Safe and sustainable: the extracranial approach toward frontoethmoidal meningoencephalocele repair

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OBJECTIVE Although rare, frontoethmoidal meningoencephaloceles continue to pose a challenge to neurosurgeons and plastic reconstructive surgeons. Especially when faced with limited infrastructure and resources, establishing reliable and safe surgical techniques is of paramount importance. The authors present a case series in order to evaluate a previously proposed concise approach for meningoencephalocele repair, with a focus on sustainability of internationally driven surgical efforts.

METHODS Between 2001 and 2016, a total of 246 patients with frontoethmoidal meningoencephaloceles were treated using a 1-stage extracranial approach by a single surgeon in the Department of Neurosurgery of the Yangon General Hospital in Yangon, Myanmar, initially assisted by European surgeons. Outcomes and complications were evaluated.

RESULTS A total of 246 patients (138 male and 108 female) were treated. Their ages ranged from 75 days to 32 years (median 8 years). The duration of follow-up ranged between 4 weeks and 16 years (median 4 months). Eighteen patients (7.3%) showed signs of increased intracranial pressure postoperatively, and early CSF rhinorrhea was observed in 27 patients (11%), with 5 (2%) of them requiring operative dural repair. In 8 patients, a decompressive lumbar puncture was performed. There were 8 postoperative deaths (3.3%) due to meningitis. In 15 patients (6.1%), recurrent herniation of brain tissue was observed; this herniation led to blindness in 1 case. The remaining patients all showed good to very good aesthetic and functional results.

CONCLUSIONS A minimally invasive, purely extracranial approach to frontoethmoidal meningoencephalocele repair may serve well, especially in middle- and low-income countries. This case series points out how the frequently critiqued lack of sustainability in the field of humanitarian surgical missions, as well as the often-cited missing aftercare and dependence on foreign supporters, can be circumvented by meticulous training of local surgeons.

https://thejns.org/doi/abs/10.3171/2017.5.PEDS1762

KEY WORDS frontoethmoidal meningoencephalocele; neural tube defects; humanitarian mission; sustainability; NGO; surgical mission; meningocele; craniofacial
the frontal and ethmoidal bones, usually covered by skin. While 75% of all meningoencephaloceles are located oc-
cipitally and 10%—12% parietally, the frontoethmoidal
defect occurs in 13%—15% of cases.1,16 Along with anen-
cephaly and spina bifida, this malformation is one of the 3
most common neural tube defects. While its etiology and
pathogenesis are still not fully understood, genetic and en-
vironmental factors, including maternal malnutrition dur-
ing pregnancy and teratogenic agents, have been discussed
as contributing factors.30,36,39,41 Frontoethmoidal meningo-
encephaloceles can be classified as nasofrontal, nasoeth-
moidal, nasoorbital, and combined types, depending on the
intra- to extracranial extent of the herniated tissues. While
the frontoethmoidal form is extremely rare in the Western
world (incidence 1 in 35,000), it affects children in poor
rural areas of lower income countries more often14,37 and
is relatively common in Southeast Asia, with an incidence
ranging from 1 in 3500 to 1 in 6000 live births.25 There is
a wide spectrum of clinical manifestations, ranging from
symptom free to occurrence of hydrocephalus, spastic pal-
sies, seizures, or intellectual disability. The malformation
is associated with craniofacial disfigurement, functional
impairment, and risk of infection of the central nervous
system. Affected patients may also suffer from telecan-
thus,3,28 amblyopia, and epiphora.8 The typical history of
the disease is progressive, with continuous herniation of
intracranial contents, decreasing life expectancy drastically.
Prognosis and treatment of affected patients depend on the
location, size, and contents of the herniation,13 but in case
of progressive externalization of tissue, a definitive surgi-
cal repair should be performed in infancy.7 A combined
intra- and extracranial approach, performed within the 1st
year of life in a multidisciplinary setting, remains the gold
standard for such correction.5,14,15,19,29

A complete extirpation of the herniated tissue is neces-
sary to prevent further infections and deformities, or even
episodes of meningitis with cerebral involvement. Un-
fortunately, such highly complex intracranial procedures
cannot be performed safely everywhere in the world, and
especially not in low-income countries with impaired
health care systems and a lack of appropriate service fa-
cilities and medical personnel.

