Operative Techniques in Otolaryngology 30 (2019) 44–53

ELSEVIER

Operative Techniques in
Otolaryngology

The endoscopic endonasal approach for pediatric craniopharyngiomas☆

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KEYWORDS
Pediatric;
Craniopharyngioma;
Sella;
Suprasella;
Vision

Craniopharyngiomas are rare but challenging tumors of the ventral skull base affecting primarily pediatric patients. In select cases, the endoscopic endonasal approach represents an appropriate surgical option when tumor resection is favored. However, nuances of the pediatric nasal corridor must be carefully considered to optimize both tumor resection and skull base reconstruction. Here we review pertinent developmental details, options for creation of an optimal endonasal corridor, principles of tumor resection, and techniques for reconstruction. Considerations for intraoperative and postoperative patient care are also reviewed.

Introduction

Craniopharyngiomas are ectodermal tumors most often arising in the sellar and suprasellar region.[1] Most commonly affecting the pediatric age group,[2] they account for 6%-13 % of all intracranial pediatric tumors[3,4] and have been histologically typed as adamantinomatous, more often affecting pediatric patients, or papillary, more often arising in adult patients.[5] They are also classified from a surgical standpoint based on their anatomical relationship to the pituitary stalk as preinfundibular, transinfundibular, and retroinfundibular.[6]

While nearly always benign, craniopharyngiomas have historically represented a significant therapeutic challenge. Often consisting of both solid and cystic components, the tumor may cause mass effect on adjacent structures and grow at widely varying rates. Patients often present with visual loss, headaches, or pituitary dysfunction.[5] Achieving gross tumor resection is challenging due to critical adjacent neurovascular structures such as the

☆ Conflicts of Interest and Financial Disclosures: None.

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http://doi.org/10.1016/j.otot.2019.01.003

pituitary gland, optic chiasm and nerves, hypothalamus, and intracranial vessels encountered in the surgical field. Additionally, there are inherent difficulties of performing endonasal surgery in a child which include a relatively smaller nasal corridor, underdeveloped or absent sphenoid sinus pneumatization, higher risk of significant blood loss, and lack of specialized endoscopic instrumentation for smaller pediatric patients.[7,8] However, the relatively smaller working distance (from the nostril to sella) in pediatric age groups as compared to adults, can be beneficial.[2]

Here we describe our standard endonasal approach for craniopharyngiomas in the pediatric population. We discuss anatomic considerations as well as the nuances of pediatric endonasal surgery, including the creation of an adequate surgical corridor, intracranial resection, and options for reconstruction.

Pediatric developmental and anatomic considerations

While many cases of childhood craniopharyngioma are diagnosed in adolescence when the tumors have grown large enough to become symptomatic, they can arise at any age. One recent case report details an endoscopic optic nerve decompression for a symptomatic tumor in a 12-month old.[9] A bimodal distribution of incidence peaks at 5 and 15 years has been described, with a mean age at diagnosis of 10.7 years.[10] In a very large German series of 496 patients under the age of 18, the vast majority (91%) were diagnosed before age 15.[11] With this epidemiology in mind, skull base and sinonasal development must be considered when planning endoscopic, endonasal approaches to these lesions.

Over 100 ossification centers in the embryonic skull fuse in various areas in utero, resulting in approximately 45 bones at birth.[12] Chondrocranial ossification of the neonatal anterior cranial base begins at the lateral most portion of the ethmoid roof and progresses toward the midline over the first 2 years of life.[13] Prior to completion of ossification, computed tomography of the anterior cranial base must be interpreted with caution as cartilage will not be visible and may look like a defect.

Ethmoid and sphenoid sinus development is ongoing during childhood. The ethmoid sinuses are pneumatized at birth and undergo significant expansion in an anterior to posterior direction until age 7-8.[14,15] By age 12, the ethmoid sinuses have reached adult proportions.[16] Sphenoid sinus development lags behind that of the ethmoids, as the earliest signs of pneumatization do not appear until age 1.[16] Between age 1 and 3 years, red marrow converts to fatty marrow within the sphenoid bone, and this process is followed by an expansion of pneumatization between 6 and 10 years.[14-16] Sphenoid sinus development is not complete until age 15-16 (Figure 1a-c).[14,16] Given that most pediatric cases present before age 15, marrow is often encountered when drilling the tuberculum sella and adjacent planum sphenoidale.

In pediatric patients, the diameter of the naris can significantly impact the use of 2 instruments in each naris. At birth, the naris is a mere 4 mm in diameter, but grows proportionately with the nasal soft tissue. In older children, its size can be estimated by checking the outer diameter of an age-appropriate endotracheal tube. Assuming that there is enough working room for an endoscopic resection, one last consideration is whether or not a nasoseptal flap reconstruction is possible. A radiographic feasibility study by Purcell et al actually found an inverse relationship between age and ratio of flap length to defect.[17] This finding was supported by Ghosh et al, who reported on the successful use of nasoseptal flap reconstruction in 7 children under age 10 (range 3-10 years) with craniopharyngiomas.[18] While there is no absolute cut-off below which nasoseptal flap reconstruction should not be undertaken, careful review of preoperative computed tomography is essential before committing to an approach. In the youngest patients, the impact of such surgery on subsequent craniofacial growth remains unknown.

Pediatric-specific equipment

With the creation of an adequate endonasal corridor, the pediatric nose allows for the use of most standard endoscopic skull base surgical instrumentation without modification. However, for particularly narrow anatomy, the use of a 2.7 mm endoscope can greatly improve technical freedom while maintaining acceptable visualization. 2.7-millimeter endoscopes are available in both 0-degree and angled iterations, though in our experience the use of a standard 4 mm scope is possible in the vast majority of cases.

