

Evolutions in the Management of Congenital Intranasal Skull Base Defects

Bradford A. Woodworth, MD; Rodney J. Schlosser, MD; Russell A. Faust, PhD, MD; William E. Bolger, MD

Background: Congenital skull base defects have traditionally been treated via an intracranial approach. Recent advances in endoscopic management have made minimally invasive extracranial approaches feasible, with less morbidity.

Objective: To determine the success of endoscopic treatment of congenital cerebrospinal fluid leaks and encephaloceles.

Main Outcome Measures: Retrospective review of congenital cerebrospinal fluid leaks and encephaloceles treated from January 1, 1992, to December 31, 2003. Data collected include demographic characteristics, presenting signs/symptoms, site of the skull base defect, surgical approach, repair technique, and clinical follow-up.

Results: Eight patients were treated via the endoscopic approach for congenital cerebrospinal fluid leaks and encephaloceles. The average age at presentation was 6 years (range, birth to 18 years). Three patients presented with meningitis (average age, 6 years), 4 had cerebrospinal fluid

rhinorrhea, and 3 developed a nasal obstruction. Five defects originated at the foramen cecum, and 3 others involved the ethmoid roof/cribriform plate only. Our endoscopic approaches were successful on the first attempt, with a mean follow-up of 19 months. One patient experienced nasal stenosis postoperatively.

Conclusions: Continuing progress in the surgical management of congenital skull base defects demonstrates that endoscopic repair is a successful alternative to traditional craniotomy approaches, with less morbidity. This technique requires meticulous preparation and precise grafting of the defect to avoid collateral damage to surrounding structures. While reduction in the risk of meningitis, intracranial complications, and facial growth abnormalities and alleviation of nasal obstruction necessitate the timely repair of these skull base defects, special considerations are discussed regarding the optimal timing of surgical intervention, operative working space, and exposure in a smaller nasal cavity.

Arch Otolaryngol Head Neck Surg. 2004;130:1283-1288

Author Affiliations:

Department of Otolaryngology–Head and Neck Surgery, Medical University of South Carolina, Charleston (Drs Woodworth and Schlosser); Departments of Otolaryngology and Pediatrics, Children's Hospital of Michigan, Detroit (Dr Faust); and Department of Surgery (Otolaryngology), Uniformed Services University of the Health Sciences, Bethesda, Md (Dr Bolger).

Financial Disclosure: None.

CONGENITAL ENCEPHALOCELES are protrusions of cranial contents beyond the normal confines of the skull. Suwanwela and Suwanwela¹ divided these entities into sincipital (also referred to as anterior or frontoethmoidal) and basal encephaloceles. Frontoethmoidal types originate between the frontal and ethmoid bones, commonly at or anterior to the foramen cecum, and appear at several exit points in the face. Basal encephaloceles are intranasal in location and more likely to be encountered by the pediatric rhinologist. Sites of skull base defects have been variously described as transethmoidal, sphenothmoidal, sphenomaxillary, sphenoorbital, transsphenoidal, and transtemporal.^{2,3} Clinically, the cribriform niche adjacent to the vertical attachment of the middle turbinate anteriorly and the superior and lat-

eral walls of the sphenoid seem to be the most common sites for these congenital defects.⁴ While it can be helpful to separate encephaloceles into categories for research and descriptive purposes, bony dehiscences are found in 14% of ethmoid bones in anatomical studies,⁵ and congenital defects can occur at any point in the skull base; thus, an individualized treatment approach is required for each patient.

**CME course available
at www.archoto.com**

Encephalocele and cerebrospinal fluid (CSF) leak management has changed dramatically during the past quarter century. Historically, neurosurgeons have approached these lesions through a bicoronal incision and frontal craniotomy, and often use a pericranial flap to reconstruct the

Table. Data for the 8 Patients Who Underwent Endoscopic Repair of Congenital CSF Leaks and Encephaloceles*

Patient No.	Presenting Age, y	Presentation	Site of the Skull Base Defect	Repair Technique	Follow-up, mo
1	5	Meningitis and CSF leak	Left anterior cribriform and FC	Fibrin glue and abdominal fat	1½
2	Birth	Nasal obstruction	Right anterior cribriform, ethmoid roof, and FC	Gelfoam (Pfizer Inc, New York, NY)	72
3	Birth	Nasal obstruction	Left anterior cribriform and FC	Temporalis fascia	7
4	8	Meningitis and CSF leak	Right anterior cribriform	Temporalis fascia	6
5	7	Meningitis	Right midcribriform	Temporalis fascia and mastoid cortical bone graft	9
6	18	CSF leak	Left ethmoid roof	Temporalis fascia	26
7	2½	Nasal obstruction	Left cribriform, ethmoid roof, and FC	Composite turbinate graft and hydroxyapatite	12
8	5	CSF leak	Left cribriform, ethmoid roof, and FC	Temporalis fascia	22

Abbreviations: CSF, cerebrospinal fluid; FC, foramen cecum.

