Chapter 11

Neural Tube Defects
Benjamin C. Warf and Louis Carter

Encephaloceles may be anterior between the forehead and nose or posterior in the occipital area or, less commonly, at the vertex or in the parietal region.

Nasal encephaloceles are discussed here since they present as a facial mass in childhood and are a concern to the parents. These masses may be large and cystic and there is a risk for rupture and infection. At one time the only accepted approach for an adequate repair was through a bifrontal craniotomy. In recent years some have suggested this is not necessary and that the repair may be done extracranially. These are thought to be in the spectrum of neural tube defects along with spina bifida (myelomeningocele), the risk for which can be reduced with dietary folic acid supplementation from the time of conception. These are common in some regions and not others. They are common in East Africa but not in West Africa. They are quite common in Southeast Asia. Spina bifida and hydrocephalus are commonly seen together as are encephaloceles and hydrocephalus. Surgery for nasal encephalocele when there is underlying hydrocephalus may result in a CSF leak after repair. It is important to get plain skull x-rays, an ultrasound and a CT scan if possible prior to surgery, not only to rule out hydrocephalus but to also rule out other midline facial masses as glioma and dermoid. Gliomas are firm and extend intracranially whereas a dermoid typically does not. Encephalocele may coexist with a glioma. A midline dermoid is less common than the lateral angular dermoid over the lateral eyebrow (Fig 1).

This is a midline dermoid in a teenage girl. It is superior to the usual nasoencephalocele. Longstanding dermoids will leave a depression on the skull but the skull is not involved.

In medical centers encephaloceles are best treated by a neurosurgeon and craniofacial plastic surgeon. When these specialties are not available then the
A general surgeon may be required to handle these cases but with great care and only if the family cannot get to a medical center for specialized treatment. Sometimes a rupture will require immediate care and this is a reason for this chapter (Fig 2).

Nasoencephalocele will usually present to the surgeon as a soft usually cystic mass between the eyes in the newborn. There may be some firm areas where there is glial tissue. There is no urgency for repair unless they are very large and obstruct vision or where they are very thin and there is a concern for rupture (Fig 2). In these cases there is a need for early decompression with repair.

There are three types of nasal encephalocele:

1. Nasofrontal
2. Nasoethmoidal
3. Naso-orbital

In a **nasofrontal encephalocele**, there is a defect between the frontal and nasal bones and the protruding mass pushes the nasal bones down and the medial orbital walls out. The intracranial defect is at the frontoethmoidal junction.

In a **nasoethmoidal encephalocele**, the defect is between the nasal bone and nasal cartilages and the mass projects below the nasal bones. This is the most common type seen in East Africa. The herniating dura passes through a defect between the frontal and ethmoidal bones.

In a **naso-orbital encephalocele** the defect is in the medial orbital walls between the lacrimal bone and frontal process of the maxilla and it presents as a mass between the nose and orbit. It may be obvious that the globe is pushed laterally. The frontal and nasal bones and the nasal cartilages are normal.
These may be seen in combination as a nasoethmoidal with a naso-orbital. In these cases the naso-orbital may be small and **missed completely** during the repair. Large encephaloceles may exert pressure on the medial orbital wall causing telecanthus (increased distance between the medial canthi) and occasionally even hypertelorism (increased distance between the orbits) when unrepaired. Other findings are a Mongoloid slant and obstruction of the nasolacrimal duct with epiphora—tearing.

Usually surgery is delayed for several months in order to obtain a CT scan and to ensure there are no other significant anomalies. If there is no visual obstruction and little chance for rupture, then surgery can safely be delayed until the child is one year of age. The CT scan will not only help rule out hydrocephalus but will also show the anatomy of the nasal encephalocele. If hydrocephalus exists, a shunt should be placed prior to the encephalocele surgery. As mentioned previously, it is not uncommon for the encephalocele to have an orbital component that may not be seen clinically on exam. Surgery must close the orbital defects with bone grafts or there will be a recurrence.
The surgical approach described here is an external approach. This approach does not require craniotomy instruments. The child must be kept warm during the procedure and blood should be available. Ideally one will have a skull model present in the theatre. If not, an anatomy textbook should be close by. The author still uses a standard bicoronal flap below the galea to gain exposure and this is usually combined with a vertical mid-line incision between the eyes in order to gain direct visualization. The pericranium can be reflected separate from the skin/galeal flap and used at the end of the case for a vascularized flap to separate the dura from the nose. An alternate method is to reflect the skin/galeal flap leaving the pericranium on the skull to just above the orbits where it is divided and reflected down exposing the orbits and any naso-orbital encephaloceles. See below.

