

Clinical Policy: Fetal Surgery in Utero for Prenatally Diagnosed Malformations

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[Coding Implications](#)

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Description

This policy describes the medical necessity requirements for performing fetal surgery. Fetal surgery becomes an option when it is predicted that there will be severe disability or mortality during delivery or after birth.¹

Policy/Criteria

- I. It is the policy of Managed Health Services that in-utero fetal surgery (IUFS) is **medically necessary** for any of the following:
 - A. Sacrococcygeal teratoma (SCT) with treatment including:
 1. Correction via a minimally invasive approach;
 2. SCT resection when meeting all of the following:
 - a. Fetuses with high-risk SCT and hydrops developing at a gestational age earlier than appropriate for delivery and neonatal care (e.g. 28-32 weeks gestation);
 - b. Does not have the following contraindications:
 - i. Type III or IV Altman-type tumors;
 - ii. Severe placentomegaly;
 - iii. Maternal cervical shortening;
 - B. Lower urinary tract obstruction without multiple fetal anomalies or chromosomal abnormalities: urinary decompression via vesico-amniotic shunting;
 - C. Congenital pulmonary airway malformation (CPAM) and extralobar bronchopulmonary sequestration (BPS), with high-risk tumors with treatment including:
 1. Resection of malformed pulmonary tissue, or placement of a thoraco-amniotic shunt when meeting all of the following:
 - a. Hydropic fetus or at risk of developing hydrops;
 - b. Failure of one round of maternal antenatal corticosteroid, unless contraindicated;
 - D. Placement of a thoraco-amniotic shunt for pleural effusion with or without secondary fetal hydrops;
Note: May include fetal pleural aspiration only when performed along with and prior to thoraco-amniotic shunt placement.
 - E. Twin-twin transfusion syndrome (TTTS): treatment approach is dependent on Quintero stage, maternal signs and symptoms, gestational age and the availability of requisite technical expertise and include either of the following:
 1. Amnioreduction;
 2. Fetoscopic laser ablation, with or without amnioreduction when pregnancy is between 16 and 26 weeks gestation;

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- F. Twin-reversed-arterial-perfusion sequence (TRAP): ablation of anastomotic vessels of the acardiac twin (laser, radiofrequency ablation);
- G. Myelomeningocele: repair when all of the following criteria are met:
 - 1. Singleton pregnancy;
 - 2. Upper boundary of myelomeningocele located between T1 and S1;
 - 3. Evidence of hindbrain herniation confirmed on fetal magnetic resonance imaging (MRI);
 - 4. Gestational age between 19 0/7 weeks and 25 6/7 weeks;
 - 5. None of the following:
 - a. Severe fetal kyphosis;
 - b. Risk of preterm birth (e.g., short cervix or previous preterm birth);
 - c. Placental abruption;
 - d. Previous hysterotomy in the active uterine segment;
- H. Fetal endoscopic tracheal occlusion (FETO) for congenital diaphragmatic hernia (CDH) when all of the following criteria are met:
 - 1. Severe left-sided CDH;
 - 2. Singleton pregnancy;
 - 3. Severe pulmonary hypoplasia defined as a quotient of the observed-to-expected lung-to-head ratios of less than 25%;
 - 4. Gestational age $\leq$ 30 weeks;

II. It is the policy of Managed Health Services that all repeat utero fetal surgery procedures require secondary review.

III. It is the policy of Managed Health Services that current evidence does not support the use of in utero fetal surgery for any of the following indications:

- A. Surgery for heart block, pulmonary valve, or aortic obstruction;
- B. Tracheal atresia or stenosis;
- C. Cleft lip and palate;
- D. In-utero stem cell transplantation;
- E. In-utero gene therapy;
- F. Amnioexchange procedure for gastroschisis.

Background*Maternal–Fetal Surgery*

Maternal–fetal surgery is a major procedure for the pregnant individual and their fetus(es), and it has significant implications and complications that could occur acutely, postoperatively, for the duration of the pregnancy, and in subsequent pregnancies. For the fetus, safety and effectiveness are variable and depend on the specific procedure, the reasons for the procedure, and the gestational age and condition of the fetus. Often neonates who have been operated on in this manner are born pre-term.

Therefore, it should only be offered at facilities with the expertise, multidisciplinary teams, services, and facilities to provide the intensive care required for these patients.¹

Fetal surgery approaches can be divided into two categories²:

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- Open fetal surgery is considered when the fetal condition is life threatening, and the intervention is felt to be the only option for fetal survival. During open fetal surgery, a hysterotomy is performed, the fetus is partially removed to expose the area that needs surgery, the fetal abnormality is corrected, and the fetus is returned to the uterus where it continues to develop until delivery.
- Fetoscopic surgery employs minimally invasive techniques and uses small fiberoptic telescopes and instruments to enter the uterus through small surgical openings to correct congenital malformations without major incisions or removing the fetus from the womb. This interim procedure is less traumatic, reduces the chances of preterm labor, and allows the fetus to remain in utero until it has matured enough to survive delivery and neonatal surgical procedures.

