Single-Stage Combined Craniofacial Repair for Frontoethmoidal Meningoencephalocele

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Introduction: Frontoethmoidal meningoencephalocele (FMEC) is a rare congenital anomaly characterized by herniation of brain tissue and meninges through a defect in the cranium and associated with facial dysmorphism. Treatment modalities include extra cranial, transcranial, or combined craniofacial approaches. The combined approach is considered the best treatment choice.

Methods: Twelve patients with FMEC aging from 6 months to 4 years were treated by single-stage combined craniofacial approach between July 2011and July 2015. They were followed up for evaluation of outcome and detection of complications.

Results: Seven patients (58.3%) were males and 5 patients (41.7%) were females. Eight patients (66.7%) were less than 2 years while 4 patients (33.3%) were between 2 and 4 years. The main presentations were external mass, telecanthus and hypertelorbitism, radiologically, frontobasal bone defect and herniated dural sac with brain tissue were detected in all patients. Excision of the mass with dural repair, craniofacial reconstruction, and medial canthopexy were done for all patients. Orbital translocation was done for 8 patients (75%), nasal reconstruction for 7 patients (58.3%), while dacryocystorhinostomy in 3 patients (25%). Venticuloperitoneal shunt was done before correction of FMEC in 1 patient (8.3%). The follow-up period ranged from 6 to 48 months with mean 29.2 months. The esthetic results were satisfactory in 9 patients (75%). Ugly facial scars were recorded in 3 patients (25%).

Conclusion: Early surgical management for FMEC is advisable to avoid deleterious effects on facial growth. Meticulous perioperative care is important for successful surgery. The authors recommend combined craniofacial approach to achieve good outcome and decrease the incidence of complications.

Key Words: Combined craniofacial approach, frontoethmoidal meningoencephalocele, hypertelorism, telecanthus

Encephalocele is defined as protrusion of cranial contents beyond the normal confines of the skull. Frontoethmoidal meningoencephaloceles (FMEC) are common in many Southeast Asian countries, with incidence 1 in 5000 live births.[1–3] A slight male AQ5 preponderance was reported.[3–5] These lesions often affect poor, rural children in developing countries but their etiology is still poorly understood.[6–8] Some authors suggest genetic predisposition, parental consanguinity, vitamin B, and folic acid deficiency due to maternal malnutrition and drugs.[7,9,10]

They may contain meninges (meningocele), brain matter, and meninges (meningoencephalocele), or they may communicate with the ventricles (meningoencephalocystocele). Frontoethmoidal myelomeningoceles were classified according to the system described by Suwanwela and Suwanwela,[8] dividing the deformity into nasofrontal, nasoethmoidal, and nasoorbital meningoencephalocoeles. Nasoethmoidal (sincipital type) is the most common and nasoorbital subtype is least common.[11]

These swellings are either sessile or pedunculated and on palpation may vary from being solid and firm to soft and cystic. The skin over the mass may be normal in appearance, thin and shiny, or thick and wrinkled. Hyperpigmentation and hypertrichosis may be noted. Visual acuity may be decreased. Strabismus and lacrimal obstructions, resulting in epiphora and/or dacryocystitis, can be observed.[12,13] Furthermore, some of these children have neurological complications or associated brain anomalies,[14,15] although most are mentally normal.[16,17]

Frontoethmoidal meningoencephaloceles are often accompanied by an increased distance between the medial orbital walls, but not the distance between the lateral orbital walls. This is called interorbital hypertelorism.[18] Encephaloceles can cause recurrent meningioencephalitis due to direct communication of the central nervous system with the external environment, facilitating the entry of pathological microorganisms. The bacteria most commonly associated with meningitis in such patients is Steptococcus pneumoniae, followed by Staphylococcus aureus then Neisseria meningitides.[19]

Surgical treatment for this malformation is a complex task, its primary aim being to close reliably the connection between the intradural and extradural spaces. The choice of the best surgical treatment for FMEC is still debated. Meticulous planning is necessary for this choice depending in the type and size of the encephaloceles and associated hydrocephalus. Most authors recommend the combined procedure.[17,20,21]

The aim of this work is to study the use of early single-stage combined craniofacial repair for frontoethmoidal meningoencephalocele and evaluate the outcome and complications in a selected group of patients in our locality.

