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REVIEW ARTICLE

Intrauterine Surgery for Spinal Defects: What is the Role of Ultrasound?

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ABSTRACT

Ultrasound imaging has led to the diagnosis of fetal anomalies that can affect many organ systems. Since the development of high-resolution real-time ultrasound, the possibility of surgical intervention before birth to correct or treat prenatally diagnosed abnormalities has been realized. Fetal surgery has become a new standard of care for the perinatal treatment of myelomeningocele for mothers and fetuses that meet the specific criteria. In this review, we will consider the role of ultrasound for open fetal surgery or minimally invasive fetal surgery approaches.

Keywords: Fetal therapies, Meningomyelocele, Ultrasonography interventional, Ultrasonography prenatal.

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INTRODUCTION

The past two decades have yielded profound advances in the fields of prenatal diagnosis and fetal intervention. Ultrasound imaging has led to the diagnosis of fetal anomalies that can affect many organ systems. Since the development of high-resolution real-time ultrasound, the possibility of surgical intervention before birth to correct or treat prenatally diagnosed abnormalities has been realized.[1,2]

The presumption has been that earlier *in utero* repair, as opposed to postnatal surgery, would provide superior outcomes for the offspring.[3] In 2011, the Management of Myelomeningocele Study (MOMS) publication (a high-quality randomized controlled trial (RCT) sponsored by the National Institutes of Health) reported a new paradigm for spina bifida treatment.[4,5]

Spina bifida is a defect of the vertebrae that results in exposure of the contents of the neural canal. In the vast majority of cases, the defect is localized to the posterior arch of the vertebrae. Open spina bifida is the most frequent lesion, resulting in 85% of dorsal defects. Synonyms of spina bifida are spinal dysraphism, rachischisis, meningocele, and myelomeningocele.[6] The neural canal may be exposed, or a thin meningeal membrane may cover the defect. More often, the lesion appears as a cystic tumor. If the lesion contains purely meninges, it is referred to as a meningocele. More frequently, neural tissue is part of the mass, and the name myelomeningocele is used.[6] The spinal level of the defect determines the degree of motor and somatosensory deficit.[7]

Spinal defects are more frequent in whites and are most prevalent among Hispanics. These differences seem to persist even after migration, suggesting a genetic rather than an environmental effect.[8] The incidence is approximately 1 to 5 per 1,000 live births.[9]

The basic purpose of fetal surgery is to prevent the progression of neurological damage resulting from exposure of the spinal cord to amniotic fluid, assuming that this injury is progressive during intrauterine life.[10-12] Currently, fetal surgery can be performed as open fetal surgery or minimally invasive fetal surgery.[5] Despite the heterogeneity of interventions and the difficulty in selecting the specific group of fetuses that would obtain real benefit from antenatal surgery, the significant change in prognostic factors of some diseases after fetal surgery highlights the relevance of antenatal intervention.[5]

Since the MOMS trial publication, open fetal surgery has become a new standard of care for the perinatal treatment of MMC for mothers and fetuses that meet the specific criteria.[4] In such studies, the maternal risk is considered high by many specialists. Minimally, invasive fetal surgery techniques are under investigation for spina bifida correction. For many fetal conditions, fetoscopic surgery is replacing open surgery. In this review, we will consider the role of ultrasound for both surgery approaches.

ULTRASOUND

The development of fetal surgery should not be disconnected from improvements in ultrasound equipment.

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Ultrasound examination is an effective technique for screening for and diagnosing neural tube defects (NTDs).[13]

Neural tube defect screening has two approaches: Measuring maternal serum alpha-fetoprotein and performing an ultrasound examination.[14] As ultrasound screening programs become more widely implemented and qualitatively improve, an increasing number of NTDs will be diagnosed in early gestation. When fetal malformations are suspected, the patients should be referred to fetal care centers. When there is an indication for fetal therapy, a subsequent referral to a highly specialized unit may be required, as many therapies cannot be offered locally with reasonably sufficient experience.[2]

First trimester transvaginal ultrasound has detection rates of 90% for anencephaly and 80% for encephalocele, but only 44% for spina bifida. The second trimester ultrasound examination increases the detection rate to approximately 92 to 95% for spina bifida and to 100% for anencephaly.[15]

Spine defects and cerebellar herniation have been identified in the first trimester.[16,17] Considering the development of new ultrasound techniques, it is expected that the diagnosis can be safely performed at 11 to 13 weeks' gestation.

Spina bifida is usually diagnosed by ultrasound between 18 and 22 weeks' gestation by the identification of indirect ultrasound signs, such as the "lemon sign," corresponding to the change in the skull shape, and the presence of ventriculomegaly associated with cerebellar herniation ("banana sign"), which is an Arnold-Chiari type II malformation.[15]

PREOPERATIVE ULTRASOUND EVALUATION

Before surgery, precise determination of the correct diagnosis is pivotal for directing possible interventions, and obstetric ultrasound remains the sole method of evaluation in many conditions. A targeted ultrasound evaluation includes confirmation of the suspected fetal diagnosis; detection of possible associated hydrops; determination of the size, extent, or severity of the lesion; measurement of the baseline cervical length; and documentation of the placental location.