*All patients had an encephalocele, except for patient 4.

skull base defect. The pitfalls of this operation include anosmia, intracranial hemorrhage or edema, epilepsy, and memory and concentration deficits.⁶ Fortunately, otolaryngologists are able to avoid many of these complications with an endoscopic approach. The endoscopic repair of CSF leaks and intranasal encephaloceles is well accepted and has become the standard of care for most adult skull base defects.⁷⁻¹⁰

Endoscopic management of congenital skull base defects presents unique challenges to the pediatric rhinologist. Most skull base defect repairs in the published literature have been performed for traumatic, iatrogenic, or spontaneous CSF leaks in adults, but there is a paucity of data concerning the management of congenital skull base defects in the pediatric population. These children present with various signs and symptoms, including CSF leaks, meningitis, nasal obstruction, and craniofacial deformity. In addition, the extremely small size of the sinonasal cavity in these patients presents significant technical challenges. This report outlines our evolution and current treatment algorithm in the endoscopic management of congenital intranasal skull base defects, based on our collective experience in treating this unusual condition over many years.

METHODS

We retrospectively reviewed all cases of congenital intranasal encephaloceles and CSF leaks treated at our institutions from January 1, 1992, to December 31, 2003. Patients met the following criteria: existence of a skull base defect with either an intranasal encephalocele or a CSF leak, presenting signs or symptoms at 18 years or younger, no history of trauma, and no history of intranasal or skull base surgery, excluding prior attempts at repair. Data collected included demographic characteristics, presenting signs and symptoms, site of the skull base defect, surgical approach, repair technique, and clinical follow-up.

The preoperative examination of all patients consisted of a thorough medical history, a physical examination, and radiographic imaging. All patients had standard computed tomographic and magnetic resonance imaging performed. Three-dimensional computed tomographic scans, triplanar reconstructions, and intraoperative computerized image guidance are useful in visualizing the skull base defects, but are not required.¹¹

The surgical technique varied slightly between surgeons, but generally followed one previously outlined.¹² By using 0°, 30°, and 70° adult and pediatric endoscopes, the meningoencephalocele sac was meticulously ablated and reduced with bipolar or suction cautery, followed by removal with through-cutting forceps. A mucosal cuff was circumferentially removed around the bony defect, with attempts to avoid collateral damage to the surrounding sinuses and otherwise healthy structures. If the defect extended into the ethmoid roof and cribriform plate, then uncinectomy and ethmoidectomy were performed to expose the defect. A large bony defect may be reconstructed using mastoid cortical bone or septal bone in an underlay fashion, followed by an overlay soft tissue graft. For smaller bony defects, a temporalis fascia, mucosa, or composite turbinate overlay graft was placed over the bony defect, followed by an absorbable gelatin sponge (Gelfoam; Pfizer Inc, New York, NY) and additional packs as needed for additional support. Lumbar drains were not routinely used.

RESULTS

Eight patients underwent endoscopic repair of congenital CSF leaks and encephaloceles (**Table**). The average age at presentation was 6 years (range, birth to 18 years). Three patients presented with meningitis (average age, 6 years), 4 had CSF rhinorrhea, and 3 developed a nasal obstruction. Five defects originated at the foramen cecum (**Figure 1** and **Figure 2**), and 3 others involved the ethmoid roof/cribriform plate only (**Figure 3**). Two patients underwent prior attempts at repair, one via a bifrontal craniotomy and pericranial flap and the other via an endoscopic approach at an outside facility. Endoscopic approaches at our institutions were successful on the first attempt, with a mean follow-up of 19 months. The only complication was in one patient who experienced nasal stenosis postoperatively. This was successfully corrected with a second procedure using a Z-plasty.