In 2008, Holm et al.16 described a series of 52 Bur-
mese patients with frontoethmoidal meningoencephalo-
celes that were operated on by means of a purely extra-
cranial approach. The surgeries were performed between
2001 and 2005 in cooperation with the local Department
of Neurosurgery of Yangon General Hospital in Myan-
mar, and were made possible by personnel and finan-
cial support from Interplast Germany, an international
nongovernmental organization (NGO) without political
or religious affiliations. The chief of the Yangon Gen-
eral Hospital’s Department of Neurosurgery (M.T.) was
trained in the new technique during a visit to Austria (see
Illustrative Case 1, below) and during 3 visits to Myanmar
by the Interplast team, which included 5 of the authors
(H.A., W.M., C.H.M., H.S., and P.H.) as well as German
and Austrian support staff. The new technique avoids the
usually necessary additional cranial opening to access the
anterior fossa. Despite the difficulties regarding long-term
follow-up, the authors concluded that this surgical ap-
proach was associated with low early postoperative com-
lications and morbidity rates, while being simple, safe,
and cheap.16 Since 2005, the project has been continued
with only marginal financial support from Interplast. In
2012, Swiss Neurosurgeons International joined forces to
boost the educational program needed to treat the popula-
tion of over 50 million people living in Myanmar. Thanks
to yearly educational visits to Myanmar and numerous fel-
lowships in Switzerland for young Burmese specialists to
deepen their skills and knowledge, the number of surgeons
able to treat frontoethmoidal meningoencephaloceles has
increased exponentially, and nationwide coverage will be
reached before the end of this decade.

The goal of the present study was to evaluate whether
the proposed treatment approach was in fact adopted by
the local medical community, and whether it continued to
prove to be a safe procedure with good outcomes in the
long run. This is one of the largest series evaluating pa-
tients with frontoethmoidal meningoencephaloceles cor-
corrected by a purely extracranial approach. Furthermore,
this project displays an example of how to overcome the
often-discussed potential lack of sustainability in the field
of medical humanitarian work, allowing the successful
treatment of otherwise often-neglected patients.

Methods

Between 2001 and 2016, a total of 246 patients with
frontoethmoidal meningoencephaloceles were treated us-
ing a 1-stage extracranial approach by a single surgeon
in the Department of Neurosurgery of the Yangon Gen-
eral Hospital in Yangon, Myanmar. To allow demonstra-
tion of surgical management, 1 patient, accompanied by
the senior local surgeon, was taken to Austria to undergo
surgery. In the past 20 years, the Munich section of Inter-
plast Germany has been performing 3–5 surgical missions
per year in Myanmar and has developed a well-organized
and widespread medical infrastructural network in the
country. With the help of local trustees, affected patients
were screened and registered in the different regions and
brought to the former capital city of Yangon for surgery.
After the patients underwent preoperative clinical and ra-
diographic examination (skull radiographs and CT scans),
their lesions were classified using the frontoethmoidal en-
cephalomeningocele classification according to Rojvachi-
ranonda et al.12

As previously described in detail,16 the procedure itself
e entails an extracranial approach through a vertical mid-
line nasal incision, dissection of the meningocele up to the
neck of the sac, amputation of the sac and herniated brain
up to the level of the inner table of the skull, repair of the
dura, bone defect closure with split-thickness calvarial
bone or cartilage graft, nasal bone reconstruction, and me-
dial canthoplasty. Local rearrangement flaps were used as
needed to achieve closure.

All patients received prophylactic perioperative antibi-
otic treatment (ampicillin) and were kept in the hospital
for 1 week after surgery. Patients were told to stay near the
hospital for another 2–4 weeks before returning to their
homes, which are frequently in rural areas of Myanmar. If
postoperative cerebrospinal fluid (CSF) leakage occurred,
bed rest was ordered and prolonged intravenous antibiotic
treatment was administered. In cases of clinical signs of elevated brain pressure, a decompressive lumbar puncture was performed. If spontaneous cessation of liquorrhea did not occur within 2 weeks after surgery, dural repair was performed by the local neurosurgical team.

Early postoperative complications and outcomes were analyzed, and the aesthetic results were evaluated by taking into consideration anatomical features, including a long nose deformity, telecanthus, and the position of the medial canthi.

