

## Nasal encephaloceles: definitive one-stage reconstruction

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✓ Nasal encephaloceles can cause complex deformities of the naso-orbital skeleton. As the encephalocele pushes through a defect in the facial skeleton it causes lateral displacement of the medial orbital walls. Correction of this skeletal deformity is necessary to achieve a normal facial contour.

Two examples of nasal encephaloceles are presented and the classification, diagnosis, and treatment of this entity are discussed. The correction of these deformities at an early age is recommended. The suggested method of reconstruction is a combined intracranial and extracranial approach with mobilization of the nasal skeleton and medial orbital walls to their normal position. The remaining defects are treated with bone grafts.

**KEY WORDS** • nasal encephalocele • encephalocele • craniofacial reconstruction • operative approach

**C**ONGENITAL nasal encephaloceles are complex problems that are best treated by the combined efforts of a neurosurgeon and a plastic surgeon. Such anomalies can present with varying degrees of herniation of cranial contents, and simple resection of the encephalocele is seldom adequate to correct the cosmetic deformity. Correction of malformation of the bony naso-orbital region usually requires an extensive craniofacial reconstructive effort. With recent advances in diagnostic and surgical techniques, it is possible to perform a thorough preoperative evaluation, and to treat the lesion with definitive one-stage reconstruction at the time of excision.

In this communication, the classification, diagnosis, and treatment of nasal encephaloceles are discussed. Two cases are summarized to illustrate current treatment methods.

### Description of the Anomaly

#### Classification

In the past, the classification of encephaloceles has been based on their location. However, this method does not distinguish the different types of nasal encephaloceles. In an attempt to clarify the classification system, in 1972, Suwanwela and Suwanwela<sup>14</sup> proposed a method based on the location of the defect within the cranium. They classified nasal encephaloceles into three subgroups according to the site of the defect in the facial skeleton: nasofrontal, nasoethmoidal, and naso-orbital.

In all three types the cranial defect is located between the frontal and ethmoid bones. However, the malformation of the facial skeleton is different in each type (Fig. 1).

In the nasofrontal encephalocele, the defect is between the frontal and nasal bones. Intracranially, the herniating dura passes through a defect in the frontoethmoidal junction. As it projects forward it causes a downward displacement of the nasal bones and a lateral displacement of the medial orbital walls (Fig. 1 *left*). In the nasoethmoidal encephalocele, the defect is situated between the nasal bone and nasal cartilages, and the herniating dura passes through a defect between the frontal and ethmoidal bones. After exiting the cranial cavity, its course is lower and, unlike the nasofrontal encephalocele, it protrudes below the nasal bone (Fig. 1 *center*). In the naso-orbital encephalocele, the defect is in the medial orbital wall between the lacrimal bone and the frontal process of the maxilla. The herniation protrudes downward behind the nasal bone and then laterally, exiting at the medial orbital walls. In this type, the frontal bone, nasal bone, and nasal cartilages are normal. Rather than presenting as a nasal mass, this type presents between the nose and lower eyelids (Fig. 1 *right*).

An additional classification system was described by Tessier<sup>15</sup> in 1976. This method describes clefts as bone defects arranged around the orbit and numbered from 0 to 14. In this system, nasal encephaloceles would be categorized as Tessier Type 14 clefts.