Creation of the endonasal corridor

After induction of general anesthesia, the patient is positioned with the head and upper body elevated to 30-45 degrees to decrease intracranial pressure and reduce bleeding. If intraoperative neurophysiologic monitoring is employed, paralytic agents are avoided. For most craniopharyngioma patients, intraoperative monitoring of somatosensory evoked potentials and electromyography for perfusion is advisable; should the tumor invade the cavernous sinus, monitoring of select cranial nerves can be considered. Corticosteroids are generally avoided unless there is significant preoperative visual dysfunction or if the patient has preoperative hypocortisolemia. The use of mannitol and other pharmacologic methods of lowering intracranial pressure are often unnecessary, but can be considered. A Foley catheter is placed to monitor urine production. Bilateral oxymetazoline- or epinephrine-soaked

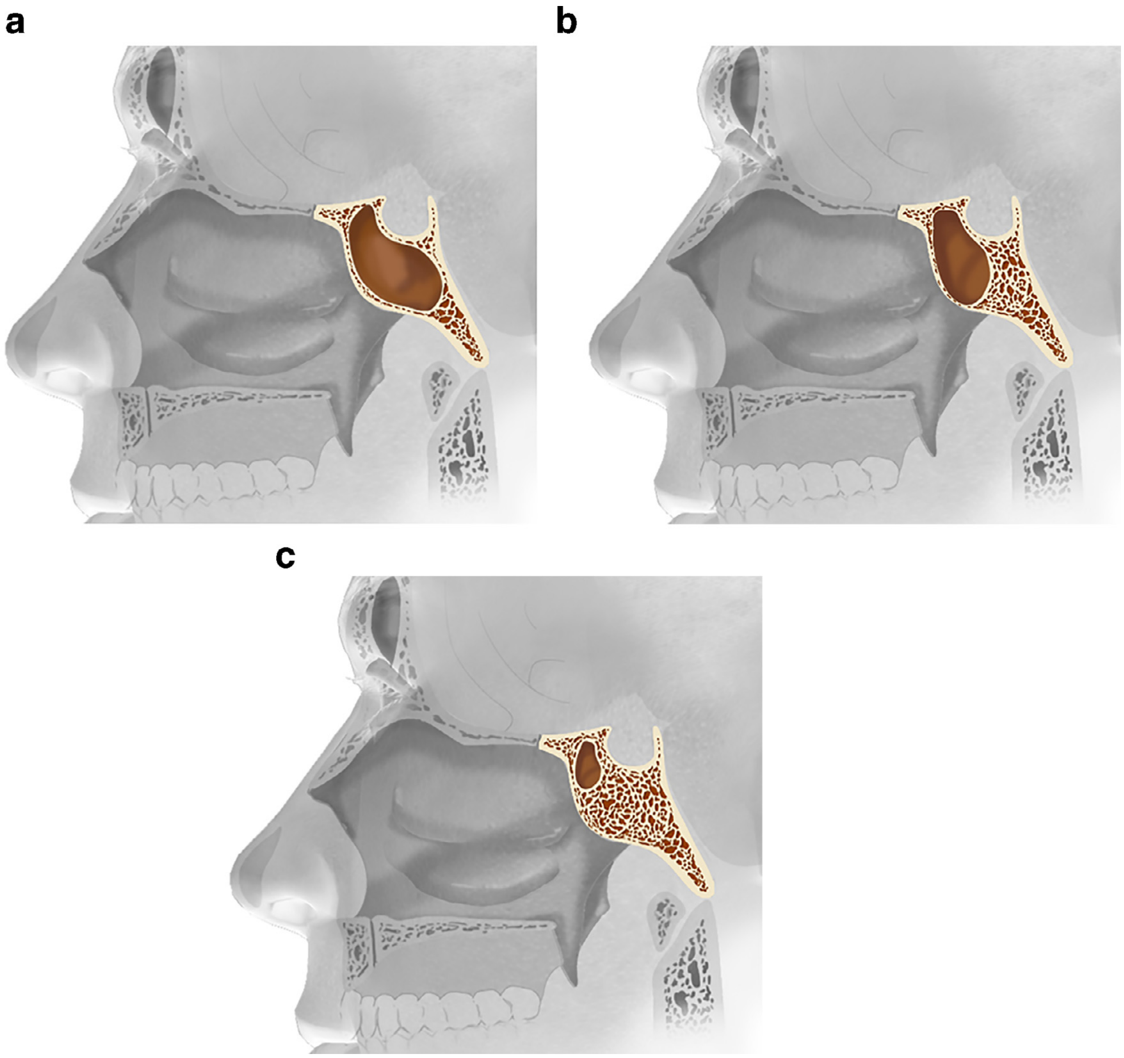

Figure 1 (a-c) Patterns of sphenoid pneumatization. Sellar (a), presellar (b), and Conchal (c). The sphenoid sinus undergoes an expansion of pneumatization between ages 6-10 and is not complete until age 15-16. The ultimate degree of pneumatization is variable.

cottonoids are placed under direct vision before the patient is prepped and draped.

The initial operative step is bilateral nasal endoscopy, both to correlate any pertinent radiographic findings such as septal deflection, and to aid in planning the surgical approach. Local anesthetic is injected bilaterally, including into the nasal septum. At our institution, we favor anatomic preservation whenever possible; as such, middle turbinectomy is rarely employed. Instead, the inferior, middle, and superior turbinates are carefully and completely lateralized to reveal the sphenoid face broadly, with care to not disrupt the cribriform plate superiorly. The sphenoid ostia are identified bilaterally.

