

Clinical Policy: Fetal Surgery in Utero for Prenatally Diagnosed Malformations

Reference Number: LA.CP.MP.129

Date of Last Revision: 02/24

[Coding Implications](#)

[Revision Log](#)

See [Important Reminder](#) at the end of this policy for important regulatory and legal information.

Description

This policy describes the medical necessity requirements for performing fetal surgery. This becomes an option when it is predicted that there will be severe disability, or if it is predicted that the fetus will not live long enough to survive delivery or after birth.¹ 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 Louisiana Healthcare Connections 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 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: 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 of the following:
 1. Amnioreduction;
 2. Fetoscopic laser ablation, with or without amnioreduction when pregnancy is between 16 and 26 weeks gestation;
 - 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. Normal fetal karyotype;
 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 ≥ 40 ;

- e. Previous hysterotomy in the active uterine segment.
- II.** It is the policy of Louisiana Healthcare Connections that all repeat utero fetal surgery procedures require secondary review.
- III.** It is the policy of Louisiana Healthcare Connections that current evidence does not support the use of utero fetal surgery 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.¹

Fetal surgery approaches can be divided into two categories²:

- 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.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 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

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

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 2022, 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 and may not support medical necessity. 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: Coverage is subject to each requested code’s inclusion on the corresponding LDH fee schedule. Non-covered codes are denoted (*) and are reviewed for Medical Necessity for members under 21 years of age on a per case basis.

| 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 |

| 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 |

| HCPCS Codes | Description |
|-------------|---|
| (*)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 |
|--|---------------|---------------|
| Converted corporate to local policy. | 10/2020 | |
| 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.” 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 Louisiana Healthcare connections that all repeat utero fetal surgery procedures require secondary review.” Reviewed by specialist. | 1/2022 | 3/26/22 |
| Annual review. Description updated with no impact on criteria. Background updated with no impact on criteria. References reviewed and updated. | 09/22 | 11/28/22 |
| Annual review. Criteria I.G.3. updated to include confirmation on fetal MRI. Added clarifying language to Criteria I.G.4. Background updated with no impact on criteria. Added CPT code 59072. ICD-10 codes removed. References reviewed and updated. Reviewed by external specialist. Added cpt codes 5900, 59074, and 59072. | 08/23 | 10/30/23 |
| Updated criteria I.G.6. to maternal body mass index of ≥ 40 and added supportive references. | 02/24 | 4/26/24 |

References

1. Committee Opinion No. 720: Maternal-Fetal Surgery for Myelomeningocele. *Obstet Gynecol.* 2017;130(3):e164 to e167. doi:10.1097/AOG.0000000000002303
2. Sampat K, Losty PD. Fetal surgery. *Br J Surg.* 2021;108(6):632 to 637. doi:10.1093/bjs/znaa153
3. Egler RA, Levine D, Wilkins-Haug L. Sacrococcygeal teratoma. UpToDate. www.uptodate.com. Updated January 30, 2023. Accessed June 21, 2023.
4. Lyttle BD. Fetal Surgery for Urinary Tract Obstruction. *Medscape*. <https://emedicine.medscape.com/article/2109522-overview>. Updated March 31, 2023. Accessed June 22, 2023.
5. Egloff A, Bulas DI. Congenital pulmonary airway malformation: Prenatal diagnosis and management. UpToDate. www.uptodate.com. Updated June 16, 2023. Accessed June 22, 2023.
6. Papanna R. Twin-twin transfusion syndrome: Management and outcome. UpToDate. www.uptodate.com. Updated December 06, 2022. Accessed May 31, 2023.
7. Miller R. Twin reversed arterial perfusion (TRAP) sequence. UpToDate. www.uptodate.com. Updated November 07, 2022. Accessed June 26, 2023.

