Transpalatal approach for the extracranial surgical repair of transsphenoidal cephaloceles in children

ERIN M. KENNEDY, M.D., DAVID P. GRUBER, M.D., DAVID A. BILLMIRE, M.D., AND KERRY R. CRONE, M.D.

Departments of Plastic Surgery and Neurosurgery, University of Cincinnati, Children’s Hospital Medical Center, and the Mayfield Clinic, Cincinnati, Ohio

The surgical treatment of transsphenoidal cephaloceles in children is controversial. Reduction and repair via a transcranial approach are associated with high postoperative rates of morbidity, mortality, and hypothalamic dysfunction. In this study, four patients, aged 3 to 35 months at surgery, underwent successful transpalatal repair of two encephaloceles and two meningoceles. Two patients presented with nasal obstruction in infancy, one presented with unexplained meningitis, and in one patient the lesion was found incidentally during evaluation for seizures. Two children had median cleft face syndrome, another had an associated Arnold–Chiari type I malformation, and the fourth had no other cranial abnormalities. All patients underwent preoperative evaluation including magnetic resonance (MR) imaging. Auditory, ophthalmological, genetic, endocrinological, or other evaluation was undertaken as indicated. Lesions were approached through the median raphe of the hard and soft palates. All cephaloceles were easily visualized and dissected after division of the nasal palatal mucosa. The dural sac and its contents were reduced by surface coagulation after division and dissection of the overlying mucosa. Once reduced, the bone defect was obliterated in three of four patients. The dura was not opened and anomalous neural elements were not resected. At follow-up evaluation, all patients demonstrated resolution of preoperative symptoms without evidence of infection or lasting morbidity. Follow-up MR imaging showed reduction in all cases. The authors conclude that this transpalatal approach is safe and reliable for the treatment of transsphenoidal cephaloceles in young children.

KEY WORDS • cephalocele • basal cephalocele • transsphenoidal cephalocele • encephalocele • transpalatal approach

Cephaloceles represent congenital herniations of intracranial contents through a bone defect. They are described as encephaloceles when the sac contains neural elements and meningoceles when no recognizable brain tissue is present within the sac. Cephaloceles are classified according to the site of bone defect and herniation. Suwanwela and Suwanwela introduced the currently used classification system, which includes sincipital (frontal), parietal, occipital, and basal defects. Basal cephaloceles have been further subclassified by Gisselsson, and more recently by Gerhardt, et al., according to the site of bone defect and the location of the cephalocele. The subclassification of Gerhardt, et al., includes transethmoidal, sphenoethmoidal, sphenoorbital, sphenomaxillary, and transsphenoidal cephaloceles.

A review of the literature describing surgical indications and management of transsphenoidal cephaloceles underscores the rarity of these lesions and the lack of uniform agreement on their indications, surgical approach, or treatment objectives. However, the available literature does report a high rate of postoperative morbidity, mortality, and permanent impairment. Within the basal variant, transsphenoidal cephaloceles are the most challenging to expose, reduce, and repair through a frontal craniotomy. For this reason, there are case reports of transnasal, transfacial, and transpalatal dissection with concomitant frontal craniotomy for reduction and repair.

We present a transpalatal approach to transsphenoidal cephaloceles without the inclusion of a frontal craniotomy. Four children underwent reduction and repair of transsphenoidal cephaloceles via this approach without mortality or permanent morbidity.

Clinical Material and Methods

Patient Demographics and Presentation

Since 1988, four patients (two boys and two girls) have presented to Children’s Hospital Medical Center in Cincinnati, Ohio, with transsphenoidal cephaloceles. All were evaluated for reduction and repair. Age at presentation ranged from birth to 34 months and age at repair from 3 to 35 months. Two patients were identified as having meningoceles and two as having encephaloceles. Presentation included nasal apnea with associated median facial cleft syndrome in two infants and unexplained meningitis in
Diagnosis and Evaluation

Diagnosis was confirmed by magnetic resonance (MR) imaging in all patients (Fig. 1). Preoperative evaluations were individualized and included genetic, ophthalmologic, auditory, endocrinological, and spinal assessments as indicated. Three patients had other intracranial radiographic abnormalities, including corpus callosum agenesis, Arnold–Chiari I malformation, pituitary dystopia, retinal retention cysts, and elongated optic nerves.

Surgical Repair

Repair of the cephalocele was coordinated by a team of neurosurgeons and plastic surgeons. Lumbar drain placement was attempted at the time of surgery in all patients and successfully placed in two. Orotrachial intubation was used. Perioperative administration of antibiotic medications was begun in the operating room and continued for 72 hours postoperatively. All patients received perioperative steroid medications. After the airway, catheters, and drains were secured, patients were placed in the Trendelenburg position. Phisohex face wash and Peridex mouthwash were used.