In the majority of cases, the anticipated reconstructive needs dictate the use of vascularized local tissue transfer. Thus, we routinely harvest a pedicled nasoseptal flap prior to entering the sphenoid sinus (Figure 2). Flap harvest in the pediatric nose is similar to that used in adult surgery. The flap is based on the posterior nasal artery, a distal branch of the sphenopalatine artery, which courses from lateral to medial across the sphenoid rostrum, inferior to the sphenoid ostium and superior to the choana. Most often, needle-tip electrocautery is used for flap elevation; the power is maintained at a very low setting (5-7 Watts) to minimize thermal injury beyond the incision or perforation of the septum. Some authors recommend cold steel incisions superiorly around the olfactory mucosa adjacent to the cribriform plate to avoid loss of olfaction. An initial incision is made at the height of the choanal arch and is carried inferiorly along the posterior margin of the nasal septum onto the nasal floor; care must be taken to make this incision over the vomer bone and hard palate and not over the muscle of the soft palate. A second incision is started at the top of the sphenoid ostium and carried anterosuperiorly and horizontally along the top of the septum, with care to maintain at least 1 cm of olfactory epithelium below the cribriform plate. The remaining borders of the flap can be modified based on the anticipated size of the bony and dural defects, but are limited by the mucocutaneous junction anteriorly, 1 cm below the cribriform plate

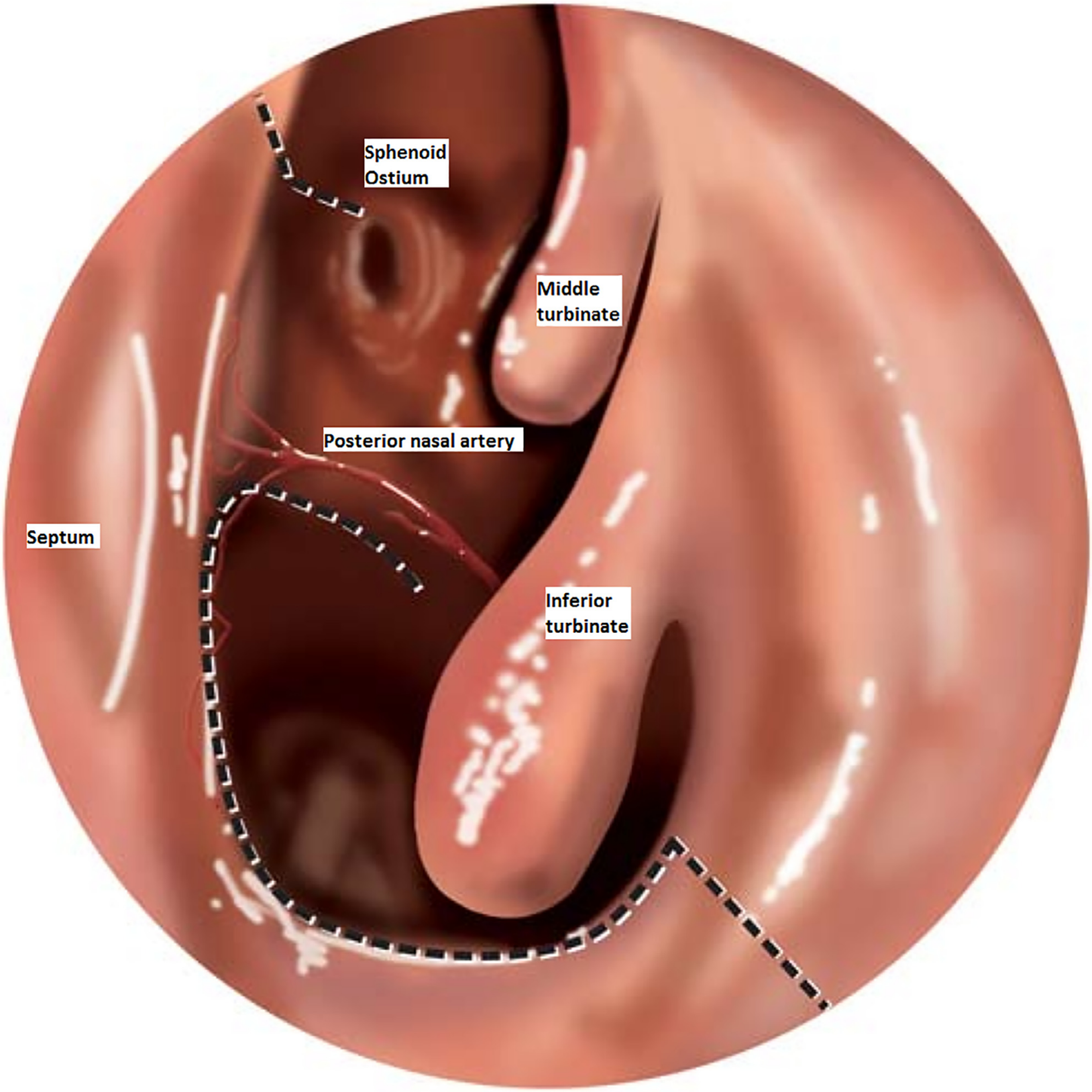

Figure 2 Nasoseptal flap incisions. Incisions can be varied to accommodate the size required. Limitations include the mucocutaneous junction anteriorly, 1 cm below the cribriform plate superiorly, and, when necessary, laterally under the inferior turbinate and onto the medial maxillary wall for extended width.

superiorly, and can safely be carried onto the nasal floor to increase flap width. The inferolateral margin can be further extended under the inferior turbinate and onto the medial maxillary wall, should further flap width be required. The flap is elevated in the subperichondrial and subperiostial plane and is elevated as far laterally off the sphenoid rostrum as possible. The mucosal surface is then marked with a surgical pen to distinguish it from the perichondrial surface, and the flap can be housed in the nasopharynx or maxillary sinus until the reconstructive portion of the case.