The fetal diagnostic confirmation of NTDs by ultrasound examination depends in part on the size and location of the defect, the position of the fetus, the volume of the amniotic fluid, maternal habitus,[18] the skill of the sonographer, and the equipment.[14] Once a diagnosis is established, ultrasound can reveal additional findings and can provide important prognostic information. This information can be used to better characterize the condition, predict outcome, and help guide families in weighing the options regarding pregnancy management and physicians in determining the potential benefits of fetal intervention.[14]

Ultrasound is utilized to obtaining prognostic information based on an assessment of the level of the spinal defect. This information is used to inform and counsel the family about the spectrum of anticipated disabilities. Magnetic resonance imaging (MRI) of the fetal brain and spine may be performed as an adjunct to ultrasound, particularly when evaluating structural brain abnormalities.[14]

The most important predictor of pediatric neurological function, with either prenatal or standard postnatal repair, is the upper level of the lesion.[10] The extent of the spinal defect is determined by serial scanning of the spinal vertebrae in a transverse plane with the ultrasound transducer held perpendicular to the surface of the fetal back. The most caudal fetal rib is identified as T12, and the superior aspect of the iliac crest is denoted as L5. The upper level of the lesion is determined to be the most cephalad vertebra affected by a dorsal defect with widening of the ossification centers.[10]

Aaronson et al[19] reported that prenatal MRI and ultrasound are equally accurate for determining the lesion level in a fetus with myelomeningocele. However, Bruner et al[20] found that only 29% of examinations performed by community physicians were successful in assigning a specific lesion level. It is clear that a significant learning effect must be considered to improve the accuracy of ultrasound evaluations.

INTRAOPERATIVE ULTRASOUND GUIDANCE

Intraoperative ultrasound guidance assists in the technical aspects of the procedure and in fetal monitoring. The fetus and placenta are located by ultrasound, and a uterine incision location remote from the placental edge is chosen by the primary surgeon.[21] The fetus is visualized by ultrasound and manually positioned within the uterus such that the myelomeningocele sac is in the center of the hysterotomy. In the case of an anterior placenta, hysterotomy may be fundal or posterior. In the case of a posterior placenta, uterine entry is anterior. Under ultrasound guidance, the surgeon places two monofilament traction sutures through the full thickness of the uterine wall, initial uterine entry is accomplished directly between the uterine traction sutures, and the uterine stapling device loaded with absorbable polyglycolic acid staples is then passed into the uterine cavity. The stapler is palpated manually, and ultrasonography is used to exclude the presence of fetal tissue; then, the stapler is used to create a 6 to 8 cm uterine incision large enough to expose the fetal myelomeningocele. During the repair, ultrasound is used to monitor fetal cardiac function and

to identify potential complications, including placental abruption.[4,21]

While the uterine incision is open, a sterile catheter is used to instill lactated Ringer's solution into the amniotic cavity. Before closure, ultrasound is utilized to ascertain the appropriate fluid volume to instill, aiming for a "low-normal" level of amniotic fluid.[21]

In minimally invasive fetal surgery, preprocedural ultrasound determination of placental location and documentation of fetal position are valuable. Once an optimal entry site is identified and the trocars are safely inserted, the amniotic fluid is replaced with carbon dioxide. However, fetal cardiac function cannot be reliably monitored by ultrasound during fetoscopic surgery. After fetoscopic defects are covered, the fluid volume is instilled, and ultrasound is used to ensure an appropriate amniotic fluid level.[22]

POSTOPERATIVE ULTRASOUND EVALUATION

After surgery, ultrasound is performed to evaluate the fetal response to the intervention, identify any procedural complications, and continue to monitor fetal growth and well-being. Ultrasound examination is an important adjunct for the clinical assessment of preterm premature rupture of membranes (PPROM), and it can also be used to monitor for chorioamniotic separation. Ultrasound evaluation of the fetal condition includes measuring the deepest vertical pocket of amniotic fluid and the bladder as well as performing a cardiac assessment. Transient mild oligohydramnios without overt rupture of membranes is not uncommon in this group of postsurgical patients, perhaps because of amniotic fluid leakage through the hysterotomy site or transient fetal renal injury with oliguria.[21]

Postoperative ultrasound assessment of the fetus includes viewing the surgical site, lateral cerebral ventricles, posterior fossa, and lower extremities.[21]

Outpatient follow-up is scheduled every week. In addition to the usual content of a prenatal visit, the fetal assessment includes measuring the amniotic fluid volume and evaluating the membrane status because oligohydramnios and chorioamniotic membrane separation are the most frequent complications after maternal-fetal surgery, and their presence may directly impact the pregnancy management. Fetal well-being is determined at every visit based on a biophysical profile. Comprehensive ultrasound is performed monthly for fetal biometry to measure the amniotic fluid index and to analyze the chorion status and the ventricular size.[4] This assessment pays considerable attention to intracranial anatomy, including the ventricles (the atrial diameter of the lateral ventricle is measured) and posterior fossa, and to the lower extremities. This prognostic information is used to inform and counsel the family.

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