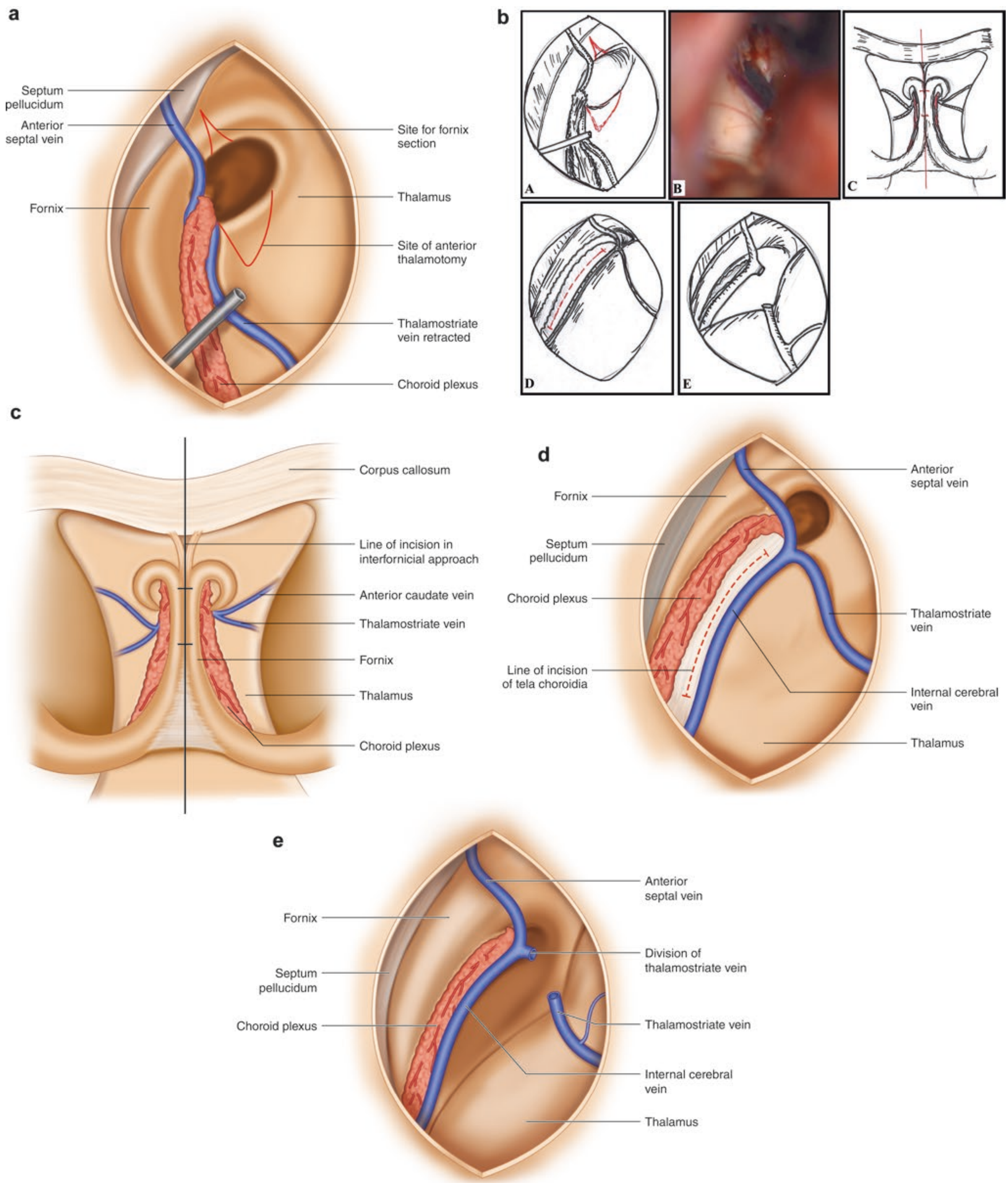


Fig. 22.6 (**a**). The foramen of Monro can be enlarged anterosuperiorly through a forniceal incision or posterolaterally through anterior thalamotomy (**b**). Intraoperative image showing small forniceal incision to enlarge the foramen (**c**). Interfornicial approach to third ventricle. The forniceal columns are separated along the midline (**d**). Subchoroidal approach to the third ventricle. Widely separated, internal cerebral veins can give easy access to the third ventricle after division of the tela choroidea (*red-dashed line*) (**e**). In other cases, the thalamostriate vein can be divided to adequately open the third ventricle

Complications

Both of these methods of dilatation of the foramen of Monro carry a definite risk of neurological deficits. Division of the fornix can result in short-term memory loss [21, 22]. Some authors report no significant risk of memory loss after unilateral forniceal division [23, 24], but others have reported transient memory loss. This transient memory loss is likely multifactorial but may be related to preoperative dysfunction of the contralateral fornix and/or due to the intraoperative manipulation. Division of the posterior inferior aspect of the foramen risks damage to the internal cerebral veins and their confluence with the thalamostriate veins [25]. The anterior nucleus of the thalamus is related to the limbic system, and unilateral removal of some of the substances has not been implicated in causing appreciable deficits [26].

Interfornicial Approach

This approach essentially involves entry to the third ventricle from above in between the two forniceal columns (Fig. 22.6c). This gives a midline exposure of the third ventricle, which allows visualization of most lesions. The interfornicial route can be explored both after a transcortical or transcallosal exposure, but the later approach comes with a distinct advantage of having a pure midline orientation.

Operative Steps

The most important structure to identify for a safe entry through the interfornicial corridor is the midline raphe. The two columns of the fornix unite together to form the body separated by a midline raphe that lies over the roof of the third ventricle. Posteriorly they again splay apart to form the crus of the fornix separated by the commissure of fornix. The structure that guides the identification of the midline raphe is the septum pellucidum. The septum pellucidum can have anatomical variability ranging from a single membranous structure to a complete cavity referred to as a cavum. From a transcallosal midline perspective, the two layers of the septum can be separated to identify the midline raphe at the base of its attachment. If a lateral ventricular entry has been achieved (by either the transcallosal or transcortical approach), the septum can be fenestrated and opened at the base to separate the two layers. It is very important to understand the midline sagittal orientation of the septum in relation to the midline raphe because in some cases of asymmetric ventriculomegaly, the septum may have a distorted shape, and there could be some displacement or stretching of one of the fornices. In these cases the midline raphe may not have a pure midline orientation. The fornices must be separated exactly along the midline raphe to avoid damage. The raphe is divided by a sharp knife and forceps starting at the level of foramen of Monro and extending approximately 2 cm posteriorly. It is again important not to extend the incision too far posteriorly beyond 2 cm to avoid damage to the hippocampal commissure, which may produce permanent memory deficits. Structures in the roof of the third ventricle including the internal cerebral veins do not pass directly underneath as they are displaced laterally in most cases by the expansion of the cyst. However, surgeons should not take this displacement for granted and should be careful while opening the roof as in some cases the veins may lie exactly in the midline. After separation, the fornices usually do not require retraction as enough space can be achieved to decompress the cyst and remove its attachment.

Complications

Even with experienced hands, some form of damage to the fornices is inevitable during this approach. Transient memory loss may result from surgical manipulation of the fornices. The extent of forniceal separation and preoperative cognitive function may have some role. As the interfornicial approach is uncommonly required for colloid cysts, there is little literature on the risk of this complication. However, in other surgical series in which an interforniceal approach was used to access other third ventricular tumors, rates of memory loss as high as 63% have been reported [27–30].

Subchoroidal Approach

An anterolateral entry to the third ventricle can be achieved by the subchoroidal trans-velum interpositum approach. This route has been proposed to avoid significant complications after retraction or division of the fornices involved in the above two approaches. Division of the tela choroidea can open the third ventricle without any need for neural structure division; however, the thalamostriate vein significantly limits the exposure. In most cases where the subchoroidal approach is indicated, there is separation of the natural corridor caused by displacement of the internal cerebral veins laterally by the cyst. In some cases, the thalamostriate vein may need to be sacrificed in order to gain adequate exposure.

Operative Steps

The choroid plexus running along the floor of the lateral ventricle is identified and is elevated to expose the fissure between the thalamus and the third ventricular roof [31]. The plexus is attached to the base by two leaflets of arachnoid, one on the medial side to the velum and the other on the lateral side to the thalamus. Sharp division of the arachnoid leaflet on either of these may open the third ventricle. In most cases the internal cerebral vein is displaced laterally in which division of the medial arachnoid leaflet may provide a natural corridor (Fig. 22.6d). In other cases, the thalamostriate vein is separated from the dorsal surface of thalamus and is mobilized to widen the corridor. If required, the thalamostriate vein can be coagulated and divided from the internal cerebral vein (Fig. 22.6e). This allows a wide anterolateral exposure of the third ventricular cavity after gentle retraction of the thalamus laterally and body of the fornix medially.

Complications

The most important potential complication arises from the occlusion of the thalamostriate vein. This may lead to catastrophic venous edema and subsequent hemorrhage in the basal ganglia; [32] however, reports of such complications are surprisingly low [12, 33]. Other complications may arise from retraction of the thalamus or inadvertent injury of the internal cerebral or posterior thalamic veins.

Selection of Approach

When the lesion is small or easily visualized through the foramen of Monro, every effort should be made to remove it through the foramen by manipulating it from all the sides (Table 22.2). A dilated foramen of Monro can be a blessing in disguise and is the most favored and routinely used corridor. Dilatation or foraminotomy of a small foramen is only indicated in a slightly bigger lesion that cannot be easily mobilized. Sizable lesions may require alternative approaches. If the lesion is filling the third ventricle and is attached along the whole length of the roof, then a midline interforniceal approach seems reasonable in which the exposure may extend more posteriorly than any other approach. This approach is again preferred in the absence of hydrocephalus as it does not require entry into the lateral ventricle. In all other cases, forniceal approaches should be avoided. Subchoroidal approach can be used for larger lesions in the middle or posterior third ventricle and is most suitable in cases with wide separation of the internal cerebral veins.

Table 22.2 Comparison of approaches to the third ventricle

Approach	Favorable situations	Unfavorable situations
Transforaminal	Small lesions	Large lesions
	Lesions visible through the foramen of Monro	Non-dilated foramen of Monro
	Widened foramen of Monro	Non-dilated ventricles
	Cysts in the anterior third ventricle	
	Dilated lateral ventricles	
Interforniceal	Lesions with wide attachment to the roof	Small lesions
	Separation of body of fornix by the cyst	Predominantly posterior lesions
	Non-dilated ventricle	Preoperative cognitive dysfunction
	Lesions not visible through foramen of Monro	
Subchoroidal	Dilated lateral ventricles	Small lateral ventricles
	Dilated third ventricle with wide separation of the internal cerebral veins	Small lesions
	Posteriorly placed lesions	Adherent choroid plexus or prior ventriculitis
	Lesions not visible through foramen of Monro	

Removal of the Cyst

Attempts should be made to completely remove the cyst using a combination of methods in order to avoid recurrence. A small (<1 cm) colloid cyst visible through the foramen of Monro is favorable for surgical resection (Fig. 22.7a). Such lesions should not be punctured as the distended capsule makes it easier to dissect it from the surrounding structures. A small probe can be passed around the lesion through the foramen along the whole circumference to free it from friable adhesions except on the superior aspect where the attachment with the internal cerebral veins lies (Fig. 22.7b) [29]. The cyst can be put on slight traction, and the attachment is sharply divided to free it completely. Larger lesions should not be removed without decompressing the cyst. The cyst wall is punctured, and the contents are aspirated by a needle or shunt tube to avoid spillage into the ventricular cavity. As the cyst is decompressed, the posterior extent of the attachment is estimated by passing a probe smoothly around the cyst. For anteriorly located lesions, the attachment can be easily coagulated and divided through the foramen (Fig. 22.7c). In larger lesions that extend to the posterior third ventricle or with tightly adhered attachments, the third ventricle should be entered by either approach as described above for direct visualization. Traction on the attachment and avulsion should be strictly avoided as it leads to tearing of the internal cerebral veins with devastating complications. Thorough irrigation is required to confirm a bloodless ventricular cavity.

Specific Challenges

Small Ventricular Cavity

A non-dilated ventricle does not allow adequate working space and requires undue manipulation and retraction of neural structures during surgery. A midline transcallosal approach is specifically helpful in this situation to visualize the foramen of Monro from above. An interfornicial corridor may be added if there are concerns of inadequate dilation of the foramen to safely deliver the cyst.

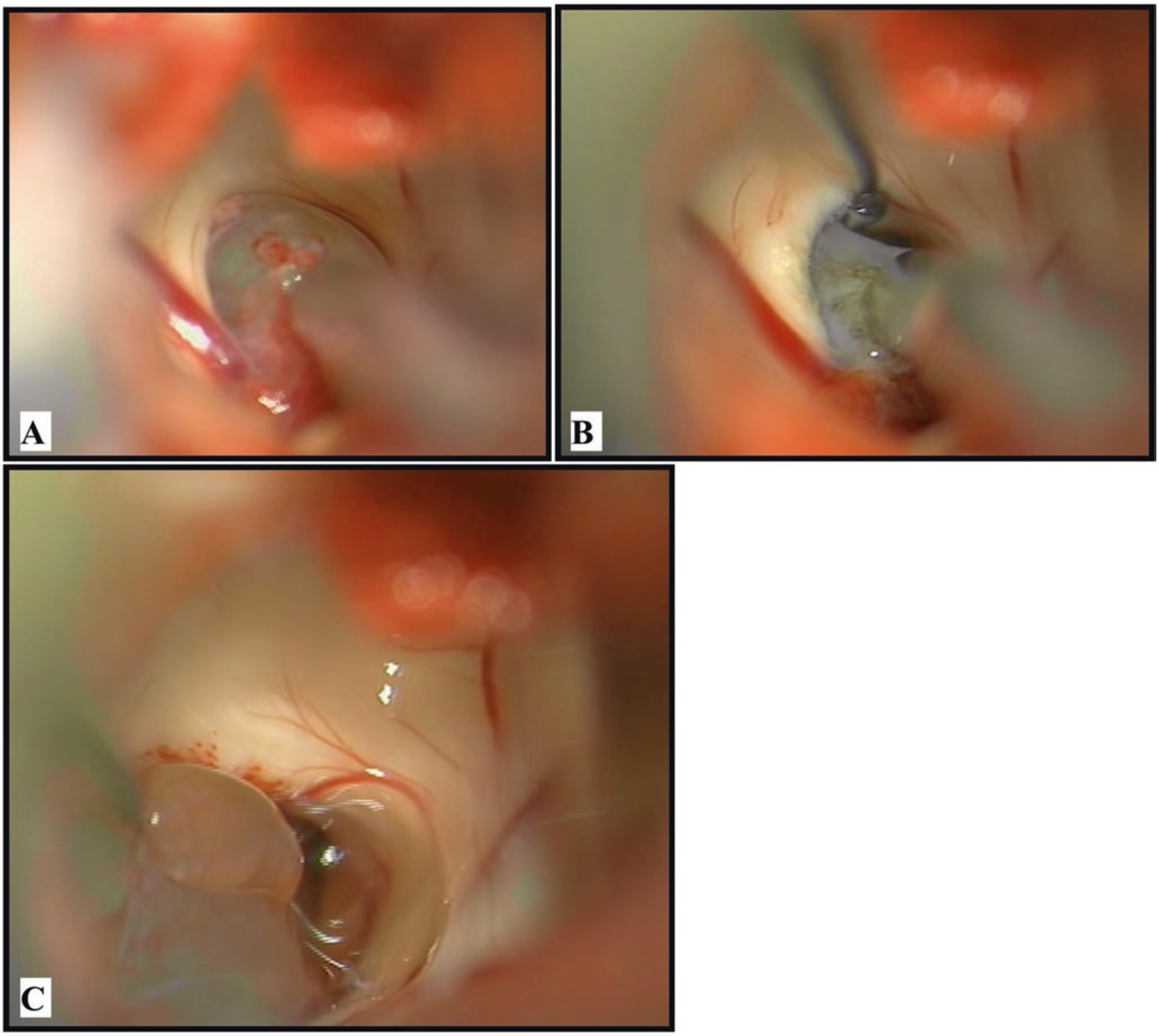


Fig. 22.7 (**a**). Intraoperative image showing a large colloid cyst visible through the foramen of Monro (**b**). After initial decompression, a probe can be passed along the circumference of the cyst to dissect it from the wall of the third ventricle (**c**). The cyst is being delivered through the foramen of Monro

Non-visualization of the Cyst

A cyst may not become obvious after entry into the lateral ventricle. Lack of cyst visualization may occur if the surgeon enters into the ventricle opposite to the side where the cyst is protruding [28]. This is of particular concern in the transcallosal approach where one may end up on either side of the lateral ventricle. Similarly, entry into the cavum of the septum pellucidum is also possible without any visible landmarks. In either case, fenestration of the septum will lead into the correct side. If the cyst is not visualized even from the correct side of the ventricle, then the cyst is probably located in the anterior and inferior parts of the foramen of Monro or in the posterior third ventricle. The anteroinferior part of the foramen of Monro is a hidden space from the surgical angle routinely employed during surgery and can be looked for by either rotating the position of the head or by passing a probe in that direction. If a cyst is found, then the foramen can be widened with limited inferior division of the fornix. A posterior location of the cyst in the third ventricle can be confirmed by passing an endoscope and if located can be approached by a subchoroidal route [34].

Nondelivery of the Cyst

A large cyst should always be decompressed before trying to deliver through the foramen. In some cases, the cyst content is too thick or calcified to be aspirated [29]. Piecemeal excision of the cyst may be attempted, but a long cottonoid should always be placed below the cyst through the foramen to avoid migration of pieces deep into the third ventricle. A densely adherent cyst should not be pulled but instead be managed under direct vision by opening of the third ventricle.

Bleeding from the Attachment

Small bleeding may occur after removal of the attachment, which usually stops with saline irrigation. Application of bipolar coagulation should be avoided unless a clear point of bleeding from small veins is visible. Most bleeding is from venous tributaries and can be managed with placement of small Gelfoam on a cottonoid. For persistent bleeding, direct application of pressure over a hemostat may be needed after opening of the third ventricle.

Surgical Outcome

Overall outcome following open surgical removal of colloid cysts is excellent. The direct visualization of the cyst attachment along with the ability of bimanual manipulation by an open surgical approach comes with the definite advantage of complete resection of the cyst with good long-term outcome. Most of the recent series [11–14, 35–37] report a gross total resection rate of more than 90%, and therefore the recurrence rate is very low to absent [12, 13, 35, 37, 38]. In their meta-analysis, Seikh et al. have analyzed 1278 patients operated by microsurgical or endoscopic techniques and found that microsurgical group had a significantly greater extent of resection, lower rates of recurrence, and lower rates of reoperation than the endoscopic group [10]. The overall complication rates were lower in the endoscopic group; however, the mortality rates were similar. One of the most important adverse outcomes is memory loss which has been reported from 9.5% of patients [11] to as high as 53% of patients in some series [35].

PEARLS
Both the transcortical and transcallosal approaches provide adequate view of the lateral ventricle and then the third ventricle. A parasagittal craniotomy placed two third anterior and one third posterior to coronal suture is mostly used for both approaches with minimal variations.
The transcortical route involves corticectomy in the middle frontal gyrus and then dissection toward the ipsilateral lateral ventricle. In the transcallosal approach, the body of the corpus callosum is exposed through anterior interhemispheric route, and then the ventricle is entered through a small callosotomy.
In most of the cases, the colloid cysts are visible in the anterior third ventricle and are removed through a transforaminal approach. In large or posteriorly located cysts, the third ventricle needs to be opened through an interfornicial or a subchoroidal approach.
Total excision of the cyst should be the goal. Initial decompression of the cyst followed by careful removal of the attachment from the roof of the third ventricle should be performed, preferably under direct vision.

Conclusion

Both the transcortical and transcallosal approaches are being used worldwide; however, evidence is accumulating in favor of the transcallosal approach as the preferred route [6, 10, 12, 38]. With proper planning and appropriate selection of surgical approach, this benign yet devastating lesion of the third ventricle can be completely removed with definitive cure.