At this point, the mucosa is elevated off of the contralateral sphenoid face (also preserving that vascular pedicle to the contralateral septum), and the bony sphenoid rostrum is removed. The sphenoidotomy is widened from the planum to the floor of the sinus and out to the lateral walls of the sphenoid. Posterior ethmoid air cells can be removed as well if wider access is required. If, however, the need for a nasoseptal flap is not clear at the outset, the surgeon can employ one of several flap-preserving approaches to the sphenoid, including a rescue flap,[19] a “raise-and-return” technique, or a “push-down” technique. At our institution,

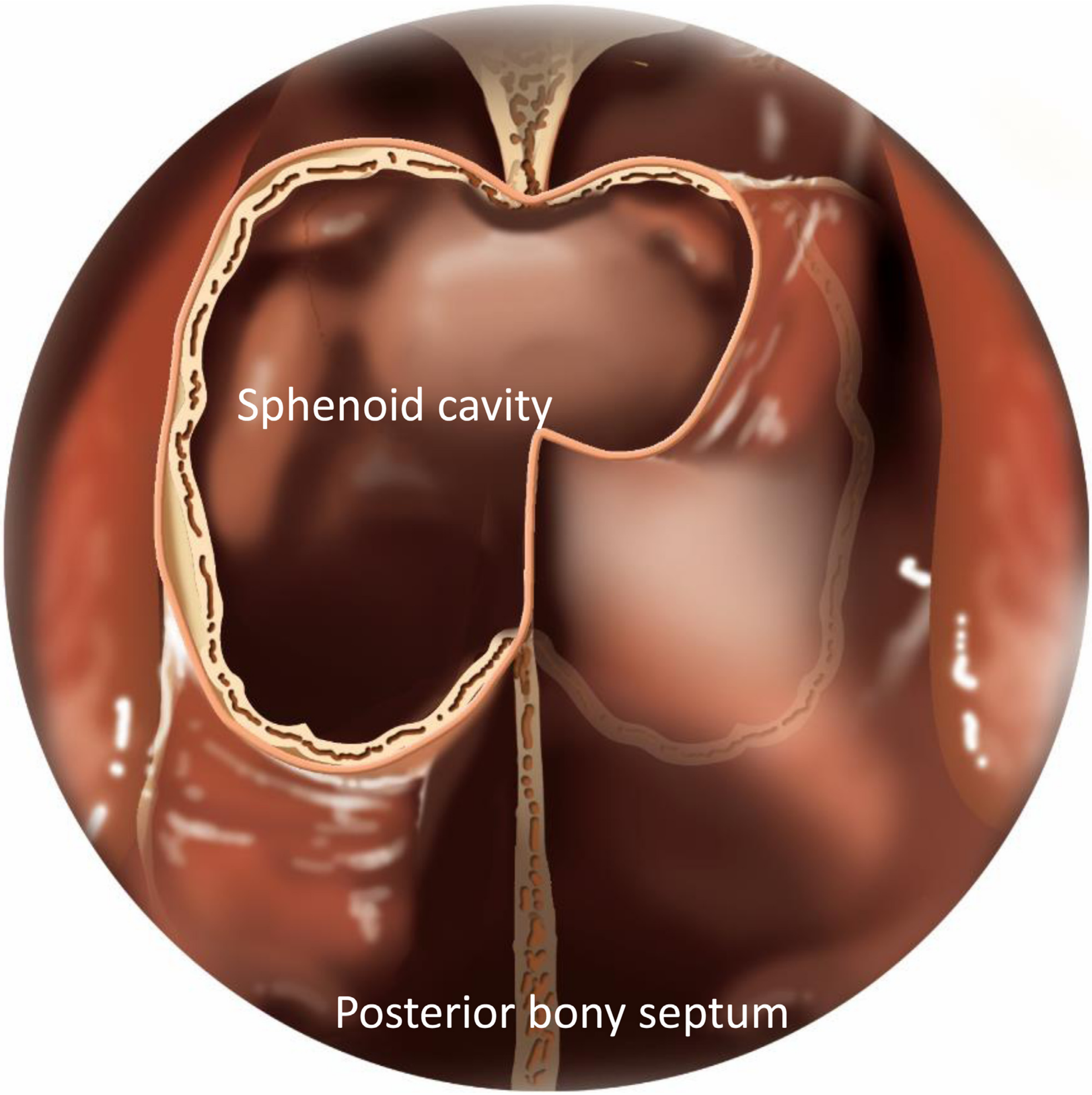

Figure 3 "1.5 with a pushdown" sellar approach. If a nasoseptal flap is not elevated at the outset, we most often employ this flap-sparing approach to the sella. A larger right-sided sphenoidotomy is created (the "1.0" side) together with a smaller left sided sphenoidotomy (the "0.5" side). Both vascular pedicles are elevated off of the sphenoid face and maintained.

we most often employ a "1.5 with a pushdown technique" (Figure 3). In this technique, a vertical mucosal incision is made through the right side on the posterior septum above the sphenoid os, at the point at which the bony septum meets the sphenoid rostrum. The bony septum is fractured off the face of the sphenoid and the mucosa is elevated off the face of the sphenoid on the left side. On the right side the mucosa along the inferior sphenoid face, including the artery, is carefully elevated off of the bone and reflected inferiorly ("pushdown"), thus preserving the nasoseptal flap pedicle. The bone over the face of the sphenoid is removed as above. The mucosa superior to the sphenoid os on the left is resected and a small posterior septectomy can be performed.

