Encephaloceles

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Introduction

Encephaloceles are the result of a congenital cranial defect that allows intracranial contents to herniate. The herniated contents may consist of the meninges (meningocele), meninges, and brain (meningoencephalocele) (see Fig. 16.1), or in severe cases meninges, brain, and part of the ventricular system (hydroencephalomeningocele). Encephaloceles are classified by their anatomic location (see Table 16.1) [1]. The vast majority (75 %) are located in the posterior cranial fossa. However, it is the anterior cranial fossa (sincipital) encephalocele that is the concern of the craniofacial team, as it is considerably more deforming.

Biology and Epidemiology

- No known genetic mutation.
- Higher incidence in Southeast Asia (1:6,000 live births) than North America (1:35,000 live births) [2].

Theories of pathogenesis

- Primary arrest of bone development allowing brain to herniate [3].
- Adhesions from brain, dura, and skin arresting bony development [4].
- Increased intracranial pressure pushes the brain through the developing cranial base, causing arrest of bone development [5].

Embryology

- Neural tube begins to close between 3rd and 4th week of fetal development.
- Neural crest cells migrate into the frontonasal and maxillary processes, differentiating into the facial bones, cartilage, and muscles.
- Abnormal development of the potential spaces between these developing structures (fonticulus frontalis, prenasal space, foramen cecum) is responsible for congenital midline masses.

Presentation

- Soft, compressible, pulsatile midline mass that transilluminates
- Occasionally may present with ulcerations and leaking CSF, which requires emergent closure

Differential Diagnosis

The three most common diagnoses of a midline mass in an infant are dermoid cyst, glioma, and encephalocele [6, 7]. History and physical exam can generally lead to the correct diagnosis; however, this is usually confirmed with imaging.

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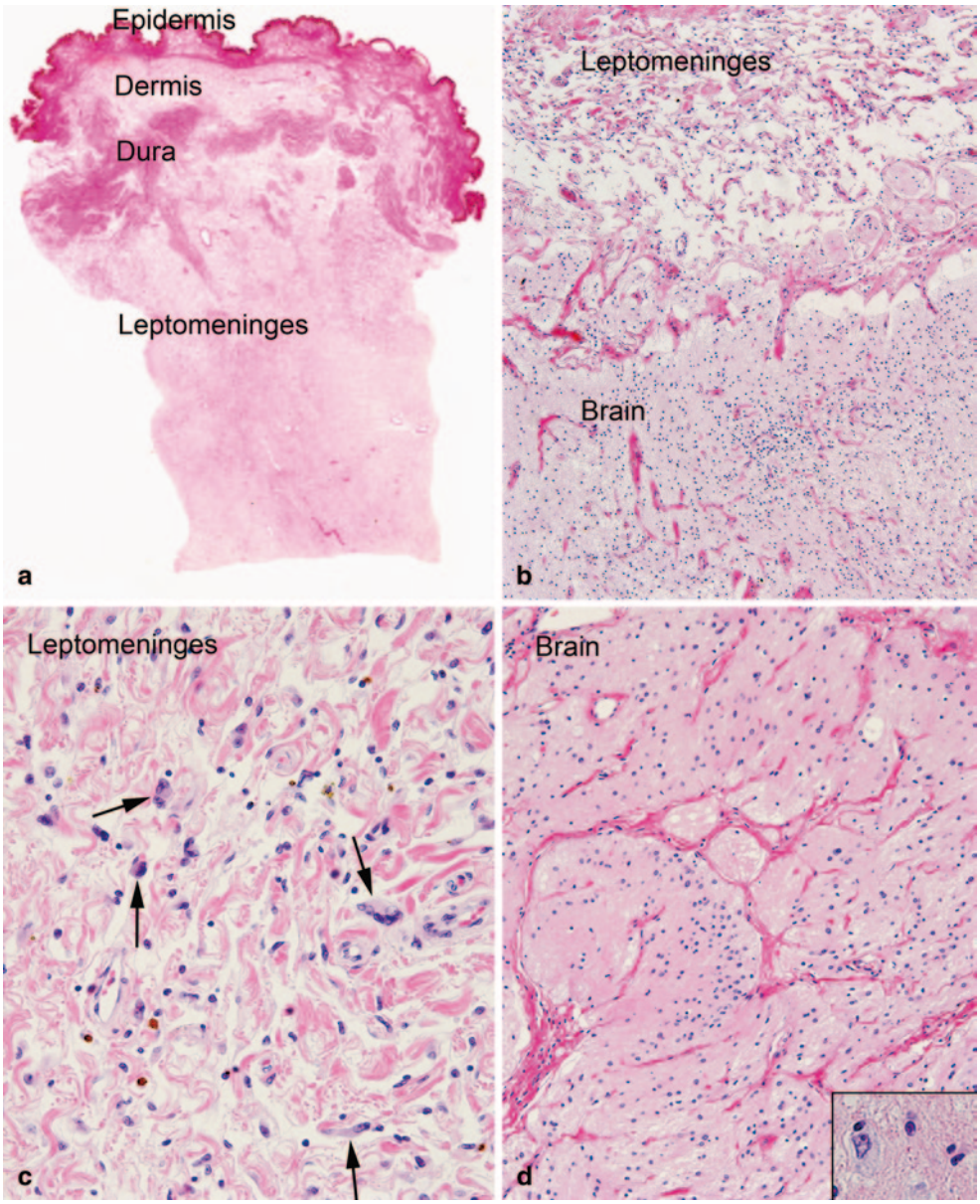