Commentary on Case Presentation in Section 22.1

Presentation

A 42-year-old man presents with headaches. His exam shows that he is otherwise neurologically intact. (See Fig. 22.1a, b).

Discussion

The patient has a colloid cyst in the anterior part of the third ventricle. The lateral ventricles are symmetric and moderately dilated. Both the transcortical and transcallosal approaches would be feasible for this patient; however, considering the thick cortical mantle and a smaller cyst size, the transcallosal approach would be more appropriate and less invasive. A preoperative venogram and intraoperative navigation would guide the appropriate placement of the craniotomy. Once inside the ventricle, the cysts should be visible through the foramen of Monro without difficulty. Considering a non-dilated foramen of Monro, the cyst needs to be decompressed initially. A careful manipulation through the foramen with the probe around the cyst wall should free it from the adhesions along the third ventricular wall. Gentle traction should enable the surgeon to visualize the attachment along the roof followed by the complete resection of the cyst wall.

22.3 Third Ventricular Colloid Cysts: Endoscopic Transventricular Approach

Christian Diniz Ferreira, Kléver Forte de Oliveira, and Samer K. Elbabaa

Brief Introduction

Colloid cysts represent about 1% of all intracranial neoplasms and 15–20% of intraventricular tumors [39, 40]. The first transcortical-transventricular approach to a colloid cyst was reported by Dandy in 1921 [41]. About 60 years later, in 1983, the first transventricular endoscopic aspiration was described for the treatment of the colloid cysts of the third ventricle [34, 39].

Surgical Technique

Anatomy

Anatomical knowledge is essential for better understanding of neuroendoscopic indications and techniques. Here we briefly review the lateral and third ventricular anatomy.

Lateral Ventricles

The lateral ventricles are two C-shaped cavities situated deep within the cerebrum, each one in a hemisphere, around the thalamus. They are lined on the inside by ependyma, an epithelial cell tissue, and filled with cerebrospinal fluid (CSF). Each lateral ventricle is a closed cavity, except for the interventricular foramen (foramen of Monro) that connects it with the third ventricle [42].

Each lateral ventricle is divided into five parts: three horns (frontal, temporal, and occipital horns), the body, and the atrium [42].

The choroid plexus is attached to the choroidal fissure, a narrow C-shaped cleft between the fornix and the thalamus (Fig. 22.8). It is seen in the medial part of the body, atrium, and temporal horn. The fornix and the thalamus are the superior and inferior limits of the choroidal fissure in the body of the lateral ventricle. The choroid plexus of the lateral ventricles goes through the foramen of Monro and continues with two parallel strands of choroid plexus in the roof of the third ventricle [42].

Fig. 22.8 Superior view of lateral ventricles, showing the following structures: thalamostriate vein (*arrow*), superior choroidal vein (*arrowhead*), body of corpus callosum (*1*), splenium of corpus callosum (*2*), head of caudate nucleus (*3*), body of caudate nucleus (*4*), collateral trigone (*5*), thalamus (*6*), and choroid plexus (*7*)

Third Ventricle

The third ventricle is a narrow, funnel-shaped, midline cavity located in the diencephalon. It communicates with each lateral ventricle through the foramina of Monro and with the fourth ventricle by the aqueduct of Sylvius, also known as the cerebral aqueduct [42].

The foramina of Monro are located at the junction of the roof and anterior wall of the third ventricle. Its anterior limit consists of the junction of the body and columns of the fornix. The anterior pole of the thalamus is its posterior limit. The choroid plexus, the distal branches of the medial posterior choroidal arteries, and the thalamostriate, superior choroidal, and septal veins are the structures that pass through the foramen [42].

The roof of the third ventricle extends from the foramina of Monro to the suprapineal recess posteriorly (Fig. 22.9). It consists of four layers: one superior formed by the fornix, two thin semiopaque membranes derived from the pia mater connected by fragile trabeculae forming the tela choroidea, and a layer of blood vessels (medial posterior choroidal arteries and their branches and the internal cerebral veins and

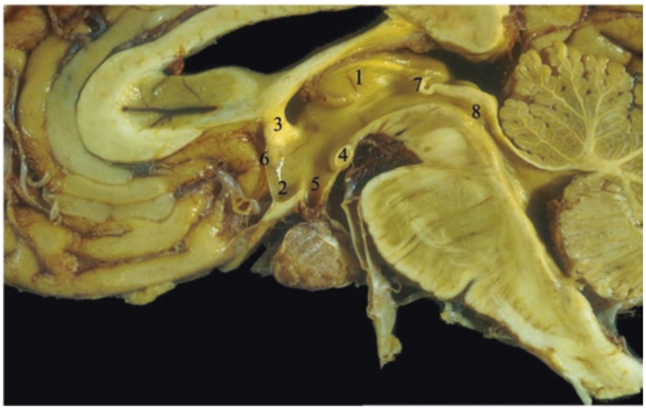


Fig. 22.9 Sagittal view of third ventricle, demonstrating the following anatomical structures: massa intermedia (*1*), optic recess (*2*), column of the fornix (*3*), mamillary body (*4*), infundibular recess (*5*), lamina terminalis (*6*), posterior commissure (*7*), and aqueduct of Sylvius (*8*)

their tributaries) between the sheets of tela choroidea. This space between the sheets of tela choroidea, located on the medial side of the body portion of the choroidal fissure in the roof of the third ventricle, below the body of the fornix, and between the superomedial surfaces of the thalami, is the velum interpositum [42].

The velum interpositum is usually a closed cavity that receives many veins from the frontal horn and the body of the lateral ventricle, which converge to form the internal cerebral veins. The internal cerebral veins arise just behind the foramen of Monro and exit the velum interpositum above the pineal body in order to enter the quadrigeminal cistern and join the great cerebral vein (vein of Galen) [42].

Endoscopic Technique

Transfrontal Approach to Lateral Ventricles

The procedure starts accessing the lateral ventricles and is performed under general anesthesia. The correct position of the head is critical to the technique's success. The head can be secured via a Mayfield head clamp or a horseshoe head holder in a neutral, slightly flexed position. A short linear or horseshoe precoronal skin incision is made. Rigid 0° viewing angle endoscopes with a working channel are commonly used for colloid cyst surgery [43].

The optimal position of the burr hole is a controversial topic. We prefer to use an entry point varying from 2 to 2.5 cm from the midline and from 0 to 1 cm anterior to the coronal suture.

In the literature, the ideal entry point varies from 2 to 7 cm from the midline and from 0 to 7 cm anterior to the coronal suture. An appropriate entry point and trajectory help to minimize the chance of leaving residual cyst and avoid iatrogenic injury to key anatomical structures such as the caudate nucleus, deep cerebral veins, and the fornices [44].

Rangel-Castilla et al. attempted to define an ideal entry point in order to get the best trajectory. Their results show that the optimal entry point varies laterally as the ventricles increase in size. In 90% of cases, it is located 4–6 cm anterior to the coronal suture [44].

Ibánez-Botella et al. suggested a frontal burr hole 1–2 cm anterior to the coronal suture and 3 cm lateral to the midline. The right side should be preferred if it provides a better approach to the lesion, always considering the asymmetries in ventricular size and lateralization of the colloid cyst. Neuronavigation provides an excellent alternative to locate the entry burr hole, especially in cases of small ventricles. However, as it is not universally available, the classic craniometric entry point is still useful [43, 44].

Other authors describe different techniques, such as the supraorbital approach reported by Delitala et al., consisting of an entry burr hole approximately 1.5 cm above the orbital rim in the mid-pupillary line, with a 15° angle from the perpendicular line in the coronal plane. They reported a better visualization of the roof of the third ventricle and lower risk of fornix injury because the foramen of Monro is approached perpendicularly and not tangentially [45].

Following the burr hole drilling, a freehand ventricular puncture is performed, creating a parenchymal corridor used to introduce the endoscope into the lateral ventricle. After dural coagulation and pial piercing, the rigid endoscope is advanced, with or without a peel-away sheath, toward the lateral ventricle, and the irrigation system with Ringer solution is started to improve visualization of the surgical field and identification of the anatomical landmarks [46].

Regardless of the chosen technique (transforaminal or transchoroidal approaches), the procedures consist of coagulation of the cyst capsule, puncture and aspiration of content, and en bloc resection of the lesion [43]. In our service, we coagulate the cyst capsule before the puncture. We cut the tip of a Fogarty catheter 4F at an oblique angle and connect it to a syringe in order to make a vacuum and initiate the aspiration process. After decompression, we proceed with the resection of the lesion.

Transventricular-Transchoroidal and Transventricular-Transforaminal Approaches

In the transventricular-transchoroidal technique, after entering the ventricle with the rigid endoscope, the opening of the choroidal fissure is performed with grasping forceps or with the coagulator introduced into the working channel of the endoscope through minimal medial to lateral movements within the endoscope. At this point, special attention must be directed to the position of the anterior septal-thalamostriate vein complex. In just under half of the cases, the confluence of this complex and the internal cerebral vein is located relatively posterior to the foramen of Monro, allowing the opening of the choroidal fissure without the need to coagulate and

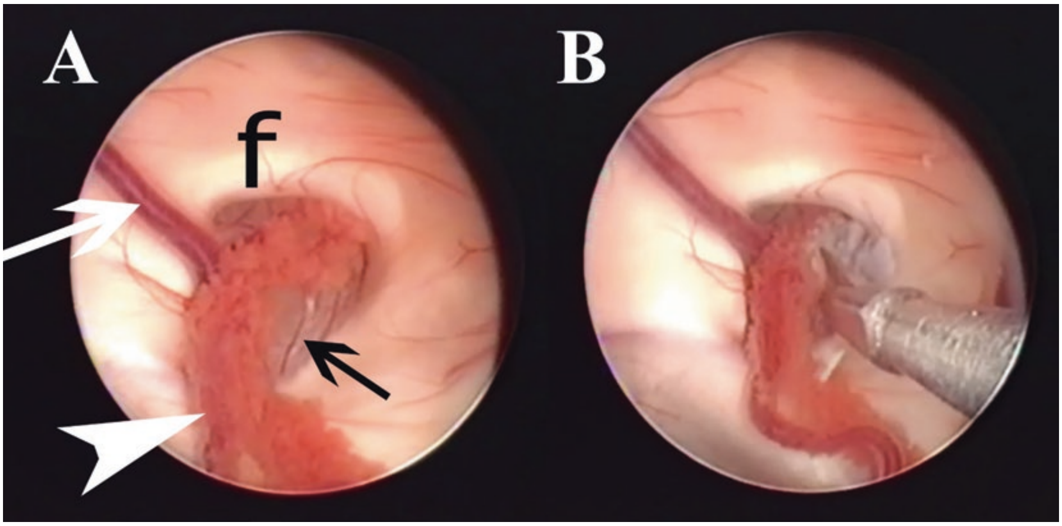


Fig. 22.10 (**a**, **b**). Endoscopic view of the foramen of Monro during a transventricular-transforaminal approach to a colloid cyst. (**a**) Ahead, the body and the columns of the fornix (*f*); coming from the medial wall of the frontal horn of the lateral ventricle, the anterior septal vein (*white arrow*); posteriorly, the choroid plexus and its superior choroidal vein (*white arrowhead*); the colloid cyst protruding through the foramen of Monro (*black arrow*). (**b**) Dissection and opening of the cyst capsule

section the anterior septal vein. However, when the confluence is adjacent to the foramen of Monro, coagulation and section of the anterior septal vein may be required before opening the choroidal fissure to avoid inadvertent rupture of the septal-thalamostriate and internal cerebral vein complex. In order to obtain better exposure of the third ventricle and its roof, the colloid cyst, and the internal cerebral vein, the choroidal fissure should be opened from the anterior septal vein to the posterior septal vein, through the tenia fornicis, between the choroid plexus and the body of the fornix [43]. A Fogarty catheter or forceps could be used to gently separate the choroid plexus from the fornix. The superior membrane of tela choroidea is then opened, and the contents of velum interpositum are exposed (medial posterior choroidal arteries and internal cerebral veins). Gentle maneuvers should be used to mobilize these structures for access and opening of the inferior membrane of the tela choroidea [46]. A transchoroidal technique is indicated only in cases with a small foramen of Monro or with a colloid cyst that protrudes minimally into the lateral ventricle [43].

The transventricular-transforaminal approach is the most commonly used approach, and it consists of resection of the colloid cyst through the foramen of Monro (Fig. 22.10a, b). It is a useful technique when the cyst sufficiently protrudes into the lateral ventricle [43, 46].

Resection Technique

Independent of the approach used, the resection technique consists of the same four steps: first, careful coagulation of the capsule and adherent choroid plexus and, as far as possible, dissection of the capsule from adjacent structures (anterior pillar of the fornix, septal-thalamostriate vein, choroid plexus). Thereafter, the capsule is opened or punctured, and the cyst contents are aspirated to enable subsequent manipulation. The third step consists of grasping the capsule with forceps, applying slow 360° angle twisting in a counterclockwise direction until a detachment of the capsule is achieved. Lastly, an en bloc removal of the lesion is performed [43].

In cases with intraventricular hemorrhage causing blood products in the sylvian aqueduct or intraventricular hemorrhagic products, although most hemorrhages can be well controlled via generous irrigation, prophylactic third ventriculocisternostomy (ETV) and/or CSF diversion via an external ventricular drain (EVD) can be performed [46].

Possible disadvantages of the transchoroidal approach are the risk of rupture of the anterior septal vein or internal cerebral vein during dissection or damage to the choroid plexus and fornix body. However, this technique allows a greater control over other important structures (body of fornix, internal cerebral vein, and choroid plexus) [43].

Iacoangeli et al. described the combined transforaminal-transchoroidal approach to be useful when the colloid cyst is firmly adhered to the tela choroidea or connected to the middle/posterior roof of the third ventricle, which makes it difficult for appropriate visualization of the insertion site of the cyst capsule. In this case, the transchoroidal route could be added to the classical transforaminal approach, as a second working channel providing complete exposure of the cyst and better visualization of the tela choroidea. The dual-route technique allows reduced traction on the foramen of Monro and avoids excessive traumatization of the fornix without increasing the risk of neurological complications [46].

Patient Selection

Many authors recommend conservative treatment for colloid cysts with a diameter less than 7 mm. Others recommend careful neurological and radiographic follow-up in cases when the cyst size is less than 10 mm without symptoms or signs of hydrocephalus or ventricular asymmetry due to the risk of sudden death [43].

Treatment options involve observation, shunting of CSF, stereotactic cyst aspiration, transcortical-transventricular microsurgery, transcallosal interforniceal or transforaminal microsurgery, and endoscopic ventricular surgery [47]. Among the surgical options, endoscopic surgery is considered by experienced endoscopic surgeons as a minimally invasive approach with less cortical injury, shorter hospital stay, satisfactory outcome, earlier return to the work, and reduced costs [48, 49].

The two most feared complications from observing asymptomatic colloid cysts are acute neurologic deterioration and sudden death. The rate of cyst growth, development of CSF obstruction, and the tendency for most colloid cysts to stop growing with increasing patient age are factors that influence the chance of cyst becoming symptomatic. Some authors argue watchful waiting if the patient remains lesions that are less than 10 mm and asymptomatic even in the presence of mild ventriculomegaly. However, if the patient becomes symptomatic, the cyst enlarges or hydrocephalus worsens, the interventional treatment should be promptly indicated [43, 47, 50]. On the other hand, some authors defend that even incidentally diagnosed cysts should be surgically treated due to the potential risks [47]. These patients, when treated using endoscopic surgery by experienced surgeons, tend to have a shorter hospital stay, greater likelihood of total removal, fewer complications, less need for a postoperative external ventricular drainage, and lower rate of recurrence than those with symptoms [51]. The presence of ventriculomegaly facilitates the endoscopic procedure. However, its absence does not contraindicate the use of endoscopic treatment [48].

The incidence of symptomatic progression in asymptomatic cysts treated with observation is estimated at 8% in 10 years [50, 51]. An increased T2 signal on magnetic resonance image (MRI) and an iso- and/or hypodense computed tomography (CT) probably reflect the ongoing cyst expansion. That could be used as surgical indication [51]. Unlike adults, colloid cysts in pediatric patients have a higher incidence of sudden neurological deterioration with potential clinical and radiographic worsening. For that reason, the surgery can be recommended early, even in some cases of incidental colloid cysts [52]. Symptomatic colloid cysts, due to hydrocephalus or local mass effect, are managed by surgical treatment. Even when asymptomatic, a large cyst blocking one or both foramina of Monro is usually surgically treated, because of the risk of sudden neurological deterioration [53].

The possibility of hemorrhagic changes within a colloid cyst may influence the treatment strategy. The cumulative lifetime risk of acute neurological deterioration or sudden death, in a 40-year-old, is estimated at 34% for an average life span of 78 years, ranging from 6% to 45% in the literature [47, 51]. Patients presenting with decreased mental status due to CSF obstruction represent a neurological emergency and should undergo emergent decompression, in order to avoid severe complications such as long-term disability or death. Ventricular shunting is the therapy for patients who will not have prompt access for surgical resection [47]. However, external ventricular drainage to emergently relief hydrocephalus, followed by surgical resection of colloid cyst in short period of time, should be preferred in order to avoid the need for permanent shunting.

Surgical Outcomes

Hellwig et al. reported 20 cases of colloid cysts endoscopically approached. Only 1 had recurrence after a 6-year follow-up, while 18 patients had improvement of symptoms soon after surgery [54]. Pinto et al. reported the use of stereotactic neuroendoscopic YAG laser in seven colloid cysts without recurrence [48].

Boogaarts et al. described 90 cases of colloid cysts treated by an endoscopic approach from 1994 to 2007, reporting total cyst removal in 41 cases (51.3%), near total removal in 5 cases (6.3%), and partial removal leading to a residual cyst in 34 cases (42.5%). According to the extent of cyst removal on magnetic resonance imaging, patients were classified into three groups: A (total cyst removal), B (persistent capsule but with isointense content to the CSF), and C (residual cyst with content signal different from CSF). Six cases needed a second endoscopic surgery, and just one needed a third intervention [39].

Detilala et al. reported a supraorbital transventricular endoscopic approach in seven patients, achieving complete removal in all of them and without cyst recurrence after 2 years of follow-up. All patients had improvement of hydrocephalus [45].

Hoffman et al. reported 58 patients who underwent endoscopic colloid cyst excisions with 45 showing total removal and only 4 with recurrence (6.9%), 3 of them belonging to the residual cyst group and 1 to the total removal group [55]. Chibbaro et al. described 29 cases of colloid cysts approached endoscopically, of which total resection was achieved in 25 cases (86.2%) and near total in 4 cases [40].

Ibáñez-Botella et al. used the Boogaarts classification to describe 24 colloid cyst surgical cases, 23 of them in the group A and only 1 in the group C [39, 43]. Iacoangeli et al. described five cases that underwent surgery with a combined endoscopic transforaminal-transchoroidal monoportal approach, without recurrence after an average follow-up of 68 months [46].

Complications

The transventricular endoscopic approach is considered to have several potential advantages in comparison to microsurgical interventions including shorter operative time and hospitalization, lower risk of cognitive and memory impairments due to forniceal injuries, lower risk of seizures due to cortex injury, and being less invasive. Nevertheless, there are some drawbacks reported such as difficulty in controlling bleeding, lower total excision rates, and risks encountered in cases when the cyst is adherent to the roof of the third ventricle [39, 40, 45].

In order to achieve appropriate hemostasis during endoscopic colloid cyst surgery, an irrigation system using Ringer solution should always be available and ready for periodic irrigation during the endoscopic procedure. Irrigation promotes the exchange of bloody CSF and helps with maintaining adequate endoscopic visibility. The outflow channel of the endoscope allows for normalizing the intraventricular pressure and intracranial pressure when long periods of irrigation are required for hemostasis [43].

Detilala et al. reported no complications in their seven patients who underwent supraorbital transventricular approach, neither of them requiring a ventricular shunt. Also, none of them presented with worsening of symptoms [45]. Hellwig et al. described 4 complications among 58 patients who underwent endoscopic colloid cyst surgery, 2 with transient memory deficits, 1 with surgical wound infection, and another with aseptic meningitis (treated with oral corticosteroids) [54].