Any obstructive septal deflections are conservatively managed at the outset. If septoplasty is required to access the sphenoid sinuses, care is taken to remove as little septal bone and/or cartilage as necessary to maintain the integrity of the nasal structure and avoid inadvertent trauma to the septum during the remainder of surgery.

If a nasoseptal flap is to be used in reconstruction, all mucosa is meticulously removed from each sphenoid sinus to avoid postoperative mucocele formation and to allow the flap to adhere to the bony skull base. Any intersinus

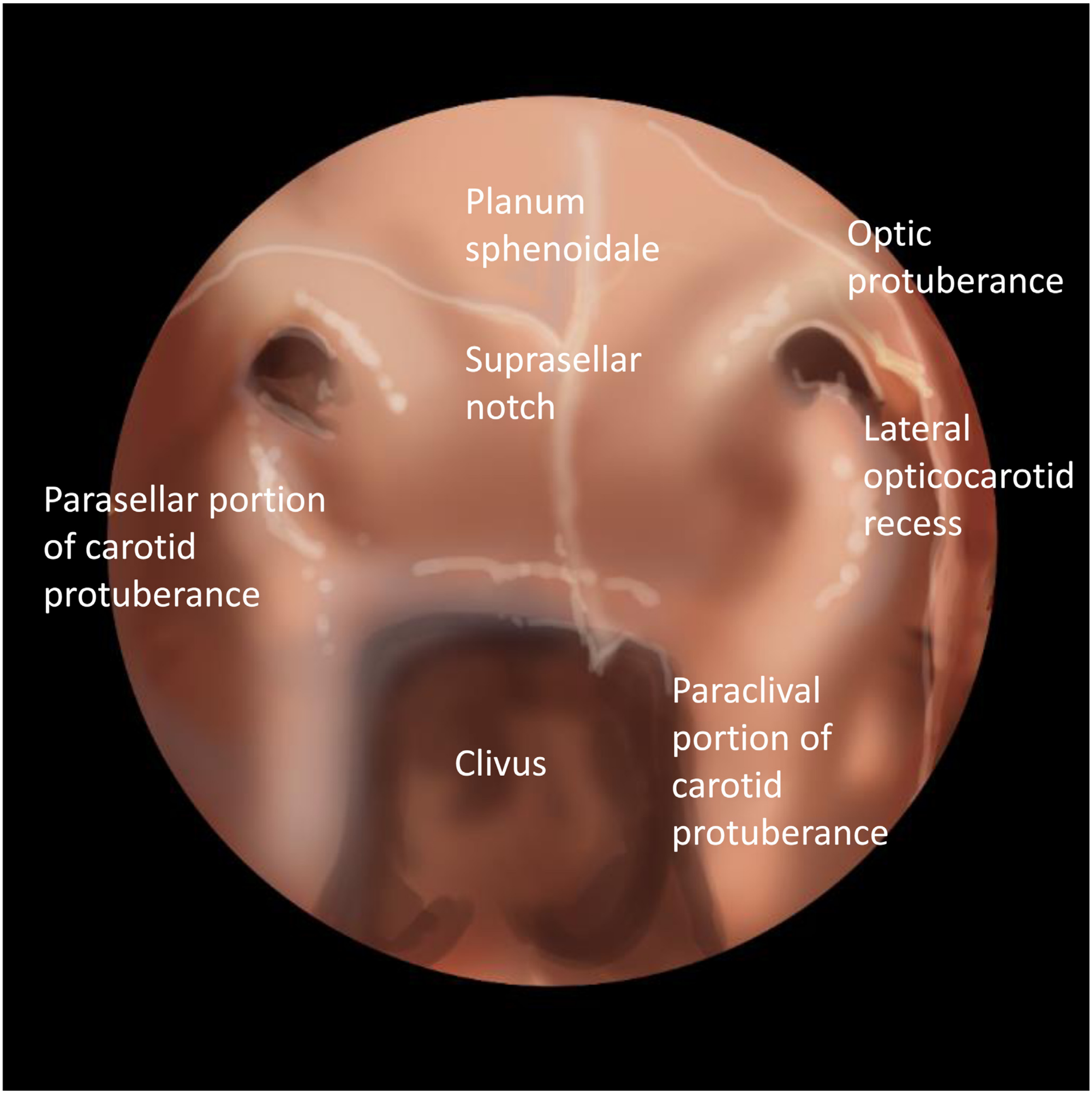

Figure 4 Surgical landmarks of the sphenoid sinus. Key landmarks include the sella, planum sphenoidale, tuberculum sella, lateral optico-carotid recesses (OCRs), optic canals, and the clinoidal carotid protuberances.

septations are carefully removed using either sharp or powered instrumentation, while noting that such septations often attach to the carotid arteries or optic nerves posterolaterally. The inferior face of the sphenoid is also removed bilaterally down to the clivus, to minimize any potential dead-space created by the clival recess and to increase the effective length of the flap.

Cranial base exposure and tumor resection

Once within the sphenoid, the key anatomical landmarks including the sella, planum sphenoidale, tuberculum sella, lateral opticocarotid recesses, optic canals, and clinoidal carotid protuberances are identified (Figure 4). The bone from mid to upper sella is removed with combination of a diamond drill and Kerrison punches. Next, the bone of the tuberculum sella and the adjacent planum is removed to expose the limbus sphenoidale. The use of a diamond bur is safer than a cutting bur when working around critical neurovascular structures and is also helpful for bony hemostasis. Continuous irrigation is used to avoid thermal injury to the optic nerves. The bone overlying the proximal optic canals is unroofed (Figure 5). The lateral opticocarotid recesses help estimate the location of the canalicular segment of the optic nerve.

