Priority Health

MEDICAL POLICY
No. 91120-R6

FETAL SURGERY:
INTRAUTERINE FETAL SURGERY;
FETOSCOPIC LASER SURGERY

Effective Date: September 1, 2025	**Review Dates: 1/93, 2/95, 12/99, 12/01, 4/02, 4/03, 3/04, 3/05, 2/06, 6/06, 6/07, 4/08, 4/09, 4/10, 4/11, 8/11, 8/12, 8/13, 8/14, 8/15, 8/16, 8/17, 8/18, 8/19, 8/20, 8/21, 8/22, 8/23, 8/24, 8/25**
Date of Origin: July 16, 1990	**Status: Current**

Summary of Changes

Changes:

- Fetal endoscopic tracheal occlusion (FETO) to treat congenital diaphragmatic hernia (CDH) is medically necessary when criteria are met.

I. POLICY/CRITERIA

Fetal surgery may be considered medically necessary when evaluated and performed at fetal surgery centers which offer comprehensive evaluation, surgery, support services and follow-up. The following conditions may be considered for fetal surgery when meeting the below associated criteria and the criteria of the fetal surgery center:

A. Vesico-amniotic shunting as a treatment of urinary tract obstruction for fetuses with hydronephrosis due to bilateral urinary tract obstruction when ALL of the following criteria are met:

1. Evidence of progressive oligohydramnios
2. Evidence of adequate renal function
3. No other lethal abnormalities or chromosomal defects
4. Male fetus
5. No other maternal complications

B. Either open in utero resection of malformed pulmonary tissue or placement of a thoraco-amniotic shunt as a treatment of congenital cystic adenomatoid malformation, extralobar pulmonary sequestration or pleural effusion when ALL of the following criteria are met:

1. Fetuses of 32 weeks' gestation or less
2. Evidence of fetal hydrops or high risk of high cardiac output
3. No other lethal abnormalities or chromosomal defects
4. No other maternal complications

C. In utero removal of sacrococcygeal teratoma (SCT) when ALL of the following criteria are met:

1. Fetuses of 32 weeks' gestation or less
2. High risk of high cardiac output
3. No other lethal abnormalities or chromosomal defects
4. No other maternal complications

D. Fetoscopic laser coagulation as a treatment for severe twin-to-twin transfusion syndrome (TTTS) as medically necessary when ALL of the following criteria are met:

1. The gestational age of the fetus is less than 26 weeks
2. There is evidence of polyhydramnios in the recipient fetus
3. The donor fetus is oligohydramniotic
4. There is an absence of other fetal abnormalities
5. No other maternal complications

E. Fetal surgery is considered medically necessary for in utero repair of myelomeningocele when the following conditions are met:

1. Singelton pregnancy; AND
2. Myelomeningocele with the upper boundary of the lesion located between T1 and S1; AND
3. Evidence of hindbrain herniation; AND
4. Gestational age of 19.0 to 25.9 weeks; AND
5. Normal fetal karyotype; AND
6. Absence of ALL of the following:
    i. Fetal anomaly unrelated to the myelomeningocele; AND
    ii. Severe fetal kyphosis; AND
    iii. Short cervix (less than or equal to 15 mm); AND
    iv. Previous pre-term birth; AND
    v. Placental abruption; AND
    vi. Maternal Body Mass Index (BMI) greater than or equal to 35; AND
    vii. Contraindications to surgery, including but not limited to previous hysterotomy in the active (upper) uterine segment.

F. <u>Fetoscopic endoluminal tracheal occlusion (FETO) may be considered medically necessary for treatment of severe congenital diaphragmatic hernia when ALL of the following criteria are met:</u>

1. <u>Singleton pregnancy</u>
2. <u>Gestational age <30 weeks</u>
3. <u>Left-sided diaphragmatic hernia</u>
4. <u>No other major structural or chromosomal defects</u>

5. Severe pulmonary hypoplasia, defined as a quotient of the observed-to-expected lung-to-head ratio of <25.0%

G. Other applications of fetal surgery are considered investigational, including but not limited to fetal surgery for aqueductal stenosis (hydrocephalus), heart block, pulmonary valve or aortic obstruction, tracheal atresia or stenosis, and cleft lip and palate. In utero gene therapy and in utero stem cell transplantation are also considered to be experimental.

II. MEDICAL NECESSITY REVIEW

Prior authorization for certain drug, services, and procedures may or may not be required. In cases where prior authorization is required, providers will submit a request demonstrating that a drug, service, or procedure is medically necessary. For more information, please refer to the Priority Health Provider Manual.

III. APPLICATION TO PRODUCTS

Coverage is subject to member's specific benefits. Group specific policy will supersede this policy when applicable.