Technique:
The skin/galeal flap is reflected to just above the orbits. The pericranium is incised above the supraorbital rim and the flap with periosteum is elevated off the supraorbital rim and into the orbit. Care is taken to elevate the supraorbital vessels with the flap. The periosteum can be carefully dissected off the medial walls of the orbit in order to visualize any orbital component to the encephalocele.
A small nasofrontal, glabellar, craniotomy is made by using a bur or side cutting saw or just a drill to make osteotomies across the root of the glabella in the frontal bone. See above. This is carried along the lateral edges of the nasal bones, just medial to the orbits, and down to the lower edge of the nasal bones. (A craniotome is an ideal instrument to make these cuts if it is available.) The nasal bones are removed giving a direct approach to the root of the encephalocele and bony defect. The mass is identified and the dura is dissected free from surrounding tissue and bone down to the anterior edge of the cribriform plate. The dura is very thin and fragile as the hernia sac is in an infant and the author has not found it easy to dissect out a “sac” of dura without creating multiple holes. It is best to save as much dura as possible, excise any atrophic neural tissue and close the sac if possible. Often a pericranial graft is necessary to close the dura. A meticulous, water-tight, closure is necessary. After carefully dissecting the dura away from the defect, one anticipates finding edges of dura to close with a pericranial graft—skull periosteum. If not possible to find good dural edges then an additional segment of skull is removed just above the craniotomy shown above to give better exposure—this is an excellent technique to give good exposure. At times it may be impossible to close the underside of the dural defect along the cribriform plate. Then a pericranial graft can be placed above the cribriform plate to completely seal off the dura. The pericranium is taken from the skull above where one elevated the galeal flap—posterior to the bicoronal incision.

A bifrontal craniotomy can always be done but this would be a challenge for most general surgeons in a district hospital. Once the pericranial patch has
been sutured to the dura, if fibrin glue is available it is ideal to seal small dural tears along the repair. If it is not available, “Super Glue” can be found anywhere in the world. The tube of Super Glue is soaked in antiseptic solution and then transferred to the sterile field. A sterile needle is used to pierce the top. The glue inside is sterile and can be used sparingly to seal defects in the dura (and even hold cranial bones together). This is an “Off Label” use of Super Glue but many neurosurgeons have used this successfully over the years without complications. Some of the newer products like “DuraSeal” have had complications with extrusion of the “DuraSeal” material. No one seems to know why. It is not recommended by the author for young children. No complications from Super Glue have been experienced.

A vascularized flap of pericranium based on the supraorbital vessels or superficial temporal vessels may be used to close the dural defect if the pedicle has not been divided. This may also be used in addition to the free pericranial graft discussed above in order to give vascularized tissue between the cranial and nasal cavities. This is taken transversely above the orbital rim based on the lateral vessels, branches of the superficial temporal artery.

Following dural closure some feel a small bony defect will close without bone grafting but most use a bone graft. The bone graft may be taken from the calvarium, iliac crest or split rib. Calvarium is preferred but calvarium cannot be easily split until the child is 4 years of age. A small full-thickness calvarial graft can be used and in most cases the donor area will close. Ideally small titanium plates are used to secure the bone graft in place but it is unlikely that district hospitals would have these. The bone graft can be stabilized with Vicryl sutures or even fibrin glue or Super Glue. Alternatively, small titanium mesh if available can be molded and placed in the defect. It can be screwed or sutured into the surrounding bone. In most cases mesh does well though there is a possibility of late extrusion.
Reconstruction of nasal encephalocele
(Reprinted with permission, Albright, Pollack & Adelson Operative Techniques of Neurosurgery, Albright, ed., Fig. 8 pg. 42, Thieme. NY: 2000.)

The author uses the following sequence in closure of the defect. First, the dura is closed directly or by pericranial graft (usually a free graft but it can be a vascularized graft). Second, the bony defect is closed by bone graft or titanium mesh. Third, but not always necessary, a vascularized pericranial flap is used to separate the skull defect from the nasal cavity (hopefully the nasal cavity has not been violated). Finally, the nasal bones are replaced.

