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Committee on Obstetric Practice
Society for Maternal–Fetal Medicine

The North American Fetal Therapy Network endorses this document. This Committee Opinion was developed by the American College of Obstetricians and Gynecologists' Committee on Obstetric Practice in collaboration with committee member Russell S. Miller, MD, and the Society for Maternal–Fetal Medicine in collaboration with member Jeffrey A. Kuller, MD.

Maternal–Fetal Surgery for Myelomeningocele

ABSTRACT: Myelomeningocele, a severe form of spina bifida, occurs in approximately 1 in 3,000 live births in the United States. The extent of disability is generally related to the level of the myelomeningocele defect, with a higher upper level of lesion generally corresponding to greater deficits. Open maternal–fetal surgery for myelomeningocele repair is a major procedure for the woman and her affected fetus. Although there is demonstrated potential for fetal and pediatric benefit, there are significant maternal implications and complications that may occur acutely, postoperatively, for the duration of the pregnancy, and in subsequent pregnancies. Women with pregnancies complicated by fetal myelomeningocele who meet established criteria for in utero repair should be counseled in a nondirective fashion regarding all management options, including the possibility of open maternal–fetal surgery. Maternal–fetal surgery for myelomeningocele repair should be offered only to carefully selected patients at facilities with an appropriate level of personnel and resources.

Recommendations

The American College of Obstetricians and Gynecologists and the Society for Maternal–Fetal Medicine make the following recommendations:

- Open maternal–fetal surgery for myelomeningocele repair has been demonstrated to improve a number of important pediatric outcomes at the expense of procedure-associated maternal and fetal risks.
- Women with pregnancies complicated by fetal myelomeningocele who meet established criteria for in utero repair should be counseled in nondirective fashion regarding all management options, including the possibility of open maternal–fetal surgery.
- Interested candidates for fetal myelomeningocele repair should be referred for further assessment and consultation to a fetal therapy center that offers this intervention and possesses the expertise, multidisciplinary team, services, and facilities to provide detailed information regarding maternal–fetal surgery and the intensive care required for patients who choose to undergo open maternal–fetal surgery.

Introduction

Myelomeningocele, a severe form of spina bifida, occurs in approximately 1 in 3,000 live births in the United States (1) and is complicated by hydrocephalus, need for ventriculoperitoneal shunt placement, motor and cognitive defects, bowel and bladder dysfunction, and social and emotional challenges. The extent of disability generally is related to the level of the myelomeningocele defect, with a higher upper level of lesion generally corresponding to greater deficits. Among newborns prenatally diagnosed with myelomeningocele, lesions are usually surgically repaired in the early neonatal period.

Fetal surgery has historically been considered a heroic intervention reserved for severe fetal presentations in which in utero therapy might favorably alter a natural history expected to result in fetal or neonatal death or severe disability. However, significant maternal and fetal risks prompted concern regarding the appropriateness of such treatments. Although open maternal–fetal surgery was originally limited to life-threatening conditions, it was considered for fetal myelomeningocele repair because results of laboratory and animal studies

demonstrated that antenatal surgery may improve neurologic outcomes. Favorable results with maternal–fetal repair were attributed to a combination of reduced direct trauma to the myelomeningocele while in utero, decreased leakage of cerebrospinal fluid, and decreased exposure of neurologic elements to potentially neurotoxic amniotic fluid. This document revises Committee Opinion No. 550, *Maternal–Fetal Surgery for Myelomeningocele*, to acknowledge the more widespread availability of prenatal myelomeningocele repair and to incorporate additional data.

Evidence Supporting Fetal Myelomeningocele Repair

After early experience in human pregnancies demonstrated feasibility, a randomized controlled trial was undertaken to evaluate the safety and efficacy of maternal–fetal surgery for myelomeningocele repair when compared with standard postnatal care (2). The support of the perinatal and maternal–fetal surgery community was key to the success of this trial, as a nationwide moratorium was honored during the recruitment period that limited fetal repair to randomization between prenatal surgery or expectant management with postnatal repair at three designated study centers.

The trial was conducted with considerable organization and oversight. All sites had experienced fetal surgeons, pediatric neurosurgeons, multidisciplinary teams, and state-of-the-art equipment. Operative procedures were standardized and involved an extensive multidisciplinary approach, including specialists to provide continuous fetal echocardiography throughout the surgery and dedicated anesthesiologists. Trial participants who underwent prenatal surgery remained near the fetal surgery center for the duration of their pregnancies.

