Medical Coverage Policy | Fetal Surgery for Prenatally Diagnosed Malformations

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OVERVIEW

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.

PRIOR AUTHORIZATION

Preauthorization is not required

POLICY STATEMENT

The following instances of fetal surgery performed for the following clinical conditions considered not medically necessary.

In utero repair of myelomeningocele in one of the following situations;

- Fetal anomaly unrelated to myelomeningocele;
- Severe kyphosis;
- Risk of preterm birth (eg, short cervix or previous preterm birth);
- Maternal body mass index of 35 or more;
- Treatment of congenital heart defect; Temporary tracheal occlusion as a treatment of congenital diaphragmatic hernia

The following fetal surgeries are considered medically necessary for Blue CHiP for Medicare and all BCBSRI commercial products.

Vesicoamniotic shunting as a treatment of urinary tract obstruction in fetuses under all of the following conditions:

- Evidence of hydronephrosis due to bilateral urinary tract obstruction;
- Progressive oligohydramnios;
- Adequate renal function;
- No other lethal abnormalities or chromosomal defects.

Open in utero resection of malformed pulmonary tissue or placement of a thoracoamniotic shunt with the following conditions:

- Congenital cystic adenomatoid malformation or bronchopulmonary sequestration is identified;
- The fetus is at 32 weeks' gestation or less;
- There is evidence of fetal hydrops, placentomegaly, and/or the beginnings of severe preeclampsia (ie, the maternal mirror syndrome) in the mother.

In utero removal of sacrococcygeal teratoma may be considered medically necessary under all of the following conditions:

- The fetus is at 32 weeks' gestation or less;
- There is evidence of fetal hydrops, placentomegaly, and/or the beginnings of severe preeclampsia (ie, maternal mirror syndrome) in the mother.

In utero repair of myelomeningocele may be considered medically necessary with all of the following conditions:

- The fetus is at less than 26 weeks' gestation;
- Myelomeningocele is present with an upper boundary located between T1 and S1 with evidence of hindbrain herniation.

MEDICAL CRITERIA

None

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 policy 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 2 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 2 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 (ie, hindbrain herniation), cognitive impairment, and disorders of cerebrospinal fluid circulation, ie, 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.

Fetal surgery is being investigated 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.

Due to a number of factors, including the rarity of the conditions and the small number of centers specializing in fetal interventions, the evidence on fetal surgery remains limited. Fetal surgery for many congenital conditions, including diaphragmatic hernia and heart defects, has not been shown to improve health outcomes in comparison with postnatal treatment. The available evidence is insufficient to demonstrate that fetal tracheal occlusion and aortic valvuloplasty provides improved health outcomes. For these and other applications of fetal surgery that are currently considered investigational, additional studies are needed to identify appropriate candidates and to evaluate longer term outcomes compared with postnatal management.

For conditions leading to fetal hydrops (certain cases of congenital cystic adenomatoid malformation, bronchopulmonary sequestration, or sacrococcygeal teratoma), for which mortality approaches 100%, fetal

surgery may be considered medically necessary. For bilateral urinary tract obstruction, evidence from retrospective and prospective cohort studies summarized in the 2011 Agency for Healthcare Research and Quality technology assessment on fetal surgery suggests that vesicoamniotic shunting improves survival, at least in the short term, although longer term follow up of patients treated with shunting and direct evaluation through randomized controlled trials is needed. As such, vesicoamniotic shunting for bilateral urinary tract obstruction may also be considered medically necessary to minimize the effects of this condition on kidney and lung development. A recent small, randomized, controlled trial evaluating the use of vesicoamniotic shunting found limited benefit from the procedure when data were analyzed by intention-to-treat analysis. However, the study's significant limitations, including low enrollment leading to early cessation of the study and significant crossover between treatment and control groups, make it difficult to generalize its finding of no significant benefit from treatment. Additional studies for these surgeries are needed to better define the appropriate surgical candidates, the most effective timing of the interventions, and the long-term health outcomes in surviving children.

Data from the MOMS trial show that prenatal repair of myelomeningocele reduces the need for shunting in the first 12 months after delivery and improves a composite measure of mental and motor function, with adjustment for lesion level, at 30 months of age. Prenatal surgery also improves the degree of hindbrain herniation and the likelihood of being able to walk independently when compared with postnatal surgery. The long-term impact on function needs to be evaluated, and benefits must be balanced against risks to mother and child. Thus, fetal surgery may be considered medically necessary following informed decision making for cases of prenatal myelomeningocele that meet the criteria of the MOMS study.

COVERAGE

Benefits vary between groups/contracts. Please refer to the appropriate Evidence of Coverage or Subscriber Agreement for applicable surgery services and services not medically necessary coverage.

CODING

BlueCHiP for Medicare and Commercial

The following CPT Code is medically necessary:

59076

The following CPT code follows the unlisted process:

59897

HCPC

The following HCPC codes are considered not medically necessary:

S2400 Repair, congenitally diaphragmatic hernia in the fetus using temporary tracheal occlusion, procedure performed in utero

S2404 Repair, myelomeningocele in the fetus, procedure performed in utero

The following HCPC codes are covered:

S2401 Repair, urinary tract obstruction in the fetus, procedure performed in utero

S2402 Repair, congenital cystic adenomatoid malformation in the fetus, procedure performed in utero

S2403 Repair, extralobar pulmonary sequestration in the fetus, procedure performed in utero

S2405 Repair of sacrococcygeal teratoma in the fetus, procedure performed in utero

S2409 Repair, congenital malformation of fetus, procedure performed in utero, not otherwise classified

RELATED POLICIES

None

PUBLISHED

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