Fig. 16.1 Encephalocele. **a** Cut section of encephalocele wall showing skin surface with *dermis* and *epidermis* a dense fibrous *dura*-like band and deep-seated *leptomeninges*. **b** *Brain* tissue covered by a layer of *leptomeninges*. **c** Leptomeningeal tissue with reactive glial cells (*arrows*) and short curvilinear bands of collagen. **d** Malformed *brain* tissue. Neurons are seen in the *inset*

- Nasal dermoid cyst
  - Most common midline mass
  - Present at birth, diagnosed in early childhood
  - Composed of ectodermal and mesodermal elements
  - Hallmark is punctum with a single hair located on the nasal dorsum
  - Often become infected and can drain sebaceous material
  - Intracranial extension cannot be ruled out on exam—imaging required for accurate diagnosis
- Nasal glioma
  - Presents as firm rubbery mass with bluish or reddish appearance
  - Composed of glial cells in a connective tissue matrix
  - Often extend intranasally
  - Does not communicate with cerebral contents, so not pulsatile and does not transilluminate
- Less common entities that occur in the midline
  - Vascular malformation
  - Teratoma
  - Sebaceous cyst
  - Neurofibroma
  - Ganglioneuroma
  - Nasal fibroma
  - Adenoma
  - Chondroma
  - Carcinoma

Diagnosis and Evaluation

Patients who present with an encephalocele are most appropriately managed by a multidisciplinary craniofacial team comprised of a craniofacial surgeon, a neurosurgeon,

Table 16.1 Classification of encephaloceles [1]

Frontoethmoidal encephaloceles
Nasofrontal
Nasoethmoidal
Nasoorbital
Cranial vault encephaloceles
Interfrontal
Anterior fontanel
Interparietal
Posterior fontanel
Temporal
Cranial base encephaloceles
Transethmoidal
Sphenoethmoidal
Transsphenoidal
Frontosphenoidal
Occipital encephaloceles
Cranioschisis
Associated with cranial/upper face clefts
Associated with basal/lower face clefts
Occipitocervical clefts
Acrania and/or anencephaly

an otolaryngologist, a geneticist, and a pediatrician. Neurological and developmental assessments and evaluation by an ophthalmologist are also essential.

Physical Examination

- Soft nasal mass in midline is the most common presentation
- Bluish appearance
- Soft, compressible, and pulsatile mass that transilluminates
- Mass increases with size with crying, Valsalva, or compression of internal jugular veins (Furstenberg test)
- "Long nose hypertelorism"—patients present with long, flat, wide noses that is more pronounced after encephalocele excision [8, 9]
  –True orbital hypertelorism is rare, but telecanthus and interorbital hypertelorism are universal
- Deformational trigonocephaly

Laboratory Data

No specific laboratory test confirms the diagnosis of encephalocele. A preoperative hemoglobin is prudent.

