ENCEPHALOCELES are developmental anomalies associated with a herniation of neural tissue through a bone defect in the skull. The occurrence rate is approximately 1 in 3000–5000 births. These entities are classified by the site of the bone defect and herniation, and occipital encephaloceles are the most common. Basal encephaloceles account for 5% of all encephaloceles and are subdivided into 4 types: transethmoidal, sphenoorbital, sphenethmoidal, and transsphenoidal. Of these, a TSEC is the rarest form and accounts for 1 in 20 (1 in 700,000 births). A sphenethmoidal encephalocele is often interpreted as an extensive form of TSEC from the clinical and pathological viewpoint. Because the bone defect is deeply located, the size of the defect is large in some cases, and the encephalocele can contain vital structures such as pituitary gland or optic chiasm, the surgical treatment of TSEC, which aims to expose, reduce, and repair the encephalocele, is still challenging. We report our experience with 7 patients surgically treated for TSEC and try to elucidate how to select the optimal surgical approach.
Methods

We reviewed 7 consecutive patients who underwent operation for TSEC at the National Center for Child Health and Development in Tokyo over the period between March 2004 and February 2012. Retrospective analysis for patient information including age, sex, presenting symptoms, surgical treatment, and clinical outcomes was performed using medical charts. Preoperative MRI and CT studies were performed in all patients to delineate the skull bone defect, to find whether the encephalocele contained vital structures such as pituitary gland or optic chiasm, or to detect other intracranial abnormalities such as corpus callosum agenesis.

Surgical repair of the encephalocele was performed by a team of neurosurgeons and plastic surgeons. Lumbar drainage was placed to remove the excess CSF during the operation and postoperatively.

For the transpalatal approach, a Dingman self-retaining retractor was placed on the patient’s mouth. The soft palate was cut at the midline while preserving the uvula. The uvula and palatal mucosal flaps were retracted laterally. The exposed hard palate was removed using rongeurs to visualize the encephalocele in the pharynx. Those procedures were not required in cases with a cleft palate. The mucosal dissection was performed using bipolar cautery to expose the bone defect and the encephalocele sac. Care was taken not to open the dura mater. If there was dehiscence, it was coagulated or sutured using 8-0 Prolene. The sac and its contents were preserved in all cases. After complete dissection of the sac from the edge of the bone defect, the sac was reduced upward into the defect. The bone defect was repaired using palatal bone, cranial bone, or titanium mesh, and covered with oxidated regenerated cellulose and fibrin glue. The palatal mucosa was closed using 3-0 Vicryl.

As for the transcranial approach, briefly, a coronal skin incision was made and a bifrontal craniotomy was performed leaving a pericranial flap. A medial supraorbital bar was removed. The frontal dura was dissected from the basal bone and the anterior edge of the bone defect of the encephalocele was reached. The sac was dissected from the wall of the bone defect and reduced intracranially. The bone defect was repaired using cranial bone, rib bone, or titanium mesh. The repaired defect was covered using a pericranial flap and sealed with fibrin glue.

A combined approach was used for TSEC with an extensive sphenoid bone defect. The encephalocele sac was dissected from the posterior edge of the bone defect via a transpalatal approach, and then from the anterior edge of the bone defect via a transcranial approach.

Postoperatively, CSF was typically drained at 5–10 ml/hr for 4–5 days, and then lumbar drainage was clamped and removed. A neuroimaging study was performed in all the patients at 2 weeks, 6 months, and 1 year after the surgery. After that, a neuroimaging study was done on a yearly basis. Patients were followed up periodically with outpatient visits.

