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Diastematomyelia

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Diastematomyelia, or split cord malformation (SCM) type I, is a congenital spinal anomaly in which the spinal cord is divided longitudinally by a fibrous or bony septum. It has been proposed that this malformation originates from one basic error that occurs at the time of closure of the primitive neurenteric canal. Formation or persistence of an accessory neurenteric canal between the yolk sac and the amnion occurs, creating an endomesenchymal tract that splits the notochord and neural plate. This defect has been reproduced in salamanders in the study of Emura, confirming the embryogenesis.

There are two types of SCM. Type I SCM consists of two hemicords, each with its own dural tube and separated by a dura-ensheathed rigid bony (osseocartilaginous) median septum. Type II SCM consists of two hemicords within the same dural tube separated by a nonrigid, fibrous median septum. Type I SCMs are usually associated with cutaneous abnormalities at the level of the split, such as a tuft of hair, nevus, or hemangioma. Type II SCMs usually have no cutaneous abnormality at the level of the split, but spina bifida occulta is usually located in the lumbosacral region.

In many instances, syringomyelia will be recognized on the preoperative magnetic resonance imaging (MRI) in association with the diastematomyelia. We generally plan to fenestrate the syrinx at the time of untethering if it is large and in the proximity of the diastematomyelia. Otherwise, it is recommended that the syrinx be followed with interval MRI scans. The syrinx rarely requires shunting and usually does not progress if the untethering is successful.

Patient Selection

The diagnosis of SCM in the newborn usually occurs after the recognition of a midline cutaneous anomaly such as an intergluteal sinus or dimple, tuft of hair, hemangioma, cutis aplasia, or subcutaneous lipoma. It may also be seen in tandem with a myelomeningocele. The recognition of an asymmetrical lower extremity motor exam in the neonate born with a myelomeningocele should alert the clinician to the possibility of an associated SCM. These infants are often studied with spinal ultrasound, which may show a low-lying conus. After the first few weeks of life, ossification of the dorsal elements limits the use of ultrasound, therefore necessitating an MRI.

Loss of continence after toilet training, an associated history of constipation, and scoliosis are the most common symptoms beyond infancy. Complaints of intermittent back or leg pain evoked by exertion may be associated. These patients may have a normal neurological exam, but physical findings such as pes cavus deformity, calf wasting, atrophy of a gluteus muscle, asymmetry of foot size, lower sacral hypesthesia, and mild scoliosis may be common. Spinal radiographs may show spina bifida occulta and urodynamic studies should show evidence of a neurogenic bladder.

If the children have been symptomatic for 6 months or less one can generally expect improvement in their symptoms. On the other hand, those symptomatic for 1 year or more may often have stabilization of their symptoms or deficits but may have less chance of neurological improvement. Once a child becomes symptomatic, the natural history is one of symptomatic progression. The best management for asymptomatic, neurologically normal children with incidental findings on imaging remains controversial. Whether these children undergo surgical correction or not, lifelong clinical followup is advised.

Preoperative Preparation

Spinal radiographs show spina bifida occulta and widened pedicles and may define the bony spicule. Urodynamic studies commonly reveal a neurogenic bladder. Axial computed tomography (CT) scanning is important for the preoperative understanding of the bony anatomy. MRI may identify a thickened filum terminale and an associated syringomyelia, which will allow the surgeon to plan for an incision long enough for simultaneous sectioning of the filum and/or fenestration of the syrinx if it is large and in the proximity of the SCM. Careful evaluation of the imaging studies preoperatively is necessary to determine the SCM type and location of the separation of the hemicords and the bony septum. CT myelography may be necessary to visualize a meningocele manqué or a fibrous band between the two hemicords.

All children should receive an antimicrobial bath the night prior to surgery. Perioperative antibiotics are given prior to the incision.

Anesthesia Considerations

The anesthesiologist should use a short-acting muscle relaxant during induction to allow for intraoperative stimulation of nerve roots if necessary and recognition of a motor response in inadvertent stimulation of the nerve roots. Intraoperative spinal cord monitoring may be of value during this repair.

Operative Procedure

Positioning

After securing the endotracheal tube and placement of a Foley catheter, the patient is placed in the prone position on chest rolls extending from the chest to the iliac crest. This prevents abdominal compression and secondary distension of the epidural venous plexus, thus decreasing venous bleeding. The arms are placed on arm boards and all pressure points are padded. The patient is then placed in a slight Trendelenburg's position to prevent excessive loss of cerebrospinal fluid during the procedure.

