

# Fetal Surgeries In Utero

Clinical Policy ID: CCP.1004

Recent review date: 2/2026

Next review date: 6/2027

Policy contains: Fetal surgery; fetoscopy; intrauterine surgery; myelomeningocele repair; spina bifida; teratoma; twin reversed arterial perfusion; twin-to-twin transfusion syndrome; urinary tract obstruction.

*FirstChoice VIP Care has developed clinical policies to assist with making coverage determinations. FirstChoice VIP Care's clinical policies are based on guidelines from established industry sources, such as the Centers for Medicare & Medicaid Services (CMS), state regulatory agencies, the American Medical Association (AMA), medical specialty professional societies, and peer-reviewed professional literature. These clinical policies along with other sources, such as plan benefits and state and federal laws and regulatory requirements, including any state- or plan-specific definition of "medically necessary," and the specific facts of the particular situation are considered, on a case by case basis, by FirstChoice VIP Care when making coverage determinations. In the event of conflict between this clinical policy and plan benefits and/or state or federal laws and/or regulatory requirements, the plan benefits and/or state and federal laws and/or regulatory requirements shall control. FirstChoice VIP Care's clinical policies are for informational purposes only and not intended as medical advice or to direct treatment. Physicians and other health care providers are solely responsible for the treatment decisions for their patients. FirstChoice VIP Care's clinical policies are reflective of evidence-based medicine at the time of review. As medical science evolves, FirstChoice VIP Care will update its clinical policies as necessary. FirstChoice VIP Care's clinical policies are not guarantees of payment.*

## Coverage policy

Fetal surgery in utero is clinically proven and, therefore, may be medically necessary to improve fetal survival for the following conditions and/or diagnoses:

- Myelomeningocele — Open maternal-fetal surgical repair (American College of Obstetricians and Gynecologists, 2017; Mazzola, 2019).
- Twin reversed arterial perfusion sequence — Cord occlusion or ablation (Society for Maternal-Fetal Medicine [Miller, 2024]).
- Twin-to-twin transfusion syndrome — Fetoscopic laser photocoagulation or radiofrequency ablation for stage II to IV presenting between 16 and 26 weeks of gestation, or for stage I if additional factors (e.g., symptomatic maternal polyhydramnios) arise (Miller, 2024).
- Twin anemia–polycythemia sequence — In utero interventions (for example, selective fetoscopic laser photocoagulation, intrauterine transfusions) when advanced disease (greater than or equal to stage II) or coexisting complications (for example, twin-to-twin transfusion syndrome) are present (American Pediatric Surgical Association, 2019; Miller, 2024).
- Lower urinary tract obstruction — Vesicoamniotic shunt with or without cystoscopy (International Lower Urinary Tract Obstruction Working Group [Mustafa, 2024]; Society for Maternal-Fetal Medicine [Norton, 2021]). Congenital pulmonary airway malformation — Thoracoamniotic shunt placement or decompression for macrocystic congenital cystic lung lesions, and high risk for fetal hydrops (American Pediatric Surgical Association, 2018b).

- High-risk sacrococcygeal teratoma — Cyst decompression, open fetal surgery for debulking, or vascular ablation (by fetoscopic laser, radiofrequency, or interstitial ablation with or without vascular coiling) for fetuses < 26 weeks of gestation (American Pediatric Surgical Association, 2018f).
- Abdominal masses — Cyst decompression to treat: bowel obstruction and resulting polyhydramnios; urinary obstruction caused by compression of the genito-urinary system; cysts greater than 4 centimeters (cm) and at greater risk of in utero torsion; or any other significant mass effect (American Pediatric Surgical Association, 2018c).
- Neck masses — Ex-utero intrapartum treatment-to-airway or ex-utero intrapartum treatment-to-resection for high-risk neck masses with polyhydramnios for a fetus < 28 weeks of gestation (American Pediatric Surgical Association, 2018d).

### Limitations

All other indications for fetal surgery are investigational/not clinically proven and, therefore, not medically necessary. Fetal surgery for some indications may be offered through participation in a clinical trial. These indications include, but are not limited to:

- Congenital diaphragmatic hernia — Fetal endoluminal tracheal occlusion (American Pediatric Surgical Association, 2018a).
- Persistent cloaca with hydrocolops (American Pediatric Surgical Association, 2018e).
- Fetoscopic repair of myelomeningocele (American College of Obstetricians and Gynecologists, 2017).
- Open surgical repair of fetal congenital heart disease (American Heart Association [Donofrio, 2014]).

### Alternative covered services

- Post-natal surgical intervention.
- Medical management.

## Background

For decades, experimental fetal surgery proved essential in studying normal fetal physiology and development and pathophysiology of congenital defects. Clinical fetal surgery started in the 1960s with intrauterine transfusions. While 80% of anomalies develop before the 12<sup>th</sup> week of gestation, ultrasound technology has improved the sensitivity for diagnosing common fetal malformations. To overcome the limitations of ultrasound, fetal magnetic resonance imaging may also be used to ensure the accuracy of the prenatal diagnosis (Maselli, 2016).

Most detected defects are best treated after birth and require a modification in the time, mode, and place of delivery for optimal obstetrical and neonatal care. The field of fetal surgery has evolved to encompass many types of fetal interventions, including less invasive options, to treat life-threatening conditions of the fetus and to improve postnatal quality of life (Koehler, 2020).

