Frontoethmoidal Meningoencephalocele: Challenges and the Tawanchai Center's Long-Term Integrated Management

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Background: *The challenges for the management of patients with fronto-ethmoidal meningoencephalocele (FEEM) include: classification, assessment and analysis of the deformities, craniofacial reconstruction and long-term management.*
Objective: *To present experience of the Tawanchai Craniofacial Center of long-term integrated management and outcome of patients with FEEM.*
Material and Method: *Medical records were reviewed of 32 patients with FEEM treated by the authors between 1993 and 2011 at the Tawanchai Center, Srinagarind Hospital; the referral center for Northeast Thailand.*
Results: *Geographic Information System (GIS) analysis was used to examine the incidence and pattern of referrals to our Center. Most of the patients had the nasoethmoidal type (12 patients) followed by the combined naso-ethmoidal/-orbital type (8 patients). The surgical procedures included craniofacial reconstruction with medial canthopexy, orbital translocation, external repair and nasal reconstruction. Ultimately, most patients were satisfied with their remedied facial appearance. The Center's interdisciplinary protocol for the care of patients with FEEM was established.*
Conclusion: *Experience demonstrated that a craniofacial center with interdisciplinary management was necessary to provide proper, early and longitudinal care and to achieve optimum outcomes for the patients with FEEM. In each case, the surgical outcome depended on the severity and classification of the deformities and the extent of associated brain anomalies. Nevertheless, in every case the final measurement should be done at the age of complete skeletal maturity. Funding from a number of sources, including the Foundation, is needed to ensure patients' access to treatment and follow-up and for the Craniofacial Cleft Center to improve the quality of treatment and programing.*

Keywords*: Frontoethmoidal meningoencephalocele, Challenges, The Tawanchai Center, Long-term integrated management*

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A meningoencephalocele is a herniation of the brain and meninges beyond the normal confines of the skull[1]. The type of skull defect may be classified as occipital, parietal, basal, and sincipital or frontoethmoidal. The site of the sincipital or frontoethomoidal meningoencephalocele (FEEM) type is at the cranial end of the defect through an internal skull defect at the area of the foramen cecum at the junction of the frontal and ethmoidal bones. The herniation of brain tissue is through the external skull defect and extending to the face[2]. Approximately 50% of the patients have an internal defect in the midline at the foramen cecum; 25% have this defect on one side of the midline and 25% on both sides[1]. FEEMs are classified according to the location of the external skull defect[3]; as naso-frontal, nasoethmoidal or naso-orbital, with some overlap or multiplicity. The contents may include the meninges (meningocele), meninges and brain (meningoencephalocele), or part of a ventricle (hydroencephalomeningocele).

Changes in facial features of patients with

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FEEM which arise from the prolapse of the sac of the meningoencephalocele may differ for each type, but the common features are medial orbital hypertelorism and an elongated midface[1,4,5]. FEEM may present with a facial mass covered with normal skin, while basal encephaloceles may present with nasal obstruction or symptoms related to herniation of basal structures[6]. Complete removal of the dysplastic tissue will allow the developing brain and eyes to mold the orbital skeleton, and allow development of: a proper nasal airway, speech, and mastication; that is, to remodel the awkward[7]. Some authors recommend early surgical correction to minimize the pressure effect of the mass on facial growth[8,9].

FEEMs have a relatively high incidence (1:5000 live births) in Southeast Asia[10] and are common in Malaysia, Thailand, and Burma. In Thailand and Burma, FEEMs occur in 1 out of 5,000-6,000 live births[1,11]. Chulalongkorn Hospital in Bangkok, Thailand, reported that most of their patients with FEEM came from the lower northern and northeast regions with the highest incidence being from Kamphaeng Phet (7.5%), Surin (6.6%) Si Sa Ket (6.6) Nakhon Ratchasima (5.7) Buri Ram (5.7) and Nakhon Sawan (5.7%) provinces[12].

The objectives of the present study were (a) to present the long-term experience of the authors and the Tawanchai Center on the challenges in evaluation and long-term, integrated management of patients with FEEM in Thailand. The results from analysis of these data can be used as current knowledge and applicable for recommendations for future clinical and surgical approaches to patients with FEEM.

Material and Method

Study Design

Medical records were reviewed of patients with FEEM seen and managed by the authors at the Tawanchai Center at Srinagarind Hospital, Khon Kaen University. Srinagarind Hospital is a university hospital and the main tertiary referral center for the northeast of Thailand, which has a population of about 22 million people. The diagnosis of FEEM was based by clinical and radiological reports. The details of the clinical presentation, analysis, reconstructive surgeries and long-term management were noted and analyzed.

The protocol of the present study was reviewed and approved by the Ethics Committee of Khon Kaen University, according to the standards set out in the Helsinki Declaration. Written informed consent was obtained from each patient.

