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Fetal surgery for myelomeningocele is effective: a critical look at the whys

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Abstract

Formerly, the disastrous cluster of neurologic deficits and associated neurogenic problems in patients with myelomeningocele (MMC) was generally thought to solely result from the primary malformation, i.e. failure of neurulation. Today, however, there is no doubt that a dimensional additional pathogenic mechanism exists. Most likely, it contributes much more to loss of neurologic function than non-neurulation does. Today, there is a large body of compelling experimental and clinical evidence confirming that the exposed part of the non-neurulated spinal cord is progressively destroyed during gestation, particularly so in the third trimester. These considerations gave rise to the two-hit-pathogenesis of MMC with non-neurulation being the first and consecutive in utero acquired neural tissue destruction being the second hit. This novel pathophysiologic understanding has obviously triggered the question whether the serious and irreversible functional loss caused by the second hit could not be prevented or, at least, significantly alleviated by timely protecting the exposed spinal cord segments, i.e. by early in utero repair of the MMC lesion. Based on this intriguing hypothesis and the above-mentioned data, human fetal surgery for MMC was born in the late nineties of the last century and has made its way to become a novel standard of care, particularly after the so-called "MOMS Trial". This trial, published in the New England Journal of Medicine, has indisputably shown that overall, open prenatal repair is distinctly better than postnatal care alone. Finally, a number of important other topics deserve being mentioned, including the necessity to work on the up till now immature endoscopic fetal repair technique and the need for concentration of these extremely challenging cases to a small number of really qualified fetal surgery centers worldwide. In conclusion, despite the fact that in utero repair of MMC is not a complete cure and not free of risk for both mother and fetus, current data clearly demonstrate that open fetal-maternal surgery is to be recommended as novel standard of care when pregnancy is to be continued and when respective criteria for the intervention before birth are met. Undoubtedly, it is imperative to inform expecting mothers about the option of prenatal surgery once their fetus is diagnosed with open spina bifida.

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