MR Imaging of Neonatal Spinal Dysraphia: What to Consider?

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• Spinal dysraphism • Diastematomyelia • Neurenteric fistula • Embryology

The development of the spinal canal and its contents is highly complex and involves multiple programmed anatomic and functional developmental and maturational processes. These processes are tightly linked to each other and interact with each other at multiple anatomic levels simultaneously.[1–3] The association of an open (non–skin covered) lumbar myelomeningocele and a Chiari II malformation is a well-known example of this multilevel interaction. In addition, malformations of the spinal canal and cord may be an isolated process involving only the neuroaxis or may be part of a complex syndrome or malformation (eg, cloacal malformation). Finally, the malformed spinal canal and cord may be secondarily injured because of prenatal, perinatal, and postnatal complications (eg, long-standing exposure of the neural tissue to the amniotic fluid, mechanical injury during delivery, or postnatal infection).

Correct and detailed knowledge about spinal malformations is essential to understand and recognize these lesions early (preferably prenatally) to counsel the parents during pregnancy, to plan possible intrauterine treatments, and to make decisions about the mode of delivery and the immediate postnatal treatment. The impact on quality of life varies significantly depending on the kind and extent of malformation. Correct classification of the identified malformation is a sine qua non. Frequently, spinal malformations are summarized as spinal dysraphism. Dysraphism is defined as an incomplete closure of a raphe or a defective fusion. Many of the malformations indeed belong to this category; however, a variety of spinal malformations may be observed that do not directly result from an incomplete closure of the neural tube during development. As two examples, early splitting of the notochord is linked to diastematomyelia (DMM), and persistence of the notochordal process may result in neurenteric fistula. Consequently, next to a detailed knowledge about the various malformations, a basic knowledge about embryology is essential.

This article discusses the imaging findings of the most frequently encountered neonatal spinal malformations and correlates these findings with the relevant embryologic processes. The presented classification is based on a correlation of clinical, neuroradiologic, and embryologic data.[1–3]

EMBRYOLOGY

The normal development of the spinal canal and its contents relies on four principal processes: (1) gastrulation with development of the notochord; (2) primary neurulation with ganglion development;

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(3) segmentation with appearance of the somites; and (4) secondary neurulation (caudal cell mass). These processes occur in a sequential order with partial overlap.[4–7] Based on these processes, malformations are classified as (1) disorders of primary neurulation, (2) disorders of secondary neurulation, or (3) anomalies of notochordal development.

During the first 10 to 14 days the fetus consists of two distinct cell layers: the ectoderm and endoderm (**Fig. 1**). Around the 14th day of life the process of gastrulation (Weeks 2–3) starts. Ectodermal cells glide between the endoderm and ectoderm along the primitive streak, which is localized along the craniocaudal axis of the dorsum of the embryonic disk (**Fig. 2**). During this phase the embryo transforms into a trilayered organism. Most of the cells migrate laterally between the endoderm and ectoderm to form the mesoderm. While the primitive streak is developing, it thickens at its cephalic end to form a structure called the Henson node (see **Fig. 2**). A portion of the invaginating cells remains in the midline and migrates along the craniocaudal axis of the primitive streak to form the notochordal process (see **Fig. 2**). After resorption of the floor of the notochordal process the resulting prochordal plate transforms into the definitive notochord (at 20 days) (**Fig. 3**). The definitive notochord defines the primitive axis and skeleton of the embryo and is eventually replaced by the vertebral column. It extends throughout the entire embryo and reaches as far as the level of the future midbrain, where it ends in the region of the future dorsum sella. Most importantly, the notochord induces the transformation of the overlying ectoderm into neuroectoderm with formation of the neural plate (see **Fig. 3**). The notochord secretes a protein called "sonic hedgehog," which plays a critical role in signaling the development of motoneurons. This induction of the neural plate signals the beginning of the primary neurulation (Weeks 3–4). Between Days 18 and 20 the neural plate transforms into a neural groove, which starts to close into a neural tube at Day 21 of gestation (**Fig. 4**). While the neural tube is closing, the neuroectoderm progressively detaches from the adjacent surface ectoderm and "dives" into the space between ectoderm and endoderm (**Figs. 5** and **6**). The adjacent surface ectoderm closes dorsally to the neural tube (see **Fig. 6**). At the cephalic and caudal end of the neural tube the anterior and posterior neuropore close at Day 25 of gestation. Simultaneously, cells at the border of the neuroectoderm and ectoderm detach to form the neural crests (see **Fig. 6**). The neural crests subsequently fragments and give rise to the primordial of the ganglia, which again give rise to the sensory innervations. The corresponding level of the neural tube and later spinal cord furnishes the motor innervations. A somite plate

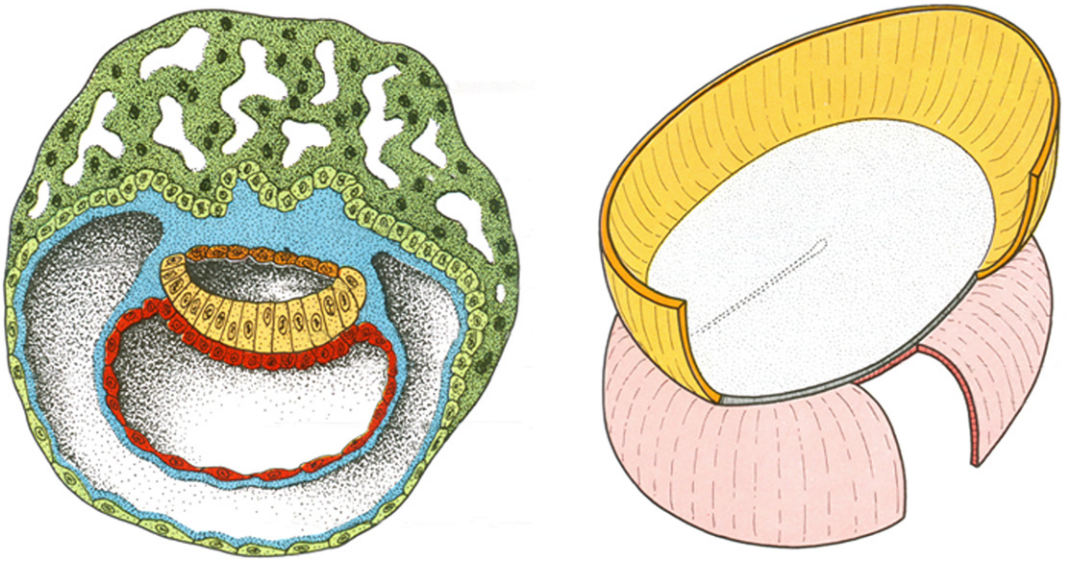

Fig. 1. During the first 10 to 14 days the embryo consists of two distinct layers: the ectoderm (*yellow*) and the endoderm (*red*). The embryonic disk is seen along the dorsum of the embryo with the developing midline primitive streak. (*From* Tuchmann-DuPlessis H, David G, Haehel P. Illustrated human embryology, embryogenesis. New York: Springer Science & Business Media (Springer-Verlag); 1982; with permission.)