Repair of Frontoethmoidal Encephalocele in the Philippines: An Account of 30 Cases Between 2008–2013

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■ BACKGROUND: **Frontoethmoidal encephalocele is a congenital abnormality of the anterior skull base involving herniation of cranial contents through a midline skull defect. Patency of the foramen cecum, along with other multifactorial variables, contributes to the development of frontoethmoidal encephaloceles. Because of limited resources, financial constraints, and lack of surgical expertise, repair of frontoethmoidal encephaloceles is limited in developing countries.**

■ METHODS: **Between 2008 and 2013 an interdisciplinary team composed of neurosurgeons, craniofacial surgeons, otolaryngologists, plastic surgeons, and nursing personnel, conducted surgical mission trips to Davao City in Mindanao, Philippines. All patients underwent a combined extracranial/intracranial surgical approach, performed in tandem by a neurosurgeon and a craniofacial surgeon, to detach and remove the encephalocele. This procedure was followed by reconstruction of the craniofacial defects.**

■ RESULTS: **A total of 30 cases of frontoethmoidal encephalocele were repaired between 2008 and 2013 (20 male; 10 female). The average age at operation was 8.7 years, with 7 patients older than 17 years. Of the 3 subtypes, the following breakdown was observed in patients: 18 nasoethmoidal; 9 nasofrontal; and 3 naso-orbital. Several patients showed concurrent including enlarged ventricles, arachnoid cysts (both unilateral and bilateral), and gliotic changes, as well as orbit and bulbus oculi (globe) deformities. There were no operative-associated mortalities or neurologic deficits, infections, or hydrocephalus on follow-up during subsequent trips.**

■ CONCLUSIONS: **Despite the limitations of performing advanced surgery in a developing country, the combined interdisciplinary surgical approach has offered effective treatment to improve physical appearance and psychological well-being in afflicted patients.**

INTRODUCTION

Encephaloceles belong to a class of congenital anomalies known collectively as neural tube defects: a diverse array of diseases pertaining to abnormal neurulation involving the neural tube or the coverings thereof. Insufficient separation of surface ectoderm from neural ectoderm, in week 4 of gestation, is the underlying mechanism by which encephaloceles form.[1] The failure of these 2 layers to separate results in an aberrant opening of the skull that permits herniation of intracranial contents, producing either a meningocele (containing meninges only) or an encephalomeningocele (containing both brain and surrounding meninges). Classification of encephaloceles varies by lesion site and includes occipital, sincipital (frontal, anterior, or frontoethmoidal), cranial vault, and basal. Epidemiologic studies regarding ethnic association to anatomic encephalocele location have found that lesions of the occipital region predominate in white races of North America, Europe, and Australia, whereas frontal encephaloceles are more common in regions of Southeast Asia.[2]

Frontoethmoidal encephaloceles account for approximately 20% of total encephalocele cases, and are further subclassified into nasofrontal, nasoethmoidal, and naso-orbital lesion types.[3] Nasofrontal lesions present as a mass located at the glabella, between the frontal and nasal bones, often resulting in

Key words
- Foramen cecum
- Frontoethmoidal encephalocele
- Neural tube defect
- Pediatric craniofacial reconstruction
- Skull base

Abbreviations and Acronyms
CSF: Cerebrospinal fluid
CT: Computed tomography

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telecanthus and inferior displacement of the nasal bones. Nasoethmoidal lesions present as a unilateral or bilateral nasal mass, often resulting in superior displacement of the nasal bones and inferior displacement of the greater alar cartilages. Naso-orbital lesions present as a mass through a defect in the medial orbital wall, often resulting in developmental abnormalities of the globe or visual changes as a result of a pressure effect on the optic nerve.[4] Common abnormalities associated with these lesions include arachnoid cysts, corpus callosum dysgenesis, enlarged and/or asymmetric ventricles, gliotic changes, and in the case of the naso-orbital subtype deformation of the globe.[5]

Cases of frontoethmoidal encephalocele have been reported across Thailand, Indonesia, India, Burma, Cambodia, and Malaysia, with incidence rates ranging from approximately 1:3000 to 1:10,000 live births.[6,7] Some countries, such as India and Thailand, are equipped with the resources and infrastructure to provide early surgical intervention and treatment; however, in other less developed countries, there are substantial barriers to care, especially for patients originating in markedly impoverished rural communities.[8] In these instances, individuals are less likely to be diagnosed at birth and may never receive formal evaluation, with many of those afflicted becoming ostracized from their communities as a result of their physical deformities.[9] Furthermore, this situation can lead to social consequences because promotion of mental health within these countries remains stigmatizing.

