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Closure of the Myelomeningocele

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Repair of the myelomeningocele in a newborn is relatively straightforward and based upon an understanding of the developmental anatomy of neurulation and what has gone wrong. Preservation of neurological function, detection of associated anomalies, and prevention of postoperative complications are the essential aims of this procedure. Pre-operative concerns include preparation of the infant for surgery and informing the family of the risks and advantages of surgery as well as what the future likely holds for the child.

Patient Selection

Fortunately, prenatal ultrasound and serum AFP have allowed time for a decision about the future of the pregnancy and, if the pregnancy is going to term, time for the family to prepare for the birth of a child with a neural tube defect. When the birth of a child with a neural tube defect is a surprise, it is important for the parents to have some time to grapple with the many new issues facing them. It is equally important to proceed with definitive treatment in a timely fashion. Fortunately, most parents today are aware that the child will be born with spina bifida and have had prior counseling. Unless the newborn is critically ill, repair of the myelomeningocele should proceed. Significant delays increase both morbidity and mortality.

Early closure of the myelomeningocele remains an important part of initial management. Although studies have not documented any increase in the deficits in the survivors of briefly delayed closure, our experience with patients transferred late and requiring delayed closure in the face of infection has been a decrease in motor function in some and an increased rate of ventriculitis (37%), as compared with that in patients with early closure (7%).

The natural history of unrepaired newborns who are fed but denied antibiotics would indicate that 40 to 60% will survive, often much more significantly impaired. If antibiotics are added to the care of the unrepaired children, the mortality and morbidity fall to levels similar to those of neonates repaired in the first 24 hours.

Optimally, we operate on the neonate soon after birth, preferably in the first few postnatal days. Prenatal diagnosis makes this increasingly more possible. The operation may safely be deferred for up to 72 hours without an increase in complications. This delay is particularly important for the unstable or critically ill newborns. These babies can usually be stabilized within 72 hours, and this time is usually well spent. A search for coexistent anomalies of other organ systems should be undertaken during this time. Severe anomalies or absence of vital organs or unrepairable cardiac defects may portend a poor outcome. Renal anomalies are common but not usually life-threatening. Although the neonate may not produce significant amounts of urine during the first 24 hours, the presence of urine in the bladder implies the presence of functioning kidneys. Ultrasonography can delineate most major renal anomalies. Syndromes related to chromosomal anomalies may not be obvious upon initial inspection but should be sought out.

Although most coexisting anomalies are not immediately life-threatening and may be dealt with without much difficulty, it is important to remember that a few newborns with myelomeningocele may have potentially fatal associated malformations and may not be saved. Intervention to prolong the lives of these infants in the setting of a dismal outlook makes little sense. They should be kept comfortable, and their families should be supported.

Preoperative Preparation

The preparation of the neonate with a myelomeningocele for surgery is usually not difficult. Most have a high hematocrit and an adequate intravascular volume, and fluid resuscitation is therefore usually not necessary. Common perioperative complications include hypothermia and hypoglycemia, both of which are easily prevented through the judicious use of heating and monitoring of serum glucose.

The placode may become desiccated with prolonged exposure to the air and should therefore be protected. Covering the placode with sterile, saline-soaked gauze is preferable. The dressing may be covered with plastic wrap to prevent rapid evaporation of the saline. Substances that are toxic to tissues and result in inhibition and delay of wound healing should not be used directly on the malformation. The use of perioperative antibiotics is left to the discretion of the surgeon. We have tended to use them.

Operative Procedure

Preservation of Neurological Function

Preservation of Neural Tissue

Prior to closure, the infant should be kept on his or her abdomen to reduce mechanical trauma to the neural tissue. During the subsequent surgical repair, great care must be taken to avoid drying, traction on neural elements, irrigation with hot saline, and excessive use of electrocautery. With magnification and microinstrumentation, the opening of significant blood vessels can be avoided. Occasionally, the entire closure can be performed without cautery.

It has clearly been shown that the exposed neural tissue is functional. Movement of muscles subserved by spinal cord segments involved in the placode as well as the presence of somatosensory evoked potentials conducted through the placode both point to the functional nature of this tissue. Even when the initial examination fails to demonstrate movement of muscles innervated by the placode, the placode should still be considered functional because more than one third of these patients subsequently gain motor functions not previously detected. Therefore, all neural tissue must be preserved.