Chibbaro et al. reported 3 complications among 29 surgical patients with patients experiencing intracerebral hemorrhage (requiring external ventricular drainage for 14 days) and another with aseptic meningitis who later developed obstructive hydrocephalus after 6 months and was treated with third ventriculostomy [40].

Ibáñez-Botella et al. described 24 patients who underwent endoscopic resection for third ventricular colloid cysts. Complications included two patients with surgical wound infections, one case of meningitis, three cases of intraventricular hemorrhage (one of which required temporary external ventricular drainage), two cases of permanent memory deficits cases, and two cases of transient memory deficits. They added that there were no statistically significant differences in complication rates when comparing the two endoscopic approaches (transventricular-transforaminal and transventricular-transchoroidal) [43].

PEARLS
The procedures consist of coagulation of the cyst capsule, puncture and aspiration of the cyst contents, and en bloc resection of the lesion.
Although the optimal position of the entry burr hole for endoscopic excision of colloid cyst remains as a controversial topic, we advocate an entry point 2.0–2.5 cm lateral to the midline and 0–1 cm anterior to the coronal suture. The right side should be preferred if it provides a suitable approach to the lesion.
Although transventricular-transchoroidal and transventricular-transforaminal approaches are comparable, the latter approach is more commonly performed for access to the colloid cyst, especially when the cyst sufficiently protrudes into lateral ventricle.
Symptomatic colloid cysts, due to hydrocephalus or local mass effect, should be managed by surgical treatment.
When the anterior septal-thalamostriate vein confluence is adjacent to the foramen of Monro, coagulation and section of the anterior septal vein may be required before opening the choroidal fissure to avoid inadvertent rupture of the septal-thalamostriate and internal cerebral vein complex.
A transchoroidal technique is indicated only in cases with a small foramen of Monro or with a colloid cyst that protrudes minimally into the lateral ventricle.
The combined transforaminal-transchoroidal approach is useful when the colloid cyst is firmly adherent to the tela choroidea or connected to the middle/posterior roof of the third ventricle, which makes it difficult for appropriate visualization of the insertion site of the cyst capsule.

Commentary on Case Presentation in Section 22.1

Presentation

A 42-year-old man presented to the neurology clinic with a 2-year history of headaches. The patient claimed to have no further history of headaches or other neurological symptoms. The headaches had worsened in the last month. At presentation, neurological examination was intact. MRI scans showed a lesion in the roof of the third ventricle that suggested a colloid cyst and a view evidencing the cyst protrusion through the foramen of Monro. (See Fig. 22.1a, b).

Discussion

This is a symptomatic adult patient presenting with a colloid cyst, diameter larger than 10 mm, and signs of hydrocephalus and mild ventricular asymmetry. As explained earlier, symptomatic colloid cysts, due to hydrocephalus or local mass effect, are considered candidates for surgical treatment. Endoscopic surgery is a very appropriate surgical technique for this clinical and radiographic scenario.

A transfrontal approach can be performed to access the lateral ventricles, preferring the right side if it provides a good approach to the lesion. As the cyst protrudes through the foramina of Monro, the transventricular-transforaminal approach can be used to resect the cyst. If the cyst is firmly adhered to the tela choroidea, a combined transforaminal-transchoroidal approach is the next step.

22.4 Colloid Cysts: Editors' Commentary

In the case provided (Fig. 22.1a, b), a relatively young man presents with progressive headaches and MRI evidence of a mass located within the anterior third ventricle causing obstructive hydrocephalus. There is symmetric ventricular enlargement suggesting obstruction of the foramina of Monro bilaterally. Although the decision of when to surgically intervene on asymptomatic patients with colloid cysts is extremely controversial, this patient is clearly symptomatic with headaches and imaging findings of hydrocephalus. Due to the potential for rapid neurologic decline and sudden death, urgent surgical intervention should be pursued in this clinical setting.

The transcortical-transventricular approach remains the most commonly practiced surgical intervention for colloid cysts. Microsurgical removal of the cyst has been associated with excellent rates of gross-total cyst removal and low rates of cyst recurrence. However, this approach carries an approximately 10% risk of postoperative seizures due to the size of the corticectomy [10]. As colloid cysts are nonneoplastic lesions, a variety of alternative surgical approaches have been utilized with the goals of acutely relieving the obstructive hydrocephalus, avoiding complications, and preventing future recurrence of hydrocephalus as well as the need for additional surgery. Due to the morbidity associated with the transcortical approach, stereotactic aspiration of the cyst contents was proposed in the 1980s as a "minimally invasive" alternative. However, likely due to the variability in the cyst internal contents and the failure to remove any of the cyst wall, cyst volume reduction was often suboptimal and recurrence rates were extremely high. Increasingly, the transcallosal interhemispheric approach has been utilized for colloid cysts as the only structure that needs to be disrupted is a small amount of the corpus callosum, and both sides of the lateral ventricle can potentially be accessed via a single approach. Similar to the transcortical approach, fine bimanual microdissection can be performed allowing the cyst to be delicately separated from the perilous diencephalic veins and fornices while carrying a lower risk of seizures. The major risk of this approach is injury to the bridging cortical veins leading to venous infarction, and the accessibility of an appropriate surgical corridor needs to be carefully determined preoperatively. In Sect. 22.2, the authors also present a variety of surgical extensions that can be utilized in patients with a small foramen of Monro or when the cyst cannot be adequately visualized upon entry into the lateral ventricle. These extensions require either partial disruption or retraction of the fornices potentially leading to memory deficits. Alternatively, one can avoid disrupting the fornices through the subchoroidal route, but this places the thalamostriate vein at risk. Although these techniques are rarely necessary in patients with symptomatic hydrocephalus, they may prove necessary for adequate cyst resection in asymptomatic patients with cysts that are enlarging.

Endoscopic resection of colloid cysts has gained rapidly increasing favor due to the ability to achieve cyst decompression and relief of hydrocephalus in a minimally invasive manner. Due to the relatively small size of most ventriculoscopes, typically 6–8 millimeter in diameter, cortical disruption is minimized, and the scope can be successfully maneuvered through the foramen of Monro into the third ventricle without causing injury to the ipsilateral fornix or venous complex. Criticisms of the endoscopic technique have appropriately focused on the relative inability to achieve gross total cyst resection due to the difficulties of performing bimanual cyst manipulation using the working channels of a coaxial endoscope.

Correspondingly, a large meta-analysis assessing the endoscopic approach demonstrated an extremely low risk of seizures (0.3%); however, gross total cyst resection was achieved in only 58% of patients overall [10]. With increased experience and improved instrumentation, the rate of gross total cyst resection has improved to >80% in modern series which further demonstrates the steep learning curve associated with minimally invasive approaches [5, 51]. As neuroendoscopic surgeons continue to push for greater extent of cyst resection, the critical question remains how much cyst wall is truly necessary to prevent recurrence without increasing the rate of potentially devastating complications. In our practice, we attempt to achieve the surgical goals of hydrocephalus relief and maximal safe cyst wall removal in as minimally invasive a manner as possible. Using the MR and CT imaging characteristics, the density of the cyst contents and the surgical instruments necessary for cyst decompression can be anticipated in the majority of patients. We typically approach the cyst initially using a 3.6 mm rigid endoscope with a working channel diameter of 1.6 mm (Little LOTTA®, Karl Storz, Tuttlingen, Germany) primarily designed for endoscopic third ventriculostomy. This comparatively thin scope allows us to safely work within even narrow ventricles and small foramina of Monro without causing injury as the endoscope is advanced into the third ventricle or due to torque movements. The restrictive working channel requires the use of smaller instruments; however, we have found that use of a side-cutting aspiration device (NICO Myriad®, Indianapolis, IN, USA; 19ga) designed specifically for this endoscope allows us to remove the cyst contents easily regardless of composition after careful coagulation of the choroid plexus overlying the cyst. The importance of approach trajectory cannot be overstated. We have found that the ideal trajectory is typically more anterior and lateral than Kocher's point and is planned with neuronavigation on an individual basis to provide access to the roof of the third ventricle. If the cortical entry point is too posterior, the working trajectory will be tangential to the anterior wall of the colloid cyst within the foramen of Monro which can make cyst puncture and decompression difficult as well as preclude resection of the posterior aspect of the cyst wall. We prefer a more lateral trajectory as this allows us to more thoroughly resect the contralateral wall of the cyst as well as perform a septum pellucidotomy to further minimize the likelihood of the patient developing symptomatic hydrocephalus in the future from potential cyst recurrence. In the rare event that the colloid cyst cannot be removed using the 3.6 mm endoscope, we typically begin upsizing to a 6 mm diameter endoscope which has an increased repertoire of working instruments. If bimanual microdissection proves necessary for either cyst removal or management of hemorrhage, we have a variety of mini-tubular retractors immediately available for conversion to microscopic technique. These tubular retractors range in diameter from 12 to 21 mm in diameter (BrainPath®, NICO Corp, Indianapolis, IN, USA; Vycor Medical, Bohemia, NY, USA) and allow for excellent visualization of the cyst using the microscope and standard microsurgical techniques. Utilization of narrow bayonetted bipolars and single shaft instruments further increases the working room within these narrow corridors. These mini-tubular retractors may represent a preferential relatively minimally invasive alternative to conventional microsurgical approaches for surgeons less comfortable with endoscopic techniques; however, no critical assessment of the extent of cyst resection or complication rates have been performed comparing the mini-tubular retractors to any of the other microsurgical or endoscopic approaches for colloid cysts [56].

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23 Intracranial Aneurysms

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Contents

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23.1 Intracranial Aneurysms: Editors' Introduction

The estimated prevalence of intracranial aneurysms is around 2–3% of the entire population [1, 2]. Most unruptured aneurysms present as incidental findings, but in a smaller subset of cases may present with transient ischemic attack or stroke, cranial nerve palsies, or other neurologic symptoms. Digital subtraction angiography remains the gold standard for detection of small aneurysms due to its sensitivity and the ability to visualize aneurysmal relationships to surrounding branch vessels. Computed tomography angiography (CTA) and magnetic resonance imaging (MRI) may play an increased role for cerebral aneurysms when endoscopic endonasal and keyhole approaches are considered as these imaging studies can provide important information on the relationship of the aneurysm to the skull base and cranial nerves. Open transcranial and endovascular approaches that are well established are not discussed in this chapter. Increasingly, keyhole and endoscopic approaches have been utilized for the treatment of ruptured and unruptured intracranial aneurysms and the indications, advantages, and disadvantages of each of these minimally invasive approaches will be discussed. The authors will also discuss the relative merits of their surgical approach to the case example below (Fig. 23.1a–e).

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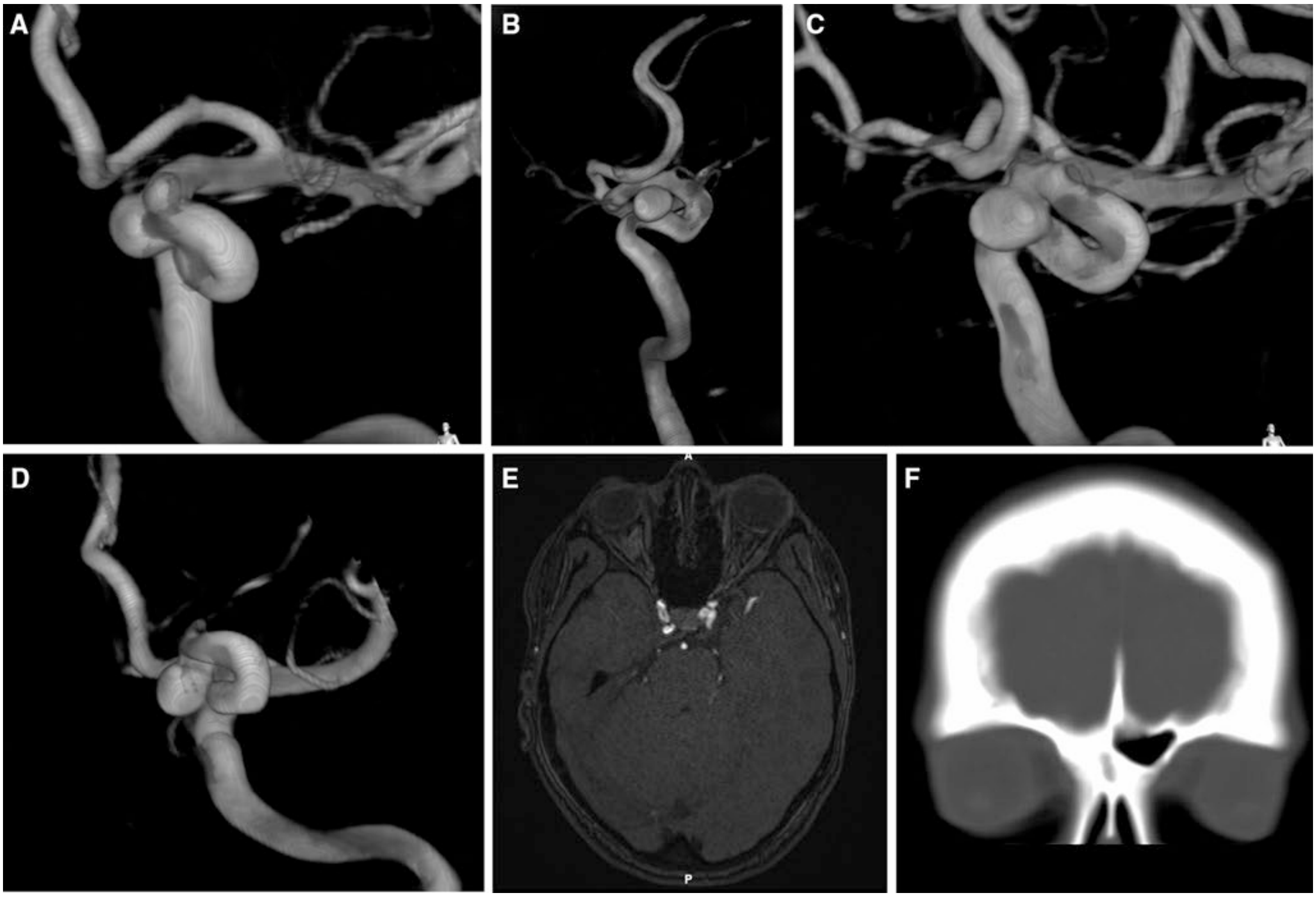

Fig. 23.1 (**a**–**f**) **CASE EXAMPLE**: intracranial aneurysm. A 58-year-old woman presents with chronic headaches and a family history of ruptured cerebral aneurysm and hypertension but is otherwise neurologically intact

23.2 Intracranial Aneurysms: Keyhole Transcranial Approaches

Nikolai J. Hopf and Robert Reisch

Introduction

Successful surgical treatment of aneurysms is technically demanding and has the potential for significant morbidity due to premature intraoperative rupture or ischemic events. Therefore, the majority of intracranial aneurysms are currently treated using endovascular techniques since this has been proven to be comparably successful but associated with a reduced morbidity and better short-term outcome when compared to standard surgical technique [3–5]. Minimally invasive neurosurgery (MIN), designed particularly to reduce approach-related morbidity, was generally thought not to be suitable for neurovascular procedures. However, it has been recently shown that MIN strategies can be successfully applied to treat a variety of unruptured as well as ruptured intracranial aneurysms [6–8].

Surgical Technique

The successful use of keyhole craniotomies is based on the understanding and consequent application of the keyhole principle [9]. When looking through a keyhole, even large objects can be seen completely when they are far enough away from the keyhole. In contrast, objects close to the keyhole are not well seen. Applied to aneurysm surgery, even large or giant aneurysms can be successfully treated using a keyhole approach when located at the cranial base. In contrast, more peripherally located large or giant aneurysms are not well suitable for a keyhole approach.

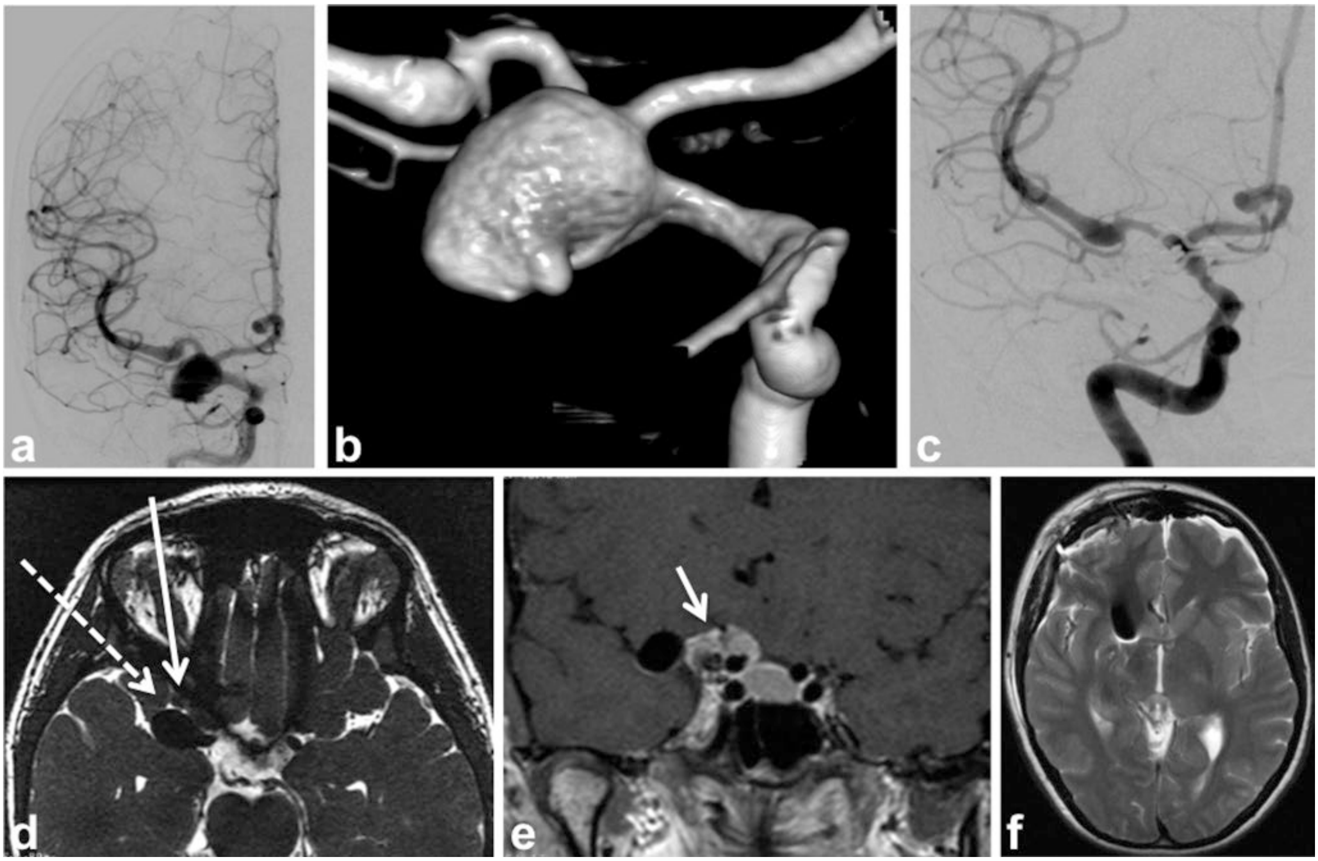

Fig. 23.2 (**a**–**f**) Digital subtraction angiography of a 28-year-old female with moderate visual impairment on the right side depicting a large aneurysm of the right internal carotid artery bifurcation in a standard anterior-posterior projection (**a**), a 3D reconstruction showing an irregular configuration with a baby aneurysm on the anterior medial surface (**b**), and after clip reconstruction through a right supraorbital keyhole craniotomy (**c**). MRI of the same patient demonstrating (**d**) on a high-resolution axial T2-weighted sequence the superiority of a supraorbital approach (*arrow*) compared to a transsylvian approach (*interrupted arrow*) to visualize and treat the aneurysm neck and (**e**) on a T1-weighted post-contrast sequence a meningioma as the probable cause of the visual impairment. (**f**). Postoperative MRI of the same patient on postoperative day 1 after the surgery demonstrating the supraorbital keyhole craniotomy on the right side and no surgical complications

Following this principle, the vast majority of unruptured and ruptured aneurysms can be successfully treated using keyhole craniotomies. One major restriction is that the trajectory to the lesion cannot be changed during the operation because of the limited opening. Therefore, thorough preoperative diagnostics and the liberal use of endoscopes to overcome this limitation are mandatory.

