# Medical Coverage Policy

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# Fetal Surgery

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## Related Coverage Resources

Ultrasound in Pregnancy (including 3D, 4D and 5D Ultrasound)

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## Overview

This Coverage Policy addresses fetal surgery performed in-utero to correct malformations of the fetus that interfere with organ development and that have potentially fatal outcomes if left untreated.

## Coverage Policy

**Fetal surgery is considered medically necessary for ANY of the following associated indication(s):**

- serial amnioreduction for twin-to-twin transfusion syndrome (TTTS)
- fetoscopic occlusion of anastomotic vessels (e.g., laser photocoagulation, radiofrequency ablation, ligation) for twin reversed arterial perfusion (TRAP sequence)
- fetal vesicoamniotic shunt procedures for bilateral fetal urinary-tract obstruction
- in-utero needle access and open resection of sacrococcygeal teratoma
- fetal thoracoamniotic shunt placement for ANY of the following indications:
  - congenital pulmonary airway malformation (CPAM)/congenital cystic adenomatoid formation (CCAM)
  - extralobar pulmonary sequestration (EPS)
  - fetal pleural effusion
- myelomeningocele repair when ALL of the following criteria are met:
  - singleton pregnancy
  - myelomeningocele with the upper boundary of the lesion located between T1 and S1
  - evidence of hindbrain herniation
  - gestational age ≥19.0 and < 26 weeks
  - normal fetal karyotype
- nonselective or selective fetoscopic laser coagulation for severe twin-to-twin transfusion syndrome (TTTS) when ALL of the following criteria are met:
  - fetal gestational age of less than 26 weeks
  - evidence of polyhydramnios in the recipient fetus
  - donor fetus is oligohydramniotic
  - evidence of abnormal blood flow documented by Doppler studies in one or both fetuses
- fetal lobectomy for congenital pulmonary airway malformation (CPAM)/congenital cystic adenomatoid formation (CCAM) when BOTH of the following criteria are met:
  - evidence of fetal hydrops
  - presence of large and multicystic or predominantly solid lesions
- fetoscopic endoluminal tracheal occlusion (FETO) for left congenital diaphragmatic hernia when ALL of the following criteria are met:
  - singleton pregnancy
  - fetal gestational age of less than 30 weeks
  - severe pulmonary hypoplasia defined as a quotient of the observed-to-expected lung-to-head ratios of less than 25%
  - no other major structural or chromosomal defects

**Fetal surgery is considered not medically necessary for ANY other indication, including the following:**

- laser, thermocoagulation or radiofrequency ablation techniques for the treatment of sacrococcygeal teratoma
- endoscopic approach (i.e., fetoscopic cystoscopy) for the treatment of lower urinary tract obstruction
- amniotic band syndrome
- aqueductal stenosis (i.e., hydrocephalus)
- cleft lip and/or cleft palate
- congenital heart defects
- in-utero gene therapy
- in-utero hematopoietic stem-cell transplantation for stem-cell-related diseases

## Health Equity Considerations

Health equity is the highest level of health for all people; health inequity is the avoidable difference in health status or distribution of health resources due to the social conditions in which people are born, grow, live, work, and age.

Social determinants of health are the conditions in the environment that affect a wide range of health, functioning, and quality of life outcomes and risks. Examples include safe housing, transportation and neighborhoods; racism, discrimination and violence; education, job opportunities and income; access to nutritious foods and physical activity opportunities; access to clean air and water; and language and literacy
skills.

## General Background

In-utero fetal surgery involves opening the gravid uterus through the less-invasive laparoscopic technique or through an open caesarian surgical incision; surgically correcting the fetal abnormality; and closing the uterus to allow gestational development to complete. Fetal surgery should be performed by highly trained physicians in advanced centers equipped to provide extracorporeal membrane oxygenation (ECMO) in Level III newborn intensive care units. The multidisciplinary approach employs pediatric surgeons, intensive care specialists, geneticists, ethicists, perinatologists, gynecological specialists, maternal/fetal specialists, pathologists and utilizes highly specialized radiology.

Fetal endoscopic surgery, a recently developed method of treating congenital conditions, can lessen maternal morbidity and additional stress to the fetus when the latter is removed from the amniotic fluid environment. Combined with the use of tocolytic drugs, this procedure may also decrease the occurrence of postoperative preterm labor.

Fetal intervention is recommended when preterm delivery is contraindicated, and the condition can be corrected allowing for normal development. Experts generally recommend early surgical intervention after a confirmed diagnosis of fetal decompensation. In general, surgery is performed prior to 32 weeks of gestation. After that time, standard treatment consists of early delivery and medically necessary interventions.

There are several contraindications to in-utero surgery, including severe congenital anomalies, chromosomal anomalies that jeopardize fetal survival, and maternal mirror syndrome. Patients

with maternal mirror syndrome are not considered candidates for prenatal intervention, as this condition may warrant immediate delivery. Maternal mirror syndrome is a maternal illness where the mother's condition mimics that of the sick fetus, as a result of severe fetal hydrops. Fetal hydrops is a condition where there is accumulation of fluid in two or more fetal compartments (e.g., abdomen, pleural space, pericardial space). With maternal mirror syndrome related to a hyperdynamic cardiovascular state, the mother develops symptoms that are similar to pre-eclampsia and may include vomiting, hypertension, peripheral edema, proteinuria and pulmonary edema. For cases of severe fetal hydrops where the cause is unknown and unable to be corrected, immediate delivery is indicated (Vidaeff, et al., 2002).

Fetal surgery has been researched for many different fetal abnormalities. However, when compared to traditional post-natal therapy, it has been shown to improve outcomes for only a few conditions that include: myelomeningocele repair, twin-to-twin transfusion syndrome, twin reversed arterial perfusion syndrome, urinary-tract obstruction, congenital cystic adenomatoid malformation, extralobar pulmonary sequestration, and sacrococcygeal teratoma. Few published studies have evaluated the safety and efficacy of fetal surgery for other conditions such as congenital heart defects, stem cell research and treatment of cleft lip and palate.

**Myelomeningocele**
Myelomeningocele, commonly referred to as spina bifida, is a neural-tube defect in which the spinal cord forms but remains open, exposing the meninges and neural tube to the intrauterine environment. The defect may include abnormal positioning of the brain (Arnold-Chiari II malformation). A variety of medical problems may result from the open neural tube and include, but are not limited to, physical and mental disabilities, deformity of the extremities, scoliosis, and urinary dysfunction or failure. Some researchers contend that intrauterine exposure may cause secondary trauma to the spinal cord.