Results

Between March 2004 and February 2012, 7 patients with TSEC were surgically treated at our institution. These patients’ characteristics are shown in Table 1. Three were male and 4 were female. The mean age was 23.9 months (range 2–69 months). Four patients presented with obstructive dyspnea, 2 with recurrent meningitis, and 1 with failure to thrive. The TSEC was found through screening of anomalies in 2 patients, including cleft lip and palate, orbital hypoplasia, or hydrocephalus. Endocrine studies revealed adrenocorticotropic hormone deficiency in 3 patients, thyroid-stimulating hormone deficiency in 3, and growth hormone deficiency in 1. Visual disturbance was noted in 3 patients with orbital hypoplasia and in 2 with morning glory syndrome.

The CT scans detected the skull bone defects, which were located within the sphenoid bone in 4 patients and extended anteriorly to the ethmoid bone in 3 (Fig. 1). The size of the sphenoid bone defect (anteroposterior and transverse diameter) ranged from 0.4 × 0.4 cm to 1.1 × 1.3 cm, and the size of the sphenoid bone defect ranged from 2.2 × 1.9 cm to 3.6 × 2.8 cm. The MRI studies revealed the encephalocele sac containing pituitary gland in 6 patients and optic chiasm in 2 patients (Fig. 2). The MRI studies also showed other intracranial anomalies, including corpus callosum agenesis in 3 patients, Chiari malformation Type I in 1, and arachnoid cyst in 1.

In 3 patients with a bone defect limited within the sphenoid bone (Cases 3, 4, and 7), it was possible to dissect the encephalocele sac circumferentially from the edge of the bone defect (Fig. 3), and to reduce the sac via a transpalatal approach. In the patient in Case 1, who had a relatively large sphenoid bone defect (1.1 × 1.3 cm) compared with the other 3 patients, dissection of the sac was performed almost circumferentially (except for the anterior tip) via the transpalatal approach. The sac was successfully reduced. The postoperative MRI studies showed reduction of the encephalocele above the level of the epipharynx in all 4 patients (Fig. 4B). Obstructive dyspnea and meningitis resolved. These patients have been stable without recurrence (follow-up ranged from 2 to 93 months).

On the other hand, in 2 patients with an extensive sphenoid bone defect (Cases 2 and 5), it was difficult to dissect the sac entirely from the bone defect via the transpalatal approach. The anterior one-third of the sac was left undetached from the bone edge at the first surgery. After a few weeks, the displacement of the cra-

![Fig. 1. 3D CT scans demonstrating a sphenoid bone defect (arrow, left) and an extensive sphenoid bone defect (arrowheads, right).](image-url)
TABLE 1: Clinical findings in 7 patients with TSEC who underwent surgical correction*

