The following Protocol contains medical necessity criteria that apply for this service. It is applicable to Medicare Advantage products unless separate Medicare Advantage criteria are indicated. If the criteria are not met, reimbursement will be denied and the patient cannot be billed. Preauthorization is not required. Please note that payment for covered services is subject to eligibility and the limitations noted in the patient’s contract at the time the services are rendered.

Description

Fetal surgery is used for specific congenital abnormalities that are associated with a poor postnatal prognosis. Prenatal surgery typically involves opening the gravid uterus (with a Cesarean surgical incision), surgically correcting the abnormality, and returning the fetus to the uterus and restoring uterine closure. Minimally invasive procedures through single or multiple fetoscopic port incisions are also being developed.

Background

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.

This Protocol will pertain to fetal surgery performed for the following clinical conditions:

1. **Fetal Urinary Tract Obstruction**

   Although few cases of prenatally diagnosed urinary tract obstruction require prenatal intervention, bilateral obstruction can lead to distention of the urinary bladder and is often associated with serious disease such as 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 decompression through percutaneous placement of a shunt or stent. Vesicoamniotic shunting bypasses the obstructed urinary tract, permitting fetal urine to flow into the amniotic space. The goals of shunting are to protect the kidneys from increased pressure in the collecting system and to assure adequate amniotic fluid volume for lung development.

2. **Congenital Diaphragmatic Hernia**

   Congenital diaphragmatic hernia (CDH) results from abnormal development of the diaphragm, which 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, and these fetuses have been considered candidates for fetal surgery. Temporary tracheal occlusion using a balloon is being evaluated for the treatment of CDH. Occluding the trachea of a fetus prevents the normal efflux of fetal lung fluid, which results in a build-up of secretions in the pulmonary tree and increases the size of the lungs,
gradually pushing abdominal viscera out of the chest cavity and back into the abdominal cavity. It is believed that this, in turn, will promote better lung maturation. Advances in imaging have resulted in the ability to detect less severe lesions, which has resulted in a decrease in mortality rates for defects detected during pregnancy. Due to these changes over time, concurrent controls are needed to adequately compare pre- and postnatal approaches.

3. **Congenital Cystic Adenomatoid Malformation or Bronchopulmonary Sequestration**

Congenital cystic adenomatoid malformation (CCAM) and bronchopulmonary sequestration (BPS) are the two most common congenital cystic lung lesions and share the characteristic of a segment of lung being replaced by abnormally developing tissue. CCAMs can have connections to the pulmonary tree and contain air, while BPS does not connect to the airway and has blood flow from the aorta rather than the pulmonary circulation. In more severe cases, the malformations can compress adjacent normal lung tissue and distort thoracic structure. CCAM lesions typically increase in size in mid-trimester and then in the third trimester either involute or compress the fetal thorax, resulting in hydrops in the infant and sometimes mirror syndrome (a severe form of preeclampsia) in the mother. Mortality is close to 100% when lesions are associated with fetal hydrops (abnormal accumulation of fluid in two or more fetal compartments). These patients may be candidates for prenatal surgical resection of a large mass or placement of a thoracoamniotic shunt to decompress the lesion.

4. **Sacrococcygeal Teratoma**

Sacrococcygeal teratoma (SCT) is both a neoplasm with the power of autonomous growth and a malformation made up of multiple tissues foreign to the region of origin and lacking organ specificity. It is the most common tumor of the newborn and generally carries a good prognosis in infants born at term. However, in utero fetal mortality approaches 100% with large or vascular tumors, which may become larger than the rest of the fetus. In this small subset, SCT is associated with fetal hydrops, which is related to high output heart failure secondary to arteriovenous shunting. In some cases, mothers of fetuses with hydrops can develop mirror syndrome.