Repair of frontoethmoidal encephaloceles in the Philippines has been limited by insufficient resources, financial constraints, and a lack of surgical expertise. A 2012 population estimate in the Philippines determined that 4.2 million families live beneath the national poverty line. In wake of the devastation of Typhoon Haiyan, thousands were displaced and the magnitude of its impact on national poverty has yet to be realized. Most health care resources within the Philippines are allocated to the capital city of Manila, on the island of Luzon, which is known for being the most densely populated area in the world. Even here, the physician/patient ratio is dramatically skewed, and the more refined subspecialties of neurosurgery and reconstructive craniofacial surgery are especially scarce. Through a nonprofit Filipino organization, a population of underserved patients with frontoethmoidal encephaloceles were identified and, since 2008, an interdisciplinary team has traveled to Davao, a city located in the southern Philippines, to evaluate these patients and to facilitate appropriate surgical repair. This team comprised a neurosurgeon, anesthesiologist, craniofacial surgeon, surgical residents, nurse anesthetist, and postoperative nurse support staff, all originating from Michigan, United States. Supplies required for preoperative preparation, surgical correction, and postoperative management were donated directly from industry. Careful attention was given to secure the appropriate equipment and pharmacologic therapies necessary to successfully manage any potential surgical or postoperative complications.

This report details 30 cases of frontoethmoidal encephalocele treated by this interdisciplinary team at the Southern Philippines Medical Center, located on the island of Mindanao. The frequency of frontoethmoidal encephalocele subtype, as well as the incidence of presenting abnormalities and comorbidities, is also described. Thorough detail is given to expound on the procedures used in the surgical correction as well as the rectification of the underlying osseous defect.

METHODS

Patient Population

During an annual trip, conducted between 2008 and 2013, 30 patients underwent repair for frontoethmoidal encephaloceles at the Southern Philippines Medical Center by a visiting medical team from the United States. Before arrival of the surgical team, all patients were locally screened and computed tomography (CT) imaging studies were performed. Patients were examined in person on the day of the surgical team's arrival and a thorough review of all patient records and CT scans was conducted. After patient assessments were completed, surgeries were then scheduled in a triage fashion, such that patients with the greatest deformity, and as a result a more complicated repair, were scheduled earlier in the trip, whereas those with less complicated disease were scheduled for later in the trip.

Classification of Frontoethmoidal Encephaloceles

Each case of frontoethmoidal encephalocele was classified by subtype via CT scans and later confirmed intraoperatively. The classification scheme that was used was originally described by Suwanwela and Suwanwela,[2,10] and is based on the anatomic site of the lesion. Each of the subtypes, nasoethmoidal (**Figure 1**), nasofrontal (**Figure 2**) and naso-orbital (**Figure 3**), was present in the patient population.

Surgical Procedure

Our surgical technique to correct the various frontoethmoidal encephalocele subtypes is detailed as follows and shown in **Figure 4**. Patients are positioned supine on the operative table and undergo endotracheal intubation as well as general anesthesia. The head and face are prepared in the usual sterile fashion and a bicoronal incision is performed in a zigzag fashion and carried down through the galea. The zigzag technique allows for improved camouflage of the resultant scar, especially when the hair is wet. The galea is then separated from the underlying pericranium via sharp dissection, with careful attention paid to avoid violating the integrity of the pericranium. A large vascularized pericranial flap is developed laterally to the superior temporal lines and posteriorly to the level of the vertex, with careful attention paid to maintain a vascularized pedicle anteriorly. This step is essential because preserving a viable pericranial flap minimizes the use of an artificial dural graft, allowing this scarce resource to be preserved for when absolutely necessary. Dissection is then continued, separating the periorbita from the orbital walls. Once the pericranial flap has been harvested, it is rolled in a wet sponge to maintain moisture. Burr holes are then placed on both sides of midline, anterior to the coronal suture. Bifrontal craniotomy is then performed and the bone flap is carefully elevated from the dura and removed. Next, bilateral superior orbital and nasal osteotomies are performed. A subfrontal approach, along the anterior skull base floor, is then used to visualize the foramen cecum and underlying encephalocele. The dura along this course can often be extremely friable, and thus, meticulous