8. 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 to 1004. doi:10.1056/NEJMoa1014379
9. Committee opinion no. 501: Maternal-fetal intervention and fetal care centers. *Obstet Gynecol*. 2011;118(2 Pt 1):405 to 410. doi:10.1097/AOG.0b013e31822c99af
10. ACOG Committee Opinion No. 439: Informed consent. *Obstet Gynecol*. 2009;114(2 Pt 1):401 to 408. doi:10.1097/AOG.0b013e3181b48f7f
11. Bulas DI, Egloff A. Bronchopulmonary sequestration: Prenatal diagnosis and management. UpToDate. www.uptodate.com. Updated June 16, 2022. Accessed June 16, 2023.
12. Araujo Júnior E, Eggink AJ, van den Dobbelen J, Martins WP, Oepkes D. 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 to 160. doi:10.1002/uog.15830
13. Morris RK, Malin GL, Quinlan-Jones E, et al. Percutaneous vesicoamniotic shunting versus conservative management for fetal lower urinary tract obstruction (PLUTO): a randomized trial. *Lancet*. 2013;382(9903):1496 to 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 to 29. doi:10.1097/AOG.0000000000001749
15. Al-Maary J, Eastwood MP, Russo FM, Deprest JA, Keijzer R. 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 to 933. doi:10.1097/SLA.0000000000001675
16. Baskin L. Fetal hydronephrosis: Etiology and prenatal management. UpToDate. www.uptodate.com. Updated December 02, 2021. Accessed May 31, 2023.
17. Araujo Júnior E, 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 to 135. doi:10.1111/jebm.12207
18. Health Technology Assessment. Fetal Surgery for Myelomeningocele. Hayes. www.hayesinc.com. Published July 23, 2018 (annual review July 26, 2022). Accessed May 30, 2023.
19. Agency for Healthcare Research and Quality. Maternal-Fetal Surgical Procedures. Technical Brief No. 5. https://effectivehealthcare.ahrq.gov/sites/default/files/pdf/fetal-surgery_technical-brief.pdf. Published April 2011. Accessed June 20, 2023.
20. Baumgarten HD, Flake AW. Fetal Surgery. *Pediatr Clin North Am*. 2019;66(2):295 to 308. doi:10.1016/j.pcl.2018.12.001
21. Fumino S, Tajiri T, Usui N, et al. Japanese clinical practice guidelines for sacrococcygeal teratoma, 2017. *Pediatr Int*. 2019;61(7):672 to 678. doi:10.1111/ped.13844
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 to 719. doi:10.1002/uog.14935
23. Wenstrom KD, Carr SR. Fetal surgery: principles, indications, and evidence. *Obstet Gynecol*. 2014;124(4):817 to 835. doi:10.1097/AOG.0000000000000476
Health Technology Assessment. Fetal surgery for congenital diaphragmatic hernia. Hayes. www.hayesinc.com. Published July 20, 2018 (annual review August 16, 2022). Accessed June 07, 2023.

24. Krispin, E., Mehollin-Ray, A.R. and Shamshirsaz, A.A Open spina bifida: In utero treatment and delivery considerations. UpToDate. www.uptodate.com. Updated April 24, 2023. Accessed January 9, 2024.
25. Yamashiro KJ, Farmer DL. Fetal myelomeningocele repair: a narrative review of the history, current controversies and future directions. *Transl Pediatr.* 2021;10(5):1497-1505. doi:10.21037/tp-20-87

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. LHCC 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 practice current at the time that this clinical policy was approved.

The purpose of this clinical policy is to provide a guide to medical necessity, which is a component of the guidelines used to assist in making coverage decisions and administering benefits. It does not constitute a contract or guarantee regarding payment or results. Coverage decisions and the administration of benefits are subject to all terms, conditions, exclusions and limitations of the coverage documents (e.g., evidence of coverage, certificate of coverage, policy, contract of insurance, etc.), as well as to state and federal requirements and applicable LHCC administrative policies and procedures.

This clinical policy is effective as of the date determined by LHCC. 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. LHCC 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.

This clinical policy does not constitute medical advice, medical treatment or medical care. It is not intended to dictate to providers how to practice medicine. Providers are expected to exercise professional medical judgment in providing the most appropriate care, and are solely responsible for the medical advice and treatment of members/enrollees. This clinical policy is not intended to recommend treatment for members/enrollees. Members/enrollees should consult with their treating physician in connection with diagnosis and treatment decisions.

Providers referred to in this clinical policy are independent contractors who exercise independent judgment and over whom LHCC has no control or right of control. Providers are not agents or employees of LHCC.

CLINICAL POLICY
Fetal Surgery in Utero



This clinical policy is the property of LHCC. Unauthorized copying, use, and distribution of this clinical policy or any information contained herein are strictly prohibited. Providers, members/enrollees and their representatives are bound to the terms and conditions expressed herein through the terms of their contracts. Where no such contract exists, providers, members/enrollees and their representatives agree to be bound by such terms and conditions by providing services to members/enrollees and/or submitting claims for payment for such services.

©2024 Louisiana Healthcare Connections. All rights reserved. All materials are exclusively owned by Louisiana Healthcare Connections and are protected by United States copyright law and international copyright law. No part of this publication may be reproduced, copied, modified, distributed, displayed, stored in a retrieval system, transmitted in any form or by any means, or otherwise published without the prior written permission of Louisiana Healthcare Connections. You may not alter or remove any trademark, copyright or other notice contained herein. Louisiana Healthcare Connections is a registered trademarks exclusively owned by Louisiana Healthcare Connections.