- **HMO/EPO:** ***This policy applies to insured HMO/EPO plans.***
- **POS:** ***This policy applies to insured POS plans.***
- **PPO:** ***This policy applies to insured PPO plans. Consult individual plan documents as state mandated benefits may apply. If there is a conflict between this policy and a plan document, the provisions of the plan document will govern.***
- **ASO:** ***For self-funded plans, consult individual plan documents. If there is a conflict between this policy and a self-funded plan document, the provisions of the plan document will govern.***
- **INDIVIDUAL:** ***For individual policies, consult the individual insurance policy. If there is a conflict between this medical policy and the individual insurance policy document, the provisions of the individual insurance policy will govern.***
- **MEDICARE:** ***Coverage is determined by the Centers for Medicare and Medicaid Services (CMS) and/or the Evidence of Coverage (EOC); if a coverage determination has not been adopted by CMS, this policy applies.***
- **MEDICAID/HEALTHY MICHIGAN PLAN:** ***For Medicaid/Healthy Michigan Plan members, this policy will apply. Coverage is based on medical necessity criteria being met and the appropriate code(s) from the coding section of this policy being included on the Michigan Medicaid Fee Schedule located at: http://www.michigan.gov/mdch/0,1607,7-132-2945_42542_42543_42546_42551-159815--,00.html. If there is a discrepancy between this policy and the Michigan Medicaid Provider Manual located at: http://www.michigan.gov/mdch/0,1607,7-132-2945_5100-87572--,00.html, the Michigan Medicaid Provider Manual will govern. If there is a discrepancy or lack of guidance in the Michigan Medicaid Provider Manual, the Priority Health contract with Michigan Medicaid will govern. For Medical Supplies/DME/Prosthetics and Orthotics, please refer to the Michigan Medicaid Fee Schedule to verify coverage.***

IV. DESCRIPTION

The objective of fetal surgery is to correct malformations of the fetus that interfere with organ development and fetal survival and have potentially fatal consequences. The specific techniques used for fetal surgery are determined by the diagnosis and specific circumstances of each individual case.

Intrauterine or in utero fetal surgery (IUFS):

Open fetal surgery involves making an incision through the abdomen of the mother and partial removal of the uterus from the body. Amniotic fluid is drained from the uterus and kept in a warmer for replacement after completion of the surgery. An incision is made in the uterus. Surgery is then performed on the fetus through the opening in the uterus to locate the abnormality and remove or fix it. Because of the nature of open fetal surgery, delivery for this child and all subsequent children of this mother will have to be performed by cesarean section.

Fetoscopic surgery: This type of surgery, which employs minimally invasive techniques, is used more often than open surgery. Surgeons can use fiberoptic telescopes and specially designed instruments to enter the uterus through small surgical openings to correct congenital malformations without major incisions or removing the fetus from the womb. This alternative is less traumatic and reduces the chances of preterm labor.

- **Fetal endoscopic tracheal occlusion (FETO) to treat congenital diaphragmatic hernia (CDH)**

Congenital diaphragmatic hernia is a birth defectof the diaphragm that causes the protrusion of abdominal viscera into the thorax during the fetal period. It impedes the development of peripheral airways and pulmonary vasculature, leading to severely deficient gas exchange and pulmonary hypertension . Severe CDH is associated with high perinatal mortality, with a rate of 75–80%, while moderate CDH has a mortality rate of ~45%. Intrauterine treatment for CDH called fetal endoscopic tracheal occlusion (FETO) has been introduced to minimize pulmonary hypoplasia. By occluding the fetal trachea with a balloon, FETO allows the accumulation of lung fluid to stretch the lungs and enables the growth of airways and vasculature. FETO may induce some complications, including preterm prelabor rupture of membranes (PPROM) and preterm birth. (Chen et al., 2023)

There are two large trials, the TOTAL trials, that demonstrate the safety and efficacy of FETO in fetuses with moderate and severe CDH. Both trials were open-label, multicenter studies that randomly assigned, in a 1:1 ratio, women carrying singleton fetuses with severe (for the severe TOTAL trial) or moderate (for the moderate TOTAL trial) isolated congenital diaphragmatic hernia on the left side to

FETO at 27 to 29 weeks (for severe CDH) or 30 to 32 weeks (for moderate CDH) of gestation or expectant care. The primary outcome for both trials was infant survival to discharge from the neonatal intensive care unit.

In the severe TOTAL trial, the trial was stopped early for efficacy after the third interim analysis. In an intention-to-treat analysis that included 80 women, 40% of infants (16 of 40) in the FETO group survived to discharge, as compared with 15% (6 of 40) in the expectant care group (relative risk, 2.67; 95% confidence interval [CI], 1.22 to 6.11; two-sided P=0.009). Survival to 6 months of age was identical to the survival to discharge (relative risk, 2.67; 95% CI, 1.22 to 6.11). The incidence of preterm, prelabor rupture of membranes was higher among women in the FETO group than among those in the expectant care group (47% vs. 11%; relative risk, 4.51; 95% CI, 1.83 to 11.9), as was the incidence of preterm birth (75% vs. 29%; relative risk, 2.59; 95% CI, 1.59 to 4.52). (Deprest et al., 2022)

The moderate TOTAL trial did not demonstrate the same benefit in survival outcomes. 62 of 98 infants in the FETO group (63%) and 49 of 98 infants in the expectant care group (50%) survived to discharge (relative risk, 1.27; 95% confidence interval [CI], 0.99 to 1.63; two sided P = 0.06). At 6 months of age, 53 of 98 infants (54%) in the FETO group and 43 of 98 infants (44%) in the expectant care group were alive without oxygen supplementation (relative risk, 1.23; 95% CI, 0.93 to 1.65). The incidence of pre-term, prelabor rupture of membranes was higher among women in the FETO group than among those in the expectant care group (44% vs. 12%; relative risk, 3.79; 95% CI, 2.13 to 6.91), as was the incidence of preterm birth (64% vs. 22%, respectively; relative risk, 2.86; 95% CI, 1.94 to 4.34), but FETO was not associated with any other serious maternal complications. (Deprest et al., 2023)