Results

Patient report

The 32 patients with FEEM included in this study were seen between 1993 and 2011. The female-to-male ratio was 1.2 to 1 [18 female (F) and 15 males (M)]. There were 12 patients with the nasoethmoidal type, 6 with the nasofrontal type, 2 with the nasoorbital type, and 8 with a combined naso-ethmoidal/-orbital type. The surgical procedures included craniofacial reconstruction with medial canthopexy in 22 patients, orbital translocation in 4, external repair in 2, and nasal reconstruction in 6 (Table 1). Fig. 1 shows the geographic distribution of the 33 patients using a Geographic Information System (GIS). Khon Kaen, Nong Bua Lam Phu and Chaiyaphum are among the provinces with the highest number of patients.

Treatment protocol by age

Infant period (the first year of age)

Emergency closure of FEEM is indicated in (a) the child born with an open FEEM to prevent meningitis and (b) the child with twisted, infracted FEEM, or obstructed vision. Craniofacial reconstruction is recommended to be performed between the age of 5 and 10 months; to lessen the risks of anesthesia and blood loss and the necessity of disturbance to subsequent growth.

Early school age (5-7 years)

Secondary craniofacial reconstruction may be indicated to manage the psychological impacts from residual FEEM. The concerns at this stage are (a) the growth of reconstructed bone and (b) any additional reconstructions needed during puberty.

Late and post Puberty (> 18 years)

The correction of the depressed nose, the long-nose deformity and the long mid-face, as well as any maxillary or orbital surgery may be indicated.

The intracranial and extracranial deformities were assessed using 3-D computer tomography (CT scan) and magnetic resonance imaging (MRI). The craniofacial reconstruction was performed by a plastic surgeon and neurosurgeon, who first made an evaluation, then planned a strategy. During the early period, traditional craniofacial reconstruction was performed using a combined intra- and extracranial approach; identifying the internal skull defect then closing the defect with bone and fascia, and finally performing orbital translocation, if indicated (Fig. 2, 1st row). Subsequently, U-shaped, medial, orbital

Table 1. Details of the 32 patients with FEEM treated by the authors at Srinagarind Hospital, Khon Kaen University, between 1993 and 2011

Patient No.	Year of Birth	Sex	Province	Classification	Treatment
1	1987	F	Udon Thani	Naso-ethmoidal	Craniofacial reconstruction, nasal reconstruction with bone graft and dermis fat graft
2	1970	F	Nong Bua Lam Phu	Naso-ethmoidal	Craniofacial reconstruction, orbital translocation, medial canthopexy, dacryocystorhinostomy, removal of right lacrimal sac
3	1969	F	Nong Bua Lam Phu	Naso-ethmoidal/ naso-orbital	-
4	1992	M	Maha Sarakham	Naso-ethmoidal/ naso-orbital	Neurosurgical excision, craniofacial reconstruction, medial canthopexy, nasal reconstruction
5	1982	F	Surin	Naso-ethmoidal	Craniofacial reconstruction
6	1993	F	Chaiyaphum	Naso-frontal	Craniofacial reconstruction with medial canthopexy, nasal reconstruction with calvarial bone graft and fascial grafts
7	1993	M	Maha Sarakham	Naso-frontal	Craniofacial reconstruction with medial canthopexy
8	1987	F	Roi Et	Naso-ethmoidal	Craniofacial reconstruction with medial canthopexy
9	1978	F	Loei	Naso-frontal	Craniofacial reconstruction with medial canthopexy
10	1994	M	Nong Khai	Naso-ethmoidal/ naso-orbital with hydrocephalus	-
11	1981	M	Khon Kaen	Undetermined	-
12	1985	F	Phetchabun	Naso-ethmoidal/ naso-orbital	Craniofacial reconstruction with medial canthopexy
13	1981	M	Chaiyaphum	Nasofrontal	Craniofacial reconstruction with median canthopexy
14	1982	M	Nong Khai	Naso-ethmoidal	Craniofacial reconstruction, medial orbital translocation with medial canthopexy
15	1982	F	Nong Bua Lam Phu	Nasofrontal	Craniofacial reconstruction, medial orbital translocation with medial canthopexy
16	1994	M	Maha Sarakham	Naso-ethmoid/ naso-orbital and cleft palate	Craniofacial reconstruction, medial orbital translocation with medial canthopexy
17	1995	F	Loei	Naso-orbital	Craniofacial reconstruction with medial canthopexy and nasal reconstruction with calvarial bone graft and fascial grafts
18	1994	F	Nong Bua Lam Phu	Undetermined	-
19	1995	M	Khon Kaen	Naso-ethmoidal with left microphthalmos	Craniofacial reconstruction with medial canthopexy
20	1986	M	Chaiyaphum	Undetermined	-
21	1996	F	Chaiyaphum	Naso-frontal	External repair
22	1995	M	Loei	Naso-ethmoidal	Craniofacial reconstruction with medial canthopexy
23	1999	F	Roi Et	Undetermined	-
24	1998	M	Loei	Naso-ethmoidal/ naso-orbital	Craniofacial reconstruction with medial canthopexy
25	1981	F	Khon Kaen	Naso-orbital	External repair
26	1977	M	Roi Et	Naso-ethmoidal	Craniofacial reconstruction with medial canthopexy
27	2004	M	Khon Kaen	Naso-ethmoidal, Microphthalmos and hydrocephalus	-
28	1987	F	Roi Et	Naso-ethmoidal. naso-orbital	Craniofacial reconstruction with medial canthopexy and nasal reconstruction