A Dingman mouth gag and throat pack were placed on the patient. Exposure was accomplished through the median raphe of the hard and soft palate (Fig. 2 left). Once the palatal mucosal flaps were retracted, palatal osteotomies were performed and the hard palate was removed (Fig. 2 right). After division and retraction of the nasal palatal mucosa, the mucosa-covered cephalocele was visualized within the epipharynx.

With the aid of the operating microscope, the cephalocele and its contents were passively reduced by applying bipolar cautery to the overlying mucosa (Fig. 3 upper). Once reduced, the mucosa was incised and dissected free of the underlying dura to allow precise definition of the bone defect and further reduction of the dural sac and its contents (Fig. 3 center). The dura was not opened and the sac and anomalous neural elements were not resected in any case. For bone defects larger than 0.5 cm, the defect was obliterated. During dissection and grafting of the osseous defect, bone contact between the defect and graft was optimized. Autogenous tissue or titanium mesh was used as graft material. Following reduction of the dural sac and bone grafting, fibrin glue was used in all cases.

Closure involved reapproximating the mucosa overlying the cephalocele and graft with No. 6-0 prolene, and palatal closure followed, as described elsewhere. If the bony palate was not used as the graft, it was replaced during closure (Fig. 3 lower). Follow-up MR imaging was performed between 6 and 12 months postoperatively.

Results

All cephaloceles were reducible using surface coagulation. The dura was not violated in any case. No attempt at resection of redundant dura or anomalous neural elements was made. Three bone defects (Cases 1, 3, and 4) were obliterated and one 0.3-cm osseous defect (Case 2) was not (Table 1). Palatal bone, titanium mesh, and a combination of mesh and cartilage were used to close the defects in three cases (Cases 1, 3, and 4). The bony palate was replaced in situ during closure in three cases.

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**Table 1**

Clinical characteristics of four patients with transsphenoidal cephaloceles

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (mos),†</th>
<th>Sex</th>
<th>Presentation</th>
<th>Location</th>
<th>Anomalies</th>
<th>Endocrine Abnormality</th>
<th>Osseous Defect Obliterated</th>
<th>Postop Course</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>11, F</td>
<td>MCFS, nasal obstruction</td>
<td>TSM</td>
<td>MCL, SMCP, hypertelorism, coanal atresia</td>
<td>anophthalmia, retinal coloboma, OC attenuation</td>
<td>ACC</td>
<td>none</td>
<td>titanium mesh</td>
</tr>
<tr>
<td>2</td>
<td>34, M</td>
<td>incidental</td>
<td>TSE</td>
<td>none</td>
<td>none</td>
<td>Chiari I, PD</td>
<td>none</td>
<td>none</td>
</tr>
<tr>
<td>3</td>
<td>7, M</td>
<td>meningitis</td>
<td>TSE</td>
<td>none</td>
<td>none</td>
<td>none</td>
<td>none</td>
<td>titanium mesh</td>
</tr>
<tr>
<td>4</td>
<td>0, F</td>
<td>MCFS, nasal obstruction</td>
<td>TSM</td>
<td>hypertelorism, notched upper lip, grooved philtrum</td>
<td>ON hypoplasia</td>
<td>ACC</td>
<td>none</td>
<td>palatal bone</td>
</tr>
</tbody>
</table>

* ACC = agenesis corpus callosum; Chiari I = Arnold–Chiari Type I malformation; CNS = central nervous system; DI = diabetes insipidus; MCFS = median cleft face syndrome; MCL = midline cleft lip; OC = optic chiasm; ON = optic nerve; PD = pituitary dystopia; SMCP = submucous cleft palate; TSE = transsphenoidal encephalocele; TSM = transsphenoidal meningocele.

† Age at presentation.
Operative times ranged from 165 to 270 minutes (mean 216 minutes). Two patients (Cases 3 and 4) were extubated in the operating room at the completion of the procedure. Patients remained in the intensive care unit for 1 to 7 days and were discharged from the hospital 7 to 12 days postoperatively. Three patients (Cases 1, 2, and 4) received blood products in the perioperative period.

One patient (Case 1) developed diabetes insipidus on postoperative Day 1, requiring two doses of vasopressin. She has subsequently done well. Another (Case 4) required reintubation on the day of surgery for an inability to protect her airway. After MR imaging and endocrinological evaluation yielded negative results, she was reextubated uneventfully on postoperative Day 3 and has done well since that time.

Follow-up evaluations ranged from 8 to 96 months. Magnetic resonance imaging showed reduction of the cephalocele in all cases (Fig. 4). There has been no worsening or noticeable improvement in the neurological or endocrinological function in any patient.