Results

A total of 246 patients with a median age of 8 years (range 75 days to 32 years) were treated using the described approach. Nasofrontal meningoencephalocele was diagnosed in 105 cases (42.7%), nasoethmoidal in 65 (26.4%), nasoorbital in 5 (2%), and combined nasoethmoidal/nasoorbital meningoencephalocele in 71 (28.9%); 71 meningoceles (combined nasoethmoidal/nasoorbital) were bilateral. Bone defect closures with tabula externa grafts were necessary in 64 cases (26%). The duration of postoperative follow-up ranged from 4 weeks to 16 years. Bone defect closure with split-thickness calvarial bone grafts were necessary in 64 cases (26%). The duration of postoperative follow-up ranged from 4 weeks to 16 years (median 4 months).

Eighteen patients (7.3%) showed signs of increased intracranial pressure postoperatively, which was the indication for performing the placement of a ventriculoperitoneal shunt. Early CSF rhinorrhea was observed in 27 patients (11%) postoperatively, but only 5 (2%) of them required operative dural repair. In the other 22 patients, liquorrhea resolved spontaneously under conservative therapy. In 8 patients, a decompressive lumbar puncture was performed. There were 8 postoperative deaths due to meningitis (mortality rate 3.3%). In 15 patients (6.1%), recurrent herniation of brain tissue was observed, and this unfortunately led to blindness in 1 case. Patient demographics and postoperative outcomes are summarized in Tables 1 and 2.

Illustrative Cases

Case 1

This 4-year-old boy presented to the neurosurgical clinic in Yangon suffering from a large frontonasal meningoencephalocele (Fig. 1A). An interdisciplinary treatment plan was established and the patient was flown to the University Hospital Innsbruck, Austria, in the company of the senior local Myanmar surgeon so that he could participate in the operation and observe the surgical procedure and perioperative care under optimal conditions (Fig. 1B–F). Follow-up 3 and 16 years postoperatively showed an acceptable aesthetic result and full functionality (3-year follow-up photograph, Fig. 1G). The patient is fully integrated into society and creates artwork for family income (Fig. 1H).

Case 2

This 5-year-old boy presented to the clinic in Yangon with a bilateral nasoethmoidal meningoencephalocele. The surgical approach included exposure of the sac and resection of the herniated tissue. After dural repair, the defect was covered with parietal tabula externa bone grafts and closed with double advancement skin flaps. At 16 years’ follow-up, the patient is socially integrated with his family, participating in traditional ox-cart racing, and has good functional and aesthetic results (Fig. 2).

Discussion

Several surgical procedures have been described to treat frontoethmoidal meningoencephaloceles, mostly based on the classic technique as described by Tessier, which includes a combined extra- and intracranial approach by means of a bicoronal incision, craniotomy, meningoencephalocele resection, creation of nasofrontal bone flaps, and facial reconstruction.7,9,11,21–23

Surgery, performed in a neurosurgical center as soon as possible to avoid progression of the deformity, is the gold standard for treatment in most Western countries.2,11

Given the high complexity of the surgery as well as the nascent state of the health care systems and lack of advanced medical infrastructure, expedited treatment of the disease might unfortunately not always be feasible in many Asian countries, such as Burma,11,16 Thailand,2,3,10,26,38,39 Cambodia,9,12,33,42 Malaysia,1 Indonesia,17,35 and Papua New Guinea.18 However, it has been shown that treating affected

<table>
<thead>
<tr>
<th>Complication</th>
<th>No. of Patients (%)</th>
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<tbody>
<tr>
<td>CSF leak</td>
<td>27 (11)</td>
</tr>
<tr>
<td>Meningitis</td>
<td>8 (3.3)</td>
</tr>
<tr>
<td>Ventriculoperitoneal shunt</td>
<td>18 (7.3)</td>
</tr>
<tr>
<td>Recurrence</td>
<td>15 (6.1)</td>
</tr>
<tr>
<td>Mortality</td>
<td>8 (3.3)</td>
</tr>
</tbody>
</table>
FG. 1. Illustrative Case 1. A: Preoperative photograph of the patient at the age of 4 years showing a large frontonasal meningoencephalocele. B–F: Intraoperative photographs. The herniated tissue was exposed through a vertical midline incision (B). After resection of the herniated tissue the bone defect was visible (C). The defect was closed with bone grafts after correction of medial hypertelorism by osteotomy and medialization of the medial orbital walls (D). The patient’s nose was reconstructed with bone and cartilage grafts (E). Skin closure was achieved with local double-advancement transposition flaps (F). G: Photograph obtained at 3-year follow-up showing the patient with improved aesthetics and a good functional result, including full vision in both eyes. H: Photograph obtained 16 years after surgery showing the patient with an example of the artwork that he creates to provide for income for his family. Figure is available in color online only.