COMMENT

OPERATIVE TIMING

The optimal timing for surgical repair of congenital skull base defects has not been definitively determined. Consideration must be given to the risks and benefits for each

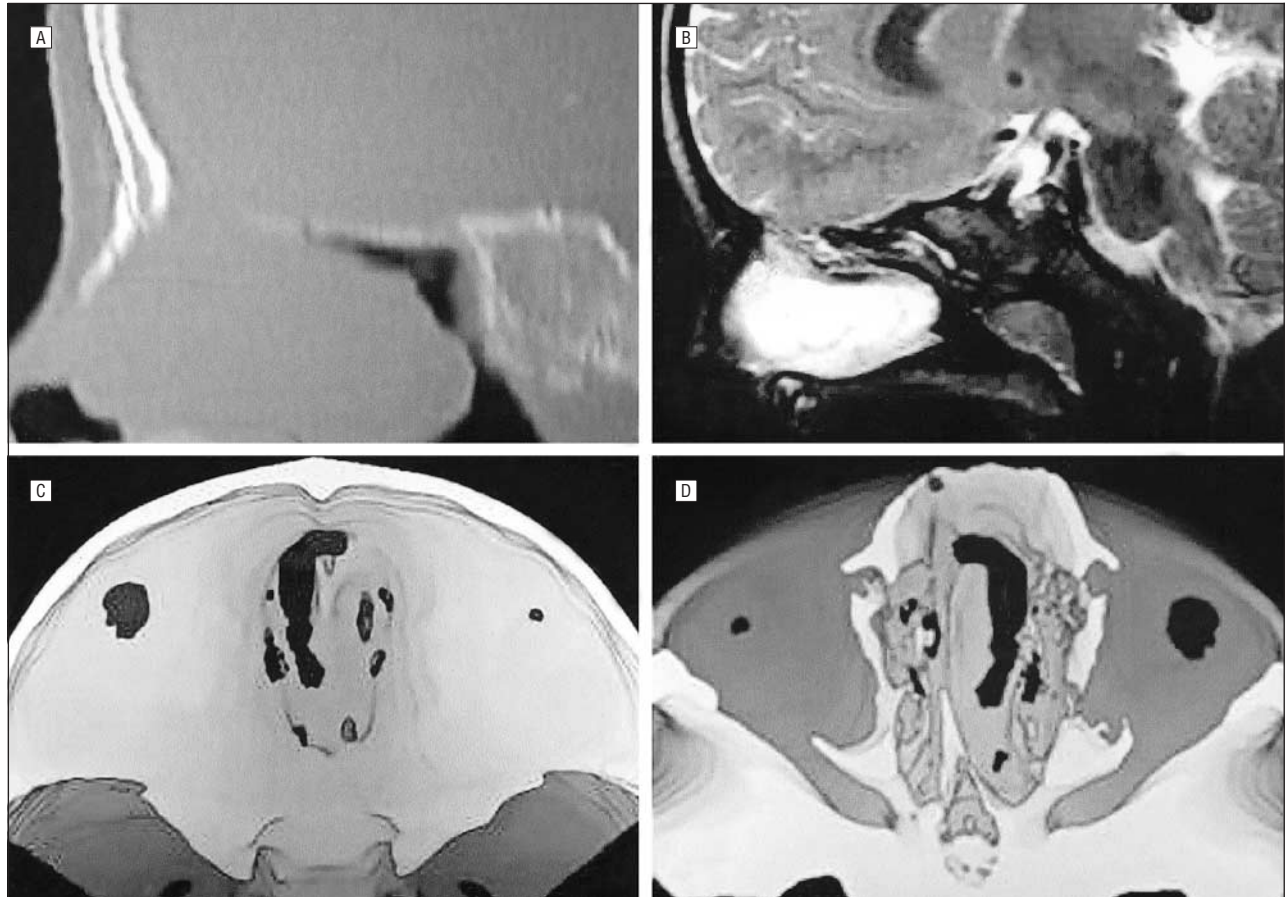


Figure 1. Views of a large encephalocele originating at the foramen cecum. Sagittal computed tomographic (CT) reconstruction (A) and T2-weighted magnetic resonance image (B) demonstrate a large encephalocele. A 3-dimensional CT reconstruction from superior (C) and inferior (D) views demonstrates the large bony defect that originates at the foramen cecum and extends into the left cribriform and ethmoid roof. Reprinted with permission from Elsevier.¹¹

patient, depending on that patient's age, size, and presenting signs and symptoms. The benefits of repairing congenital skull base defects include closing any CSF leaks, decreasing the risk of meningitis, preventing further craniofacial deformity, and alleviating nasal obstruction by removal of any obstructing encephalocele. These benefits must be weighed against the risks and difficulty in performing surgery at an early age.