Conditions that may be considered for fetal surgery include the following:

- Cardiac malformations occur in approximately 3.5 per 1000 pregnancies, but only a select proportion of them possibly benefit from in utero surgery (Gardiner, 2016).
- Myelomeningocele is a neural tube defect and the most severe form of spina bifida. It is the most commonly observed malformation of the central nervous system, affecting more than 1,000 fetuses annually in the United States (American College of Obstetricians and Gynecologists, 2017) .
- Twin-to-twin transfusion syndrome is a complication in which twins sharing a common placenta experience unequal blood exchange through placental arteriovenous anastomoses, resulting in different growth rates and complications. This condition occurs in approximately 10% to 15% of identical twins

who share a placenta. There are five stages of twin-to-twin transfusion syndrome, each reflecting characteristics of pregnancy changes based on the extra blood supplied from the donor twin to the recipient twin. Treatment goals include stopping the sharing of blood and halt progression of the condition, preserving the survival of the healthier twin, and easing maternal pain or discomfort (Children’s Hospital of Philadelphia, 2022e).

- Twin reversed arterial perfusion sequence is a rare condition of monochorionic twin pregnancies in which one twin has an absent or non-functioning heart and receives all of its perfusion from the structurally normal “pump” twin joined by a large blood vessel between their umbilical cords. The acardiac or “parabiotic” twin is not viable and can endanger the pump twin, causing an increased risk of congestive heart failure, intrauterine growth restriction, preterm premature rupture of membranes, and preterm delivery. Intrauterine treatment encompasses hysterotomy and removal of the acardiac twin and cord occlusion methods that interrupt cord blood flow to the acardiac fetus. The goals of treatment are to preserve the survival of the pump twin and approach a term delivery (Children’s Hospital of Philadelphia, 2025d).
- Urinary tract infection or obstruction in the fetus occurs when obstruction to the flow of urine out of the bladder causes 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. Treatments aim to restore urinary flow through the urethra to the amniotic fluid space. Prenatal surgical options include vesicoamniotic shunt placement and amnioinfusion (Children’s Hospital of Philadelphia, 2025b).
- Fetal tumors and cysts (mostly abdominal) are rare and mostly benign. The most commonly diagnosed tumors are fetal teratomas, of which sacrococcygeal teratoma is the most prevalent. In most cases, treatment can be performed successfully in the neonatal period, but some may benefit from fetal intervention. Laparoscopic excision, ultrasound-guided needle aspiration, and drain placement are treatment options depending on the organ involved, the size of the cyst, and symptoms (Children’s Hospital Colorado, 2025c).
- Congenital diaphragmatic hernia occurs when the diaphragm fails to close during prenatal development. This allows abdominal organs to move into the chest, which can affect lung and heart development. Approximately 83% of babies with congenital diaphragmatic hernia have a defect on the left side of the diaphragm. Affected newborns require aggressive surgical repair at delivery and often extracorporeal membrane oxygenation to support heart and lung development. The goal of fetal treatment is to reverse lung damage resulting from lung compression. Fetoscopic endoluminal tracheal occlusion applies a balloon to block the fetus’s trachea to prevent fluid from escaping and to increase pressure to stimulate lung growth (Children’s Hospital of Philadelphia, 2025a).

## Findings

The evidence for the safety and effectiveness of fetal surgery is of low quality consisting of primarily retrospective observational studies and few randomized controlled trials, which prevents determining the superiority of any intrauterine procedure for a particular condition. Fetal surgical intervention can be considered when preterm delivery is contraindicated and the condition can be corrected allowing for normal development to either improve fetal survival or alter the natural history of a condition and improve postnatal outcome. While early surgical intervention may be recommended after a confirmed diagnosis of fetal decompensation, the benefit of early intrauterine procedures as prophylaxis is unclear. In the latter part of pregnancy, standard treatment consists of early delivery and medically necessary interventions rather than fetal surgery.

### Guidelines

Several professional society guidelines have been developed in response to the necessity for correction of various fetal conditions for which surgery is recommended.

The American Pediatric Surgical Association Fetal Diagnosis and Therapy Committee issued expert guidance on the following indications:

- Abdominal masses — Cyst decompression to treat: bowel obstruction and resulting polyhydramnios; urinary obstruction caused by compression of the genito-urinary system; cysts greater than 4 cm to prevent in utero torsion; or any other significant mass effect (2018c).
- Congenital diaphragmatic hernia — Fetal endoluminal tracheal occlusion is offered for severe left-sided congenital diaphragmatic hernia via participation in the TOTAL trial (2018a).
- Congenital pulmonary airway malformation — Thoracoamniotic shunt or decompression for congenital pulmonary airway malformation at high-risk for developing hydrops (2018b).
- High-risk neck masses with polyhydramnios— Ex-Utero Intrapartum Treatment-to-airway or Ex-Utero Intrapartum Treatment-to-resection for high-risk neck masses (defined as tracheoesophageal displacement index > 12 millimeters, magnetic resonance imaging stage II, teratoma pathology) for a fetus less than 28 weeks of gestation (2018d).
- Persistent cloaca with hydrocolops — Limited data exist to support prenatal decompression for persistent cloaca with hydrocolops (2018e).
- Sacrococcygeal teratoma — Cyst decompression, open fetal surgery for debulking, ablation (fetoscopic laser, radiofrequency, or interstitial with or without vascular coiling) for vascular flow interruption may be considered (2018f).
- Twin-twin transfusion syndrome — Fetoscopic coagulation of vascular anastomoses is the standard of care for advanced stage twin-twin transfusion syndrome, frequently during 16 to 26 weeks of gestation. Fetoscopically-guided radiofrequency ablation, amnioreduction, and septostomy may also be considered (2019).

For prenatal repair of myelomeningocele, the American College of Obstetricians and Gynecologists (2017) recommends open maternal-fetal surgery. Fetoscopic repair was not recommended outside of a research protocol due to limited published data on maternal and fetal outcomes. Similarly, the Congress of Neurological Surgeons recommends open fetal spina bifida repair of myelomeningocele in patients who meet maternal and fetal criteria for prenatal surgery, as specified in the Management of Myelomeningocele Study inclusion criteria, to reduce the risk of developing shunt-dependent hydrocephalus (Level I recommendation). The need for permanent cerebrospinal fluid diversion and other relevant maternal and fetal factors should be considered when deciding the optimal method of myelomeningocele closure (Mazzola, 2019).

