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Surgical management of clival chordomas in children

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KEYWORDS
Chordoma;
Clivus;
Endoscopic endonasal surgery;
Pediatric tumors

Objective: Describe the surgical management of clival chordomas in the pediatric population, with an emphasis on techniques of endoscopic endonasal surgery.
Finding: Endoscopic endonasal approaches to the clivus provide access from posterior clinoids to foramen magnum. The clivus is divided into thirds (superior, middle, and inferior), each with corresponding vascular and neural structures. The abducens nerve is at greatest risk for injury due to spread of tumor within the interdural space to Dorello's canal. Despite decreased pneumatization of the sinuses in pediatric patients, the same techniques can be applied to this population. The contralateral transmaxillary approach enhances the ability to achieve a gross total resection when tumor extends laterally into the petrous apex. A multi-layer reconstruction with vascularized tissue minimizes the risk of cerebrospinal fluid leak.
Conclusion: Endoscopic endonasal surgery can be safely applied to the pediatric population and provides the best opportunity for complete surgical excision for most tumors.

Introduction

Chordomas are rare neoplasms with an incidence of 0.08 per 100,000.[1] Although they are most common in the third, fourth, and fifth decades of life, chordomas occur in children and represent one of the more common skull base neoplasms encountered in the pediatric population.[2] Chordomas are usually slow growing but locally aggressive malignant tumors with low propensity for metastasis.[3] In the clivus, they typically present as a midline extradural tumor of the bone. They have a propensity for lateral infiltration within the bone and growth within the interdural space. The most common presenting symptoms are diplopia and headache, but lower cranial nerve palsies are more common in children since craniocervical junction involvement is more common. Cranial nerve involvement, particularly the abducens (VI) nerve, is a consequence of interdural growth into Dorello's canal or compression by intradural tumor in the posterior fossa.

Pediatric chordomas can be more aggressive and are often advanced when discovered in the pediatric population.[3] The optimal treatment is a gross total resection via endoscopic endonasal surgery (EES) that is supplemented with an open approach as necessary. Treatment at a multidisciplinary skull base center with extensive experience in EES for both adult and pediatric patients provides the

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Table 1 Divisions of clivus

Division of clivus	Associated vessels	Associated nerves	Regions accessed
Upper (superior)	Basilar artery/ apex, posterior cerebral arteries, superior cerebellar arteries	Oculomotor nerve	Midbrain, floor of 3rd ventricle
Middle	Basilar artery/trunk, anterior inferior cerebellar arteries	Abducens nerve	Pons, petrous apex
Lower (inferior)	Vertebrobasilar junction	Hypoglossal nerve	Medulla, petroclival synchondrosis, jugular tubercle, occipital condyle

best opportunity for complete excision of tumor with the least morbidity. Postoperative radiation therapy, preferably proton beam therapy, is provided postoperatively for most patients. Proton beam therapy provides a more conformal dose distribution and may limit the dose to adjacent neural tissue. The rare poorly differentiated subtype generally occurs in children under age 5 and may recur rapidly regardless of treatment.[4] Chemotherapeutic response has been reported in this group.[5]

The principles of EES apply to both adult and pediatric populations.[2] Generally, the same surgical techniques and tools can be used. In very young patients, small nasal apertures limit instrumentation and there is increased risk of soft tissue injury from powered instrumentation. Rarely, we have used a sublabial approach with entry into the nasal cavity for passage of instruments. The greatest challenge with EES in pediatric patients is incomplete development of paranasal sinuses with poor pneumatization. This makes identification of sphenoid landmarks difficult or impossible, increasing the risk of neurovascular injury. Greater attention to hemostasis is necessary in the pediatric population due to smaller blood volumes. Reconstruction of large and inferior clival defects is also more difficult with a higher risk of postoperative cerebrospinal fluid (CSF) leak. The growth of the midface (nasal septum) is delayed relative to the cranium and a nasoseptal flap does not provide the same degree of coverage as an adult until about age 14.[6]

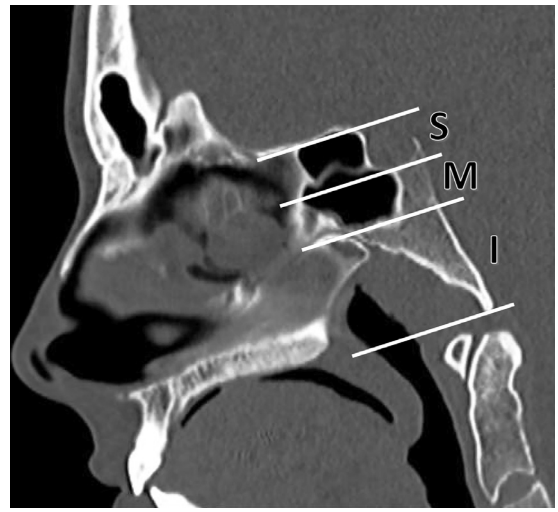

Figure 1 [3D] Anatomic classification of clivus. The junction of the superior clivus (S) and middle clivus (M) is at the floor of the sella. The junction of the middle clivus and inferior clivus (I) is at the floor of the sphenoid sinus.

