margins develop laterally and fuse dorsally to form the neural tube. Neural tube formation begins in the cervical region near the center of the embryo and advances in both directions—caudally and cephalically—until by the end of the fourth week of gestation, the ends of the neural tube, the anterior and posterior neuropores, close.

Most authorities believe the primary defect in neural tube malformations is a failure of neural tube closure. However, some evidence indicates that the defects are a result of splitting of the already closed neural tube as a result of an abnormal increase in cerebrospinal fluid (CSF) pressure during the first trimester.

Etiology

There is evidence of a multifactorial etiology, including drugs, radiation, maternal malnutrition, chemicals, and possibly a genetic mutation in folate pathways in some cases, which may result in abnormal development. There is also evidence of a genetic component in the development of SB; myelomeningocele may occur in association with syndromes, such as trisomy 18, PHAVER (limb **p**terygia, congenital **h**eart **a**nomalies, **v**ertebral defects, **e**ar anomalies, and **r**adial defects) syndrome, and Meckel-Gruber syndrome (Shaer, Chescheir, and Schulkin, 2007). Additional factors predisposing children to an increased risk of NTDs include prepregnancy maternal obesity, maternal diabetes mellitus, low maternal vitamin B_{12} status, maternal hyperthermia, and the use of AEDs in pregnancy. The genetic predisposition is supported by evidence of the risk of recurrence after one affected child (3% to 4%) and a 10% risk of recurrence with two previously affected children (Kinsman and Johnston, 2016).

The degree of neurologic dysfunction depends on where the sac protrudes through the vertebrae, the anatomic level of the defect, and the amount of nerve tissue involved. The majority of myelomeningoceles (75%) involve the lumbar or lumbosacral area (Fig. 30-6). Hydrocephalus is a frequently associated anomaly in 80% to 90% of the children. About 80% of patients with myelomeningocele have an associated type II Chiari malformation (Kinsman and Johnston, 2016). There is some evidence that prolonged exposure of the myelomeningocele sac to amniotic fluid predisposes to the development of hindbrain herniation and Chiari