Diagnostic studies should include an MRI, MR angiography (MRA), as well as a high-resolution superselective catheter digital subtraction angiography (DSA) with 3D reconstruction (Fig. 23.2a–c). The DSA is recommended to make the decision of the preferred treatment modality (i.e., surgical or endovascular). It also provides additional important information on flow dynamics for detailed surgical planning and anticipation of intraoperative challenges. The MRI is important to determine the best approach and to recognize the exact anatomical relationship between the aneurysm and the involved cisterns, vessels, nerves, skull base, and the brain. Therefore, it should include high-resolution T2-weighted images, preferably a CISS sequence (Fig. 23.2d–f). Recently, contrast enhancement of the aneurysm wall depicted by high-resolution MRI is thought to demonstrate instability of aneurysms [10]. In selected aneurysms with close relation to the skull base, preoperative evaluation should include high-resolution CT imaging.

Specific pre- or intraoperative measures for brain relaxation are not necessary as a routine using keyhole approaches. However, early access to the cisterns followed by slow and patient removal of adequate amounts of cerebrospinal fluid (CSF) is important for the successful use of keyhole approaches. The subtemporal keyhole approach (STKA), in particular on the dominant side, is the only keyhole approach where a perioperative lumbar drain seems to be helpful to gain early and adequate brain relaxation. In patients with an acute subarachnoid hemorrhage (SAH), opening of the intraventricular space toward the outer CSF spaces is often very helpful, for example, via the lamina terminalis. With adequate positioning and early CSF egress, brain retractors are generally not necessary. Even before opening the cisterns or in patients with an acute SAH, the tendency of the brain to herniate seems to be less with keyhole approaches compared to standard craniotomies.

All minimally invasive cerebrovascular procedures are performed under multimodal electrophysiological monitoring consisting of somatosensory-evoked potentials (SEP) and motor-evoked potentials (MEP) for all supratentorial aneurysms and additionally continuous electromyography (EMG) of the according cranial nerves for complex and large aneurysms of the proximal internal carotid artery (ICA), posterior communicating artery (PCoA), and all infratentorial aneurysms [11]. This is particularly important during temporary clipping or transient cardiac arrest. Furthermore, indocyanine green (ICG) angiography and micro-Doppler sonography are used in the majority of cases. Intraoperative angiography is required only in select complex cases, independent of the use of keyhole or standard approaches.

The liberal use of endoscopes is mandatory to overcome the inability to visualize the aneurysm from different angles when using limited keyhole craniotomies. The preferred technique is endoscopic-assisted microsurgery (EAM), meaning that the endoscope is used intermittently during specific stages of the operation [12]. The goal is to gain optimal visual information of the individual anatomy as well as particular pathology that is not provided by the straight visual trajectory of the operating microscope (Fig. 23.3a–f). Therefore, in most instances, a 4 mm endoscope with a 30° viewing angle is used. In some instances, the 2.7 mm endoscope with a slightly inferior image quality is more suitable, particularly in cases of small surgical windows (e.g., the optico-carotid window when approaching the basilar artery) or large aneurysms obstructing the operating field. Because of the relatively short time of the use, fixation of the endoscope is generally not necessary. However, an ergonomic setup with the endoscopic image depicted on a monitor in straight vision for the surgeon is mandatory to avoid mechanical or thermal damage with the endoscope. Full-endoscopic technique for transcranial procedures should be applied only in highly selected cases as long as stereoscopic vision with adequate resolution is not provided by endoscopes. Therefore, the operating microscope is still the preferred imaging tool for transcranial aneurysm surgery, even when using keyhole approaches.

In addition to the routine use of endoscopes, specific surgical instruments are required for unrestricted and meticulous dissection when using keyhole craniotomies. Therefore, delicate tube shaft instruments are recommended rather than standard microsurgical instruments. In addition, modified clips and clip appliers with minimal obstruction of the surgeon's view along the instrument and the ability to reach around corners or follow non-straight trajectories are of great help. This can be achieved, for example, with a clip applier that connects from inside the clip lock, a reduced size of the clip lock (Fig. 23.4), and the use of malleable materials.

Proximal and distal vascular control is mandatory for a safe and successful procedure. This is well achieved for most aneurysms of the anterior circulation using a supraorbital keyhole approach (SOKA) because it is a subfrontal rather than a transsylvian approach. This implies that proximal control of the intracranial ICA, anterior cerebral artery (ACA), and middle cerebral artery (MCA) can be achieved very early on without excessive opening of the Sylvian fissure, which is regularly necessary when using a standard pterional approach. Gaining proximal control of supra- and paraclinoidal ICA aneurysms is difficult through keyhole as well as standard approaches. Rather than drilling large amounts of the skull base, proximal control may be achieved by an endovascular balloon catheter. For large or giant aneurysms, deflation of the aneurysm by the suction-decompression technique or short-term cardiac arrest using adenosine as well as the use of endoscopes to inspect the aneurysm from all sides is mandatory.

Intraoperative premature rupture is managed similar to standard craniotomies by (a) control of the bleeding site using one or two suction devices, (b) optional temporary clipping of the feeding vessels and/or rupture site, (c) completion of the dissection of the aneurysm neck, and (d) definite clipping of the aneurysm. Anticipation of this situation and preparation of appropriate temporary clips is the key to minimize blood loss and occlusion time of relevant vessels. Temporary cardiac arrest achieved by adenosine is an additional, very effective option to deal with intraoperative rupture in case of inadequate proximal control or when managing large and complex aneurysms.

Postoperative care includes short-term postoperative intensive care observation. Patients are generally mobilized 4 h after surgery. Postoperative non-contrast-enhanced CT imaging to screen for complications and a catheter angiography to confirm complete occlusion of the aneurysm are recommended in patients with no intraoperative imaging. Any perioperative medications (e.g., antiepileptic drugs, steroids, or antibiotics) are optional.

Suitable keyhole approaches for aneurysm surgery are the supraorbital keyhole approach (SOKA), the pterional keyhole approach (PTKA), the interhemispheric keyhole approach (IHKA), the subtemporal keyhole approach (STKA), the retrosigmoid keyhole approach (RSKA), and the median inferior suboccipital keyhole approach (MISKA). The most frequently used keyhole approach in our own series for aneurysms is the SOKA.

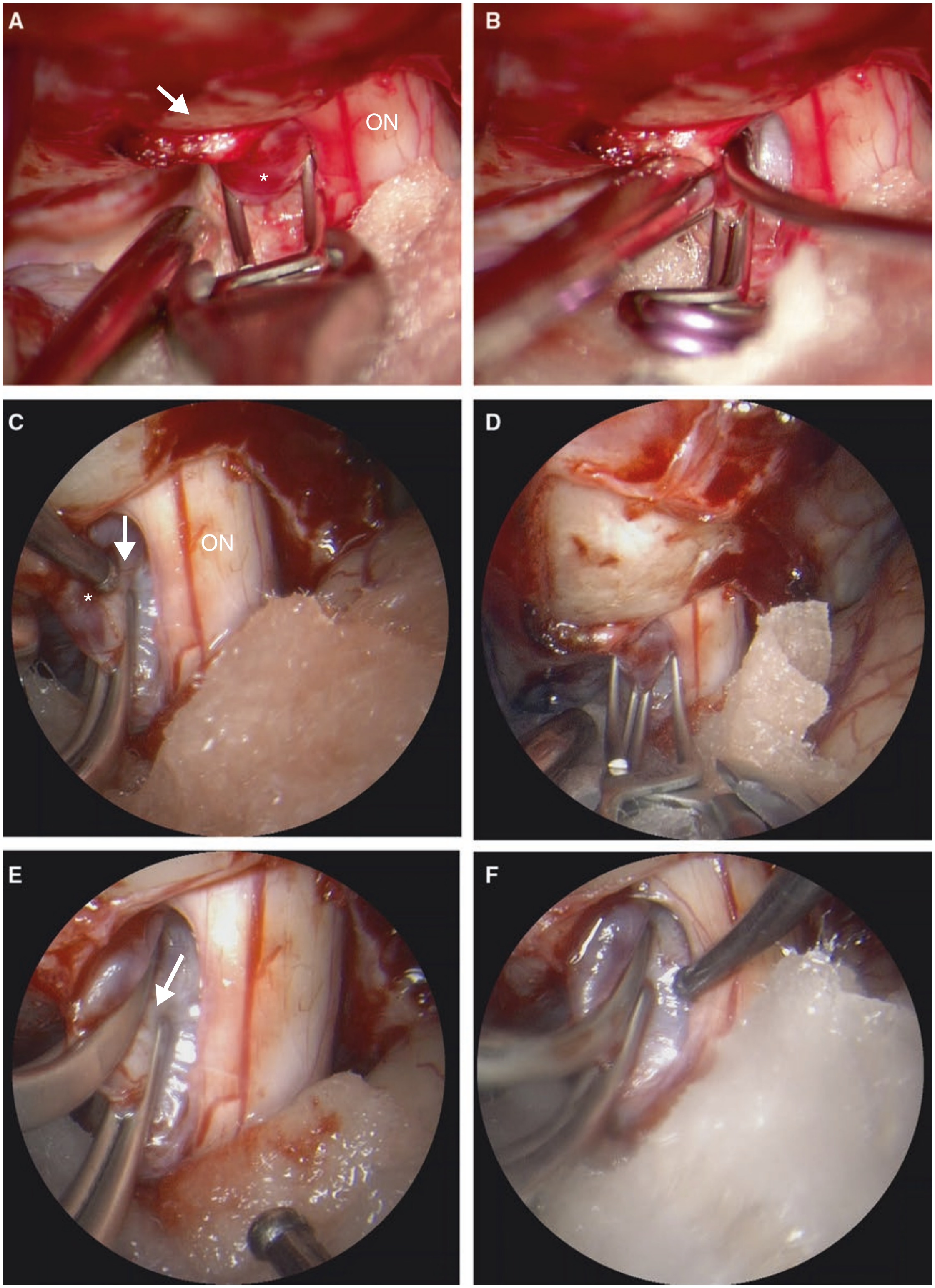

Fig. 23.3 (**a**–**f**) Clipping of a paraophthalmic aneurysm of the left internal carotid artery via a left supraorbital keyhole craniotomy demonstrating the limited visualization of the aneurysm neck with the operating microscope during (**a**) and after (**b**) clipping despite partial removal of the anterior clinoid process (*arrow*), endoscopic view demonstrating incomplete clipping of the neck (**c**) (*arrow*), placing of a second clip under endoscopic control (**d**), a persisting small remnant of the neck (**e**) (*arrow*), and complete occlusion after clip repositioning (**f**). *ON* optic nerve, * aneurysm

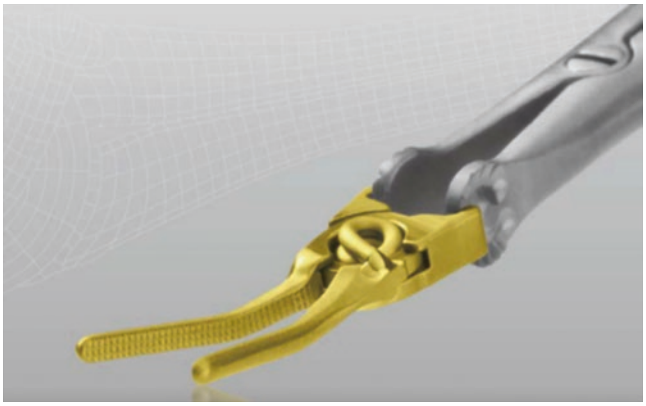

Fig. 23.4 Clip applier for minimal invasive aneurysm surgery with a reduced lock size and the applier connecting from inside the clip (Courtesy of Peter Lazic Inc., Tuttlingen, Germany)

Supraorbital Keyhole Approach

The supraorbital keyhole approach (SOKA) is performed with the patient in a supine position. The neck is slightly extended with the eyebrow being the highest point, and the head is rotated between 15° and 45° to the contralateral side providing a perpendicular view to the aneurysm. The skin incision does usually not have to exceed 3–4 cm in length and should be within the eyebrow for cosmetic reasons just lateral to the supraorbital foramen extending to the lateral margin of the eyebrow. Then, the junction of the frontal muscle, orbicularis oculi muscle, and temporal muscle is exposed, and a pericranial flap consisting of the muscle and periosteum is created using an incision parallel and approximately 2 cm superior to the orbital rim, but not exceeding the superior temporal line laterally to avoid injury of the facial nerve. The flap is completed by cutting along the superior temporal line in an inferior direction (Fig. 23.5a). The temporal muscle is then detached from the temporal line and a burr hole made as close as possible to the cranial base lateral to the superior temporal line (Fig. 23.5b). The craniotomy is performed by first cutting toward medially and parallel to the frontal skull base and completed by a C-shaped cut starting again from the burr hole and ending at the medial end of the first cut (Fig. 23.5c). The size of the craniotomy is typically 1.5 × 2.5 cm. After elevation of the bone flap, the dura is elevated from the cranial base to drill down the inner edge and bony protrusions (jugae) of the orbital roof. The dura is then opened in a C-shaped manner toward the skull base. The initial dissection is along the sphenoid ridge toward the optic nerve. The arachnoid is opened lateral to the optic nerve to gain broad access to the basal cisterns. After removing adequate amounts of the CSF, the brain will fall back usually enabling further dissection without the need for brain retractors. However, for aneurysms of the anterior communicating artery (ACoA), retractors may be used intermittently to prevent partial removal of the gyrus rectus to achieve adequate dissection and safe clipping.

Closing should be performed only after meticulous elimination even of minor venous oozing, and the intradural space should be filled with irrigation fluid or artificial CSF. Tack-up sutures are generally not necessary due to the small size of the craniotomy. However, watertight closure of the dura is recommended to prevent extensive postoperative swelling, discomfort, or wound healing disturbances. Because the craniotomy is not covered by hair, re-fixation of the bone flap and coverage of the burr hole with mini-plates, filling of the bony gap with bone cement or collected bone dust, and tight subcuticular suture are key aspects for a perfect cosmetic outcome.

Patient Selection

Keyhole craniotomies can be used in all patients, independent on age, location of the aneurysm, presence of SAH, or comorbidities. The particular keyhole craniotomy is selected based on the location and configuration of the aneurysm as well as the individual anatomy of the patient. The most versatile approach is the SOKA, which may be used for aneurysms of the A1 and proximal A2 segment of the anterior cerebral artery (ACA), anterior communicating artery (ACoA), internal carotid artery (ICA), posterior communicating artery (PCoA), middle cerebral artery (MCA) (Fig. 23.6), distal basilar artery (BA), and proximal superior cerebellar artery (SCA) and the P1 or proximal P2 segment of the posterior cerebral artery (PCA). The PTKA can be used alternatively to the SOKA but is associated with more manipulation of the temporal muscle and thus postoperative discomfort and swelling. However, inferiorly pointing PCoA aneurysms or MCA aneurysms well inferior to the sphenoid wing are better visualized using a PTKA. The IHKA may be used for selected aneurysms of the ACoA and is recommended for aneurysms of the A2 and distal segments of the ACA. The STKA is most frequently used for selected aneurysms of the BA tip or PCA located at the P2 segment and distally. The RSKA may be used for aneurysms of the proximal BA, anterior inferior cerebellar artery (AICA), and distal vertebral artery (VA) and selected aneurysms of the posterior inferior cerebellar artery (PICA). The MISKA is used for aneurysms of the proximal VA and PICA (Table 23.1).

Following the keyhole principle, even large or giant aneurysms located at the cranial base, for example, arising from the ICA or ACoA, can be successfully treated using a SOKA (Fig. 23.6) or PTKA. However, more peripherally located large or giant aneurysms, in particular those arising from distal branches of the MCA, are not well suitable for a keyhole approach.

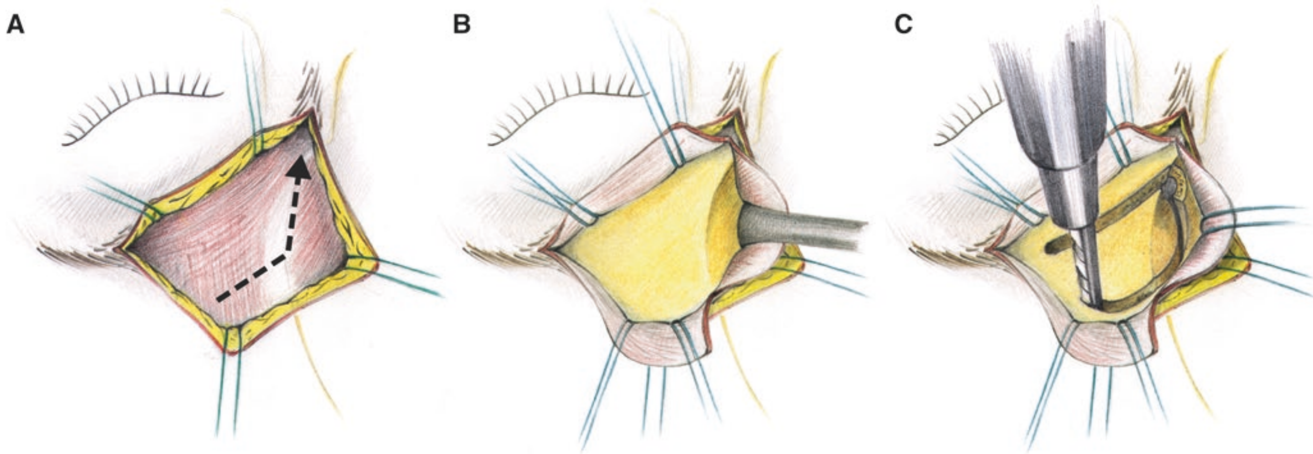

Fig. 23.5 (**a**–**c**) Drawing of the surgical steps of a right-sided supraorbital keyhole craniotomy showing the cutting line (*arrow*) in the frontalis muscle perpendicular to its fibers from medial to lateral but turning inferiorly when reaching the superior temporal line (**a**), detaching the temporal muscle from the superior temporal line (**b**), and performing the craniotomy starting from the burr hole just lateral to the temporal line and performing first the inferior cut before completing the craniotomy in a C-shaped manner from lateral to medial (**c**) (Copyright retained by Dr. Robert Reisch)

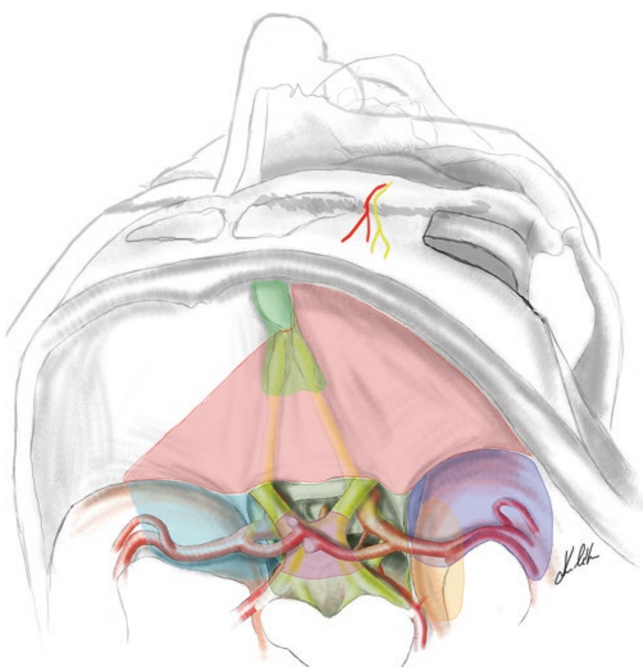

Fig. 23.6 Drawing demonstrating the accessible vascular structures through a right-sided supraorbital keyhole craniotomy (Courtesy of Peter Kurucz, MD, Stuttgart, Germany, and Budapest, Hungary)