Preservation of Vascular Supply

Preservation of the vascular supply to the placode is essential if this tissue is to survive. Unlike the normal spinal cord, the blood supply to the placode does not enter exclusively through the vertebral foramina along the nerve roots. Many large vessels pass directly through the laterally reflected dura mater and supply the myelomeningocele. Those supplying the junction between the neurulated spinal cord and the placode seem to be at greatest risk. Rarely is it possible to preserve all of these vessels, and, fortunately, they can sometimes be sacrificed if necessary without apparent injury to the placode. Nonetheless, great care must be exercised to preserve these vessels while mobilizing the dura for closure (**Fig. 15–1A**).

Inclusion Dermoid

Great care should be exercised in separating the edge of the placode from the contiguous cutaneous epithelium (**Figs. 15–2A** and **15–2B**). Some pearls of epidermoid tumors may already reside within the placode. Retained fragments, possibly even a single cell, could, if imbricated within the closure, produce an inclusion epidermoid tumor (**Figs. 15–3A** and **15–3B**). These inclusion dermoids produce not only tumors but associated desquamation debris, which may also stimulate an intense arachnoiditis. Later in the child's life, a tethered cord release in the face of the scar produced by this inflammatory process can be extremely difficult.

Missed Abnormalities

Split Cord (Diastematomyelia)

Both the rostral and caudal ends of the closure site should be closely inspected prior to the closure of the placode to identify associated tethering, bony spurs, or fibrous bands. Cranially, removal of an additional lamina may be necessary to adequately visualize the adjacent spinal cord (**Figs. 15–3C** and **15–4**). Hemimyelomeningoceles may also be readily visualized by examining the adjacent spinal cord. The presence of an asymmetrical neurological deficit preoperatively should alert the surgeon to the possibility of a hemimyelomeningocele or an associated split cord malformation.

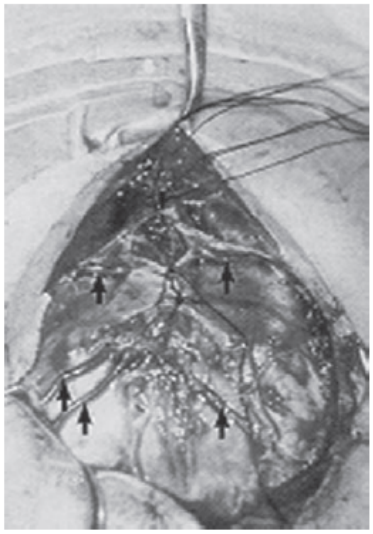
Thickened Filum Terminale

Caudal to the placode a thickened filum terminale may often be present (**Figs. 15–1B, 15–5A,** and **15–6A**). This should be sectioned if present. Spinal cord tethering in these patients may result as much from a missed thickened filum as from adhesions to the placode.

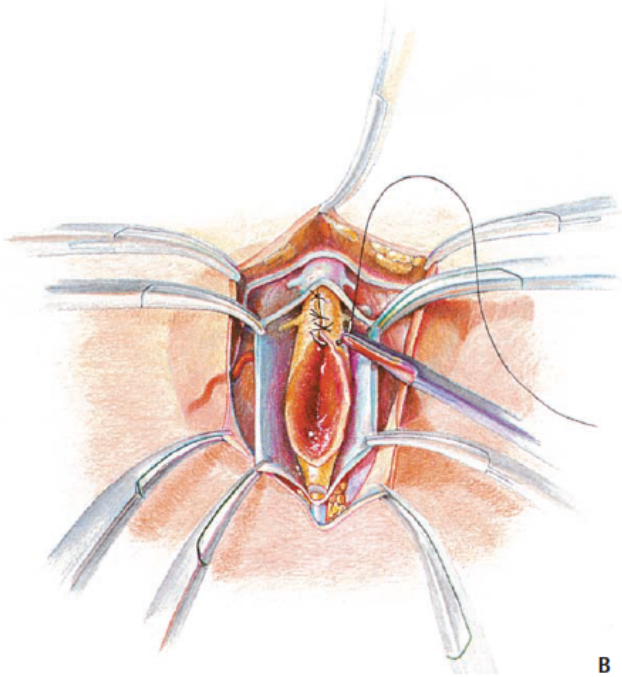
Anatomical Reconstruction

The "different types" of myelomeningocele are best understood in terms of an archetypal anatomical deformity and variation about that archetype. The basic deformity consists of an open neural placode, which represents the embryologic form of the caudal end of the spinal cord prior to neurulation (**Fig. 15–5A—15–5C**). A narrow groove passes down the placode in the midline. This represents the primitive ventral sulcus, and it is directly continuous with the central canal of the closed spinal cord above (and occasionally below) the neural placode. Cerebrospinal fluid passes down the central canal of the spinal cord and discharges from a small pit at the upper end of the placode to bathe the external surface of the exposed neural tissue. This fluid does not indicate rupture of subarachnoid space ventral to the myelomeningocele.