Inclusion criteria for this trial were stringent, requiring a singleton gestation, myelomeningocele with an upper boundary located between T1 and S1, evidence of hindbrain herniation on fetal magnetic resonance imaging, gestational age between 19 0/7 weeks and 25 6/7 weeks at randomization, and a normal karyotype. Major trial exclusion criteria included anomalies unrelated to the myelomeningocele, severe kyphosis, risk of preterm birth (such as short cervix or prior preterm birth), placental abruption, contraindication to surgery (such as previous hysterotomy in the active uterine segment), and a maternal body mass index of 35 or more.

The investigators (2) found that open maternal–fetal surgery for myelomeningocele repair improved a number of important pediatric outcomes at the expense of procedure-associated maternal and fetal risks. When compared with standard postnatal repair, prenatal surgery reduced the rate of death or cerebrospinal shunt requirement at 12 months of age, decreased the rate of hindbrain herniation at 12 months of age (3), doubled the rate of independent ambulation at 30 months of age, improved 30-month neuromotor outcomes, and produced a level of function that was more often better than expected according to anatomical levels (Table 1). However, prenatal surgery also was associated with higher rates of obstetric and maternal complications, including preterm birth, chorion–amnion separation, spontaneous membrane rupture, oligohydramnios, placental abruption, pulmonary edema, maternal transfusion at delivery, and an increased incidence of uterine thinning or dehiscence of the uterine scar at delivery (4). It is postulated that uterine thinning or dehiscence is likely to happen because the hysterotomy location for fetal myelomeningocele repair is dictated by placental location and, therefore, may require a fundal or posterior approach. Long-term follow-up from the trial of children ages 5–8 years is ongoing, as is maternal follow-up including subsequent pregnancy outcomes.

Despite this randomized trial, a recent Cochrane review concluded that high-quality data comparing prenatal to postnatal myelomeningocele repair are limited to this single small trial and, thus, represent insufficient evidence to recommend drawing firm conclusions regarding prenatal myelomeningocele repair (5). Furthermore, because the trial was conducted in rigorous fashion, another concern is that study outcomes may represent a best-case scenario that cannot be reproduced outside of a trial setting. However, authors of another study recently published a single-center posttrial experience involving 100 cases of prenatal myelomeningocele repair, reporting short-term outcomes that were comparable with the original trial (6). This experience was notably from a primary study center in the original trial, which limits conclusions regarding contemporary outcomes at nontrial centers now offering fetal myelomeningocele repair. Nonetheless, there is still value in the observation that equivalent outcomes were achieved outside of the trial setting.

Practice Implications

Based upon the results of the randomized trial, the American College of Obstetricians and Gynecologists and the Society for Maternal–Fetal Medicine recommend that women with pregnancies complicated by fetal myelomeningocele who meet established criteria for in utero repair should be counseled in a nondirective fashion regarding all management options, including the possibility of open maternal–fetal surgery. This consultation should include a discussion of risks and benefits to the woman and her fetus, including limited data regarding potential implications for long-term maternal reproductive health and future pregnancies after open maternal–fetal surgery (7). In addition to the need for cesarean delivery with the index pregnancy after open maternal–fetal surgery, discussion should emphasize the need for cesarean delivery with all future pregnancies similar to that recommended for patients with a prior classical hysterotomy. Interested candidates should be referred for further assessment and consultation 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)	<.001
Death or ventriculoperitoneal shunt (12 months of age)	53/78 (68%)	78/80 (98%)	0.70 (0.58–0.84)	<.001
30-month outcome Bayley Mental Developmental Index and motor function	148.6+/-57.5	122.6+/-57.2	—	.007
Hindbrain herniation (12 months)	45/70 (64%)	66/69 (96%)	0.67 (0.56–0.81)	<.001
Walking independently (30 months)	26/62 (42%)	14/67 (21%)	2.01 (1.16–3.48)	.01
Preterm birth (<37 weeks)	62/78 (80%)	12/80 (15%)	—	<.001
Preterm birth (<30 weeks)	10/78 (13%)	0/80	—	—
Pulmonary edema	5/78 (6.4%)	0/80	—	.03
Oligohydramnios	16/78 (20.5%)	3/80 (3.8%)	5.47 (1.66–18.04)	.001
Placental abruption	5/78 (6.4%)	0/80	—	.03
Chorion–amnion separation	20/78 (26%)	0/80 (0%)	—	<.001
Spontaneous membrane rupture	36/78 (46%)	6/80 (8%)	6.15 (2.75–13.78)	<.001
Hysterotomy site: thin or dehiscence	27/76 (35.5%)	0/80	—	—
Transfusion at delivery	7/78 (9.0%)	1/80 (1.3%)	7.18 (0.90–57.01)	.03

*A total of 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.