The most serious risk in patients with skull base defects is meningitis or other intracranial complications. Prevention of these potentially fatal conditions is of paramount importance; however, the risk of meningitis has never been fully assessed. It is unclear to what degree the risk of meningitis increases with defects or encephaloceles that are actively leaking CSF vs those that are not. The risk of ascending meningitis in the acute-care setting with an active traumatic CSF leak may be as high as 10%.¹³ Following successful nonsurgical management of traumatic CSF leaks, a 29% incidence of meningitis has been reported during long-term follow-up.¹⁴ However, to our knowledge, the long-term risk of meningitis in a patient with an encephalocele has not been quantified. Because congenital encephaloceles are rare lesions, an adequate sample size for risk assessment may not be feasible. Some insight can be gleaned, however, from our review. In our series, 2 patients with active leaks presented with meningitis at the ages of 5 and 8 years. A third

patient with a nonleaking encephalocele presented with meningitis at the age of 7 years. Thus, intact mucosal membrane and dura separating the intracranial contents from the nasal cavity may not always be adequate to prevent meningitis, but from our limited series, the risk for meningitis in the absence of a CSF leak seems to be low for the first 5 years of life. While parents of these children must clearly understand the warning signs of meningitis, we believe it is probably safe to delay definitive surgical repair until the child is of sufficient size to make endoscopic repair feasible (typically aged 2-3 years). Actively leaking defects should be repaired earlier if possible, because the risk of meningitis in the presence of dural and mucosal defects is probably higher.

Traditionally, intranasal encephalocele repairs have been delayed longer than their frontoethmoidal counterparts, because neurosurgeons perceive a lower risk of meningitis and craniofacial malformation during development.^{15,16} However, to our knowledge, the risk of facial dysmorphism with intranasal encephaloceles during development has not been adequately characterized. David¹⁷ notes that, in a series of frontoethmoidal encephaloceles, telecanthus was reported in all cases. These encephaloceles involved external exit points that splay the involved bones apart. The intranasal or basal type encephaloceles do not have external bony defects and usually cause mass effect in the intranasal cavity only. Ob-