METHODS

This is a prospective study that was done during the period from July 2011 to July 2015, after approval by the Institutional Human Research and Ethics Committee. Informed consent was obtained from the parents or the legal guardian of the patient.

This study included 12 patients with FMEC that were selected to be new patients not previously operated with age not less than 6 months and weight not less than 6 kg. Previously operated patients were excluded.

All of our patients underwent a clinical evaluation including family history, cranio-facial examination, and ophthalmologic study. Plain X rays, three-dimensional computed tomography study were done for all patients. MRI was done in some patients. Routine laboratory investigations were done. Preoperative and postoperative photos were taken for each patient.

A standard craniofacial approach was done for all patients. Soft tissue and skeletal procedures were done when indicated. These procedures included excision of the mass with dural repair, medial canthopexy, nasal reconstruction, dacryocystorhinostomy, craniofacial reconstruction, and box osteotomy for orbital translocation. Also, venticuloperitoneal shunt would be done if there was hydrocephalus. Rib bone graft was used for closure of the bony defect (Fig. 1). Naso-lacrimal drainage was done using silicon rods. Squint was corrected by ophthalmologists 2 to 3 months after the primary repair.

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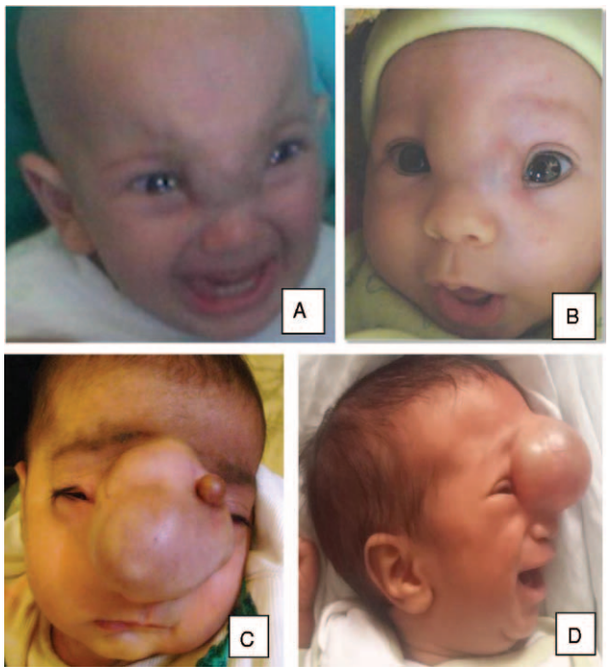

FIGURE 1. Preoperative photos of some patients of our series. (A) Preoperative photos patient no. 1. (B) Preoperative photos patient no. 2. (C) Preoperative photos patient no. 3. (D) Preoperative photos patient no. 4.

All patients were followed up for evaluation of the outcome and detection of complications. The esthetic outcome was evaluated by both parents of the patient and surgeons. Visual analogue scale was used by parents of the patients for measuring the overall satisfaction of the outcome. Evaluation of the esthetic outcome by the surgeons was classified into 3 categories: a poor outcome means that the postoperative facial appearance of patient was still incompatible with a normal social life without improvement; an average result means that the patient's facial appearance was improved but with some abnormalities; a good result means the facial appearance was normal with a few scars.

RESULTS

This prospective study included 12 patients of FMEC treated between July 2011 and July 2015 at Sohag University Hospitals in collaboration between the Neurosurgical and Maxillofacial Surgery Departments. Regarding sex of the patients, 7 patients (58.3%) were males and 5 patients (41.7%) were females (male/female ratio 1.4:1). Their ages ranged between 6 months to 4 years. Eight patients (66.7%) were less than 2 years while 4 patients (33.3%) were between 2 and 4 years (Table 1).

