COVERAGE:

Fetal Surgery is considered medically necessary for the following conditions:

- Vesicoamniotic shunting as a treatment of urinary tract obstruction
- Temporary tracheal occlusion as a treatment of congenital diaphragmatic hernia
- Either open in utero resection of malformed pulmonary tissue or placement of a thoracoamniotic shunt as a treatment of either congenital cystic adenomatoid malformation or extralobar pulmonary sequestration.
- Surgical intervention consisting of in utero removal of sacrococcygeal teratoma for fetuses with a gestational age of 32 weeks or less and evidence of fetal hydrops, placentomegaly and/or the beginnings of maternal mirror syndrome.

As other techniques of fetal surgery are developed, they will be considered appropriate for benefits. Perhaps the most appropriate approach to the entire field of fetal surgery may be the identification of qualified fetal surgery centers rather than attempting to evaluate the benefits of each new surgical procedure itself.

DESCRIPTION:

Most fetal anatomic malformations are best managed after birth. However, advances in methods of prenatal diagnosis (particularly prenatal ultrasound) have led to a new understanding of the natural history and physiologic outcomes of certain congenital anomalies. Fetal surgery is the logical extension of these diagnostic advances, related in part to technical advancement in anesthesia, tocolysis, and hysterotomy. Fetal surgery has been most thoroughly researched for the following clinical conditions:

- Urinary tract obstruction
- Congenital diaphragmatic hernia
- Congenital cystic adenomatoid malformation and extralobar pulmonary sequestration
- Sacrococcygeal teratoma

Fetal surgery typically involves opening the uterus with either a traditional cesarean surgical incision or through single or multiple fetoscopic port incisions. The fetal abnormality is surgically corrected, the fetus (if removed from the uterus for the intervention) is returned to the uterus and then the uterus is closed for completion of gestational development.
Fetal surgery is a specialized technique that requires a multidisciplinary approach. Adzick and Harrison, two of the pioneers in fetal surgery, have published the following (Adzic NS, Harrison HR. Fetal surgical therapy. Lancet, 1994;343: 897-902.):

“The promise of fetal therapy is that the earliest possible intervention for a life-threatening fetal disorder may produce the best results. Because fetal surgery jeopardizes the pregnancy and may put the mother as well as the fetus at risk, it should be considered only in centres that are committed to a program of continuing research together with cautious clinical application. A fetal treatment centre requires the close collaboration of dedicated pediatric surgeons, perinatal obstetricians, sonographers, echocardiographers, neonatalogists, intensive care specialists, geneticists, ethicists, neonatal and obstetric nurses, and a compassionate nurse coordinator. The fetal treatment team should be committed to having this innovative therapy reviewed by uninvolved professional colleagues (institutional review board). To publish all results (bad as well as good), to avoid media reports until cases are peer reviewed, and to test the validity and cost-effectiveness of this approach in properly controlled trials.”

RATIONALE:

This policy is based in part on a 1999 and a 2000 TEC assessment that offered the following conclusions on different applications of fetal surgery:

1. Fetal Urinary Tract Obstruction

Few cases of prenatally diagnosed urinary tract obstruction require prenatal intervention. However, bilateral obstruction is often associated with serious disease (i.e. pulmonary hypoplasia secondary to oligohydramnios). Therefore, fetuses with bilateral obstruction, oligohydramnios, adequate renal function reserve, and no other lethal or chromosomal abnormalities may be candidates for fetal surgery. The most common surgical approach is vesicoamniotic shunting by means of shunt or stent placement. The shunting procedure bypasses the obstructed urinary tract, permitting fetal urine to flow into the amniotic space. Small case series have shown improved survival associated with fetal surgery.

2. Congenital Diaphragmatic Hernia (CDH)

CDH is a defect that permits abdominal viscera to enter the chest, frequently resulting in hypoplasia of the lungs. CDH can vary widely in severity, depending on the size of the hernia and the timing of herniation. For example, late herniation after 25 weeks of gestation may be adequately managed postnatally. In contrast, liver herniation into the chest prior to 25 weeks of gestation is associated with a poor prognosis; these fetuses have been considered candidates for fetal surgery. Temporary tracheal occlusion prevents the normal efflux of fetal lung fluid, which in turn enhances positive pressure in the growing lungs, promoting lung growth and ultimately reducing abdominal viscera back into the abdominal cavity. One small case series reports a significant improvement in survival with fetal surgery.

3. Congenital Cystic Adenomatoid Malformation (CCAM) or Extralobar Pulmonary Sequestration (EPS)
CCAM and EPS are the two most common congenital cystic lung lesions. Survival is poor when associated with fetal hydrops before 32 weeks’ gestation. These patients may be candidates for prenatal surgical resection of a large mass or placement of a thoraco-amniotic shunt for a large uniilocular cystic lesion. Small case series reports that prenatal intervention has been attempted in 22 fetuses with a survival rate of 73%.

4. Sacrococcygeal Teratoma (SCT)

SCT is both a neoplasm with the power of autonomous growth and a malformation made up of multiple tissues foreign to the sacrococcygeal region lacking organ specificity. With an incidence of 1 in 35,000-40,000 live births, SCT is the most common tumor of the newborn. It occurs with greater frequency in females (3:1). Believed to arise from pluripotent cells during embryogenesis, SCT is closely attached to the coccyx in all cases and may present as a presacral (intrapelvic) tumor and/or as an external tumor.

Case reports as well as retrospective studies from several medical centers demonstrate that the usual clinical outcome in the subset of patients with sacrococcygeal teratoma, fetal hydrops, and pulmonary immaturity is very poor, with mortality of 100% when SCT is diagnosed before 25 weeks gestation. In utero surgical treatment of fetuses with SCT, pulmonary immaturity, ultrasonographic evidence of hydrops, and/or placentomegaly appears to offer a dramatic reduction in the risk of death.

Other Fetal Conditions

It is anticipated that additional experience in the area of fetal surgery may establish the clinical indications for surgical approaches such as Myelomeningocele, Aqueductal Stenosis and other fetal conditions. The most appropriate approach to the entire field of fetal surgery may be the identification of qualified fetal surgery centers rather than attempting to evaluate the benefits of each new surgical procedure itself.

DISCLAIMER:

State and federal law, as well as contract language, including definitions and specific inclusions/exclusions, takes precedence over Medical Policy and must be considered first in determining coverage. The member’s contract benefits in effect on the date that services are rendered must be used. Any benefits are subject to the payment of premiums for the date on which services are rendered. Medical technology is constantly evolving, and we reserve the right to review and update Medical Policy periodically. HMO Blue Texas physicians who are contracted/affiliated with a capitated IPA/medical group must contact the IPA/medical group for information regarding HMO claims/reimbursement information and other general polices and procedures.

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