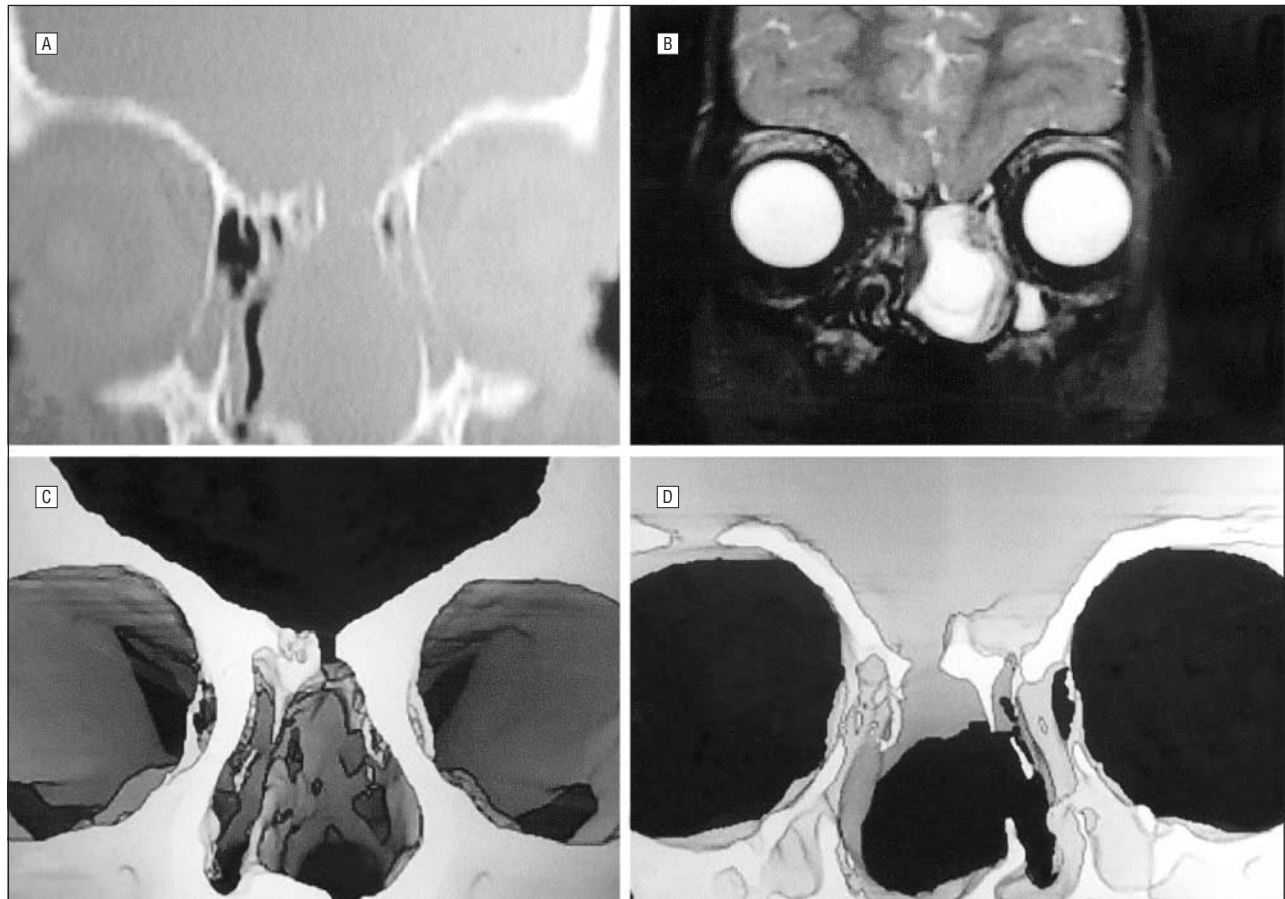


Figure 2. Views of a large encephalocele sac. Coronal computed tomographic (CT) reconstruction (A) and T2-weighted magnetic resonance image (B) of the same patient from Figure 1 again show the large encephalocele sac in the left nasal cavity. A 3-dimensional CT reconstruction from anterior (C) and posterior (D) views demonstrates the skull base defect and deviation of the nasal septum to the contralateral side. Reprinted with permission from Elsevier.¹¹



Figure 3. Coronal computed tomographic scan of a funnel-shaped bony skull base defect limited to the ethmoid roof without foramen cecum involvement. The septum is at the midline, and the skull base is extremely low lying.

viously, larger encephaloceles would have the capacity to splay the nasal bridge if located anteriorly. One of our patients with a large encephalocele involving the foramen cecum, left anterior cribriform plate, and ethmoid roof presented with telecanthus at the age of 2½ years (Figures 1 and 2). In contrast, another patient underwent repair at the age of 23 months, without evidence

of telecanthus. While the second encephalocele also involved the foramen cecum and the encephalocele sac completely filled the nasal cavity, his bony defect was significantly smaller, probably decreasing his risk of developing telecanthus (**Figure 4**). Earlier endoscopic removal of large encephaloceles with significant bony skull base defects may reduce the risk of developing telecanthus, because facial growth has been shown to realign itself after surgery in infancy.¹⁸

SURGICAL CONSIDERATIONS

Once the timing of surgical repair has been decided, there are several technical considerations that are unique to the endoscopic repair of congenital defects. First is the exposure of the skull base defect. Large encephaloceles often result in severe septal deviations and lateralization of the turbinates, resulting in excellent exposure of the skull base, an improved working space, and the ability to perform endoscopic repair in younger children. Defects limited to the ethmoid roof with a midline septum, normally positioned turbinates, and no involvement of the foramen cecum provide limited exposure for repair in young children.