Modified 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. MOMS Investigators. N Engl J Med 2011;364:993–1004.

a fetal therapy center that offers this intervention and possesses the expertise, multidisciplinary team, services, and facilities to provide detailed counseling and the intensive care required for women if they choose to undergo open maternal–fetal surgery (8). Ideally, maternal–fetal surgery for myelomeningocele repair should be offered to patients meeting the inclusion criteria outlined in the aforementioned randomized trial (2), and it is recommended that pregnancies not meeting these established criteria only be considered for therapy under an institutional review board-approved research study.

Fetoscopic Repair of Fetal Myelomeningocele

Active research is ongoing into minimally invasive fetoscopic approaches to myelomeningocele repair. Although the allure of such approaches is a reduction in maternal and obstetric risk, published data are limited (9, 10). A recent retrospective study described experience developing a two-port fetoscopic technique and suggested that maternal–fetal complications may be comparable to those reported with open repair, yet with the potential to achieve vaginal delivery in the index pregnancy and reduced long-term maternal risks (10). However, study limitations included a small sample size and lack of information regarding long-term neurodevelopmental outcomes. At this time, fetoscopic fetal myelomeningocele repair cannot be recommended outside of an institutional review board-approved investigational setting at a center with an appropriate level of expertise, resources, and research oversight.

Conclusions

Open maternal–fetal surgery for myelomeningocele repair is a major procedure for the woman and her affected fetus. Although there is demonstrated potential for fetal and pediatric benefit, there are significant maternal implications and complications that may 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 with the best and most experienced surgeons. Maternal–fetal surgery for myelomeningocele repair should only be offered to carefully selected patients at facilities with an appropriate level of personnel and resources.

References

1. Parker SE, Mai CT, Canfield MA, Rickard R, Wang Y, Meyer RE, et al. Updated national birth prevalence estimates for selected birth defects in the United States, 2004–2006. National Birth Defects Prevention Network. Birth Defects Res A Clin Mol Teratol. 2010;88:1008–16. ⇦
2. 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. MOMS Investigators. N Engl J Med 2011;364:993–1004. ⇦
3. Sutton LN, Adzick NS, Bilaniuk LT, Johnson MP, Crombleholme TM, Flake AW. Improvement in hindbrain herniation demonstrated by serial fetal magnetic resonance imaging following fetal surgery for myelomeningocele. JAMA 1999;282:1826–31. ⇦
4. Johnson MP, Bennett KA, Rand L, Burrows PK, Thom EA, Howell LJ, et al. The Management of Myelomeningocele Study: obstetrical outcomes and risk factors for obstetrical complications following prenatal surgery. Management of Myelomeningocele Study Investigators. Am J Obstet Gynecol 2016;215:778.e1–9. ⇦
5. Grivell RM, Andersen C, Dodd JM. Prenatal versus postnatal repair procedures for spina bifida for improving infant and maternal outcomes. Cochrane Database Systematic Review 2014, Issue 10. Art. No.: CD008825. Available at: http://onlinelibrary.wiley.com/doi/10.1002/14651858.CD008825.pub2/abstract. Retrieved May 26, 2017. ⇦
6. Moldenhauer JS, Soni S, Rintoul NE, Spinner SS, Khalek N, Martinez-Poyer J, et al. Fetal myelomeningocele repair: the post-MOMS experience at the Children's Hospital of Philadelphia. Fetal Diagn Ther 2015;37:235–40. ⇦
7. Wilson RD, Lemerand K, Johnson MP, Flake AW, Bebbington M, Hedrick HL, et al. Reproductive outcomes in subsequent pregnancies after a pregnancy complicated by open maternal–fetal surgery (1996–2007). Am J Obstet Gynecol 2010;203:209.e1–6. ⇦
8. Cohen AR, Couto J, Cummings JJ, Johnson A, Joseph G, Kaufman BA, et al. Position statement on fetal myelomeningocele repair. MMC Maternal–Fetal Management Task Force. Am J Obstet Gynecol 2014;210:107–11. ⇦
9. Pedreira DA, Zanon N, Nishikuni K, Moreira de Sá RA, Acacio GL, Chmait RH, et al. Endoscopic surgery for the antenatal treatment of myelomeningocele: the CECAM trial. Am J Obstet Gynecol 2016;214:111.e1–11. ⇦
10. Belfort MA, Whitehead WE, Shamshirsaz AA, Bateni ZH, Olutoye OO, Olutoye OA, et al. Fetoscopic open neural tube defect repair: development and refinement of a two-port, carbon dioxide insufflation technique. Obstet Gynecol 2017;129:734–43. ⇦

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