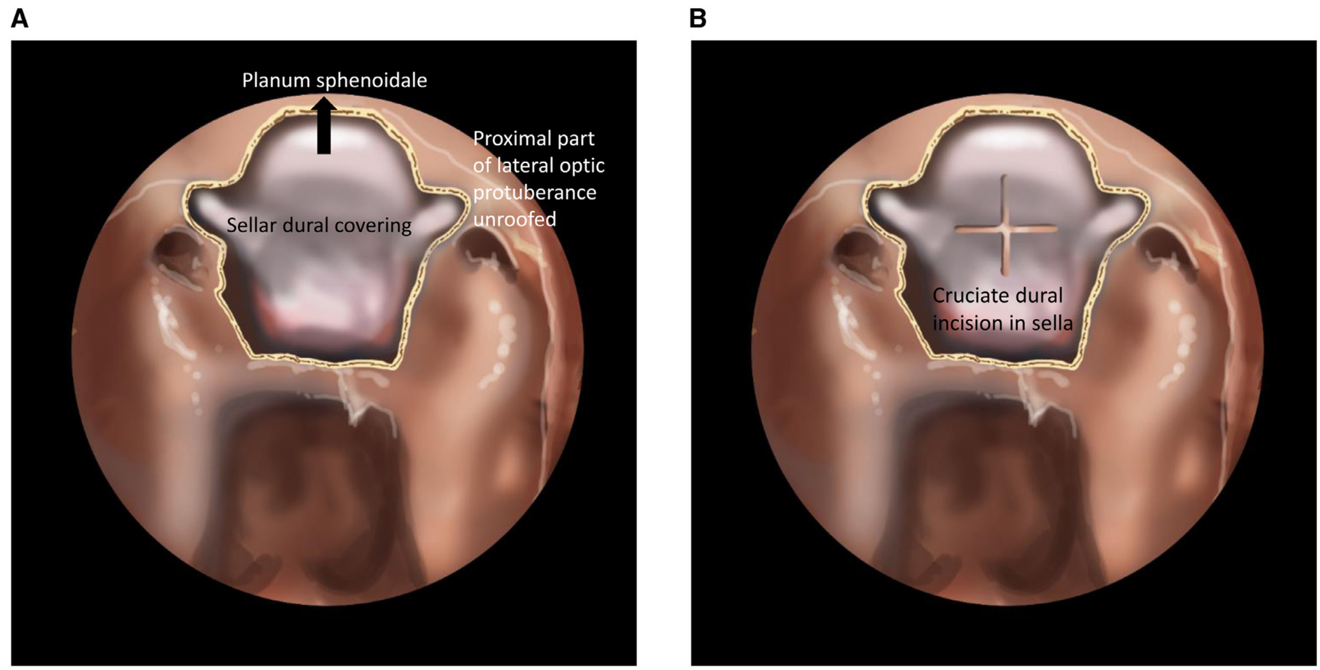

Figure 5 Dural exposure. A highpowered diamond drill with continuous irrigation is used to remove the sphenoid bone posteriorly. The bone from the mid- to upper sella is removed, followed by that of the tuberculum sella and adjacent planum to expose the limbus sphenoidale. Finally, the bone overlying the proximal optic canals (shown in the figure as "star" mark bilaterally) is unroofed in a "T-shirt" pattern.

In sphenoid sinuses with limited aeration and anatomical landmarks, neuro-navigation and microdoppler are useful adjuncts to determine the location of the optic nerve and the carotid artery. A key to unlocking wide access to the optic-carotid cistern is removal of the lateral strut of the tuberculum sella. Neuronavigation also provides useful information regarding the size of the bony osteotomies required along the skull base to allow access to the craniopharyngioma.

Once the bony removal is completed, one can proceed with the resection of the tumor, which has been previously described in detail.[20,21] A vertical incision is made in the midline from the superior aspect of the pituitary gland up to the inferior aspect of the optic chiasm. Care must be taken to avoid injury to the chiasm when making the dural incision as it may be displaced anteriorly, prefixed, or displaced inferiorly onto the diaphragm sella. A strategy we use is to make an initial small incision and, upon development of a plain in the arachnoid between the chiasm and dura, to use endoscopic scissors to complete the dural opening. The superior intercavernous sinus must be controlled by either bipolar electrocautery or injection of hemostatic agents into the sinus. If needed, the dural opening can be expanded laterally with the use of microscissors or Kerrison punches. Once the durotomy is completed, initial arachnoid dissection is centered at the midline, identifying and mobilizing the superior hypophyseal branches to the optic apparatus superolaterally. The cyst capsule is coagulated and opened to perform central debulking of the craniopharyngioma (Figure 6a and b). Care is taken to minimize as much as possible the spillage of cyst fluid into the subarachnoid space. After adequate central debulking is completed, extracapsular dissection is then performed under direct visualization using sharp dissection. This is often aided by the use of a 30-degree endoscope to visualize the lateral and posterior borders of the tumor capsule. Blunt dissection is minimized to avoid inadvertent avulsion of small perforating vessels. An attempt to preserve the infundibulum may be made if the stalk is not extensively invaded. However, the stalk is transected when necessary in order to achieve gross total resection, as anatomical stalk preservation does not necessarily predict functional preservation.[22]