Another surgical consideration is the specific site of these skull base defects. We generally observed 2 types. The first originated at the foramen cecum, with a devi-

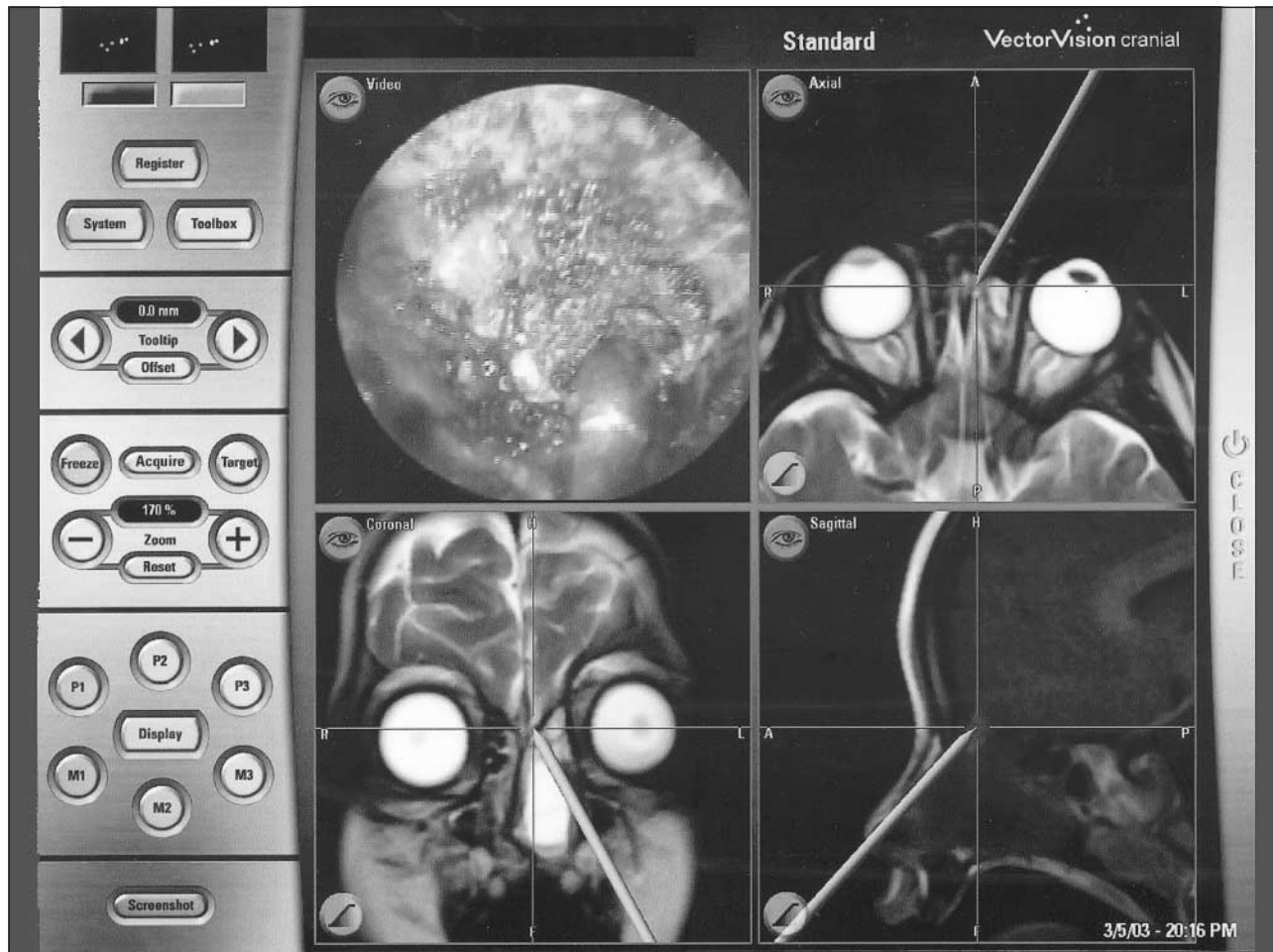


Figure 4. Triplanar magnetic resonance imaging demonstrates a large meningoencephalocele sac that originated through a small bony defect approximately 1.5 mm in diameter at the foramen cecum.

ated crista galli and variable posterior extension into the cribriform or ethmoid roof (Figures 1, 2, and 4). The second was an isolated ethmoid roof defect with a normal foramen cecum. The skull base in these patients was extremely low lying, with a funnel-shaped bony defect (Figure 3). Most of our patients had the first type of defect. This anterior location makes surgical repair more difficult, and may require the use of 70° endoscopes for adequate visualization.

