Surgical Treatment of Diastematomyelia

WILLIAM F. MEACHAM, M.D.
Division of Neurological Surgery, Department of Surgery, Vanderbilt University School of Medicine,
Nashville, Tennessee

DIASTEMATOMYELIA, a form of spinal dysraphism, is characterized by a division of the spinal cord or cauda equina resulting from a bony or cartilaginous spur which transfixes the neural elements and dura. Occasionally a complete septum may exist, rather than a spur, dividing the spinal canal into two separate bony compartments for a short distance (Fig. 1).

The cause of diastematomyelia is not known, but the frequency with which it is encountered in association with various forms of spina bifida and other congenital anomalies of the vertebral column suggests that it is due to a mal-arrangement of mesenchymal cells protruding into the ventral neural tissue in the early period of differentiation of the neural tube.

Transfixion of the neural tissues by the projecting spur results in a low anatomical position of the cord and impairs the normal ascent of the cord, which, by continued growth against the spur, results in a cleft or division of the dura and the neural parenchyma over a distance of several segments.

Occurring in the spinal areas from the mid-thoracic to the low lumbar segments, diastematomyelia is most frequently located in the lumbar area (Fig. 1). The disorder should be suspected in those individuals harboring any of the cutaneous signs of spina bifida occulta (midline dimpling of the skin, areas of local hypertrichosis, lipomatosous masses over the spine, and midline cutaneous vascular nevi). Neurologically, there may be disturbances of sphincter control, weakness of the distal musculature, deformities of the feet, and the absence of deep tendon reflexes in the legs. Sensory loss may be detected in the sacral (saddle) area in some patients.

The diagnosis may be made by the disclosure of a midline area of calcific density in the anteroposterior projection by x-ray, which also may indicate widening of the canal in this local area. With the use of myelography, the septum or spur can be graphically demonstrated as a central filling defect in the canal at its widest portion. Diastematomyelia must be distinguished from diplomyelia. In diplomyelia true doubling of the cord occurs, and each portion is invested with its own pial covering but shares a common arachnoid and dura; moreover, each cord is rotated approximately 90° so that the ventral columns face each other (Fig. 1). This condition has been thought to represent an incipient form of twinning. Obviously, many variations can occur, so that an individual patient may show salient features of each disorder, such as diplomyelia with intervening spur formation and separate dural compartmentation.

The surgical correction of diastematomyelia is recommended as a prophylactic measure against further progressive neurological damage. It should not be performed with the mistaken concept that dramatic reversal of the existing neurological deficits will occur, although there may be continuing improvement over a long period of time. It should also be emphasized that the neurological dysfunction which develops latest in the preoperative period is likely to show the earliest clinical improvement.

Incision

The surgical correction of diastematomyelia requires a generous exposure through a midline incision that provides access to the spinous processes and laminae for one or two segments above and below the site of the lesion (Fig. 2).

Extradural Exposure and Removal of Spur

The paraspinal muscles on each side are freed and retracted laterally as in any standard laminectomy. The spinous process and lamina at the site of the bony spicule are removed and the dura, dural cleft, and bony spur exposed. The laminectomy is then
Surgical Treatment of Diastematomyelia

Fig. 1. Mechanism of diastematomyelia.
Fig. 2. Incision and exposure of the spinous processes and laminae.
extended above and below the lesion until the dural cleft has been completely exposed; the cleft usually will extend cephalad to the spur (Fig. 3). The dura surrounding the bony spur or septum is then gently separated from the spur by blunt dissection. A septal elevator is ideal for this purpose. With a small tipped rongeur, the spur is then removed piecemeal until the base of the spur has been made flush with the anterior wall of the spinal canal (Fig. 3). Occasionally, it is difficult to remove the spur completely by this method; in such a case final removal is deferred until the dural compartment has been opened.

**Intradural Dissection and Removal of Base of Spur**

The dura is opened longitudinally above and below the cleft which is incorporated in the dural opening. All interdural adhesions at the cleft site are divided. The island of dura and the remnants of the spur can then be removed down to the level of the anterior dura.

Adhesions between the arachnoid and dura at the cleft site are carefully separated so that the two halves of the cord can be retracted to facilitate removal of the deepest portion of the base of the spur (Fig. 4).

**Closure of the Dura**

The halves of the cord, now mobilized, will juxtapose and allow the posterior dura to be closed in linear fashion. The defect in the anterior dura at the base of the spur is allowed to remain open (Fig. 5).

The remainder of the wound is closed in layers by the operator’s preferred method.

**Alternate Method for Intradural Removal**

An alternate method of management employs a wide dural opening after the exposure is considered adequate and the cord exposed well above and below the spur. The dural reflection at the cleft is then carefully divested of adhesions, mobilized, and separated from the spur. By retracting the halves of the cord away from the spur, access to the spur can be obtained from first one side and then the other, and the piecemeal removal carried out down to the anterior canal wall (Fig. 6A).

On rare occasions the bony spur may be found so attenuated at its base that a gentle rocking motion applied to the apex of the spur will break it free at the base. This, however, should not be attempted unless the surgeon is convinced that the thinness of the spur is sufficient to guarantee success.

**Special Situations**

*Asymmetrical Splitting of Dura and Cord.*

There are situations in which exposure of the area will reveal the “halves” of the cord to be quite disproportionate, so that the division is closer to one-fourth and three-fourths (Fig. 6B). The technique to be used is identical with that already described, but the surgeon must use redoubled precautions to avoid damage to the lesser segment.

*Involvement of the Cauda Equina.* Where the spicule has penetrated the area of the conus medullaris, there will be division of the terminal conus and the proximal cauda equina. One or more roots of the cauda equina may be found coursing over the spicule and adherent to it; even though these roots appear to be attenuated, severely stretched, and useless (Fig. 6C), they should be meticulously dissected free and repositioned in the dural compartment. The remainder of this special operative procedure is carried out in the fashion described.

One final word of caution. The surgeon must keep in mind throughout the operation that the handling of sensitive neural tissues already compromised in function must be done with the utmost gentleness. Excessive retraction and manipulation of the cord at the site of existing injury could tragically augment the clinical deficit already in existence. The use of small, delicate instruments, adequate lighting, generous exposure, and extremely careful technique, enhanced by the use of an operating loupe, will help produce a good result.
Fig. 3. Exposure of the dural cleft and spur; excision of the posterior portion of the spur.
Fig. 4. Division or separation of dural and arachnoidal adhesions to expose base of the spur.
Fig. 5. Closure of the dura.
Surgical Treatment of Diastematomyelia

Fig. 6. A. Intradural removal of spur, alternate method. B. Asymmetrical splitting of dura and cord. C. Involvement of cauda equina.