A meta-analysis by Chen and colleagues (2022) which analyzed a total of 1187 CDH fetuses from 20 studies confirmed the findings from the TOTAL trials. FETO significantly reduced 1-month (OR, 0.56 (95% CI, 0.34-0.93); P = 0.02, number needed to treat (NNT) = 7.67) and 6-month (OR, 0.34 (95% CI, 0.18-0.65); P = 0.0009, NNT = 5.26) CDH mortality. Subgroup analysis suggested that the effects of FETO on the rates of pulmonary hypertension and ECMO usage were significant in severe CDH but not in moderate CDH. FETO was also associated with an increased risk of preterm prelabor rupture of membranes before 37 weeks' gestation (OR, 4.94 (95% CI, 2.25-10.88); P < 0.0001, number needed to harm (NNH) = 3.13) and preterm birth before 37 weeks (OR, 5.24 (95% CI, 3.33-8.23); P < 0.00001, NNH = 2.79). However, FETO was not associated with severe complications, such as preterm birth before 32 weeks, placental abruption or chorioamnionitis.

Guidelines/Position Statements

- Diagnosis and Management of Congenital Diaphragmatic Hernia: a 2023 Update from the Canadian Congenital Diaphragmatic Hernia Collaborative (Puligandla et al., 2024)
    - "Fetal endoscopic tracheal occlusion (FETO) should be considered a treatment option and discussed with parents for all cases of severe CDH." (strength of consensus 4, level of evidence A)
    - "FETO may be considered as a treatment option for moderately severe CDH." (strength of consensus 4, level of evidence B-R)

Aqueductal stenosis (hydrocephalus)

Fetal severe central nervous system (CNS) ventriculomegaly is associated with poor neurologic outcomes. Ventriculomegaly refers to ventricular enlargement that is diagnosed prenatally. It is one of the most common fetal anomalies. The diagnosis is made by ultrasound when the arterial diameter of the ventricle is more than 10 mm. Once it is diagnosed, further evaluation by detailed ultrasound, fetal MRI, and genetic studies is required (Alluhaybi, 2022). Severe ventriculomegaly is often secondary to an underlying malformation, deformation, or disruption, and the prognosis is tied to the primary diagnosis such as hydranencephaly, schizencephaly, or Dandy-Walker malformation. Aqueductal stenosis (AS), on the other hand, results in noncommunicating hydrocephalus in an otherwise normally developing brain. Fetal aqueductal stenosis (FAS) is multifactorial in etiology, with acquired causes resulting in intrinsic obstruction being more common prenatally. These include obstruction to the aqueduct secondary to infection or intraventricular hemorrhage or an aqueductal web. Extrinsic compression causes include tectal plate mass/thickening, tectal plate dysplasia, or periaqueductal vascular malformation. Because of its associated poor prognosis, fetal severe ventriculomegaly was an obvious target in early attempts at in utero intervention. (Emery et al., 2020)

In the early 1980s, fetal intervention was explored with the intention of improving outcome (Manning, 1986). However, patient selection was poor. Prenatal surgical management of fetal ventriculomegaly is still limited and associated with high risks. In the prenatal management of fetal ventriculomegaly, the efficacy of intrauterine ventricular shunting procedures is still limited.

Heart block

Congenital heart block (CHB) is a rare fatal condition that may eventually lead to fetal demise, neonatal death, or permanent pacemaker implantation. Incidence of CHB is approximately 1 in 20,000–30,000 live births. CHB may occur in a structurally normal heart (isolated CHB) as a complication of maternal autoimmune disease or in fetuses with congenital heart defects (complex CHB). Prenatal diagnosis of CHB can be achieved early in the second trimester, either incidentally

during intermittent auscultation or during anatomical survey ultrasound, and can be confirmed by fetal echocardiogram with Doppler techniques to determine level of heart block and verify any underlying major structural heart lesions. Several studies have investigated a possible role of immediate post-diagnosis fetal therapy to improve fetal and neonatal outcomes of CHB. Treatment options include fluorinated and non-fluorinated corticosteroids, immunoglobulins or combined treatment. The aim of treatment is to reverse or downgrade CHB and to prevent intrauterine progression of the disease which can be manifested as hydrops fetalis, pericardial effusion, or cardiomegaly, which impacts overall survival rate and lines of treatment. (Michael et al., 2019)

In a systematic review and meta-analysis by Michael and colleagues (2019), the effect of antenatal fluorinated corticosteroids on fetal morbidity and mortality was studied. 12 studies were included in the analysis. The authors found that compared to no treatment, there was no significant difference in incidence of fetal death (OR 1.10, 95%CI 0.65–1.84), neonatal death (OR 0.98, 95%CI 0.41– 2.33), or need for pacing (OR 1.46, 95%CI 0.78–2.74). Heart block downgrade was significantly higher in treatment group (9.48%vs.1.76%, OR 3.27, 95%CI 1.23–8.71). This treatment was found to have no significant effect on improving fetal morbidity and mortality and was associated with a higher incidence of feal and maternal complications.