The anatomical location and available exposure of a given defect will subsequently influence the surgical approach and technique. We advocate a precise surgical technique that minimizes trauma to surrounding tissues and sinuses within the cavity, which may otherwise result in “collateral damage.” Small defects that originate at the foramen cecum can be repaired without even performing an uncinectomy or turbinectomy (Figure 4 and **Figure 5**). An extension of this concept can be applied to peripheral submucosal components of these encephaloceles. Encephaloceles that originate at the foramen cecum often dissect submucosally along the anterior-superior septum, the nasal bones, and even the upper lateral cartilages. Early in our experience, we resected these submucosal extensions, similar to resection of a neoplastic process. This extensive resection resulted in nasal stenosis in one patient who subsequently required a Z-

plasty for repair. We approach these patients by precisely identifying the bony skull base defect at the neck of the encephalocele. Mucosa is removed for approximately 5 mm surrounding the bony defect, but submucosal extensions beneath the nasal bones, upper lateral cartilages, and anterior septum are left undisturbed. Preserving mucosa over peripheral components of the encephalocele sac does not seem to compromise the skull base repair. Adequate repair at the skull base eliminates the risk of intracranial complications, and the submucosal extensions seem to atrophy and the mucosa returns to normal with time. Thus, we recommend aggressive ablation and resection of the neck of the encephalocele sac at the level of the bony skull base defect, but total resection of the encephalocele is not always necessary and may impose potential harm to the patient. In addition, it is unlikely that transcranial approaches from above excise these peripheral extensions beneath the nasal bones and upper lateral cartilages. In our 23-month-old patient, peripheral mucosa underneath the nasal bones and upper lateral cartilages was preserved. Amputation and repair of the skull base defect was appropriately performed, and the mucosa returned to a normal endoscopic appearance by 6 months postoperatively.

The final surgical consideration is the availability of appropriate surgical instrumentation. All of our repairs

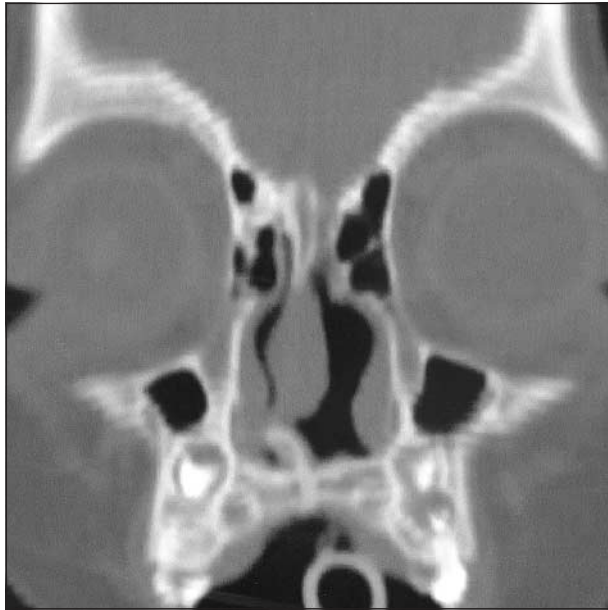


Figure 5. Coronal computed tomographic scan of the patient from Figure 4 at 6 months postoperatively demonstrates removal of the encephalocele and repair of the skull base. "Collateral damage" was avoided, with an intact uncinata and no surrounding iatrogenic sinusitis. At age 23 months, the patient underwent repair using a temporalis fascia overlay graft. Submucosal extensions of the encephalocele sac beneath the nasal bones and upper lateral cartilages were not removed.

were done using standard 4.0- or 2.7-mm endoscopes and standard endoscopic equipment. The youngest patient described in the literature was in our series and was aged 23 months at the time of repair (B.A.W. and R.J.S., unpublished data, 2003). While his repair was done using 2.7-mm endoscopes, others have reported endoscopic repair of an intranasal encephalocele using 0.9-mm needle endoscopes on a 2½-year-old child.¹⁹ While the use of special endoscopes, such as those used for otoendoscopy, may facilitate earlier correction in the future, we have demonstrated the feasibility of repair at the age of 23 months. Development of additional pediatric-sized giraffe instruments and cautery will help make successful endoscopic repair an option at even earlier ages.

At this point, we agree with others that congenital intranasal encephaloceles rarely need to be treated immediately after birth, because they are nearly always covered with healthy skin or at least an epidermal layer.²⁰ Waiting until the child is aged 2 to 3 years for significant facial growth will facilitate an endoscopic technique; however, a child who develops meningitis, a CSF leak, or a cosmetic deformity may need surgical intervention at an earlier age. Two patients presenting at birth because of nasal obstruction were able to be followed up and repaired at a later age without development of meningitis, a CSF leak, or facial deformity.