Table 23.1 Keyhole approaches for aneurysms

Keyhole approach	Abbreviation	Target vessels
Supraorbital	SOKA	ACA (A1, prox. A2), ACoA, ICA, PCoA, MCA, dist. BA, prox. SCA, PCA (P1, prox. P2)
Pterional	PTKA	ACA (A1, prox. A2), ACoA, ICA, PCoA, MCA, dist. BA, prox. SCA, PCA (P1, prox. P2)
Subtemporal	STKA	Dist. BA, PCA (P2 and ff)
Anterior interhemispheric	IHKA	ACoA, ACA (dist. A2 and ff)
Retrosigmoid	RSKA	Prox. BA, AICA, dist. VA, PICA
Median inferior suboccipital	MISKA	Prox. VA, PICA

A1 first segment of the anterior cerebral artery, *A2* second segment of the anterior cerebral artery, *ACA* anterior cerebral artery, *ACoA* anterior communicating artery, *AICA* anterior inferior cerebellar artery, *BA* basilar artery, *ICA* internal carotid artery, *MCA* middle cerebral artery, *P1* first segment of the posterior cerebral artery, *P2* second segment of the posterior cerebral artery, *PICA* posterior inferior cerebellar artery, *SCA* superior cerebellar artery, *VA* vertebral artery, *dist*. distal, *prox*. proximal, *ff* following

Illustrative Case

A 45-year-old otherwise healthy female suffered from recurrent episodes of headaches. MRI was suspicious for a MCA aneurysm on the right side. DSA confirmed the aneurysm at the origin site of the early temporal branch of the M1 segment with a broad neck and a size of 9 mm (Fig. 23.7a). A second aneurysm was seen between the SCA and PCA on the right side (Fig. 23.7b). Indication for treatment was based on the young age as well as a high rupture risk due to location (posterior circulation), size (MCA aneurysm >7 mm), multiplicity (SCA and MCA aneurysms), and additional risk factors (smoker). Surgical clipping was offered for both aneurysms via a single subfrontal approach because of the high location of the basilar tip (Fig. 23.7c). Endovascular treatment was thought to be well suitable only for the SCA aneurysm because of the broad neck and mismatch between

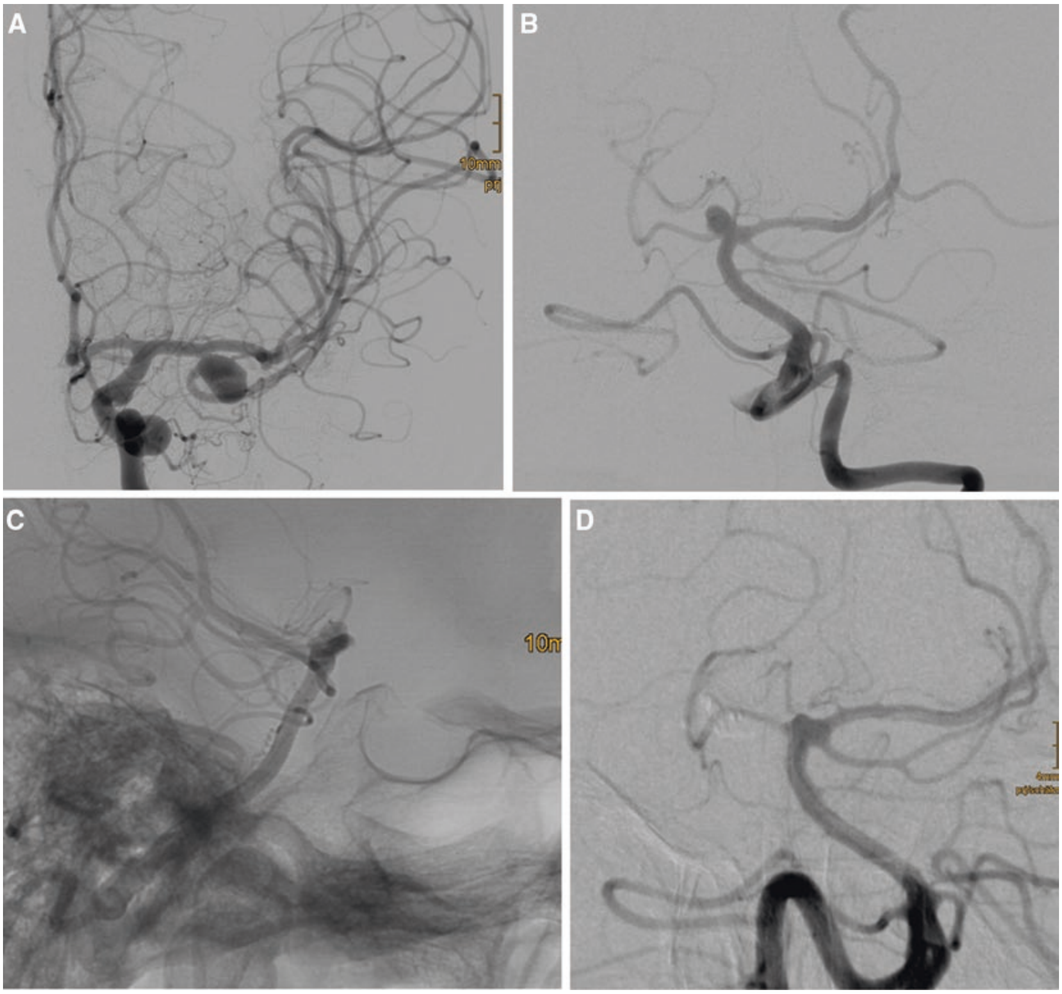

Fig. 23.7 (**a**–**d**) Angiography demonstrating an aneurysm arising from the middle cerebral artery at the origin of the early temporal branch on the right side (**a**) and the superior cerebellar artery (**b**), with the basilar artery tip being well above the dorsum sellae (**c**). (**d**) Angiography of the posterior circulation demonstrating complete occlusion of the aneurysm

the small size of the carrying vessel compared to the size of the MCA aneurysm. Surgical clipping of both aneurysms was attempted via a right-sided SOKA. After straight forward clipping of the MCA aneurysm, only the neck of the SCA aneurysm could be visualized through the optico-carotid window using the operating microscope (Fig. 23.8a). A 4 mm 0° endoscope was used to provide a comprehensive overview of the entire pathology and its relation to the surrounding structures (Fig. 23.8b). The definite clipping was achieved under the stereoscopic image of the operating microscope (Fig. 23.8c) followed by close endoscopic inspection confirming complete occlusion of the aneurysm and no impairment of any perforators (Fig. 23.8d). The postoperative course was uneventful. On discharge, the patient was in good general condition and neurologically intact. Postoperative DSA confirmed complete occlusion of both aneurysms (Fig. 23.7d).

Surgical Outcomes

Surgical outcome using keyhole craniotomies is at least comparable with that achieved with standard or extended craniotomies. Fischer et al. reviewed 1000 consecutive patients in which 1297 aneurysms were surgically treated in 1062 operations. The outcome from 942 keyhole craniotomies was compared to the outcome of 120 operations where standard or extended craniotomies had been used in the same institution [6]. In this series, 61.5% of the patients presented with ruptured aneurysms. The authors concluded that keyhole craniotomies are suitable for adequate treatment of unruptured as well as ruptured aneurysms, showing a trend for fewer complications in the keyhole craniotomy group. Reisch et al. confirmed these findings recently by analyzing outcomes in 793 patients treated with endoscopic-assisted microsurgical (EAM) technique on 989 aneurysms using a

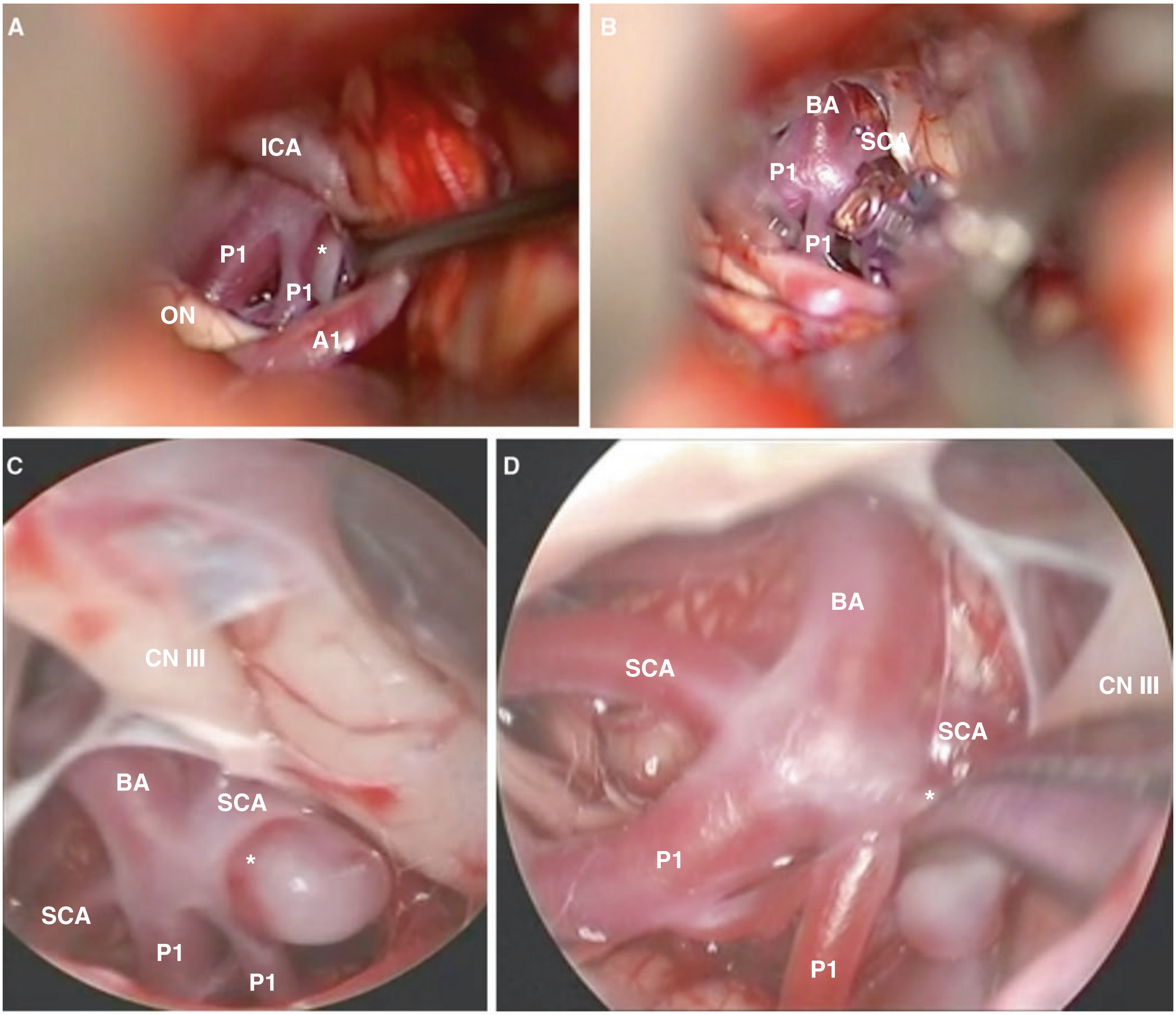

Fig. 23.8 (**a**–**d**) Intraoperative view through the operating microscope via a right supraorbital keyhole craniotomy of the basilar tip and only aneurysm neck before (**a**) and after (**b**) clipping and endoscopic view of the same situation demonstrating a much better overview in the depth before (**c**) and after (**d**) clipping. *A1* first segment of the anterior cerebral artery, *BA* basilar artery, *CN III* cranial nerve III, *ICA* internal carotid artery, *ON* optic nerve, *P1* first segment of the posterior cerebral artery, *SCA* superior cerebellar artery, * aneurysm neck

SOKA [8]. Similarly, 60% of the patients suffered from a SAH. Patients with unruptured aneurysms showed good outcome in 96.5% defined as a score ≤2 on the modified Rankin scale. A current study on the outcome after clipping of unruptured aneurysms using standard craniotomies reports good outcomes in 73.2% of patients treated in ten high-volume academic medical centers in New York state with a variation between 44.6% and 91.1% [13]. In this study, good outcome was defined as discharge to home under self-care.

Potential limitations that may arise with the use of keyhole craniotomies, such as the inability to view the aneurysm from different angles, can be overcome by using endoscopes. Accordingly, endoscopes provided unique visual information that led to clip repositioning in 19.1% of 793 operations using EAM technique [8]. In addition, endoscopes enable the successful use of keyhole craniotomies for patients with multiple aneurysms. This is particularly important if there is no matching best craniotomy for all the aneurysms intended to treat. In a series of 15 patients with bilateral MCA aneurysms, complete occlusion of all aneurysms could be achieved with EAM technique in 10 patients using a unilateral SOKA [14]. Three of these patients had ruptured aneurysms. The outcome was good in all ten patients. Permanent morbidity consisted of anosmia in one patient and hyposmia as well as a minor visual field deficit in another patient.

Concerning postoperative cosmetic results, discomfort, and morbidity, 94.8% of 375 patients treated via a SOKA for various lesions were very satisfied (84%) or satisfied (8.8%) with the cosmetic result [15]. No pain or discomfort was reported by 77% and only little discomfort in further 12.3% of the patients. Morbidity included frontal hypesthesia (initially 8.3%, permanent: 3.4%), hyposmia (initially 2.9%; permanent, 2.1%), and frontalis muscle palsy (initially 5.6%; permanent, 2.1%) in this particular group of patients.

PEARLS
Keyhole craniotomies are suitable for unruptured and ruptured intracranial aneurysms of any location
Anticipation of the detailed individual relation of the aneurysm to the surrounding structures is mandatory, because the trajectory of approach cannot be changed during surgery when using limited openings
Specific single tube shaft instruments and endoscopes help to overcome limitations of keyhole craniotomies
Early CSF egress from the basal cisterns is the key for adequate brain relaxation and generally prevents the need for brain retractors in aneurysm surgery
The supraorbital keyhole approach (SOKA) is the most versatile keyhole approach for intracranial aneurysms and enables early control of the feeding vessel in aneurysms of the anterior circulation
Cosmetic results are generally very good, and complications tend to be less using keyhole craniotomies compared to standard approaches

Commentary on Case Presentation in Chapter 23

Discussion

In the case example (Fig. 23.1a–d), the aneurysm is located at the supraclinoid left ICA, arising from the area of origin of the superior hypophyseal artery and pointing medially. This implies that the aneurysm dome may extend medially to the ipsilateral optic nerve. This relationship can be best confirmed on a T2-weighted high-resolution MRI.

Suitable keyhole approaches for transcranial surgical treatment of this particular aneurysm are the contralateral PTKA and contralateral SOKA providing a fairly straight trajectory to the aneurysm through the prechiasmatic window (Fig. 23.9a, b). Contraindications of a contralateral trajectory would be a prefixed chiasm or upward bulging of the skull base in the region of the tuberculum sellae or planum sphenoidale. Both unfavorable factors are not present in this particular case. A large frontal sinus would be in favor for a PTKA. This is also not present. Thus, both keyhole approaches may be used. However, adequate overview in the hidden area of the anterior part of the aneurysm neck will be difficult with either approach. Therefore, endoscopic-assisted technique is recommended.

An important issue to consider is proximal control in these ICA aneurysms. This cannot be easily achieved with either approach. Therefore, extracranial dissection of the ICA at the neck or installation of a balloon catheter for the time of the surgical procedure has to be considered. In addition, instruments suitable for keyhole approaches, in particular the clip system, will be of great help since the approach through the prechiasmatic window is very narrow and the final position of the clip lock is anticipated to be in close proximity to the left optic nerve (Fig. 23.9c).

In summary, a SOKA on the contralateral right side is the preferred approach over the PTKA for minimal invasive surgical treatment of this particular left-sided ICA aneurysm because of the less invasive character and superior cosmetic outcome of this particular approach. Endoscopic-assisted technique is strongly recommended to achieve adequate visualization of the entire aneurysm neck and a balloon catheter within the extracranial ICA for proximal control.

23.3 Intracranial Aneurysms: Endoscopic Endonasal Approaches

Pradeep Setty, Juan C. Fernandez-Miranda, Eric W. Wang, Carl H. Snyderman, and Paul A. Gardner

Introduction

Intracranial aneurysm clipping via an endoscopic endonasal approach (EEA) was first described in the literature by Kassam et al. in 2006 [16]. In this instance, an EEA was successfully utilized to gain access to a large, previously coiled, unruptured vertebral artery aneurysm to both trap it with aneurysm clips as well as to decompress the adjacent brainstem from the significant mass effect via aneurysmorrhaphy. Since this early report, several authors have subsequently published numerous cases of successful clipping of selected intracranial aneurysms through an EEA [17–25]. These reports have encompassed aneurysms of various sizes and locations, including those from both the anterior and posterior circulation as well as both ruptured and unruptured aneurysms. In our experience, the EEA when utilized by experienced surgical teams in well-selected cases can allow for successful clipping of intracranial aneurysms that would otherwise be extremely challenging to access and treat.

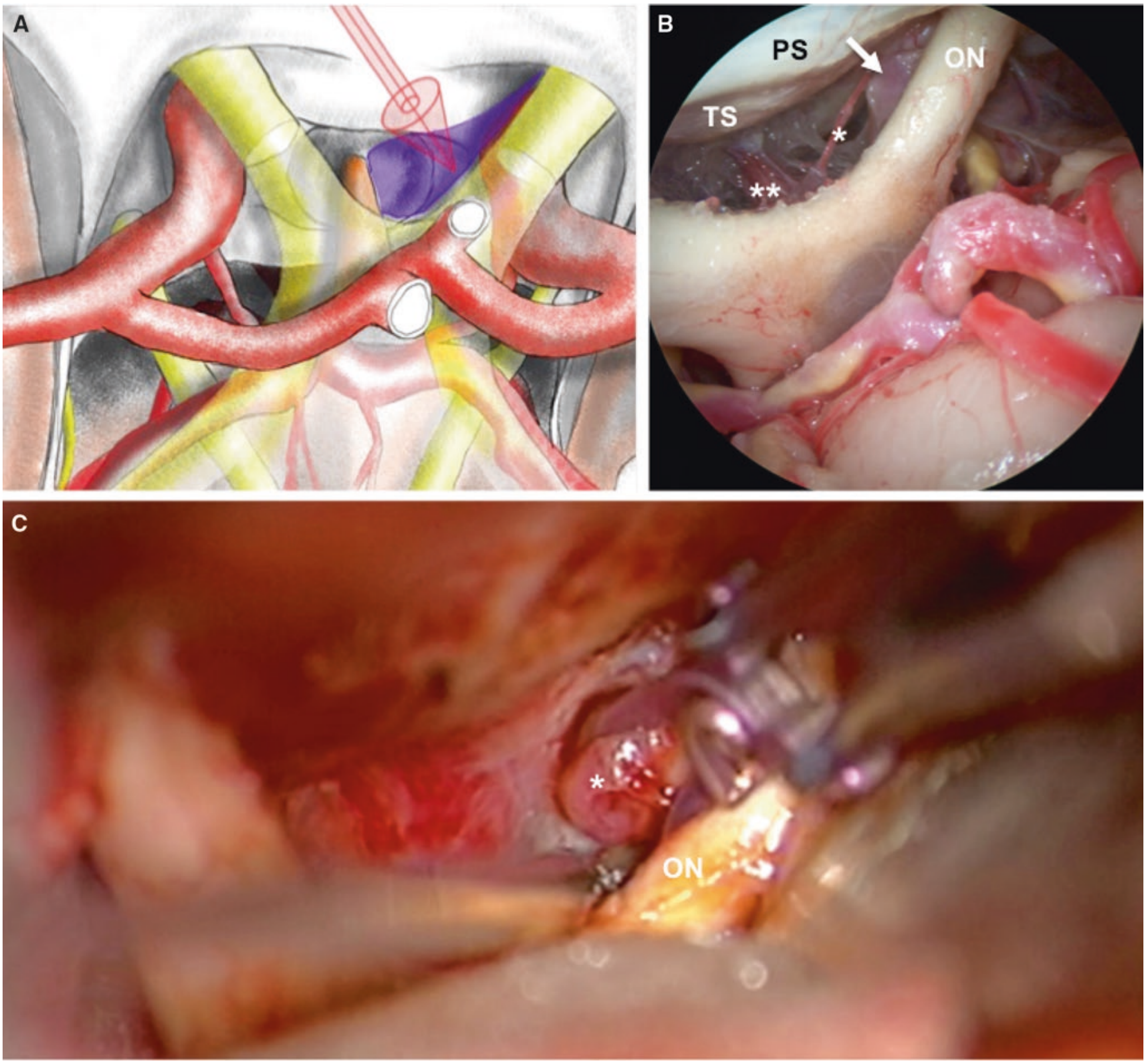

Fig. 23.9 (**a**) Drawing of the contralateral approach to the medial aspect of the internal carotid artery. (**b**) Endoscopic picture demonstrating the mesial wall of the proximal contralateral internal carotid artery (*arrow*) in a fresh cadaver dissection. (* superior hypophyseal artery, ** pituitary stalk, *ON* optic nerve, *PS* planum sphenoidale, *TS* tuberculum sellae). (**c**) Intraoperative view showing surgical clipping of an aneurysm of the medial aspect of the internal carotid artery through a contralateral supraorbital keyhole craniotomy in a patient with an acute subarachnoid hemorrhage (*ON* optic nerve, * aneurysm)

The endonasal corridor is generally limited to ventral aneurysms near midline, such as paraclinoidal aneurysms with a medial trajectory [17, 21, 24]. The main advantage in these cases is that EEA allows for a more direct surgical corridor to the aneurysm, eliminating the need for any brain retraction (or even contact) and allowing the surgeon to minimize or even eliminate any manipulation of the complex network of adjacent and often intervening neurovascular structures in the subarachnoid space, such as the optic nerve. The EEA also allows for early identification and access to proximal vessels (e.g., cavernous internal carotid artery [ICA], basilar trunk), allows for improved vascular control, and provides optimal visualization of the operative field due to the superior illumination of deep structures provided by the endoscope, potentially allowing the surgeon to better identify and avoid the small adjacent perforating arteries that can create devastating complications if inadvertently clipped with the aneurysm. Contrary to popular belief, the EEA is not contraindicated for ruptured aneurysms, as several reports exist of successful endoscopic clipping in the setting of subarachnoid hemorrhage [17, 21–24]; however, intraoperative rupture is more likely and needs to be considered [26, 27]. Finally, EEA provides excellent cosmetic results (due to the lack of skin incision), shorter operative times, and potential for easier postoperative recovery, thereby shortening overall hospitalization.

