Endoscopic Endonasal Repair of Congenital Defects of the Anterior Skull Base: Developmental Considerations and Surgical Outcomes

William C. Gump¹

¹ Division of Pediatric Neurosurgery, Norton Neuroscience Institute and Kosair Children's Hospital, Louisville, Kentucky, United States

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Address for correspondence William C. Gump, MD, Division of Pediatric Neurosurgery, Norton Neuroscience Institute and Kosair Children's Hospital, 210 East Gray Street, Suite 1102, Louisville, KY 40202, United States (e-mail: william.gump@nortonhealthcare.org).

Abstract

Techniques of endoscopic endonasal surgery, initially developed primarily for intracranial neoplasms, have been adapted to treat a wide variety of pathologies previously addressed with open craniotomy including congenital and acquired defects of the anterior skull base. Congenital defects can lead to herniation of leptomeninges containing cerebrospinal fluid alone or with brain tissue. Specific types of encephalocele can be defined on the basis of the associated abnormal bony anatomy. Endoscopic endonasal surgery represents a relatively recent development in the treatment of these entities. Technical considerations include relatively younger age range of the patient population, dimensions of preexisting bony defect, volume of herniated meninges and brain tissue, and distorted anatomy from abnormal development of the affected craniofacial skeleton. Recent highly detailed anatomical studies have quantitatively verified the utility of endoscopic endonasal surgery in the pediatric population. Particular attention has been directed toward adequacy of nasoseptal flap reconstruction in pediatric patients. Several reports have described patients with encephalocele of the anterior cranial fossa successfully treated with endoscopic surgery. The literature on endoscopic repair of congenital encephalocele is reviewed. Outcomes have generally been reported as favorable, although long-term follow-up and systematic studies have not been pursued.

Keywords

- ▶ encephalocele
- ▶ endoscopic surgery
- ▶ endonasal surgery

Introduction

Anterior Skull Base Development

Developmental considerations are important in young patients for whom significant anatomical maturation has yet to occur. The complexity of anterior skull base anatomy is reflected in the highly detailed but incomplete understanding of its prenatal and postnatal development that has emerged over decades of research. A comprehensive review of this topic is beyond the scope of this article but is available elsewhere.¹

Briefly, the first evidence of skull formation emerges at ~ 4 weeks postconception (pc) when mesenchyme derived from the paraxial mesoderm and neural crest condenses to form the base of the ectomeningeal capsule. The timing is remarkable in that this process takes place well after the primordial brain, cranial nerves, eyes, and major intracerebral blood vessels have begun differentiation. Conversion of the ectomeninx mesenchyme into cartilage constitutes the beginning of the chondrocranium, starting on day 40 pc. The anterior (rostral) prechordal region of the cranial base

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is of neural crest origin, and the posterior (caudal) chordal region is of mesodermal origin. The interface between these regions becomes the junction of the basisphenoid and basisoccipital bones.

There are ~ 110 ossification centers in the embryonic human skull that fuse to produce 45 bones in the neonatal skull. Ossification centers begin as early as 8 weeks pc, including the medial and lateral pterygoid plates, and continue to appear up to birth. Unossified chondrocranial remnants persistent at birth include the alae and septum of the nose, the sphenoccipital and sphenopetrous junctions, the apex of the petrous bone, between the separate parts of the occipital bone, and in the foramen lacerum. By young adulthood, 22 skull bones are recognized.

An enlarged foramen cecum is often implicated as a site of herniation in anterior encephalocele formation. This structure is formed by the articulation of the ethmoid bone with the frontal crest of the frontal bone and can vary in size. It is frequently closed or very small, transmitting an emissary vein from the nose to the superior sagittal sinus.²⁻⁴ Absence of complete ossification on computed tomography (CT) scan after age 4 years can be considered abnormal; differential diagnosis of an open foramen cecum would include cephalocele, nasal glioma, metabolic disorder, or dysplasia.

Classification of Anterior Cranial Fossa Encephaloceles

Meningocele refers to herniation of leptomeninges and cerebrospinal fluid (CSF) alone; encephalocele includes herniation of brain tissue as well. Either can be associated with CSF leak. Treatment approaches to these entities are functionally identical, and a distinction is often not drawn in the clinical literature.

The incidence of specific types of encephalocele varies by geographic location and race. Anterior encephaloceles occur with the greatest frequency in Southeast Asia, parts of Russia, and central Africa, where they are seen in up to 1 in 3,500 live births and are the most common type of encephalocele observed. In contrast, anterior encephalocele is seen in only 1 in 35,000 live births in North America, where occipital encephalocele comprises ~ 85% of all encephaloceles seen.⁵

The most widely used classification scheme divides anterior fossa encephaloceles into sincipital and basal categories.⁶ These categories, along with their subtypes, are defined based on the specific location of the associated bony defect. Sincipital encephaloceles, which are associated with a skull defect at the foramen cecum, are anterior to the cribriform plate. These can be nasofrontal, nasoethmoidal, nasoorbital, or interfrontal. Basal encephaloceles, protruding through the cribriform plate or planum sphenoidale, are classified as sphenopharyngeal, sphenoorbital, sphenomaxillary, or sphenoeethmoidal.