Continuing progress in the surgical management of congenital skull base defects demonstrates that endoscopic repair is a successful alternative to traditional cra-

niotomy approaches, with less morbidity. Endoscopic techniques can be performed in fairly young children, but require meticulous preparation and precise grafting of the defect to avoid collateral damage to surrounding structures. Timely repair of these skull base defects will hopefully reduce the risk of meningitis, intracranial complications, and facial growth abnormalities and alleviate nasal obstruction. While our report is preliminary, our observations will be enriched with further follow-up and replication of our experience at other institutions.

Submitted for Publication: March 22, 2004; accepted June 16, 2004.

Correspondence: Rodney J. Schlosser, MD, Department of Otolaryngology–Head and Neck Surgery, Medical University of South Carolina, 135 Rutledge Ave, Suite 1130, PO Box 250550, Charleston, SC 29425 (schlosser@musc.edu).

REFERENCES

1. Suwanwela C, Suwanwela N. A morphological classification of sincipital encephalomeningoceles. *J Neurosurg.* 1972;36:201-211.
2. David DJ. Cephaloceles: classification, pathology, and management—a review. *J Craniofac Surg.* 1993;4:192-202.
3. David DJ, Simpson DA. Frontoethmoidal meningoencephaloceles. *Clin Plast Surg.* 1987;14:83-89.
4. Lund VJ. Endoscopic management of cerebrospinal fluid leaks. *Am J Rhinol.* 2002; 16:17-23.
5. Ohnishi T. Bony defects and dehiscences of the roof of the ethmoid cells. *Rhinology.* 1981;19:195-202.
6. McCormack B, Cooper PR, Persky M, Rothstein S. Extracranial repair of cerebrospinal fluid fistulas: technique and results in 37 patients. *Neurosurgery.* 1990; 27:412-417.
7. Mattox DE, Kennedy DW. Endoscopic management of cerebrospinal fluid leaks and cephaloceles. *Laryngoscope.* 1990;100:857-862.
8. Stankiewicz JA. Complications of endoscopic intranasal ethmoidectomy. *Laryngoscope.* 1987;97:1270-1273.
9. Lanza DC, O'Brien DA, Kennedy DW. Endoscopic repair of cerebrospinal fluid fistulae and encephaloceles. *Laryngoscope.* 1996;106:1119-1125.
10. Marshall AH, Jones NS, Robertson IJ. Endoscopic management of basal encephaloceles. *J Laryngol Otol.* 2001;115:545-547.
11. Schlosser RJ, Faust RA, Phillips CD, Gross CW. 3-Dimensional computed tomography of congenital nasal anomalies. *Int J Pediatr Otorhinolaryngol.* 2002; 65:125-131.
12. Schlosser RJ, Bolger WE. Management of multiple spontaneous nasal meningoencephaloceles. *Laryngoscope.* 2002;112:980-985.
13. Brodie HA. Prophylactic antibiotics for posttraumatic cerebrospinal fluid fistulae: a meta-analysis. *Arch Otolaryngol Head Neck Surg.* 1997;123:749-752.
14. Bernal-Sprekelsen M, Bleda-Vazquez C, Carrau RL. Ascending meningitis secondary to traumatic cerebrospinal fluid leaks. *Am J Rhinol.* 2000;14:257-259.
15. Hoving EW, Vermeij-Keers C. Frontoethmoidal encephaloceles, a study of their pathogenesis. *Pediatr Neurosurg.* 1997;27:246-256.
16. Rapport RL II, Dunn RC Jr, Alheady F. Anterior encephalocele. *J Neurosurg.* 1981; 54:213-219.
17. David DJ. New perspectives in the management of severe cranio-facial deformity. *Ann R Coll Surg Engl.* 1984;66:270-279.
18. Rahman NU. Nasal encephalocele: treatment by trans-cranial operation. *J Neurol Sci.* 1979;42:73-85.
19. Zhang XM, Gan JY, Guo JB, Li TY, Wen WP, Jiang FL. Pediatric intranasal and skull base needle endoscopic surgery and examination [in Chinese]. *Lin Chuang Er Bi Yan Hou Ke Za Zhi.* 2000;14:349-351.
20. Hoving EW. Nasal encephaloceles. *Childs Nerv Syst.* 2000;16:702-706.