Despite these successful reports, many surgeons remain hesitant due to the fact that traditional, open transcranial approaches for cerebrovascular lesions are well studied and have a long, well-documented history of being safe and effective with acceptable morbidity and mortality rates. In addition, most cerebrovascular surgeons are far more familiar and comfortable with the anatomy and surgical techniques of open transcranial surgery. Further concerns regarding the feasibility of performing the delicate maneuvers required of cerebrovascular surgery through the long, deep, and narrow endonasal corridor have also discouraged some from utilizing EEAs for aneurysm clipping. In many cases, the lack of availability and the design infancy of appropriate instrumentation, such as endoscopic clip appliers, have limited neurosurgeons from attempting such cases. One great fear of many neurosurgeons is managing and controlling vascular catastrophes, such as intraoperative aneurysm rupture or inadvertent carotid artery injury [26, 27]. Furthermore, the two-dimensional visualization provided by the endoscope may also create difficulty for surgeons with limited experience in endoscopy. Finally, protrusion of aneurysm clips into the sphenoid sinus can hinder reconstruction of the skull base and increase the likelihood of CSF leak, meningitis, and even clip exposure.

Despite these limitations and the current controversy that surrounds this topic, the EEA can be safely and effectively utilized to access well-selected aneurysms and be a valuable addition to the armamentarium of cerebrovascular surgeons. It must be emphasized, however, that this technique should be limited to surgical teams with extensive experience and comfort in EEA as well as open vascular surgery and only performed in select cases after careful scrutiny. Finally, the indications of EEA for aneurysms may continue to grow beyond what we currently describe as more surgical teams are gaining familiarity with its nuances as well as the continued technological advancement of endoscopic equipment.

Surgical Technique

Preoperative Management

At first evaluation, patients must be divided into two key categories, those with ruptured or unruptured aneurysms. Patients presenting with ruptured aneurysms and subarachnoid hemorrhage must first be evaluated for hydrocephalus and, if necessary, CSF diversion performed at the surgeon's discretion. At this point, a diagnostic cerebral angiogram (DSA) is completed with 3-D reconstruction to both evaluate endovascular options as well as provide detailed anatomy for surgical planning. CT angiography (CTA) is also performed for intraoperative image guidance and to better understand the relationship of the aneurysm to the skull base and sinuses. In addition, if the aneurysm is in a location that may necessitate pituitary gland manipulation, a preoperative pituitary panel may be considered. All other usual subarachnoid protocols are also performed including tight blood pressure control, arterial line placement, and central venous access prior to surgery.

Unruptured aneurysms require similar workup for surgical planning and evaluation of endovascular treatment options. A full pituitary panel is also performed to evaluate for preoperative pituitary gland function. Depending on the patient's overall health, medical and/or cardiac stratification is also required as unruptured aneurysms are usually treated electively.

Technical Nuances

There are several major principles that are required of EEA for aneurysm clipping. Firstly, wide skull base exposure provides the surgeon with the ability to utilize a 0° endoscope in nearly all instances, thereby minimizing the risk of spatial disorientation that may accompany the use of angled endoscopes. Secondly, a two-surgeon, four-handed dynamic endoscopy technique, utilizing a binaural approach, in which one surgeon is fully dedicated to driving the endoscope and maintaining the optimal view while the other surgeon bimanually dissects, improves surgical guidance as one surgeon can continually move the endoscope in any direction that is necessary to maintain optimal, uninterrupted visualization and enhanced depth perception for the surgeon that is performing bimanual dissection while minimizing spatial disorientation [28, 29]. In addition, communication between surgeons and familiarity working as a skull base team is absolutely essential.

A high-speed drill is used to thin the bone of the skull base to permit the use of a Kerrison rongeur to remove the egg-shelled bone off of the dura. This technique allows for safe exposure of the skull base dura and can also be employed to skeletonize the paraclival and paraclinoidal segments of the ICA. The use of a 6- or 8-French Fukushima-style teardrop suction in the non-dominant hand of the dissecting surgeon allows for continuous flow regulation of the suction, while sharp and blunt dissection can be performed with the dominant hand, ensuring a clear surgical field without risking damage from uncontrolled suctioning of critical structures. Just as in open transcranial surgery, temporary clipping and trapping with burst suppression may be utilized when needed. Bleeding from the sphenoid mucosa can usually be controlled with warm water irrigation; more torrential bleeding from the cavernous sinus or basilar plexus is controlled with flowable Gelfoam and gentle pressure, while bipolar cautery is reserved for arterial bleeding. In addition, we prefer the use of a single shaft clip applier, as conventional clip appliers, such as those that are typically used in open transcranial surgery, are difficult to utilize in the narrow endonasal corridor. In general, standard aneurysm clips are used; however, low profile clips may ease reconstruction.

Various skull base techniques may be employed for closure, depending upon if a CSF leak is present and, if so, whether this is a high or low flow leak. The vascularized nasoseptal flap, pedicled from the sphenopalatine artery, is the mainstay of reconstruction [30, 31]; however, other adjuncts such as free flaps, abdominal fat grafts, fascia lata, pericranium, inferior turbinate flaps, and temporoparietal fascial grafts may also be combined or substituted depending on the situation. In general, a multilayer reconstruction is required, especially if the clips are protruding into the sphenoid sinus. This usually includes a fat graft surrounding and padding the clips, auto- or allograft overlying them (before or after the fat graft, depending on degree of protuberance), and finally a vascularized flap. Finally, as these are considered level V cases [32], it is imperative that the entire skull base team has a strong anatomic knowledge of the skull base and adjacent neurovascular structures as well as extensive experience and familiarity working together in the two-surgeon, four-handed technique.

Initial Exposure

The patient is placed in the supine position with the patient's right side on the edge of the operating table. The bed is then turned 90° with the left side of the patient toward anesthesia. The head is fixated in a three-point Mayfield holder and placed in gentle extension and then rotated, and the chin angled toward the patient's right side. Neuronavigation with a fine cut CTA is then registered. If an image-guided MRI is available, this is also fused to the CTA for improved soft tissue navigation. Neurophysiological monitoring is utilized throughout the surgery. This consists of somatosensory evoked potentials (SSEPs) and, depending on the location of the aneurysm, can also include electromyography for cavernous sinus cranial nerve monitoring and/or brainstem auditory evoked responses (BAER) as well. Motor evoked potentials (MEPs) are usually not used given the lack of involvement of more distal perforators. Nasal pledgets are soaked with 0.05% oxymetazoline and are placed into each nasal passage. These pledgets are left in place, while the midface, abdomen (fat), and right lateral thigh (fascia lata) are prepped and draped. Two high-definition (HD) monitors are placed just to the left of the patient's head, directly across from and at eye level of each surgeon, allowing each surgeon a direct view of one of the monitors. A third-generation cephalosporin is given for antibiotic prophylaxis prior to removal of the pledgets from the nares.

A 0° rigid endoscope is introduced into the nasal cavities to inspect the nasal passages. Generally, the right middle turbinate is resected, followed by harvesting of a right-sided pedicled nasoseptal flap; however, this can be done on the left side as well depending on the patient's anatomy and location of the aneurysm. The nasoseptal flap is then tucked into the nasopharynx until it is needed at the end of the case. If the flap is prohibitive of a lower transclival approach/vertebrobasilar aneurysm, an ipsilateral medial maxillary antrostomy can be performed so that the flap can be stored in the maxillary sinus during the exposure, dissection, and clipping. The posterior nasal septum is disarticulated from the sphenoid rostrum, and a wide sphenoidotomy is performed. Posterior ethmoidectomies are usually added to widen visualization for paraclinoidal {or anterior communicating artery (ACoA)} aneurysms. At this point all sphenoid septa are drilled flush with the sella or skull base until the floor of the sella is exposed. Image guidance and micro-Dopplers are then utilized to confirm the location of both parasellar carotid arteries. The mucosa of the sphenoid sinus is stripped, and the bone of the sellar floor is removed until the medial cavernous sinuses are exposed laterally. Bilateral middle clinoidectomies [33] are performed, and drilling continues toward the planum until the superior intercavernous sinus (SIS) is exposed superiorly.

Paraclinoidal Aneurysms

Once the sella is fully exposed, attention is turned to obtaining proximal vascular control of the cavernous ICA, which can be done by performing a transcavernous approach with removal of the bone overlying the parasellar ICA (Fig. 23.10), careful opening into the medial cavernous sinus, dissection across the ICA, and packing of the CS (cavernous sinus) with flowable Gelfoam. Depending on the aneurysm location, proximal control can be obtained on the paraclinoidal ICA for aneurysms distal to the dural ring/diaphragma or on the paraclival ICA if the aneurysm or a portion of its neck is proximal to the dural ring/diaphragma. The suprasellar dura and distal dural ring are then carefully opened, providing access for distal vascular control and allowing exposure to the ophthalmic artery and aneurysmal neck, respectively. After appropriate exposure and proximal and distal vascular control are achieved, aneurysmal dissection can begin. Due to the midline trajectory of the EEA, medially projecting paraclinoidal aneurysms, typically classified as superior hypophyseal artery (SHA) aneurysms, are ideal as they require minimal or no manipulation of the optic apparatus which often obscures the neck of the aneurysm from a transcranial approach. In addition, the improved visualization

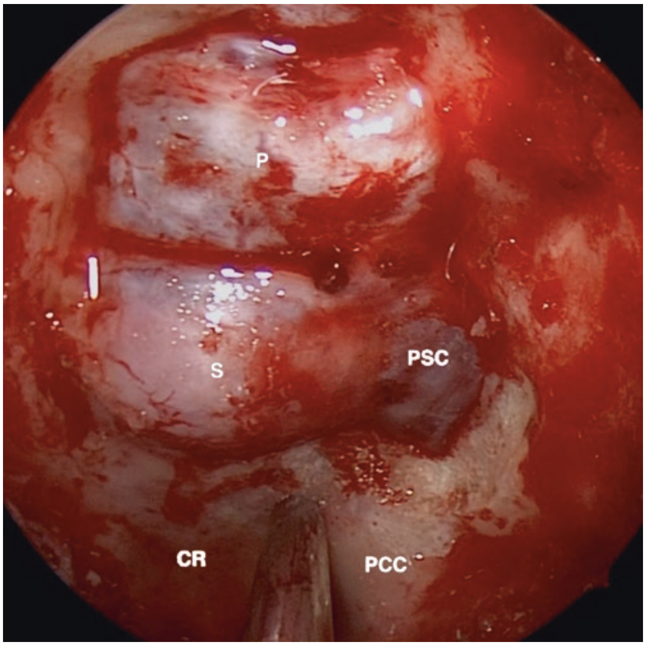

Fig. 23.10 Endoscopic endonasal exposure of sellar and planar dura along with removal of bone over the left parasellar ICA (*CR* clival recess, *P* planum sphenoidale dura, *PCC* paraclival ICA, *PSC* parasellar ICA, *S* sellar dura)

allows for better identification of the SHA as well as its perforators (Fig. 23.11a), reducing the likelihood of inadvertent clip occlusion that could result in pituitary dysfunction or vision loss. The EEA can also be used for a medially pointing ophthalmic artery aneurysm, which would point more superiorly than an SHA aneurysm. As a result, distal control is easier to obtain by dissecting inferior to these aneurysms. Often, simple straight clips are all that are necessary for both of these aneurysms as the clip blades introduced from this corridor are usually perpendicular to the neck of a medially projecting aneurysm and parallel to the course of the supraclinoidal ICA through the ipsilateral nostril (Fig. 23.11b); however, a curved clip may be preferred for aneurysms with a wide neck or one directed in an anteroposterior trajectory. After adequate clip placement, meticulous hemostasis is obtained and intraoperative angiography used to confirm adequacy of treatment. Of note, there is nascent endoscopic technology that will allow for intraoperative endoscopic fluorescence angiography, allowing for early evaluation of clip placement, verification of perforator preservation, and patency of major vasculature.

Closure depends on the projection of the clip spring. Often, the clip(s) are sticking out of the dura and into the sinus, but if a clip lies flat against the planum dura, the reconstruction may be simpler. In such a case, a piece of collagen matrix (DuraGen®, Plainsboro, NJ, USA) is tucked inside the edges of the dura with a slit as needed for the clips, and a fascia lata autograft or other allograft is placed over the clips, widely covering the dural opening after all surrounding mucosa has been removed. If the projection is significant such that the fascia is significantly tented outward, fat is first placed around the clip spring(s) before the fascia is placed (Fig. 23.12a). The nasoseptal flap is mobilized out of the nasopharynx to cover the skull base defect with careful attention paid to ensure that the flap is in direct contact with the bony edges of the defect without any folds to provide a corridor for a CSF fistula (Fig. 23.12b). Surgicel is laid on the flap edges, which are then covered with tissue glue and Gelfoam. At this point, Merocel® nasal tampons (Invotec International, Inc., Jacksonville, FL, USA) are used to keep the flap in position, and Doyle and Silastic splints (Micromedic, Inc., St. Paul, MN, USA) are sutured onto the nasal septum.

An inherent advantage of the EEA is the ability to achieve proximal ICA control without the need for an anterior clinoidectomy or neck incision, one of which would be necessary from a transcranial approach and the latter of which does not address collateral supply. In addition, the lateral position of the cranial nerves in the cavernous sinus provides an advantage to the EEA for aneurysms that extend into or involve the medial cavernous ICA, as the proximal ICA is reached through the medial compartment of the cavernous sinus, reducing the risk of cranial nerve palsies that can occur when as entering through the roof of the sinus in a transcranial approach. The EEA also provides several other advantages to paraclinoidal aneurysms such as the elimination of brain retraction, easier recovery, and improved postoperative cosmesis. The main disadvantage of the EEA in this instance, however, is the difficulty of distal vascular control, especially in larger or giant aneurysms. In such a case, a craniotomy should be considered for distal control (with or without EEA). In addition, laterally projecting paraclinoidal aneurysms are generally not amenable to an EEA approach as the optic nerve prevents adequate access, which is much simpler to obtain through a standard craniotomy.

Anterior Communicating Artery Aneurysms

After the initial sellar exposure has been performed, drilling is continued anteriorly until the posterior portion of the planum sphenoidale is removed. In addition, it is important to remove the bone over the medial optic canal. The suprasellar dura is then opened in the midline, immediately superior to the SIS (superior intercavernous sinus), and extended laterally immediately above the optic canals to the medial falciform ligament, exposing the suprachiasmatic region and the anterior communicating artery complex. The A1s can be identified as they cross over the optic nerves/genu; the interhemispheric fissure is then carefully dissected, allowing visualization of the bilateral A1 and A2 segments for both

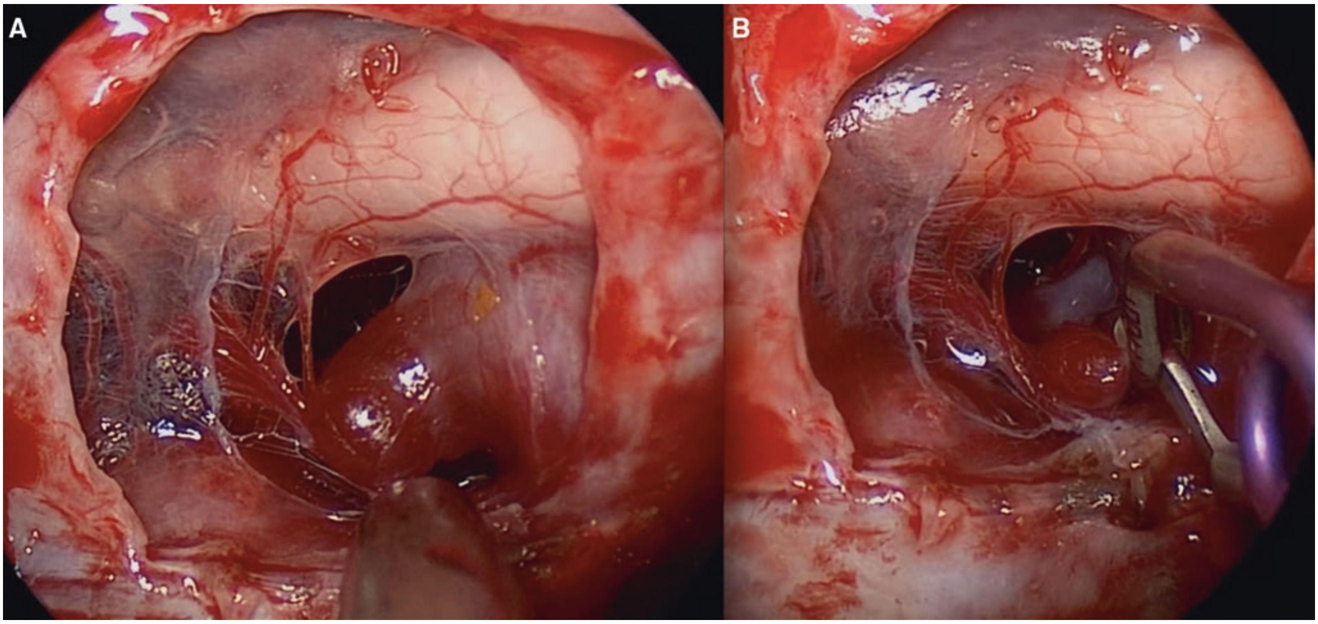

Fig. 23.11 (**a**) Left superior hypophyseal artery aneurysm after dissection. (**b**) Left superior hypophyseal artery clipped with a simple, straight clip

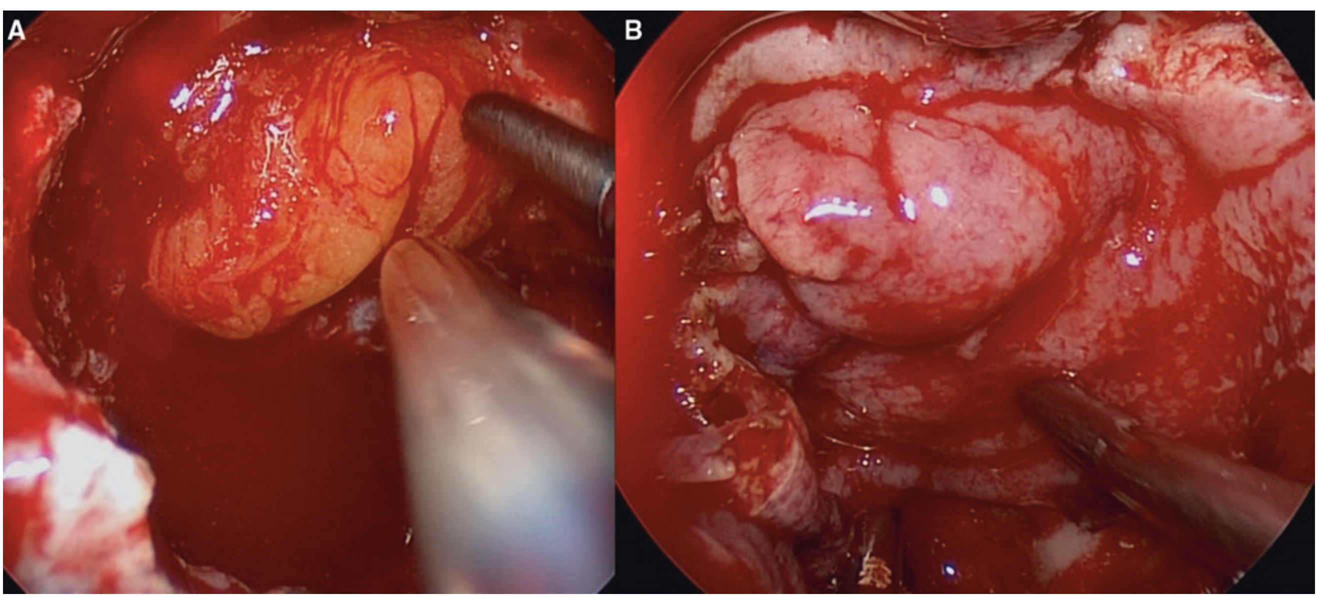

Fig. 23.12 (**a**) Abdominal fat graft placed over dural defect, covering the aneurysm clip springs. (**b**) Placement of the vascularized, pedicled nasoseptal flap over entire bony defect

proximal and distal vascular control. After vascular control has been obtained, attention can be turned to dissecting the neck and dome of the aneurysm as well as any perforating branches that may be present. Interhemispheric dissection can be challenging endonasally, and clip placement may require creativity given the limitation of direction of clip application. Once the dissection is completed, the clip can be placed across the neck of the aneurysm. Meticulous hemostasis is obtained, and adequacy of treatment is assessed as discussed above. Closure is similar to as listed in the previous section, though properly selected clips are less likely to project through the dural opening. A lumbar drain may also be placed postoperatively at the surgeon's discretion.