Transclival approaches

The transclival approach is part of the sagittal plane approaches and provides access to the posterior cranial fossa from the posterior clinoids to foramen magnum. It is useful to divide the clivus into thirds using endonasal landmarks (Figure 1). Each third is associated with specific nerves and vessels (Table 1). The superior clivus extends from the posterior clinoids to the floor of the sella. The middle clivus extends from the floor of the sella to the floor of the sphenoid sinus and provides access to the petrous apex and Dorello's canal. The lower clivus extends from the floor of the sphenoid sinus to foramen magnum and provides access to the medial jugular tubercles and occipital condyles laterally.

Preoperative evaluation and preparation

The initial diagnosis of chordoma is usually made on the basis of radiologic imaging (Figure 2). Both computed tomography (CT) and magnetic resonance imaging (MRI) provide valuable and complementary information.[7] Chordomas are osteolytic lesions with the bulk of the tumor located in the midline. Tumor may involve the upper, middle, or lower thirds of the clivus or craniocervical junction with lateral extension to the petrous apex, jugular tubercle, and occipital condyle. Extraosseous growth of tumor can occur in all directions. On noncontrast CT, chordomas are heterogenous with extensive lytic bone destruction. Intratumoral calcifications may be present. With MRI, chordomas have variable signal intensity on T1 but are characteristically high T2 signal intensity. Contrast enhancement with Gadolinium is typical but not universal.

Review of preoperative scans should include assessment of anatomical variations with special attention to pneumatization of the sphenoid bone, and inflammatory sinus disease. If necessary, sinusitis should be treated with antibiotics prior to surgery.

Biopsy of a clival tumor prior to definitive surgical resection is generally not necessary if radiologic imaging is consistent with a chordoma. With reliable pathology, a

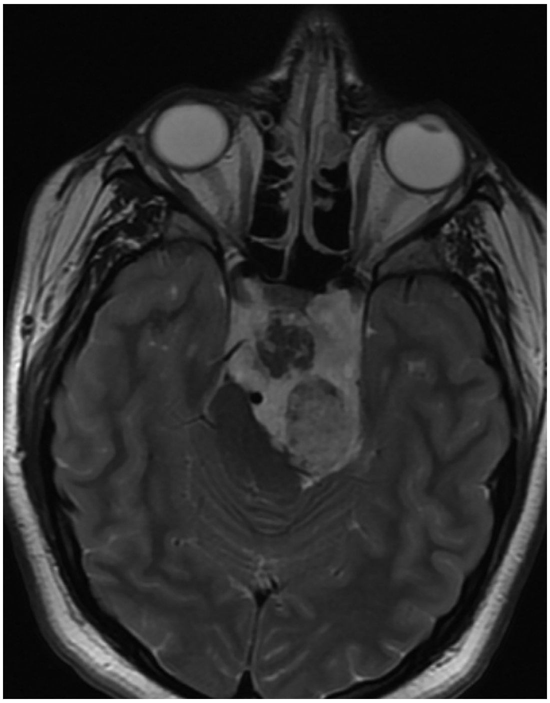

Figure 2 MRI of chordoma in 14-year-old male with left abducens palsy. The tumor is characteristically bright on T2-weighted image. There is encasement of the cavernous segments of both internal carotid arteries and the basilar artery and marked compression of the brainstem.

frozen section can be obtained at the time of surgery to confirm the diagnosis. If there is concern about the diagnosis, definitive surgery can be delayed pending final diagnosis.

Neurophysiological monitoring of brain and cranial nerve function is routinely employed in all transclival skull base procedures.[8] Somatosensory evoked potentials (SSEP) provide an overall measure of cortical function and global ischemia. Brainstem evoked response audiometry (BSER) provides assessment of brainstem function. Free-run electromyography (EMG) is helpful in identification and avoidance of cranial nerve injury, specifically the abducens nerve.[9,10]

Intraoperative image-based navigation is a valuable adjunct to surgery and aids with the identification of key anatomical landmarks and tumor margins. CT angiogram can be fused with MRI to provide optimal visualization of vascular structures, bony landmarks, and tumor margins.

Operative technique

Devices, instrumentation, and materials

1. Oxymetazoline 0.05% solution
2. 0.5% xylocaine with epinephrine (1:200,000)
3. Stryker navigation system (Stryker, Kalamazoo, MI)
4. Karl Storz rod lens endoscopes (0°, 45°) with high definition camera (KARL STORZ Endoscopy America, Inc., El Segundo, CA)
5. SPIWay protective nasal sleeves (SPIWay, LLC, San Clemente, CA)
6. Karl Storz skull base instrumentation set (KARL STORZ Endoscopy America, Inc., El Segundo, CA)
7. KLS Martin dissector set (KLS Martin, Jacksonville, FL)
8. Feather blade (Mizuho America, Inc., Union City, CA)
9. Karl Storz Take-Apart bipolar electrocautery (KARL STORZ Endoscopy America, El Segundo, CA)
10. Endo-Pen bipolar electrocautery (Sutter Medical, Atlanta, GA)
11. Stryker TPS drill with 4 mm coarse diamond extended drill bits (Stryker, Kalamazoo, MI)
12. Ultrasonic aspirator with bone tip (Stryker, Kalamazoo, MI)
13. Doppler probe (Koven Technology, Inc., St. Louis, MO)
14. Kartush stimulating electrode for neurophysiologic monitoring (Medtronic Surgical Technologies, Jacksonville, FL)
15. Surgifoam (Ethicon US, LLC, Bridgewater, NJ), Floseal (Baxter, Deerfield, IL)
16. Indocyanine Green (ICG) fluorescent dye with Karl Storz ICG endoscope (KARL STORZ Endoscopy America, El Segundo, CA)
17. V-Loc suture (Ethicon US, LLC, Bridgewater, NJ)
18. DuraGen collagen graft (Integra LifeSciences, Plainsboro, NJ)
19. Merocel tampons (Medtronic Surgical Technologies, Jacksonville, FL)
20. Doyle nasal splints (Boston Medical Products, Inc., Shrewsbury, MA)

