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WOMEN'S HEALTH CARE PHYSICIANS

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Committee on Obstetric Practice

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Maternal–Fetal Surgery for Myelomeningocele

ABSTRACT: Myelomeningocele, the most severe form of spina bifida, occurs in approximately 1 in 1,500 births in the United States. Fetuses in whom myelomeningocele is diagnosed typically are delivered at term and are treated in the early neonatal period. A recent randomized controlled trial found that fetal surgery for myelomeningocele improved a number of important outcomes, but also was associated with maternal and fetal risks. Maternal–fetal surgery is a major procedure for the woman and her fetus, and it has significant implications and complications that occur acutely, postoperatively, for the duration of the pregnancy, and in subsequent pregnancies. Therefore, it should only be offered at facilities with the expertise, multidisciplinary teams, services, and facilities to provide the intensive care required for these patients.

Myelomeningocele, the most severe form of spina bifida, occurs in approximately 1 in 1,500 births in the United States and is complicated by hydrocephalus, the need for a ventriculoperitoneal shunt, motor and cognitive defects, bowel and bladder injury, and social and emotional challenges. The extent of the effect of the condition is related to the level of the myelomeningocele defect; a higher level of lesion affects more nerves and results in more deficits. Fetuses in whom myelomeningocele is diagnosed typically are delivered at term and are treated in the early neonatal period.

Fetal surgery typically has been undertaken as a heroic intervention because of the risks for the fetus and the mother. Open fetal procedures have been considered and offered when the fetal condition is life threatening, and the intervention is felt to be the only option for fetal survival. Maternal–fetal surgery was considered in this nonlethal condition because the results of laboratory and animal studies demonstrated that repair of myelomeningocele during pregnancy may improve the outcome in these pregnancies. These results were likely in part by normalizing the anatomy and decreasing the exposure of the spinal cord to the amniotic fluid. Early work in human pregnancies demonstrated feasibility, and a randomized, controlled trial was undertaken to evaluate the safety and efficacy of maternal–fetal surgery for myelomeningocele (1). The support of the perinatal and maternal–fetal surgery communities was key to the success of this trial because a moratorium was in place on this procedure throughout the trial.

The authors (1) found that fetal surgery for myelomeningocele improved a number of important outcomes, but it also was associated with maternal and fetal risks. Prenatal repair before 26 weeks of gestation resulted in a reduction in the rate of death or the need for a shunt at 12 months of age, decreased the rate of hindbrain herniation by one third at 12 months, doubled the rate of the ability to walk independently, and produced a level of function that was two or more levels better than expected according to anatomic levels. However, prenatal surgery also was associated with high rates of preterm birth, fetal bradycardia, oligohydramnios, placental abruption, pulmonary edema, maternal transfusion at delivery, and an increased incidence of uterine thinning or dehiscence at the uterine scar, which occurred in 35% of the cases. The latter is likely because of the location of the fetal surgery incision on the uterus. The location of the incision typically is through a muscular portion of the uterus and may be posterior or fundal, depending on the location of the placenta. Long-term follow-up of these children at 5–8 years of age, including urologic outcomes, is ongoing.

The trial was undertaken in a rigorous fashion; therefore, the outcomes are likely the best-case scenario. All sites had experienced fetal surgeons, multidisciplinary teams, and state-of-the-art equipment. To participate in the trial, an experienced fetal surgeon was required to

Table 1. Management of Myelomeningocele Study Trial Outcomes*

	Prenatal Surgery	Postnatal Surgery	Relative Risk (95%CI)	*P* Value
Perinatal death	2/78 (3%)	2/80 (2%)	1.03 (0.14–7.10)	1.00
Ventriculoperitoneal shunt placement	31/78 (40%)	66/80 (82%)	0.48 (0.36–0.64)	<0.001
Death or ventriculoperitoneal shunt (12 months of age)	53/78 (68%)	78/80 (98%)	0.70 (0.58–0.84)	<0.001
30-month outcome Bayley Mental Developmental Index and motor function	148.6+/-57.5	122.6+/-57.2	—	0.007
Hindbrain herniation (12 months)	45/70 (64%)	66/69 (96%)	0.67 (0.56–0.81)	<0.001
Walking independently (30 months)	26/62 (42%)	14/67 (21%)	2.01 (1.16–3.48)	0.01
Preterm birth (<37 weeks)	62/78 (80%)	12 (15%)	—	<0.001
Preterm birth (<30 weeks)	10/78 (13%)	0/80	—	—
Pulmonary edema	5/78 (6.4%)	0/80	—	0.03
Oligohydramnios	16/78 (20.5%)	3/80 (3.8%)	5.47 (1.66–18.04)	0.001
Placental abruption	5/78 (6.4%)	0/80	—	0.03
Hysterotomy site: thin or dehiscence	27/76 (35.5%)	0/80	—	—
Transfusion at delivery	7/78 (9.0%)	1 (1.3%)	7.18(0.90–57.01)	0.03

*183 women were randomized. The primary outcome reported is on 158 children at 12 months of age and 134 children at 30 months of age. The remaining patients are still being monitored for outcome and will be reported when available.

Data from Adzick NS, Thom EA, Spong CY, Brock JW 3rd, Burrows PK, Johnson MP, et al. A randomized trial of prenatal versus postnatal repair of myelomeningocele. N Engl J Med 2011;364:993–1004. [PubMed] [Full Text]

assist an approved fetal surgeon on five cases and then perform five cases with an approved fetal surgeon as an assistant. Operative procedures were standardized and involved an extensive multidisciplinary approach, including specialists to provide continuous fetal cardiac echocardiography throughout the surgery and dedicated anesthesiologists. The trial participants who underwent surgery during pregnancy remained near the hospital site for the duration of their pregnancies with protocols for follow-up for both groups of patients.

Inclusion criteria for this trial were stringent, requiring a singleton gestation with myelomeningocele between T-1 and S-1, evidence of hindbrain herniation, gestational age between 19.0 weeks and 25.9 weeks, normal karyotype, no anomalies unrelated to the myelomeningocele, no evidence of severe kyphosis, and a maternal body mass index less than 35. Based on the criteria set from this trial, the Committee on Obstetric Practice recommends that women who meet these criteria should be made aware of these findings and counseled regarding the option of maternal–fetal surgery, including both the risks and benefits to the woman and the baby (Table 1). This discussion also should include discussion of implications for future pregnancies.

Maternal–fetal surgery is a major procedure for the woman and her fetus, and it has significant implications and complications that occur acutely, postoperatively, for the duration of the pregnancy, and in subsequent pregnancies. It is a highly technical procedure with potential for significant morbidity and possibly mortality, even in the best and most experienced hands. Maternal–fetal surgery for myelomeningocele should only be offered at facilities with the expertise, multidisciplinary teams, services, and facilities to provide the intensive care required for these patients. Rigorous patient care selection that offers the best outcomes with the least risk is important.

Reference

1. Adzick NS, Thom EA, Spong CY, Brock JW 3rd, Burrows PK, Johnson MP, et al. A randomized trial of prenatal versus postnatal repair of myelomeningocele. N Engl J Med 2011;364:993–1004. [PubMed] [Full Text]

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