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (mos), Sex</th>
<th>Signs &amp; Sxs</th>
<th>Associated Anomalies</th>
<th>Endocrine Dysfunction</th>
<th>Skull Bone Defect Location (AP &amp; transverse diameter)</th>
<th>Involvement of Vital Structures</th>
<th>Surgical Approach</th>
<th>FU (mos)</th>
<th>Recurr</th>
<th>FU Status</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>2, M</td>
<td>obstructive dyspnea, failure to thrive</td>
<td>coloboma of optic nerve (morning glory syndrome)</td>
<td>none</td>
<td>sphenoid (1.1 x 1.3 cm)</td>
<td>pituitary gland</td>
<td>transpalatal</td>
<td>93</td>
<td>no</td>
<td>Sxs resolved, GH therapy started 2 yrs postop</td>
</tr>
<tr>
<td>2</td>
<td>2, F</td>
<td>obstructive dyspnea</td>
<td>coloboma of optic nerve (morning glory syndrome), cleft lip &amp; palate, CC agenesis</td>
<td>none</td>
<td>sphenethmoid (2.2 x 1.9 cm)</td>
<td>pituitary gland, optic chiasm</td>
<td>transpalatal or transcranial</td>
<td>90</td>
<td>yes</td>
<td>underwent several ops during 4 yrs, GH therapy started 1 yr after last op</td>
</tr>
<tr>
<td>3</td>
<td>36, F</td>
<td>meningitis, rhinorrhea</td>
<td>rt orbital hypoplasia</td>
<td>none</td>
<td>sphenoid (0.7 x 0.6 cm)</td>
<td>none</td>
<td>transpalatal</td>
<td>81</td>
<td>no</td>
<td>stable w/ Sxs resolved</td>
</tr>
<tr>
<td>4</td>
<td>2, F</td>
<td>incidentally found when screening for HC</td>
<td>orbital hypoplasia, CM-I</td>
<td>ACTH/TSH deficiency</td>
<td>sphenoid (0.4 x 0.4 cm)</td>
<td>pituitary gland</td>
<td>transpalatal</td>
<td>69</td>
<td>no</td>
<td>stable</td>
</tr>
<tr>
<td>5</td>
<td>13, M</td>
<td>obstructive dyspnea, cleft lip &amp; palate, CC agenesis</td>
<td>cleft lip &amp; palate, CC agenesis</td>
<td>ACTH/TSH deficiency</td>
<td>sphenoeothmoid (2.9 x 2.1 cm)</td>
<td>pituitary gland, optic chiasm</td>
<td>transpalatal or transcranial</td>
<td>54</td>
<td>yes</td>
<td>underwent several ops during 1.7 yrs</td>
</tr>
<tr>
<td>6</td>
<td>69, M</td>
<td>found when screening for cleft lip &amp; palate</td>
<td>cleft lip &amp; palate, rt orbital hypoplasia, CC agenesis, arachnoid cyst</td>
<td>ACTH/TSH/GH deficiency</td>
<td>sphenoeothmoid (3.6 x 2.8 cm)</td>
<td>pituitary gland</td>
<td>combined (transpalatal &amp; transcranial)</td>
<td>43</td>
<td>no</td>
<td>stable</td>
</tr>
<tr>
<td>7</td>
<td>6, F</td>
<td>obstructive dyspnea, meningitis</td>
<td>none</td>
<td>none</td>
<td>sphenoid (0.4 x 0.6 cm)</td>
<td>pituitary gland</td>
<td>transpalatal</td>
<td>2</td>
<td>no</td>
<td>stable w/ Sxs resolved</td>
</tr>
</tbody>
</table>

* ACTH = adrenocorticotropic hormone; AP = anteroposterior; CC = corpus callosum; CM-I = Chiari malformation Type I; FU = follow-up; GH = growth hormone; HC = hydrocephalus; Recurr = recurrence; Sxs = symptoms; TSH = thyroid-stimulating hormone.
nial bone graft and a prolapse of the encephalocele were detected, causing recurrent meningitis or obstructive dyspnea. Transcranial repair of the encephalocele was added. However, it was difficult to manipulate the posterior third of the encephalocele from the bone edge. These patients experienced several relapses, with a prolapse of the encephalocele due to osteolysis of the bone graft, and underwent transpalatal or transcranial repairs within 4 and 1.7 years, respectively.

One patient with an extensive sphenoethmoidal bone defect was treated via a combined transpalatal and transcranial approach (Case 6). The circumferential dissection of the encephalocele sac from the edge of the bone defect was made posteriorly via the transpalatal approach, and then anteriorly via the transcranial approach. The sac was reduced above the level of the epipharynx (Fig. 4D). A pericranium-pedicled cranial bone graft was inserted to seal the osseous defect via the transcranial approach (Fig. 4E). The patient has been stable without relapse for 3.6 years since the surgery.

In all cases there were no complications during the operation. The encephalocele sac and its contents were preserved in all cases. The mean follow-up was 61.7 months (range 2–93 months). None of the patients experienced neurological deterioration or developed diabetes insipidus. Growth hormone replacement therapy was started in 2 patients several years after the initial operation. There has been no worsening or significant improvement in endocrinological function in the other patients.