Preparation

1. The patient is positioned in a supine position with the endotracheal tube secured to the left side of the mouth. An orogastric tube is inserted for aspiration of stomach contents during surgery.
2. A pediatric Mayfield head clamp with doughnut ring support is applied with age-appropriate pressure with neck slightly extended and rotated toward the surgeons.
3. The set-up of the operating theater assumes two right-handed surgeons working from the patient's right side. The location of personnel and equipment is shown in Figure 3.
4. The nasal cavity is decongested with pledgets soaked in 0.05% oxymetazoline.
5. Registration of the image-based navigation system is performed with attachment of the tracker to the Mayfield head holder.
6. Needle electrodes for neurophysiologic monitoring includes SSEP, BSER and EMG (abducens nerve, lower cranial nerves as needed).
7. The surgical site is prepped, including harvest sites for fat and fascia lata.

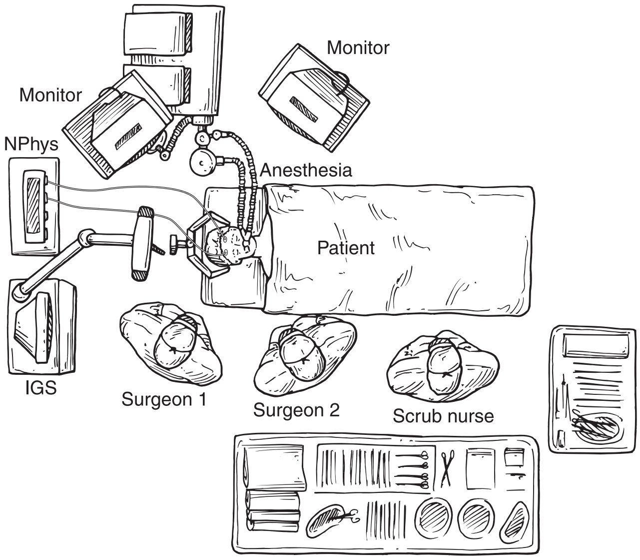

Figure 3 Typical arrangement of operating room for 2 right-handed surgeons. Separate viewing monitors (M) provide good ergonomics. IGS: image-guided system; NPhys: neurophysiology.

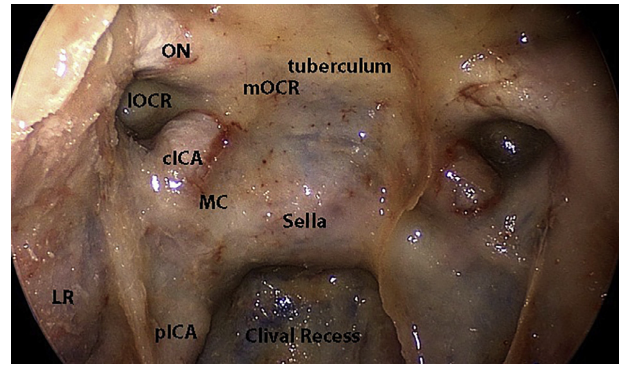

Figure 4 In a well-pneumatized sinus, bony landmarks are readily apparent: sella, clival recess, cavernous (cICA), and paraclival (pICA) segments of internal carotid artery, middle clinoid (MC), optic nerve (ON), medial optic-carotid recess (mOCR), lateral optic-carotid recess (lOCR), and tuberculum.

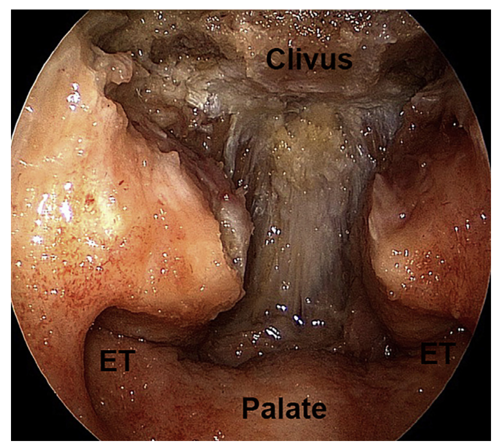

Figure 5 The lower clivus is bounded by the floor of the sphenoid sinus, plane of the palate, and the Eustachian tubes. Removal of the mucosa exposes the longus capitis and rectus capitis muscles.