Reconstruction

Once the removal of the tumor has been completed, we proceed with the reconstruction of the dural defect. Among centers performing this type of surgery, there are significant variations in techniques and materials used for dural reconstruction. A common theme, however, is a stable primary dural repair followed by a pedicled vascularized mucosal flap. For the primary dural repair, we use a fascia lata bilayer inlay/onlay "button" graft as we have previously described[23] (Figure 7a-c). Using a separate set of sterile instruments, the fascia lata is harvested and the small lateral thigh incision closed without use of drain. The size of the dural defect is measured using the tips of a pituitary rongeur as a caliper, and an appropriately sized button graft is fashioned. The 2 layers of fascia lata are sutured together using 4-0 Nurolon suture. This helps to

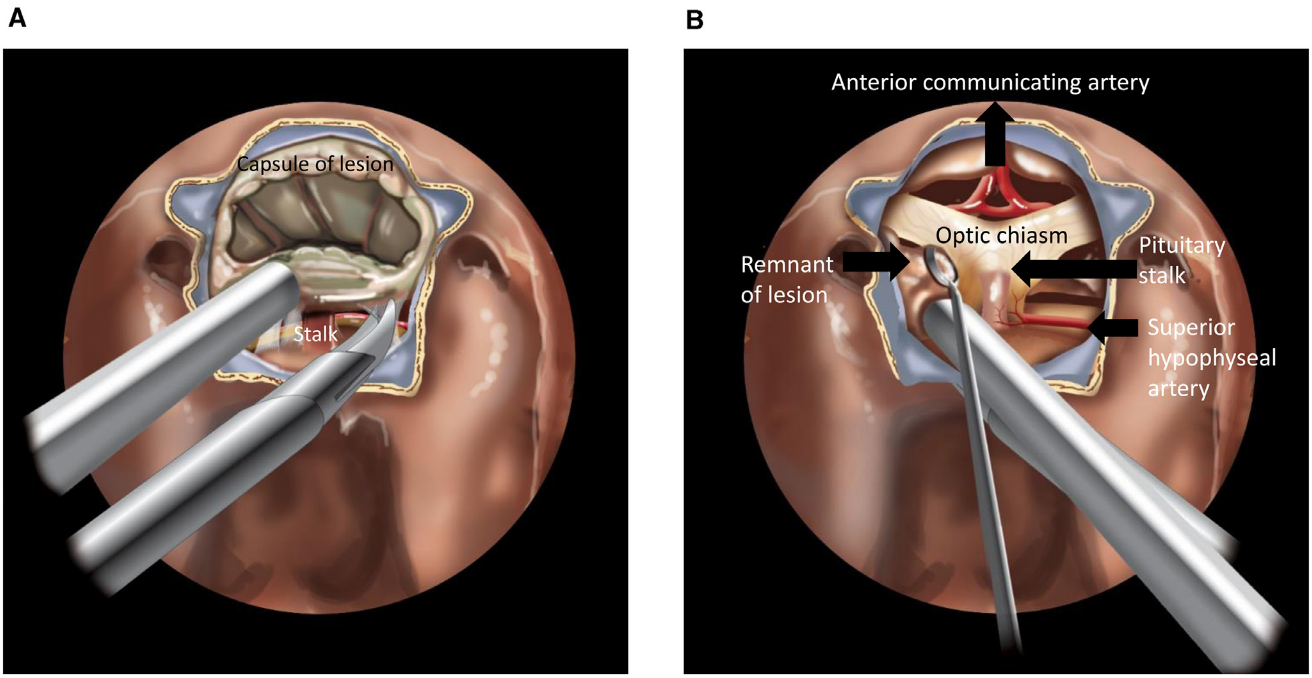

Figure 6 (a) Tumor dissection. After durotomy, the cyst capsule is coagulated and opened to perform central tumor debulking, followed by extracapsular microdissection using sharp technique. (b) Tumor dissection. Laterally, tumor is carefully dissected to achieve complete gross resection. A1 and A2 denote the first (horizontal) and second (vertical) segment of anterior cerebral artery, ACom denotes Anterior communicating artery, DS denotes Dorsum sella and the "star" mark denotes area of third ventricle.

hold both layers in close approximation to the dura and doubles the surface area for healing. The button graft can also be safely applied around critical neurovascular structures and the optic nerves. Once in adequate position, typically there is no evidence of Cerebrospinal fluid (CSF) egress and the graft transmits the normal dural pulsations, indicating a water-tight closure and that a good seal had been achieved. The nasoseptal flap is then placed over the dural reconstruction and surrounding sphenoid bone with care to ensure that there is no dead space between the flap and bone. Polyethylene glycol hydrogel glue is applied to secure the edges of the nasoseptal flap. Absorbable nasal packing may be used to buttress the graft. The middle and inferior turbinates are then medialized. Absorbable nasal packing is placed in the middle meatus bilaterally to keep the middle turbinates medialized.

Prior to extubation, discussion with the anesthesia providers is crucial to avoid bucking or Valsalva during emergence, which could elevate intracranial pressure and threaten the dural reconstruction. Additionally, positive pressure ventilation must be avoided after extubation. As such, extubation at a moderate depth of anesthesia with the patient spontaneously ventilating is optimal. Either topical or intravenous lidocaine may reduce the airway reflex during emergence[24] as may the administration of remifentanil or dexmedetomidine during emergence, with the latter demonstrating less respiratory depression.[25] Before extubation and after, the patient's head of bed is kept elevated to 45 degrees to further reduce intracranial pressure.

Postoperative care

The patient is monitored in a neurosurgical critical care or intermediate care setting, with particular attention paid to monitoring endocrine function. Urine output is monitored together with sodium levels to detect diabetes insipidus, and cortisol levels are followed serially. Gross visual acuity and motor function are assessed as early after emergence from anesthesia as possible. The patient is monitored closely for clear rhinorrhea or salty-tasting postnasal drainage which may suggest a CSF leak; if suspected, early operative exploration and revision is highly recommended. We often opt for immediate postoperative imaging to assess for decompression or to detect residual tumor, with all patients undergoing imaging at 3 months for surveillance.

