

Clinical Policy: Fetal Surgery in Utero for Prenatally Diagnosed Malformations

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[Coding Implications](#)

[Revision Log](#)

Description

This policy describes the medical necessity requirements for performing fetal surgery. This becomes an option when it is predicted that the fetus will not live long enough to survive delivery or after birth. Therefore, surgical intervention during pregnancy on the fetus is meant to correct problems that would be too advanced to correct after birth.

Policy/Criteria

- I. It is the policy of Pennsylvania Health and Wellness[®] (PHW) that in-utero fetal surgery (IUFS) is **medically necessary** when meeting the following criteria (A and B):
 - A. Treatment is for one of the following indications (1-8):
 1. In-utero removal of sacrococcygeal teratoma (SCT) associated with fetal hydrops related to high output heart failure secondary to arteriovenous shunting through the tumor; or
 2. Vesico-amniotic shunting as a treatment of lower urinary tract obstruction when there are no other lethal abnormalities or lethal chromosomal abnormalities; or
 3. Open or in-utero resection of malformed pulmonary tissue, or placement of a thoraco-amniotic shunt as a treatment of either a congenital pulmonary airway malformation (previously referred to as congenital cystic adenomatoid malformation or bronchopulmonary sequestration) when fetus has evidence of fetal hydrops (hydrops fetalis); or
 4. Amnioreduction alone as a treatment of twin-twin transfusion syndrome (TTTS); or
 5. Fetoscopic or open laser ablation of anastomotic vessels, with or without amnioreduction, as a treatment for TTTS, when both of the following are met:
 - a. Severe TTTS is confirmed clinically and by ultrasound between 16 and 26 weeks' gestation; and
 - b. The benefits of laser surgery outweigh the risks to a pre-viable fetus who is not a candidate for delivery and for whom the mortality rate is otherwise high; or
 6. Ablation of anastomotic vessels in acardiac twins with twin-reversed-arterial-perfusion (TRAP); or
 7. The ex utero intrapartum treatment (EXIT) procedure for congenital pulmonary airway malformation/congenital cystic adenomatoid malformation or bronchopulmonary sequestration; or
 8. Myelomeningocele repair when all of the following criteria are met:
 - a. Singleton pregnancy;
 - b. Upper boundary of myelomeningocele located between T1 and S1;
 - c. Evidence of hindbrain herniation;
 - d. Gestational age 19.0 to 25.9 weeks;
 - e. Normal fetal karyotype; and
 - f. None of the following:
 - i. Severe kyphosis;
 - ii. Risk of preterm birth(e.g., short cervix or previous preterm birth);

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- iii. Placental abruption;
 - iv. Maternal body mass index of ≥ 35 ;
 - v. No previous hysterotomy in the active uterine segment.
- B.** Member does not have placentomegaly or severe pre-eclampsia.
- II.** It is the policy of PHW that in utero fetal surgery is **investigational** for any of the following indications:
- A.** Open or endoscopic fetal surgery for congenital diaphragmatic hernia (CDH), including temporary tracheal occlusion;
 - B.** Surgery for heart block, pulmonary valve, or aortic obstruction;
 - C.** Tracheal atresia or stenosis;
 - D.** Cleft lip and palate;
 - E.** In-utero stem cell transplantation;
 - F.** In-utero gene therapy;
 - G.** Amnioexchange procedure for gastroschisis.

Background

Maternal–Fetal Surgery

Maternal–fetal surgery is a major procedure for the mother and her fetus, 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 babies 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.

Fetal surgery approaches can be divided into two categories:

- 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. 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 is intended to allow the fetus to remain in utero until it has matured enough to survive delivery and neonatal surgical procedures.

In some cases, surgery on the fetus is scheduled to coincide with delivery. The planned surgery is done on the fetus after Cesarean section, but before the cord is cut, so that the fetus continues to be sustained by the mother's placenta and doesn't have to breathe on his or her own. This method, known as an EXIT (ex utero intrapartum treatment) is usually employed when the fetus suffers from a congenital defect that blocks the airway, such as a cervical teratoma. EXIT gives surgeons time to perform multiple procedures to secure the baby's airway, so that by the time the cord is cut and the baby has to breathe; he or she has an unblocked airway.

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Twin reversed-arterial-perfusion

Twin reversed-arterial-perfusion (TRAP) sequence is a serious complication of monozygotic twin pregnancies, affecting 1% of monozygotic twins. Inadequate perfusion of the recipient twin is responsible for the development of a characteristic and invariably lethal set of anomalies, including the acardius fetal malformation (acardiac twins) and acephalus. Typically, the pump twin is structurally normal, but it is at risk for in utero cardiac failure and without treatment dies in 50 to 75% of cases, particularly if the recipient twin weighs more than half as much as the pump twin.

Guideline Recommendations

The American College of Obstetricians and Gynecologists and the Society for Maternal–Fetal Medicine make the following recommendations:

- Open maternal–fetal surgery for myelomeningocele repair has been demonstrated to improve a number of important pediatric outcomes at the expense of procedure-associated maternal and fetal risks.
- Women with pregnancies complicated by fetal myelomeningocele who meet established criteria for in utero repair should be counseled in nondirective fashion regarding all management options, including the possibility of open maternal–fetal surgery.
- Interested candidates for fetal myelomeningocele repair should be referred for further assessment and consultation to a fetal therapy center that offers this intervention and possesses the expertise, multi-disciplinary team, services, and facilities to provide detailed information regarding maternal–fetal surgery and the intensive care required for patients who choose to undergo open maternal–fetal surgery.⁹

Coding Implications

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CPT® Codes	Description
59076	Fetal shunt placement, including ultrasound guidance
59897	Unlisted fetal invasive procedure, including ultrasound guidance

HCPCS Codes	Description
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

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HCPCS Codes	Description
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

ICD-10-CM Diagnosis Codes that Support Coverage Criteria

ICD-10-CM Code	Description
D43.4	Neoplasm of uncertain behavior of spinal cord
O30.021- O30.029	Conjoined twin pregnancy [twin reversed arterial perfusion (TRAP)]
O35.0XX0- O35.9XX9	Maternal care for known or suspected fetal abnormality and damage
O36.20X0- O36.23X9	Maternal care for hydrops fetalis
O43.021 - O43.023	Fetus-to-fetus placental transfusion syndrome
Q05.0-Q05.9	Spina Bifida
Q33.0	Congenital cystic lung
Q33.2	Sequestration of lung
Q33.3	Agenesis of lung
Q33.6	Congenital hypoplasia and dysplasia of lung
Q34.0-Q34.9	Other Congenital malformations of respiratory system
Q62.31- Q62.39	Other obstructive defects of renal pelvis and ureter
Q64.2	Congenital posterior urethral valves
Q64.31- Q64.39	Other atresia and stenosis of urethra and bladder neck
Q89.4	Conjoined twins
Q89.8	Other specified congenital malformations

Reviews, Revisions, and Approvals	Date	Approval Date

References

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