

CASE: TWO-MONTH-OLD CHILD WITH SPASTIC LOWER EXTREMITIES AND SWELLING ON THE BACK OF THE NECK

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T2 Weighted sagittal MR of the cervical spine. Note defect in the posterior elements from C5 through T1 (Large white arrows). Neural elements (black arrowheads) are extending through the defect into a fluid-filled sac. There is associated descent of the cerebellar tonsils, the lower limit of which lies at the level of C2.

DIAGNOSIS

Cervical spinal dysraphism with myelo-meningocele and Arnold-Chiari II malformation.

COMMENT

Spinal dysraphism results from a defect in the closure of the neural tube, which usually occurs by the 28th day of gestation. The effects are variable, with the spectrum ranging from asymptomatic spina bifida occulta (only a bony defect in the posterior elements and no associated neurological deficits) to a myelo-meningocele (all the contents of the spinal canal protrude through the defect). Prognosis depends on severity, site and adequacy of surgical closure. The defect can occur at any level in the spine, but 70% occur below the level of L2, with the higher locations associated with a worse prognosis. Cervical and sub-occipital lesions have the worst prognosis. Associated abnormalities include hydro-cephalus, Chiari II malformation, diastematomyelia, spinal dermoid/lipoma, and hydromyelia. Myelo-meningocele is the commonest congenital abnormality of the central nervous system. Fifteen percent die before their 10th birthday. About half of those afflicted have learning difficulties. Ten percent have a family history of similar defects. Presence of a neural tube defect in a sibling or parent imparts a 3-7% risk of similar occurrence in a subsequent fetus. Antenatal diagnosis is based on a combination maternal serum alpha fetoprotein levels and ultrasound examination. Prophylaxis is possible by treating women in the child bearing age with folic acid supplements.

REFERENCE

Radiology Review Manual, Dahnert W., 5th Edition. 2003. Williams & Wilkinson USA.