

## Clinical Policy: Fetal Surgery in Utero for Prenatally Diagnosed Malformations

Reference Number: OH.CP.MP.129

Date of Last Revision: 07/23

[Coding Implications](#)

[Revision Log](#)

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

### Policy Statement

In compliance with Ohio Medicaid, Buckeye Health Plan must ensure coverage of medically necessary procedures. The plan covers all the services in the amount, duration, and scope that is no less than that covered by FFS Ohio Medicaid and in accordance with 42 CFR 438.210, with limitations, exclusions, and clarifications provided in the Ohio Medicaid Managed Care Provider Agreement and the Ohio Administrative Code.

### 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.<sup>8</sup> Therefore, surgical intervention during pregnancy on the fetus is meant to correct problems that would be too advanced to correct after birth.

### Procedure

- Buckeye Health Plan will not impose hard limits or restrictions on coverage of medically necessary services.
- Prior to making a determination regarding coverage of services and procedures, Buckeye Health Plan will conduct a medical necessity review for all requests to include non-covered services and any request for services over an established benefit amount(s).
- Coverage determinations are not limited to only certain diagnoses, but rather all requests will be reviewed for inclusion of coverage based on individual medical conditions and needs of mother and fetus.
- All codes covered on the Ohio Medicaid FFS schedule will be covered by Buckeye Health Plan.
- Buckeye Health Plan will include as medically necessary procedures interventions for amniotic band syndrome.
- Buckeye Health Plan will ensure all members under age 21 have access to all services that are available in accordance with federal EPSDT requirements found at 42 U.S.C. 1396d(r). This would include medically necessary services covered by Ohio Medicaid as well as any medically necessary screening, diagnostic and treatment services available to Ohio Medicaid consumers.

### Policy/Criteria

- I. It is the policy of Buckeye Health Plan and plans affiliated with Centene Corporation® 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:
    - 1. Amnioreduction; or
    - 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;
    - 4. Gestational age 19.0 to < 26 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  $\geq 35$ ;
      - e. Previous hysterotomy in the active uterine segment.
- II.** It is the policy of Buckeye Health Plan, a health plan affiliated with Centene Corporation, that all repeat utero fetal surgery procedures require secondary review.
- III.** It is the policy of Buckeye Health Plan and plans affiliated with Centene Corporation that current evidence does not support the use of utero fetal surgery for 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.<sup>8</sup>

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 sacroccygeal teratoma (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. 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.<sup>9</sup>

### *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.<sup>19</sup>

### *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.

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.<sup>10</sup>

#### *Twin-twin transfusion syndrome (TTTS)*

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 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.<sup>18</sup>

#### *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.<sup>12</sup>

#### *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.<sup>8</sup>

**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 through O30.029   | Conjoined twin pregnancy [twin reversed arterial perfusion (TRAP)] |
| O31.031 through O31.039   | Twin pregnancy, monochorionic/diamniotic                           |
| O35.0XX0 through O35.9XX9 | Maternal care for known or suspected fetal abnormality and damage  |
| O36.20X0 through O36.23X9 | Maternal care for hydrops fetalis                                  |
| O43.021 through O43.029   | Fetus-to-fetus placental transfusion syndrome                      |

| ICD-10-CM Code        | Description  |
|-----------------------|--|
| Q05.0 through 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 through Q34.9   | Other congenital malformations of respiratory system   |
| Q62.31 through Q62.39 | Other obstructive defects of renal pelvis and ureter   |
| Q64.2                 | Congenital posterior urethral valves                   |
| Q64.31 through 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 |
|--|---------------|---------------|
| Original Centene Policy CP.MP.129 created and approved.  | 09/16         | 09/16         |
| Centene Policy CP.MP.129 updated with OH Addendum and approved by Ohio Department of Medicaid.   | 02/23         | 02/23         |
| Policy moved to Ohio Specific template and OH Addendum language integrated into policy template as Policy Statement and Procedure. Reviewed for content and no changes required. References reviewed and updated. No material changes in review criteria made. | 07/23         | 07/23         |