Pulmonary or aortic valve obstruction

Fetal critical pulmonary stenosis (CPS) is associated with an elevated risk of morbidity and mortality in fetuses and neonates. Untreated CPS may develop into pulmonary atresia with an intact ventricular septum (PAIVS), defined by right ventricle (RV) and tricuspid valve (TV) proportions ranging from standard to varying degrees of hypoplasia. Unfortunately, because of impaired biventricular circulation, most fetuses with severe RV hypoplasia die after birth or in utero. Although postnatal interventions can be performed after the infant is delivered, RV function may remain inadequate, and the possibility for postnatal RV development is limited. (Mendel et al., 2024) Recent research has focused on fetal pulmonary valvuloplasty (FPV) as a potential treatment option for CPS.

In a systematic review by Mendel and colleagues (2024), 8 studies on fetal pulmonary valvuloplasty were included with the primary outcome of mortality. The results showed that the FPV procedure was performed at a mean gestational age of 26.28 weeks (95%CI: 24.83–27.73) and was successful in 87.6 % (95 % CI: 78.3–96.3 %) of patients. A total of 52.9 % patients attained biventricular circulation postnatally (95 % CI: 31.2–74.7 %). Successful FPV was associated with a slightly higher overall mortality rate [periprocedural death 4.7 % (95%CI: 0–10.7 %) and postnatal death 8 % (95%CI: 3–13 %)] compared to the three currently available definitive therapies, namely the Fontan procedure [10 % (95%CI: 4–17 %)], 1.5V repair [11 % (95%CI: 5–17 %), and 2V repair [8 % (95%CI: 1–15 %)].

The most severe congenital heart defect involves severe aortic stenosis (AS) presenting in mid-gestation that progresses to growth restriction of the left ventricle, commonly known as hypoplastic left heart syndrome (HLHS). HLHS is a lethal disease if it remains untreated, and palliative surgery resulting in univentricular circulation (UVC) is currently the only available treatment option. Despite improved postnatal surgical and clinical management of HLHS, morbidity and mortality rates in patients with UVC remain high .Traditional management advocates in-utero observation and postnatal palliative surgery, but advances in fetal cardiac intervention have changed the trajectory of this patient cohort presenting with fetal AS. At present, there are still concerns over technical failure, fetal death, maternal complications and postnatal mortality associated with FAV. Controversies on the risks and benefits of FAV, as well as ethical concerns, are nurtured by a lack of randomized controlled trials (RCTs), low numbers of subjects reported, and short follow-up with limited assessment of the overall success of achieving postnatal biventricular circulation (BVC). (Vorisek et al., 2022)

Vorisek and colleagues (2022) conducted a meta analysis to study postnatal circulation in patients with aortic stenosis undergoing fetal aortic valvuloplasty. 7 studies including 266 fetuses were included who underwent FAV with median follow up per study from 12 months to 13.2 years. There were no maternal deaths and only one case of FAV-related maternal complication was reported. Hydrops was present in 29 (11%) patients. The pooled prevalence of BVC and univentricular circulation (UVC) among liveborn patients was 45.8% (95% CI, 39.2–52.4%) and43.6% (95% CI, 33.9–53.8%), respectively. The pooled prevalence of technically successful FAV procedure was 82.1% (95% CI, 74.3–87.9%), of fetal death it was 16.0%. The pooled prevalence of BVC and UVC among liveborn patients who had technically successful FAV was 51.9% (95% CI, 44.7–59.1%) and39.8% (95% CI, 29.7–50.9%), respectively. The authors concluded that this study showed a BVC rate of 46% among liveborn patients with AS undergoing FAV, which improved to 52% when subjects underwent technically successful FAV. Given the lack of randomized clinical trials, results should be interpreted with caution. Currently, data do not suggest a true benefit of FAV for achieving BVC.

Tracheal atresia or stenosis

Tracheal atresia is an extremely rare condition whereby a partial or total obstruction of the trachea is seen. It is almost always lethal, with just a handful of cases that have ended with a positive outcome. Under normal circumstances, the fluids secreted by the lung are eliminated in the amniotic cavity. In patients with laryngeal or tracheal atresia, there is persistent retention of the liquid inside the lungs, accounting for the hyperechogenic ultrasound aspect and the gross hypervoluminous aspect of the lungs. As a result, the heart is usually tiny, compressed and shifted. Additionally, the intrathoracic pressure is increased, leading to cardiac failure, anemia, ascites and hydrops. (Georgescu et al., 2021)

Unfortunately, there is no robust literature describing in utero surgical correction of tracheal atresia or stenosis as a standard or even experimental intervention. Most interventions are postnatal and involve EXIT-to-airway strategies or palliative care, depending on the severity and associated anomalies.

Cleft lip and palate

Cleft lip and/or palate (CL/P), in its different variations, is the most common facial congenital malformation worldwide, occurring in approximately 1 in 1,000 live births. The malformation can affect several different areas including feeding, dental development, facial growth, speech development, hearing, aesthetics, and psychosocial well-being. Clefts including the palate are typically associated with problems concerning feeding, hearing, speech, and facial growth. (Cornefjord et al., 2023)

Current standard of care is surgical correction of the cleft lip and/or palate postnatally. The available published literature on in-utero surgical management is extremely limited and primarily consists of animal model studies.

