

Clinical Policy: Fetal Surgery in Utero for Prenatally Diagnosed Malformations

Reference Number: CP.MP.129

Last Review Date: 07/20

[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 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 health plans affiliated with Centene Corporation[®] that in-utero fetal surgery (IUS) 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.** 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 is between 16 and 26 weeks gestation;
 - E.** Twin-reversed-arterial-perfusion sequence (TRAP): ablation of anastomotic vessels of the acardiac twin (laser, radiofrequency ablation);
 - F.** 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. Previous hysterotomy in the active uterine segment.
- II.** It is the policy of health plans affiliated with Centene Corporation that in utero fetal surgery is **investigational** for any of the following indications:

CLINICAL POLICY

Fetal Surgery in Utero

- 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 allows 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.⁹

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-

CLINICAL POLICY

Fetal Surgery in Utero

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.¹⁹

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.¹⁰

Twin-twin transfusion syndrome

TTTS occurs in approximately 10–15% 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 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.¹⁸

Twin reversed-arterial-perfusion

Twin reversed-arterial-perfusion sequence (TRAP) 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 (RFA).¹²

Guideline Recommendations

The American College of Obstetricians and Gynecologists and the Society for Maternal–Fetal Medicine have developed recommendations for myelomeningocele repair. Open maternal–fetal surgery for myelomeningocele repair has been demonstrated to improve a number of important

CLINICAL POLICY

Fetal Surgery in Utero

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.⁸

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, when performed |

| 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)] |
| 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 |

| ICD-10-CM Code | Description |
|----------------|--|
| 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 |
|---|-------|---------------|
| Policy adopted from HN NMP344 Fetal Surgery in Utero for Prenatally Diagnosed Malformations. | 09/16 | 10/16 |
| Removed gestational age requirements from sections: IA.1, treatment of sacrococcygeal teratoma and IA.3 congenital pulmonary airway malformation or bronchopulmonary sequestration. Removed specific criteria from IA.2, Vesico-amniotic shunting as a treatment of lower urinary tract obstruction to allow procedure per the discretion of the treating surgeon. Updated background with more recent recommendations from ACOG committee opinion. Code updates. | 09/17 | 10/17 |
| 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. | 08/18 | 08/18 |
| 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. | 10/18 | |
| SCT: removed requirement for hydrops and included option for minimally invasive approach. CPAM/BPS: removed requirement for hydrops. Specialist review. | 08/19 | 08/19 |
| References reviewed and updated | 07/20 | 07/20 |

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; 364:993–1004.
2. American Congress of Obstetricians and Gynecologists (ACOG). Committee Opinion. Maternal-Fetal Intervention and Fetal Care Centers. Number 501. August 2011. Reaffirmed 2017.
3. American Congress of Obstetricians and Gynecologists (ACOG). Informed consent. ACOG Committee Opinion No. 439. American College of Obstetricians and Gynecologists. *Obstet Gynecol* 2009; 114:401-8. Reaffirmed 2015.

CLINICAL POLICY

Fetal Surgery in Utero

4. Araujo Júnior E, Eggink AJ, van den Dobbelsteen 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 Aug;48(2):151-60. 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 Jun 15. doi: 10.1111/jebm.12207. [Epub ahead of print].
6. Hayes Medical Technology Directory. In *Utero Fetal Surgery for Myelomeningocele*. Update August 22, 2019.
7. Walsh WF, Chescheir NC, Gillam-Krakauer M, et al. *Maternal-Fetal Surgical Procedures. Technical Brief No. 5.* Agency for Healthcare Research and Quality. April 2011. AHRQ Publication No. 10(11)-EHC059-EF.
8. The American College of Obstetricians and Gynecologists. *ACOG Committee Opinion. Maternal–Fetal Surgery for Myelomeningocele. Number 720.* Sept 2017.
9. Egler RA, Levine D, Wilkins-Haug L. Sacrococcygeal germ cell tumors. In: UpToDate, Simpson LL, Pappo VA. (Ed). Accessed June 30, 2020.
10. Egloff A, Bulas DI. Prenatal diagnosis and management of congenital pulmonary airway malformation. In; UpToDate. Levine D, Wilkins-Haug L (Ed) Accessed June 30, 2020.
11. Bulas DI, Egloff A. Prenatal diagnosis and management of bronchopulmonary sequestration. In: UpToDate. Levine D, Wilkins-Haug L (Ed) Accessed June 30, 2020.
12. Miller R. Diagnosis and management of twin reversed arterial perfusion (TRAP) sequence. In: UpToDate. Simpson LL, Levine D (Ed). Accessed June 30, 2020.
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; 382:1496.
14. Belfort MA, Olutoye OO, Cass DL, et al. Feasibility and Outcomes of Fetoscopic Tracheal Occlusion for Severe Left Diaphragmatic Hernia. *Obstet Gynecol.* 2017 Jan;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 Dec;264(6):929-933.
16. Baskin L. Overview of fetal hydronephrosis. In: UpToDate, Mattoo TK, Wikins-Haug L, Wicox D. (Ed), Accessed June 30, 2020.
17. Papanna R. Twin-twin transfusion syndrome: Management and outcome. In: UpToDate, Levine D, Simpson LL (Ed). Accessed June 30, 2020. .
18. American Congress of Obstetricians and Gynecologists (ACOG). *Practice Bulletin. Multifetal Gestations: Twin, Triplet, and Higher-Order Multifetal Pregnancies. Number 169.* October 2016. Reaffirmed 2016.
19. Derderian SC, Hirose S. *Fetal Surgery for Urinary Tract Obstruction.* Medscape, Last updated Dec 2017. Accessed June 30, 2020.
20. Baumgarten HD, Flake AW. *Fetal Surgery.* *Pediatric Clinics of North America.* 2019; 66(2), Pages 295-308.
21. Fumino S, Tajiri T, Usui N, et al. Japanese clinical practice guidelines for sacrococcygeal teratoma,2017. *Pediatrics International* (2019) 61, 672678.

CLINICAL POLICY

Fetal Surgery in Utero

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 Jun;47(6):712-9.
23. Wenstrom KD, Carr SR. Fetal Surgery: Principles, Indications, and Evidence. *Obstetrics & Gynecology*: October 2014 - Volume 124 - Issue 4 - p 817835.

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 practice current at the time that this clinical policy was approved. "Health Plan" means a health plan that has adopted this clinical policy and that is operated or administered, in whole or in part, by Centene Management Company, LLC, or any of such health plan's affiliates, as applicable.

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 Health Plan-level administrative policies and procedures.

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.

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. This clinical policy is not intended to recommend treatment for members. Members 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 the Health Plan has no control or right of control. Providers are not agents or employees of the Health Plan.

CLINICAL POLICY

Fetal Surgery in Utero

This clinical policy is the property of the Health Plan. Unauthorized copying, use, and distribution of this clinical policy or any information contained herein are strictly prohibited. Providers, members 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 and their representatives agree to be bound by such terms and conditions by providing services to members and/or submitting claims for payment for such services.

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

©2016 Centene Corporation. All rights reserved. All materials are exclusively owned by Centene Corporation 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 Centene Corporation. You may not alter or remove any trademark, copyright or other notice contained herein. Centene[®] and Centene Corporation[®] are registered trademarks exclusively owned by Centene Corporation.