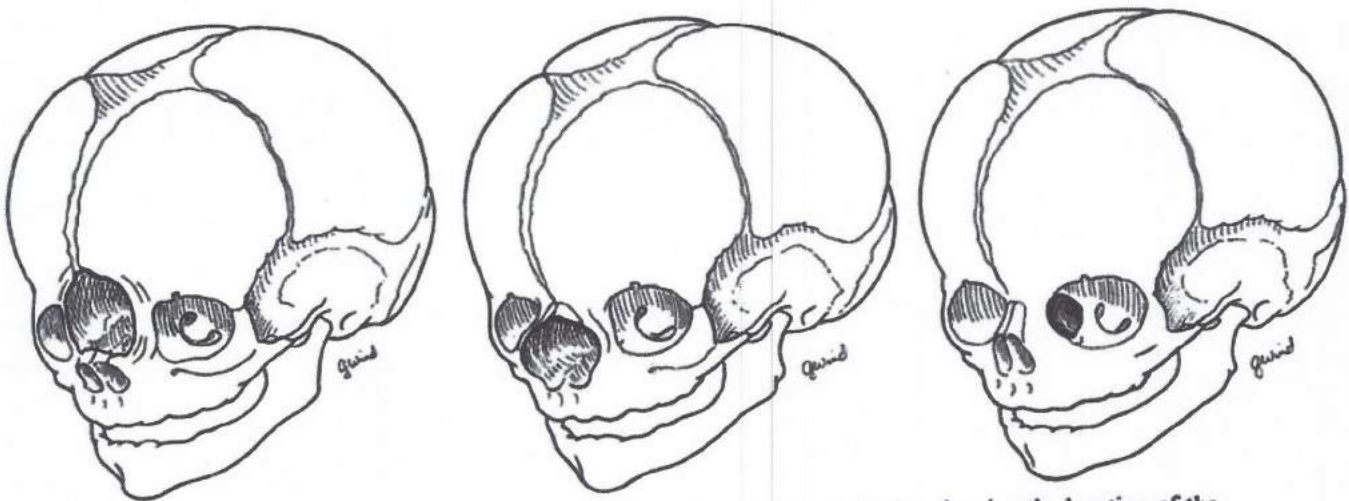


FIG. 1. Examples of the three-group classification of nasal encephaloceles, showing the location of the bone defect: nasofrontal (left), nasoethmoidal (center), and naso-orbital (right).

### Clinical Features

Nasal encephalocele can present as a single space-occupying lesion or as multiple masses in the frontal naso-orbital area. The most common site is in the midline at the root of the nose between the medial canthi. The mass may be soft and cystic or hard and firm, and may range in size from a few millimeters to over 10 cm in width. The large masses usually have thin shiny skin, and may obstruct vision and the nasal airway. With large encephaloceles, secondary facial deformities may be present due to pressure on adjacent structures. The nose is usually broad, flat, and elongated in the upper half. The encephalocele exerts pressure on the anterior medial orbital wall, causing a lateral displacement. This results in telecanthus and can also lead to true hypertelorism, a lateral translocation of the entire orbit. A Mongoloid slant may also be present due to the inferior displacement or lengthening of the medial orbital walls. Displacement of the lacrimal and nasal bone may obstruct the nasolacrimal ducts with resultant epiphora. When nasal encephaloceles are very large, breakdown in the overlying skin may occur with leakage of cerebrospinal fluid and subsequent infection. If diagnosed and corrected early, the deformity can be effectively limited to the medial orbital walls.

### Diagnosis

Nasal encephaloceles must be differentiated from tumors such as dermoid cysts and nasal gliomas, and the diagnosis may be difficult when the communication between the sac and the intracranial contents is small.<sup>1,2</sup> However, investigations may define this communication, along with any associated deformities. Plain skull radiographs are helpful in outlining the cranial defect as well as the orbital alignment. Computerized tomography (CT) and magnetic resonance (MR) imaging are also helpful in visualizing the anatomy. With these preoperative studies, the diagnosis and classification can

be documented accurately. Preoperative assessment of the orbital walls and nasal bone positions is invaluable in formulating strategy for reconstructive surgery.

### Treatment

The treatment plan is designed according to the particular lesion and anatomical deformity. Two cases will be presented to illustrate this point.

### Illustrative Cases

#### Case 1

This baby girl, referred at 1 week of age, was born with a large midline nasal mass that was increasing in size (Fig. 2 left).

**Examination.** She exhibited a large sac-like mass in the frontonasal area that transilluminated. Due to the large size of the mass, the visual field of the right eye was almost completely obscured. The distal half of the nose was flattened and there was partial obstruction of the nasal airway. The medial canthi were widely separated, but the orbits appeared normal.

Plain radiographs revealed a frontonasal defect with widening of the medial orbital walls. The position of the lateral orbital walls was normal and the cribriform plate was minimally displaced inferiorly. A CT scan (Fig. 3) and MR imaging confirmed that the mass was an encephalocele with the communicating defect situated above the nasal bones.