For twin-twin transfusion syndrome in monochorionic diamniotic twin pregnancies, the Society for Maternal-Fetal Medicine issued recommendations based on Quintero staging in fetuses presenting between 16 and 26 weeks of gestation. For advanced stages II-IV twin-twin transfusion syndrome, the Society recommends fetoscopic laser photocoagulation. For Stage I twin-twin transfusion syndrome, management is more nuanced. The Society recommends expectant management with at least weekly fetal surveillance for asymptomatic patients continuing pregnancies complicated by stage I twin-twin transfusion syndrome, and consideration of fetoscopic laser surgery for stage I twin-twin transfusion syndrome complicated by additional factors such as maternal polyhydramnios symptoms (Miller, 2024).

For twin reversed arterial perfusion, the Society for Maternal-Fetal Medicine recommends fetal therapy for patients with advanced stage (stage ≥ II) identified in the second or early third trimester. Fetal therapy may consist of expectant management, fetoscopic laser surgery, fetal transfusion therapy, and delivery (Miller, 2024).

For lower urinary tract obstruction, the International Lower Urinary Tract Obstruction Working Group issued expert consensus guidance in support of fetal intervention to improve the chance of perinatal survival compared

to conservative management. Vesicoamniotic shunt should be the primary fetal intervention offered, and several different types of shunt are available that might affect efficacy. Serial amnioinfusion should be offered only under research protocols. The group issued no specific recommendations for fetal cystoscopy but indicated that both shunt and cystoscopy and cystoscopy alone have been used at the experts' institutions (Mustafa, 2024). In addition, the Society for Maternal-Fetal Medicine (Norton, 2021) lists cystoscopy with or without ablation of the valve, vesicoamniotic shunting, or amnioinfusion as potential interventions.

For treatment of fetal cardiac disease, the American Heart Association supports fetal interventions that treat primary extracardiac anomalies that affect heart function, such as meningomyeloceles, large congenital pulmonary airway malformation with signs of hydrops, giant sacrococcygeal teratomas, and severe diaphragmatic hernia. The optimal techniques for open surgical repair of fetal congenital heart disease have not been developed and should be pursued on an investigational basis, after carefully weighing the risks to both fetus and mother (Donofrio, 2014).

### Evidence review

#### Cardiac malformations

A systematic review and single-arm meta-analysis examined 485 fetuses with congenital heart disease from 12 observational studies. In the majority of studies, fetal cardiac intervention targeted severe disease, with an emphasis on critical aortic stenosis and its progression toward hypoplastic left heart syndrome. The most applied technique was fetal aortic valvuloplasty, typically ultrasound-guided and sometimes with atrial septoplasty or other adjunct procedures. The pooled overall survival rate was 57.4% (95% confidence interval 39.8% to 73.3%), suggesting moderate survival following prenatal intervention. The pooled perinatal mortality rate was 31.5% (95% confidence interval 21.0% to 44.2%), indicating significant risks associated with these complex procedures. The authors noted substantial heterogeneity across studies, reflecting variability in clinical practice, patient selection, and procedural techniques (de Azevedo Teixeira, 2025).

A qualitative analysis of nine studies (n = 101) addressing fetal cardiac interventions found post-valvuloplasty benefits to right ventricular growth and hemodynamic flow in pulmonary stenosis and benefits to left ventricular growth and pressure in critical aortic stenosis, but also high complication rates, morbidity, and mortality. The most common complications were pericardial effusion requiring drainage and bradycardia requiring treatment. Fetal cardiac intervention was usually undertaken once a substantial risk of morbidity and mortality for the fetus was determined, but uniform criteria for fetal cardiac intervention are needed to avoid unnecessary procedures (Diniz, 2023).

#### Myelomeningocele/spina bifida

The first published randomized controlled trial was the Management of Myelomeningocele Study (n = 158 fetuses) that compared the outcomes of prenatal and postnatal myelomeningocele repair. Shunt replacement rates were higher following postnatal surgery (82% versus 40%). At 30 months, the prenatal surgery group had a higher composite score for mental development and motor function ( $P = .007$ ), and improved hindbrain herniation by 12 months and ambulation by 30 months. Rates of preterm delivery and uterine dehiscence at delivery were higher in the prenatal group (Adzick, 2011). As a result, open classical hysterotomy has become the preferred technique for open fetal spina bifida repair, although it carries the risk of uterine dehiscence/rupture in the index and subsequent pregnancies. Newer fetoscopic approaches may mitigate maternal risk while preserving fetal benefits.

A systematic review and proportional meta-analysis examined the safety and efficacy of various perinatal surgical approaches to open spina bifida repair, including open and fetoscopic maternal–fetal surgery and postnatal surgery. The analysis included 72 cohort studies and six randomized controlled trials representing participants at 28 fetal centers, and the following procedures: open classical hysterotomy; open minihysterotomy;

hybrid fetoscopy; percutaneous fetoscopy; and postnatal surgery. As the majority of studies were non-comparative, only indirect comparisons of outcomes among surgical approaches can be made, and superiority of any one approach cannot be determined (Kunpalin, 2025).

For maternal outcomes associated with prenatal surgery, no cases of maternal death were reported. Common maternal complications included preterm prelabor rupture of membranes and preterm birth. Preterm prelabor rupture of membranes occurred in approximately one-third of pregnant women who underwent fetal open spina bifida repair, and in 32% to 80% among those who underwent fetoscopic surgery. Of those who underwent open classical hysterotomy and later became pregnant again, placenta accreta spectrum disorder and uterine rupture occurred in 4% and 9%, respectively, in any subsequent pregnancy (Kunpalin, 2025).