The size of the sac on the baby's back at the time of birth is dependent upon the amount of spinal fluid that is collected ventral to the neural placode. The majority of lesions will be flush with the baby's back. A smaller number of placodes are raised far above the surface of the back by marked expansion of the subarachnoid space. Generally, however, both types are grouped under the myelomeningocele heading. In most cases, the spinal cord rostral to the neural placode is normal in gross form. Anomalies such as split cord and absence of a segment of the spinal cord can exist above the neural placode, however. Concurrent arteriovenous malformations and lipomas of the spinal cord are also possible. Occasionally, the neural placode is in a totally disorganized state. In these cases, the neural placode appears to have undergone intrauterine infarction so that portions of it are severely dysplastic and reduced to a simple membrane. This would support the concept that myelomeningocele is indeed a progressive intrauterine disease.



A



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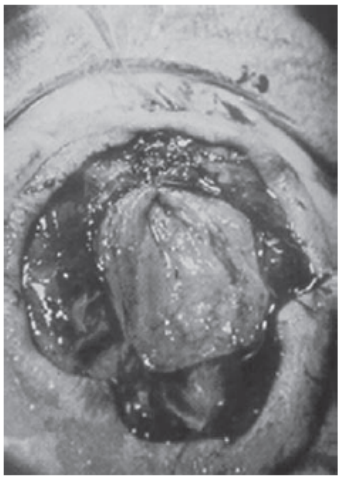

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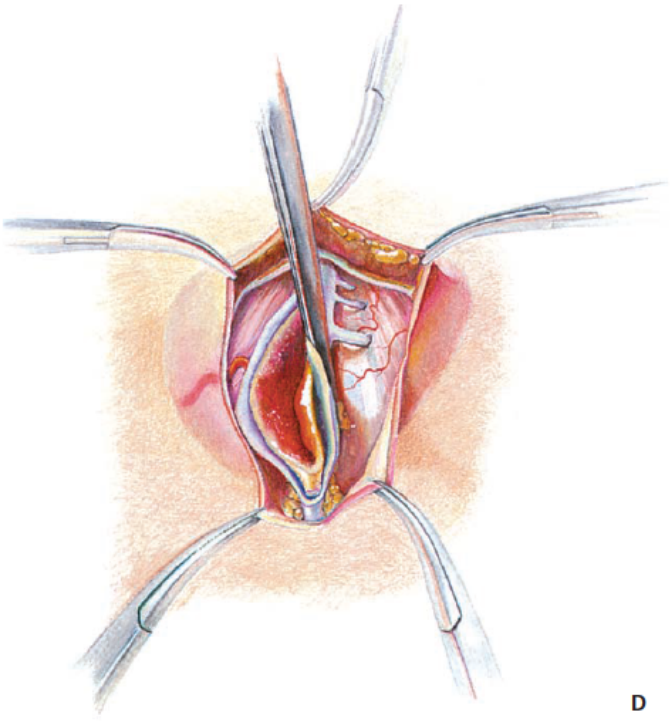
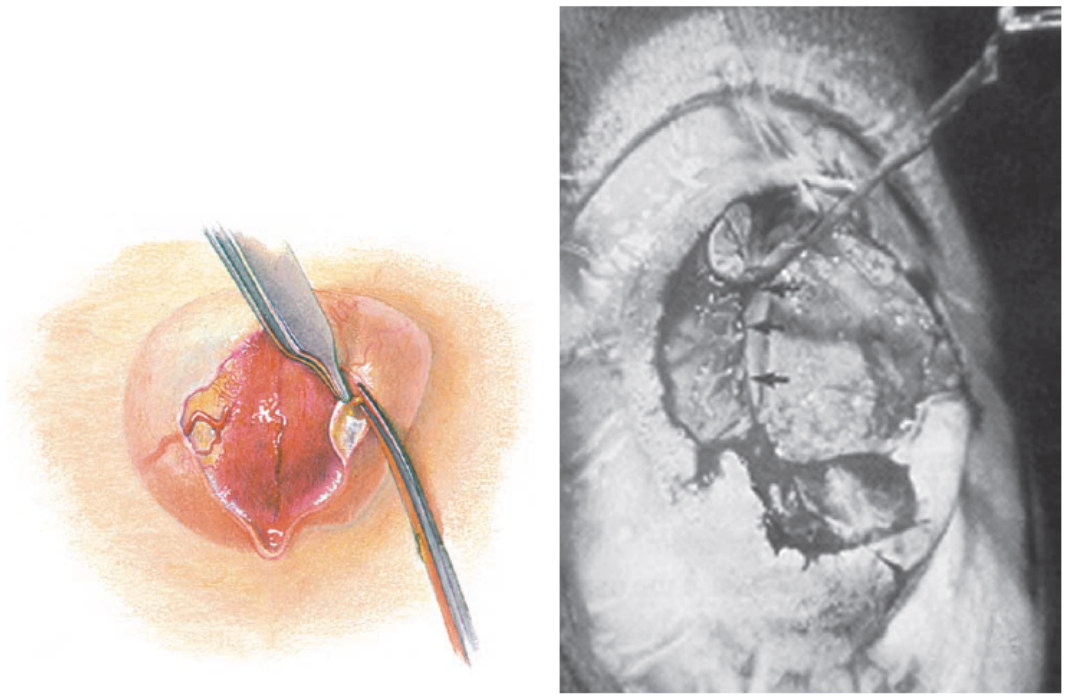