Surgical approach

Transclival approach (middle and lower clivus)[10]

1. The nasal cavity is examined with a 0° endoscope for anatomical variations/ deformities, presence of rhinosinusitis, and other pathology that may have an impact on the surgery. Turbinates are lateralized, and the nasal septum is infiltrated with local anesthetic with epinephrine for hemostasis. The right middle turbinate can be partially resected to make room for the endoscope; this can even be done bilaterally for very small nasal passages.
2. A nasoseptal flap is elevated from the nasal septum.[11] For clival defects, the flap is extended to include mucosa from the nasal floor and inferior meatus. This creates a wider flap; this maximizes the vertical dimension of the flap since it will be oriented horizontally. Laterality of the flap depends on the location of the tumor and nasal/septal anatomy. With a transclival approach, the flap interferes with the approach. A wide middle meatal antrostomy is performed and the flap can be tucked into the sinus to get it out of the way. For lower clival or craniocervical junction tumors, the flap can be placed into the upper sphenoid sinus.
3. The posterior half of the cartilaginous and bony septum is removed. Releasing incisions for a reverse septal flap are made and contralateral posterior septal mucosa is transposed and used to cover the donor site of the nasoseptal flap anteriorly.[12]
4. SPIWay protective nasal sleeves are inserted bilaterally if the nares are not too small.
5. The rostrum of the sphenoid bone is removed in the midline with bone rongeurs or by drilling to create openings into the sphenoid sinus. The opening is maximally enlarged with Kerrison rongeurs; intrasinus septations are reduced and mucosa is stripped to expose the underlying bone. Key landmarks are identified with the aid of surgical navigation: sella, clival recess, internal carotid arteries, optic canals, lateral optic-carotid recess (Figure 4). In pediatric patients, pneumatization of the sphenoid sinus is often limited and bony landmarks may not be apparent. In such patients, dissection commences in the midline with exposure of the sella using available landmarks and image guidance to carefully drill down to the dura. Epidural dissection along the floor of the sella identifies the entrance of the paraclival ICA laterally.
6. Exposure of the lower clivus requires resection of the nasopharyngeal mucosa and underlying muscles (longus capitis, rectus capitis) (Figure 5). Needle-tip monopolar electrocautery is used to incise the soft tissues medial to the fossa of Rosenmuller. Inferiorly, a transverse incision is made at the level of C1 (anterior arch). The soft tissues are resected using through-cutting instruments or electrocautery. If not involved with tumor, these tissues can also be reflected inferiorly as an in-

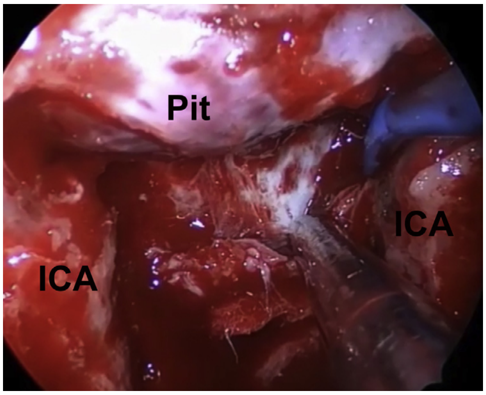

Figure 6 The bone of the middle and lower clivus between the paraclival segments of the internal carotid arteries (ICA) is drilled to expose the dura overlying the brainstem. The outer layer of dura is stripped to expose the basilar plexus and tumor growing between the layers of dura. A Kartush stimulator is used to identify the course of the abducens nerve within Dorello's canal. Pit: pituitary gland.

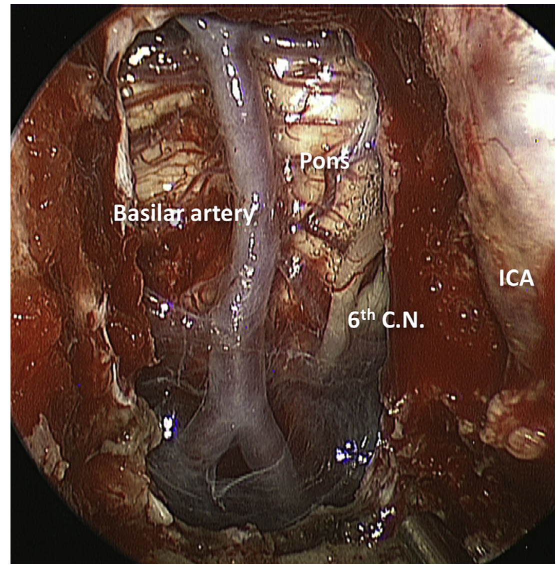

Figure 7 The abducens nerve (sixth C.N.) exits the brainstem at the vertebrobasilar junction and courses superolaterally to pass interdurally and enter Dorello's canal. ICA: internal carotid artery.

verted "U-shaped", retropharyngeal (RP) flap. This can help with reconstruction of lower clival or craniocervical junction defects and separate the oropharynx and nasopharynx. Nodules of tumor that extend into the retropharyngeal space can often be delivered en bloc via traction on involved fascia. This can even be supplemented by pressing on the tumor via the oropharynx to deliver it rostrally into the field. The basopharyngeal fascia is very adherent to the underlying bone and is difficult to dissect. It is helpful to drill the outer cortical bone of the clivus to elevate the fascia.