EEAs for anterior communicating artery aneurysms are generally limited to small midline aneurysms projecting either superiorly or inferiorly. Anteriorly projecting aneurysms pose a challenge as the dome can block direct visualization of the aneurysm neck. Posteriorly projecting aneurysms offer limited control of perforating branches, increasing the risk of inadvertent vascular injury. In addition, the location of the optic chiasm and nerves creates another challenge as it limits clip placement for posteriorly projecting aneurysms. Frontal lobe herniation upon dural opening can also limit exposure, making aneurysm clipping nearly impossible without CSF diversion and adequate brain relaxation. Finally, aneurysms located deep in the interhemispheric fissure may require prohibitively difficult dissection from an endonasal approach, which should be carefully assessed for on the preoperative CTA or angiogram.

Posterior Circulation Aneurysms

As mentioned above, for midbasilar or vertebrobasilar junction aneurysms, a medial maxillary antrostomy is performed on the ipsilateral side of the pedicled nasoseptal flap, allowing the flap to be safely stored in the maxillary sinus and providing unobstructed access to the nasopharynx and clivus. After completion of the sellar exposure as described above, the soft tissue of the nasopharynx is dissected with a needle tip monopolar cautery and elevated inferiorly away from the clivus leaving a cuff or flap inferiorly. Bone is then drilled between the paraclival segments of the carotids arteries and as inferiorly as necessary for proximal access (Fig. 23.13). If needed, the inferior bony exposure can be widened beyond the vertical plane of the paraclival ICA by exposing and drilling the medial jugular tubercle just below foramen lacerum. The paraclival carotid arteries can also be skeletonized to define the lateral limits of exposure and allow for gentle mobilization, if needed. The extent of bony clival removal is dependent upon the location of the aneurysm that is being treated. For basilar apex, superior cerebellar artery (SCA), or posterior cerebral artery (PCA) aneurysms, only the upper half of the clivus is necessary to remove; however, depending on the location of the aneurysm, this often includes performing posterior clinoidectomies via an *interdural* pituitary transposition [32] to achieve adequate visualization. This is in contrast to the approach for an anterior inferior cerebellar artery (AICA), midbasilar, or vertebrobasilar aneurysm, which requires greater exposure of the inferior clival dura but generally does not necessitate posterior clinoidectomies.

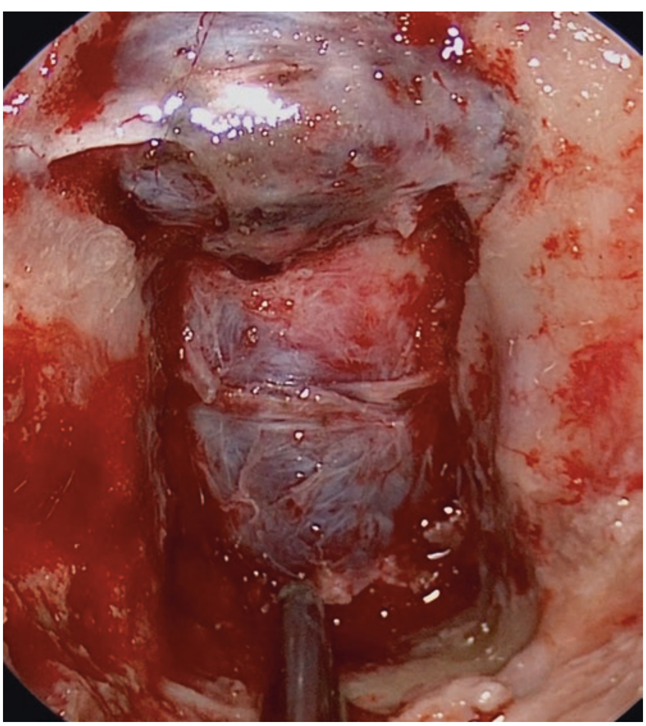

Fig. 23.13 Endoscopic endonasal transclival approach with *interdural* pituitary transposition for exposure of clival dura

Once the dura is exposed, a micro-Doppler is utilized to approximate the location of the basilar artery. The outer, periosteal layer of dura is stripped widely, and the basilar plexus embolized with flowable Gelfoam. The dura is then sharply opened in a vertical fashion at a safe distance from the basilar artery. The dural edges are then resected to provide wide exposure of the interpeduncular, prepontine, or premedullary cisterns, depending on the target. At this point, the aneurysm is visualized (Fig. 23.14a, b), and proximal and distal vascular control is readily available at the proximal basilar trunk and distal terminal branch vessel, respectively (Fig. 23.15a). Perforating branches should also be visible and dissected away if possible. One factor to consider during basilar apex aneurysm clipping is the selection of a bayonetted clip, which prevents the clip applier from obstructing visualization of clip placement given the location of the basilar apex behind the mobilized pituitary gland (Fig. 23.15b). Once the clip is placed, the brainstem can be decompressed, if needed, by resection of the aneurysmal dome or aneurysmorrhaphy. After meticulous hemostasis is obtained, a piece of collagen (DuraGen®, Plainsboro, NJ, USA) is gently tucked into the dural edges as an inlay if the surgeon feels that this can safely be performed without damaging the nearby sixth cranial nerve (Fig. 23.16). A harvested piece of fascia lata is used as a dural onlay, widely covering the defect as well as the sellar and paraclival ICAs (Fig. 23.16b). Abdominal fat graft is placed to buttress the layers in place and fill the space between the paraclival ICAs, creating a flatbed for the flap (Fig. 23.16c). The nasoseptal flap is placed over the entire defect (Fig. 23.16d), and the remainder of the closure is as previously described. Following the case, a lumbar drain is placed for any posterior fossa endonasal dural defect.

The transclival EEA and/or *interdural* pituitary transposition provides wide visualization of the interpeduncular, prepontine, and ventral pontomedullary cisterns. As a result,

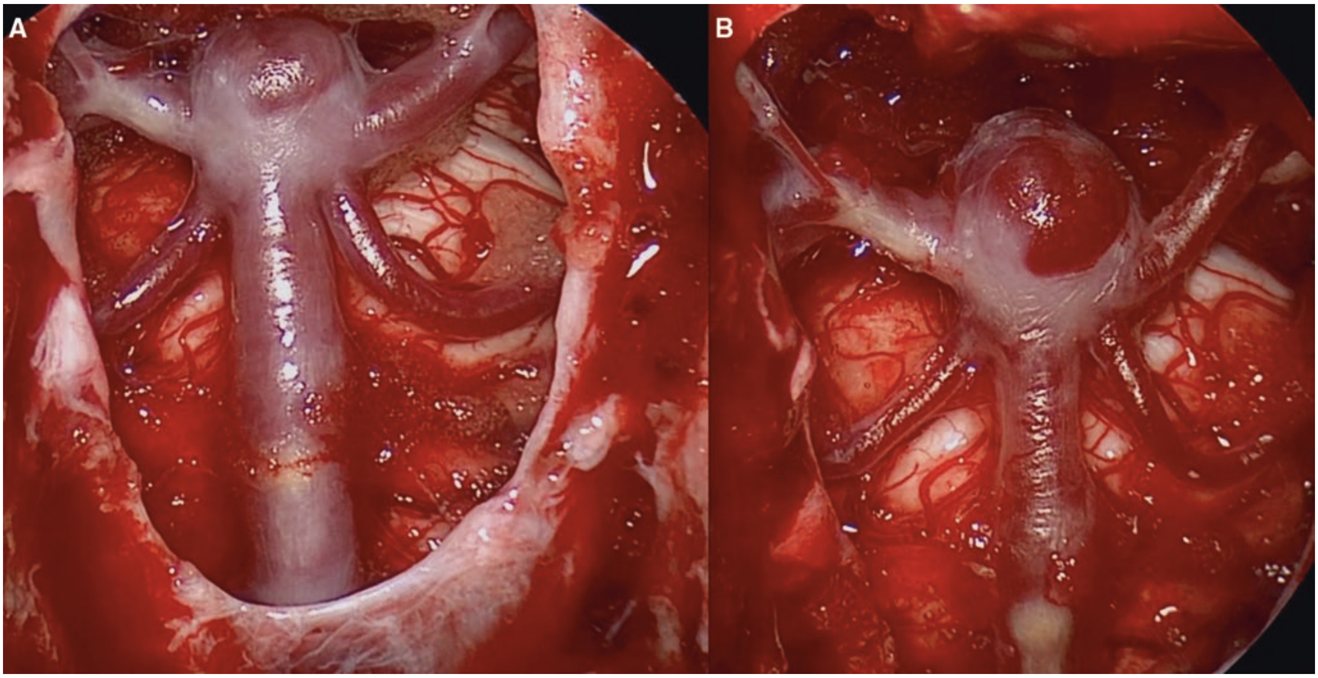

Fig. 23.14 (**a**, **b**) Exposure of a basilar apex aneurysm with access to proximal and distal vessels via EEA

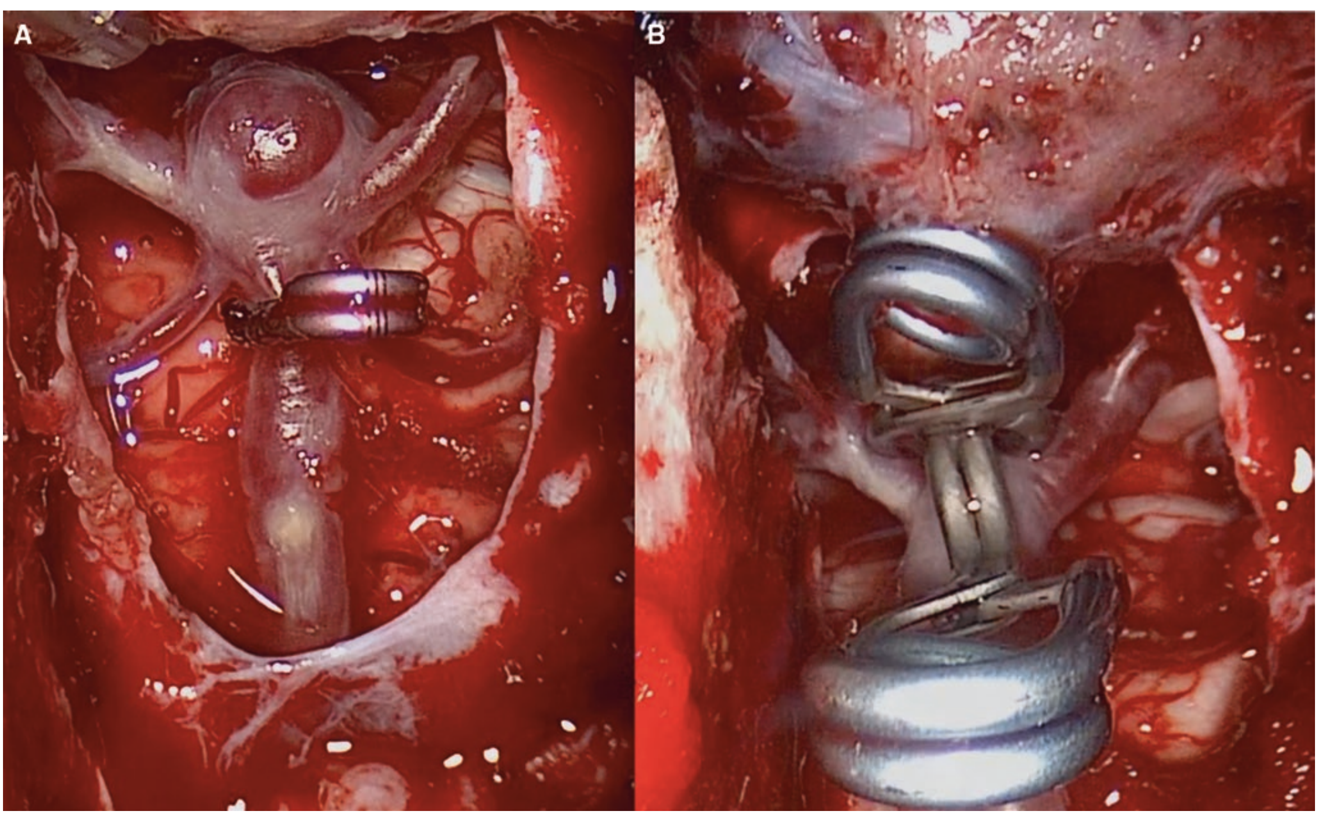

Fig. 23.15 (**a**) Proximal control obtained via temporary clipping of basilar trunk. (**b**) Placement of bayonetted clips across the neck of a basilar apex aneurysm

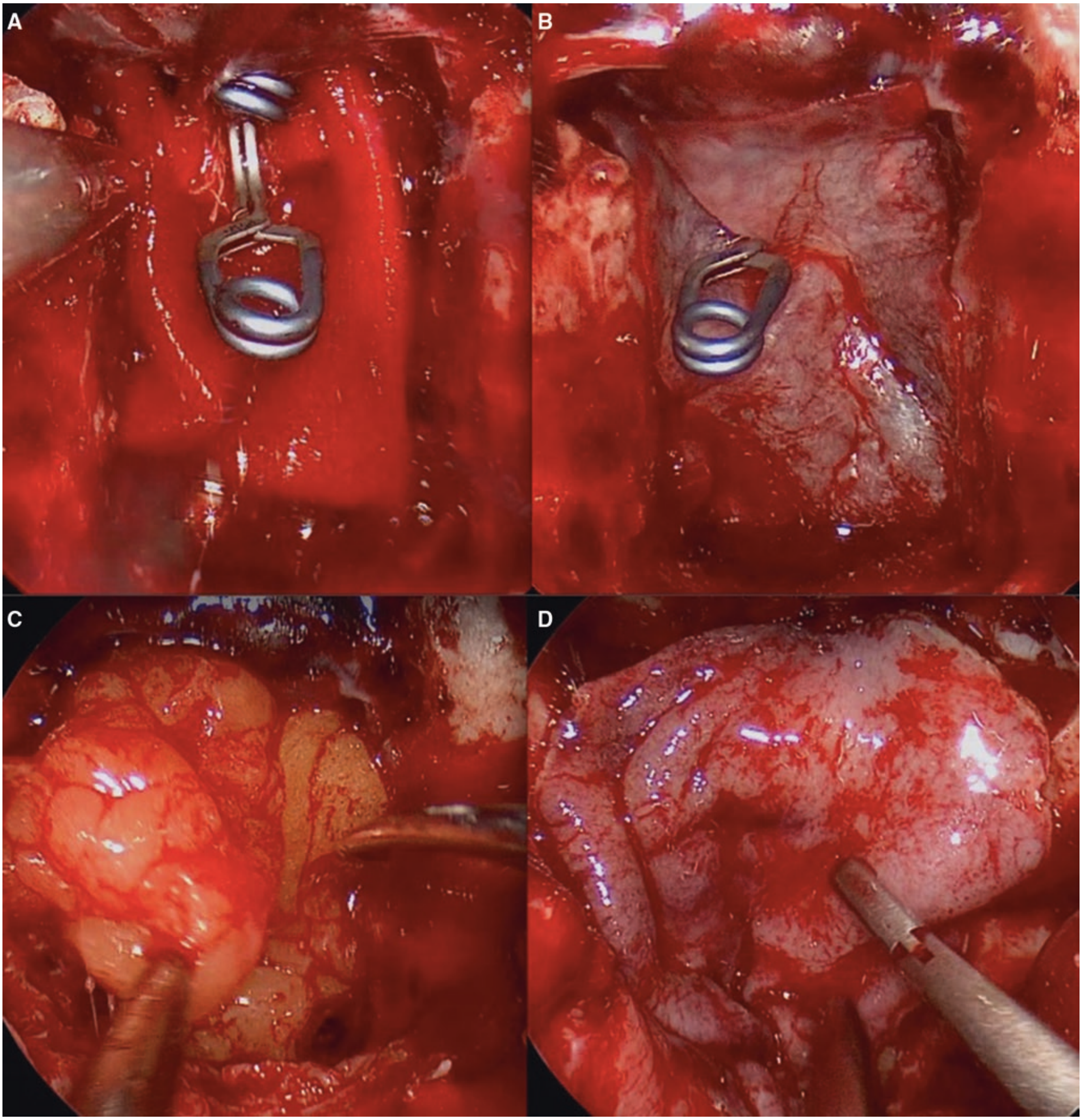

Fig. 23.16 (**a**) Placement of collagen inlay over transclival dura defect. (**b**) Harvested fascia lata graft placed as an onlay with aneurysm clips protruding. (**c**) Abdominal fat graft used to obliterate dead space created by transclival defect. (**d**) Placement of vascularized, pedicled nasoseptal flap over entire bony defect

access to the vertebrobasilar junction, basilar trunk, and terminal vessels can be directly obtained without any brain retraction and minimal neurovascular manipulation. In fact, the EEA provides a unique access to the midbasilar trunk and AICA, what Drake referred to as “no man’s land” [34]. This exposure, however, is generally not favored for high-riding basilar apex aneurysms, above the posterior clinoid processes, as distal vascular control becomes difficult, aneurysm access is limited, and transcranial approaches are more than adequate.