Many small naso-orbital encephaloceles are missed initially. In these encephaloceles the dural sac must be carefully dissected out of the orbit and the defect bone grafted. It is often easier to place a thin graft on the orbital side.

Preoperatively, if telecanthus is identified, medial orbital osteotomies need to be done to move the canthi in. This procedure by Sargent is referenced below. Normally, the author does not do these osteotomies in young children. In the young children he identifies the medial canthal ligaments and brings these together in the midline. This is best performed with small wire sutures through each canthus and twisted together in the midline. The medial canthi can be identified by grasping and pulling on the tissue until there is pull on the tarsal plates in both upper and lower eyelids. In older children that require osteotomies, it is best if a neurosurgeon is available.

The nasal bones are then replaced and plated or wired or sutured back to the skull. If bone graft has been used for the defects above, then bone would likely be available to place a dorsal strut as a cantilever bone graft for nasal definition.
Skin is closed in two layers. The excess skin is excised, though it will shrink down if just closed. Postoperatively the patient is continued on antibiotics for a few days and the head is kept elevated.

At times hydrocephalus will not be identified preoperatively and a CSF leak will develop. If there is a leak without hydrocephalus, a lumbar drain can be placed and left in a few days.

**Summary**

Most surgeons in district hospitals will not operate on a nasal encephalocele and that will be a wise decision. What happens if it ruptures, then one will be forced to operate and close the defect. For this reason this chapter was written.

**Meningomyelocele**

Meningomyelocele is very common in East Africa and Southeast Asia. It was once common in the Western world but in recent years young women in the West have been instructed to take folic acid prior to conception and this has reduced the risk. Moreover, when a fetus is found to have a meningomyelocele in the early months of gestation, the embryo is often aborted.

In Africa where there are few neurosurgeons or plastic surgeons, the general surgeons are consulted to close these defects. This section deals with skin closure in large meningomyeloceles, but it is most important to get a secure, water tight, closure of the dura. Usually wide dissection and mobilization of the surrounding skin is sufficient to close these adequately. The author’s (Warf) technique is as follows:

“Finally, the skin must be closed in a way that avoids tension and ischemia at the suture line. Some surgeons advocate swinging in a flap of lumbodorsal fascia as an extra layer of closure over the dura. See below. This is optional, in my opinion, but can be helpful especially if the dural closure is tenuous. This is a pedicle flap based on perforators. It is based on one side of the defect and the fascial flap turned over to cover the entire defect. One must take enough fascia distally so than one may leave 2-3 cm. of fascia at the base for blood supply and still cover the defect.

Bluntly undermine the skin circumferentially around the defect. As much as possible, preserve the columns of tissue that may contain its blood supply. Undermine far laterally to mobilize as much skin as possible. Bring the skin edges together with interrupted 3-0 or 4-0 Vicryl stitches through the relatively tough layer at which the dura, fascia, and skin had been originally fused. Although not essential, I leave a small round Jackson Pratt drain subcutaneously that exits through a distant stab incision to be connected to bulb suction. This prevents fluid accumulation beneath the flap that can
compromise wound healing. After the skin edges are approximated, trim redundant (and especially abnormal) skin and “dog ears”, and close the skin with a 4-0 monofilament on a cutting needle. Cover with a dry dressing and keep the infant prone or on the side for several days in the post-operative period to avoid pressure on the closure that can compromise its vascular supply.

Sometimes, a local skin flap advancement or rotation is necessary to accomplish a relatively tension-free closure. A suture line that is under too much tension is doomed to dehiscence. If closure of skin is technically impossible, which is rarely the case, a fall back is to create relaxing incisions lateral to the defect in the flank regions. This allows closure of the defect over the dura. The relaxing incisions can then heal in by secondary intention with wet-to-dry dressing changes.” See Fig 17. This creates two large bipedicle flaps.

![Fig 13](image13.png) ![Fig 14](image14.png) ![Fig 15](image15.png)

Lipomeningocele with breakdown of dural closure: Required a turn over flap of lumbar fascia to secure water-tight closure. In Fig 2 the forceps are holding the fascia. Fig 3 shows the completed closure with the back fascia sutured down to the dura along the bony defect. The fascia was based superior to the defect and turned down to reach the inferior margin without tension.