**Operations.** The encephalocele was approached via a bicoronal scalp incision. A frontal bone flap was raised in two sections due to the open metopic suture. The dural defect was identified and the dura opened. Cerebral tissue and a left frontal subarachnoid cyst herniated through a large defect in the nasofrontal region. The dura was dissected free from the anterior cranial fossa and the portion of the brain extending into the encephalocele was excised. The dural defect was repaired

## One-stage reconstruction of nasal encephaloceles

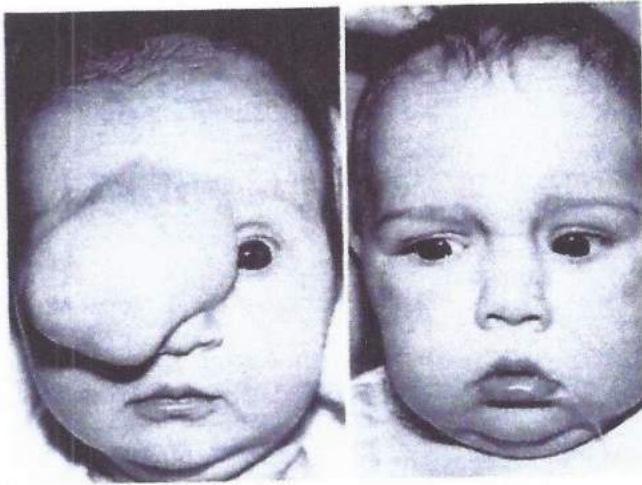


FIG. 2. Case 1. *Left:* Photograph of the patient at 1 week old showing a large nasofrontal encephalocele. *Right:* Post-operative photograph of the patient 8 months after definitive one-stage reconstructive surgery.

with a pericranial graft. The encephalocele sac was then approached extracranially by making a transverse incision and dissecting the sac away from the overlying skin. The sac was excised, allowing excellent exposure of the defect at the frontonasal junction and the medial orbital walls. An extensive circumferential subperiosteal orbital dissection was performed, and dissection of the lateral nasal wall mucosa allowed movement of the nasolacrimal ducts. The caudal two-thirds of the nasal bones were present but not connected to the frontal

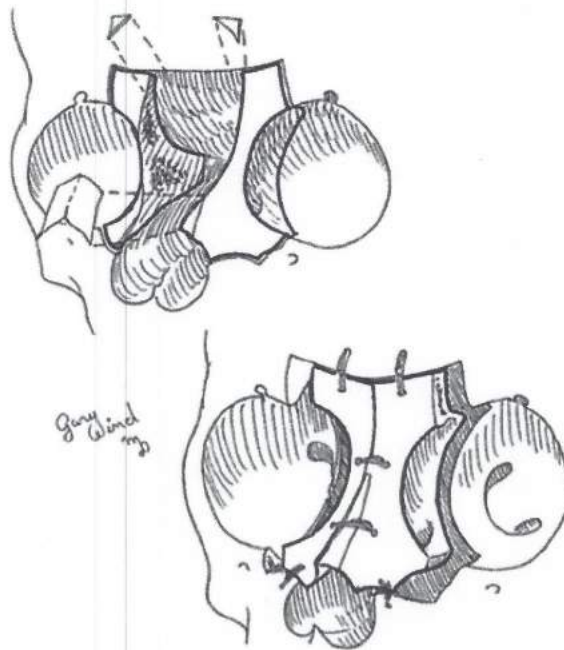
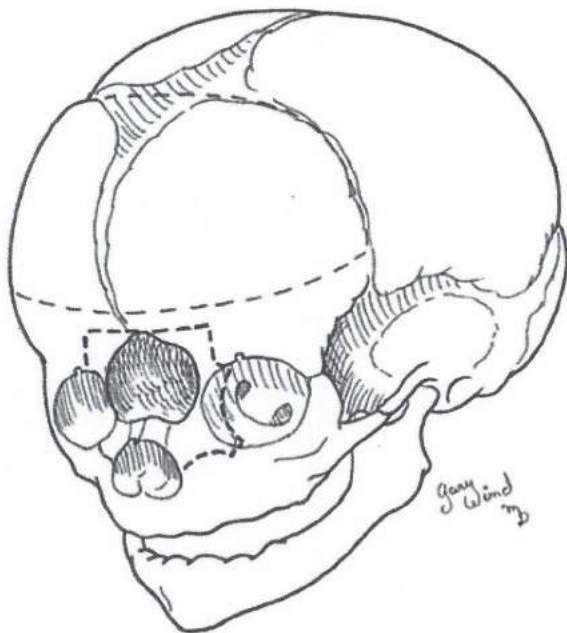


FIG. 4. Case 1. *Left:* The nasofrontal encephalocele bone defect is shown. *Dashed lines* indicate the extent of osteotomies for mobilization of the medial orbital walls. *Upper Right:* The medial orbital walls are centrally mobilized and stabilized. *Lower Right:* The nasal bone is repositioned or reconstructed, and the remaining defects are treated with bone grafts.

