Medical Coverage Policy

Effective Date.....7/15/2025
Next Review Date.....7/15/2026
Coverage Policy Number..... 0175

Fetal Surgery

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Genetic Testing for Reproductive Carrier Screening and Prenatal Diagnosis
Recurrent Pregnancy Loss: Diagnosis and Treatment
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will be denied as not covered. Coverage Policies relate exclusively to the administration of health benefit plans. Coverage Policies are not recommendations for treatment and should never be used as treatment guidelines. In certain markets, delegated vendor guidelines may be used to support medical necessity and other coverage determinations.

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):

- 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.

Health disparities in fetal surgery exist, particularly for conditions like myelomeningocele (MMC) and are influenced by socioeconomic and racial factors. Retrospective analysis of registry data evaluating demographics and socioeconomic variables of women who were offered and opted for open maternal-fetal surgery (OMFS) for MMC compared to postnatal MMC surgery has found that 20% of women underwent OMFS and 74% received postnatal repair. Of these women, 71% were White, 14% were Hispanix/Latinx, 12% were Black, and 3% identified as other or multiple races. Among women who underwent OMFS, 16% were White, 1.7% were Hispanic/Latinx, 0.7% were Black, and 1% identified as other or multiple races. Having private or United States Department of Defense insurance was associated with higher odds of being eligible for OMFS compared with women who were uninsured or had Medicaid. Women who underwent OMFS were more likely to reside in zip codes with a higher median household income and less-distressed communities (Harbert, et al., 2022; Foy, et al., 2021).

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 stress to the fetus when the condition 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 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 because 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 • Myelomeningocele with upper boundary located between T1 and S1 • Evidence of hindbrain herniation • Gestational age of 19.0 to 25.9 weeks at randomization • Normal karyotype • U.S. residency • Maternal age of at least 18 years	• Unrelated fetal anomaly • Severe kyphosis • Risk of preterm birth (including short cervix and previous preterm birth) • Placental abruption • Body-mass index of 35 or more • 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.

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.

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 in-utero surgical intervention (needle access and open resection) may be performed to prevent the development of fetal hydrops. These are extremely vascular tumors. Fetal hydrops develops because 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.

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).

The existing, peer-reviewed literature evaluating the use of FETO for left congenital diaphragmatic hernia consists of open label randomized trials and systematic reviews and meta-analysis of randomized controlled trials and cohort studies (n=41–196). Study results are homogenous and demonstrate a higher survival rate to discharge and at six months without supplemental oxygen in the FETO group compared to expectant care for singleton pregnancies, a fetal gestational age of <30 weeks, a diagnosis of severe pulmonary hypoplasia, and no other major structural or chromosomal defects (Provinciatto, et al., 2024; Deprest, et al., 2021a; Deprest, et al., 2021b; Grivell, et al., 2015; Ruano, et al., 2012).

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.

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 like 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 <u>not</u> 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 and 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:

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

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

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Revision Details

Type of Revision	Summary of Changes	Date
Annual Review	Removed policy statement for: • 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	7/15/2025
Annual Review	No clinical policy statement changes	07/15/2024