Medical Coverage Policies

Fetal Surgery for Prenatally Diagnosed Malformations

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**Description:**

Fetal surgery is being investigated for specific congenital abnormalities that are associated with a poor postnatal prognosis. Prenatal surgery typically involves a Cesarean incision to surgically correct 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.

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 addresses fetal surgery performed for the following clinical conditions considered not medically necessary:

**Congenital Diaphragmatic Hernia (CDH)**

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.

**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.

**Cardiac Malformations**

In utero interventions are being investigated for severe narrowing in one of the cardiac outflow tracts (aortic valve or pulmonary valve) that cause 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.

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 has not been shown to improve health outcomes in comparison with postnatal 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.

**Medical Criteria:**

Not applicable.

**Policy:**

Vesico-amniotic shunting as a treatment of urinary tract obstruction is covered for all BCBSRI products. (S2401)

Open in utero resection of malformed pulmonary tissue or placement of a thoraco-amniotic shunt for Congenital Cystic Adenomatoid Malformation (CCAM) (S2402) or bronchopulmonary sequestration (BPS) (S2403) is covered for all BCBSRI products.

In utero removal of sacrococcygeal teratoma is covered for all BCBSRI products. (S2405)

Other applications of fetal surgery are not medically necessary for all BCBSRI products, including but not limited to, temporary tracheal occlusion as a treatment of congenital diaphragmatic hernia (S2400), treatment of congenital heart defects, or fetal surgery for myelomeningocele (S2404) which have all not been shown to improve health outcomes in comparison with postnatal treatment.

Amnioreduction and fetoscopic laser therapy as a treatment of twin-twin transfusion are not addressed in this policy.

**Coverage:**

Benefits may vary between groups/contracts. Please refer to the appropriate Evidence of Coverage, Subscriber Agreement, or Rite Care contract for the applicable surgery benefits/coverage.

**Coding:**

CPT

59076
59897

HCPCS

**Effective 02/01/2011** the following codes will be 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

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

Also known as:

Congenital Cystic Adenomatoid Malformation, Fetal Surgery
Congenital Diaphragmatic Hernia, Fetal Surgery
Extralobar Pulmonary Sequestration, Fetal Surgery
Fetal Surgery
In Utero Fetal Surgery
Sacrococcygeal Teratoma, Fetal Surgery
Temporary Tracheal Occlusion
Thoraco-amniotic Shunt
Urinary Tract Obstruction, Fetal Surgery
Vesico-amniotic Shunting
Myelomeningocele, Fetal Surgery

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Back to Previous Page