Pathogenesis of Anterior Encephalocele

Secondary encephalocele refers to extracranial herniation of leptomeninges, brain, and CSF due to an acquired skull defect, most commonly traumatic or iatrogenic. In primary encephalocele, the defect or its congenital substrate is present at birth, presumably from disordered development.⁷ The etiology of primary encephalocele formation remains incomplete-

ly understood, although there are currently two predominant theories. One is that structural weakness in the ossifying bone, particularly around the foramen cecum in the region of the junction between the membranous frontal bone and endochondral ethmoid bone formation, could permit herniation of neural elements.^{8,9} The other theory presumes a delay in closure of the rostral neural tube.¹⁰ However, the fact that most encephaloceles are skin covered argues that the pathology does not result from a simple failure of neurulation. Anterior encephaloceles are much more likely to contain only gliotic nonfunctional neural tissue compared with occipital encephaloceles.

Anterior encephalocele manifests across a broad clinical spectrum, from barely perceptible midface, intranasal, or intrapharyngeal masses to severe craniofacial deformity. Nasal masses typically show a positive Furstenberg sign (increased swelling and pulsatility associated with Valsalva or jugular vein compression) that distinguishes them from other entities such as nasal glioma.⁵

Treatment

Treatment can be performed electively if the encephalocele is not associated with a CSF leak. However, a significant treatment consideration is the potential effect of the encephalocele mass on the development of the craniofacial skeleton that may prompt earlier intervention. Direct surgical repair is the mainstay of treatment. Bifrontal craniotomy exposes the patient to the risks of blood loss, brain retraction, disruption of growth centers, and injury to the supraorbital/supratrochlear neurovascular complexes associated with standard surgical exposure of the anterior skull base from above. Endoscopic endonasal surgery (EES) avoids many of these potential morbidities.

Numerous reports have described good results with EES in pediatric patients for a variety of skull base pathologies, predominantly benign and malignant neoplasm.¹¹⁻¹³ Other pathologies less frequently treated with this modality have included cavernous malformation,¹⁴ arteriovenous malformation, hypertrophic pituitary gland, and Rathke cleft cyst. EES has been successfully used for odontoidectomy in children, thereby avoiding the morbid transoral corridor.^{15,16} Studies specific to pediatric craniopharyngioma have shown good outcomes^{17,18} and may suggest EES is the preferred approach for this pathology, despite its anatomically heterogeneous presentations.¹⁹

Technical Considerations in Pediatric Endoscopic Endonasal Surgery

Microscopic transnasal surgery in children has been limited by the dimensions of the nares, and a sublabial exposure has historically been favored for pediatric transsphenoidal approaches. This degree of limitation is minimal for endoscopic techniques.^{20,21} Multiple recent quantitative anatomical studies of surgical corridors in children have demonstrated quantitatively that EES techniques are minimally restricted in pediatric patients when compared with adults.^{11,22-24} The nasal aperture in children is almost always sufficient to

accommodate standard endoscopes and instrumentation.⁹ Intraoperative image guidance has been shown to be a safe and effective adjunct to EES in the pediatric population.^{25,26}

Reconstruction of skull base defects to prevent CSF leak is important to isolate the sinonasal and cranial cavities. Larger defects and high-flow CSF leaks associated with expanded EES approaches require a robust, vascularized multilayer closure that is now most commonly achieved using a nasoseptal flap.²⁷ In this technique, a vascularized pedicled flap of the nasoseptal mucoperiosteum and mucoperichondrium is based on the nasoseptal and terminal internal maxillary arteries.²⁸ The flap is harvested before septectomy and placed temporarily in the nasopharynx prior to final positioning.

The nasoseptal flap has some limitations in pediatric patients. In a series of six patients undergoing this procedure for skull base reconstruction, two of three patients age < 14 years did not have sufficient septal flap tissue to completely cover the defect; one of these patients subsequently developed a CSF leak requiring revision surgery with fat graft and lumbar drainage.²⁹ This particular study also analyzed adult and pediatric CT scans to calculate sizes of obtainable nasoseptal flaps relative to estimated dimensions of transcribiform, transsellar, and transclival defects from expanded EES. The authors concluded that the age limitations for transcribiform and transsellar defect reconstruction were > 9 years and > 7 years, respectively. Nasoseptal flap coverage was predicted to be insufficient for a transclival defect in any pediatric patient. Alternative reconstruction techniques include temporoparietal fascial flap or pericranial flap coverage, although endoscopic variants of these techniques have not been specifically studied in children.^{30–33}

Reported Cases

Several case reports and limited case series have included patients with congenital anterior fossa encephalocele or various posttraumatic or postsurgical pathologies repaired via EES.

