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.

MEDICAL CRITERIA
Not applicable

PRIOR AUTHORIZATION
Not applicable

POLICY STATEMENT
Medicare Advantage Plans and Commercial Products
Fetal surgery for prenatally diagnosed malformations is considered medically necessary for the following procedures:

- Vesicoamniotic shunting for the treatment of urinary tract obstructions,
- Open in-utero resection of malformed pulmonary tissue or placement of a thoracoamniotic shunt,
- In-utero removal of sacrococcygeal teratoma
- In-utero repair of myelomeningocele when the fetus is less than 26 weeks gestation and is present with an upper boundary located between T1 and S1 with evidence of hindbrain herniation

All other fetal surgeries are not covered for Medicare Advantage Plans and not medically necessary for Commercial Products as the evidence is insufficient to determine that the technology results in an improvement in the net health outcome.

COVERAGE
Benefits vary between groups and contracts. Please refer to the appropriate Benefit Booklet, Evidence of Coverage, or Subscriber Agreement for applicable surgery not medically necessary/not covered benefits/coverage.

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 the middle of the second 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 (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 several potentially lethal congenital heart disorders, including critical aortic stenosis with evolving hypoplastic left heart syndrome (HLHS), HLHS with intact atrial septum, and critical pulmonary stenosis or pulmonary atresia.1 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. Pulmonary atresia with intact ventricular septum can evolve into right ventricular hypoplasia; fetal pulmonary valvuloplasty may result in biventricular circulation. Critical aortic stenosis with impending HLHS is a very narrow aortic valve that develops early during gestation that may result in HLHS, a complex spectrum of cardiac anomalies characterized by hypoplasia of the left ventricle and aorta, with atretic, stenotic, or hypoplastic atrial and mitral valves. In-utero aortic balloon valvuloplasty relieves aortic stenosis with the goal of preserving left ventricular growth and halting the progression to HLHS. HLHS with intact trial septum is a variant of HLHS that occurs in about 22% of all HLHS cases in which blood flow across the foramen ovale is restricted, leading to left atrial hypertension and damage to the pulmonary vasculature, parenchyma, and lymphatics. For HLHS with intact atrial septum, fetal balloon atrial septostomy is designed to reduce the left atrial restriction.

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 is limited. Fetal surgery for many congenital conditions, including congenital diaphragmatic hernia (CDH) 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 for CDH and fetal intervention for evolving hypoplastic left heart syndrome (HLHS) and critical pulmonary stenosis or pulmonary atresia provides improved health outcomes.

For these and other applications of fetal surgery that are currently considered not medically necessary, 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. 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. 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. 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 management of myelomeningocele study (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.

The evidence related to the use of fetal surgery is limited by the rarity of the conditions treated and the extremely specialized nature of the procedures, although RCTs have been conducted for several conditions.

CODING

Medicare Advantage Plans and Commercial Products

The following code(s) are medically necessary for Medicare Advantage Plans and Commercial Products:

59076 Fetal shunt placement, including ultrasound guidance
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
S2404 Repair, myelomeningocele 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

The following HCPCS code(s) are not covered for Medicare Advantage Plans and not medically necessary for Commercial Products:

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

The following code follows the Unlisted Procedures process:

59897 Unlisted fetal invasive procedure including ultrasound guidance

RELATED POLICIES

Unlisted Procedures

PUBLISHED

Provider Update, August 2026
Provider Update, July 2025
Provider Update, December 2024
Provider Update, April 2023
Provider Update, June 2022

REFERENCES

1. Schidlow DN, Tworetzky W, Wilkins-Haug LE. Percutaneous fetal cardiac interventions for structural heart disease. Am J Perinatol. Aug 2014;31(7):629-636. PMID 24922056
2. 2.Morris RK, Malin GL, Quinlan-Jones E, et al. The Percutaneous shunting in Lower Urinary Tract Obstruction (PLUTO) study and randomised controlled trial: evaluation of the effectiveness, cost-effectiveness and acceptability of percutaneous vesicoamniotic shunting for lower urinary tract obstruction. Health Technol Assess. Dec 2013;17(59):1-232. PMID 24331029
3. 3.Rocha LA, Byrne FA, Keller RL, et al. Left heart structures in human neonates with congenital diaphragmatic hernia and the effect of fetal endoscopic tracheal occlusion. Fetal Diagn Ther. 2014;35(1):36-43. PMID 24356206
4. Shan W, Wu Y, Huang G, et al. Foetal endoscopic tracheal occlusion for severe congenital diaphragmatic hernia--a systemic review and meta-analysis of randomized controlled trials. J Pak Med Assoc. Jun 2014;64(6):686-689. PMID 25252491
5. White SB, Tutton SM, Rilling WS, et al. Percutaneous in utero thoracoamniotic shunt creation for fetal thoracic abnormalities leading to nonimmune hydrops. J Vasc Interv Radiol. Jun 2014;25(6):889-894. PMID 24702750
6. Van Mieghem T, Al-Ibrahim A, Deprest J, et al. Minimally invasive therapy for fetal sacrococcygeal teratoma: case series and systematic review of the literature. Ultrasound Obstet Gynecol. Jun 2014;43(6):611-619. PMID 24488859
7. Moldenhauer JS, Soni S, Rintoul NE, et al. Fetal Myelomeningocele Repair: The Post-MOMS Experience at the Children's Hospital of Philadelphia. Fetal Diagn Ther. Aug 15 2014. PMID 25138132
8. Bennett KA, Carroll MA, Shannon CN, et al. Reducing perinatal complications and preterm delivery for patients undergoing in utero closure of fetal myelomeningocele: further modifications to the multidisciplinary surgical technique. J Neurosurg Pediatr. Jul 2014;14(1):108-114. PMID 24784979
9. Pedra SR, Peralta CF, Crema L, et al. Fetal interventions for congenital heart disease in Brazil. Pediatr Cardiol. Mar 2014;35(3):399-405. PMID 24030590

10. Chaturvedi RR, Ryan G, Seed M, et al. Fetal stenting of the atrial septum: technique and initial results in cardiac lesions with left atrial hypertension. Int J Cardiol. Oct 3 2013;168(3):2029-2036. PMID 23481911
11. Kalish BT, Tworetzky W, Benson CB, et al. Technical challenges of atrial septal stent placement in fetuses with hypoplastic left heart syndrome and intact atrial septum. Catheter Cardiovasc Interv. Jul 1 014;84(1):77-85. PMID 23804575
12. American College of O, Gynecologists. ACOG Committee opinion no. 550: maternal-fetal surgery for myelomeningocele. Obstet Gynecol. Jan 2013;121(1):218-219. PMID 23262964
13. Biard JM, Johnson MP, Carr MC et al. Long-term outcomes in children treated by prenatal vesicoamniotic shunting for lower urinary tract obstruction. Obstet Gynecol 2005; 106(3):503-8. Kilby M, Khan K, Morris K et al. PLUTO trial protocol: percutaneous shunting for lower urinary tract obstruction randomised controlled trial. BJOG 2007; 114(7):904-5, e1-4. ve Stimulation (PTNS) (L31523) Rhode Island

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