7. The bone of the middle and lower clivus is then drilled between the paraclival ICAs. In young patients, the synchondrosis of the occipital and sphenoid bones is prominent and can be followed in a posterior-superior direction as the bone is drilled. Tumor-involved bone is removed to the underlying dura.
8. Clival chordomas often spread between the periosteal and meningeal layers of the dura within the basilar plexus. The outer layer of dura can be stripped to expose this space and remove tumor (Figure 6). Bleeding from the basilar plexus is controlled with injection of morselized Gelfoam (Surgifoam [Ethicon US, LLC, Bridgewater, NJ], Floseal [Baxter, Deerfield, IL]) into the space using a suction instrument attached to a syringe.
9. The abducens nerve is associated with the middle clivus (Figure 7). It passes interdurally just above the approximate midpoint of the paraclival ICA (over the petrosal process) and is at risk from drilling and dissection in this area. Deep to the artery, it enters Dorello's canal. Interdural tumor often follows the course of the nerve laterally and must be carefully dissected to avoid injury to the nerve.
10. If there is tumor involvement of the inner layer of dura, resection of dura, and intradural dissection are performed after bone margins have been cleared circumferentially. The tumor is typically soft and yields to gentle suction and blunt dissection technique, but sharp dissection is critical for arachnoid adhesions. It is critical to avoid pulling of tumor since it may be adherent to underlying vessels. Intradural bleeding is controlled with precise bipolar electrocautery and warm saline irrigation. The abducens nerve exits the brainstem at the vertebrobasilar junction and courses superolaterally (Figure 7). A Kartush nerve stimulator employed as a dissecting instrument aids localization of the nerve (Figure 6). The nerve is very fragile and is easily injured. Measurement of a nerve stimulation threshold before and after dissection provides prognostic information following manipulation of the nerve.

Pituitary transposition (superior clivus)

1. If tumor involves the superior clivus, a pituitary transposition is performed to gain access to the dorsum sella and posterior clinoids deep to the pituitary gland.[13]
2. The bone overlying the sella and cavernous ICAs is thinned with a drill and carefully elevated with Kerrison rongeurs, recognizing the course of the cavernous ICA. It is helpful to also remove the bone of the tuberculum sellae so that the transposed pituitary gland is not compressed against the skull base.
3. Small incisions are made between the pituitary gland and cavernous ICA to open the cavernous sinus (Figure 8). Bleeding is controlled by injection of Surgifoam or Floseal. The incision is extended vertically using a hook feather blade (Mizuho America, Inc., Union City, CA).
4. The dura is elevated from the dorsum sella with the gland. Careful suction dissection of the cavernous sinuses reveals the inferior hypophyseal arteries which are cauterized and transected or carefully elevated. This

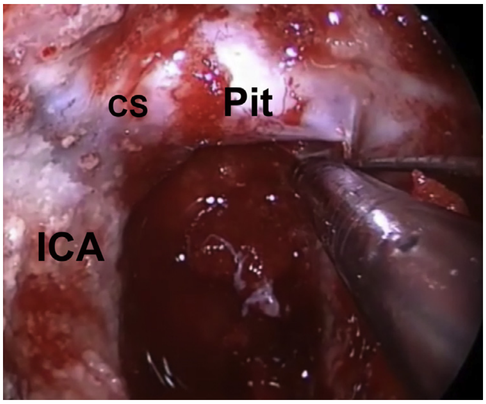

Figure 8 The cavernous sinus (CS) is opened with a right-angle knife between the pituitary gland (Pit) and cavernous segment of internal carotid artery (ICA).

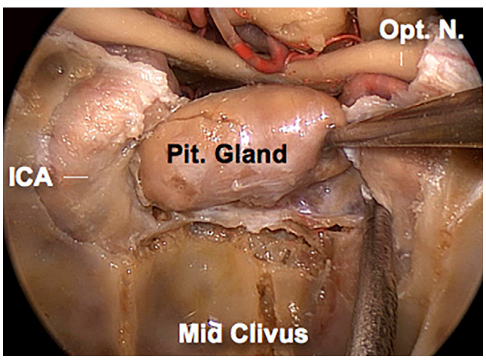

Figure 9 The pituitary gland (Pit) is transposed superiorly to expose the dorsum sella and posterior clinoids. The inferior hypophyseal artery may be sacrificed on one or both sides. Opt. N.: optic nerve; ICA: internal carotid artery.

allows further mobilization of the gland (Figure 9). Unlike the anterior gland, the posterior gland is not protected by dura and is easily injured, resulting in diabetes insipidus.

5. The posterior clinoids are fully exposed and the bone is divided in the midline with Kerrison rongeurs (Figure 10). Each posterior clinoid is removed individually, carefully dissecting and then rotating the bone away from the ICA as it is extricated.

Contralateral transmaxillary approach (petrous apex)

1. The contralateral transmaxillary (CTM) approach provides additional access to the petrous apex for chordomas that infiltrate the bone deep to the petrous ICA (Figure 11).[14] It extends the angle of the approach by about 26° and increases the reach along the posterior surface of the petrous bone by approximately 1.4 cm. For this description, *ipsilateral* refers to the side of the tumor whereas *contralateral* refers to the side of the CTM approach (anterior maxillotomy).
2. After a nasoseptal flap is elevated contralateral to the tumor-involved petrous apex, a medial maxillectomy is performed on the same side as the flap. The inferior turbinate is resected, and the bone of the medial

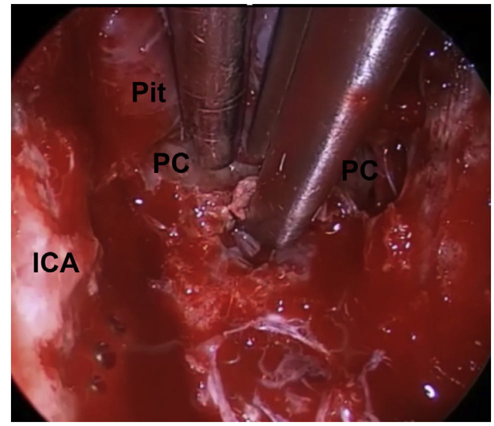

Figure 10 The posterior clinoids (PC) are divided in the midline with a Kerrison rongeur before removing. Pit: pituitary gland; ICA: internal carotid artery.