The rates of perinatal and infant death were similar across all surgical approaches. The most common neonatal complications were mostly consequences of preterm birth. The percutaneous fetoscopy group had the highest risk of respiratory distress syndrome, necrotizing enterocolitis, periventricular leukomalacia, intraventricular hemorrhage, neonatal sepsis, and postnatal surgical wound revision. Compared with postnatal surgery, prenatal surgery was associated with lower need for cerebrospinal fluid diversion surgery at 12 months after birth and higher rates of walking with or without assistive devices at 30 months after birth. The one exception was a lower rate of ambulation at 30 months in the hybrid fetoscopy group. The rates of clean intermittent catheterization for bladder management at 30 months after birth were similar for open classical hysterotomy, percutaneous fetoscopy, and postnatal surgery (Kunpalin, 2025).

Findings from another systematic review and meta-analysis of 32 studies found that, compared to fetoscopic surgery, the open approach was associated with statistically significant improvement in maternal, fetal, and neonatal outcomes with respect to premature rupture of membranes, premature placental abruption, oligohydramnios, birth weight, prematurity less than 37 weeks, surgical time, neonatal surgical wound dehiscence, hydrocephalus, reversal of brainstem herniation, and maintenance or improvement in motor function (all  $P < .05$ ). Only neonatal sepsis in the fetoscopic surgery group was not statistically significant ( $P < .05$ ) (de Oliveira Júnior, 2025).

#### Twin reversed arterial perfusion sequence

A systematic review of 26 studies comparing surgery (cord occlusion or ablation) with conservative management showed superior survival for surgical cases ( $P = .008$ ). Survival was better with ablation than with cord occlusion ( $P = .01$ ) (Mone, 2016).

A meta-analysis of ten studies ( $n = 156$  monochorionic twin pregnancies) found an overall neonatal survival rate after intrafetal laser of 79%. Gestational age at treatment (before or after 16 weeks) did not affect neonatal survival rate. The benefit of early treatment needs to be confirmed in randomized controlled studies (Vitucci, 2022).

In a meta-analysis of 120 studies consisting of 757 pregnancies complicated by twin reversed arterial perfusion sequence, the pump twin's overall survival rate was 77.4%. Of these, 48% underwent radiofrequency ablation and 29% underwent fetoscopic laser ablation. Radiofrequency ablation had the highest technical success (96.5%), though microwave and monopolar ablation each achieved 100% in fewer total cases, while suture cord ligation demonstrated 87.5%. Pump twin survival did not significantly differ across methods, with radiofrequency ablation yielding the highest pump twin survival rate (82.1%). Interventions generally occurred between 18.07 and 22.41 weeks of gestation, and deliveries between 29.59 and 34.74 weeks. Complications included preterm labor (32%), preterm premature rupture of membranes (16.9%), maternal hemorrhage (2.3%), and chorioamnionitis (0.9%). These data suggest that while radiofrequency ablation offers strong technical success, no single modality clearly improves overall pump twin survival, highlighting the need for further prospective research (Stellon, 2025).

## Twin-to-twin transfusion syndrome

A systematic review and meta-analysis of 26 studies examined the survival rates of twins according to the Quintero stage of twin-to-twin transfusion syndrome in participants treated with fetoscopic laser coagulation of placental anastomoses. There was no significant difference in the incidence of double survival ( $P = .933$ ), at least one survivor ( $P = .688$ ), and no survivors ( $P = .866$ ) between stages I and II twin-to-twin transfusion syndrome. There was no significant difference in the incidence of double survival ( $P = .201$ ), at least one survivor ( $P = .380$ ), and no survivors ( $P = .947$ ) between stages III and IV twin-to-twin transfusion syndrome. However, all three survival outcomes were significantly superior in pregnancies with stages I/II compared to pregnancies with advanced stages III and IV (all  $P < .001$ ) (D'Antonio, 2025).

A meta-analysis of five studies compared the outcomes of fetoscopic laser photocoagulation versus expectant management for stage I twin-to-twin transfusion syndrome. There were no significant differences between interventions in terms of at least one fetal survival at birth, gestational age at delivery, preterm premature rupture of membranes less than 32 weeks, preterm birth less than 32 weeks, or single and dual fetal survival (Nassr, 2023).

A systematic review of 17 studies comparing the outcomes of monochorionic pregnancies treated with radiofrequency ablation ( $n = 320$ ) or bipolar cord occlusion ( $n = 480$ ) determined the co-twin death rates, live birth rates, and neonatal death rates were not significantly different between groups. The rate of preterm premature rupture of membranes was significantly lower in the radiofrequency ablation group (17.7% versus 28.2%,  $P = .01$ ) (Gaerty, 2015).

## Urinary tract obstruction

Severe fetal lower urinary tract obstruction is associated with high perinatal mortality and morbidity, primarily due to pulmonary hypoplasia and chronic kidney disease. From the following analyses, vesicoamniotic shunting has demonstrated a reduction in postnatal mortality, and fetal cystoscopy has emerged as a potential treatment alternative.

A systematic review and meta-analysis of 49 observational studies compared obstetrical, survival, and nephro-urological outcomes of prenatal treatment of fetuses diagnosed with lower urinary tract obstruction. Included were 644 (76%) vesicoamniotic shunting procedures and 198 (24%) fetal cystoscopy interventions. Both procedures offered comparable rates of intrauterine fetal death, postnatal death, and long-term renal function. Both interventions were associated with significant perinatal risks, including high rates of preterm birth and procedure-related complications, fetal abdominal wall or intestinal problems, and chorioamnionitis. For vesicoamniotic shunting, shunt migration, obstruction, and displacement occurred in more than 40% of cases. Fetal cystoscopy avoided some of the mechanical complications of shunting, but urological fistula formation occurred in approximately 74% of cases in studies reporting this outcome, along with a significantly higher rate of termination of pregnancy, which the authors surmised may be attributed to the dual role of fetal cystoscopy as both a diagnostic and therapeutic tool (Paraboschi, 2025).

