Pediatric Neurology: Chapter 102. Spinal cord malformations (Handbook of Clinical Neurology)

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Malformations of the spinal cord are one of the most frequent malformations. They should be clearly divided into two completely different families of malformations: open dysraphisms and occult dysraphisms. Open dysraphism mostly consists in myelomeningocele (MMC). Its incidence is 1/1000 live births with a wide variation. Folic acid supplementation has been shown to reduce its risk. In most cases, the diagnosis is done prenatally by serum screening and ultrasound and may lead to termination of pregnancy. In case of decision to continue pregnancy, surgical treatment must be achieved during the first days of life, and in 50 to 90% of cases, a ventricular shunt must be installed. The follow-up of these children must be continued throughout life looking for late complications (Chiari II and syringomyelia, vertebral problems, neuropathic bladder, tethered cord). Occult dysraphisms are a heterogeneous group of malformations. Lipomas (filum and conus) are the most frequent and their treatment remains controversial. Diastematomyelia, neurenteric cysts, dermal sinus, and more complex forms (Currarino syndrome) belong to this group. Most of them can and must be diagnosed prenatally or at birth by careful examination of the lower back for the cutaneous stigmata of the disease to decrease the risk of neurological, urological, or orthopedic permanent handicap.

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