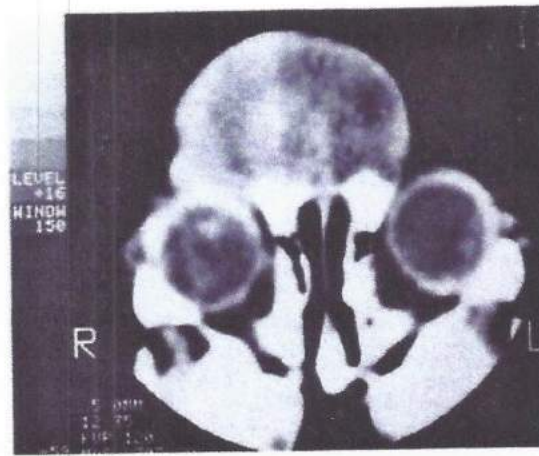


FIG. 3. Case 1. Computerized tomography scan showing the nasal encephalocele with widening of the medial orbital walls.

bone. The cribriform plate was minimally displaced inferiorly.

Vertical osteotomies were made in the medial thirds of the supraorbital and infraorbital rims, in both medial orbital walls and orbital roofs extending into the bone defect, and in the medial third of the orbital floor, extending to the vertical medial wall osteotomy (Fig. 4 *left*). The medial orbital walls were then centrally mobilized and the nasal bone repositioned (Fig. 4 *right*). The defects created in the orbital rims and medial walls were grafted with contoured calvarial bone. The medial walls and bone grafts were wired together and to the frontal bar in order to maintain position. Contoured



FIG. 5. Case 2. *Left:* Photograph of the patient at 6 months old, showing the nasoethmoidal encephalocele. *Right:* Postoperative photograph of the patient 1 year after definitive one-stage reconstructive surgery.

calvarial grafts were placed against the lateral orbital walls to decrease the orbital dimension. The medial canthi were correctly positioned and wired directly into bone with transnasal wires. The frontal bone plates were replaced and wired to the frontal bar and the lateral canthi were repositioned. The scalp incision was closed, and the excess nasal skin and soft tissue were excised transversely. The postoperative course was uneventful. At her 1-year follow-up examination, the patient had maintained the anatomical correction (Fig. 2 right).

#### Case 2

This baby boy, referred at 6 months of age, was born with a large eccentric intercanthal mass (Fig. 5 left).

**Examination.** Objectively, the mass occupied the frontonasal region, was compressible, and transilluminated irregularly. The intercanthal distance was 38 mm, and both medial canthi were displaced inferiorly. Plain radiographs showed widening of the medial orbital walls and a low cribriform plate. The lateral orbital walls were in a normal position. Computerized tomography revealed that the mass communicated with the frontal lobe and confirmed the diagnosis of an encephalocele.

**Operation.** The encephalocele was approached via a bicoronal scalp incision and a frontal bone flap was removed, leaving a supraorbital bar. The dura was opened and the mass resected from its origin at the base of the frontal lobes. Likewise, it was dissected extradurally from the anterior cranial fossa, then excised. The sac exited between the nasal bone and nasal cartilages and had two lobes: one located to the right and one to the left of the midline. However, there was no intervening tissue between these lobes. Later histological analysis of the specimen showed glial elements.

The sac was then approached extracranially through a transverse incision in the overlying skin. The mass

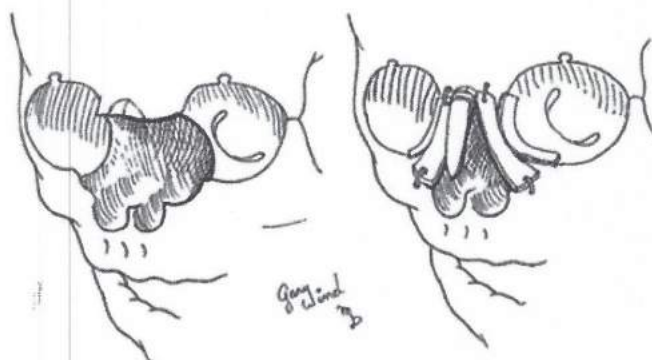


FIG. 6. Operative diagrams in Case 2. *Left:* The large nasoethmoidal encephalocele bone defect is shown. *Right:* The defect in the naso-orbital skeleton is shown reconstructed with contoured calvarial bone grafts.

was dissected away from the skin and excised from both the intracranial and extracranial aspects. The dura was closed, and a large bone defect remained. The nasal bone and portions of the medial orbital walls were absent and the interorbital distance was larger than normal (Fig. 6 left).

