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Spinal Dysraphism: A Review of Clinical Manifestations and Surgical Treatment Options

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Spinal dysraphism is used to describe a group of developmental abnormalities in which there is defective closure of the neural tube. The authors review the clinical manifestations and surgical treatment of occult and open spinal dysraphism.

KEYWORDS spinal dysraphism, tethered cord, spina bifida

The term spinal dysraphism is commonly used to describe a group of developmental abnormalities in which there is a defective closure of the neural tube. Two forms of spinal dysraphism exist: a closed form referred to as spina bifida occulta, and an open form known as spina bifida aperta. In occult spinal dysraphism, there is a congenital absence of a spinous process and variable amounts of lamina resulting in neural tissue that lies just deep to an intact layer of skin. In contrast, open dysraphism is characterized by neural tissue that is exposed to the external surface of skin. Whereas spina bifida aperta is an entity that is evident at birth, spina bifida occulta frequently goes undiagnosed and may manifest later in life, if at all.[1,2] Consequently, the true incidence of spinal dysraphism is not known, though it is estimated to be approximately 0.05 to 0.25 per 1000 births.[1] Moreover, extending the term spina bifida occulta to include benign bone cleft lesions in the L5 or S1 spinous process, the estimated incidence increases to roughly 20% among the total population.[1] This article will review the more common clinical presentations of the two types of spinal dysraphism, while discussing associated neurosurgical procedures.

Embryology

Development of the central nervous system begins around day 18 postconception. At this time, a process of neurulation occurs during which a flat plate of neuroectodermal cells overlying the midline notochord of the embryo is induced to fold onto itself, creating a neural groove (Fig. 1). As the neural groove deepens, neural folds develop laterally. These folds continue to elevate, moving toward each other at the midline, until their outer edges are approximated. This results in fusion of the neural folds, thus producing a closed neural tube.[3] This closure causes the neuroectodermal cells of the neural folds to dissociate from the surrounding cutaneous ectodermal cells with which they were contiguous, in a process called dysjunction.[2,3]

The neural tube first closes in the upper cervical region, and from this point closure extends both cephalically and caudally. Complete closure of the neural tube is thought to occur in at least five waves of neurulation along the craniocaudal axis.[3] The caudal-most portion of the neural tube following this primary neurulation reaches the level of L1 or L2. The region of the neural tube that develops caudal to this is formed during secondary neurulation, which comprises a canalization process and a regression process. During canalization, the tail bud, which forms caudal to the neural tube after neurulation, develops vacuoles within it. These vacuoles coalesce to form a canal that connects the tail bud to the more rostral neural tube. Finally, in a process termed regression, the terminal filum and the cauda equina are formed from the caudal region of the neural tube.[1]

By day 52 postconception, both primary and secondary neurulation are complete. During the fetal period, growth of the vertebral canal occurs at a faster rate than growth of the neural tube, thus accounting for the relative "ascent" of the spinal cord so that the infantile conus medullaris lies at the level of L2 to L3. Defects that occur in primary or secondary neurulation give rise to the group of disorders categorized as spinal dysraphism.

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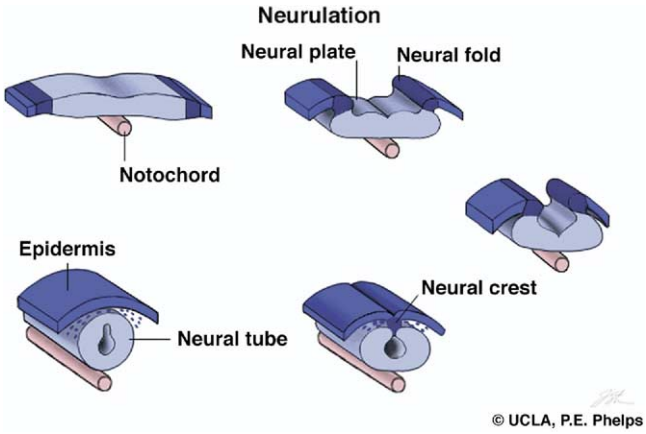

Figure 1 Neurulation: A flat plate of neuroectodermal cells is induced to fold onto itself. (Color version of figure is available online.)