**References**

1. Ohio Administrative Code 5160-1-14 EPSDT. <https://codes.ohio.gov/ohio-administrative-code/rule-5160-1-14>
2. Ohio Administrative Code 5160-26-03 Managed healthcare programs- covered services. <https://codes.ohio.gov/ohio-administrative-code/rule-5160-26-03>
3. Ohio Administrative Code 5160-26-03.1 Managed healthcare programs- primary care and utilization management. <https://codes.ohio.gov/ohio-administrative-code/rule-5160-26-03.1>
4. Ohio Department of Medicaid Fee Schedule: <https://medicaid.ohio.gov/resources-for-providers/billing/fee-schedule-and-rates/fee-schedule-and-rates>
5. 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
6. 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
7. ACOG Committee Opinion No. 439: Informed consent. *Obstet Gynecol.* 2009;114(2 Pt 1):401-408. doi:10.1097/AOG.0b013e3181b48f7f
8. 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):151-160. doi:10.1002/uog.15830

9. 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
10. Health Technology Assessment. Fetal Surgery for Myelomeningocele. Hayes. [www.hayesinc.com](http://www.hayesinc.com). Published July 23, 2018 (annual review September 08, 2021). Accessed June 24, 2022.
11. 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](https://effectivehealthcare.ahrq.gov/sites/default/files/pdf/fetal-surgery_technical-brief.pdf). Published April 2011. Accessed June 27, 2022.
12. Committee Opinion No. 720: Maternal-Fetal Surgery for Myelomeningocele. *Obstet Gynecol*. 2017;130(3):e164-e167. doi:10.1097/AOG.0000000000002303
13. Egler RA, Levine D, Wilkins-Haug L. Sacrococcygeal germ cell tumors. UpToDate. [www.uptodate.com](http://www.uptodate.com). Updated January 28, 2021. Accessed June 27, 2022.
14. Egloff A, Bulas DI. Congenital pulmonary airway malformation: Prenatal diagnosis and management. UpToDate. [www.uptodate.com](http://www.uptodate.com). Updated April 29, 2022. Accessed June 24, 2022.
15. Bulas DI, Egloff A. Bronchopulmonary sequestration: Prenatal diagnosis and management. UpToDate. [www.uptodate.com](http://www.uptodate.com). Updated June 16, 2022. Accessed June 23, 2022.
16. Miller R. Diagnosis and management of twin reversed arterial perfusion (TRAP) sequence. UpToDate. [www.uptodate.com](http://www.uptodate.com). Updated May 17, 2022. Accessed June 24, 2022.
17. 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
18. 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
19. 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):929-933. doi:10.1097/SLA.0000000000001675
20. Baskin L. Fetal hydronephrosis: Etiology and prenatal management. UpToDate. [www.uptodate.com](http://www.uptodate.com). Updated December 02, 2021. Accessed June 22, 2022.
21. Papanna R. Twin-twin transfusion syndrome: Management and outcome. UpToDate. [www.uptodate.com](http://www.uptodate.com). Updated May 11, 2022. Accessed June 24, 2022.
22. 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
23. Derderian SC. Fetal Surgery for Urinary Tract Obstruction. *Medscape*. <https://emedicine.medscape.com/article/2109522-overview>. Updated November 05, 2019. Accessed June 27, 2022.
24. Baumgarten HD, Flake AW. Fetal Surgery. *Pediatr Clin North Am*. 2019;66(2):295-308. doi:10.1016/j.pcl.2018.12.001
25. Fumino S, Tajiri T, Usui N, et al. Japanese clinical practice guidelines for sacrococcygeal teratoma, 2017. *Pediatr Int*. 2019;61(7):672-678. doi:10.1111/ped.13844

26. 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
27. Wenstrom KD, Carr SR. Fetal surgery: principles, indications, and evidence. *Obstet Gynecol.* 2014;124(4):817-835. doi:10.1097/AOG.0000000000000476
28. Health Technology Assessment. Fetal surgery for congenital diaphragmatic hernia. Hayes. [www.hayesinc.com](http://www.hayesinc.com). Published July 20, 2018 (annual review September 02, 2021). Accessed June 22, 2022.

### **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. Buckeye 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 Buckeye 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/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 Buckeye Health Plan has no control or right of control. Providers are not agents or employees of Buckeye Health Plan.

This clinical policy is the property of Buckeye Health Plan. 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.

**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.

©2018 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.