Traditional treatment consists of surgical repair after delivery, with ventriculoperitoneal shunting. In-utero surgical repair to the fetus has been proposed as a way to improve neurological outcomes; however, the procedure's long-term effects on brain function have not been determined. Reduction in hindbrain herniation has been reported by some authors (Adzick, et al., 2011; Sutton, et al., 1999) as well as reduction in shunt-dependent hydrocephalus (Adzick, et al., 2011; Tulipan, et al., 2003; Bruner, et al., 1999).

Three types of fetal surgery are performed to treat myelomeningocele: fetoscopic myelomeningocele repair; maternal hysterotomy; and microsurgical, three-layered, fetal myelomeningocele repair (fetal patch repair). Myelomeningocele repair consists of closing the dura and skin over the exposed spinal cord.

Maternal complications associated with myelomeningocele repair have been reported and include uterine rupture, placental abruption and maternal bowel obstruction, which may occur as a result of post-hysterotomy adhesions. There is also increased risk of oligohydramnios, pre-term uterine contractions, delivery at earlier estimated gestation and smaller birth weight.

Data evaluating in-utero repair of myelomeningocele is limited; however, there is some evidence to support improved clinical outcomes. Johnson et al. (2003) (n=50) reported overall perinatal survival of 94% with reversal of hindbrain herniation in all fetuses. Ventriculoperitoneal shunting was required in 43% of the fetuses compared to 68–100% in historical controls. Better-than-predicted leg function was demonstrated in 57% of thoracic- and lumbar-level patients. In 2006, Johnson et al. reported the neurodevelopmental and cognitive outcomes in children two years of age who underwent myelomeningocele repair in-utero. Neurodevelopmental deficits were noted but did not appear to be worsened by fetal surgery. The deficits were considered characteristic of children with spina bifida.

Data from the Management of Myelomeningocele Study (MOMS) compared the results of prenatal and postnatal myelomeningocele repair. After recruiting 183 of the planned 200 subjects, the trial was stopped due to significantly improved clinical outcomes for the prenatal surgery group compared to the post-natal treatment group. In 2011, Adzick and colleagues published the results of this trial which included 158 subjects who completed up to 12 months follow-up; 134 of those subjects were also available for evaluation at 30 months. Individuals were randomized to receive myelomeningocele repair in-utero or repair following delivery. Inclusion and exclusion criteria were as follows (See Table 1):

**TABLE 1:**

| Inclusion Criteria MOMS Trial | Exclusion Criteria MOMS Trial |
|---|---|
| • Singleton pregnancy<br>• Myelomeningocele with upper boundary located between T1 and S1<br>• Evidence of hindbrain herniation<br>• Gestational age of 19.0 to 25.9 weeks at randomization<br>• Normal karyotype<br>• U.S. residency<br>• Maternal age of at least 18 years | • Unrelated fetal anomaly<br>• Severe kyphosis<br>• Risk of preterm birth (including short cervix and previous preterm birth)<br>• Placental abruption<br>• Body-mass index of 35 or more<br>• Contraindication to surgery (e.g., including previous hysterotomy in the active uterine segment) |

The primary outcomes measured included fetal death or the need for cerebrospinal fluid shunt by the age of 12 months and at 30 months; a composite score of the Mental Development Index of the Bayley Scales of Infant Development II; and the child's motor function, with adjustment for lesion level. Secondary outcome measures included maternal, fetal, and neonatal surgical and pregnancy complications, and neonatal morbidity and mortality as well as several other secondary outcomes. The authors reported the following results:

- The first primary outcome, fetal death or the need for cerebrospinal fluid shunt by the age of 12 months, was significantly better in the prenatal surgery group (68%) compared to the postnatal surgery group (98%) ($P < 0.001$).
- The rates of actual shunt placement were 40% for the prenatal surgery group compared to 82% in the postnatal surgery group.
- At 12 months of age, the number of infants who had no evidence of hindbrain herniation was higher in the prenatal surgery group compared to the postnatal surgery group (36% versus 4%, respectively).
- At 12 months, the prenatal surgery group also demonstrated lower rates of brainstem kinking, abnormal fourth ventricle location and syringomelia.

The secondary outcome, made up of data from the Bayley Mental Developmental Index and the difference between the functional and anatomical lesion, was calculated at 30 months and was significantly better in the prenatal surgery group (mean 148.6 vs. mean 122.6, $P < 0.007$). In the post hoc analysis, the authors reported that subjects in the prenatal surgery group were more likely to have a level of function two or more levels better than their anatomical level (32% vs., 12%, $P < 0.005$), and were more likely to ambulate without orthotics or other devices (42% vs. 21%, $P < 0.01$). The authors noted the prenatal surgery group had significantly better motor function scores on the Bayley and Peabody motor scales, although this same group had more severe anatomical lesion levels at baseline. Between groups, cognitive scores were not significantly different. The authors acknowledged the prenatal surgery group had significantly higher rates of pre-term birth and uterine dehiscence at delivery; early intervention was associated with both maternal and fetal morbidity. Nonetheless, prenatal surgery for

myelomeningocele reduced the need for shunting and improved motor outcomes at 30 months follow-up. When considering prenatal myelomeningocele repair, the potential benefits of prenatal surgery must be balanced against the risks of prematurity and maternal morbidity. The authors agreed additional follow-up is necessary to assess long-term outcomes and to evaluate the effect of prenatal intervention on bowel and bladder continence, sexual function and mental capacity (Adzick, et al., 2011).

The American College of Obstetricians and Gynecologists (ACOG) and Society for Maternal-Fetal Medicine (SMFM) published a joint committee opinion (ACOG, updated 2017, reaffirmed 2022) acknowledging publication of the MOMS trial and the rigorous requirements for the study. The duo further noted that maternal fetal surgery has significant implications and complications that may occur acutely, postoperatively, for the duration of the pregnancy and in subsequent pregnancies. The Committee recommends that treatment is only offered at facilities with the expertise, multidisciplinary teams, services and facilities to provide the intensive care required for these patients.

The Congress of Neurological Surgeons has published the following evidence-based guidelines and recommendations:

- Management of Patients With Myelomeningocele: Whether Prenatal or Postnatal Closure Affects Future Ambulatory Status
  - When possible, for prenatally diagnosed fetuses with MM who meet maternal and fetal Management of Myelomeningocele Study inclusion criteria, prenatal closure of MM should be performed, which may improve ambulatory status for patients in the short term (at 30 mo of age) (Level II*).
  - Long term benefit for ambulatory status with prenatal closure is unknown. Children who have had either prenatal or postnatal closure should be carefully followed for the development of tethered spinal cord with the associated loss of ambulatory function (Level III) (Bauer, et al., 2019).