FIG. 2. Illustrative Case 2. A: Preoperative photograph of the patient at the age of 5 years showing a bilateral nasoethmoidal meningoencephalocele. B: Intraoperative photograph showing the bone defect after resection of the herniated tissue through a nasal incision. The defect was subsequently repaired using bone grafts. C: Early postoperative photograph showing an acceptable cosmetic result. D: Photograph obtained 16 years after surgery showing the patient participating in traditional ox-cart racing. Figure is available in color online only.
patients under such circumstances can be made possible through the work of joint ventures between local surgeons and NGOs. To do this effectively, highly motivated local surgeons remain an indispensable resource.\textsuperscript{23,43} These need to be provided with continuous development opportunities, including training with visiting professionals as well as rotations at centers of excellence, with international staff and volunteers ultimately only serving in supporting roles.\textsuperscript{4}

Regarding frontoethmoidal meningoencephalocele repair, Oucheng et al.\textsuperscript{27} published a series of 200 cases that were treated by a combined neurosurgical-craniofacial approach in Cambodia and showed that good results can be achieved as long as certain surgical and perioperative principles—including clear indications, proper surgical technique, and postoperative care—are adhered to. However, they used a combined intra- and extracranial approach, which is more complex than a solely extracranial procedure. A few years later, Rifi et al.\textsuperscript{31} analyzed a series of 60 cases in Africa and were also able to show the feasibility of a 1-step combined procedure, elucidating the importance of repair at an early age as well as the interdisciplinary approach.

In 2008, Holm et al.\textsuperscript{16} introduced the purely extracranial approach for the correction of frontoethmoidal meningoencephaloceles and demonstrated the feasibility and safety of performing the surgery in middle- and low-income countries. While they initially encountered a relatively high incidence of postoperative CSF leaks, the technique has evolved, and leakage rates could be reduced to 11\% in the current series, which is comparable to reports by other groups (Table 3).\textsuperscript{11,20,24,25,27,31} This improvement might be explained by the learning curve as well as the improved availability of adequate instrumentarium and medication in Myanmar. In most patients with a postoperative CSF leak, the leakage ceased spontaneously within 2 weeks after the operation, and craniotomy and secondary dural repair became necessary in only 5 patients (2\% of cases). In 8 cases (3.3\%), ascending meningitis led to postoperative deaths, which might have been due, at least in part, to the suboptimal condition of perioperative hygiene in a developing country hospital. Given the constraints of the medical system, the mortality rate might be difficult to reduce much further; nonetheless, the perioperative management might be improved. Only ampicillin was used for preoperative antibiotic prophylaxis. It was the drug of choice due to its availability, broad-spectrum coverage, and low cost. However, adding sulbactam to extend coverage presents a feasible option for reducing complications. Furthermore, earlier intervention in cases of CSF leaks, which is a widely discussed topic in the literature, will be given consideration in the future. Overall, when comparing the results of this study to reports in the published literature, we found that similar incidences of early postoperative complications were observed by other groups (Table 4).\textsuperscript{2,3,12,20,21,24,27,28,31}

TABLE 3. Incidence of postoperative CSF leak associated with different operative techniques

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Op Technique</th>
<th>No. of Patients w/ CSF Leak (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mahapatra et al., 1994</td>
<td>2-step procedure in 4 cases; 1-step in 26 cases; combined intra- &amp; extracranial approach; no details of technique available</td>
<td>6/30 (20.0)</td>
</tr>
<tr>
<td>Fuente del Campo et al., 1989</td>
<td>Combined intra- &amp; extracranial approach; specific approach in respect to pathology; use of bone graft</td>
<td>2/63 (3.2)</td>
</tr>
<tr>
<td>Mahatumarat et al., 2003</td>
<td>Combined intra- &amp; extracranial approach; “Chula technique”</td>
<td>3/108 (2.8)</td>
</tr>
<tr>
<td>Kumar et al., 2009</td>
<td>Combined intra- &amp; extracranial approach; “HULA technique”</td>
<td>0/12 (0)</td>
</tr>
<tr>
<td>Oucheng et al., 2010</td>
<td>Combined intra- &amp; extracranial approach</td>
<td>24/200 (12)</td>
</tr>
<tr>
<td>Present study</td>
<td>Purely extracranial approach</td>
<td>27/246 (11.0)</td>
</tr>
</tbody>
</table>

TABLE 4. Early postoperative complications after the correction of frontoethmoidal meningoencephaloceles