**Discussion**

Using the expertise of both craniofacial plastic surgery and neurosurgery, the goals in our approach are to expose the ventral aspect of the cephalocele by dissection of the overlying mucosa, reduce the sac by bipolar electrocautery, and seal the osseous defect. The bone or mesh graft is secured by primary repair of the overlying mucosal layers and palatal tissues. Our intent is not to restore all herniated elements to the middle fossa but to elevate the sac beyond the confines of the epipharynx, seal the defect to prevent recurrence, and alleviate any symptoms of respiratory distress or rhinorrhea. Repair also minimizes subsequent risks of rhinorrhea or meningitis. We have found this approach to be safe and efficacious. In follow-up evaluations ranging from 8 to 96 months, there have been no recurrences and the only complication was a single, self-limited episode of diabetes insipidus encountered early in the postoperative course of the patient in Case 1.

**Incidence of Cephaloceles**

Cephaloceles are an uncommon entity that occur in approximately one in every 3000 to 5000 live births. Basal cephaloceles are less common with an estimated incidence of one in every 35,000 live births. The trans-
Teratogens may affect the failure of developing ossification. Late first trimester exposure to x-ray or radiation, trypan blue dye, or excessive doses of vitamin A is known to influence the developing chondrocranium and to induce cephaloceles in experimental studies. The pathophysiological mechanism of these lesions is not completely understood. Teratogens may affect the normal separation of ectodermal elements from neural crest tissues, leaving an osseous defect. Alternatively, the defect may represent persistence of the craniopharyngeal canal, failure of developing ossification centers to fuse properly, or progressive cranial thinning secondary to hydrocephalus. The resulting sphenoidal defect can engender a herniation of intracranial tissue including the third ventricle, segments of the hypothalamus and pituitary axis, anterior cerebral arteries, or optic chiasm into the epipharynx.

The clinical presentation of a patient with a transsphenoidal cephalocele is in part dependent on age. Without the characteristic facies, the diagnosis can be delayed into adolescence or adulthood when an unexplained rhinorrhea, meningitis, or progressive visual field defect prompts evaluation and diagnosis. Even when the diagnosis is made in adults, Smith and colleagues found that 10 of 16 adults presenting with a sphenoidal cephalocele had hypertelorism. Skull films may demonstrate the osseous defect in the lateral view. With characteristic facial malformations, diagnosis is usually made in infancy or early childhood. Respiratory difficulties due to epipharyngeal obstruction prompt evaluation in most cases.

Although none of the children in our series demonstrated preoperative endocrine dysfunction, earlier studies suggest that this is the exception rather than the norm. Hypothyroidism, growth-hormone deficiency, hypogonadism, or diabetes insipidus has been documented as an accompanying disorder in patients with sphenoidal encephaloceles.

Advanced imaging studies are necessary to confirm the diagnosis of transsphenoidal cephalocele as well as to define any neural or vascular elements that may be included in the herniation. Computerized tomography and MR imaging also provide information about the osseous defect and uninvolved brain. Even with the availability of such imaging, reports persist of patients with “nasal polyps” in whom a biopsy specimen is obtained, with resultant rhi-
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norhea or meningitis. Therefore, the potential inclusion of the anterior cerebral arteries or hypothalamic–pituitary elements within these cephaloceles mandates a thorough radiographic evaluation. The historical recommendations for angiography and plain films are obviated by the currently available MR technology.

Treatment of Transsphenoidal Cephaloceles

The treatment, indications, and surgical approaches for transsphenoidal cephaloceles remain controversial. Intervention is indicated for respiratory obstruction and for rhinorhea or meningitis ascribed to the lesion. In asymptomatic individuals or those with endocrine changes or stable visual deficits, indications for repair are less clear. Historical experience has discouraged elective intervention in these lesions largely because of high morbidity and mortality rates and a questionable effect on future function. In reviews by Yokota, et al., in 1986 and David in 1993, mortality rates approached 50% and morbidity or long-term severe disability 70%. Defects in these cases were repaired via a transcranial approach with frontal lobe retraction and attempted elevation of herniated elements into the sella, with or without resection of the cephalocele and its contents.

This conservative view is not universally accepted, however, and others advocate repair in all cases to minimize the lifetime risks of infection, enlargement, or trauma to the sac and its contents. Although the indications for repair are not universally accepted, current literature does support a transcranial approach for the treatment of these difficult lesions, with or without extracranial dissection and exposure. However, case reports have outlined successful extracranial repair of a sphenoidoethmoidal variant via an endoscope and also transorally in two children with cleft palates and sphenoidoethmoidal cephaloceles.

We conclude that the transpalatal approach, as presented herein, for the treatment of transsphenoidal cephaloceles is an effective intervention resulting in minimal morbidity. Preoperative evaluation of these lesions using high-quality computerized tomography and MR imaging as well as multidisciplinary testing is essential to confirm the extent of the lesion and associated abnormalities and to plan the safest possible repair.

References


Address reprint requests to: Kerry R. Crone, M.D., Editorial Office, 231 Bethesda Avenue, P.O. Box 670515, Cincinnati, Ohio 45267–0515.