TABLE 1. Sociodemographic Characteristics of the Patients of the Study

Item	No. of Patients (%)
Sex	
Male	7 (58.3%)
Female	5 (41.7%)
Age	
Less than 2 years	8 (66.7%)
Between 2 and 4 years	4 (33.3%)

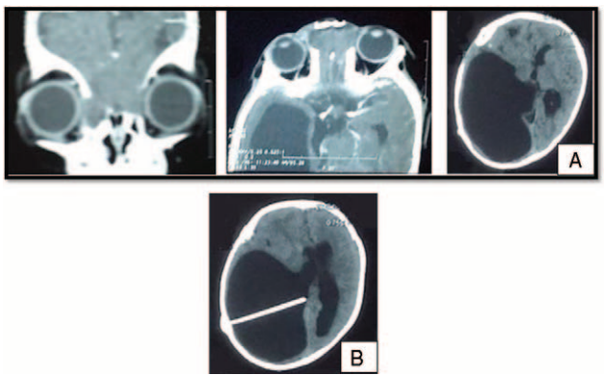

FIGURE 2. Computed tomography (CT) brain showing frontobasal bone defect and hydrocephalus. (A) Preoperative CT brain (coronal and axial cuts) showing frontobasal bone defect and hydrocephalus. (B) Postoperative axial CT scan with right v-p shunt.

All patients presented with external mass and telecanthus. Hypertelorbitism occurred in 8 patients (66.7%) and long nose deformity in 4 patients (33.3%). Hydrocephalus was found in 1 patient (8.3%). Ocular anomalies occurred in the form of lacrimal drainage dysfunction in 5 patients (41.7%), microphthalmia in 1 patient (8.3%), and squint in another patient (8.3%) (Fig. 1). Computed tomography and MRI showed Herniated dural sac with brain tissue in all patients. Frontobasal bone defect was detected in all patients, it was unilateral in 9 patients (75%) and bilateral in 3 patients (25%). Hydrocephalus was detected in 1 patient (8.3%) (Figs. 2, 3 and Table 2).

Regarding the surgical procedures that were done for the correction of the FMEC, excision of the mass with dural repair, craniofacial reconstruction, and medial canthopexy were done for all patients. Craniofacial reconstruction to close the bony defect was done using rib bone graft in all patients. Orbital translocation was done for 8 patients (75%), nasal reconstruction for 7 patients (58.3%), while dacryocystorhinostomy in 3 patients (25%). Ventriculo-peritoneal shunt was done before correction of FMEC in 1 case (8.3%) that was associated with hydrocephalus (Figs. 2, 4, 5 and Table 3).

The follow-up period ranged from 6 to 48 months with a mean of 29.2 months, during which the patients were evaluated as regarding the esthetic outcome and the complications.

The parents of all patients were satisfied by the esthetic results. The mean level of satisfaction was 83.75% using a visual analogue scale. Surgeons found that 9 patients (75%) had good esthetic outcome while only 3 patients (25%) had average results. Regarding the complications, 3 patients (25%) had ugly facial scars.

DISCUSSION

Planning for surgery and the age of operation is very important in management of FMEC. Most authors agree that FMEC should be treated as soon as possible to avoid deleterious effects on facial

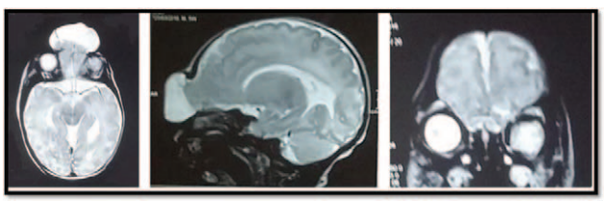

FIGURE 3. Preoperative MRI brain T2, axial, sagittal, and coronal cuts showing herniated dural sac with brain tissue.