Limitations of the evidence include heterogeneity of study designs, small sample sizes, and a high risk of bias. Factors such as the severity of obstruction, presence of associated anomalies, and gestational age at diagnosis can significantly impact outcomes and influence patient selection. The authors recommended both well-designed randomized controlled trials to directly compare these procedures and wide-spread adoption of standardized classification systems based on disease severity (Paraboschi, 2025).

A systematic review/meta-analysis of 10 studies ( $n = 355$ ) of fetuses with lower urinary tract obstruction found perinatal survival following vesico-amniotic shunt performed in the second trimester was higher than those cases treated conservatively (57.1% versus 38.8%, odds ratio 2.54, 95% confidence interval 1.14 to 5.67) (Saccone, 2020).

## Fetal tumors/cysts

In a systematic review of 27 articles (62 fetuses with sacrococcygeal teratomas) for solid tumors, open fetal surgical intervention appears to be superior to percutaneous intervention with regard to survival to discharge (50% vs. 39.5%,  $P = .02$ ), fewer interventions (0% vs. 31.6%,  $P = .01$ ), and lower procedural failure rates (11.1% vs. 55.9%,  $P = .02$ ) (Menchaca, 2023).

A review of 92 non-randomized studies ( $n = 380$ ) analyzed 324 observed and 56 aspirated cysts. Cysts that underwent ultrasound-guided aspiration had a significantly lower rate of postnatal surgery ( $P < .001$ ) than those treated conservatively. The rate of prenatal torsion in simple cysts  $\geq 40$  mm was lower in aspirated cysts ( $P = .03$ ) (Tyraskis, 2017).

A systematic review/meta-analysis of 34 studies ( $n = 954$ ) showed that, in fetuses undergoing prenatal aspiration of the cyst, recurrence was 37.9%. Rates of ovarian torsion and intracystic hemorrhage diagnosed after birth were 10.8% and 12.8%, respectively, and 21.8% had surgery after birth (Bascietto, 2017).

A review of 59 studies ( $n = 70$ ) of fetuses with pericardial teratoma compared prenatal treatment and non-intervention. Of those treated who were hydropic at intervention, 75.0% had a favorable outcome, compared to 30.8% in controls who developed hydrops (Nassr, 2017).

## Congenital diaphragmatic hernia

In randomized controlled trials analyzed in the following systematic reviews, fetoscopic endoluminal tracheal occlusion improved fetal survival at birth but increased the risk for preterm delivery in fetuses with isolated congenital diaphragmatic hernia. Mild to severe tracheal injury occurred in up to 12% of cases. Fetoscopic endoluminal tracheal occlusion is usually reserved for the most severe cases at risk for pulmonary hypoplasia and death at birth. A lack of standardization in prenatal evaluation and postnatal management approaches may affect the external validity of trial results, but multidisciplinary management and access to extracorporeal membrane oxygenation appear to optimize postnatal outcomes. Data on long-term outcomes are needed.

In a systematic review and meta-analysis of four randomized controlled trials, fetoscopic endoluminal tracheal occlusion was associated with a higher survival rate in fetuses with severe, predominately left-sided congenital diaphragmatic hernia but not with more moderate cases. Fetoscopic endoluminal tracheal occlusion was associated with a higher risk of premature rupture of membranes and preterm delivery. There were no differences between cases and matched controls (expectant management) in terms of the need for supplemental oxygen at both birth and discharge or in the incidence of pulmonary hypertension. The optimal timing of balloon insertion could not be determined (Li, 2022).

A systematic review and meta-analysis of five studies ( $n = 192$ ) and an individual participant analysis ( $n = 150$ ) revealed a significant survival benefit when the procedure was provided in an integrated prenatal and postnatal setting compared to a nonintegrated setting (70.7% vs. 45.7%,  $P = .003$ ). Increased availability of extracorporeal membrane oxygenation was a strong determinant of postnatal survival (odds ratio = 18.8,  $P = .049$ ) (Sferra, 2022).

A systematic review of eight cohort studies and two randomized controlled trials ( $n = 449$ , of whom 228 survived to discharge) found tracheal complications of varying morbidity occurred in 6% of infants born alive and in 12% of those who survived to discharge. The spectrum of tracheal morbidities ranged from mild symptoms (e.g., effort-induced barking cough) to those requiring tracheostomy or tracheal stenting (Tho, 2023).

A reanalysis of data from the two Tracheal Occlusion To Accelerate Lung Growth trials on fetal surgery for congenital diaphragmatic hernia reported an absolute increase in the survival to discharge of 13% (95% confidence interval -1% to 28%,  $P = .059$ ) and 25% (95% confidence interval 6% to 46%,  $P = .0091$ ) for fetuses with moderate and severe pulmonary hypoplasia, respectively. Differences in outcomes between trials may be

related to differences in local practices and underlying disease severity in study populations. The effect of gestational age at the time of balloon insertion on outcomes was uncertain (Van Calster, 2022).

### Thoracic lesions

Congenital thoracic malformations are a group of developmental disorders involving lung parenchyma, arterial supply, and venous drainage. Congenital pulmonary airway malformation, the most common form, and bronchopulmonary sequestration occur at different stages during fetal lung development. In the postnatal period, surgical resection is the treatment of choice for the symptomatic infant with respiratory distress. If there is a risk for fetal hydrops and postnatal treatment is not an option, congenital pulmonary airway malformation can be managed prenatally using corticosteroids, drainage, or fetal surgery to prevent fetal demise (Mehta, 2023).

Results of a systematic review found low level evidence supporting trans-amniotic needle decompression or thoracoamniotic shunting for improving survival when a fetus has a space-occupying cystic lung lesion and is determined to be high risk for hydrops based on imaging characteristics. Fetal lobectomy may be considered for the fetus who does not respond appropriately to a non-operative fetal intervention (e.g., maternal steroid therapy) and is less than 32 weeks of gestational age (Downard, 2017).