Transnasal repair of iatrogenic CSF rhinorrhea from intracranial or endoscopic sinus surgery in adults was reported as early as 1990.^{34,35} A report of a posttraumatic pseudomeningocele traversing the cribriform plate in a 7-year-old boy described endoscopic repair of this defect using auricular cartilage in the bony defect, reinforced with temporalis fascia and muscle packing. The patient was asymptomatic without complications 15 months after surgery.³⁶ Intrathecal fluorescein was used in these cases to confirm the site of CSF leak or pseudomeningocele.

A subsequent case series described four patients, from 12 months to 13 years of age, with CSF rhinorrhea that was associated with a nasal glioma in three patients and a congenital anterior meningocele in the other.³⁷ All patients underwent transnasal endoscopic resection of the intranasal mass, with concurrent repair of the associated skull base defect using a combination of free mucosal flaps, pedicled flaps, conchal cartilage, fibrin glue, and nasal packing. One patient who was 12 months old at the time of surgery required a second endoscopic procedure 2 weeks later for

recurrent CSF leak. A slight deformation of the nasal bones described in the patient on preoperative examination was reported as gradually resolved with facial growth over the subsequent 12 months. There was no further recurrence of CSF leak in any of the patients over follow-up of 1 to 3 years.

A review of EES in 13 pediatric patients from 2000 included four cases of spontaneous CSF leak, three of which were posttraumatic/iatrogenic.³⁸ The lone congenital meningocele was treated successfully without complication or recurrence over 1 year postoperatively, as were two of the three other patients. One iatrogenic skull base defect was described as being too large for endoscopic repair. A subsequent review by the same surgical group in 2006 included 12 patients, ages 1 to 14 years, undergoing EES for spontaneous anterior meningoencephalocele (seven cases) or posttraumatic CSF rhinorrhea (five cases).³⁹ Intrathecal fluorescein was used intraoperatively to identify the site of CSF leak and verify a satisfactory seal at the conclusion of surgery. Closure techniques included a combination of dural substitute, abdominal fat, middle concha bone, mucoperichondrium, and middle turbinate to seal the CSF fistula and pack the space. There were no complications and no reoperations over 12 to 72 months of postoperative follow-up. All patients underwent endoscopic endonasal evaluation 1 month postoperatively with findings of normal nasal mucosa.

Another similar case series that included 11 pediatric patients, mean age 6 years at the time of surgery, treated with EES over 8 years reported no surgical complications and no postoperative adverse events over a mean follow-up interval of 46 months.⁷ These operations also used a variety of closure and packing techniques as previously described to prevent CSF leak. Comparable results were described in another report of two cases in patients 15 months and 6 years of age⁴⁰ and in a report of a patient age 2 years.¹⁰

A relatively large case series from 2010 included 28 pediatric patients with 18 congenital and 10 posttraumatic anterior skull base defects who were considered for possible endoscopic repair.⁴¹ Endoscopic techniques were similar to those described in the reports just cited. Three patients in this group required combined open and endoscopic surgery due to the size and complexity of their skull base defects. This series is remarkable for reporting one mortality in a patient requiring combined repair, occurring 2 years postoperatively due to meningitis. There were also four iatrogenic frontal or ethmoidal mucocoeles observed postoperatively; otherwise, there were no recurrences or complications at 9- to 57-month follow-up.

The most thorough discussion of complications associated with EES has been in a report from 2013 that encompasses 171 endoscopic endonasal procedures performed in 133 pediatric patients for a variety of anterior skull base pathologies.¹¹ Of these patients, 112 overall had a diagnosis of skull base tumor and 21 had a diagnosis of bony abnormality. Reported complication rates were relatively low in the bony abnormality group, with no reported cases of postoperative diabetes insipidus (17.9% in the tumor group; 10.7% permanent), syndrome of inappropriate antidiuretic hormone (2.7% in the tumor group), new anterior pituitary endocrinopathy

(12.5% in the tumor group), meningitis (4.5% in the tumor group), or cranial nerve palsy (7.1% transient, 2.7% permanent in the tumor group). Risk factors for increased complications in this study were diagnoses of craniopharyngioma or angiofibroma.

Few patients < 1 year of age undergoing EES have been reported, although technical barriers were described as minimal. The youngest reported patient to undergo EES for meningocele repair was 2 months old at the time of surgery.⁴² Control magnetic resonance imaging done 8 months postoperatively was described as normal, and no complications were reported over 2 years. A previously reported patient was 5 months old at surgery⁴³ with no complications at 3-month follow-up.

Older children and adults with nontraumatic, presumed congenital, anterior skull base defects repaired endoscopically have also been reported. Case reports including patients ages 17, 47, and 26 years have described uncomplicated postoperative courses.^{44,45}

Conclusions

EES is a well-established treatment modality for a variety of anterior skull base pathologies including tumors and traumatic injuries. The utility of EES includes treatment of congenital anterior fossa encephalocele, with overall favorable results that are difficult to compare directly with transcranial repair. The reported complication rate is low. Young age is not a contraindication to EES, although most case reports describe patients who were older than infancy at the time of operation. Described outcomes have been very good. Dedicated study may be required to quantify any potential advantage of EES over transcranial repair of congenital skull base defects.

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