

Clinical Policy: Fetal Surgery in Utero for Prenatally Diagnosed Malformations

Reference Number: PA.CP.MP.129

Effective Date: 01/18

Date of Last Revision: 8/31/2021

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 PA Health and Wellness[®] (PHW) that in-utero fetal surgery (IUFS) is **medically necessary** for any of the following:
 - A. Sacrococcygeal teratoma (SCT): SCT resection or a minimally invasive approach;
 - B. Lower urinary tract obstruction without multiple fetal abnormalities or chromosomal abnormalities: urinary decompression via vesico-amniotic shunting
 - C. Congenital pulmonary airway malformation (CPAM) and extralobar bronchopulmonary sequestration (BPS), with high risk tumors: resection of malformed pulmonary tissue, or placement of a thoraco-amniotic shunt;
 - D. Placement of a thoraco-amniotic shunt for pleural effusion with or without secondary fetal hydrops;
 - 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:
 1. Amnioreduction; or
 2. Fetoscopic laser ablation, with or without amnioreduction when member/enrolee is between 16 and 26 weeks gestation;
 - F. Twin-reversed-arterial-perfusion (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;
 4. Gestational age 19.0 to < 26 weeks;
 5. Normal fetal karyotype; and
 6. None of the following:
 - a. Severe kyphosis;
 - b. Risk of preterm birth(e.g., short cervix or previous preterm birth);
 - c. Placental abruption;
 - d. Maternal body mass index of ≥ 35 ;
 - e. History of previous hysterotomy in the active uterine segment.

II. It is the policy PHW that all repeat utero fetal surgery procedures require secondary review.

III. 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.

Sacroccygeal germ cell tumors

The prenatal diagnosis of SCT typically occurs during the second trimester during routine sonography. Prenatal diagnosis and close monitoring have improved outcomes for fetal SCT, but overall perinatal mortality remains high. The major goal is to identify fetuses at increased risk of fetal demise because of hydrops fetalis and intervene as appropriate. Hydrops fetalis is a condition of excess fluid accumulation in the fetus that results in significant fetal demise and neonatal mortality. 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). Contraindications to open fetal surgery for SCT include type III or IV Altman type tumors, severe placentomegaly, cervical shortening, and maternal medical issues.¹⁰

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.²⁰

Congenital pulmonary airway malformation

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 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 hemodynamic alterations from vena cava obstruction, cardiac displacement/compression and require prenatal intervention. The presence of hydrops is a sign for impending fetal demise (risk of death approaches 100 percent 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 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.¹¹

Twin-twin transfusion syndrome

TTTS occurs in approximately 10–15% of monochorionic–diamniotic 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 twin–twin transfusion syndrome 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 twin–twin transfusion syndrome diagnosis and management.¹⁹

Twin reversed-arterial-perfusion

Twin reversed-arterial-perfusion (TRAP) sequence is 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 (RFA).¹³

Guideline Recommendations

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

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 2020, 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.

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

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
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)]

CLINICAL POLICY
Fetal Surgery in Utero



ICD-10-CM Code	Description
O31.031- O31.039	Twin pregnancy, monochorionic/diamniotic
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 - O43.029	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	Revision Date	Approval Date
Reworded section I.A# 1-5 for clarification. Removed EXIT procedure from I.A. as the procedure is an “ex utero procedure.” Updated background information. Removed I.B, to defer to the discretion of the treating surgeon. References reviewed and updated.	09/18	10/18
Myelomeningocele repair: clarified that “no history of previous hysterotomy in the active uterine segment should be “history of previous hysterotomy in the active uterine segment. References reviewed and updated.	12/18	03/19
SCT: removed requirement for hydrops and included option for minimally invasive approach. CPAM/BPS: removed requirement for hydrops. Specialist review.	05/2020	8/16/2020
Annual review. References reviewed and updated. Coding reviewed. Changed “review date” in the header to “date of last revision” and “date” in the revision log header to “revision date.” Replaced all instance of “member” with “member/enrollee.” Replaced Pennsylvania with PA in Section I under Policy/Criteria; Added, “D. Placement of a thoraco-amniotic shunt for pleural effusion with or without secondary fetal hydrops,” to criteria set I. Added criteria set, “II. It is the policy of health plans affiliated with Centene Corporation that all repeat utero	8/13/2021	9/14/2021

Reviews, Revisions, and Approvals	Revision Date	Approval Date
fetal surgery procedures require secondary review.” Reviewed by specialist.		