In 2024, we deleted several older references and added two indications to the coverage section:

- Thoracoamniotic shunt placement or decompression for macrocystic congenital cystic lung lesions and high risk for fetal hydrops (Downard, 2017).
- Fetal tracheal occlusion for isolated congenital diaphragmatic hernia and high risk for pulmonary hypoplasia (Li, 2022; Van Calster, 2022).

In 2025, we updated the policy to include more specific staging criteria for twin-to-twin transfusion syndrome treatment and added coverage for in utero interventions for twin anemia-polycythemia sequence when advanced disease or coexisting complications are present, based on new guidance from the Society for Maternal-Fetal Medicine (Miller, 2024). Additionally, we added recent findings from Stellon (2025) to the literature review section on twin-reversed arterial perfusion sequence.

In 2026, we updated the references, deleted older, redundant references, and added several new guidance documents from the American Pediatric Surgical Association Fetal Diagnosis and Treatment Committee, the American Heart Association, and the International Lower Urinary Tract Obstruction Working Group. We added several new indications to coverage that align with the new guideline recommendations.

## References

On December 11, 2025, we searched PubMed and the databases of the Cochrane Library, the U.K. National Health Services Centre for Reviews and Dissemination, the Agency for Healthcare Research and Quality, and the Centers for Medicare & Medicaid Services. Search terms were “fetal therapies” (MeSH), “fetal surgery,” “myelomeningocele,” “tumors,” “twin reversed arterial syndrome,” “twin-to-twin transfusion syndrome,” “urinary tract obstruction,” “cardiac anomal\*,” “teratoma,” and “diaphragmatic hernia.” We included the best available evidence according to established evidence hierarchies (typically systematic reviews, meta-analyses, and full economic analyses, where available) and professional guidelines based on such evidence and clinical expertise.

Adzick NS, Thom EA, Spong CY, et al. A randomized trial of prenatal versus postnatal repair of myelomeningocele. *N Engl J Med*. 2011;364(11):993-1004. Doi: 10.1056/NEJMoa1014379.

American College of Obstetricians and Gynecologists. Committee opinion no. 720: Maternal-fetal surgery for myelomeningocele. *Obstet Gynecol*. 2017;130(3):e164-e167. Doi: 10.1097/AOG.0000000000002303.

- American Pediatric Surgical Association Fetal Diagnosis and Treatment Committee. Prenatal Counseling Series. Congenital diaphragmatic hernia. [https://apsaped surg.org/wp-content/uploads/2020/09/APSA-CDH-Brochure\\_Updated-Jul2018\\_FNL.pdf](https://apsaped surg.org/wp-content/uploads/2020/09/APSA-CDH-Brochure_Updated-Jul2018_FNL.pdf). Published 2018. (a)
- American Pediatric Surgical Association Fetal Diagnosis and Treatment Committee. Prenatal Counseling Series. Congenital pulmonary airway malformation. [https://apsaped surg.org/wp-content/uploads/2020/09/APSA-CPAM-Brochure\\_FNL.pdf](https://apsaped surg.org/wp-content/uploads/2020/09/APSA-CPAM-Brochure_FNL.pdf). Published 2018. (b)
- American Pediatric Surgical Association Fetal Diagnosis and Treatment Committee. Prenatal Counseling Series. Fetal abdominal cysts. [https://apsaped surg.org/wp-content/uploads/2020/09/APSA-Fetal-Abdominal-Cysts-Broch\\_FNL.pdf](https://apsaped surg.org/wp-content/uploads/2020/09/APSA-Fetal-Abdominal-Cysts-Broch_FNL.pdf). Published 2018. (c)
- American Pediatric Surgical Association Fetal Diagnosis and Treatment Committee. Prenatal Counseling Series. Fetal neck masses. [https://apsaped surg.org/wp-content/uploads/2020/09/APSA-Fetal-Neck-Masses-Broch\\_FNL.pdf](https://apsaped surg.org/wp-content/uploads/2020/09/APSA-Fetal-Neck-Masses-Broch_FNL.pdf). Published 2018. (d)
- American Pediatric Surgical Association Fetal Diagnosis and Treatment Committee. Prenatal Counseling Series. Persistent cloaca. [https://apsaped surg.org/wp-content/uploads/2020/09/APSA-Persistent-Cloaca-Brochure\\_FNL.pdf](https://apsaped surg.org/wp-content/uploads/2020/09/APSA-Persistent-Cloaca-Brochure_FNL.pdf). Published 2018. (e)
- American Pediatric Surgical Association Fetal Diagnosis and Treatment Committee. Prenatal Counseling Series. Sacrococcygeal tumors. [https://apsaped surg.org/wp-content/uploads/2020/09/APSA-Sacrococcygeal-Tumors-Broch\\_FNL\\_1.pdf](https://apsaped surg.org/wp-content/uploads/2020/09/APSA-Sacrococcygeal-Tumors-Broch_FNL_1.pdf). Published 2018. (f)
- American Pediatric Surgical Association Fetal Diagnosis and Treatment Committee. Prenatal Counseling Series. Twin-twin perfusion syndrome. [https://apsaped surg.org/wp-content/uploads/2020/09/APSA-Twin-Twin-Transfusion-Brochure\\_FNL.pdf](https://apsaped surg.org/wp-content/uploads/2020/09/APSA-Twin-Twin-Transfusion-Brochure_FNL.pdf). Published 2019.
- Bascietto F, Liberati M, Marrone L, et al. Outcome of fetal ovarian cysts diagnosed on prenatal ultrasound examination: Systematic review and meta-analysis. *Ultrasound Obstet Gynecol*. 2017;50(1):20-31. Doi: 10.1002/uog.16002.
- Children's Hospital Colorado. Fetal abdominal cysts. <https://www.childrenscolorado.org/conditions-and-advice/conditions-and-symptoms/conditions/fetal-abdominal-cysts/>. Published 2025.
- Children's Hospital of Philadelphia. Congenital diaphragmatic hernia (CDH). <https://www.chop.edu/conditions-diseases/congenital-diaphragmatic-hernia-cdh>. Last reviewed September 17, 2025. (a)
- Children's Hospital of Philadelphia. Lower urinary tract obstruction (LUTO). <https://www.chop.edu/conditions-diseases/lower-urinary-tract-obstruction-luto>. Published 2025. (b)
- Children's Hospital of Philadelphia. Sacrococcygeal teratoma (SCT). <https://www.chop.edu/conditions-diseases/sacrococcygeal-teratoma-sct>. Published 2025. (c)
- Children's Hospital of Philadelphia. Twin reversed arterial perfusion sequence (TRAP sequence). <https://www.chop.edu/conditions-diseases/twin-reversed-arterial-perfusion-sequence-and-bipolar-cord-coagulation-for-acardiac-acephalic-twins>. Published 2025. (d)
- Children's Hospital of Philadelphia. Twin-twin transfusion syndrome (TTTS). <https://www.chop.edu/conditions-diseases/twin-twin-transfusion-syndrome-ttts>. Published 2025. (e)
- D'Antonio F, Marinceu D, Eltaweel N, Prasad S, Khalil A. Survival rates in pregnancies complicated by twin-to-twin transfusion syndrome undergoing laser therapy: A systematic review and meta-analysis. *Am J Obstet Gynecol MFM*. 2025;7(5):101503. Doi: 10.1016/j.ajogmf.2024.101503.