Surgical Technique

Extensive preparation and draping should be performed to allow for extension of the incision as needed or placement of a second incision if the filum terminale is located a distance from the SCM. A midline incision is made down to the level of the thoracolumbar fascia (**Fig. 21–1**). The laminae at this level are always malformed and often incomplete. Careful dissection through the fascia and exposure of the bony anatomy is required to avoid inadvertent penetration through an incomplete arch and damage to the spinal cord.

Laminectomy at the level of the malformation, one level above and one level below, is often adequate for the repair. The dura of both hemicords is then exposed (**Fig. 21–2A**). The canal at this level is normally quite wide. The bony septum of the SCM can be quite vascular, which is easily controlled with bone wax and Gelfoam. The bony septum is then removed with a small rongeur (**Fig. 21–2B**) or a high-speed drill (**Fig. 21–2C**) down to the posterior longitudinal ligament. A diamond bur helps to minimize bleeding from the septum. If there is a thickened filum causing tethering of the spinal cord, it should be sectioned at the same setting. This may require a separate incision and laminectomy depending on the location.

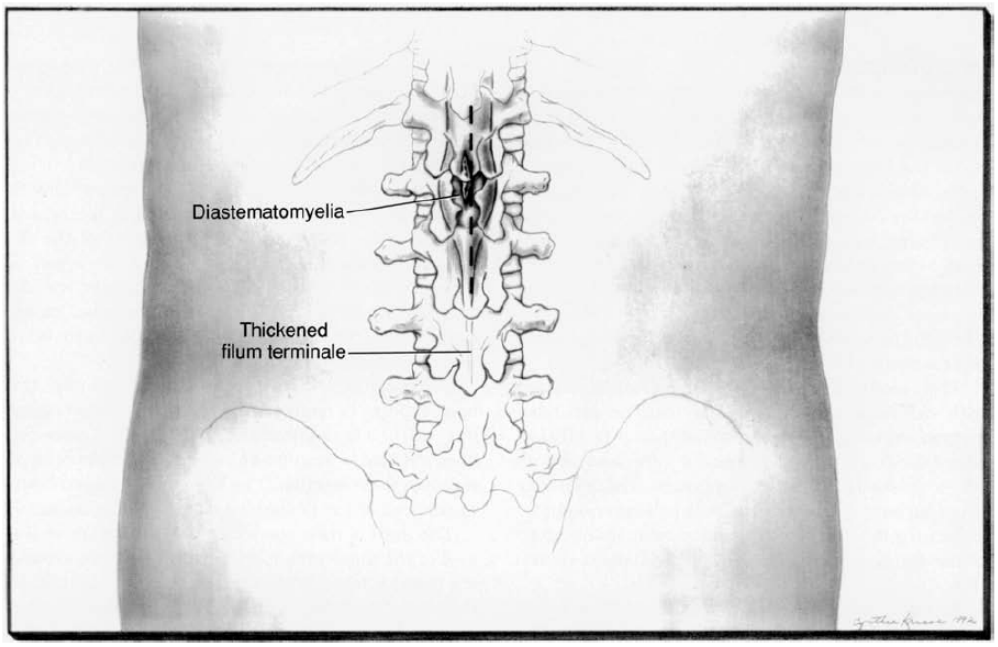

Figure 21–1 A midline skin incision (*dashed line*) is made over the level of the diastematomyelia. Note the accompanying thickened filum terminale, which may require a second incision.