- Incidence of Shunt-Dependent Hydrocephalus in Infants With Myelomeningocele After Prenatal Versus Postnatal Repair
  - Prenatal repair of MM is recommended for those fetuses who meet maternal and fetal MOMS specified criteria for prenatal surgery to reduce the risk of developing shunt-dependent hydrocephalus (level I). Differences between prenatal and postnatal repair with respect to the requirement for permanent cerebrospinal fluid (CSF) diversion should be considered alongside other relevant maternal and fetal factors when deciding upon a preferred method of MM closure (Tamber, et al., 2019).

- Incidence of Tethered Cord Syndrome in Infants With Myelomeningocele With Prenatal Versus Postnatal Repair
  - Continued surveillance for tethered cord syndrome and/or the development of inclusion cysts in children with prenatal and postnatal closure of myelomeningocele is indicated (Level II) as there is evidence that prenatal closure may increase the risk of recurrent tethered cord over the baseline rate seen with postnatal closure (Mazzola, et al., 2019).

    *Demonstrating the highest degree of clinical certainty, Class I evidence is used to support recommendations of the strongest type, defined as Level I recommendations. Level II recommendations reflect a moderate degree of clinical certainty and are supported by Class II evidence. Level III recommendations denote clinical uncertainty supported by Class III evidence. evidence.

**Twin-to-Twin Transfusion Syndrome**
Twin-to-twin transfusion syndrome (TTTS) is a condition in which abnormal chorionic vessels in the placenta connect the circulatory systems of two fetuses. As a result, the placenta does not correctly supply oxygen and nutrients to the fetuses' circulation and causes an uneven blood flow to the twins. One twin (the recipient) receives excess blood, and the other (the donor) receives insufficient blood. Increased blood flow to the recipient results in hypervolemia, polyuria and polyhydramnios and, subsequently, in cardiac overload and congestive heart failure. The decreased blood flow to the donor results in hypovolemia, oliguria and oligohydramnios and, subsequently, in anemia and growth retardation. Although it occurs most frequently in twin pregnancies, it may occur in triplet or higher order multiple gestations provided that at least two of the fetuses are monochorionic (Quintero, 2003).

Standard interventions include selective termination, amnioreduction and fetoscopic laser surgery performed percutaneously or through open surgery.

The most severe cases are those diagnosed prior to 25 weeks of gestation. If TTTS is diagnosed in the second trimester and left untreated, the mortality rate rises to 80–90%. By 28 weeks of gestation, chances for survival improve, although the surviving fetus is prone to neurological damage and developmental impairment.

One established therapy for TTTS, amnioreduction, seeks to equalize the volume of amniotic fluid between the twins. This treatment involves serial amniocentesis and is recommended for pregnancies of gestation later than 26 weeks if delivery is not an option. Amnioreduction does not correct the underlying vascular abnormality.

Fetoscopic laser surgery corrects the underlying circulatory imbalance and is preferred treatment, depending on gestational age, location of the placenta and stage of TTTS (CHOP, 2020). The surgery may be performed through an open approach or percutaneously. Laser energy is used to ablate the placental anastomoses, thus interrupting fetal blood-flow transfusion and restoring the circulatory balance. The reported survival rates have improved in recent years and average 70% double twin survival with survival of at least one twin in > 90% of cases following fetoscopic laser treatment (Bamberg and Hecher, 2019).

The laser ablation, which is followed by amnioreduction, may be nonselective or selective. In nonselective laser treatment, all anastomosed vessels that cross the inter-twin septum are ablated, thereby creating a dichorionic placenta. In the selective approach, the ablation is limited to the participating vessels. Fetal and neonatal survival rates following selective ablation are higher than those following nonselective ablation, with a lower rate of spontaneous abortion.

While some research indicates that serial amniocenteses of the polyhydramniotic sac may stabilize the pathophysiological balance, other studies have shown that methods of interrupting the abnormal vascular connections may improve outcomes. In the peer-reviewed, published literature, no single therapy is associated with superior outcomes. Both amnioreduction and laser surgery have resulted in perinatal survival rates of 60–65% (Fisk and Galea, 2004). Roberts et al. (2014) concluded that endoscopic laser coagulation of anastomotic vessels should be considered in the treatment of all stages of TTTS to improve neurodevelopmental outcomes. Further research to assess the effect of treatment on milder and more severe forms of TTTS and long-term survival outcomes are still required.

Published studies evaluating treatment for twin-to-twin transfusion syndrome consist of prospective, retrospective and randomized trials (Kweon, et al., 2019; Kowitt, et al., 2012; Salomon, et al., 2010; Cincotta, et al., 2009; Rossi, et al., 2008; Crombleholme, et al., 2007; Graef, et al., 2006; Bussey, et al., 2004; Senat, et al., 2004; Quintero, et al., 2000). Several

studies lend support to improved health outcomes, including perinatal survival and survival without neurological complications. The World Association of Perinatal Medicine (WAPM) consensus group published recommendations and guidelines for perinatal evaluation and treatment of TTTS (WAPM, 2011) supporting fetoscopic laser coagulation as the treatment of choice and amnioreduction as a palliative treatment that may prolong pregnancy by reducing the risk of polyhydramnios and relieve maternal discomfort. In 2013 (reaffirmed 2014), the Society for Maternal-Fetal Medicine (SMFM) published clinical guidelines for TTTS. Within the guidelines, the SMFM notes perinatal survival for Stage I cases is approximately 86% and that more than three quarters of cases regress or remain stable without invasive intervention. As a result, many of Stage I cases can be managed conservatively. The natural history of an advanced Stage (>III) is bleak. When invasive intervention is warranted, fetoscopic laser photocoagulation is considered, by most experts, as the best available approach for Stage II, III and IV TTTS in pregnancies continuing at 26 weeks. However, the meta-analysis did not support significant survival benefit and long-term neurologic outcomes in the Eurofetus trial. Results were no different than in non-laser treated controls. Despite laser treatment, TTTS is associated with perinatal mortality of 30-50% and long-term neurologic handicap of 5-20%. The SMFM recommends extensive counseling to couples with pregnancies complicated by TTTS.

**Twin Reversed Arterial Perfusion (TRAP) Syndrome**
Twin reversed arterial perfusion (TRAP) syndrome is a condition in which an acardiac/acephalic twin receives all of its blood supply from a normal twin, the so-called "pump" twin. Blood enters the acardiac twin by retrograde flow via the umbilical artery and exits via the umbilical vein. The extra work places an increased demand on the heart of the pump twin, resulting in cardiac failure.