de Azevedo Teixeira LK, Provinciatto H, Callado GY, de Oliveira Nieblas C, Granese R, Araujo Júnior E. Fetal surgery for congenital heart diseases: A systematic review and single-arm meta-analysis. *Adv Ther.* 2025;42(12):6150-6162. Doi: 10.1007/s12325-025-03388-0.

de Oliveira Júnior JP, Morais BA, Fernandes MNF, et al. Open repair versus fetoscopic fetal surgery: Which is the best approach for intrauterine myelomeningocele correction? Systematic review and meta-analysis. *Childs Nerv Syst.* 2025;41(1):256. Doi: 10.1007/s00381-025-06915-0.

Diniz AMB, Manso PH, Santos MV, Rodrigues AJ, Sbragia L. A systematic review of benefits and risks of fetal surgery for congenital cardiac defects such as pulmonary valve stenosis and critical aortic stenosis. *Braz J Cardiovasc Surg.* 2023;38(3):398-404. Doi: 10.21470/1678-9741-2022-0273.

Donofrio MT, Moon-Grady AJ, Hornberger LK, et al. Diagnosis and treatment of fetal cardiac disease: A scientific statement from the American Heart Association. *Circulation.* 2014;129(21):2183-242. Doi: 10.1161/01.cir.0000437597.44550.5d.

Downard CD, Calkins CM, Williams RF, et al. Treatment of congenital pulmonary airway malformations: A systematic review from the APSA outcomes and evidence based practice committee. *Pediatr Surg Int.* 2017;33(9):939-953. Doi: 10.1007/s00383-017-4098-z.

Gaerty K, Greer RM, Kumar S. Systematic review and meta-analysis of perinatal outcomes after radiofrequency ablation and bipolar cord occlusion in monochorionic pregnancies. *Am J Obstet Gynecol.* 2015;213(5):637-643. Doi: 10.1016/j.ajog.2015.04.035.

Gardiner H, Kovacevic A, Tulzer G, et al; Fetal Working Group of the AEPC. Natural history of 107 cases of fetal aortic stenosis from a European multicenter retrospective study. *Ultrasound Obstet Gynecol.* 2016;48(3):373-381. Doi: 10.1002/uog.15876.

Koehler SM, Knezevich M, Wagner A. The evolution of fetal surgery. *Journal of Fetal Surgery.* 2020;1(1). Open access pub website. <https://openaccesspub.org/jfs/article/548>.

Kunpalin Y, Karadjole VS, Medeiros ESB, et al. Benefits and complications of fetal and postnatal surgery for open spina bifida: Systematic review and proportional meta-analysis. *Ultrasound Obstet Gynecol.* 2025;66(2):135-146. Doi: 10.1002/uog.29240.

Li Q, Liu S, Ma X, Yu J. Fetal endoscopic tracheal occlusion for moderate and severe congenital diaphragmatic hernia: A systematic review and meta-analysis of randomized controlled trials. *Pediatr Surg Int.* 2022;38(9):1217-1226. Doi: 10.1007/s00383-022-05170-7.

Mazzola CA, Assassi N, Baird LC, et al. Congress of Neurological Surgeons systematic review and evidence-based guidelines for pediatric myelomeningocele: Executive summary. *Neurosurgery.* 2019;85(3):299-301. Doi: 10.1093/neuros/nyz261.

Maselli KM, Badillo A. Advances in fetal surgery. *Ann Transl Med.* 2016;4(20):394. Doi: 10.21037/atm.2016.10.34.

Mehta PA, Sharma G. Congenital pulmonary airway malformation. In: *StatPearls* [Internet]. Treasure Island (FL): StatPearls Publishing; 2025 Jan-. <https://www.ncbi.nlm.nih.gov/books/NBK551664/>. Updated August 7, 2023.

Menchaca AD, Olutoye OO, Cass DL, Marwan AI. Percutaneous versus open fetal surgical intervention for sacrococcygeal teratomas: Is less really more? *Fetal Diagn Ther.* 2023;50(5):313-331. Doi: 10.1159/000533634.