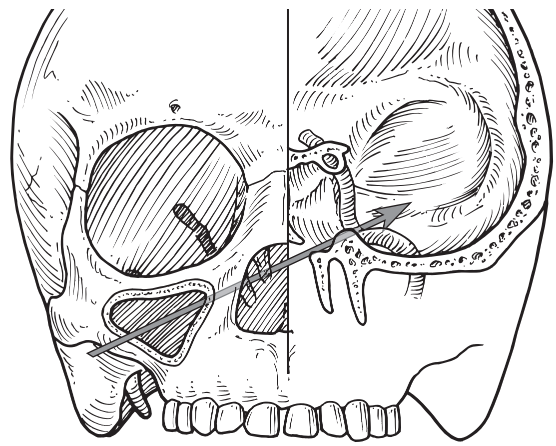

Figure 11 The contralateral transmaxillary approach improves the angle of approach to the petrous apex by approximately 25°.

maxilla is removed to the nasal floor posteriorly. The nasolacrimal duct is preserved anteriorly. The sphenopalatine foramen is identified posteriorly and the bone overlying the vascular pedicle is carefully removed without injury to the vessels.
3. An anterior maxillotomy is performed by making a mucosal incision in the gingivolabial sulcus and elevating the periosteum to the level of the infraorbital nerve. A drill or Kerrison rongeurs are used to maximize the opening into the maxillary sinus, especially laterally. In pediatric patients, care is taken to avoid injury to the roots of unerupted permanent teeth.
4. Once the endonasal drilling of the clivus is complete and the paraclival ICA has been skeletonized, the CTM approach is employed to gain additional access to the tumor-involved petrous apex for drilling or tumor dissection (Figure 12). It is helpful to place a SPIWay nasal sleeve through the anterior maxillotomy to protect the labial tissues and facilitate passage of instruments. The endoscope (0° or 45°) is shifted to the contralateral nasal passage while instruments are passed through the CTM corridor and/or ipsilateral nostril.

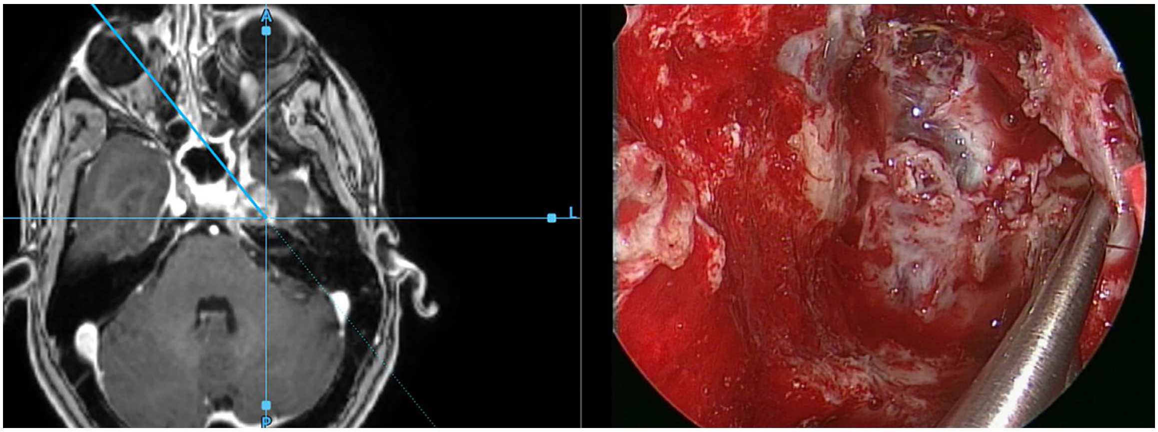

Figure 12 A contralateral transmaxillary approach provides access to tumor-involved bone deep to the paraclival and petrous segments of the internal carotid artery. Image guidance demonstrates the angle and reach of the approach. The left internal carotid artery (paraclival segment) has been lateralized to provide additional exposure.