If this condition is left untreated, mortality is 50–75% and occurs frequently, especially when the size of the acardiac/acephalic twin is greater than half that of the pump twin. Twin death occurs in 64% of cases in which the ratio of acardiac to pump twin weight is greater than 50%. The mortality for the pump twin increases to 90% for weight ratios greater than 75. Evaluation consists of three parts: umbilical-cord Doppler velocimetry and echocardiography to determine reversed flow; determination of twin weight ratios; and determination of mono- or diamniotic gestation.

To interrupt the vascular connection between the twins and promote survival of the pump twin, various treatment methods have been used, including hysterotomy and selective removal of the anomalous twin. Fetoscopic occlusion of the anastomotic vessels using ultrasound-guided embolization, ligation of the umbilical cord (e.g., laser photocoagulation) or radiofrequency cord ablation have been described in the literature. Selective removal and embolization have been associated with high morbidity and unreliable outcomes. Radiofrequency ablation, a newer technique, is less invasive compared to photocoagulation and some fetal specialty surgery centers have had promising results using this technique. The results of some studies indicate outcomes are improved with umbilical-cord laser photocoagulation. During this procedure, the umbilical cord root to the affected fetus is coagulated. Quintero and colleagues (2006) reported, and other authors agree, that the surgical approach and technique for treatment of TRAP sequence should be tailored to the specific clinical presentation.

Evidence in the medical literature evaluating treatment for TRAP sequence is limited and consists mostly of retrospective cohort, reviews, case reports, case series involving small populations, and registry data (Zhang, et al., 2018; Sugibayashi, et al., 2016; Anca, et al., 2015; Cabassa, et al., 2013; Lee, et al., 2013; Pagani, et al., 2013; Quintero, et al., 2006; Weisz, et al., 2004; Ville, et al., 1994). The results of these studies however do support improved perinatal survival and favorable clinical outcomes.

**Fetal Urinary-Tract Obstruction**

Lower urinary obstruction in the fetus is an obstruction to the flow of urine out of the bladder, causing backup of urine and damage to the kidneys. The most common cause of bladder obstruction is posterior urethral valves in males although the condition may be linked to a genetic abnormality. The patient selection criteria for intervention are based upon fetal-urine electrolyte studies, beta²-microglobulin levels and the use of ultrasound. The severity of damage at birth depends on the type, degree and duration of the obstruction. In as many as 90% of all fetuses diagnosed with urinary-tract dilatation, intervention is not required (Harrison, 1996). Conditions of minimal renal dysfunction and normal pulmonary development can be treated after delivery. Unilateral obstruction does not lead to oligohydramnios (decrease in amniotic fluid). However, bilateral urinary obstruction in the fetus is often associated with serious adverse outcomes, such as pulmonary hypoplasia secondary to oligohydramnios. Oligohydramnios early in the second trimester results in fetal mortality rate near 100% (Clayton and Brock, 2018). Some authors have investigated endoscopic surgery (i.e., fetoscopic cystoscopy with laser) to visualize the posterior urethral valves, however, the data is limited and further studies are needed to support safety and efficacy. The most common surgical approach to repair the obstruction is vesicoamniotic shunting by means of a shunt or a stent inserted into the urinary tract above the obstruction and then passed through the abdominal wall to drain into the amniotic sac. This method of treatment restores amniotic fluid, preventing pulmonary hypoplasia. In the event that the shunt becomes displaced, or if it cannot be inserted, and if the fetus age is than 22 weeks of gestation, the authors recommend creating a surgical opening in the bladder (vesicostomy). Fetuses with severe renal damage are not considered candidates for this procedure as it is not clear whether decompression can reverse the renal damage.

Evidence demonstrating that early surgical intervention results in improved survival is mainly in the form of small case series with few RCTs. Early surgical intervention is not curative, further evaluation and surgical treatment following delivery are necessary (Wu and Johnson, 2009). Morris et al. (2013) published the results of a RCT (n=31) involving pregnancies complicated by isolated fetal lower urinary tract obstruction with treatment by either conservative management (n=15) or vesicoamniotic shunt placement (n=16) to assess the effect of treatment on survival. Improved survival was associated with renal morbidity. The primary outcome of the trial was survival of the baby to 28 days postnatal. Twelve live births occurred in each group with eight of the shunt group babies and four of the conservative group babies surviving to 28 days; the difference was not statistically significant. At one year follow-up, one baby subsequently died in each subgroup. Overall, the outcomes were poor with only two babies, both from the shunt group, surviving with normal renal function. In the authors opinion, by the time of diagnosis, renal damage may have already taken place and may have been irreversible.

The European Reference Network for Rare Kidney Diseases established a work group to develop recommendations regarding the clinical definition, diagnosis and management of prenatally detected fetal lower urinary tract obstruction (LUTO). The work group recommendations include but are not limited to the following:

- Recommendation 6: Fetuses with prenatal LUTO must be referred to a tertiary obstetric centre with multidisciplinary expertise in prenatal and postnatal management of obstructive uropathies.
- Recommendation 7: Based on existing evidence that vesico- amniotic shunt placement increases perinatal survival in fetuses with LUTO, fetal intervention should be offered in selected cases. However, parents should be made aware of the residual risk of long- term mortality and kidney function impairment (Capone, et al., 2022).

**Congenital Pulmonary Airway Malformation (CPAM)/Congenital Cystic Adenomatoid Malformation (CCAM)**

Congenital Pulmonary Airway Malformation (CPAM), previously termed congenital cystic adenomatoid malformation (CCAM), is a benign cystic pulmonary mass that may lead to fetal hydrops and pulmonary hypoplasia. The CPAM is typically unilateral and unilobular and receives blood supply from the pulmonary vasculature. The condition may result in air trapping and progressive respiratory compromise. Prenatally the lesions are classified as microcystic or macrocystic based on ultrasound examination (Zobel, 2019). Large lesions may cause mediastinal shift and fetal hydrops, pulmonary hypoplasia and persistent pulmonary hypertension. The mortality rate approaches 100% for cases in which both CPAM and fetal hydrops are present. Fortunately, fetal hydrops occurs in fewer than 10% of cases. Most lesions can be successfully treated after birth, and some may resolve prior to birth. It is rare, however, that resolution of hydrops occurs in conjunction with regression of the lesion (Adzick, 1998). When large lesions are identified prior to 26 weeks of gestation, the disease progresses rapidly, ultimately resulting in fetal demise.