- Miller RS, Miller JL, Monson MA, Porter TF, Običan SG, Simpson LL. Society for Maternal-Fetal Medicine Consult Series #72: Twin-twin transfusion syndrome and twin anemia-polycythemia sequence. *Am J Obstet Gynecol*. 2024;231(4):B16-B37. Doi: 10.1016/j.ajog.2024.07.017.
- Mustafa HJ, Khalil A, Johnson S, et al. Fetal lower urinary tract obstruction: International Delphi consensus on management and core outcome set. *Ultrasound Obstet Gynecol*. 2024;64(5):635-650. Doi: 10.1002/uog.27684.
- Mone F, Devaseelan P, Ong S. Intervention versus a conservative approach in the management of TRAP sequence: A systematic review. *J Perinat Med*. 2016;44(6):619-629. Doi: 10.1515/jpm-2015-0165.
- Nassr AA, Hessami K, Zargarzadeh N, et al. Fetoscopic laser photocoagulation versus expectant management for stage I twin-to-twin transfusion syndrome: A systematic review and meta-analysis. *Prenat Diagn*. 2023;43(9):1229-1238. Doi: 10.1002/pd.6413.
- Nassr AA, Shazly SA, Morris SA, et al. Prenatal management of fetal intrapericardial teratoma: A systematic review. *Prenat Diagn*. 2017;37(9):849-863. Doi: 10.1002/pd.5113.
- Norton ME, Cheng Y, Chetty S, et al. SMFM Fetal Anomalies Consult Series #4: Genitourinary anomalies. *Am J Obstet Gynecol*. 2021;225(5):B2-B35. Doi: 10.1016/j.ajog.2021.06.009.
- Paraboschi I, Pierucci UM, Peycelon M, Pelizzo G, Ruano R. Fetal cystoscopy and vesicoamniotic shunting in lower urinary tract obstruction: A meta-analysis of long-term outcome and current technical limitations. *Fetal Diagn Ther*. 2025:1-24. Doi: 10.1159/000547182.
- Saccone G, D'Allessandro P, Escolino M, et al. Antenatal intervention for congenital fetal lower urinary tract obstruction (LUTO): Systematic review and meta-analysis. *J Matern Fetal Neonatal Med*. 2020;33(15):2664-2670. Doi: 10.1080/14767058.2018.1555704.
- Sferra SR, Miller JL, Cortes MS, et al. Postnatal care setting and survival after fetoscopic tracheal occlusion for severe congenital diaphragmatic hernia: A systematic review and meta-analysis. *J Pediatr Surg*. 2022;57(12):819-825. Doi: 10.1016/j.jpedsurg.2022.05.011.
- Stellon MA, Joshi DS, Beninati MJ, et al. Management of twin-reversed arterial perfusion (TRAP) sequence: A systematic review and meta-analysis. *Fetal Diagn Ther*. 2025;52(2):207-222. Doi: 10.1159/000542841.
- Tho ALW, Rath CP, Tan JKG, Rao SC. Prevalence of symptomatic tracheal morbidities after fetoscopic endoluminal tracheal occlusion: A systematic review and meta-analysis. *Arch Dis Child Fetal Neonatal Ed*. 2023;109(1):52-58. Doi: 10.1136/archdischild-2023-325525.
- Tyraskis A, Badalis S, David AL, Eaton S, De Coppi P. A systematic review and meta-analysis on fetal ovarian cysts: Impact of size, appearance and prenatal aspiration. *Prenat Diagn*. 2017;37(10):951-958. Doi: 10.1002/pd.5143.
- Van Calster B, Benachi A, Nicolaidis KH, et al. The randomized tracheal occlusion to accelerate lung growth (total)-trials on fetal surgery for congenital diaphragmatic hernia: Reanalysis using pooled data. *Am J Obstet Gynecol*. 2022;226(4):560.e1-560.e24. Doi: 10.1016/j.ajog.2021.11.1351.
- Vitucci A, Fratelli N, Fichera A, Sartori E, Prefumo F. Timing of intra-fetal laser therapy for twin reversed arterial perfusion (TRAP) sequence: Retrospective series and systematic review and meta-analysis. *Int J Gynaecol Obstet*. 2022;159(3):833-840. Doi: 10.1002/ijgo.14221.

## Policy updates

1/2013: initial review date and clinical policy effective date: 6/2014

10/2019: Policy references updated.

2/2021: Policy references updated.

2/2022: Policy references updated.

2/2023: Policy references updated.

2/2024: Policy references updated. Coverage modified.

2/2025: Policy references updated. Coverage modified.

2/2026: Policy references updated. Coverage modified.

## Related Codes

Below are the most commonly submitted codes for the service(s)/item(s) subject to this policy CCP.1004. This is not an exhaustive list of codes. Providers are expected to consult the appropriate coding manuals and bill accordingly.

Code	Code Description
59001	Amniocentesis; therapeutic amniotic fluid reduction (includes ultrasound guidance)
59072	Fetal umbilical cord occlusion, including ultrasound guidance
59076	Fetal shunt placement, including ultrasound guidance
59074	Fetal fluid drainage (e.g., vesicocentesis, thoracocentesis, paracentesis), including ultrasound guidance
59897	Unlisted fetal invasive procedure, including ultrasound guidance, when performed
S2401	Repair, urinary tract obstruction in the fetus, procedure performed in utero
S2402	Repair, congenital cystic adenomatoid malformation in the fetus, procedure performed in utero
S2403	Repair, extralobar pulmonary sequestration in the fetus, procedure performed in utero
S2404	Repair, myelomeningocele in the fetus, procedure performed in utero
S2405	Repair of sacrococcygeal teratoma in the fetus, procedure performed in utero
S2409	Repair, congenital malformation of fetus, procedure performed in utero, not otherwise classified
S2411	Fetoscopic laser therapy for treatment of twin-to-twin transfusion syndrome
S2400	Repair, congenital diaphragmatic hernia in the fetus using temporary tracheal occlusion, procedure performed in utero