Table 1. Cont.

Patient No.	Year of Birth	Sex	Province	Classification	Treatment
29	2004	M	Nong Bua Lam Phu	Naso-ethmoidal	Craniofacial reconstruction with medial canthopexy
30	2006	M	Nong Khai	Naso-ethmoidal, cleft lip and cleft palate	Craniofacial reconstruction with medial canthopexy, cleft lip and palate repair
31	2009	M	Roi Et	Naso-ethmoidal	-
32	1970	F	Chaiyaphum	Naso-ethmoidal/ naso-orbital	Craniofacial reconstruction with medial canthopexy, nasal reconstruction

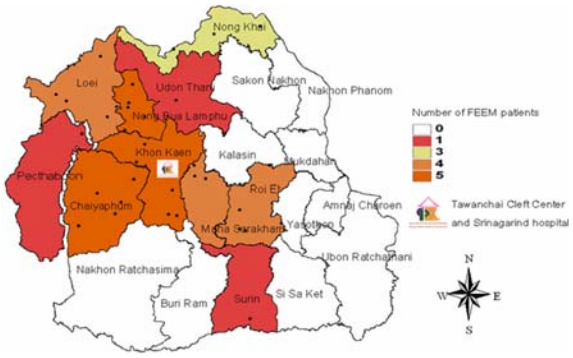

Fig. 1 Geographic distribution of the 32 patients with FEEM treated at Srinagarind Hospital between 1993 and 2011

translocation was used (Fig. 2, 2nd row). At present, on most of the patients the inverted T-shaped medial orbital translocation can be used as well as a more limited intracranial procedure using a naso-frontal bone flap (Fig. 2, 3rd row). Nasal reconstruction-as well as orbital, maxillary and mandibular surgery-may be performed, if indicated, at skeletal maturity age (Fig. 2, 4th row).

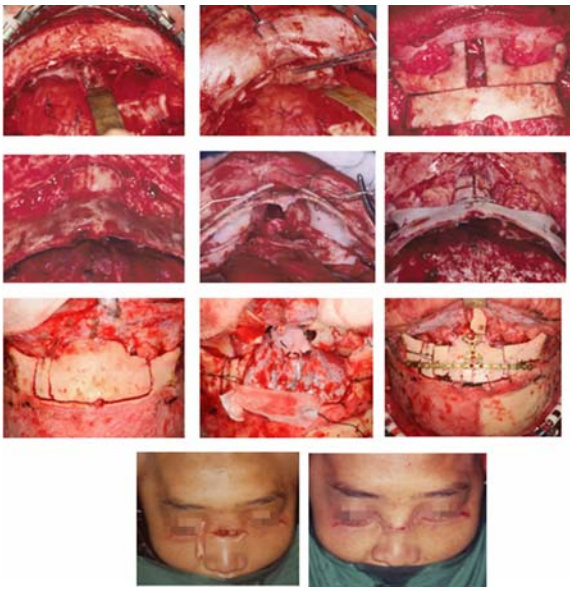

Fig. 2 Intraoperative photos of craniofacial reconstruction. First row: the traditional craniofacial reconstruction using a combined intra- and extracranial approach and orbital translocation. Second row: U-shaped medial orbital translocation. Third row: inverted T-shaped medial orbital translocation and more limited intracranial procedure by nasofrontal bone flap. Fourth row: nasal reconstruction with bone graft

Patient Report

Patient No. 2

A female patient, born in 1970 in Nongbualampoo province, presented with naso-orbital FEEM. The surgical reconstruction included closure defect with orbital translocation in 1990, medial canthopexy with dacryocystorhinostomy (DCR) in 2000 and removal of the right lacrimal sac in 2008. She was satisfied with her corrected facial appearance and was married and had two normal children.

