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Maternal, Fetal, and Neonatal Care in Open Fetal Surgery for Myelomeningocele

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ABSTRACT

Fetal myelomeningocele closure (fMMC) was demonstrated in a randomized, prospective clinical trial to improve outcomes for children diagnosed prenatally. Complex care of the maternal/fetal dyad undergoing fetal surgery requires a well-coordinated multidisciplinary team. Nurses in many roles are essential members of the team that cares for these women across the continuum. In this article we discuss the care of the woman, fetus, and family from initial contact through the discharge of the neonate.

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Spina bifida is a general term used to describe any birth defect involving incomplete closure of the neural tube. The neural tube is the embryonic structure that develops into the fetal brain, spinal cord, and the meninges that surround and protect the brain and spinal cord. In spina bifida, the neural tube fails to close in the first trimester of pregnancy. Myelomeningocele is the most common and severe form of spina bifida, occurring in 3.4 per 10,000 live births in the United States (Boulet et al., 2008). Myelomeningocele is characterized by defects in the skin, vertebrae, and meninges resulting in exposure of the spinal cord to the intrauterine environment. The open lesion may be flat, or the meninges and spinal cord may protrude in a sac on the fetal back resulting in nerve damage below the level of the lesion and in most cases development of the Arnold-Chiari II malformation. The Arnold-Chiari II malformation encompasses a group of anomalies including hindbrain herniation (displacement of the cerebellum into the spinal canal) that can lead to hydrocephalus and is associated with developmental brain abnormalities. Brain stem dysfunction related to hindbrain herniation includes apnea, bradycardia, vocal cord paresis, and poor or absent suck and swallow coordination. Children with spina bifida often experience lifelong disabilities including paralysis of the lower extremities related to the location of the lesion, lack of bladder and bowel control, and learning disabilities.

Standard ultrasound surveillance in pregnancies complicated by fetal myelomeningocele suggest that damage to the spinal cord and development of the Arnold-Chiari II malformation are progressive, as noted by loss of lower limb movement and increasing hydrocephalus. Using animal models, the progressive damage has been demonstrated to be related to exposure of the spinal cord to potentially harmful elements in the amniotic fluid and trauma to the exposed spinal cord (Meuli et al., 1995; Meuli et al., 1996).

Maternal /fetal surgery for myelomeningocele was developed to decrease the progression of neurologic damage to the fetus. The spinal cord is closed to prevent leakage of the cerebral spinal fluid and the exposure of the nerve roots to the amniotic fluid. This is done in an effort to improve quality of life for the child and family. Historically, the risk of performing open fetal surgery was considered less than the risk of the impending morbidity of the fetus and the mother. More than 200 mothers

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received maternal/fetal surgery for spina bifida in the United States between 1997 and 2003, and the findings were compared against historical controls (Danzer, Gerdes, et al., 2009; Fichter et al., 2008; Johnson et al., 2006). Promising results from these cases (Danzer, Finkel, et al., 2009; Fichter et al.; Johnson et al.) led to the development of the Management of Myelomeningocele Study (MOMS), a prospective randomized clinical trial sponsored by the Eunice Kennedy Shriver National Institute of Child Health and Human Development. The trial compared fetal myelomeningocele closure (fMMC) to standard postnatal myelomeningocele closure (MMC) in the neonate. The trial began enrolling participants in March 2003 and was conducted at three fetal treatment centers. The goal was to enroll 200 women and their children in the study: 100 receiving prenatal surgery and 100 receiving postnatal repair. Due to the demonstrated efficacy of prenatal surgery within the intervention group, the clinical trial was suspended early in December 2010 after enrolling only 183 women. The initial findings showed a decrease in the need for ventriculoperitoneal (VP) shunt placement by the age of 12 months and improved ambulation at 30 months (Adzick et al., 2011).

One of the most promising findings associated with fMMC closure is the improvement in hindbrain herniation (Danzer, Finkel, et al., 2009). Unfortunately, the surgery cannot restore lost motor or nerve function in the lower spine and is associated with increased maternal morbidity and rate of prematurity (Adzick et al., 2011). Although fMMC has been shown to improve short-term outcomes for some children, not every child benefits from fMMC closure, nor is every woman carrying a fetus with spina bifida a candidate for fetal surgery.

Due to the findings of the MOMS trial, open maternal/fetal surgery can now be offered for a non-life-threatening condition with potentially significant benefits for the fetus or child. In addition, the MOMS trial investigators developed standards of care that were closely evaluated for safety and efficacy. To duplicate these positive outcomes in other clinical environments, it is important that the standards of care developed by the protocol be implemented (Deprest et al., 2010).

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Initial Contact with Fetal Center

A large number of women currently elect prenatal first- and second-trimester screening that ultimately allows for early prenatal diagnosis of fetal anomalies such as myelomeningocele. When a fetal diagnosis of myelomeningocele is rendered, the family may be referred to a fetal treatment center by their care provider or self-referral.

The woman's initial contact with the fetal center is a lengthy telephone screen with a nurse coordinator to determine eligibility for an evaluation at the fetal center. The screening begins with an assessment of the family's anxiety level, their understanding of the diagnosis, and their readiness to learn. A basic maternal medical, surgical, and obstetric history and a psychosocial assessment are obtained during the initial screening. Information about family support, other children, the woman and her partner's careers, marital status, and financial stability is equally important in assessing a family's ability to pursue fMMC. This phone call also allows for an opportunity for the nurse coordinator to provide support and basic education to the family. The pathophysiology of myelomeningocele, outcomes based on lesion level, degree of ventriculomegaly, the Arnold-Chiari II malformation, and the need for placement of a ventriculoperitoneal (VP) shunt is reviewed. A brief discussion of fMMC closure may ensue with emphasis placed on two significant results of the MOMS trial: (a) reversal of the hindbrain herniation that resulted in a decreased need for VP shunting and (b) improved mobility. Additional results derived from the MOMS trial including outcomes and risks for prematurity are reviewed. Clarification of misinformation is imperative. Families need to understand that fMMC will not "cure" spina bifida. Fetal myelomeningocele closure is not to be undertaken to prevent mortality, but to possibly prevent the lifelong morbidities associated with open spina bifida (Bebbington, Danzer, Johnson, & Adzick, 2011). The family needs to recognize that their child will always have spina bifida and may still have special needs. The woman and her family are directed to view an educational DVD on the web at http://www.fetalsurgery.chop.edu. In addition, educational material regarding the results of the MOMS trial is mailed to the woman allowing them to review the information prior to their evaluation.

If relocation of the family for evaluation and possible fMMC procedure will be necessary, travel, lodging, and resources are facilitated and coordinated. These arrangements may be expedited

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