In utero gene therapy

For inherited genetic diseases, fetal gene therapy offers the potential of prophylaxis against early, irreversible and lethal pathological change. Massaro (2018) studied neuronopathic Gaucher disease (nGD), caused by mutations in GBA. In adult patients, the milder form presents with hepatomegaly, splenomegaly and occasional lung and bone disease; this is managed, symptomatically, by enzyme replacement therapy. Using this model, fetal intracranial injection of adeno-associated virus (AAV) vector reconstituted neuronal glucocerebrosidase expression. Mice lived for up to at least 18 weeks, were fertile and fully mobile. Neurodegeneration was abolished and neuroinflammation ameliorated. Neonatal intervention also rescued mice but less effectively. To date, no clinical trials with human participants have been completed.

The fetal window offers unique advantages, including enhanced tissue accessibility and biodistribution, immune tolerance to new therapeutic molecules, and has the potential to prevent irreversible organ damage before birth. However, this approach requires ethical considerations including risks to both the fetus and mother, complexities of informed consent, and broad societal implications. (Jeanne et al., 2025)

In utero stem cell transplantation

The widespread availability of high-resolution ultrasound scanning and the rapid advances in prenatal molecular diagnostic techniques, particularly testing for cell free fetal DNA in the maternal blood; this means that congenital disorders are

increasingly diagnosed early in gestation. In utero stem cell transplantation offers the promise of treatment, or even cure, for such genetic disorders. In utero stem cell transplantation is preferably achieved via a single injection, ideally into the umbilical vein at the intrahepatic portion or placental cord insertion, which is a route that is commonly used in fetal medicine practice globally to transfuse the fetus with anemia. There are no large studies reporting on the risks of prenatal stem cell transplantation. Individual case reports from as early as 1967 used such disparate routes of administration, cell types, and conditions treated. (Sagar et al., 2019)

Fetoscopic laser surgery is a procedure for ablating abnormal vascular connections between monozygotic twins who share a single placenta and are affected by twin-twin transfusion syndrome (TTTS). TTTS is a serious and potentially fatal complication resulting from an imbalance in net blood flow from one twin, the donor, to the other, the recipient. Laser energy is used to ablate the placental connecting vessels, thereby interrupting fetofetal blood flow and restoring circulatory balance. The goal of this procedure is to decrease the risk of intrauterine fetal and neonatal death and improve the outcomes of the surviving infants.

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, beta^2-microglobulin levels and the use of ultrasound. The severity of damage at birth depends on the type, degree and duration of the obstruction. 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.

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.

Congenital cystic adenomatoid malformation (CCAM)/congenital pulmonary airway malformation (CPAM): Congenital cystic adenomatoid malformation, recently termed Congenital Pulmonary Airway Malformation (CPAM) is a benign cystic pulmonary mass that may lead to fetal hydrops and pulmonary hypoplasia. The CCAM/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. 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 CCAM/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, 1996). When large lesions are identified prior to 26 weeks of gestation, the disease progresses rapidly, ultimately resulting in fetal demise.

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. Cardiac failure manifests as hydrops, association of fetal hydrops and SCT is usually fatal and always fatal prior to 30 weeks gestation (Gharpure, 2013). SCT types depend on their location, severity, and appearance: Type I: Almost completely external (outside the fetus) and attached to the tailbone; Type II: Mostly external, with a small part of the tumor growing inside the fetus; Type III: Visible externally, but with the tumor extending from the pelvis into the abdomen; Type IV: Completely internal. For some high-risk SCT fetuses, in utero open fetal surgery is an option at specialized centers. Although criteria for open fetal surgery vary across centers, most include fetuses with high-risk SCT and hydrops developing at a gestational age earlier than appropriate for delivery and neonatal care (eg, 28 to 32 weeks gestation) (UptoDate, 2024).

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. 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.

In-utero correction involves placement of a thoracoamniotic shunt and is supported mainly by evidence on the form of case reports and reviews (Adzick, et al, 1998; Adzick, et al, 2003).

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. The persistence of pleural effusion in early pregnancy interferes with normal lung development and often results in pulmonary hypoplasia. 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.

Twin-to-twin transfusion syndrome:
Twin-twin transfusion syndrome (TTTS) results from unbalanced blood flow through vascular arteriovenous anastomoses, which are present in the placenta of the majority of monochorionic multiple pregnancies. With preferential blood flow in severe cases, one twin becomes the donor and the other is the recipient, which may result in wide discordance in fetal growth.

The recipient twin, the larger fetus in the amniotic sac with polyhydramnios, typically has a large umbilical cord, abdominal circumference, kidneys, and bladder. Excessive volume can lead to cardiovascular decompensation with cardiomegaly, tricuspid regurgitation, and ventricular hypertrophy; hydrops fetalis may also develop. Polycythemia in the recipient fetus can lead to thrombosis or hyperbilirubinemia after birth.

The donor twin is the smaller fetus and is in the oligohydramniotic sac. This fetus may have severe growth restriction with anemia, hypovolemia, and renal insufficiency.