Patient No. 4

A male patient, born in 1993 in Mahasarakam province, presented with naso-ethmoidal/-orbital FEEM, amblyopia and right strabismus. The surgical reconstruction included first neurosurgical repair during the infancy period, craniofacial reconstruction in 1995 and calvarial bone graft (with right medial canthopexy) in 2011. He continued his education until he finished primary school level. At the age of 18 years, his family and he were satisfied with his facial

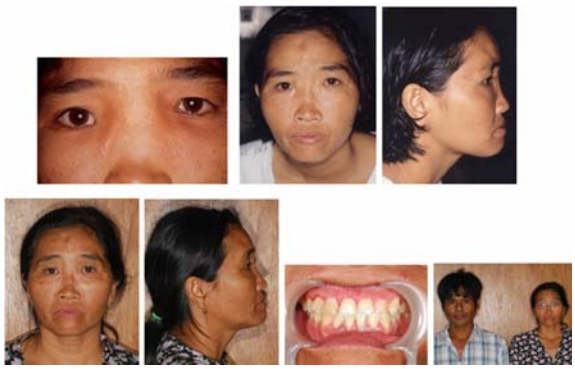

Fig. 3 Patient No. 2 presented with naso-orbital FEEM, treated by closure defect with orbital translocation, medial canthopexy with dacryocystorhinostomy (DCR) and removal of the right lacrimal sac. Last follow-up photos at 41 years of age with her husband show final satisfactory facial appearance

appearance.

Patient No. 6

A female patient, born in 1993 in Chaiyapoom, presented with naso-orbital FEEM. Interestingly, she had a mother with a right unilateral cleft lip. The child's surgical reconstruction included external repair before the age of 1 year and nasal reconstruction with calvarial bone graft the age of 18 years. In 2011, she was studying at secondary level in a professional college and had a satisfactory facial appearance.

Patient No. 12

A female patient, born in 1985 in Petchaboon province, presented with epilepsy, naso-ethmoidal/-orbital FEEM, right nasal strabismus and chronic pansinusitis. The surgical reconstruction was combined craniofacial reconstruction. At the last follow-up in 2011, at the age of 26 years, she had an acceptable facial appearance and had borne a child. Unfortunately, she had persistent epilepsy but was continuing her medical treatment for a neurologic condition.

Patient No. 19

A male patient, born in 1995 in Khon Kaen, presented with naso-ethmoidal FEEM and left microphthalmos. He had undergone surgical craniofacial reconstruction with medial canthopexy. He discontinued his education after finishing primary school and receives disability support. At follow-up, when 16 years of age, he had a satisfactory facial appearance.

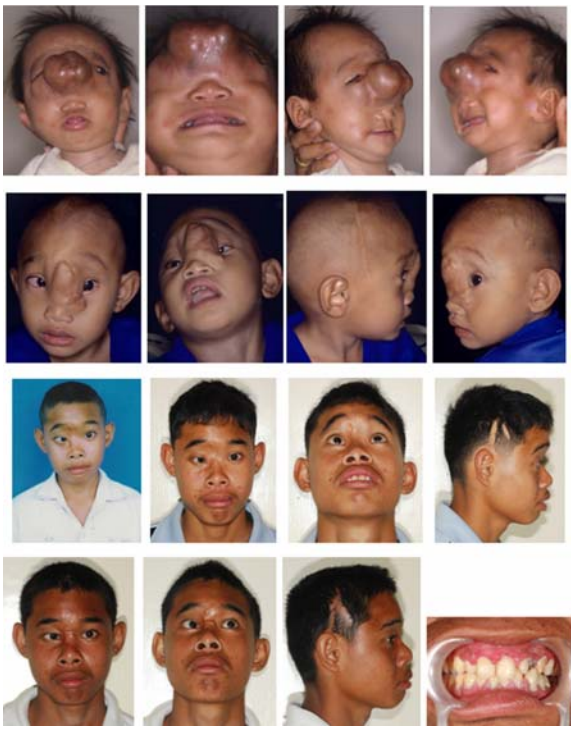

Fig. 4 Patient No. 4, presented with naso-ethmoidal/-orbital FEEM, amblyopia and right strabismus. It was treated first using neurosurgical repair, craniofacial reconstruction, and nasal reconstruction with calvarial bone graft with right medial canthopexy. Follow-up photos are at age of 18 years, showing the final satisfactory facial appearance

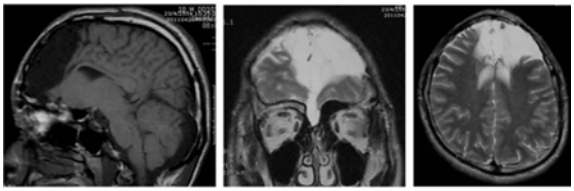

Fig. 5 MRI brain of patient No. 4 after surgical correction at the age of 18. Sagittal T1W(a), Coronal T2W(b) and Axial T2W show a large cystic lesion replacing the damaged brain tissue in the anterior part of the bilateral frontal lobe, predominately on the left side, which represents a porencephalic cyst