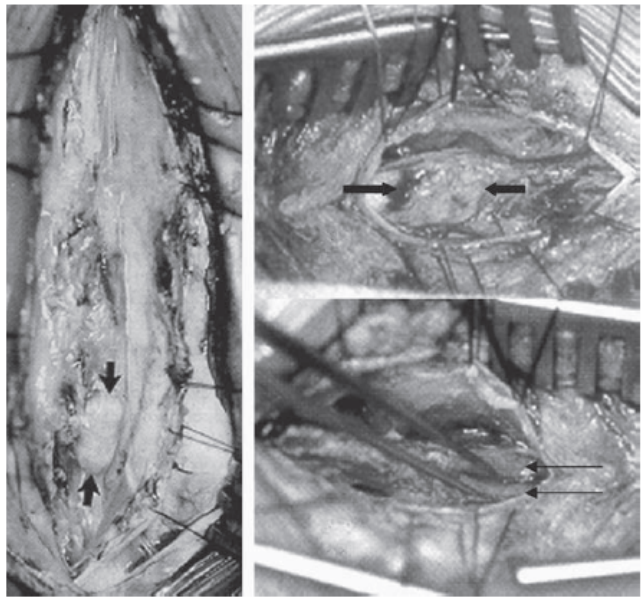
D

Figure 15–1 **(A)** Vessels entering the placode (*arrows*) are preserved during reconstruction of the neural tube. **(B)** A drawing shows the free edges of the dura being held open as the neural tube is reconstructed. The arrow indicates the point where the thickened filum was cut. **(C)** A photograph showing the beginning of reconstruction of the neural tube at the rostral portion of the myelomeningocele. **(D)** The epidural space lateral to the neural tissue is opened; it is important not to carry this dissection ventral to the neural tissue.



A B

Figure 15–2 (A) A drawing shows the incision being made at the junction of normal and abnormal thin skin. **(B)** A photograph at surgery shows the abnormal thin skin being cut free at the junction (*arrows*) of the abnormal skin and the placode.



C

Figure 15–3 (A,B) Intraoperative photographs show inclusion dermoid tumors with marked arachnoiditis. **(C)** Photograph shows a split cord malformation (*arrows*) proximal to the myelomeningocele.

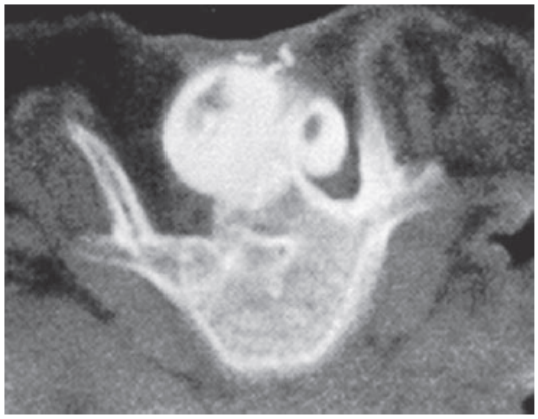


Figure 15–4 A contrast computed tomography scan shows a hemimyelomeningocele, a split cord malformation; the hemi cord on the left had a myelomeningocele.