Surgeons who see a large volume of such cases have learned that with wide dissection and mobilization of surrounding skin and fascia, the defects may be closed. It is important to keep the dissection beneath the superficial fascia (Scarpa’s fascia) which will give fasciocutaneous flaps. In some cases, it the superficial fascia is tenuous, it may be necessary to dissect beneath the muscle fascia.

**To demonstrate the methods of closure a relatively small meningocele will be used. This one could be easily closed by undermining and a midline closure with the technique described above.**
Discussed below are various methods to close the skin when undermining is not enough. If one has limited experience in flap surgery one can do relaxing incisions along the mid-axillary line as discussed in the technique above. The open areas left by the advancement of these flaps can be grafted or left open to heal by secondary intention. This technique may be the best method for one not trained in flap surgery when the defect is quite large as Fig 25. The figure below shows the incisions in the mid-axillary line and the dual blood supply from the thoracodorsal perforators superiorly and the lumbosacral and superior gluteal perforators inferiorly.

Shows the bilateral bipedicile flaps—blood supply from superior and inferior. The lateral incisions (black arrows/blue lines) are actually in the mid-axillary line. The tissue between these incisions and the defect is raised beneath the superficial fascia (Scarpa’s fascia in the back). Blood supply comes into the flaps from perforators, superior (thoracodorsal) and inferior (lumbosacral and superior gluteal). The closure is in the midline. The defects created laterally can be grafted or allowed to heal by secondary intention. This is the easiest way to close these defects if one does not have experience with other flap techniques.
Rhomboid flap Closure: Angles in the residual defect after closure of the meningomyelocele are 60° and 120° (these may have to be created for the rhomboid flap to fit well). Flaps are raised beneath the superficial fascia or even deep muscle fascia to capture more perforators. The tips of each flap in the flanks will reach the apices of the defect. Often if the skin is supple, one flap will be sufficient. In the case below, the one flap was not quite enough and a second flap was necessary. As in all flaps the surrounding skin outside the flaps must be elevated and mobilized for closure. The dissection should be carried deep to the superficial fascia and just above the muscle fascia.

Rhomboid flaps: Complete undermining of the left sided flap was almost enough to gain complete closure.

Lumbar perforator flaps
The lumbar perforator flaps are very reliable and can be extended all the way to the mid-axillary line. The dissection should begin distally (laterally) and continued medially in a fasciocutaneous plane—beneath the superficial fascia (Scarpa’s). Several cm. from the midline, the dissection should be continued in a deep fascial plane—deep to the muscle fascia. It would not be wrong to include muscle fascia the entire length of the flap, but the flap will be slightly more bulky, will not advance as easily and there will be more bleeding with dissection in this deep plane. These flaps may be very wide. In such cases a small backcut may be necessary at the black arrow. This backcut across the base of the flap must be only 1-2 cm. and only through the skin so that the blood supply to the flap is not compromised. One flap or two flaps may be taken. If two are used, the resulting suture line will be in the midline after rotation of the flaps.

The above flap can be used for more inferior defects than our example—note the location of the arrow. Small red dots indicate the location of perforators. Usually the flap is turned on the medial perforators. This flap will be rotated in such a way that the lateral tip will end up in the superior midline. See chapter on Pressure sores and reference here:

The keystone flap is described in the Perforator Flap chapter. One may use one flap or two if necessary, as shown below.

Approximate location of perforators marked with dots. These are island flaps based on deep perforators. The island may extend to the mid-axillary line. The island should be mobilized by blunt dissection so that the perforators are not damaged.
Once dura is closed satisfactorily, then one of the methods above may be used for skin closure. In this case very large flaps should be used; however, the defect will likely be much smaller after dural closure. Relaxing incisions in the mid-axillary line bilaterally may be safest and easiest for the non-plastic surgeon.

A newer method for closure has been described recently that also relies on the lumbosacral and thoracodorsal perforators:


This method includes the formation of triangular flaps with careful measurements. One will need to review this article carefully and have it close at hand in the operating theatre as it requires geometric planning of the incisions.

Conclusion:

Most defects will be closed primarily by experienced surgeons. The rhomboid flaps, the lumbar artery perforator flaps (Fig 18, 21) and the newer keystone perforator island flaps are recommended if the defect cannot be closed primarily or cannot be closed without tension. The method of closure using bilateral lateral relaxing incisions in the mid-axillary line (Fig 17) is a rather simple but excellent method for one who does not have experience with flaps.