Clinical Presentation and Surgical Management of Occult Spinal Dysraphism

Lipomyeloschisis

Spina bifida occulta presenting with lipomatous accumulations within the spinal cord represents the most common manifestation of closed spinal dysraphism.[2] While at least six forms have been described, only three of these will be discussed here as they hold greater clinical significance given their potential role in the etiology of a tethered cord. The first type is the lipomyelomeningocele, a subcutaneous lipoma that extends through a defect in the lumbosacral fascia and lamina, past the dura and pia mater, to compress an abnormally low spinal cord.[1] These patients often present early in life with signs of spinal cord compression. The second spinal lipoma associated with tethered cord is the intradural lipoma, which appears most commonly in the thoracic spinal cord region and, as the name implies, is an intramedullary structure. This also

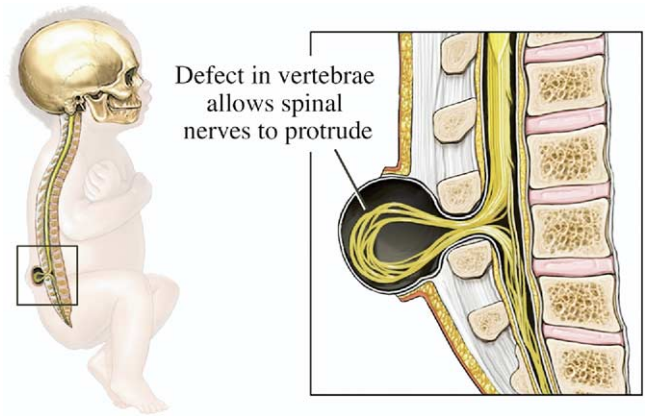

Figure 2 Open myelomeningocele. (Color version of figure is available online.)

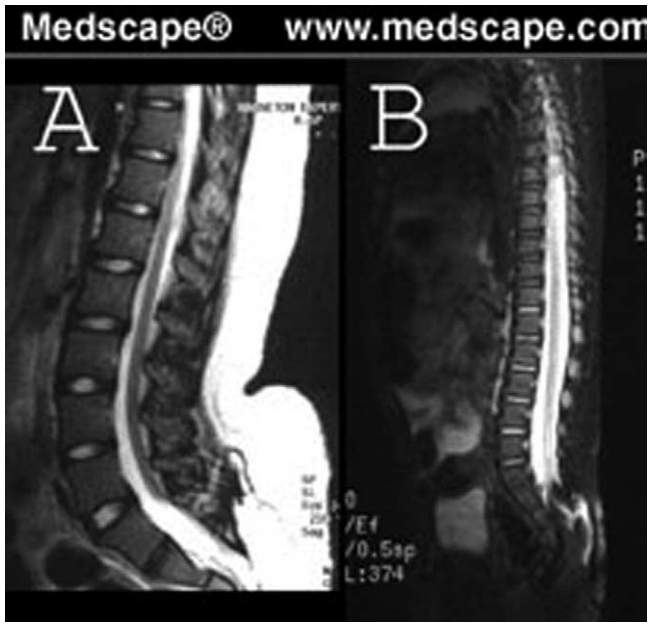

Figure 3 T2 weighted sagittal MRI showing tethered cord and lipomyelomeningocele.

manifests as spinal cord compression, yet it is not involved with cutaneous or bony anomalies. The fatty filum is the third clinically significant spinal lipoma, and it refers to a fatty infiltration of the filum terminale that can be appreciated on an unenhanced CT or an MRI.[1]

In infancy, spinal lipomas frequently present as a palpable mass in the back, and often it is not until later in life that pain, motor deficits, urinary incontinence, and foot deformities become apparent.[1,2,4,5] This delayed onset of signs and symptoms may be explained by the pathophysiology of the condition. It is possible that prolonged impairment of the mobility of the spinal cord and nerve roots results in a longitudinal stretching of the cord that compromises the blood supply to the neural tissues.[5,6] This

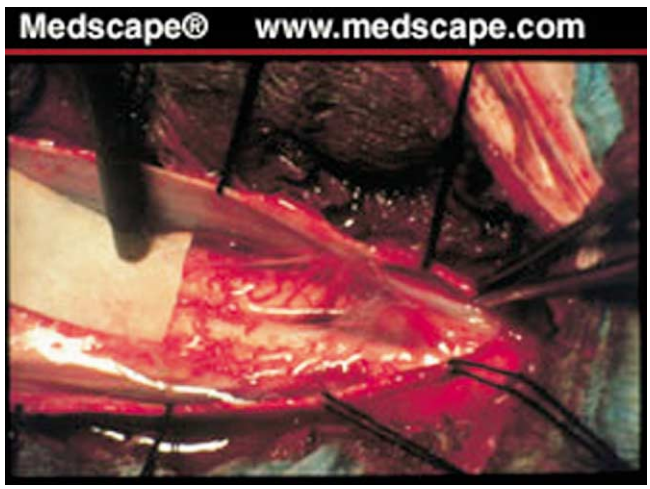

Figure 4 Intraoperative photograph of diastematomyelia. (Color version of figure is available online.)