Imaging Evaluation

- Computed tomography (CT) scan is the imaging modality of choice.
  – Analyzing both bone and brain windows in the axial, coronal, and sagittal planes, as well as three-dimensional reconstructions, are necessary for understanding the complex bony and intracranial anatomy involved.
  – Useful for assessing the potential presence of hydrocephalus.
  – Sagittal reconstructions helpful for evaluating the presence of Chiari I malformation, relevant to patients at risk for hydrocephalus (can also be seen on magnetic resonance imaging, MRI).
  – Essential for planning a successful operative intervention.
- MRI.
  – Provides the most detailed soft tissue images and is useful in distinguishing between soft tissue masses.
- Ultrasound.
  – May be useful in evaluating for hydrocephalus, but often redundant if CT or MRI performed in initial evaluation.

Pathology

Diagnosis can be made with a combination of history, physical exam, and imaging. Biopsies prior to definitive repair are unnecessary and should be discouraged. Histopathologically, meningoceles consist of leptomeningeal membranes with or without glial tissue and meningoencephaloceles of malformed brain tissue and the leptomeningeal membranes covering it (Fig. 16.1). Ependymal tissue may be be seen in hydroencephalomeningoceles.

Treatment

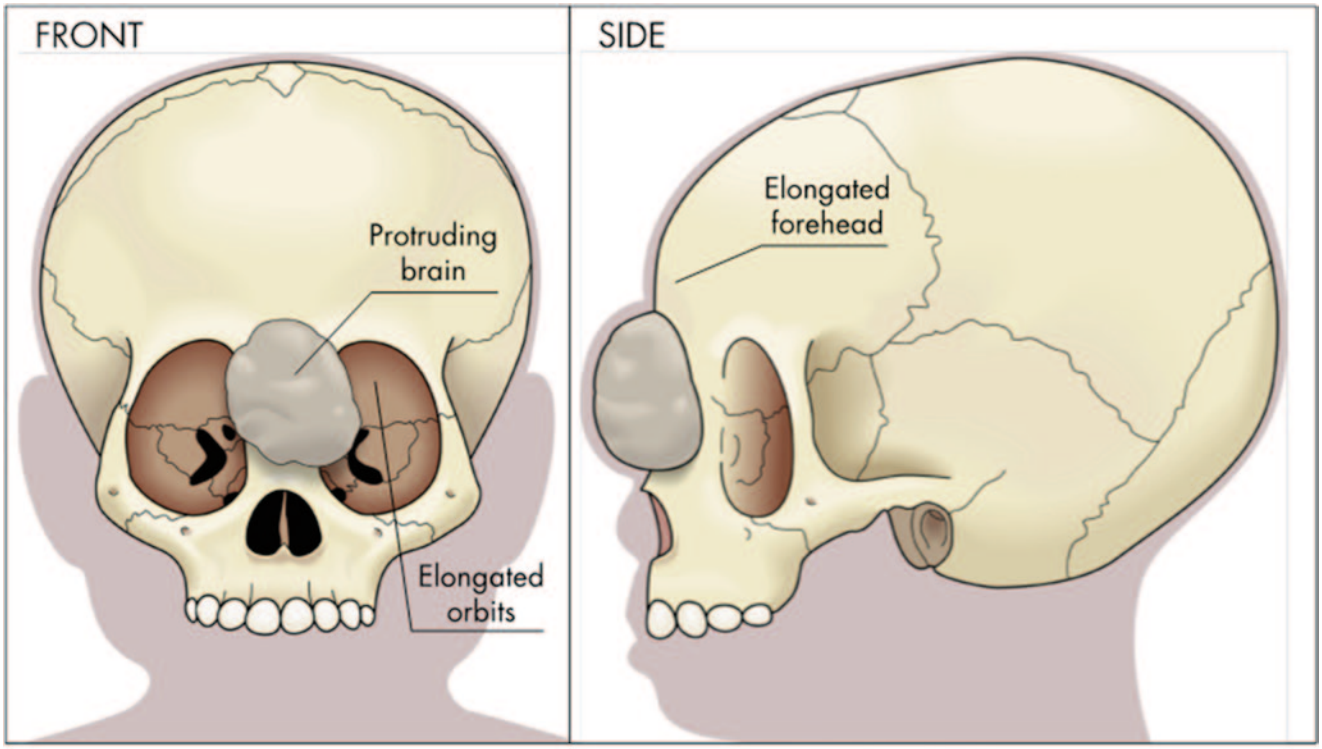
Medical

No medical intervention exists to treat this anatomical abnormality.