Reconstruction

1. If the clival chordoma is small and dissection remains extradural, no reconstruction is necessary. The defect can be covered with fibrin glue or Gelfoam for temporary protection and hemostasis.
2. Reconstruction with vascularized tissue is preferred when there is a dural defect or exposure of the carotid arteries. The reconstruction consists of four layers: inlay collagen graft, onlay fascial graft, fat graft, and vascularized flap.
3. Following hemostasis, a collagen graft (Duragen, Integra LifeSciences, Plainsboro, NJ) is tucked intradurally between the brain and dura. The graft should be in contact with the dura circumferentially.
4. Fascia lata is harvested from the thigh using a vertical incision. The graft is harvested from the middle third of the fascia with preservation of the ileo-tibial band laterally. A generous graft is obtained, large enough to cover the surrounding bone for several centimeters. The areolar fat is stripped from the surface of the fascia and it is placed extradurally, extending up the sides of the clival defect to cover the paraclival ICAs (Figure 13). With low clival defects, the inferior margin of the fascia lata is tucked deep to the nasopharyngeal mucosa (retropharyngeal [RP] flap). Alternatively, the fascia lata can be sutured to the mucosal margin of the nasopharyngeal tissue using an absorbable V-Loc suture.[15] This suture has tiny barbs along its length, holding the suture in place and allowing one to run the suture without the need for tying knots.
5. A fat graft is harvested from the leg incision or separate periumbilical incision and placed centrally in the clival defect superficial to the fascia lata (Figure 14). It should just be large enough to fill the defect and create a flat surface for the nasoseptal flap.
6. The nasoseptal flap is transposed to cover the inferior and central parts of the fascia lata (Figure 15). It is oriented horizontally due to the inferior location of the defect.
7. Small postage stamp-size pieces of Surgicel are placed around the edges of the flap and tissue glue or small pieces of Gelfoam soaked in thrombin solution cover the surgical site. Merocel tampons are then inserted bilaterally, taking care to avoid displacement of the reconstructive tissues. In small children, strings attached to the tampons can be cut short so that the child can't pull on them.
8. If a CTM approach is used, the mucosal incision is closed with interrupted absorbable suture using a vertical mattress stitch.
9. A lumbar spinal drain is placed at the conclusion of the surgery if the dura was transgressed.

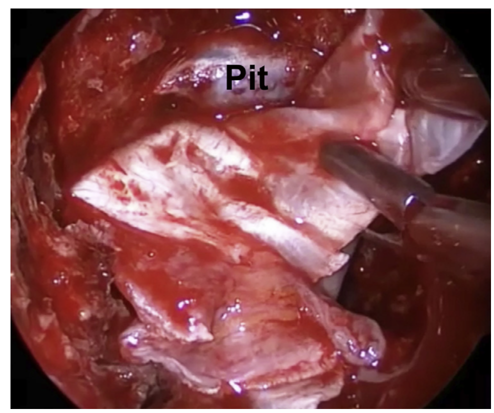

Figure 13 Following placement of an intradural collagen graft, a fascia lata graft covers the entire defect including the exposed carotid arteries. Pit: pituitary gland.

Postoperative care

Immediate concerns are safety of the airway and neurological function.[16] Ideally, patients should be extubated in the operating room unless sedation is needed for immediate

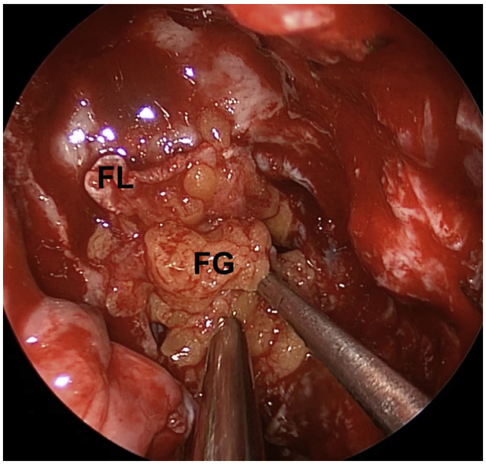

Figure 14 A small fat graft (FG) is placed superficial to the fascia lata (FL) in the clival defect to create a flat surface for the vascularized flap and prevent herniation of the brainstem.

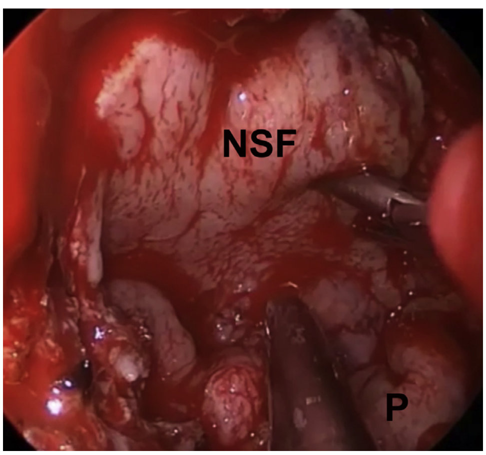

Figure 15 A vascularized nasoseptal flap (NSF) covers the fat and fascia lata grafts. P: pedicle of nasoseptal flap.

postoperative imaging (commonly employed in children). Close monitoring of neurological function is performed in an Intensive Care Unit (ICU) postoperatively. A CT without contrast obtained within 12 hours rules out a significant hematoma or pneumocephalus. Early MRI is useful in assessing the completeness of tumor resection; early reoperation for residual tumor is an option and is recommended if complete resection can be achieved.

Clival defects are the highest risk group for postoperative CSF leak following EES. The risk is minimized with a multilayer vascularized reconstruction and postoperative CSF diversion with a lumbar drain. In a randomized controlled trial of adult patients undergoing EES, lumbar drainage decreased the risk of postoperative CSF leak for transclival EES from 35%-8%.[17] The protocol for lumbar drainage at our institution is 5-10 cc/hour (weight dependent) for 72 hours. The drain is typically removed after 72 hours to minimize the risk of infection. In small children in whom removal of nasal packing is difficult, packing is removed under anesthesia; the integrity of the reconstruction can be assessed at the same time with endoscopy.

Nasal hygiene is limited to saline spray in young patients. Older children will tolerate saline irrigations. Serial endoscopic debridement of nasal crusts following discharge is performed when possible but is often limited due to noncompliance.