Patient No. 28

A female patient, born 1987 in Roi Et province, presented with naso-ethmoidal meningoencephalocele. The surgical reconstruction included craniofacial reconstruction, medial orbital translocation, medial cantopexy and calvarial bone graft in 2001. At the time of the last follow-up in 2011, she had a satisfactory

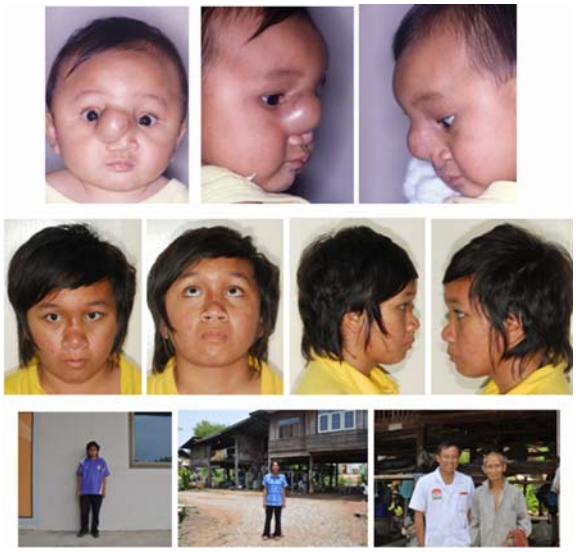

Fig. 6 Photos of patient No. 6, presented with naso-orbital FEEM, treated by external repair and nasal reconstruction with calvarial bone graft. Follow-up photos, at the age of 18 years, at her college and with her father and mother at the village, show a satisfactory facial appearance

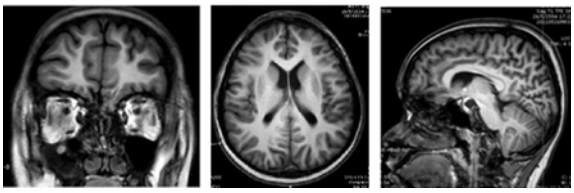

Fig. 7 MRI of patient No. 6 after surgical correction at 18 years of age. Coronal T1W (a), Sagittal T1W (b) and Axial T1W (c) images show a slightly, low-lying, cribiform plate and absent frontal air sinus. No herniation of the brain tissue is identifiable

facial appearance and had recently married.

Patient No. 32

A female patient presented at 41 years of age. She was born with naso-ethmiodal/-orbital FEEM and anomalies of the hands and feet. She lived with her mother and sister in a village in Chaiyapum province, finished primary school and was a laborer. Due to a lack of financial resources and inadequate information about treatment options, her deformities were left untreated until 2011 when she met with team of Tawanchai Foundation and decided to go to Srinagarind Hospital for surgical correction of her facial deformities.

The review and analysis of treatment and outcome of these patients has lead to the development

Fig. 8 Patient No. 12 presented with epilepsy, naso-ethmoidal/-orbital FEEM, right nasal strabismus and chronic pansinusitis, treated by craniofacial reconstruction and medial canthopexy. The last follow-up photos, taken when she was 26 years of age, with her mother and her daughter, show an acceptable facial appearance

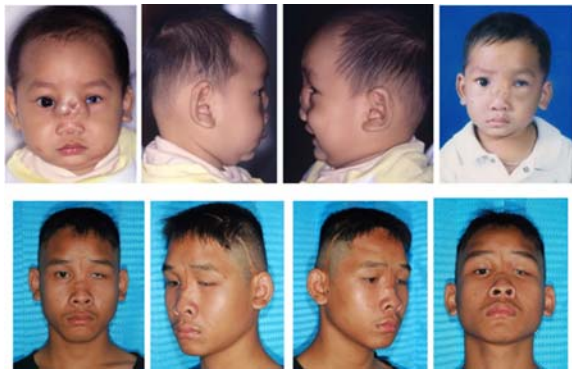

Fig. 9 Patient No. 19 presented with naso-ethmoidal FEEM and left microphthalmos. He was treated by craniofacial reconstruction with medial canthopexy. Follow-up at 16 years of age show a satisfactory facial appearance

of the Tawanchai Craniofacial Center's interdisciplinary care protocol for patients with FEEM (Table 2).

Discussion

Suwanwela and Suwanwela[3] and by Meyer[13] used the bone defects associated with FEEM to classify FEEMs by into naso-frontal, naso-ethmoidal and naso-orbital. The morphology of facial bone defects has variation according to the type. In the naso-frontal type, the defects are at the junction of the frontal and nasal bones. The nasal bones are attached to the inferior margin of the defect and the meningoencephaloceles presents at the root of the nose above the nasal bones.