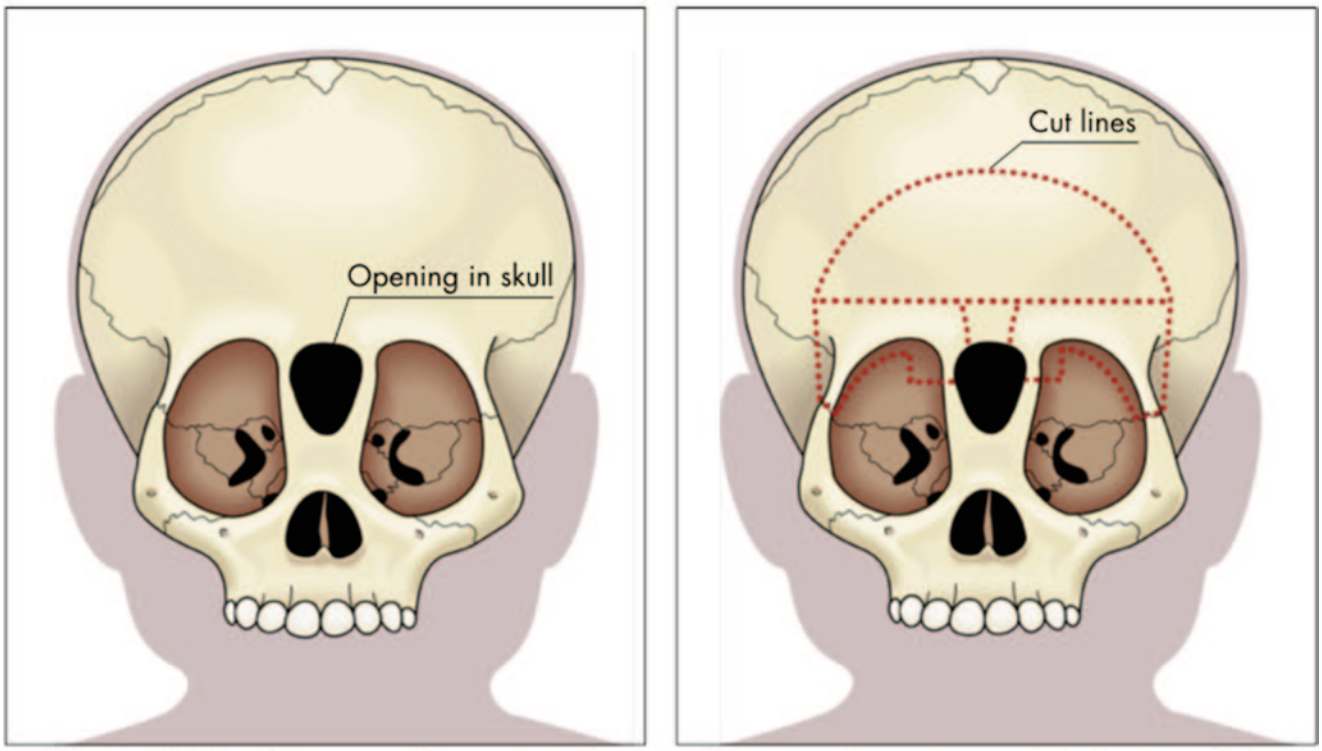
Surgical

Operative intervention provides definitive correction of this problem (see Fig. 16.2). Successful correction follows the following principles [10]:

- Accurate diagnosis, delineation of anatomy, and surgical planning



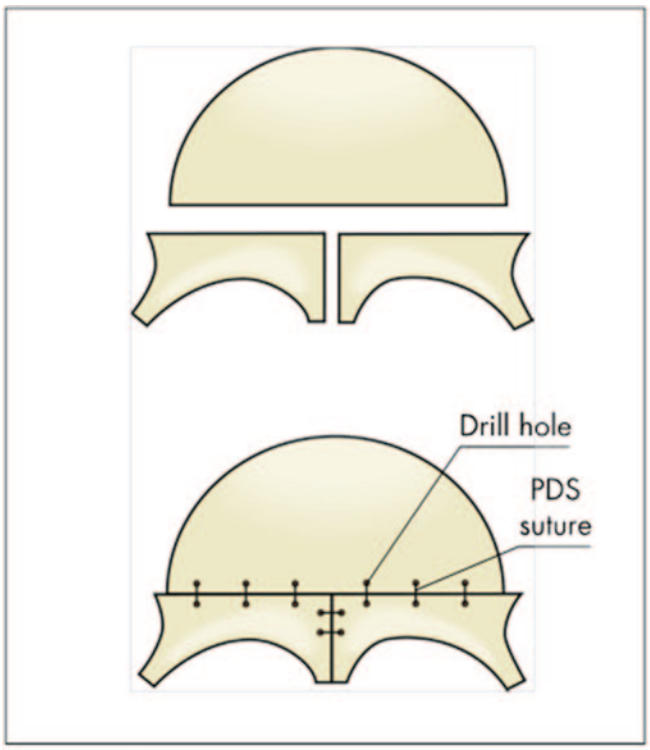
A frontoethmoidal encephalocele is a protrusion of the brain through an opening in a skull due to a birth defect. This also causes an elongation of the orbits and the forehead.



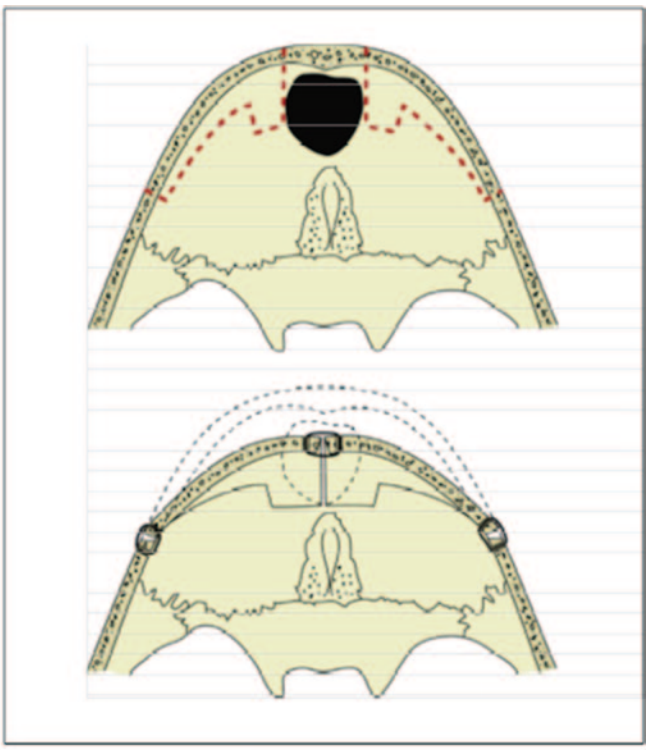
The encelphalocele is removed exposing the opening in the skull.

Lines are drawn along which the skull will be cut.

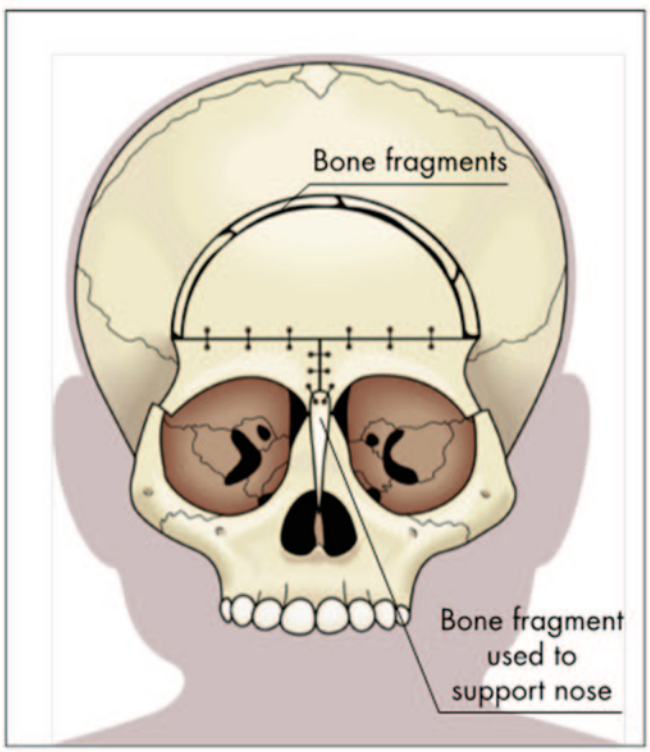
Fig. 16.2 Reconstruction of frontoethmoidal encephaloceles