References

1. 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.
2. Committee opinion no. 501: Maternal-fetal intervention and fetal care centers. *Obstet Gynecol*. 2011;118(2 Pt 1):405-410. doi:10.1097/AOG.0b013e31822c99af.
3. ACOG Committee Opinion No. 439: Informed consent. *Obstet Gynecol*. 2009;114(2 Pt 1):401-408. doi:10.1097/AOG.0b013e3181b48f7f.
4. Araujo Júnior E, Eggink AJ, van den Dobbelen J, et al. Procedure-related complications of open vs endoscopic fetal surgery for treatment of spina bifida in an era of intrauterine myelomeningocele repair: systematic review and meta-analysis. *Ultrasound Obstet Gynecol*. 2016;48(2):151-160. doi:10.1002/uog.15830.
5. Araujo E Júnior, Tonni G, Martins WP. Outcomes of infants followed-up at least 12 months after fetal open and endoscopic surgery for meningomyelocele: a systematic review and meta-analysis. *J Evid Based Med*. 2016;9(3):125-135. doi:10.1111/jebm.12207.
6. Fetal Surgery for Myelomeningocele. Hayes. www.hayesinc.com. Published July 23, 2018 (annual reviewe October 29, 2020). Accessed June 15, 2021.
7. Maternal-Fetal Surgical Procedures. Technical Brief No. 5. Agency for Healthcare Research and Quality. https://effectivehealthcare.ahrq.gov/sites/default/files/pdf/fetal-surgery_technical-brief.pdf. Published April 2011. Accessed June 16, 2021.
8. Committee Opinion No. 720: Maternal-Fetal Surgery for Myelomeningocele. *Obstet Gynecol*. 2017;130(3):e164-e167. doi:10.1097/AOG.0000000000002303.
9. Egler RA, Levine D, Wilkins-Haug L. Sacrococcygeal germ cell tumors. UpToDate. www.uptodate.com. Updated January 28, 2021. Accessed June 15, 2021.
10. Egloff A, Bulas DI. Congenital pulmonary airway malformation: Prenatal diagnosis and management. UpToDate. www.uptodate.com. Updated April 30, 2021. Accessed June 16, 2021.
11. Bulas DI, Egloff A. Bronchopulmonary sequestration: Prenatal diagnosis and management. UpToDate. www.uptodate.com. Updated April 9, 2021. Accessed June 30, 2020.
12. Miller R. Diagnosis and management of twin reversed arterial perfusion (TRAP) sequence. UpToDate. www.uptodate.com. Updated December 1, 2020. Accessed June 16, 2021.
13. Morris RK, Malin GL, Quinlan-Jones E, et al. Percutaneous vesicoamniotic shunting versus conservative management for fetal lower urinary tract obstruction (PLUTO): a randomised trial. *Lancet*. 2013;382(9903):1496-1506. doi:10.1016/S0140-6736(13)60992-7.
14. Belfort MA, Olutoye OO, Cass DL, et al. Feasibility and Outcomes of Fetoscopic Tracheal Occlusion for Severe Left Diaphragmatic Hernia. *Obstet Gynecol*. 2017;129(1):20-29. doi: 10.1097/AOG.0000000000001749.
15. Al-Maary J, Eastwood MP, Russo FM, et al. Fetal Tracheal Occlusion for Severe Pulmonary Hypoplasia in Isolated Congenital Diaphragmatic Hernia: A Systematic Review and Meta-analysis of Survival. *Ann Surg*. 2016 ;264(6):929-933. doi:10.1097/SLA.0000000000001675.
16. Baskin L. Overview of fetal hydronephrosis. UpToDate. www.uptodate.com. Updated June 17, 2019. Accessed June 16, 2021.

17. Papanna R. Twin-twin transfusion syndrome: Management and outcome. UpToDate. www.uptodate.com. Updated March 10, 2021. Accessed June 16, 2021.
18. Committee on Practice Bulletins—Obstetrics; Society for Maternal–Fetal Medicine. Practice Bulletin No. 169: Multifetal Gestations: Twin, Triplet, and Higher-Order Multifetal Pregnancies. *Obstet Gynecol*. 2016;128(4):e131-e146. doi:10.1097/AOG.0000000000001709.
19. Derderian SC, Hirose S. Fetal Surgery for Urinary Tract Obstruction. Medscape. <https://emedicine.medscape.com/article/2109522-overview>. Updated November 2019. Accessed June 16, 2021.
20. Baumgarten HD, Flake AW. Fetal Surgery. *Pediatr Clin North Am*. 2019;66(2):295-308. doi:10.1016/j.pcl.2018.12.001.
21. Fumino S, Tajiri T, Usui N, Tamura M, Sago H, Ono S, Nosaka S, Yoneda A, Souzaki R, Higashi M, Sakai K, Takahashi K, Sugiura T, Taguchi T. Japanese clinical practice guidelines for sacrococcygeal teratoma, 2017. *Pediatr Int*. 2019 Jul;61(7):672-678. doi: 10.1111/ped.13844. Epub 2019 Jun 18. PMID: 30903638.
22. Sananes N, Javadian P, Schwach Wernech Britto I, et al. Technical aspects and effectiveness of percutaneous fetal therapies for large sacrococcygeal teratomas: cohort study and literature review. *Ultrasound Obstet Gynecol*. 2016;47(6):712-719. doi:10.1002/uog.14935.
23. Wenstrom KD, Carr SR. Fetal Surgery: Principles, Indications, and Evidence. *Obstet Gynecol*. 2014;124(4):817-835. doi:10.1097/AOG.0000000000000476.