Clinical Policy: Fetal Surgery in Utero for Prenatally Diagnosed Malformations

Reference Number: CP.MP.129
Effective Date: 10/16
Last Review Date: 09/16

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 (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 at < 30 weeks gestation; or
2. Vesico-amniotic shunting as a treatment of lower urinary tract obstruction when all of the following are met:
   a. Bladder distension; and
   b. Bilateral hydronephrosis due to urinary tract obstruction; and
   c. Evidence of progressive oligohydramnios; and
   d. Adequate renal function; and
   e. 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 cystic adenomatoid malformation or extralobar pulmonary sequestration when both of the following are met:
   a. Fetus is < 32 weeks' gestation; and
   b. 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 15 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 cystic adenomatoid malformation or extralobar pulmonary 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; and
c. Evidence of hindbrain herniation; and
d. Gestational age 19.0 to 25.9 weeks; and
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);
   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 health plans affiliated with Centene Corporation 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.

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 (ACOG), noted that fetal surgery for myelomeningocele improved a number of important outcomes, but also was associated with maternal and fetal risks. They recommend that women who meet the criteria, noted within this policy, should be made aware of these findings and counseled regarding the option of maternal–fetal surgery, including both the risks and benefits to the woman and the baby, as well as the implications for future pregnancies. [3]

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 2015, 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
59076	Fetal shunt placement, including ultrasound guidance
59897	Unlisted fetal invasive procedure, including ultrasound guidance

HCPCS Codes	Description
S2400	Repair, congenital diaphragmatic hernia in the fetus using temporary tracheal occlusion, procedure performed in utero
S2401	Repair, urinary tract obstruction in the fetus, procedure performed in utero

HCPCS Codes	Description
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)]
O35.0XX1-O35.9XX9	Maternal care for known or suspected fetal abnormality and damage
O36.20XX1-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-Q33.9	Congenital malformations of lung
Q34.0-Q34.9	Other Congenital malformations of respiratory system
Q60.0-Q64.9	Congenital malformations of the urinary system
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

References

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4. American Congress of Obstetricians and Gynecologists (ACOG). Committee Opinion. Number 550, January 2013. Maternal–Fetal Surgery for Myelomeningocele. Reaffirmed 2015.
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6. 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].
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8. 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. Available at: http://www.effectivehealthcare.ahrq.gov/ehc/products/90/649/TechBrief5_Maternal-Fetal-Surgery.pdf

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.

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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.