Standard interventions include amnioreduction and fetoscopic laser surgery performed percutaneously or through open surgery. The most severe cases are those diagnosed prior to 25 weeks of gestation.

The most widely used 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. Survival rates have been reported to be between 50–65% with this intervention (Children's Hospital of Philadelphia [CHOP]).

Fetoscopic laser surgery corrects the underlying circulatory imbalance. 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 average 67%, with 80% of pregnancies having at least one survivor; laser photocoagulation is associated with reduced neurologic morbidity (CHOP).

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. These include, but are not limited to, physical and mental disabilities, deformity of the extremities, scoliosis, and urinary dysfunction or failure. Some researchers contend that the intrauterine exposure causes 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. 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. 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. ACOG published a committee opinion (ACOG, 2013) acknowledging publication of the MOMS trial and the rigorous requirements for the study. ACOG noted further 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.

V. CODING INFORMATION

ICD-10 Codes:
Not specified - see criteria

HCPCS/CPT Codes:
** PA rules do not apply*

36460* Transfusion, intrauterine, fetal
76941* Ultrasonic guidance for intrauterine fetal transfusion or cordocentesis, imaging supervision and interpretation

59012* Cordocentesis (intrauterine), any method
59015* Chorionic villus sampling, any method
59030* Fetal scalp blood sampling

59070* Transabdominal amnioinfusion, including ultrasound guidance
59072* Fetal umbilical cord occlusion, including ultrasound guidance
59074* Fetal fluid drainage (eg, vesicocentesis, thoracocentesis, paracentesis), including ultrasound guidance
59076 Fetal shunt placement, including ultrasound guidance
59897 Unlisted fetal invasive procedure, including ultrasound guidance (*Use this code in lieu of S codes for Priority Medicare or Medicaid claims. Explanatory notes must accompany all claims)*

S2401 Repair, urinary tract obstruction in the fetus, procedure performed in utero
S2402 Repair, congenital cystic adenomatoid malformation in the fetus, procedure performed in utero
S2403 Repair, extralobar pulmonary sequestration in the fetus, procedure performed in utero
S2404 Repair, myelomeningocele in the fetus, procedure performed in utero
S2405 Repair of sacrococcygeal teratoma in the fetus, procedure performed in utero
S2411 Fetoscopic laser therapy for treatment of twin-to-twin transfusion syndrome
**"S" codes not covered for Priority Medicare, Medicaid*

Not covered:
S2400 Repair, congenital diaphragmatic hernia in the fetus using temporary tracheal occlusion, procedure performed in utero
S2409 Repair, congenital malformation of fetus, procedure performed in utero, not otherwise classified

VI. REFERENCES

Adzick NS. Management of fetal lung lesions. Clin Perinatol. 2009 Jun;36(2):363-76.

Alluhaybi AA, Altuhaini K, Ahmad M. Fetal Ventriculomegaly: A Review of Literature. Cureus. 2022 Feb 18;14(2):e22352. doi: 10.7759/cureus.22352. PMID: 35223331; PMCID: PMC8860673.

Araujo Júnior E, Tonni G, Chung M, Ruano R, Martins WP. Perinatal outcomes and intrauterine complications following fetal intervention for congenital heart disease: systematic review and meta-analysis of observational studies. Ultrasound Obstet Gynecol. 2016 Oct;48(4):426-433. doi: 10.1002/uog.15867. Epub 2016 Sep 15. PMID: 26799734.

Biard JM et al. Long-term outcomes in children treated by prenatal vesicoamniotic shunting for lower urinary tract obstruction. Obstet Gynecol. 2005 Sep;106(3):503-8.

Clayton DB, Brock JW 3rd. In utero intervention for urologic diseases. Nat Rev Urol. 2012 Feb 21;9(4):207-17. doi: 10.1038/nrurol.2012.9. PMID: 22349659.

Evans LL, Harrison MR. Modern fetal surgery-a historical review of the happenings that shaped modern fetal surgery and its practices. Transl Pediatr. 2021 May;10(5):1401-1417. doi: 10.21037/tp-20-114. PMID: 34189101; PMCID: PMC8192985.

Friedman KG, Sleeper LA, Freud LR, Marshall AC, Godfrey ME, Drogosz M, Lafranchi T, Benson CB, Wilkins-Haug LE, Tworetzky W. Improved technical success, postnatal outcome and refined predictors of outcome for fetal aortic valvuloplasty. Ultrasound Obstet Gynecol. 2018 Aug;52(2):212-220. doi: 10.1002/uog.17530. Epub 2018 Jul 4. PMID: 28543953.

Gharpure V. Sacrococcygeal teratoma. J Neonatal Surg. 2013 Apr 1;2(2):28. PMID: 26023448; PMCID: PMC4420369

Hirose S, Farmer DL. Fetal surgery for sacrococcygeal teratoma. Clin Perinatol. 2003 Sep;30(3):493-506. doi: 10.1016/s0095-5108(03)00059-9. PMID: 14533891.

Hedrick HL, Flake AW, Crombleholme TM, Howell LJ, Johnson MP, Wilson RD, Adzick NS. Sacrococcygeal teratoma: prenatal assessment, fetal intervention, and outcome. J Pediatr Surg. 2004 Mar;39(3):430-8; discussion 430-8. Hirose S, Farmer DL. Fetal surgery for sacrococcygeal teratoma. Clin Perinatol. 2003 Sep 1;30(3):493-506.

