Clinical Policy: Fetal Surgery in Utero for Prenatally Diagnosed Malformations

Reference Number: LA.CP.MP.129 Coding Implications
Date of Last Revision: 07/24 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. Fetal surgery becomes an option when it is predicted that there will be severe disability or mortality during delivery or after birth.[1]

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) with treatment including:
   1. Correction via a minimally invasive approach;
   2. SCT resection when meeting all of the following:
      a. Fetuses with high-risk SCT and hydrops developing at a gestational age earlier than appropriate for delivery and neonatal care (e.g. 28-32 weeks gestation);
      b. Does not have the following contraindications:
         i. Type III or IV Altman-type tumors;
         ii. Severe placentomegaly;
         iii. Maternal cervical shortening;

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. None of the following:
   c. Severe kyphosis (≥30 degrees);
   d. Risk of preterm birth (e.g., short cervix or previous preterm birth);
   e. Placental abruption;
   f. Maternal body mass index of ≥ 40;
   g. Previous hysterotomy in the active uterine segment.

H. Fetal endoscopic tracheal occlusion (FETO) for congenital diaphragmatic hernia (CDH) when all of the following criteria are met:
   1. Severe left-sided CDH;
   2. Severe pulmonary hypoplasia defined as a quotient of the observed-to-expected lung-to-head ratios of less than 25%;
   3. Gestational age ≤ 30 weeks.

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. Surgery for heart block, pulmonary valve, or aortic obstruction;
B. Tracheal atresia or stenosis;
C. Cleft lip and palate;
D. In-utero stem cell transplantation;
E. In-utero gene therapy;
F. 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 categories[2]:

- 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 2023, 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

CPT® Codes	Description
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
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	Effective 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	
Annual review. Description updated with no impact to criteria. Under I.A. added “with treatment including”. Added criteria to I.A.1.-I.A.2. to include: Correction via a minimally invasive approach; SCT resection when meeting all of the following: Fetuses with high-risk SCT and hydrops developing at a gestational age earlier than appropriate for delivery and neonatal care (eg. 28-32 weeks gestation); Does not have the following	07/24	9/24/24	10/25/24

Reviews, Revisions, and Approvals	Revision Date	Approval Date	Effective Date
contraindications: Type III or IV Altman-type tumors; Severe placentomegaly; Maternal cervical shortening. Removed indication I.F.5. Normal fetal karyotype. Quantified criteria I.F.5.c. to include (≥30 degrees). Added criteria I.G. Fetal endoscopic tracheal occlusion (FETO) for congenital diaphragmatic hernia (CDH) when all of the following criteria are met: Severe left-sided CDH; Severe pulmonary hypoplasia defined as a quotient of the observed-to-expected lung-to-head ratios of less than 25%; Gestational age ≤ 30 weeks. Removed III.A. Open or endoscopic fetal surgery for congenital diaphragmatic hernia (CDH), including temporary tracheal occlusion. References reviewed and updated. Reviewed by external specialist.			

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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.

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