Current treatment includes medical therapy, single-needle thoracentesis, thoracoamniotic shunts or open fetal surgery for patients at risk of or who already have developed hydrops (Zobel, et al., 2019). Steroids are effective for treatment of large microcystic lesions. However, thoracentesis and shunting are typically employed for treatment of large multicystic lesions. Resection of CPAM reverses hydrops and improves survival (Adzick, 2009; Adzick, 2003, Adzick, et al., 1998). Treatment for a fetus with fetal hydrops and a large multicystic lesion involves resecting the large, cystic pulmonary lobe (lobectomy). A single, large cyst may be treated by means of a thoracoamniotic shunt. Thoracoamniotic shunting appears to be beneficial in preventing lung hypoplasia in affected fetuses with CPAM (Muntean, et al., 2023; Morikawa, et. al., 2003). Fetal thoracentesis alone is minimally effective for treatment because cystic fluid reaccumulates; nonetheless, the procedure is often performed prior to resection or shunting. Catheter shunt placement has improved neonatal outcomes in some clinical studies. Other treatment options are to terminate the pregnancy or to continue observation.

**Extralobar Pulmonary Sequestration (EPS)**
Bronchopulmonary sequestration is a condition characterized by the presence of nonfunctioning lung tissue which is not connected to the tracheal bronchial tree. It may be intralobar or extralobar. The ability to determine the actual type of sequestration is very limited unless extralobar pulmonary sequestration (EPS) is associated with pleural effusion or is located in the abdomen. No diagnostic landmarks have been found that can identify intralobar sequestration. If not corrected, bronchopulmonary sequestration results in abnormal respiratory functioning and ultimately in fetal hydrops. Large lesions may cause esophageal compression, which may interfere with fetal swallowing of amniotic fluid and eventually result in polyhydramnios. Fetal hydrops develops secondary to vena caval obstruction and cardiac compression. Bronchopulmonary sequestration may also result in a tension hydrothorax from associated fluid or lymph secretion. Treatment includes thoracentesis or in-utero correction involving placement of a thoracoamniotic shunt, both supported mainly by evidence in the form of case reports and reviews (Zobel, et al., 2019; Adzick, 2003; Adzick, et al., 1998).

**Sacrococcygeal Teratoma (SCT)**
A sacrococcygeal teratoma is a tumor derived from more than one embryonic germ layer. Most tumors are benign, but the odds of malignancy increase with increasing age. In many cases, the abnormal size of the uterus (from either the tumor or polyhydramnios) leads to diagnosis by ultrasound. Less commonly, presentation may include maternal pre-eclampsia.

The standard treatment is complete excision after birth if not detected prenatally. When SCT is detected prenatally, early surgical intervention may be performed to prevent the development of fetal hydrops. These are extremely vascular tumors. Fetal hydrops develops as a result of vascular shunting between low-pressure vessels within the tumor, leading to cardiovascular collapse in

cases of large lesions. Left uncorrected, SCT, when it occurs in conjunction with high output failure that is associated with placentomegaly or hydrops, results in 100% fetal mortality.

Additional methods that have been proposed for treating SCT have involved the use of laser ablation, radiofrequency ablation and thermocoagulation. In laser ablation, the vessels leading to the tumor are ablated with the use of a laser. Radiofrequency ablation employs radiofrequency energy for the same purpose. This technique may be performed under ultrasound guidance with minimal access. In thermocoagulation, another minimal-access method, an insulated wire is passed through a needle into the SCT, heating the vessels until blood flow diminishes. Authors propose coagulating the vessels decreases the blood supply to the tumor, decreases cardiovascular demand, and ultimately reverses the fetal hydrops. While minimal access techniques may reduce complications (e.g., preterm labor, premature rupture of membranes) that are often associated with more invasive techniques, these techniques do not support superior outcomes compared to those for percutaneous drainage and open resection (Van Mieghem, et al., 2014). Within a systematic review, Van Mieghem et al. (2014) reported that minimally invasive treated procedures led to a survival rate of 30% while open fetal surgery led to a 55% survival rate. Litwinska et al. (2019) concluded the reported survival from intratumor laser or radiofrequency ablation was about 50%. They cautioned that survival does not mean success, and it remains uncertain whether such interventions are beneficial.

Although there are few published clinical trials, it has been proposed that in-utero resection may reverse the physiologic effects of the tumor and improve fetal survival in a previable fetus. Evidence is mainly in the form of case reports, case series and cohort studies. Surgical resection in cases with evidence of fetal hydrops, placentomegaly and gestational age prior to 32 weeks has shown favorable outcomes compared to cases with untreated fetal hydrops (Hedrick, et al., 2004).

**Aqueductal Stenosis (Hydrocephalus)**
Stenosis of the aqueduct of Sylvius leads to congenital hydrocephalus. The aqueduct of Sylvius is a space that connects the third and fourth ventricles of the brain and allows for flow of cerebrospinal fluid. Obstruction of the flow dilates the ventricles and leads to compression of the brain, eventually compromising brain function. When hydrocephalus is diagnosed, the treatment options include termination or continuation of the pregnancy with monitoring for progression of the disease and detection of additional anomalies. Traditionally, the condition is detected and then treated after birth with a shunt procedure. Researchers suggest that decompressing the ventricles may prevent adverse effects on the developing brain, although in-utero treatment with ventriculoamniotic shunts has not led to improved perinatal outcomes.

If isolated hydrocephalus occurs, it is followed with serial ultrasounds because with increasing length of gestation, the outcome is variable and worsening developmental outcomes may result. Nonetheless, outcomes after early shunting and delivery have been poor; hence, such treatment is not recommended until 32 weeks of gestation.

A moratorium, initially implemented at the fourth annual meeting of the International Fetal Medicine and Surgery Society in 1985, still remains in effect on percutaneous shunting for fetal hydrocephalus.

**Congenital Diaphragmatic Hernia (CDH)**
Congenital diaphragmatic hernia (CDH) is a condition that results in abdominal viscera entering the chest cavity through an opening, or hernia, in the diaphragm. It frequently results in pulmonary hypoplasia and pulmonary hypertension. Outcomes can vary widely, however, depending on the size of the hernia and the timing of herniation. Prognosis depends on the degree of liver herniation, the presence or absence of other anomalies, and the lung-to-head ratio.

Although the condition is correctable after birth, most babies die because of underdeveloped lungs.

In cases without liver herniation, in-utero correction involves reduction of the viscera, reconstruction of the diaphragm, and enlargement of the abdomen to accept the herniated organs. The surgical correction performed on a fetus with liver herniation involves temporary occlusion of the fetal trachea to expand the lungs, thus displacing the viscera back into the abdomen and hastening fetal lung growth. At birth, the tracheal occlusion is then reversed, and the hernia is repaired.

The goal of fetal intervention for CDH is to prevent or reverse hypoplasia and restore adequate lung growth. Three surgical approaches have been attempted in the human fetus for CDH and include: open tracheal clipping, application of a tracheal clip using the fetal endoscopic approach (FETENDO clip), and tracheal balloon occlusion (Arca and Teich, 2004). Reported survival rates for CDH vary widely. Open fetal surgery has failed to demonstrate any advantage and is high risk to both mother and fetus. The use of balloons, sponges or clips generally results in larger but abnormal lungs (Chung, 2012).