Luks FI, Johnson A, Polzin WJ; North American Fetal Therapy Network. Innovation in maternal-fetal therapy: a position statement from the North American Fetal Therapy Network. Obstet Gynecol. 2015 Mar;125(3):649-652.

Manning FA, Harrison MR, Rodeck C. Catheter shunts for fetal hydronephrosis and hydrocephalus. Report of the International Fetal Surgery Registry. N Engl J Med. 1986 Jul 31;315(5):336-40. doi: 10.1056/NEJM198607313150532. PMID: 3724830.

Moise KJ Jr, Moldenhauer JS, Bennett KA, Goodnight W, Luks FI, Emery SP, Tsao K, Moon-Grady AJ, Moore RC, Treadwell MC, Vlastos EJ, Wetjen NM. Current Selection Criteria and Perioperative Therapy Used for Fetal Myelomeningocele Surgery. Obstet Gynecol. 2016 Mar;127(3):593-597.

Morris RK1, Malin GL, Quinlan-Jones E, Middleton LJ, Diwakar L, Hemming K, Burke D, Daniels J, Denny E, et al. The Percutaneous shunting in Lower Urinary Tract Obstruction (PLUTO) study and randomised controlled trial: evaluation of the effectiveness, cost-effectiveness and acceptability of percutaneous vesicoamniotic shunting for lower urinary tract obstruction. Health Technol Assess. 2013 Dec;17(59):1-232.

Nationwide Children's Sacrococcygeal Teratoma (SCT). Available at https://www.nationwidechildrens.org/conditions/sacrococcygeal-teratoma-sct (Accessed May 28, 2025).

University of California, Davis. Fetal Care and Treatment Center. Sacrococcygeal teratoma. Available at (Accessed May 28, 2025).

Trigo L, Chmait RH, Llanes A, Catissi G, Eixarch E, Van Speybroeck A, Lapa DA. Revisiting MOMS criteria for prenatal repair of spina bifida: upper gestational-age limit should be raised and assessment of prenatal motor function rather than anatomical level improves prediction of postnatal function. Ultrasound Obstet Gynecol. 2024 Jan;63(1):53-59. doi: 10.1002/uog.27536. PMID: 37970655.

Van Mieghem T, Al-Ibrahim A, Deprest J, Lewi L, Langer JC, Baud D, O'Brien K, Beecroft R, Chaturvedi R, Jaeggi E, Fish J, Ryan G. Minimally invasive therapy for fetal sacrococcygeal teratoma: case series and systematic review of the literature. Ultrasound Obstet Gynecol. 2014 Jun;43(6):611-9. doi: 10.1002/uog.13315. Epub 2014 May 8. PMID: 24488859.

Wu S, Johnson MP. Fetal lower urinary tract obstruction. Clin Perinatol. 2009 Jun;36(2):377-90.

Yadav DK, Acharya SK, Bagga D, Jain V, Dhua A, Goel P. Sacrococcygeal Teratoma: Clinical Characteristics, Management, and Long-term Outcomes in a Prospective Study from a Tertiary Care Center. J Indian Assoc Pediatr Surg. 2020 Jan-Feb;25(1):15-21. doi: 10.4103/jiaps.JIAPS_219_18. Epub 2019 Nov 27. PMID: 31896894; PMCID: PMC6910050.

Aqueductal Stenosis (Hydrocephalus)

Emery, S. P., Narayanan, S., & Greene, S. (2020). Fetal aqueductal stenosis: Prenatal diagnosis and intervention. Prenatal diagnosis, 40(1), 58–65. https://doi.org/10.1002/pd.5527

Cleft Lip and Palate

Cornefjord, M., Arnebrant, K., Guné, H., Holst, J., Klintö, K., Stiernman, M., Svensson, H., Wiedel, A. P., & Becker, M. (2023). A systematic review of differences in outcome between one and two stage palate repair in cleft lip and palate. Journal of plastic surgery and hand surgery, 58, 132–141. https://doi.org/10.2340/jphs.v58.13368

Fetal Endoscopic Tracheal Occlusion

Chen, Y.et al. (2023). Fetal endoscopic tracheal occlusion for congenital diaphragmatic hernia: systematic review and meta-analysis. Ultrasound in obstetrics & gynecology : the official journal of the International Society of Ultrasound in Obstetrics and Gynecology, 61(6), 667–681. https://doi.org/10.1002/uog.26164

Deprest, J. A et al. and TOTAL Trial for Moderate Hypoplasia Investigators (2021). Randomized Trial of Fetal Surgery for Moderate Left Diaphragmatic Hernia. The New England journal of medicine, 385(2), 119–129. https://doi.org/10.1056/NEJMoa2026983

Deprest, J. A., et al, & TOTAL Trial for Severe Hypoplasia Investigators (2021). Randomized Trial of Fetal Surgery for Severe Left Diaphragmatic Hernia.