TABLE 2. Clinical Characteristics and Radiological Characteristics

Item*	Frequency (%)
Clinical characteristics	
External mass	12 (100%)
Telecanthus	12 (100%)
Hypertelorbitism	8 (66.6%)
Long nose deformity	4 (33.3%)
Lacrimal drainage dysfunction	5 (41.7%)
Microphthalmia	1 (8.3%)
Squint	1 (8.3%)
Radiological characteristics	
Herniated dural sac with brain tissue	12 (100%)
Unilateral frontobasal bone defect	9 (75%)
Bilateral frontobasal bone defect	3 (25%)
Hydrocephalus	1 (8.3%)

*Multiple items can be found in the same patient.

growth.[21,22] Care should be taken to reduce the risks of anaesthesia and surgery, which can be more common and severe in young babies.[14] Some schools do not prefer performing a complete facial skeletal correction before the age of 3 to 4 years, and therein comes the role of late surgery for secondary facial deformity correction.[19]

In our series, we avoided treating children less than 6 month of age or weighed less than 6 kg. Prolonged surgery (1-stage correction), blood loss, hypothermia, and electrolyte disturbances are the important intraoperative complications in the pediatric neurosurgery. We controlled blood loss by preparation cross matched packed RBCs, infiltration of vasoconstrictive agents (epinephrine 1: 200,000) into the scalp 7 to 10 minutes before scalp incision and

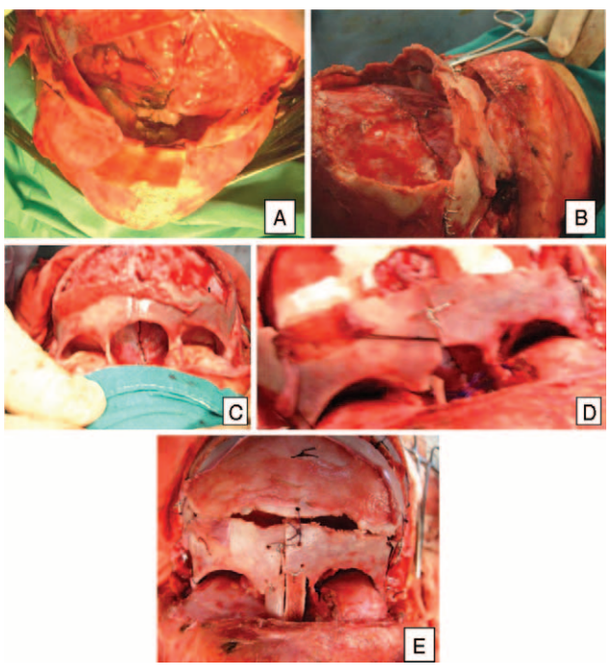

FIGURE 4. The surgical procedure. (A) Craniotomy, excision of the mass. (B) Dural repair. (C) Frontobasal bone defect and orbital hypertelorism. (D) Bilateral medial canthopexy. (E) Orbital box osteotomy, medial orbital translocation, and split rib bone graft for the reconstruction of the defect.

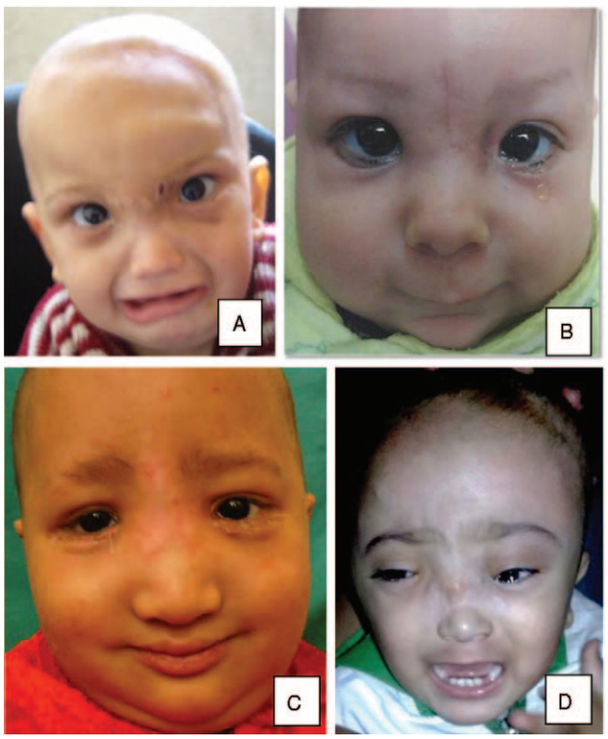

FIGURE 5. Postoperative photos of some patients of our series. (A) Postoperative photos patient no. 1. (B) Postoperative photos patient no. 2. (C) Postoperative photos patient no. 5. (D) Postoperative photos patient no. 6.