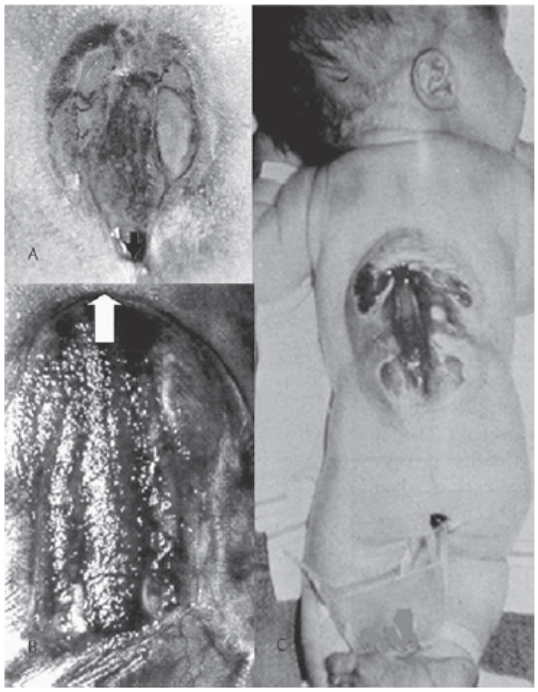


Figure 15–5 (A) A terminal myelomeningocele; the arrow indicates the area of a thickened filum terminale. **(B)** A larger myelomeningocele shows the ventral sulcus in the midline and the arrow indicates the entrance to the central canal of the adjacent normal cord. **(C)** An infant with a large thoracolumbar myelomeningocele and a kyphotic deformity at the junction of the thoracic and lumbar spines.

The functional motor and sensory levels are related to but not always consistent with the anatomical level of the lesion. Often, function is preserved below the anatomical segments involved. Again, functional asymmetry occurs and should raise the question of an additional lesion such as a split cord.

The normal anatomical structures derived from the neural tube are almost always present but are open in the midsagittal plane (unneurulated), and the dorsal roots are therefore displaced laterally. Because the neural crest is usually involved in the defective neurulation, the dorsal roots are often attenuated or absent. The exposed neural surface is the ependymal surface of the neural placode and is continuous with the central canal of the spinal cord rostrally. The lateral edges of the neural tissue are developmentally the alar (sensory) plate with the dorsal root entry zones at the lateral edge. The medial portion of the placode is basal (motor) plate and contains the anterior motor horns. Ventral to the placode along either side of the midline the motor roots exist from the placode. The sensory roots enter the cord at the periphery of the placode lateral to the motor roots. The dorsal root ganglions are usually reduced or absent, so the dorsal roots are small or absent. The ventral surface of the placode is covered with pia-arachnoid, which is directly contiguous with the arachnoid membrane of the sac. The sac usually encloses an intact subarachnoid space. An understanding of this anatomy is essential to reconstituting the spinal cord and its coverings.

Dissection of the myelomeningocele begins at the junction of the abnormal covering epithelium and the normal skin near the rostral end of the placode (**Figs. 15–2A** and **15–2B**). This junction should be incised around the entire circumference of the myelomeningocele. Once this has been completed, the dissection is carried toward the neural placode. Dividing the epithelial junction from the neural tissue requires care because on the one hand, this is the region where dorsal roots and segmental vasculature enter the neural placode, and on the other hand, any residual skin elements may grow to become inclusive epidermoid tumors (**Figs. 15–3A** and **15–3B**). The use of magnification enables one to dissect free any nerve roots that are adherent to surrounding tissues. When this has been completed, the neural tissue may float freely on an arachnoid enclosed sac of cerebrospinal fluid. Once the neural tissue is freed, every attempt should be made during anatomical reconstruction of the spinal cord to prevent later retethering of the placode.

Although pial-to-pial closure of the placode into a tubular structure has not completely prevented retethering, it may reduce the incidence of this complication (**Figs. 15–1B** and **15–1C**). More importantly, it makes untethering of the spinal cord later considerably easier to perform. The reapproximated neural tube is usually adherent only along the dorsal closure line (**Figs. 15–7A—15–7D**). In contrast, leaving the placode open allows the unclosed neural tissue to become densely adherent over the entire exposed ependymal area of the placode; the laterally displaced dorsal roots are usually caught in the scar and require tedious dissection to free them (**Figs. 15–7E—15–7G**).