Deprest et al. (2021b) published an open-label, randomized, multicenter, parallel-group, superiority trial that studied fetoscopic endoluminal tracheal occlusion (FETO) for infants with severe pulmonary hypoplasia due to isolated congenital left diaphragmatic hernia. Pregnant women (n=80) over the age of 18 with singleton pregnancies, gestational age of less than 29 weeks six days, congenital diaphragmatic hernia on the left side with no other major structural or chromosomal defects, and severe pulmonary hypoplasia were randomized 1:1 into either FETO intervention group or expectant care. For the FETO group, fetoscopic placement of a tracheal balloon was conducted at 27 weeks zero days to 29 weeks six days of gestation. Reversal of occlusion, either by fetoscopy or by ultrasound-guided puncture of the balloon, was scheduled at 34 weeks zero days to 34 weeks six days of gestation. If preterm birth was imminent, emergency balloon retrieval was performed in utero, at the time of delivery while the umbilical cord still connected the infant to the placenta, or by direct puncture immediately after delivery. The primary outcome evaluated was infant survival to discharge from the neonatal intensive care unit (NICU). Operative and pregnancy complications, fetal survival, survival to six months of age, and neonatal complications were also measured. From the FETO group, 40% survived to discharge from the NICU while only 15% survived in the expectant care group. Those that survived discharge from the NICU also survived to six months of age in both groups. Additional outcomes from the FETO group include: 47% preterm, pre-labor rupture of membranes; 75% preterm birth; median gestational age 34 weeks four days; median birth weight 481 grams lower than in the expectant care group; one neonatal death after emergency delivery because of fetoscopic placental laceration from the balloon removal; and one neonatal death due to problems associated with balloon removal. The expectant care group had 11% preterm, pre-labor rupture of membranes, 29% preterm births and median gestational age of 38 weeks three days. The authors also noted that no other serious complications occurred in the women, and there were no obvious between-group differences in the duration of stay in the NICU, the duration of ventilatory support, or the incidence of complications related to preterm birth.

Deprest et al. (2021a) published results of a multicenter, open-label, adaptive, parallel-group, superiority randomized controlled trial for FETO in infants with moderate pulmonary hypoplasia due to isolated congenital left CDH. Pregnant women (n=196) over the age of 18 with singleton pregnancies, gestational age at randomization less than 31 weeks five days, CDH on the left side with no other major structural or chromosomal defects, and moderate pulmonary hypoplasia (defined as the quotient of observed-to-expected lung-to-head ratios of 25.0% to 34.9%, irrespective of liver positions, or 35.0 to 44.9% with intrathoracic liver herniation) were randomized 1:1 into either FETO intervention group or expectant care. For the FETO group,

fetoscopic placement of a tracheal balloon was conducted at 30 weeks zero days to 31 weeks six days of gestation. Reversal of occlusion, either by fetoscopy or by ultrasound-guided puncture of the balloon, was scheduled at 34 weeks zero days to 34 weeks six days of gestation. If preterm birth was imminent, emergency balloon retrieval was performed in utero, at the time of delivery while the umbilical cord still connected the infant to the placenta, or by direct puncture immediately after delivery. The primary outcomes evaluated were infant survival to discharge without bronchopulmonary dysplasia and survival to six months without oxygen supplementation. Surgical and pregnancy complications, fetal and neonatal survival, and complications in early infancy were also measured. From the FETO group, 63% survived to discharge from the NICU and 54% survived without oxygen supplementation at six months of age. Fifty percent survived to discharge in the expectant care group and 44% survived without oxygen supplementation at six months of age. There were no obvious between-group differences in the duration of NICU stay or neonatal complications. Additional outcomes from the FETO group include: 44% preterm, pre-labor rupture of membranes; 64% preterm birth; one unexplained fetal death; two problematic balloon removals with one resulting in infant death; and tracheomalacia diagnosed in one infant and suspected in another. The expectant care group had 12% preterm, pre-labor rupture of membranes, 22% preterm births and one unexplained fetal death.

A systematic review and meta-analysis by Grivell et al. (2015) compared the effects of prenatal versus postnatal interventions for CDH on perinatal mortality and morbidity, longer-term infant outcomes and maternal morbidity. The review also looked to compare the effects of different prenatal interventions with each other. The review included three studies involving 97 women. Two trials examined in-utero fetal tracheal occlusion with standard (postnatal) care in fetuses with severe diaphragmatic hernia. The authors noted that while the trials utilized fetal interventions that were similar, how access was gained to the fetus and timing and mode of delivery varied. As a result, the trials were not combined in meta-analysis and the results were examined in separate comparisons. The third and remaining trial examined the effect of antenatal corticosteroids versus placebo. There was no clear difference in the incidence of perinatal mortality (primary infant outcome) between the group of women who received antenatal corticosteroids and the placebo control.

Minimally invasive approaches using smaller instrumentation are now being investigated. A randomized controlled trial published by Ruano and colleagues (2012) evaluated a minimal access approach using fetal endoscopic tracheal occlusion (FETO) for fetuses with severe CDH. Subjects were randomized to undergo a minimal approach (FETO) (n=20) or to undergo standard postnatal management (n=21). With the FETO technique, a smaller diameter fetoscope was placed percutaneously under ultrasound guidance. The authors hypothesized a smaller diameter fetoscope would result in less complications and improved perinatal survival rates. Fetuses who underwent FETO were delivered by ex-utero intrapartum therapy (EXIT). Fourteen were planned at an average of 37-38 weeks, four were delivered at 34-36 weeks after premature rupture of membranes and five were emergent due to preterm contractions. Fetuses in the control group were delivered by Cesarean section; 15 were planned at 38 weeks gestation and four were emergent. Balloon removal occurred at the time of delivery. The authors noted current guidelines recommend removal six weeks following the initial placement to improve neonatal outcome. In the intention-to-treat analysis, ten of 20 infants in the FETO group survived, while one of 21 infants in the control group survived.