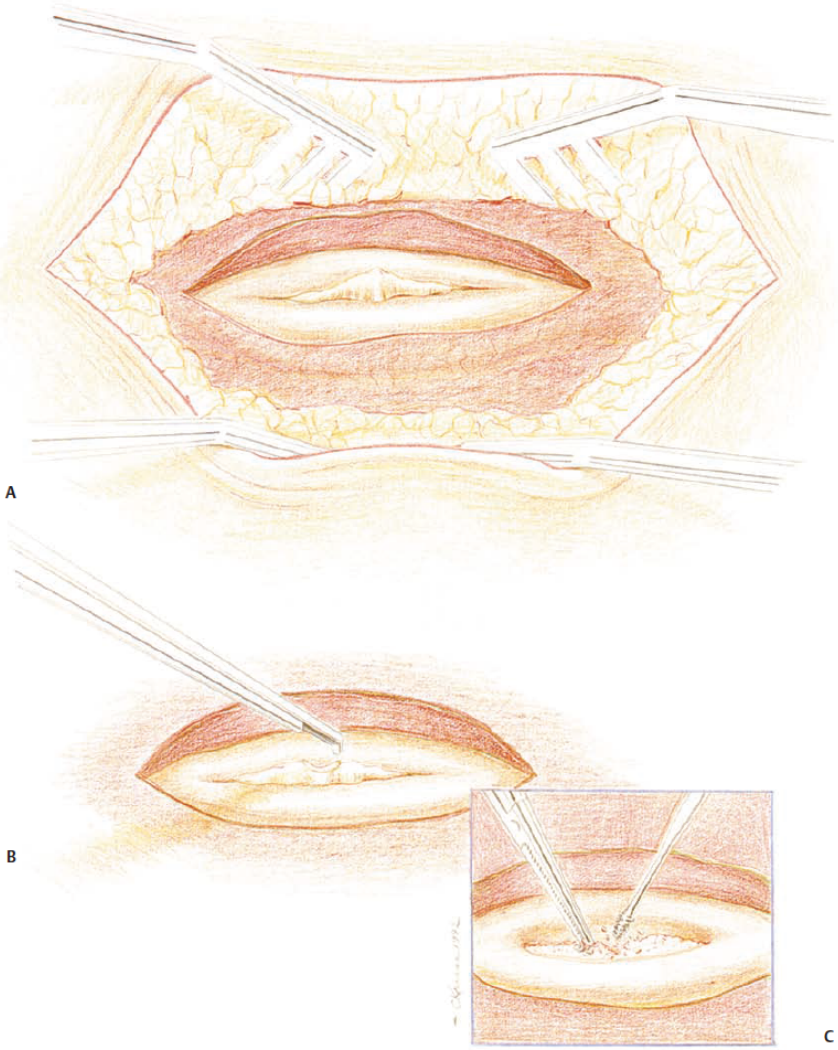

Figure 21–2 **(A)** Removal of the dysmorphic laminae allows exposure of the diastematomyelia. Periosteal elevators and other dissecting instruments should be used with care because the laminar arches may be incomplete. **(B)** The dorsal portion of the diastematomyelia septum is removed with a fine rongeur. **(C)** A high-speed air drill is used to bur away the residual septum of bone. (*continued*)

Next, the dura is opened in the midline above the level of the malformation, ellipsing the defect, leading into a midline dural incision below the level of the defect (**Fig. 21–2D**). The ventral segment of the dura separating the two hemicords is approximated, if possible, with fine prolene sutures and inverted knots. This is done in an effort to recreate the floor of the spinal canal (**Fig. 21–2E**) and prevent the regrowth of the bony septum in a young child. Next, the hemicords are explored for tethering adhesions ventral and dorsal, which are taken down with sharp dissection using microscissors and/or a knife. Strict hemostasis is obtained, the intradural space is irrigated with saline, and the dura is closed with 4–0 Gore-Tex suture (**Fig. 21–2F**). This creates one thecal sac containing both hemicords (**Fig. 21–2G**).

The epidural space is irrigated with bacitracin in saline. The epidural veins are coagulated as necessary for hemo-