The central canal is reconstructed throughout its entire length so that the neural placode becomes a tube. Closing

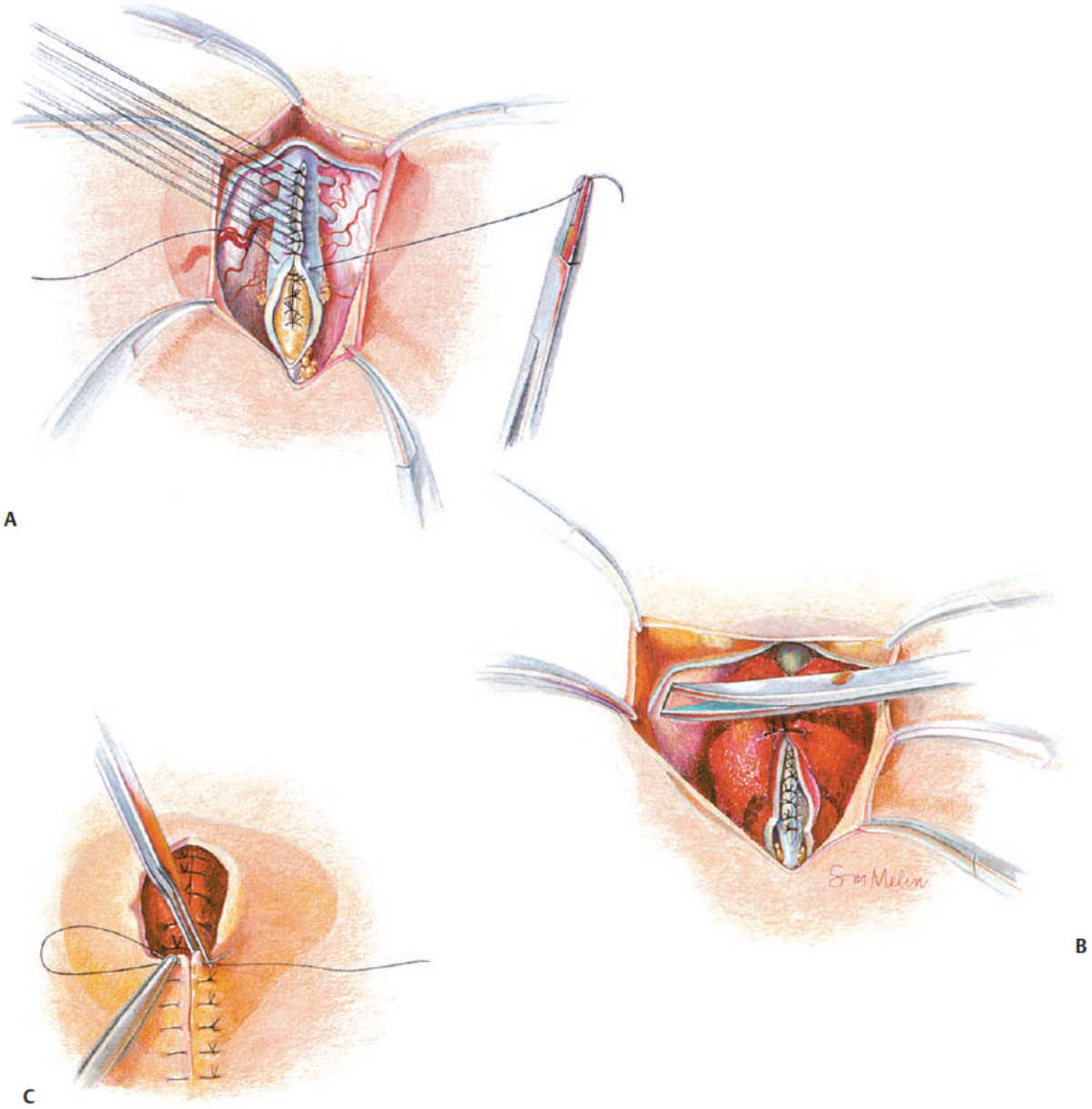


Figure 15–6 **(A)** A drawing shows the suturing of the dura. We now prefer a running locked suture rather than the interrupted suture. **(B)** Dissection in the plane between the subcutaneous fat and the muscle fascia allows the skin to be mobilized to aid closure. **(C)** A drawing shows closure of the skin.