Sacrococcygeal germ cell tumors

The prenatal diagnosis of sacrococcygeal teratoma (SCT) typically occurs during the second trimester during routine sonography. Despite improved outcomes for SCT with prenatal diagnosis and close monitoring, perinatal mortality remains high. Identifying fetuses at increased risk of fetal demise due to hydrops fetalis and intervening appropriately is the primary goal. Hydrops fetalis is a condition of excess fluid accumulation in the fetus that results in significant fetal demise and neonatal mortality. Criteria for open fetal surgery varies, but most centers include fetuses with high-risk SCT and hydrops that have developed at a gestational age too early for appropriate delivery and neonatal care. Type III or IV Altman type tumors, severe placentomegaly, cervical shortening, and maternal medical issues are all contraindications for open fetal surgery for SCT.^{2,3}

Lower Urinary Tract Obstruction

The prenatal diagnosis of lower urinary tract obstructions typically occurs during the first or second trimester during routine sonography. Outcomes range from clinically insignificant to in-utero fetal demise. Vesicoamniotic shunts can be a temporizing measure and provide a survival advantage in a select cohort of fetuses with urinary tract obstruction.⁴

Congenital pulmonary airway malformation (CPAM)

CPAM is one of the most common lung lesions diagnosed prenatally, although the birth prevalence is quite low. Prenatal diagnosis is typically made by ultrasonography. CPAMs presenting prenatally are classified as macrocystic or microcystic based on ultrasound appearance. Approximately 50% of the masses resolve before delivery, while the remainder persists until delivery. Hydrops can develop with either micro or macrocystic lesions due to compression of lymphatic structures or due to hemodynamic alterations from vena cava obstruction or cardiac displacement/compression.⁵

The presence of hydrops is a sign for impending fetal demise (risk of death approaches 100% in the absence of intervention), and thus it is an indication for fetal intervention. For hydropic fetuses over 32 to 34 weeks of gestation, early delivery with immediate postnatal resection is a reasonable option. Ex utero intrapartum therapy (EXIT) has been used to stabilize fetuses with large lesions expected to have difficulty breathing at delivery. In EXIT, the fetus is partially delivered and intubated without clamping the umbilical cord. Uteroplacental blood flow and gas exchange are maintained by using inhalational agents to provide uterine relaxation and

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amnioinfusion to maintain uterine volume. This provides time for resection of the lung mass prior to complete delivery of the infant. For hydropic fetuses between 20 and 32 weeks of gestation, the choice of the best invasive approach depends on the type of anomaly (macro-versus microcystic). Drainage procedures are used for CPAMS with dominant cysts, while solid masses are treated by resection or ablation.⁵

Twin-twin transfusion syndrome (TTTS)

TTTS occurs in approximately 10 to 15% of monochorionic–diamniotic twin pregnancies and results from the presence of arteriovenous anastomoses in a monochorionic placenta. In the affected pregnancy, there is an imbalance in the fetal–placental circulations, whereby one twin transfuses the other. It usually presents in the second trimester. Once the diagnosis of TTTS has been made, the prognosis depends on gestational age and severity of the syndrome. Staging is commonly performed via the Quintero staging system, and treatment is by laser coagulation or amnioreduction, often in collaboration with an expert in TTTS diagnosis and management.⁶

Clinical studies have shown that in severe cases of twin–twin transfusion syndrome diagnosed at very early gestational ages (<26 weeks), selective fetoscopic laser coagulation, compared with serial amnioreduction, is associated with higher survival of at least one fetus, reduced neurologic morbidity among survivors, and prolongation of gestation resulting in a more advanced gestational age at delivery.³²

Twin reversed arterial perfusion (TRAP)

TRAP sequence is a rare unique serious complication of monochorionic twin pregnancy in which a twin with an absent or a nonfunctioning heart (“acardiac twin”) is perfused by its co-twin (“pump twin”) via placental arterial anastomoses. The acardiac twin usually has a poorly developed heart, upper body, and head. The pump twin is at risk of heart failure and problems related to preterm birth. Current treatment modalities target occlusion of the umbilical cord of the acardiac twin and include laser coagulation, bipolar cord coagulation, and radiofrequency ablation.⁷