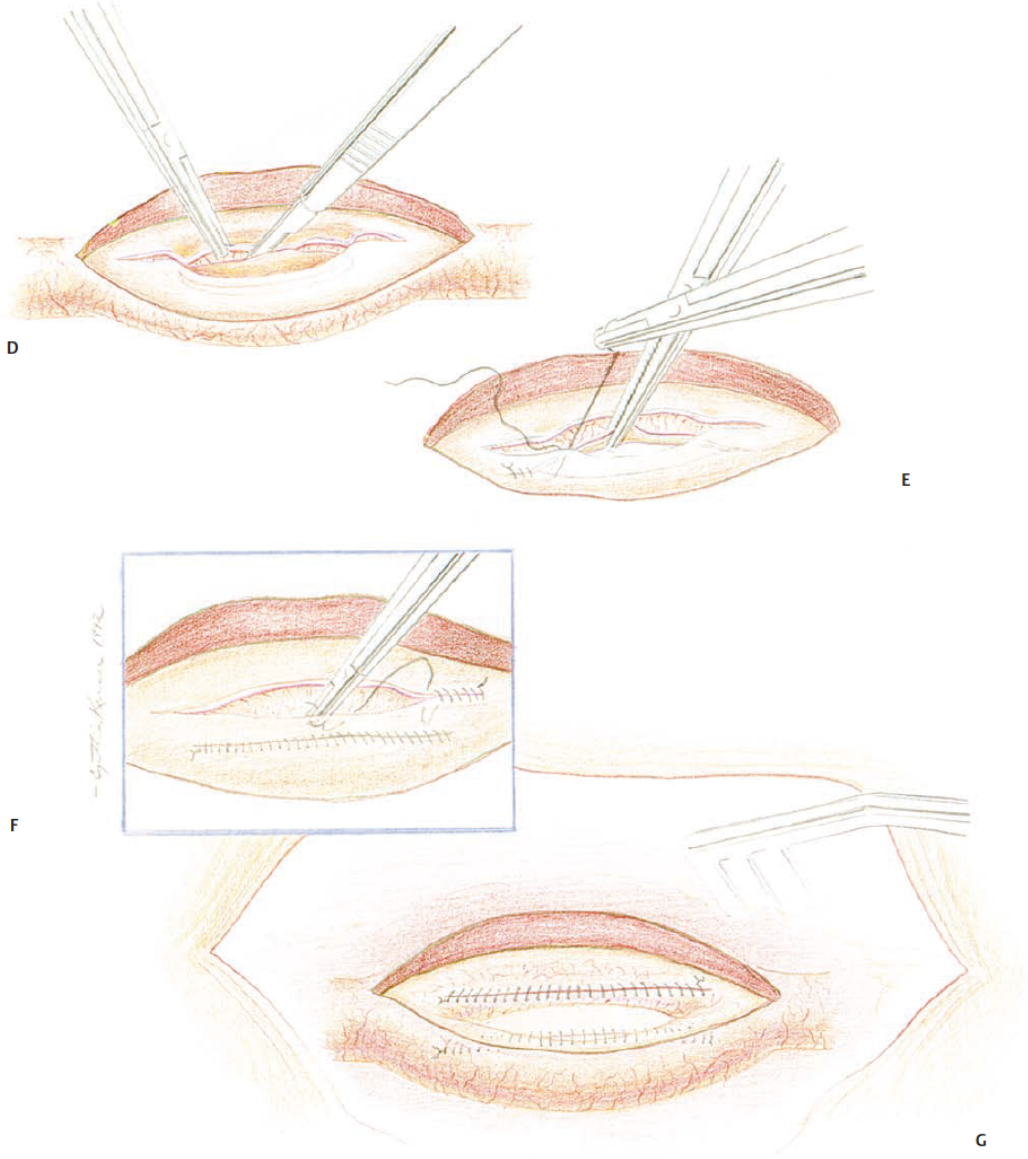

Figure 21–2 *(continued)* **(D)** Once the septum has been removed, the dura is opened in an elliptical fashion. **(E)** The ventral dura is closed primarily when possible. **(F)** Completion of dorsal dural closure. **(G)** After the ventral and dorsal incisions are closed, the two hemicords are contained within a single thecal sac.

stasis and a Valsalva maneuver is performed to ensure watertight closure of the dural repair. A dural sealant may be used as well. Next, layered closure of the paraspinous muscle and fascia is performed with interrupted absorbable sutures. The subcutaneous tissue is approximated with absorbable sutures. The skin is closed with absorbable suture and covered with Dermabond (Ethicon Inc., Somerville, NJ).

Postoperative Management Including Possible Complications

If anesthesia support is available, an epidural catheter may be placed under direct visualization at the time of closure to allow for postoperative pain control and potentially earlier mobilization. This catheter may then be removed after 24 to 48 hours, once pain is controlled by oral and/or intravenous medications. The patient is kept in the horizontal position for 24 to 48 hours postoperatively, thereby preventing spinal headaches. The patient is discharged once at baseline neurological status. It is important to remember to place all patients on a stool softener as constipation secondary to narcotic medications is a common complication. Once postoperative muscle spasm has resolved in the child presenting with scoliosis, a repeat x-ray of the spine will generally show several degrees of improvement in the scoliosis. This improvement may be significant enough to obviate the need for spinal instrumentation.

Acknowledgment

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