Discussion

Encephaloceles are developmental anomalies associated with a herniation of neural tissue through a bone defect in the skull. Of all encephaloceles a TSEC is the rarest form, and its occurrence rate is 1 in 700,000 births.4 Previous studies reported that conservative treatment of TSEC led to progression of symptoms such as visual disturbance, endocrine dysfunction, or recurrent meningitis.2,3,7,12,13 To resolve or prevent those symptoms, surgical correction was generally recommended.5,6,13

However, because the bone defect is deeply located, the defect extends from the sphenoid to the ethmoid bone in some cases, and the encephalocele can contain vital structures such as pituitary gland or optic chiasm, the surgical treatment of TSEC remains challenging and the optimal approach has not yet been determined.

In previous series of patients treated via the transcranial approach, the mortality rate approached 50% and the morbidity rate 70%.2,15 These unsatisfactory results are considered to be partly due to insufficient dissection of the encephalocele at the posterior part of the bone defect, and to resection of the encephalocele and its contents. When the dissection of the encephalocele sac is insufficient and not circumferentially done around the bone defect, it is difficult to reduce the sac completely. In such a case, to elevate the sac above the confines of the epipharynx, resection of the encephalocele might be necessary, which can cause lethal hypothalamic damage. Also, being forced to reduce the sac without circumferential dissection can cause damage to its contents, which may lead to visual or hypothalamic dysfunction.3,15

Insufficient dissection makes it difficult to seal the osseous defect, which may lead to a recurrent prolapse of the sac as described in our series. Thus, circumferential dissection of the sac from the bone defect and definition of the bone edge is considered to be important for reducing the sac and its contents safely, and sealing the osseous defect appropriately, which eventually prevent a recurrence with a prolapse of the sac.

For this purpose, in a case of TSEC with a bone defect limited within the sphenoid bone, the transpalatal approach is sufficient to expose it, as shown in previous series as well as in ours.2,13 The transcranial approach is not considered to be suitable for this type of TSEC because the sac of the TSEC lies deep and low, and exces-
Intrusive retraction of the frontal lobe is needed to dissect the posterior part of the sac, as previously mentioned.13,15

On the other hand, in a case of TSEC with an extensive sphenoethmoidal bone defect, dissection of the anterior part of the sac is difficult to achieve via an extracranial transpalatal approach alone, which necessitates that it be combined with the intracranial transfrontal approach as in our series. Kohan et al.6 also reported successful treatment of 4 patients with TSEC by using combined intracranial and transpalatal encephalocele correction. Although not detailed in all 4 cases, an illustrative 3D CT scan showed an extensive sphenoethmoidal bone defect. All 4 cases in their series were associated with cleft palate. In our series all 3 patients with cleft palate had an extensive sphenethmoidal bone defect, and the 4 patients without cleft palate had a relatively small bone defect within the sphenoid bone. The patients with TSEC associated with cleft palate may tend to have a more extensive skull bone defect, and may need a combined approach to correct the encephalocele.

Regarding the lumbar drain, we usually placed it before the incision was made for an encephalocele repair. Intraoperatively it is useful to drain the CSF from the lumbar drain and shrink the sac, which makes dissection easier. Postoperatively draining the CSF facilitates the healing of the dehiscence of the dural sac, if any, and prevents meningitis.

**Conclusions**

The transpalatal and transcranial combined approach is useful for treatment of TSEC with an extensive sphenoethmoidal bone defect, to dissect the sac and expose the bone defect circumferentially. If dissection is not sufficient, relapse with a prolapse of the encephalocele can ensue. The transpalatal approach is sufficient by itself for TSEC with a bone defect limited within the sphenoid bone.

**Disclosure**

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 to the study and manuscript preparation include the following. Conception and design: Ogiwara. Acquisition of data: Ogiwara. Analysis and interpretation of data: Ogiwara. Drafting the article: Ogiwara. Critically revising the article: Morota. Reviewed submitted version of manuscript: Morota. Approved the final version of the manuscript on behalf of both authors: Ogiwara.

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