Even with a gross total resection, the risk of tumor recurrence remains high following surgery. Most patients with chordoma will benefit from postoperative radiation therapy, either proton beam therapy or intensity modulated radiation therapy (IMRT), depending upon availability.[18] Potential benefits of proton beam therapy include greater conformality of the radiation field adjacent to the brainstem with decreased morbidity. The benefits of radiation therapy must be balanced by the risks in a young population. Small chordomas with gross total resection may be closely monitored for recurrence, especially if they show a low degree of molecular derangement.[19] Postoperative radiation therapy in pediatric chordoma is generally recommended, but can be withheld if radical resection is achieved, especially in incidentally discovered tumors. Poorly differentiated tumors should be considered for chemotherapy in addition to radiation therapy.

Onset of new symptoms, especially cranial nerve deficit, and endoscopic examination may suggest recurrence of tumor. Tumor surveillance consists primarily of serial imaging with MRI, at a minimum of 6-12 month intervals. Depending on the age of the patient, this may require sedation. Seeding, though rare, can occur, typically on the side of blind instrumentation (left nasal passage for right-handed surgeons). This may be decreased by the use of protective nasal sleeves (SPIWay, LLC, San Clemente, CA), but should be monitored closely, even with endoscopy for aggressive tumors.

Complications and management

CSF leak

Postoperative CSF leak can be difficult to diagnose in pediatric patients but is more common than in adults, likely due to smaller septal flaps and larger tumors.[20] With clival defects, drainage is often postnasal without evidence of rhinorrhea. Complaints of a salty taste or nocturnal cough may be the only symptoms. Physical examination is limited by nasal packing and patient compliance. Suspicious drainage from the nose can be tested for beta-2-transferrin to confirm. A CT scan may be obtained to look for evidence of increased or new pneumocephalus. A CT cisternogram is usually not necessary unless the patient has already failed attempts at repair and the site of the leak is not apparent.

If a CSF leak is suspected, endoscopic examination under anesthesia is warranted. All packing material is removed, and the surgical site is carefully inspected with an endoscope. A Valsalva maneuver ("count to 10") is helpful in identifying drainage of CSF. If a leak is confirmed, it is usually limited to a small area, most often at the inferior margin of the reconstruction. Repositioning of tissues, augmentation with additional fat or fascia, suturing of the fascia lata to the nasopharyngeal mucosa, or elevation of a lateral nasal wall flap (inferior turbinate flap)[21] are all options. In recalcitrant cases, we have even employed an extracranial pericranial flap passed along the ethmoid roof on one side.[22] A lumbar spinal drain is usually placed at the time of the repair.

Infection

The risk of meningitis is very low (<3%) in patients undergoing EES.[23] It is important to continue oral antibiotic prophylaxis for the duration of nasal packing. Delay in diagnosis and management of a CSF leak is the biggest risk factor for postoperative meningitis. Lumbar drainage without surgical repair of a CSF leak is to be avoided since this prolongs the opportunity for infection. Necrosis of a nasoseptal flap is a rare complication that can present with delayed infection of the surgical site with meningitis.[24]

CN injury/diplopia

The abducens nerve is at greatest risk for injury with transclival approaches. In patients with preoperative diplopia, the risk of injury is increased, and patients and parents should be counseled about the possible need for corrective surgery. Ophthalmology should be consulted early for postoperative evaluation and counseling, patching or prism lenses if there is diplopia due to an abducens palsy.

Nasal morbidity

Nasal morbidity related to EES includes loss of olfaction, persistent crusting or rhinosinusitis, septal perforation or deformity, and cosmetic deformity. Nasal dorsum collapse has been observed in a small percentage of patients undergoing reconstruction with a nasal septal flap.[25] Long term effects of EES on midfacial growth have been poorly documented and remain a potential concern in young patients, but early radiographic studies do not show a significant impact.[26]

Outcomes

Pediatric chordomas are generally considered to be more aggressive with reported rates of recurrence of 16%-68%.[27] In a limited review of our experience with 10 pediatric chordomas, 2 were recurrences at time of surgery.[27] Of the remaining 8 cases, gross total resection was achieved in 4, and near-total resection (>90%) was achieved in the remaining 4 patients. We observed a recurrence rate of 30%, which is well within the reported range in the literature and does not differ from the adult population. While low prevalence makes it difficult to reach conclusions, it may be that, other than the poorly differentiated tumors, pediatric chordoma can have a promising prognosis with complete resection.

Conclusions

Pediatric clival chordomas are ideally suited for EES due to their central location and infiltrative behavior. An endoscopic endonasal approach provides the best opportunity for a gross total resection with the least morbidity for the patient. Endonasal transclival approaches provide access to the entire clivus from posterior clinoids to foramen magnum and craniocervical junction. An interdural pituitary transposition provides access to the upper clivus with preservation of pituitary function. A contralateral transmaxillary approach augments access to the petrous apex for tumors that extend laterally. A multilayer reconstruction using fascia, fat, and a vascularized flap minimizes the risk of a CSF leak postoperatively as does lumbar drainage. Most patients will benefit from adjunctive radiation therapy due to the high risk of local recurrence.

Disclosures

Dr. Gardner and Dr. Snyderman are consultants for SPI-Way, LLC.

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