Effects of prenatal myelomeningocele closure on the need for a CSF shunt

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Management of Myelomeningocele Study (MOMS) investigators analyzed updated data on the effects of prenatal myelomeningocele closure on the need for placement of a cerebrospinal fluid (CSF) shunt within the first 12 months of life. These researchers reaffirm the initial MOMS finding that prenatal repair of a myelomeningocele results in less need for a shunt at 12 months and introduce the new finding that prenatal repair reduces the need for shunt revision in those infants who do require shunt placement. The researchers also found that patients with extensive hydrocephalus at the time of the prenatal evaluation did not show improved outcomes following prenatal surgery.

Full details of the new analyses can be found in "Prenatal surgery for myelomeningocele and the need for cerebrospinal fluid shunt placement" by Noel Tulipan and colleagues, published today online, ahead of print, in the *Journal of Neurosurgery: Pediatrics*.

The researchers present final outcome data for the Management of Myelomeningocele Study (MOMS), a multicenter randomized clinical study in which the safety and efficacy of prenatal and postnatal closure of myelomeningoceles were compared. Preliminary data from this study on 158 infants were published in The *New England Journal of Medicine* (*NEJM*) in 2011; the present paper contains final outcome data on the need for cerebrospinal fluid (CSF) shunt placement up to 12 months of age for all 183 infants included in the study.

Myelomeningocele is the most common and severe form of spina bifida,

a neural tube defect in which spinal vertebrae fail to develop normally during the first few weeks of gestation. In the normal spine, vertebrae encircle the spinal canal, a passageway from the base of the skull to the sacrum. The spinal canal is lined by membranes, known as meninges, which contain the spinal cord surrounded by CSF. In cases of myelomeningocele, some vertebrae are malformed, leaving a gap in the bones that allows a portion of the cord, CSF, and meninges to bulge outward and form a pouch beyond the infant's skin. This pouch provides little protection for the spinal cord and nerves. Damage to the cord and nerves can occur before birth as well as afterward, and can result in sensory or motor nerve deficits as well as loss of bladder or bowel control.

A common consequence of myelomeningocele is hydrocephalus, a buildup of CSF in the brain. Hydrocephalus can cause debilitating symptoms, such as intellectual and motor disabilities, and if left untreated can even lead to death. Hydrocephalus is usually treated by placement of a shunt to divert excess CSF from the brain to other parts of the body, where it is subsequently reabsorbed by the body. In the present study, the researchers examined what benefits prenatal surgical closure of the myelomeningocele has on the need for shunt placement during infancy in patients with spina bifida.

The *NEJM* report was based on 158 cases and evaluated the primary composite outcome of fetal loss, neonatal death, or the need for a CSF shunt by the time the infant reached 12 months of age (either actual shunt placement or meeting the prespecified criteria for shunt placement). The researchers demonstrated that prenatal repair of the myelomeningocele resulted in less need for a shunt at 12 months of age.

In the present analysis, the researchers examined updated data in all 183 children (91 who were treated prenatally and 92 who were treated after birth). The primary outcome was met significantly less frequently in

infants whose myelomeningocele had been repaired prenatally (73% compared with 98% in the postnatal treatment group). The researchers also found that the actual rates of shunt placement were lower in infants treated prenatally (44% compared with 84% in the postnatal treatment group), as were the rates of shunt revision among those infants who had already received a shunt (15% in the prenatal treatment group compared to 40% in the postnatal treatment group).

The researchers found that a substantial number of infants met the criteria for shunt placement but did not receive one. Therefore the researchers redefined the original study's criteria for shunt placement based on the more overt signs of hydrocephalus that are now used by surgeons to decide when a shunt should be inserted. The researchers then redefined the primary outcome based on the new criteria. The new primary outcome was also met significantly less frequently in infants whose myelomeningocele was repaired prenatally (50% in the prenatal group and 87% in the postnatal group).

The researchers found two prenatal characteristics that had a significant impact on the effectiveness of prenatal surgery: the size of the lateral ventricles within the brain at the time of prenatal screening ultrasonography (19-25 weeks gestational age) and the gestational age at enrollment. Among infants whose ventricles measured less than 10 mm at the time of screening, 20% in the prenatal group and 79% in the postnatal group received shunts by 12 months of age; similarly, in infants whose ventricles had measured 10 mm up to 15 mm, 45% in the prenatal group and 86% in the postnatal group received shunts during their first year of life. No significant advantage of prenatal surgery was identified in infants whose ventricles measured 15 mm or larger at the time of screening: 79% of infants in the prenatal group and 87.5% in the postnatal group later received shunts. The researchers also found that for those infants enrolled at 23 weeks of gestation or earlier the shunt placement rate was 31% in the prenatal repair group and 86% in the

postnatal repair group while those enrolled at 24 weeks or later had shunts placed at a rate of 60% in the prenatal group and 82% in the postnatal group. Similar results were found for the new primary outcome.

Finally, the researchers examined several risk factors present at the time of screening in patients randomized to prenatal repair that indicated the need for future shunt placement. In the prenatal surgery group, larger ventricles were associated with an increased rate of shunt placement and meeting the new primary outcome.

Ventricles increase in size throughout fetal development in patients with spina bifida. Based on the findings, the researchers suggest that it may be best to perform prenatal surgery early—when the ventricles are still small. They state that fetuses with ventricles smaller than 10 mm are the best candidates for prenatal surgery and that perhaps this procedure should take place earlier than 20 weeks. The researchers also advise that caution be exercised when recommending prenatal closure of a myelomeningocele in a fetus whose ventricles are 15 mm or larger, because of the limited advantage of prenatal surgery in this group.

More information: Tulipan N, Wellons JC III, Thom EA, Gupta N, Sutton LN, Burrows PK, Farmer D, Walsh W, Johnson MP, Rand L, Tolivaisa S, D'Alton ME, Adzick NS, for the MOMS Investigators: Prenatal surgery for myelomeningocele and the need for cerebrospinal fluid shunt placement *Journal of Neurosurgery: Pediatrics*, published online, ahead of print, September 15, 2015; DOI: 10.3171/2015.7.PEDS15366

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