5. **Myelomeningocele**

Myelomeningocele is a neural tube defect in which the spinal cord forms abnormally and is left open, exposing the meninges and neural tube to the intrauterine environment. Myelomeningocele is the most common cause of spina bifida, and depending on the location, results in varying degrees of neurologic impairment to the legs and bowel and bladder function, brain malformation (i.e., hindbrain herniation), cognitive impairment, and disorders of cerebrospinal fluid circulation, i.e., hydrocephalus requiring placement of a ventriculoperitoneal shunt. Traditional treatment consists of surgical repair after term delivery, primarily to prevent infection and further neurologic dysfunction. Fetal surgical repair to cover the exposed spinal canal has been proposed as a means of preventing the deleterious exposure to the intrauterine environment with the hope of improving neurologic function and decreasing the incidence of other problems related to the condition.

6. **Cardiac Malformations**

In utero interventions are being investigated for severe narrowing in one of the cardiac outflow tracts (aortic valve or pulmonary valve), a condition which causes progressive damage to the heart in utero. In utero intervention has been proposed for the following lethal cardiac conditions: critical pulmonary stenosis, critical aortic stenosis, and hypoplastic left heart syndrome (HLHS). Critical pulmonary stenosis or atresia with intact ventricular septum is characterized by a very narrow pulmonary valve without a connection between the right and left ventricles. Critical aortic stenosis with impending HLHS is a very narrow aortic valve that develops early during gestation that may result in HLHS. In utero aortic balloon valvuloplasty has been suggested as a way to relieve aortic stenosis in an attempt to preserve left ventricular growth and halt
the progression to HLHS. HLHS with intact atrial septum describes the absence of a connection between the left and right atrium. A subset of fetuses with HLHS will present with severe cyanosis and require immediate postnatal intervention to survive. In utero septostomy has been performed in an attempt to improve postnatal survival for this condition.

Policy (Formerly Corporate Medical Guideline)

Vesicoamniotic shunting as a treatment of urinary tract obstruction may be considered medically necessary in fetuses under the following conditions:
- Evidence of hydronephrosis due to bilateral urinary tract obstruction; AND
- Progressive oligohydramnios; AND
- Adequate renal function; AND
- No other lethal abnormalities or chromosomal defects.

Open in utero resection of malformed pulmonary tissue or placement of a thoracoamniotic shunt may be considered medically necessary under following conditions:
- Congenital cystic adenomatoid malformation or bronchopulmonary sequestration is identified; AND
- The fetus is at 32 weeks’ gestation or less; AND
- There is evidence of fetal hydrops, placentomegaly, and/or the beginnings of severe pre-eclampsia (i.e., the maternal mirror syndrome) in the mother.

In utero removal of sacrococcygeal teratoma may be considered medically necessary under following conditions:
- The fetus is at 32 weeks’ gestation or less; AND
- There is evidence of fetal hydrops, placentomegaly, and/or the beginnings of severe pre-eclampsia (i.e., the maternal mirror syndrome) in the mother.

In utero repair of myelomeningocele may be considered medically necessary under the following conditions:
- The fetus is at less than 26 weeks’ gestation; AND
- Myelomeningocele is present with an upper boundary located between T1 and S1 with evidence of hindbrain herniation.

In utero repair of myelomeningocele is considered investigational in the following situations:
- Fetal anomaly unrelated to myelomeningocele; OR
- Severe kyphosis; OR
- Risk of preterm birth (e.g., short cervix or previous preterm birth); OR
- Maternal body mass index of 35 or more.

Other applications of fetal surgery are investigational, including but not limited to, temporary tracheal occlusion as a treatment of congenital diaphragmatic hernia or treatment of congenital heart defects.

Policy Guideline

After 32 weeks’ gestation, fetal lung maturity is adequate to permit Cesarean section and management of congenital cystic adenomatoid malformation, bronchopulmonary sequestration, or sacrococcygeal teratoma postnatally.
Services that are the subject of a clinical trial do not meet our Technology Assessment Protocol criteria and are considered investigational. For explanation of experimental and investigational, please refer to the Technology Assessment Protocol.

It is expected that only appropriate and medically necessary services will be rendered. We reserve the right to conduct prepayment and postpayment reviews to assess the medical appropriateness of the above-referenced procedures. Some of this Protocol may not pertain to the patients you provide care to, as it may relate to products that are not available in your geographic area.

References

We are not responsible for the continuing viability of web site addresses that may be listed in any references below.