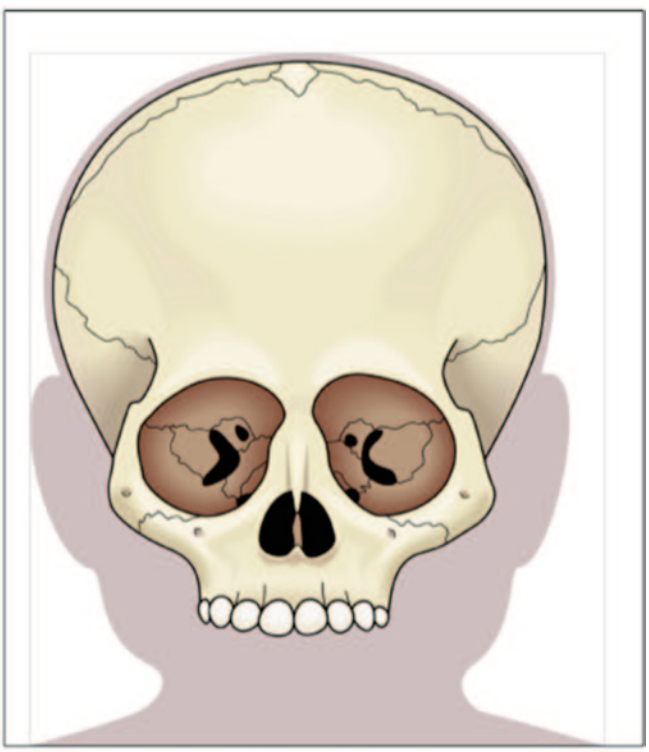
Pieces of the skull are trimmed and shaped and then held together with wire.



The reconstucted forehead and nose are placed back on the skull.



Left over bone is used to fill gaps. Another bone fragment is used to support the nose.



As the patient heals, the bone fragments fuse together again.

Fig. 16.2 (continued)

- Single-stage operation with both craniofacial surgeon and neurosurgeon present
- Osteotomies and bone movements that correct all deformities, including the trigonocephalic head shape and interorbital hypertelorism
- Nasal reconstruction that avoids the long-nose hypertelorism deformity
- Skin closure that removes abnormal skin and places incisions in advantageous locations

Proper anatomical relationships need to be restored. A frontonasoethmoidal encephalocele displaces the frontal bone cephalad and the nasal bones caudad; the medial orbital walls are anterolaterally displaced. Specifics of the technique used by the authors are described in the following sections [11].

Positioning

- Supine, using a Mayfield headrest to secure the cranium.
- Pins are placed posterior enough to avoid interference with the coronal flap and parietal bone graft harvest.
- Circum-mandibular 28-gauge dental wire is used to secure the endotracheal tube.
- Tarsorrhapy sutures placed for corneal protection.
- If hydrocephalus present, or if dural dissection difficult, consider placement of external ventricular drain (in mid-pupillary line, just anterior to coronal suture, preferably on right (nondominant) side, inserted perpendicular to skull to a depth of 3–5 cm then affixed in place during case).

Coronal Exposure

- Wavy-line coronal incision.
- Combination of local anesthesia with epinephrine and use of electrocautery obviate the need for hemostatic clips at the scalp edge.
- Subperiosteal dissection performed anteriorly, elevating the temporalis muscles with the flap.
- Pericranial flap based on the supraorbital vessels is preserved for potential use in closing the dural defect.
- The encephalocele sac is bluntly dissected from the pericranium of the frontal bone and the periosteum of the nasal and orbital bones.
- Periorbita elevated circumferentially taking care to preserve the lacrimal system.
- Medial canthi detached for repositioning later.