**Amniotic Band Syndrome (ABS)**
Amniotic band syndrome (ABS), also referred to as amnion disruption sequence, constriction ring syndrome or annular constriction rings, is an abnormality that occurs in approximately 1:1,200 to 15,000 live births. The exact cause is unknown; however, authors have proposed that early rupture of the amnion without damage to the chorion sac results in oligohydramnios and formation of amniotic bands. Oligohydramnios results in abnormal pressure on the fetal distal

extremities. Amniotic bands may result in ring constrictions, limb auto-amputations, pseudo-syndactylism and other fetal defects, but does not cause increased risk for the mother during pregnancy. In many cases, ABS is associated with congenital anomalies that are beyond surgical repair, although some cases may result in the isolated constriction of an extremity without amputation. Isolated extremity ABS is not a life-threatening condition (Keswani, et al., 2003). There is currently no effective treatment for ABS, and reconstructive surgery is typically performed in the postnatal period. According to the literature, bands may be snipped after birth, and Z-plasty may be performed on the affected limb. Surgical release of the bands in-utero has been proposed by some authors to avoid amputation or permanent damage to the extremity. Nevertheless, histologic changes, neurological paresis, contractures or hypoplasia persist despite surgical release. Attempts at identifying patient selection criteria for in-utero surgery are currently being investigated. However, at present, there is no prenatal classification available (Singh and Gorla, 2019; Hüsler, et al., 2009).

The evidence in the peer-reviewed scientific literature consists mainly of case reports and is insufficient to support improved patient outcomes with in-utero surgical release of amniotic bands. The reported clinical outcomes vary and may include salvage of an intact limb, a viable extremity with limited function, and a grossly deformed extremity requiring postnatal amputation (Keswani, et al., 2003). Ronderos-Dumit et al. (2006) reported on a case of constriction amniotic bands involving both legs of a fetus with compromising blood flow to the distal extremity. The constriction ring was successfully released in-utero, although the baby underwent Z-plasty of the compromised leg in the postnatal period. While successful lyses of amniotic bands have been reported, further clinical trials are warranted to support the benefit of in-utero surgical release and the avoidance of limb dysfunction.

**Pleural Effusions**
Isolated fetal pleural effusions have an incidence rate of approximately 1:10,000 to 15,000 pregnancies and may be bilateral, but are most commonly unilateral. There are a variety of causes which include congenital abnormalities and chromosomal abnormalities. Congenital hydrothorax is a rare disorder and is defined by the accumulation of fluid in the pleural cavity. Congenital chylothorax, defined as accumulation of chyle in the thoracic cavity, is also a frequent cause of pleural effusions (Harrison and Adzick, 1991). The persistence of pleural effusion in early pregnancy interferes with normal lung development and often results in pulmonary hypoplasia. Mediastinal compression resulting from effusion can cause hemodynamic compromise leading to fetal hydrops and perinatal death. Prenatal intervention is dependent on the severity of fluid accumulation and the gestational age of the fetus at the time of diagnosis. In some cases, spontaneous resolution occurs and no intervention other than observation is indicated. Poor outcomes are generally associated with isolated hydrothorax, and neonatal death rates vary from 55% when diagnosis is made prior to 32 weeks' gestation to 30% when the diagnose is made later (Prontera, et al., 2002). When the condition is associated with hydrops, mortality rates approach 100%. Treatment consists of draining the intrathoracic fluid by the insertion of pleuro-amniotic shunts or by thoracentesis, where liquid is drained after single or multiple transthoracic punctures. Authors agree when diagnosed in early pregnancy (i.e., prior to 32 weeks) the initial treatment of choice is thoracentesis; however, most effusions reaccumulate and often cause fetal hydrops. When reaccumulation of fluid occurs, shunting is recommended. When hydrothorax is diagnosed later in pregnancy (close to term), the treatment is ultrasound-guided thoracentesis or transthoracic puncture immediately after birth.

Successful placement of pleuro-amniotic shunts is supported in the published scientific literature (Chon, et al., 2019; Rocha, et al., 2006; Smith, et al., 2005; Wilson, et al., 2004; Nicolaides and Azar, 1990)**.** Published evidence is however limited to case reports (Hamada, et al., 2001), small case series and retrospective reviews, and the indications for pleuro-amniotic shunting are not well-defined. Nevertheless, authors agree the presence of fetal hydrothorax-induced hydrops or

polyhydramnios are indications for shunting. Some authors have recommended shunting for primary fetal hydrothorax with evidence of effusion under tension, even without hydrops.

**Miscellaneous Conditions**
In-utero fetal surgery has been performed for correction of other fetal abnormalities, such as complete heart block (open or percutaneous placement of pacemaker), treatment of hypoplastic left heart syndrome (laser atrial septotomy), pulmonary-aortic obstruction (percutaneous placement of a balloon catheter to open the stenotic heart valve [i.e., balloon valvuloplasty procedures]), tracheal-atresia stenosis (fetal tracheostomy), cleft lip and palate (in-utero correction to avoid scarring), and fetal stem-cell transplantation for related stem-cell disease (to decrease fetal rejection and need for immuno-suppression). In addition, some authors have investigated in-utero gene therapy for disorders that result in irreversible illness or death in the pre- or neonatal period (e.g., Type 2 Gaucher's Disease, Krabbe's disease, Hurler's Disease). Several concerns exist with in-utero gene therapy regarding safety and efficacy, and further clinical investigation is necessary to support improved patient outcomes. Presently, in-utero gene therapy is not an established treatment modality. Evidence in the published, peer-reviewed scientific literature is inadequate to support improved perinatal outcomes with the use of in-utero fetal surgery to treat these conditions.

**Congenital heart disease:** Diniz et al (2023) conducted a systematic literature review to demonstrate the benefits and risks of fetal interventions in the two most prevalent congenital heart diseases (CHDs), pulmonary stenosis and pulmonary atresia with an intact ventricular septum, but also critical aortic stenosis and hypoplastic left heart syndrome. Nine studies that met the selection criteria were included. The authors reported that fetal cardiac surgery increased right ventricular growth and hemodynamic flow in pulmonary stenosis, whereas in critical aortic stenosis it enabled growth of the left ventricle and increases left ventricular pressure. However, it has a high complication rate, along with considerable morbidity and mortality.

Mendel et al. (2023) conducted a systematic review and meta-analysis to know the outcomes of fetal aortic valvuloplasty in critical aortic stenosis patients. A total of 389 fetal subjects from 10 cohort studies were included in this systematic review and meta-analysis.

- Fetal aortic valvuloplasty (FAV) was successfully done in 84% patients. Most studies reported this procedure performed in 23 or 26 weeks of gestational age. The pooled mortality prevalence from a total of 389 fetal subjects undergoing aortic valvuloplasty was 20%, mostly from intrauterine death. Of fetal aortic valvuloplasty survivors, 33% patients achieved postnatal biventricular circulation. Meanwhile, 27% of them remained in univentricular circulation.
- There were some complications reported related to the procedure. Bradycardia and pericardial effusion requiring treatment were two most common complications, happening in 50–60% patients. Only one maternal complication presented, which was placental abruption in one patient.
- The authors noted a limitation of their analysis is that given the special populations, the participants in this meta-analysis are extremely constrained. Randomized controlled trials are not available yet, hence FAV is still considered as an experimental intervention.