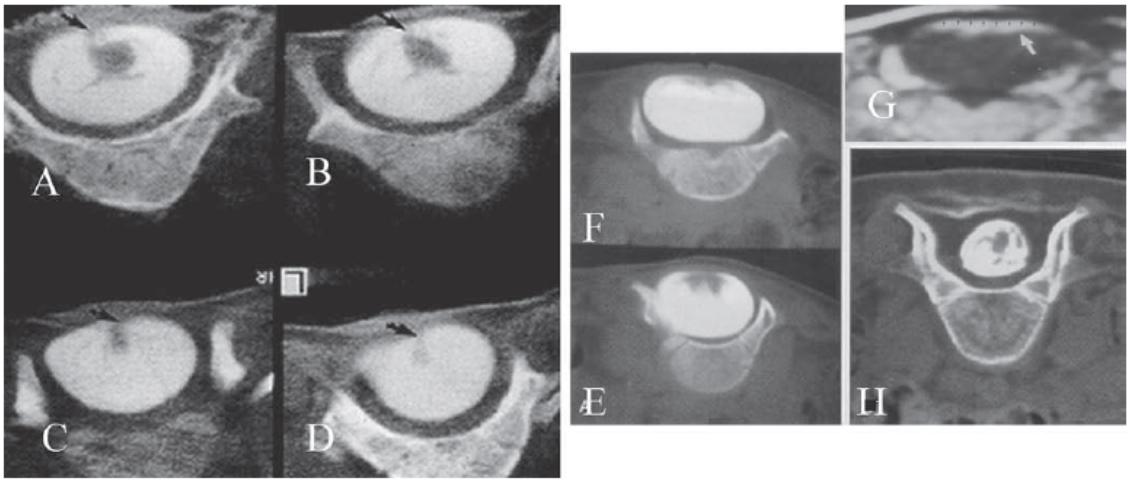


Figure 15–7 (A–D) A contrast computed tomography (CT) myelogram shows a reconstructed neural tube that has retethered along the suture line to the overlying dural closure. **(F–G)** Contrast CT myelograms showing neural placodes adherent to the overlying dural closure. **(H)** A contrast CT myelogram showing a free-floating reconstructed neural tube.

the neural placode into a neural tube and folding the arachnoid sac around the tube encloses the cord within an envelope of cerebrospinal fluid. By suspending the closed neural tube in an intact cerebrospinal fluid compartment, we hope to decrease the possibility of scarring and adherent neural elements that might later result in tethering of the spinal cord as the child grows. Magnetic resonance imaging has become the imaging modality of choice, and a postoperative study can demonstrate the reconstructed neural tube (**Figs. 15–8A—15–8D**), but not as clearly as a contrast computed tomography myelogram.

The open edges of the dura mater attach to the underside of the skin lateral to open skin edge. To ensure adequate dura mater for closure, the most lateral extent of the dura must be found and detached at that point (**Fig. 15–1B**). No dissection in the epidural space ventral to the neural tissue should be attempted (**Fig. 15–1D**). The dura is usually very thin under the spinal cord, and if torn is difficult to repair. During dural closure, the neural tissue may become included in the suture. Therefore, care should be taken to avoid this preventable complication.

Once the dura mater is free, it is closed in the midline. This layer should be closed "watertight" if possible. We prefer a running locked nonabsorbable suture of 5 or 7–0 (**Fig. 15–6A**). The dural closure must not constrict the underlying neural elements or interfere with the blood supply to the reconstructed cord. Potential recovery may be lost to ischemia or infarction if dural or fascial coverings constrict the underlying tissues.

Mobilization and midline approximation of lateral para spinal muscle fascia are optional and not essential (**Fig. 15–6B**). It may not be easy to obtain significant lateral tissues. Muscle closure at the lumbosacral level is often difficult because the fascia of sacrum and ileum are densely adherent to the bones.

Thoracic and upper lumbar myelomeningoceles can be difficult to repair if associated with a kyphotic deformity (**Fig. 15–5C**). To allow skin closure without compression may require a kyphectomy. It has the benefits of making the closure easier, giving the patient a flat back, and converting muscles from flexors of the spine to extensors, which prevents progression of the deformity.