The medial canthal ligaments were isolated and tagged, and extensive subperiosteal mobilization of the orbital contents was carried out bilaterally. The medial orbital walls were reconstructed with tailored calvarial bone grafts (Fig. 6 right). The medial canthi were then repositioned and wired directly into the medial orbital walls, decreasing the interorbital distance. A contoured calvarial bone graft was wired to the supraorbital bar to restore the nasal bone. A pericranial flap was used to seal off the anterior cranial fossa from the nose and orbits. The lateral canthi were wired into position, the bone flap was returned, and the scalp was closed. The excess nasal skin was removed and the incision closed transversely. The postoperative course was uneventful and the patient continues to do well 3 years after his surgery (Fig. 5 right).

#### Discussion

There is an unusual geographic distribution of nasal encephaloceles. In North America the lesions are rare; however, in Southeast Asia they are quite common. In Thailand, for example, they occur in one of 6000 live births,<sup>11</sup> with a 9.5:1 ratio of anterior to posterior lesions.<sup>2</sup> In North America, however, posterior lesions are much more common than anterior ones.<sup>7</sup> There does not appear to be any pattern of genetic inheritance or environmental factors affecting the incidence of nasal encephaloceles.

In general, the prognosis for patients with these lesions is excellent.<sup>7,9</sup> Hydrocephalus, increased intracranial pressure, and major neurological deficits are seldom a problem. However, these lesions can create a significant cosmetic deformity of the nose and orbits.

The anatomy of these deformities is complex. The herniating mass exits through a cranial defect that in-

## One-stage reconstruction of nasal encephaloceles

variably corresponds to the foramen cecum at the junction of the frontal and ethmoid bones. Only the distal pathway varies as it exits the facial skeleton, which is the basis of the three-group classification. The pressure of the herniating encephalocele causes a widening of the medial orbital walls with a resultant telecanthus and nasal deformity. Definitive treatment of this deformity requires correction of the encephalocele, and a thorough mobilization and reconstruction of the involved naso-orbital skeleton along with correction of the telecanthus.

In nasoethmoidal and naso-orbital encephaloceles, large defects may be present in the medial orbital walls. The encephalocele herniates through these defects, pushing against the periorbital. The dissection of the encephalocele sac may be difficult in this area and is best approached extracranially. The periorbital is first dissected out in a normal adjacent area to find the appropriate plane. The encephalocele sac can then be more easily separated without injury to structures in the periorbital. The displaced bone elements and defects are reconstructed by a combination of osteotomies and bone grafting. The displaced bone structures are returned to their normal position and the defects in the medial orbital walls and nasal bone are reconstructed with bone grafts. With this approach the normal bone anatomy can be restored.

Removal of any extruded cerebral tissue and repair of the dura and anterior cranial fossa is essential. In addition to the displaced bone elements, there may be large dural defects, and an attempted "lesser" procedure, such as a subcranial approach, could very easily become an intracranial one due to a low-lying cribriform plate. Therefore, an intracranial-extracranial approach not only is necessary, but is the safest technique.

In the past, nasal encephaloceles were initially managed by neurosurgeons, and the residual deformity was treated by plastic surgeons at a later date. Charoonsmith and Suwanwela<sup>2</sup> recommended late secondary bone reconstruction for patients 3 years of age and under. However, since the development of modern craniofacial surgical techniques, definitive one-stage reconstruction of the deformity, as reported by Tessier<sup>15</sup> and others,<sup>3-6,12,13,16</sup> is the current treatment of choice.

An important observation illustrated in the cases presented is the fact that a telecanthus is usually present rather than a hypertelorism.<sup>4-6,16</sup> This corresponds to the early deforming forces of the encephalocele. True hypertelorism, or a lateral translocation of the entire orbit, is uncommon with these lesions but may be present in the older patient due to continued forces during early growth.<sup>10</sup> If treatment is instituted early, it can usually be limited to movement of the deformed naso-orbital skeleton. After the appropriate osteotomies are made, the residual defects are then reconstructed with calvarial bone grafts. As seen in the cases presented, movement of the medial orbital walls with bone grafting and canthopexy provided definitive one-stage correction.

## Conclusions

For cases of nasal encephalocele, an early intracranial-extracranial approach with definitive reconstruction of the nasal, orbital, and canthal deformity is recommended. The nasal encephalocele can be safely treated at an early age with good results. This treatment can stop progression of the deformity and can potentially restore normal growth forces.

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