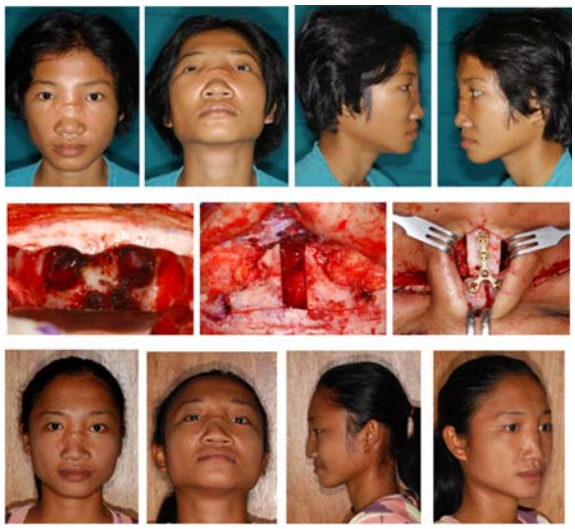

Fig. 10 Patient No. 28 presented with naso-orbital type FEEM. It was treated by craniofacial reconstruction with medial orbital translocation, medial cantopexy and calvarial bone graft. Intraoperative photos show internal skull defects, medial orbital translocation and nasal reconstruction. Follow-up photos, at the age of 24 years, show a satisfactory facial appearance

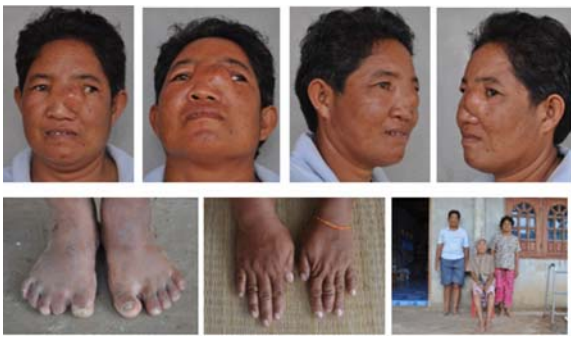

Fig. 11 Patient No. 32 presented at 41 years of age with naso-ethmiodal/-orbital FEEM and anomalies of the hands and feet. The photos were taken in the village wither sister and elderly mother

Fig. 12 Photos of a disable book and the funding from the Tawanchai Foundation for Clefts in helping the patients with FEEM in Thailand to increase accessibility to treatment, improve quality of care and provide educational support

Table 2. Interdisciplinary care protocol at The Tawanchai Craniofacial Center for patients with FEEM

Age	Treatment	Team Members
Prenatal	Prenatal imaging, and counselling	Multidisciplinary
Newborn	Feeding, management of associated anomalies, genetic counselling, providing information Emergency neurosurgery, if indicated	Multidisciplinary, including a plastic surgeon, neurosurgeon
>12 months	Intracranial and extracranial deformities assessed by CT and MRI Craniofacial reconstruction	Plastic surgeon, neurosurgeon, radiologist
4-6 years (preschool)	Evaluation of THAICLEFT 5-year-index, secondary craniofacial reconstruction or reconstruction of lacrimal system (if indicated)	Plastic surgeon, neurosurgeon, psychiatrist and multidisciplinary team
All ages	Treatment of hydrocephalus or other intracranial problems associated with this deformity	Plastic surgeon, oral surgeon and multidisciplinary team
18-21 years (Skeletal maturity, adulthood)	Evaluation of THAICLEFT 19-year-index. Definite nasal reconstruction, including orbital, maxillary, or mandibulary surgery, if indicated	Plastic surgeon, oral surgeon and multidisciplinary team

In the naso-ethmoidal type, the defects are between the nasal bones and the nasal cartilages, either uni- or bi-laterally. If the meningoencephalocele is large, the facial defect extends laterally. In the naso-orbital type, the defects are bilobed, through holes in the medial wall at the junction of the frontal process of the maxilla and the lacrimal bones and meningoencephaloceles can cause proptosis and displacement of the eye. FEEM may also be associated with craniofacial deformity consisting of medial orbital hypertelorism, secondary

trigonocephaly, orbital dystopia, elongation of the face, nasal deformity, and dental malocclusion[14].

The etiology of FEEM includes largely undefined ethnic, genetic and environmental factors. Some research has revealed involvement of paternal age[9,11,15], a multi-factorial genesis[16] and a combination of environmental factors with a genetic predisposition[17-19]. An epidemiological study in Burma reported a higher prevalence of FEEM among the poor, in rural communities and among rice farmers[11] so that vitamin deficiency and fungal agents have been suggested as the main associated factors.