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Rhinorrhea</th>
<th>Meningitis</th>
<th>Hydrocephalus</th>
<th>Death</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mahapatra et al., 1994</td>
<td>6/30 (20)</td>
<td>1/30 (3.3)</td>
<td>0/30 (0)</td>
<td>0/30 (0)</td>
</tr>
<tr>
<td>Boonvisut et al., 2001</td>
<td>3/145 (2.1)</td>
<td>0/145 (0)</td>
<td>1/145 (0.7)</td>
<td>0/145 (0)</td>
</tr>
<tr>
<td>Pinzer et al., 2006</td>
<td>1/30 (3.3)</td>
<td>0/30 (0)</td>
<td>0/30 (0)</td>
<td>1/30 (3.3)</td>
</tr>
<tr>
<td>Leelanukrom et al., 2007</td>
<td>3/102 (2.9)</td>
<td>1/102 (1.0)</td>
<td>4/102 (3.9)</td>
<td>0/102 (0)</td>
</tr>
<tr>
<td>Gollogly et al., 2008</td>
<td>10/128 (7.8)</td>
<td>3/128 (2.3)</td>
<td>0/128 (0)</td>
<td>4/128 (3.1)</td>
</tr>
<tr>
<td>Kumar et al., 2009</td>
<td>0/12 (0)</td>
<td>0/12 (0)</td>
<td>0/12 (0)</td>
<td>0/12 (0)</td>
</tr>
<tr>
<td>Oucheng et al., 2010</td>
<td>24/200 (12)</td>
<td>6/200 (3)</td>
<td>4/200 (8)</td>
<td>3/200 (6)</td>
</tr>
<tr>
<td>Rifi et al., 2015</td>
<td>4/60 (6.7)</td>
<td>0/60 (0)</td>
<td>2/60 (3.3)</td>
<td>1/60 (1.7)</td>
</tr>
<tr>
<td>Present study</td>
<td>27/246 (11)</td>
<td>8/246 (3.3)</td>
<td>18/246 (7.3)</td>
<td>8/246 (3.3)</td>
</tr>
</tbody>
</table>

Values presented as the number of patients (\%).
living in remote areas without any established infrastructures makes it very difficult to locate patients and examine them again. In this series also, only a few patients had appropriate follow-up beyond 4–6 weeks. Overall, 15 patients came back to Yangon with recurrent herniation, and they subsequently underwent revision surgery. It is difficult to say whether the purely extracranial approach was associated with an overall higher incidence of recurrence, as has been debated by previous authors. However, the experience of the local surgeons shows that patients who encounter any problems or complications tend to come back to the place where they underwent surgery. One can thus speculate that patients who were doing well during the first 6 weeks after surgery and did not come back continued to do well, but unfortunately there is no guarantee of this, and this uncertainty regarding longer-term outcomes is a major limiting factor of the presented series.

The described approach does show, however, that even complex surgical tasks can be taught to motivated local surgeons who may subsequently continue their efforts and provide safe and efficient care to their people with only minimal additional support.

Conclusions

Frontoethmoidal meningoencephalocele repair remains a challenging surgical task. A minimally invasive purely extracranial approach may provide a useful option, especially in middle- and low-income countries with inadequate infrastructure and monetary funds. The frequently criticized lack of sustainability in the field of humanitarian surgical missions, as well as the often-cited missing aftercare and dependence on foreign supporters, could be circumvented by meticulous teaching of local surgeons.

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**Disclosures**
The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

**Author Contributions**
Conception and design: Heidekrueger, Broer. Acquisition of data: Heidekrueger, Thu, Schucht, K Aung, Mg Ag, Thu Soe Myint, Juran. Analysis and interpretation of data: Heidekrueger, Juran, T Aung. Drafting the article: Heidekrueger, Ehrl. Critically revising the article: Thu, Mühlbauer, Holm-Mühlbauer, Schucht, Anderl. Reviewed submitted version of manuscript: Mühlbauer, Holm-Mühlbauer, Schucht, Anderl, Schoeneich, K Aung, Mg Ag, Thu Soe Myint, Juran, T Aung, Ehrl, Ninkovic. Approved the final version of the manuscript on behalf of all authors: Heidekrueger. Statistical analysis: Heidekrueger. Administrative/technical/material support: Thu, Mühlbauer, Holm-Mühlbauer, Schucht, Anderl, Schoeneich, K Aung, Mg Ag, Thu Soe Myint, Ninkovic. Study supervision: Broer.

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