using bone wax. Hypothermia was avoided by wrapping the patient by warming blanket, warming the IV fluids and transfused blood. Electrolyte disturbance was managed by intraoperative monitoring. All patients were admitted to pediatric intensive care unit for 48 hours at least for observation of the vital signs, level of consciousness, and postoperative care. Other authors prefer delaying surgical treatment to 8 to 10 months of age to minimize these complications.[11]

The choice of the best surgical treatment for FMEC is debated. Most authors consider that the best method is the "combined" procedure.[11,15,21,23] In our study, the combined craniofacial procedure was used for all patients because intracranial approach guaranteed a secure closure of the dura, removal the encephalocele, and repair the skull defect. Also, the facial approach was important to achieve correction of the associated facial deformities in the same surgery, especially hypertelorism and nasal reconstruction.

TABLE 3. The Surgical Procedures

Procedure*	Frequency (%)
Excision of the mass with dural repair	12 (100%)
Craniofacial reconstruction	12 (100%)
Medial canthopexy	12 (100%)
Orbital translocation	8 (75%)
Nasal reconstruction	7 (58.3%)
Dacryocystorhinostomy	3 (25%)
Venticuloperitoneal shunt	1 (8.3%)

*Multiple procedures can be done for the same patient.

Others used either cranial, facial approaches, or combined procedures according to the patient malformation pattern.[22] Some claim that FMEC can be corrected only by an extracranial procedure.[4,24] We think this was done in developing countries in past years where surgical expertise was lacking and perioperative care was deficient.

In our series 1 patient has hydrocephalus and was treated by V-P shunt before the correction of FMEC as we think that shunting prior to definitive surgery prevents the risk of postoperative CSF leak and facilitates the surgical approach. Other authors think that the presence of hydrocephalus does not necessarily cause problems or require treatment.[23]

In our series, nasal reconstruction was performed in the same surgery in 7 patients. Costo-chondral grafts were used for reconstruction. Some authors recommend performing nasal reconstruction first to avoid the long-nose deformity or later after the age of skeletal maturity for definite reconstruction.[20]

In our series a single-stage repair was done for all patients. We think that performing full correction of the FMEC and associated deformities in 1 stage is better. Postponing the skeletal correction can increase the deformity. Multiple stages are associated with more complications especially dural tears and CSF leak. Also, growth of the craniofacial bones can be affected by repeated interference with their blood supply. Although single-stage correction is associated with prolonged operative time, close intraoperative and postoperative monitoring can decrease complications. The financial costs and psychological impact on the patient and his family are more with repeated surgeries. In accordance with our opinion, some authors believe that best results would be produced in terms of facial correction when 1-stage reconstruction is done at the time of the encephalocele correction.[23,25,26] In contrary, Charoonsmith and Suwanwela have suggested that only encephalocele correction is needed initially and that the facial deformity would regress by itself in the developing face and the bony defect would close. They have advised that surgery of the nose and the facial bones be kept to a minimum. The correction of hypertelorism can be undertaken at the age of 3 years if it has not corrected by itself.[27]

The most feared complications in FMEC are CSF leak and infection. In our study none of these complications was recorded, as we used the combined craniofacial approach, performing watertight closure of the dura, and applying fibrin glue with or without dural graft. Others found that postoperative morbidity in FMEC treatment is usually due to infection and CSF leaks due to the use of the transfacial approach, and inadequate base reconstruction with no watertight dura closure.[4,5,11,28]

All our patients had postoperative facial scars that were ugly in only 3 patients (25%). These scars were due to the use of the facial incisions in the combined craniofacial approach. We tried to minimize the incidence of ugly nasal scars by placing the incisions at the borders of the nasal subunits which may extend laterally and by using local care. The esthetic outcome was satisfactory for patients' parents and also for the surgeons.

CONCLUSION

Early surgical management for FMEC is advisable to avoid deleterious effects on facial growth. Meticulous perioperative care is important for successful surgery. We recommend combined craniofacial approach to achieve good outcome and decrease the incidence of complications.

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