The pathogenesis of meningoencephaloceles has been suggested by the mechanisms occurring between the beginning of the 2nd month and the end of the 3rd month of intrauterine life[20] and an observed localized deficiency in the mesoderm combined with abnormal adhesion of the neuroectoderm (nervous tissue) to the surface ectoderm (epithelial layer) in the midline just after closure of the neural folds[21] during the final part of neural tube formation. Sternberg theorized that a disturbance at the site of the final closure of the rostral neuropore was between the nasal fields and resulted in sustained connections between the neurectoderm and the surface ectoderm, creating a midline mesodermal (skull) defect[22] and resulting in the formation of a FEEM[23]. It was postulated that a disturbance in this separation process could be due to a lack of apoptosis[24-28]. Moreover, the separation process can be disturbed at any of the sites of neural tube closure, leading to the various classified types of FEEM.

The natural history of FEEM varies among patients[29]. It has been reported that a majority of affected children with FEEM are mentally normal[30] and that associated congenital brain anomalies are uncommon[31,32]; however, in the present study some of the patients had delayed development and insufficient intelligence to continue their education (patient No. 4, 12, 19). Associated brain anomalies were also found in some patients and persisted until the long-term follow-up (patient No. 19) as in some previous studies in which some children had neurological complications or associated brain anomalies[33,34]. An extensive evaluation is mandatory in every patient with FEEM, in order to arrive at an accurate diagnosis, thoroughly delineate the malformed anatomy, classify the deformities, evaluate the associated anomalies, make a prognosis, conceive of treatment options, conduct surgical planning, and determine the outcomes to measure throughout the treatment trajectory.

Children with FEEM should have early surgical correction to treat and prevent facial deformities, impairment of binocular vision, increasing size of the FEEM by secondary herniation of intracranial contents, and risk of infection of the central nervous. The treatment of associated brain anomalies (such as hydrocephalus) should be the first priority[9] and subsequently a one-stage reconstructive procedure can be performed[33,35-37]. The objectives of the reconstruction are (a) closure of open skin defects to prevent infection and desiccation of viable brain tissue (b) removal or invagination of nonfunctional extracranial cerebral tissue and (c) water-tight closure of the dura and craniofacial reconstruction with particular emphasis on exact skeletal reconstruction. For closure of dural and bony defects, a transcranial approach is used in most cases of FEEM; however, in cases with a lower level of the cribriform plate, a subcranial approach[34] can be also used. FEEM often coincides with an increased distance between the medial orbital walls or medial orbital hypertelorism so medial orbital translocation is indicated[38,39]. Treatment of patients with FEEM should be undertaken by a multidisciplinary craniofacial team at an early age to avoid further distortion of the facial anatomy during growth[5,9,33,35]. The authors recommend delaying surgical treatment to the age of 5-10 months to minimize complications from anesthesia (*i.e.,* blood loss and hypothermia) and from the operation itself (*i.e.,* CSF leaks or infection)[40]. Other anomalies such as mental retardation, epilepsy, and ocular problems[17] have been reported.

The surgical techniques to reconstruct the deformities caused by FEEM include (a) combined intra- and extra-cranial procedures (bicoronal insicion, naso-frontal bone flap and facial reconstruction)[8,9,30,33,34,40,41], or (b) an extracranial procedure only[17,39,42,43] according to the pattern of the patient's malformation and the availability of neurosurgical expertise.

Nasal reconstruction may be performed first to avoid the long-nose deformity or later during the age of skeletal maturity for definite reconstruction. Cranial bone or costo-chondral grafts with or without fascia or dermis fat graft may be used. The planning for surgical correction of facial skin includes (a) removal of abnormal skin (b) correction of nasal bifidity by a midline scar (c) placing the incisions at the borders of the nasal subunit which may extend laterally and (d) the excision of excess skin and transition flap to correct the position of the medial canthal tendon.

Management by a multidisciplinary craniofacial team with a long-term protocol for holistic care-according to the natural history of the disease and outcome evaluation from birth to skeletal maturity after the age of 18 years-is necessary. Optimum results with less morbidity should be prioritized and delivered with appropriate timing[44]. The measurement of surgical outcomes can be challenging, depending on the severity of the deformity and particularly the extent of associated brain anomalies; thus, long-term assessment is most appropriate.

FEEMs appear to have a more favorable outcome than occipital or parietal meningoencephaloceles: an overall mortality of 7-20% with a favourable developmental outcome has been reported[33]. The type of FEEM and associated brain anomalies may therefore be important when predicting the outcome. The patient with naso-frontal FEEM may have a more predictable outcome as the face is usually in a more normal position, the eyes are unaffected and the intellectual development may be within normal limits. The patient with nasoethmoidal FEEM may have a less predictable outcome and more long-term problems due to the characteristic long face, distorted lacrimal ducts, and variable orbital involvement. The patient with naso-orbital FEEM with only a small volume of dysplastic brain involvement may also have a more predictable outcome: the prognosis and final outcome may be determined by the presence of associated hydrocephalus or brain anomalies[39,45].

