

Commentary on:*Delayed Repair of Myelomeningoceles*by **Watson et al.** pp. 428-430.**Concezio Di Rocco, M.D.**Pediatric Neurosurgery
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Myelomeningocele: An Overview

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Myelomeningocele, the most severe form of spina bifida, is a defect of primary neurulation that results from the failed fusion in the caudal region of the neural tube, the so-called neuropore, at 25–28 days of gestation (12). The resulting unclosed segment of the neural plate (the neural placode) floats on top of a cerebrospinal fluid–filled, membrane-bound sac (9). Established risk factors, such as previous affected pregnancies, inadequate maternal intake of folic acid, pregestational diabetes, use of some antiepileptic drugs, and some genetic pathways, such as folate–homocysteine pathway genes, have been linked to primary neurulation failure. Other suspected maternal risk factors are vitamin B₁₂ status, obesity, hyperthermia, and diarrhea (10).

Prevalence varies across time, by region, and by both race and ethnicity, with a general worldwide decreasing incidence trend since the early 1980s (10). This is generally attributed to improved nutrition and dietary fortification and, in developed countries, to highly predictive prenatal testing (maternal serum α -fetoprotein, ultrasound, amniocentesis, and prenatal magnetic resonance imaging), resulting in selective abortion (2).

Neurologic impairment is primarily related to the arrested development of the neural placode, often resulting in motor and sensory deficits in the lower limbs and in bladder/bowel continence deficits. Secondary neurologic deficits may be often linked to Chiari II malformation, hydrocephalus, syringomyelia, and scoliosis. In long-term surviving patients, further impairments can result from spinal cord tethering and orthopedic problems such as foot or ankle deformities, dislocated hips, and joint tightness or contractures. Moreover, untreated open spinal dysraphisms have a high rate of infections and subsequent meningitis.

In the past few decades, controversies have been raised on the optimal management of children affected by myelomeningocele, ranging from prenatal surgery to post-natal selective treatment (7). Even an active termination of life has been discussed in very severe forms of myelodysplasia (15). However, the great majority of the neurosurgical community, as well as the members of concerned societies, currently agrees that children affected by myelomeningocele should undergo an early surgical repair of the lesion. The goals of surgical management are preserving the functional neural tissues, reducing the risk of cerebrospinal fluid leaks and infections, and avoiding spinal cord tethering. The usual timing for surgical treatment is within 48–72 hours from birth (8), in order to minimize the risk of infections and further damage to exposed neural structures (13). Although the typical scenario is a delayed-onset hydrocephalus, if macrocrania, tense fontanelle, and ventriculomegaly are evident at birth, concomitant ventriculoperitoneal shunting or endoscopic third ventriculostomy should be considered.

Prenatal repair of myelomeningocele has gained popularity since it was first reported in 1994 (4), with the rationale that an early repair would protect the neural tube from the injury resulting from prolonged exposure of neural elements to the intrauterine environment. According to the “two-hit” theory (6), the deterioration in lower limb movements observed in sonograms of an affected fetus after 17–20 weeks of gestation could be the result of exposure of neural tissue to amniotic fluid, meconium, or direct trauma. In utero repair is usually performed by hysterotomy because the early experiences with endoscopy were unsatisfactory. Recently, a randomized trial of prenatal versus postnatal repair of myelomeningocele (1) showed a reduced need for

Key words

- Myelomeningocele
- Pediatric neurosurgery
- Spina bifida
- Spinal dysraphism



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shunting and improved motor outcomes at 30 months and higher maternal and fetal risks in prenatally treated patients.

In developed countries, delayed repair of myelomeningocele is therefore confined to rare instances such as postponed surgery for infections, parental denial of early treatment, or children who migrated from developing countries with still insufficient expertise and inadequate facilities (5).

Some interesting techniques have been proposed to face such a late repair, especially in the case of a large myelomeningocele, like the use of tissue expanders (11) or modified skin flaps, including the epithelium overlying the dysraphism (5). In this issue of **WORLD NEUROSURGERY**, Watson et al. describe the surgical technique they developed to treat 97 children affected by myelomeningocele untreated at birth in a 10-year experience of medical missions in Guatemala. A first aspect to be considered is that, even if left untreated and probably with inconsistent medical assistance for at least 6 months from birth, presumably many patients were alive, in contrast to Lorber's reported mortality rate in untreated patients during his "selective treatment" policy trial (7).

The paper by Watson et al. focuses on the technical aspects of the delayed closure of myelomeningocele and includes no details on the clinical and neurologic status of the children, apart from the need for shunting in the majority of cases. As well outlined in the paper, delayed myelomeningocele closure differs from

perinatal closure, because the sac is epithelialized and may have undergone changes associated with previous infections. The key point of the procedure is, indeed, the creation of a dissecting plane between the reepithelialized skin and the placode in order to excise the former, to perform an optimal skin closure, and to preserve the latter from delayed injuries. Actually, the goals of delayed treatment are to prevent the deterioration of the neurologic deficits related to spinal cord tethering and to avoid sac rupture. No data except the early postsurgical complications are available on the follow-up of patients of this case series.

However, in spite of the above-mentioned, ethically questionable positions on the selective treatment or euthanasia of these children, 75% of the patients treated at birth can reach their early adult years, with 85% of these survivors either attending or having graduated from high school or college, 80% being able to maintain social bladder continence via clean intermittent catheterization, nearly 90% reporting acceptable levels of bowel continence, and 75% being able to walk, often requiring braces and crutches (3). Intelligence Quotient levels above 80 (normal intelligence) are reported in 75% of shunt-free patients. However, 86% of surviving patients are shunt dependent, 60% of them having a normal intelligence (14). Shunt malfunction, problems associated with Chiari II malformation, respiratory failure secondary to chest deformity, urinary sepsis, and more rarely, cardiac or renal failure, are the most frequent causes of death in childhood or young adulthood in myelomeningocele patients.

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