Nasal Exposure

- Circumferential skin incision made around the encephalocele, leaving enough skin to close the final defect

Frontal Craniotomy

- Marchac template used to determine the position of the craniotomy
- 1–2 cm frontoorbital bandeau preserved
- Bandeau removed before encephalocele resection
- Dura opened on superior and lateral border of sac, prepared for possible superior sagittal sinus ligation, with amputation of herniated cranial tissue (taking care to preserve arterial anatomy) and meticulous hemostasis
- Sac transected
- Dura closed primarily, reinforced with DuraGen, cadaveric dermis and pericranial flap as necessary, along with possible dural glue, such as Tisseal

Cranial Remodeling

- Trigonocephaly and interorbital hypertelorism must be corrected.
- Central segment of bandeau resected.
- Two hemibandeau segments rotated and advanced medially to close the bony defect and secured to the frontal bone.
- Calvarial defects closed with a combination of bone graft and bone mush harvested from inner table of the frontal bone.

Nasal Reconstruction

- Correction of the long-nose deformity is essential
- Dorsal onlay graft of calvarial bone or costal cartilage attached as a cantilever to bandeau/neofrontal construct with lag screws

Medial Canthal Repositioning

- 30-gauge wire used to transnasally reposition the medial canthal ligaments
- Slight overcorrection necessary as canthal height will drift caudally and laterally as swelling subsides

Skin Closure

- External ventriculostomy drain placed before skin closure.
- Coronal incision closed in two layers over a closed suction drain.
- H-shaped excision of excess skin over the encephalocele addresses both vertical and horizontal tissue excess and allows for repositioning of the medial brow.
- Gentle compression dressing placed prior to extubation.

Endoscopic Management of Encephaloceles

Basal encephaloceles are located in the anterior skull base and herniate through the cribiform plate or posterior to it. Therefore, the masses are confined to the nasal cavity and are potentially amenable to endoscopic repair. Historically these lesions have been removed and the skull base defects repaired using external approaches including lateral rhinotomy and craniotomy. However, advancements in endoscopic equipment and technique has made endoscopic repair of spontaneous cerebrospinal fluid leaks and encephaloceles involving the anterior skull base standard of care in the adult population [12, 13]. Currently there is less experience with repair of these lesions in the pediatric patient population, but

there is evidence to support this treatment option in young patients.

- Successful endoscopic repair demonstrated in children as young as 1.5 months [14].
- Delay in repair can increase risk of complications such as meningitis.
- Multiple small case series have shown that endoscopic techniques are effective for repair of encephaloceles in pediatric patients [14–21].

Technique

- Localization of encephalocele by preoperative CT and MRI imaging.
- Image guidance can assist in intraoperative localization.
- 0, 30, and 70° endoscopes in 2.7 mm and 4 mm sizes depending on age of the child.
- Pediatric sinus instruments.
- Lumbar drain dependent on presence of hydrocephalus or other indicator of increased intracranial pressure [14].
- Intrathecal fluorescein can be used to identify the breach in the skull base in the setting of CSF leak; however, use in pediatric patients is limited.
- Bipolar cauterization or Coblation™ technology for reduction of the encephalocele for visualization of the skull base defect requiring repair [22].
- Repair is carried out using underlay or overlay techniques.
- Materials used include temporalis fascia, fascia lata, autologous fat, dural substitutes, bone, and cartilage.
- Pedicled septal flaps and turbinate flaps provide vascularized tissue.
- Absorbable packing such as gelfoam used to hold repair in place, followed by the placement of nonabsorbable packs which are left in place for 1–2 weeks.
- Patients left on antibiotics as long as packing in place.
- Patients should be followed up long-term with serial nasal endoscopy and repeat imaging if clinical evaluation indicates.