Nasal debridement is attempted at 1 week, 3 weeks, and 7 weeks postoperatively, but may represent a challenge in the pediatric population. Though many pediatric patients are tolerant of endoscopy and gentle suction, any undue healing may require operative intervention for debridement of dense crusting or for division of synechiae. These can be avoided by employing tissue-preserving techniques during surgery, minimizing exposed bone when possible, and optimizing postoperative nasal hydration. We routinely use a humidified face tent during postoperative admission, as well as frequent topical nasal saline sprays and, when possible, nasal saline irrigations until follow up.

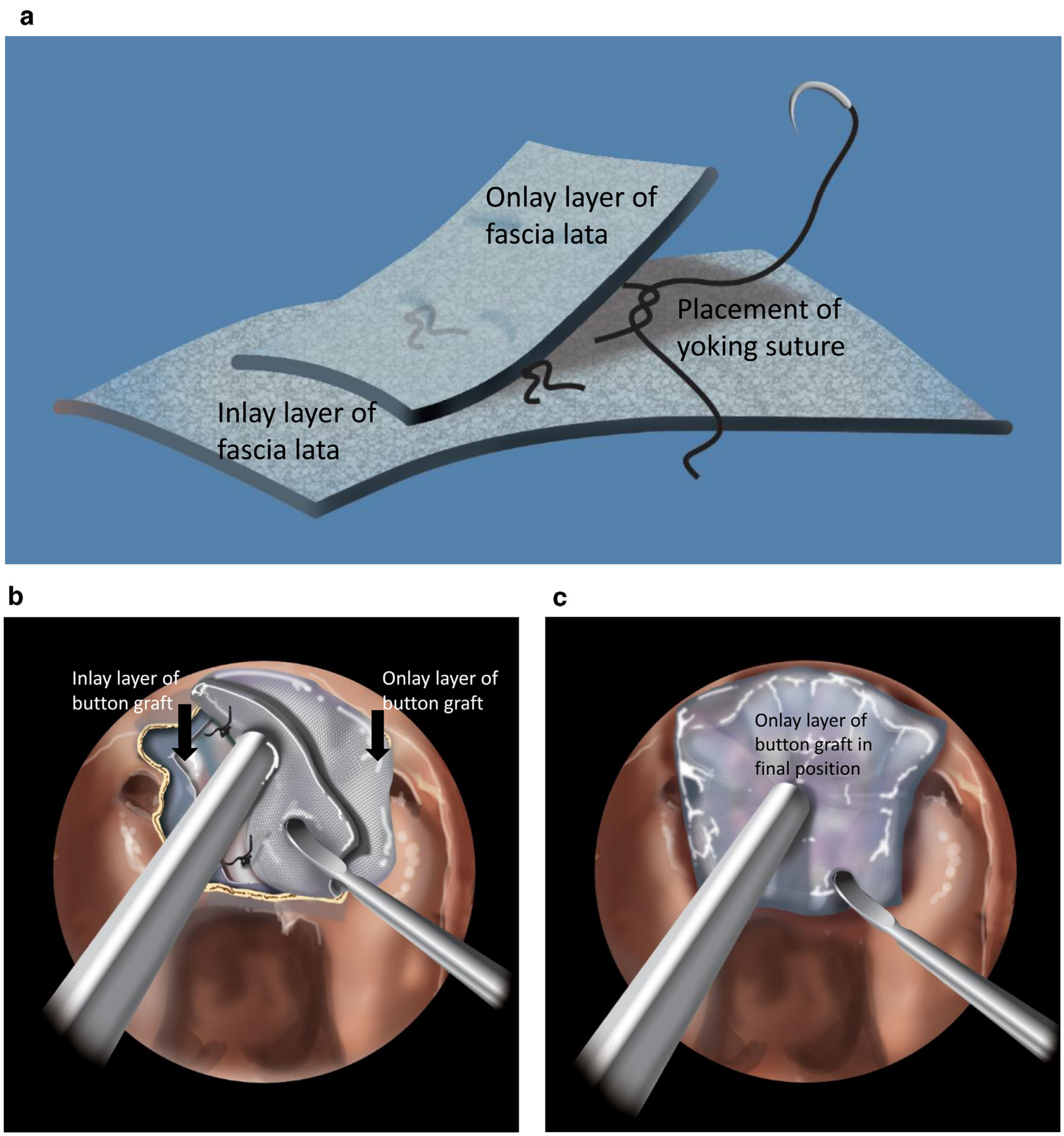

Figure 7 (a-c) Dural reconstruction. We most often employ a fascia lata inlay-onlay graft, or "bilayer button" technique. The dural defect is measured and the graft is created on the back table, with the 2 layers secured to one another by 4 sutures approximating the size of the dural defect. Both the inlay and onlay components of the graft are thus secured at the site of the defect, with the pliability of the fascia allowing the graft to be durable in multiple spatial planes.

Conclusion

Pediatric craniopharyngiomas continue to represent a significant therapeutic challenge and require the consideration of open transcranial, endoscopic endonasal, and stereotactic radiation treatment modalities.[26-28] Even within the confines of the pediatric nasal corridor, the endoscopic endonasal approach represents a viable option for appropriately selected tumors[29] and can be optimized by meticulous attention to the creation of an adequate nasal corridor, microsurgical tumor resection, and robust skull base reconstruction.

Conflict of Interest

None.

Disclosure

The authors reported no proprietary or commercial interest in any product mentioned or concept discussed in this article.

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