The New England journal of medicine, 385(2), 107–118. https://doi.org/10.1056/NEJMoa2027030

Puligandla, P., et al. Canadian Congenital Diaphragmatic Hernia Collaborative (2024). Diagnosis and management of congenital diaphragmatic hernia: a 2023 update from the Canadian Congenital Diaphragmatic Hernia Collaborative. Archives of disease in childhood. Fetal and neonatal edition, 109(3), 239–252. https://doi.org/10.1136/archdischild-2023-325865

Heart Block

Michael, A., et al., & Middle-East Obstetrics and Gynecology Graduate Education (MOGGE) Foundation Research Group (2019). Use of antenatal fluorinated corticosteroids in management of congenital heart block: Systematic review and meta-analysis. European journal of obstetrics & gynecology and reproductive biology: X, 4, 100072. https://doi.org/10.1016/j.eurox.2019.100072Hydrocephalus

von Koch CS, Gupta N, Sutton LN, Sun PP. In utero surgery for hydrocephalus. Childs Nerv Syst. 2003 Aug;19(7-8):574-86. doi: 10.1007/s00381-003-0775-4. Epub 2003 Jul 25. PMID: 12955423.

In Utero Gene Therapy and Stem Cell Transplantation

Chauhan DP, Srivastava AS, Moustafa ME, Shenouda S, Carrier E. In utero gene therapy: prospect and future. Curr Pharm Des. 2004;10(29):3663-72. doi: 10.2174/1381612043382828. PMID: 15579062.

Sagar, R., Götherström, C., David, A. L., & Westgren, M. (2019). Fetal stem cell transplantation and gene therapy. Best practice & research. Clinical obstetrics & gynaecology, 58, 142–153. https://doi.org/10.1016/j.bpobgyn.2019.02.007

Jeanne, M., & Chung, W. K. (2025). Opportunities and Challenges of Fetal Gene Therapy. Prenatal diagnosis, 45(6), 764–771. https://doi.org/10.1002/pd.6809\

Massaro G, Mattar CNZ, Wong AMS, Sirka E, Buckley SMK, Herbert BR, Karlsson S, Perocheau DP, Burke D, Heales S, Richard-Londt A, Brandner S, Huebecker M, Priestman DA, Platt FM, Mills K, Biswas A, Cooper JD, Chan JKY, Cheng SH, Waddington SN, Rahim AA. Fetal gene therapy for neurodegenerative disease of infants. Nat Med. 2018 Sep;24(9):1317-1323. doi: 10.1038/s41591-018-0106-7. Epub 2018 Jul 16. PMID: 30013199; PMCID: PMC6130799.

Zanjani ED, Anderson WF. Prospects for in utero human gene therapy. Science. 1999 Sep 24;285(5436):2084-8. doi: 10.1126/science.285.5436.2084. PMID: 10523208.

Myelomeningocele

Adzick NS, Thom EA, Spong CY, Brock JW 3rd, Burrows PK, Johnson MP, Howell LJ, Farrell JA, Dabrowiak ME, Sutton LN, Gupta N, Tulipan NB, D'Alton ME, Farmer DL; MOMS Investigators. A randomized trial of prenatal versus postnatal repair of myelomeningocele. N Engl J Med. 2011 Mar 17;364(11):993-1004.

American College of Obstetricians and Gynecologists. Maternal Fetal Surgery for Myelomeningocele repair. Number 720, September 2017. Committee Opinion; 130 (3); e164-e167

Pulmonary or Aortic Valve Obstruction

Mendel, B., Kohar, K., Yumnanisha, D. A., Djiu, R. J., Winarta, J., Prakoso, R., & Siagian, S. N. (2023). Impact of fetal pulmonary valvuloplasty in in-utero critical pulmonary stenosis: A systematic review and meta-analysis. International journal of cardiology. Congenital heart disease, 15, 100485. https://doi.org/10.1016/j.ijcchd.2023.100485

Vorisek, C. N., Zurakowski, D., Tamayo, A., Axt-Fliedner, R., Siepmann, T., & Friehs, I. (2022). Postnatal circulation in patients with aortic stenosis undergoing fetal aortic valvuloplasty: systematic review and meta-analysis. Ultrasound in obstetrics & gynecology : the official journal of the International Society of Ultrasound in Obstetrics and Gynecology, 59(5), 576–584. https://doi.org/10.1002/uog.24807

Tracheal Atresia or Stenosis

Georgescu, T., Radoi, V., Radulescu, M., Ilian, A., Toader, O. D., Pop, L. G., & Bacalbasa, N. (2021). Prenatal Diagnosis and Outcome of Tracheal Agenesis as Part of Congenital High Airway Obstruction Syndrome. Case Presentation and Literature Review. Medicina (Kaunas, Lithuania), 57(11), 1253. https://doi.org/10.3390/medicina57111253

Twin-to-Twin Infusion

Lopriore E et al. Neonatal outcome in twin-to-twin transfusion syndrome treated with fetoscopic laser occlusion of vascular anastomoses. J Pediatr. 2005 Nov;147(5):597-602.

WAPM Consensus Group on Twin-to-Twin Transfusion, Baschat A, Chmait RH, Deprest J, Gratacós E, Hecher K, Kontopoulos E, Quintero R, Skupski DW, Valsky DV, Ville Y. Twin-to-twin transfusion syndrome (TTTS). J Perinat Med. 2011 Mar;39(2):107-12