Myelomeningocele

Per the American College of Obstetricians and Gynecologists (ACOG) and the Society for Maternal–Fetal Medicine (SMFM), open maternal–fetal surgery for myelomeningocele repair has shown improvement in pediatric outcomes, but poses procedure-associated maternal and fetal risks. According to ACOG and SMFM recommendations for myelomeningocele repair, women who meet specific criteria for in utero repair should be counseled about all management options, including open maternal-fetal surgery. A referral for additional assessment and consultation to a fetal therapy center should be completed for candidates interested in fetal myelomeningocele repair. These centers have the expertise, resources, and multi-disciplinary team to provide the information and intensive care needed for patients choosing to undergo open maternal-fetal surgery.¹

Congenital diaphragmatic hernia

CDH is the result of abnormal development of the diaphragm. The resulting defect permits abdominal viscera to enter the chest, displacing the lungs and heart within the thoracic cavity. This interferes with normal lung development and frequently leads to lung hypoplasia. CDH

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varies in its severity, depending on the size of the hernia and timing of the herniation. The defect may range from a small window within the diaphragm to complete absence thereof.

The pathophysiology of CDH is a combination of lung hypoplasia and immaturity associated with persistent pulmonary hypertension of newborn (PPHN) and cardiac dysfunction. Some fetuses may be candidates for post-natal management. While others with more severe presentations and therefore poorer prognosis, like large defects that occur earlier in gestation, may be considered candidates for fetal surgery. The two primary approaches for treatment of CDH in-utero are surgical repair of the herniated diaphragm, and ligation of the fetal trachea, with subsequent stimulation of lung growth. The latter is referred to as fetal endoscopic tracheal occlusion (FETO). The goal of FETO is to prevent or reverse pulmonary hypoplasia and restore adequate lung growth for neonatal survival.^{27, 33-35}

Coding Implications

This clinical policy references Current Procedural Terminology (CPT[®]). CPT[®] is a registered trademark of the American Medical Association. All CPT codes and descriptions are copyrighted 2025, American Medical Association. All rights reserved. CPT codes and CPT descriptions are from the current manuals and those included herein are not intended to be all-inclusive and are included for informational purposes only. Codes referenced in this clinical policy are for informational purposes only. Inclusion or exclusion of any codes does not guarantee coverage. Providers should reference the most up-to-date sources of professional coding guidance prior to the submission of claims for reimbursement of covered services.

Note: Per IHCP, the following codes are noncovered:

- **59072** – Umbilical cord occlusion with ultrasound
- **S2401** – Fetal surgery for urinary tract obstruction
- **S2402** – Fetal surgery for congenital cystic malformation
- **S2403** – Fetal surgery for pulmonary sequestration
- **S2404** – Fetal surgery for myelomeningocele
- **S2405** – Fetal surgery for sacrococcygeal teratoma
- **S2409** – Fetal surgery, not otherwise classified
- **S2411** – Fetoscopic laser therapy for twin-to-twin transfusion syndrome [39] [40]

CPT [®] Codes	Description
59001	Amniocentesis; therapeutic amniotic fluid reduction (includes ultrasound guidance)
59076	Fetal shunt placement, including ultrasound guidance
59897	Unlisted fetal invasive procedure, including ultrasound guidance, when performed
59072	Fetal umbilical cord occlusion, including ultrasound guidance
59074	Fetal fluid drainage (eg, vesicocentesis, thoracocentesis, paracentesis), including ultrasound guidance

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

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HPCPS Codes	Description
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

Reviews, Revisions, and Approvals	Revision Date	Approval Date
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Important Reminder

This clinical policy has been developed by appropriately experienced and licensed health care professionals based on a review and consideration of currently available generally accepted standards of medical practice; peer-reviewed medical literature; government agency/program approval status; evidence-based guidelines and positions of leading national health professional organizations; views of physicians practicing in relevant clinical areas affected by this clinical policy; and other available clinical information. The Health Plan makes no representations and accepts no liability with respect to the content of any external information used or relied upon in developing this clinical policy. This clinical policy is consistent with standards of medical

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This clinical policy is effective as of the date determined by the Health Plan. The date of posting may not be the effective date of this clinical policy. This clinical policy may be subject to applicable legal and regulatory requirements relating to provider notification. If there is a discrepancy between the effective date of this clinical policy and any applicable legal or regulatory requirement, the requirements of law and regulation shall govern. The Health Plan retains the right to change, amend or withdraw this clinical policy, and additional clinical policies may be developed and adopted as needed, at any time.

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Note: For Medicaid members/enrollees, when state Medicaid coverage provisions conflict with the coverage provisions in this clinical policy, state Medicaid coverage provisions take precedence. Please refer to the state Medicaid manual for any coverage provisions pertaining to this clinical policy.

Note: For Medicare members/enrollees, to ensure consistency with the Medicare National Coverage Determinations (NCD) and Local Coverage Determinations (LCD), all applicable NCDs, LCDs, and Medicare Coverage Articles should be reviewed prior to applying the criteria

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set forth in this clinical policy. Refer to the CMS website at <http://www.cms.gov> for additional information.

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