Postoperative Monitoring

- Intensive care unit (ICU) monitoring with frequent neurologic assessments performed for 24 h
- Closed suction drain monitored for evidence of cerebrospinal fluid (CSF) leak and removed between postoperative days 3 to 5
- External ventriculostomy usually removed by postoperative day 5 following successful clamping trial, but options of endoscopic (endoscopic third ventriculocisternostomy/choroid plexus cauterization) or CSF diversion (ventriculoperitoneal shunt) surgical treatments exist should hydrocephalus develop
- Periorbital edema universally present for first 72 h
- 3-day steroid taper used to address edema

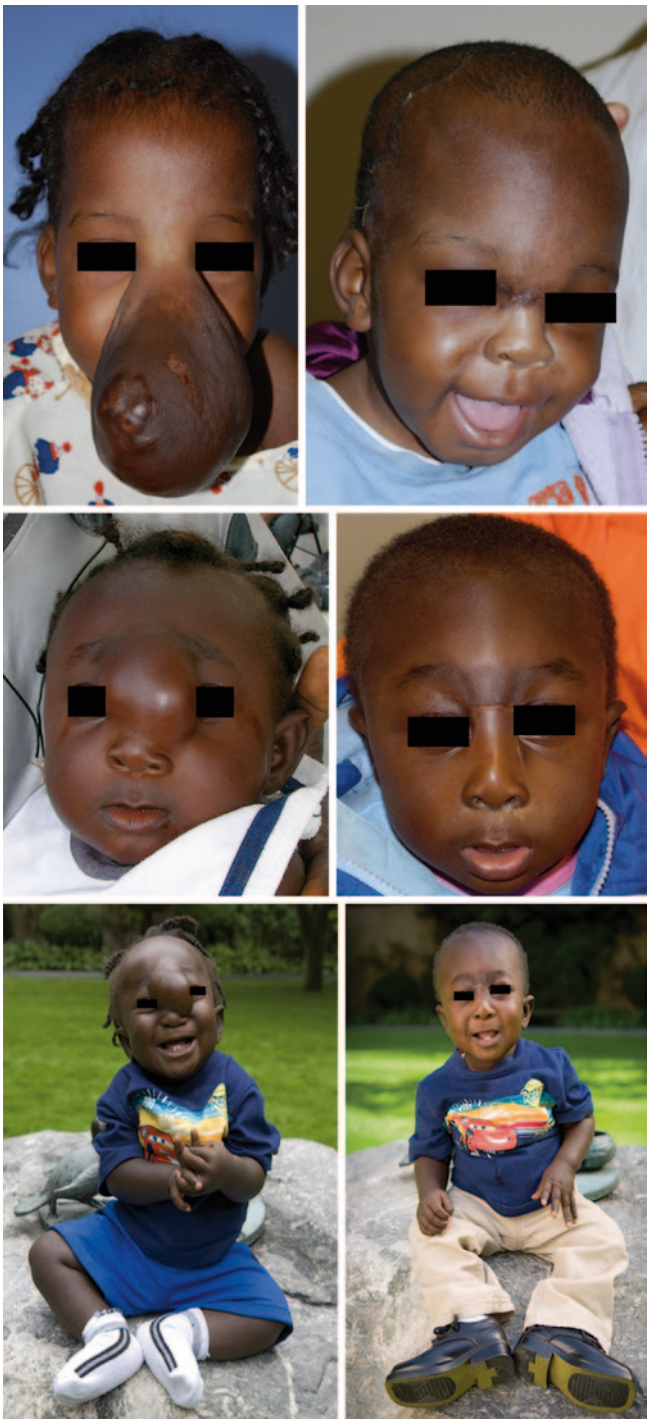


Fig. 16.3 Patients before and after operative intervention

Adjuvant Treatment

Postoperative hydrocephalus is managed by neurosurgery as necessary with conversion of external ventriculostomy to an internal shunt. Some children are at risk of seizure and a course of antiepileptic medication may be indicated.

Outcome

The prognosis of patients who have had anterior encephaloceles corrected is generally good, although mental retardation, epilepsy, and ocular problems have been reported in this population [23, 24]. Children who have the encephalocele repaired well before puberty may require further augmentation of the nasal dorsum later in life as the reconstructed nose is unlikely to grow. Current refinements in technique have shown that a one stage intracranial and extracranial approach is safe and is associated with high parental satisfaction rates of the improvement in appearance (Fig. 16.3) [11, 25, 26].

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