The American Heart Association published a Scientific Statement on Diagnosis and treatment of fetal cardiac disease (Donofrio, et al., 2014). It notes the following:

- Invasive fetal interventions currently exist for the treatment and management of primary extracardiac anomalies. Fetal surgery can be performed with hysterotomy and exposure of the fetus or through laparoscopic techniques with a closed uterus, depending on the anomaly present. Fetal surgery may be reasonable to consider in select congenital anomalies, including large congenital cystic adenomatoid malformations with signs of

hydrops, giant sacrococcygeal teratomas, severe congenital diaphragmatic hernia, and meningomyeloceles (Donofrio, et al., 2014).

**National Institute of Health and Care Excellence (NICE):** In utero fetal surgery has been performed in countries outside the U.S. and is generally regulated by professional societies/organizations similar to those of the U.S. While the conditions for which this type of surgery is being performed vary, it is recommended the procedures be performed in centers specializing in invasive fetal medicine. The National Institute of Health and Care Excellence (NICE) has developed guidelines regarding performance of in utero surgery to treat some fetal anomalies, such as pulmonary atresia, aortic stenosis, twin to twin transfusion syndrome and fetal tumors. According to these guidelines, the following recommendations were given:

- percutaneous fetal balloon valvuloplasty for pulmonary atresia or aortic stenosis has not been proven safe and effective (NICE, 2012; NICE 2018)
- percutaneous laser therapy for sacrococcygeal teratomas, cervical teratomas, cystic hygromas and CCAM has not been proven safe and effective (NICE, 2012)
- fetoscopic prenatal repair of open neural tube defects in the fetus has not been proven safe and effective (NICE, 2020)
- open prenatal repair of open neural tube defects in the fetus has been proven safe and effective (NICE, 2020)
- intrauterine laser ablation of placental vessels for treatment of twin-to-twin transfusion syndrome has been proven safe and effective (NICE, 2012)

## Medicare Coverage Determinations

| | Contractor | Determination Name/Number | Revision Effective Date |
|---|---|---|---|
| NCD | | No determination found | |
| LCD | | No determination found | |

Note: Please review the current Medicare Policy for the most up-to-date information.
(NCD = National Coverage Determination; LCD = Local Coverage Determination)

## Coding Information

**Notes:**

1. This list of codes may not be all-inclusive since the American Medical Association (AMA) and Centers for Medicare & Medicaid Services (CMS) code updates may occur more frequently than policy updates.
2. Deleted codes and codes which are not effective at the time the service is rendered may not be eligible for reimbursement.

**Considered Medically Necessary when criteria in the applicable policy statements listed above are met:**

**Serial Amnioreduction for Twin-to-Twin Transfusion Syndrome (TTTS)**

| CPT®* Codes | Description |
|---|---|
| 59001 | Amniocentesis; therapeutic amniotic fluid reduction (includes ultrasound guidance) |

**Fetoscopic Occlusion of Anastomotic Vessels (e.g., laser photocoagulation, radiofrequency ablation, ligation) for Twin Reversed Arterial Perfusion (TRAP sequence)**

| CPT®* Codes | Description |
|---|---|
| 59072 | Fetal umbilical cord occlusion, including ultrasound guidance |

**Fetal Vesicoamniotic Shunt Procedures for Bilateral Fetal Urinary Tract Obstruction**

| CPT®* Codes | Description |
|---|---|
| 59076 | Fetal shunt placement, including ultrasound guidance |

| HCPCS Codes | Description |
|---|---|
| S2401 | Repair, urinary tract obstruction in the fetus, procedure performed in utero |

**In-utero Needle Access and Open Resection for Sacrococcygeal Teratoma**

| HCPCS Codes | Description |
|---|---|
| S2405 | Repair of sacrococcygeal teratoma in the fetus, procedure performed in utero |

**Fetal Thoracoamniotic Shunt Placement**

| CPT®* Codes | Description |
|---|---|
| 59076 | Fetal shunt placement, including ultrasound guidance |

| HCPCS Codes | Description |
|---|---|
| S2403 | Repair, extralobar pulmonary sequestration in the fetus, procedure performed in utero |

**Myelomeningocele Repair**

| CPT®* Codes | Description |
|---|---|
| 59897 | Unlisted fetal invasive procedure, including ultrasound guidance, when performed |

| HCPCS Codes | Description |
|---|---|
| S2404 | Repair, myelomeningocele in the fetus, procedure performed in utero |

**Nonselective or Selective Fetoscopic Laser Coagulation for Twin-to-Twin Transfusion Syndrome**

| CPT®* Codes | Description |
|---|---|
| 59001 | Amniocentesis; therapeutic amniotic fluid reduction (includes ultrasound guidance) |
| 59897 | Unlisted fetal invasive procedure, including ultrasound guidance, when performed |

| HCPCS Codes | Description |
|---|---|
| S2411 | Fetoscopic laser therapy for treatment of twin-to-twin transfusion syndrome |

**Fetal Lobectomy for Congenital Cystic Adenomatoid Formation (CCAM)/Congenital Pulmonary Airway Malformation (CPAM)**

| CPT®* Codes | Description |
|---|---|
| 59897 | Unlisted fetal invasive procedure, including ultrasound guidance, when performed |

| HCPCS Codes | Description |
|---|---|
| S2402 | Repair, congenital cystic adenomatoid malformation in the fetus, procedure performed in utero |

**Fetoscopic Endoluminal Tracheal Occlusion (FETO) for Left Congenital Diaphragmatic Hernia**

| CPT®* Codes | Description |
|---|---|
| 59897 | Unlisted fetal invasive procedure, including ultrasound guidance, when performed |

| HCPCS Codes | Description |
|---|---|
| S2400 | Repair, congenital diaphragmatic hernia in the fetus using temporary tracheal occlusion, procedure performed in utero |

**Considered Not Medically Necessary when used to report any procedure listed as such in this policy including, but not limited to: laser, thermocoagulation, or radiofrequency ablation techniques for the treatment of sacrococcygeal teratoma or endoscopic approach (i.e., cystoscopy) for the treatment of lower urinary tract obstruction:**

| CPT®* Codes | Description |
|---|---|
| 59897 | Unlisted fetal invasive procedure, including ultrasound guidance, when performed |

| HCPCS Codes | Description |
|---|---|
| S2409 | Repair, congenital malformation of fetus, procedure performed in utero, not otherwise classified |



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## Revision Details

| Type of Revision | Summary of Changes | Date |
|---|---|---|
| Annual Review | • No clinical policy statement changes | 07/15/2024 |