Intraoperative Rupture

Intraoperative rupture is the greatest fear of any aneurysm surgeon, and this fear is amplified with an endoscopic approach. However, the concepts of management are identical to open surgery. The most critical factor for success in this situation is the use of team surgery with significant experience in endoscopic endonasal surgery. The ability to work seamlessly together typically takes many cases, and endoscopic endonasal surgery is well known to have a long learning curve [35–37]. The second surgeon, the endoscopist, becomes critical in the setting of intraoperative rupture and plays multiple roles, the least of which is keeping the endoscope clear. Dynamic endoscopy is critical for close inspection of the vasculature and bleeding site during control of hemorrhage as well as any subsequent clip placement. The endoscope is constantly maneuvered to give the best view while staying on the edge of the working field to not hamper maneuverability. In addition, team surgery at its finest becomes paramount in the setting of potential disaster by providing constant feedback, troubleshooting, and modulation of enthusiasm (often the enemy of a good outcome).

In this team setting, all of the same techniques for management of intraoperative rupture can be applied. Case selection is critical to assure adequate proximal and distal control prior to aneurysm manipulation. As usual, once rupture occurs, applying suction or a cottonoid to the site of rupture is used to control hemorrhage and identify the site of rupture. Maintaining this control usually requires great facility with a slotted suction to maintain pressure on the site of bleeding while the other hand is used to apply a clip proximal to the site of rupture. Packing off of the bleeding site, which may be a natural reflex when working in a deep space, should be avoided and can lead to significant intracranial hemorrhage.

Postoperative Management

Patients are taken to the ICU for initial postoperative management, and ruptured aneurysms are continued on a typical subarachnoid protocol. If a lumbar drain was placed after surgery, drainage of 10–15 cc/hr. of CSF is continued for 72 h. In the setting of a rupture, CSF diversion is always performed with a low threshold for conversion to shunt given the increased risk of CSF leak with endonasal approaches [24]. Nasal packing is left in place for 7 days, at the discretion of the ENT co-surgeon. A third-generation cephalosporin is continued intravenously for 48 h after surgery and then transitioned to an oral second-generation cephalosporin or equivalent until nasal packing is removed. Prolactin and morning cortisol levels are also drawn on postoperative days 1 and 2 to evaluate for pituitary function. Vigilant evaluation for a CSF leak is also undertaken; if a leak is suspected, the patient is returned to the operating room within 24 h for inspection and repair. After hospital discharge, the patient returns to the clinic for removal of nasal packing, endoscopic inspection of the reconstruction for CSF leak, and a full pituitary panel. Repeat DSA approximately 5–7 days; following a rupture may help stratify patients based on the evidence of vasospasm and lead to earlier transition from the ICU.

Complication Avoidance and Management

Complication avoidance begins with the assembly of a skull base team that has extensive experience working together on similar approaches. It is strongly recommended that teams start with more basic cases and gradually grow together through the learning curve where the team can work together seamlessly before attempting cases as challenging as cerebrovascular surgery via EEA [28]. After an appropriate skull base team has been assembled, development of contingency plans becomes the consideration. The same basic principles of vascular control, sharp microdissection, hemostasis, and adequate exposure and visualization must be achieved. If any of these factors cannot be reached, a plan to abort the EEA and revert to or combine with an open craniotomy must be in place and ready to be implemented at any time. This includes prepping for an open craniotomy and having appropriate instrumentation open and available at all times. Just as in open cerebrovascular surgery, skull base exposures should be performed by accessing proximal control first. Prior to aneurysmal exposure, the cavernous ICA should be exposed for paraclinoidal aneurysms, while access to the vertebral arteries and basilar trunk need to be obtained prior to posterior circulation aneurysm exposure. It is imperative that only after these steps are completed that dissection can continue to reach the aneurysm as they can minimize bleeding from an intraoperative rupture and prevent an uncontrollable surgical disaster.

Finally, multiple steps should be taken throughout the case in anticipation of skull base reconstruction. This begins by prepping the lateral thigh and abdomen for fascia lata and fat, respectively; elevating a large enough vascularized, pedicled nasoseptal flap to cover the entire skull base defect or sphenoid; as well as vigilant postoperative monitoring for CSF leaks or clip exposure, both of which should have a low threshold for return to the operating room and repositioning or bolstering of the flap.

Patient Selection

Detailed examination of the DSA and CTA must be undertaken before considering an EEA for aneurysm clipping, as indications are specific for aneurysms that are amenable to this approach. Sinus anatomy must also be closely examined to understand the relationship of the sinuses to the aneurysm. Any active sinus infection is a contraindication to EEA which could delay surgery for elective aneurysms and cause consideration of another approach for ruptured aneurysms that need to be treated immediately. Body habitus should also be considered, as morbid obesity may be a relative contraindication to EEA for aneurysm clipping. The necessity of a large skull base defect and sizable dural opening in the setting of occult intracranial hypertension heightens the risk for postoperative CSF leak, which can be extremely challenging to repair even with the assistance of a vascularized, pedicled flap and the addition of aneurysm clips to the defect. In evaluating the angiogram, ICA anatomy is of major importance. Patients with "kissing carotids" may not be candidates for EEA if the surgical corridor is too narrow to safely work within. In general, EEA candidates consist of aneurysms of the ventral midline skull base, specifically paraclinoidal aneurysms, aneurysms originating from the vertebrobasilar junction, basilar trunk or terminal basilar branches, and anterior communicating artery aneurysms.

Small- or medium-sized paraclinoidal aneurysms with a medial trajectory, medially projecting distal cavernous ICA, superomedial pointing ophthalmic artery aneurysms, or "carotid cave" aneurysms with question of subarachnoid extension are generally the most amendable to an endoscopic endonasal approach [17, 21, 24]. In these instances, the endonasal approach allows for early proximal vascular control at the cavernous ICA, requiring minimal or no manipulation of the optic apparatus and allowing the aneurysm to be approached through the medial wall of the cavernous sinus, which is known to have a low level of morbidity. A transcranial approach to such an aneurysm requires an anterior clinoidectomy and possible neck dissection to obtain proximal vascular control, mobilization, and manipulation of the ipsilateral optic nerve with potential vision loss, as well as opening of the roof of the cavernous sinus, which is associated with high rates of cranial nerve palsy and morbidity. Utilizing an endoscopic endonasal approach for large or giant paraclinoidal aneurysms should, however, be undertaken with caution. In such cases, distal vascular control is extremely difficult, or impossible, and may require a craniotomy. Laterally pointing paraclinoidal aneurysms are not accessible via an EEA due to interposition of the optic nerve between the surgeon and aneurysm.

Anterior communicating artery aneurysms may be candidates for EEA if they are small, midline, and projecting either superiorly or inferiorly [18, 20, 25]. Anterior pointing aneurysms pose an issue as the EEA trajectory may cause the dome of the aneurysm to block visualization of the neck. Additionally, posteriorly directed aneurysms are risky, as visualization of perforating arteries is limited. Dissection into the interhemispheric fissure is challenging via EEA, and therefore aneurysms located deep in the interhemispheric fissure should utilize a different approach. Finally, herniation of the frontal lobes through the dural opening may block the necessary access, requiring aggressive brain relaxation or CSF diversion. Otherwise, the EEA may need to be aborted in favor of a transcranial approach.

Several posterior circulation aneurysms can be candidates for EEA [16, 22, 23]. Paramedian, proximal aneurysms of the PCAs, SCAs, and AICAs are anatomically suitable for an endoscopic endonasal transclival approach. Low-lying basilar apex aneurysms may be approached through EEA via a transclival route (Fig. 23.17a, b); however, aneurysms located above the posterior clinoid processes are a contraindication for EEA as visualization of the aneurysm dome, and even neck access is limited. Therefore a transcranial approach should instead be considered when addressing high-riding basilar apex aneurysms.

Surgical Outcomes

A review of the recent meta-analysis by Vaz-Guimaraes et al. [38] is listed in Table 23.2. There are currently ten reports of EEA for aneurysm clipping, consisting of 22 patients and 24 total aneurysms, with two patients undergoing clipping of two aneurysms in the same case. Separating these 24 aneurysms by location, the most common was paraclinoidal with 12 (50%). This was followed by basilar apex with 3 (13%), anterior communicating artery with 3 (13%) and the remainder involving the posterior circulation including basilar trunk location with 2 (8%), posterior cerebral artery with 2 (8%), posterior inferior cerebellar artery with 1 (4%), and vertebral artery with 1 (4%). Presenting symptoms were as follows: 11 (50%) were incidental findings, 7 (32%) were ruptured with subarachnoid hemorrhage, and 4 (18%) were presented with focal deficits from mass effect. Successful clipping, defined by no need for further surgical or endovascular intervention, was accomplished in 22 (92%) of 24 aneurysms with only 2 (8%) requiring further treatment, one by repeat clipping via EEA and the other by endovascular coiling.

All 15 (100%) anterior circulation aneurysms were successfully clipped without the need for further treatment; however, 2 (15%) did require repeat EEA for CSF leak. No other complications were seen in this group. Both (22%) of the aneurysms that required further treatment were among the 9 posterior circulation aneurysms, while 3 (33%) of these 9 were found to have postoperative CSF leak and 4 (44%) with perforating branch stroke. Anterior and posterior circulation aneurysms in this group did not have a statistically significant difference in size and had an overall median diameter of 6.8 mm. Posterior circulation aneurysms did, however, demonstrate a significantly higher rate of perforator injury, CSF leak, and need for additional treatment.

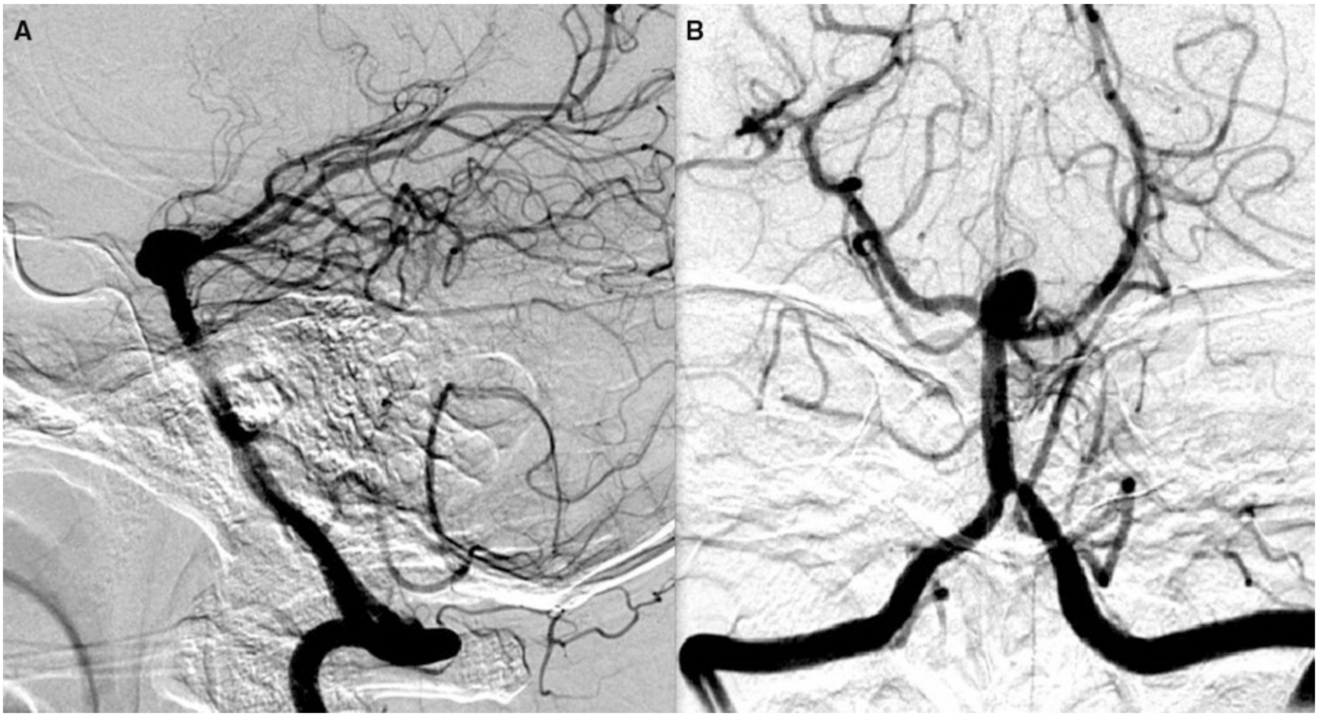

Fig. 23.17 (**a**, **b**) Preoperative angiogram demonstrating a low-riding basilar apex aneurysm, note the height of the aneurysm in relation to the sella and posterior clinoid processes

PEARLS
Wide endonasal exposure is essential
Must utilize bimanual-binaural microdissection with dynamic endoscopy in a two-surgeon, four-handed technique
The same principles of open transcranial cerebrovascular surgery apply to endoscopic endonasal cerebrovascular surgery, including proximal and distal vascular control and sharp microdissection
A well-developed, pedicled vascularized nasoseptal flap is essential in skull base reconstruction to avoid CSF leaks
Multilayered reconstruction is necessary for adequate clip coverage

Commentary on Case Presentation in Chapter 23

Discussion

The case illustration (Fig. 23.1a–d) presents an incidental finding of a left paraclinoidal ICA aneurysm with a medial trajectory. As previously discussed, we find this to be an ideal candidate for an EEA and clipping of this aneurysm. Combining with a transcavernous approach, the EEA provides direct access to this aneurysm with adjacent proximal control and allows for excellent visualization of both the aneurysmal neck and dome. Perforating branches can also be identified and dissected away after proximal control is obtained, minimizing the risk of perforator injury. In addition, an EEA spares the patient the need for an anterior clinoidectomy and lowers the risk of damage to the optic apparatus or cranial nerves of the lateral wall of the cavernous sinus, both of which cannot be completely avoided in a transcranial approach. The major difficulty in an endoscopic endonasal approach for this aneurysm is obtaining distal vascular control and skull base reconstruction; however, these are best addressed after carefully opening the distal dural ring and visualizing the distal ICA and harvesting a vascularized, pedicled nasoseptal flap, respectively. In addition, if opening of the distal dural ring does not provide adequate distal vascular control, the case must either be done without distal control or an open craniotomy can be done in combination with the EEA. As a result, in experienced hands, this patient is a strong candidate for an EEA and aneurysm clipping.

Conclusions

The endoscopic endonasal approach for intracranial aneurysms remains a controversial treatment for rare, well-selected aneurysms in the hands of an experienced team.

Table 23.2 EEA for aneurysm clipping

Author	Clinical presentation	Location	Size (mm)	Projection	Complication	Outcome
Kassam (2006)	Focal deficit	Vertebral artery	11	Ventral	None	Complete recovery
Kassam (2007)	Incidental finding	Superior hypophyseal	5	Medial	None	Complete recovery
Kitano (2007)	Incidental finding	AComm	N/A	N/A	None	Complete recovery
Ensenat (2011)	SAH	PICA	1.2	Ventral	CSF leak	Complete recovery
Froelich (2011)	Incidental finding	AComm	7	Superior	None	Complete recovery
Germanwala (2011)	SAH	Ophthalmic/ paraclinoidal	5/10	Superomedial/ posteromedial	None	Complete recovery
Drazin (2012)	SAH	Basilar trunk	4	Ventral	None	Complete recovery
Sommana (2015)	SAH	Basilar apex	10	Posterosuperior	None	Endovascular coiling for residual
	SAH	Basilar apex	5	Superior	Lacunar stroke	Neurologic disability
	Focal deficit	PCA	9.4	Superior	Stroke CSF leak Meningitis	Neurologic disability
Gardner (2015)	Incidental finding	Ophthalmic	3.5	Medial	None	Complete recovery
	CN palsy	PCA	19	Ventral	CSF leak Meningitis Lacunar stroke	Mild disability
	Incidental finding	Superior hypophyseal	5	Medial	CSF leak	Complete recovery
	Incidental finding	Basilar apex	9	Posterosuperior	Lacunar stroke	Complete recovery
	Vision loss and hypopituitarism	Ophthalmic/ ophthalmic	Giant/5	Superomedial	None	Complete recovery
	Incidental finding	Ophthalmic	6	Medial	CSF leak Meningitis	Complete recovery
	SAH	Ophthalmic	7	Superomedial	None	Complete recovery
	Incidental finding	Ophthalmic	4	Medial	None	Complete recovery
	Incidental finding	Superior hypophyseal	11	Medial	None	Complete recovery
	Incidental finding	Superior hypophyseal	N/A	Inferomedial	None	Complete recovery
Yildirim (2015)	Incidental finding	AComm	N/A	Anterosuperior	None	Complete recovery

Reprinted with permission from Vaz-Guimaraes et al. [38]

Anterior circulation aneurysms, especially medially directed paraclinoidal aneurysms, are the best suited for this treatment option. As with open surgery, posterior circulation aneurysms have higher complication rates including CSF leak and perforator infarct.

23.4 Intracranial Aneurysms: Editors' Commentary

As expressed in the preceding discussions, one of the most critical aspects of both keyhole craniotomy and endoscopic endonasal approaches (EEA) for the treatment of cerebral aneurysms is patient selection. Thorough planning with close examination of preoperative films is necessary to anticipate the spatial relationship of the aneurysm neck and dome with surrounding neurovascular structures and the skull base. Many aneurysms may not be amenable to one or either of these approaches and attempting a one-size-fits-all approach can be catastrophic. Nonetheless, with sound patient selection, thorough knowledge of skull base anatomy, and meticulous surgical technique, good outcomes can be obtained via both keyhole craniotomy and EEA for unruptured and ruptured aneurysms.

EEA does present some distinct advantages for medially projecting supraclinoid aneurysms. EEA obviates the need for brain retraction and can provide good cosmetic results as well. EEA may eliminate the need to perform anterior clinoidectomy or cervical neck incision for proximal control. Proximal control is most commonly attained at the clinoidal segment of the carotid artery or at the paraclival portion. However, distal control can sometimes be challenging to obtain depending on the anatomy and size of the aneurysm as well as its relationship to the optic nerve. In addition, there is less maneuverability in the trajectory of clip appliers. To compensate, there are tubular shaft clip appliers available with straight and angled heads. As documented in the preceding chapters, increased risk of CSF leak remains a major concern. This is particularly the case for ophthalmic segment (C6) aneurysms because the aneurysm neck resides close to the distal dural ring, and clip head protrusion into the sphenoid sinus poses a reconstructive challenge. This may be less of an issue in anterior communicating artery aneurysms. The wider availability of low profile aneurysm clips can significantly reduce clip head protrusion and will likely become the standard clip used in EEA cases. One of the major limitations in anterior communicating artery aneurysms is the unpredictable relationship of the ACOM complex and aneurysm neck to the optic chiasm and interhemispheric fissure, both of which can impede surgical access to the neck. Very careful study of high-resolution MR imaging is necessary to determine if EEA is feasible in these cases.

The supraorbital keyhole approach has well-established outcomes for clipping of intracranial aneurysms of the supraclinoid ICA, anterior communicating artery, and less commonly, basilar, superior cerebellar artery, and middle cerebral artery bifurcation aneurysms. The mini-pterional keyhole approach may provide more direct visualization of MCA bifurcation aneurysms with similar cosmetic result. The surgical trajectory of the supraorbital approach may provide better access if clipping of additional contralateral aneurysms is needed. Significant experience is required before tackling ruptured aneurysm cases with either of these minimally invasive approaches.

The case example (Fig. 23.1a–e) highlights an unruptured, medially projecting supraclinoid aneurysm most consistent with a superior hypophyseal artery origin. The patient has a family history of subarachnoid hemorrhage so it is reasonable to consider treatment. One must weigh the risks and benefits of treating this via an endovascular approach by coiling or flow diversion compared to a surgical approach. Among the approaches discussed, the EEA might provide more direct access to obtain proximal control without the need for anterior clinoidectomy or cervical carotid exposure. In addition, it may provide a more direct line of sight of the aneurysm neck, and the aneurysm is recessed enough from the posterior wall of the sphenoid sinus that the clip head may not protrude significantly into the sinus. However, the supraorbital keyhole approach with endoscopic assistance may have the benefit of shorter operative time for exposure and fewer issues with reconstruction compared with EEA. Although there may not be a definitive "best" surgical approach, having significant experience in each of these approaches is necessary before attempting to treat such an aneurysm.

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