Closure of the skin should be performed in the midsagittal plane whenever possible. Future untetherings or orthopedic procedures will be facilitated by a simple midline closure (**Fig. 15–6C**). Mobilization of the skin should also include the subcutaneous fat layer because the vascular supply to the skin comes through this layer. Blunt dissection in the plane between the muscle and subcutaneous fat is the best method to preserve the blood supply (**Fig. 15–6B**). Some consideration of cosmesis should be given here, but this is not a major consideration if it poses any added stress to neural tissue.

Hydrocephalus

The timing of shunt placement is a matter of some debate. Approximately 20 to 30% of patients with myelomeningoceles do not need a shunt, and therefore we have advocated delaying a shunt procedure until well after the initial closure. In the presence of obvious severe hydrocephalus at birth, however, it would seem to make little sense to delay and subject the patient to a second anesthetic. Placement of the shunt at the time of initial closure in these cases is safe and reduces the risk of cerebrospinal fluid leakage or wound breakdown postoperatively.

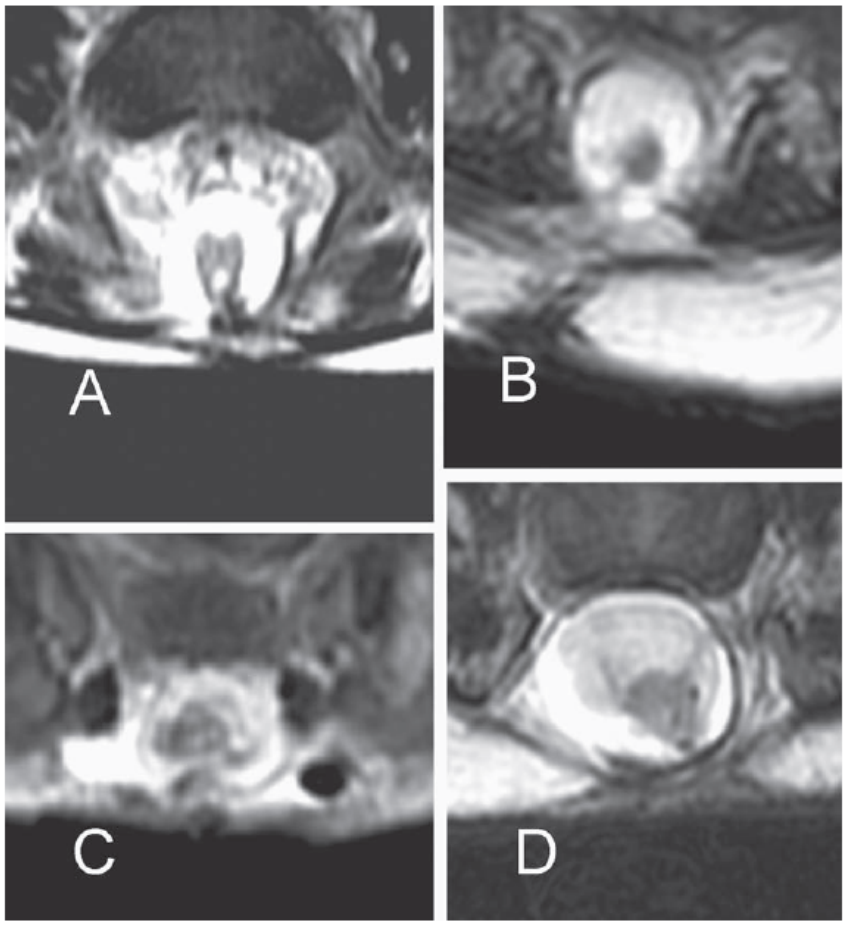


Figure 15–8 (A–D) Four neonates with myelomeningoceles. Postoperative magnetic resonance imaging axial T2 scans show the reconstructed neural tubes.

Postoperative Management

Management of the Closure Site

A variety of techniques have been employed to protect the closure site postoperatively, including placing the patient prone or suspending the patient from a sling. These maneuvers are of little value. We simply place the patient in a bassinet postoperatively and allow him or her to be held in the mother's arms without restrictions. We have not encountered any significant problems using this regimen.

Care of the Patient

At present, the patient is given intravenous fluids with 10% dextrose for the first 24 hours and then is given to the mother for feeding. Daily inspection of the closure is recommended for signs of infection, separation of the skin edges, or leakage of cerebrospinal fluid.

During the hospital stay, instruction is given to the parents to prepare them for caring for the baby in the home. This is the ideal time for the parents to become familiar with the team that will assist them in the care of the baby as an outpatient. It is essential that the parents gain confidence in their own ability to care for the baby and are aware that the team is always available for support.