The Craniofacial Center and interdisciplinary management is important for the provision of proper and longitudinal care for patients with FEEM. The limitation of healthcare resources in Thailand and other developing countries can result in limitations to access to proper management. Funding from other resources, in our situation, including from the Tawanchai Foundation for Cleft Lip, Cleft Palate and Craniofacial Deformities, may be crucial for increasing accessibility, organizational development, interdisciplinary team management and improving the overall quality of the treatment program. For example, GIS data has provided useful information for treatment-planning for the family of patients with cleft lip/plate and is applicable to other craniofacial deformities, including FEEM. Information includes preparation of outreach programming, referral, continual treatment and partnership with other emerging local cleft centers[46].

Conclusion

The understanding of pathology and its classification, and having a competent plastic surgery and neurosurgery team are important for craniofacial reconstruction and achievement of an optimum outcome for patients with FEEM[47]. The Craniofacial Center with its interdisciplinary management provides proper early and longitudinal care, with ongoing psychological interventions for the patient and family as well as providing for functional needs. Since the measurement of surgical outcomes is challenging-dependent on the severity of the deformity and the extent of associated brain anomalies-very long-term treatment until the age of complete skeletal maturity is needed. Moreover, adequate funding from other resources, including foundations, is needed to ensure accessibility for patients, enable organizational development and improve the quality of treatment programing.

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Potential conflicts of interest

None.

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ฟรอนโตเอตมอยดอล เมนิงโกเอนเซฟาโลซีล: ความท้าทายและการดูแลแบบบูรณาการระยะยาวของศูนย์ตะวันฉาย

บวรศิลป์ เชาวน์ชื่น, ไชยวิทย์ ธนไพศาล, ปรารถนา เชาวน์ชื่น, พิชเยนทร์ ดวงทองพล

ภูมิหลัง: *ความท้าทายของการดูแลผู้ป่วยฟรอนโตเอตมอยดอล เมนิงโกเอนเซฟาโลซีล (โรคงวงช้าง) คือ การจำแนกชนิด การประเมินความพิการ การเสริมสร้างศีรษะและใบหน้า และการดูแลระยะยาว*

วัตถุประสงค์: *เพื่อนำเสนอประสบการณ์ของศูนย์ความพิการศีรษะและใบหน้าตะวันฉายในด้านการดูแลแบบบูรณาการระยะยาวในผู้ป่วยโรคงวงช้างในประเทศไทย*

วัสดุและวิธีการ: *การศึกษาเป็นการทบทวนบันทึกทางการแพทย์ของผู้ป่วยโรคงวงช้าง จำนวน 32 ราย ที่ได้รับการรักษาโดยผู้นิพนธ์ในศูนย์ตะวันฉาย โรงพยาบาลศรีนครินทร์ ซึ่งเป็นศูนย์กลางการส่งต่อผู้ป่วยของภาคตะวันออกเฉียงเหนือในระหว่างปี พ.ศ. 2536-2554*

ผลการศึกษา: *ระบบภูมิสารสนเทศได้ถูกนำมาใช้ เพื่อวิเคราะห์อุบัติการณ์และระบบการส่งต่อผู้ป่วยมายังศูนย์การดูแล ผู้ป่วยส่วนใหญ่จำแนกเป็นชนิด เอโซเอตมอยด์ (12 ราย) และเนโซเอตมอยด์ร่วมกับเนโซออร์บิตอล (8 ราย) การผ่าตัดรักษาประกอบด้วยการเสริมสร้างศีรษะและใบหน้าร่วมกับการยึดตรึงเอ็นหัวตา การเลื่อนตำแหน่งของกระบอกตา การซ่อมแซมภายนอก และการเสริมสร้างจมูก ผู้ป่วยส่วนใหญ่มีผลลัพธ์ด้านภาพลักษณ์ของใบหน้าที่ดีและเป็นที่พึงพอใจ และมีการสร้างแนวทางการดูแลแบบสหวิทยาการของศูนย์ตะวันฉาย*

สรุป: *ศูนย์การดูแลศีรษะและใบหน้าที่ประกอบด้วยการดูแลแบบสหวิทยาการมีความจำเป็นในการดูแลผู้ป่วยงวงช้างทั้งในระยะแรกและระยะต่อเนื่อง และต่อการได้รับผลการรักษาที่เหมาะสม ผลลัพธ์ของการผ่าตัดผู้ป่วยโรคงวงช้างขึ้นกับความรุนแรงและชนิดของความพิการ ความพิการร่วมทางสมอง และควรมีการประเมินเมื่อการเจริญเติบโตของกระดูกใบหน้าเกิดขึ้นโดยสมบูรณ์แล้ว กองทุนจากการจัดตั้งมูลนิธิจะมีส่วนช่วยในการเข้าถึง การรักษาเพิ่มขึ้น สนับสนุนการจัดการของศูนย์การดูแลผู้ป่วย และคุณภาพของการรักษาให้ดียิ่งขึ้น*