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Subject: In Utero (Intrauterine) Fetal Surgery

THIS MEDICAL COVERAGE GUIDELINE IS NOT AN AUTHORIZATION, CERTIFICATION, EXPLANATION OF BENEFITS, OR A GUARANTEE OF PAYMENT, NOR DOES IT SUBSTITUTE FOR OR CONSTITUTE MEDICAL ADVICE. ALL MEDICAL DECISIONS ARE SOLELY THE RESPONSIBILITY OF THE PATIENT AND PHYSICIAN. BENEFITS ARE DETERMINED BY THE GROUP CONTRACT, MEMBER BENEFIT BOOKLET, AND/OR INDIVIDUAL SUBSCRIBER CERTIFICATE IN EFFECT AT THE TIME SERVICES WERE RENDERED. THIS MEDICAL COVERAGE GUIDELINE APPLIES TO ALL LINES OF BUSINESS UNLESS OTHERWISE NOTED IN THE PROGRAM EXCEPTIONS SECTION.

DESCRIPTION:

In utero (intrauterine) fetal surgery is performed to correct a malformation of a fetus that interferes with organ development and that can have potentially fatal consequences. Intrauterine fetal surgery involves opening of the gravid uterus using a minimally invasive endoscopic technique (single or multiple fetoscopic port incisions) or a traditional cesarean surgical incision to surgically correct a fetal abnormality.

Advances in methods of prenatal diagnosis, particularly prenatal ultrasound has led to an understanding of the physiologic outcomes of certain congenital anomalies. Surgical correction of an anatomic malformation in utero remains a highly invasive procedure. At present, there is broad consensus that the potential indications for subjecting mother and fetus to such a high level of risk remain limited to conditions that irreversibly interfere with the development of fetal organs. Fetal surgery requires a multidisciplinary approach (e.g., pediatric surgeons, perinatal obstetricians, sonographers, echocardiographers, neonatologists, intensive care specialists, geneticists, ethicists, neonatal, and obstetric nurses) and must be performed in an appropriate environment for optimum patient care.

This guideline addresses fetal surgery performed for the following:

- Fetal urinary tract obstruction
- Congenital diaphragmatic hernia
- Congenital cystic adenomatoid malformation and extralobar pulmonary sequestration
- Sacrococcygeal teratoma
- Myelomeningocele (spina bifida)
- Other neurological conditions.

Fetal Urinary Tract Obstruction

Few cases of prenatally diagnosed urinary tract obstruction require prenatal intervention. Bilateral obstruction is often associated with serious disease such as pulmonary hypoplasia secondary to

oligohydramnios. Fetuses with bilateral obstruction, oligohydramnios, adequate renal function reserve, and no other lethal or chromosomal abnormalities may be candidates for fetal surgery. The most common surgical approach is vesicoamniotic shunting by means of shunt or stent placement. The shunting procedure bypasses the obstructed urinary tract, permitting fetal urine to flow into the amniotic space.

Congenital Diaphragmatic Hernia (CDH)

CHD is a defect that permits abdominal viscera to enter the chest, frequently resulting in hypoplasia of the lungs. CDH can vary widely in severity, depending on the size of the hernia and the timing of herniation. For example, late herniation after 25 weeks of gestation may be managed postnatally. In contrast, liver herniation into the chest prior to 25 weeks of gestation is associated with a poor prognosis; these fetuses have been considered candidates for fetal surgery. Temporary tracheal occlusion prevents the normal efflux of fetal ling fluid, which in turn enhances positive pressure in the growing lungs, promoting lung growth and ultimately reducing abdominal viscera back into the abdominal cavity.

Congenital Cystic Adenomatoid Malformation (CCAM) or Extralobar Pulmonary Sequestration (EPS)

CCAM and EPS are the two most common congenital cystic lung lesions. When associated with fetal hydrops before 32 weeks gestation, the survival is poor. These patients may be candidates for prenatal surgical resection of a large mass or placement of a thoraco-aminotic shunt for a large unilocular cystic lesion.

Sacrococcygeal Teratoma (SCT)

Sacrococcygeal teratoma is both a neoplasm with the power of autonomous growth and a malformation made up of multiple tissues foreign to the region of origin lacking organ specificity. It is the most common tumor of the newborn. Postnatal SCT carries a good prognosis with morbidity and mortality determined largely by extent of local disease and malignant potential. In utero, fetal mortality has approached 100% when SCT is associated with fetal hydrops, which is related to high output heart failure secondary to arteriovenous shunting through the tumor. Intrauterine surgery may result in prenatal resolution of hydrops, healthy long-term survival, and normal development.

Myelomeningocele (spina bifida

Myelomeningocele is a neural tube defect in which the spinal cord forms abnormally and is left open, exposing the meninges and neural tube to the intrauterine environment. This exposure may cause secondary trauma to the spinal cord, and depending on the location of the myelomeningocele, results in varying degrees of neurologic impairment to the legs and bowel and bladder function, brain malformation (e.g., hindbrain herniation), and disorders of cerebrospinal fluid circulation (e.g., hydrocephalus requiring placement of a ventriculoperitoneal shunt). Traditional treatment consists of surgical repair after term delivery, primarily to prevent infection and further neurologic dysfunction. Fetal surgical repair to cover the exposed spinal canal has been proposed as a means of preventing exposure to the intrauterine environment with the hope of improving neurologic function and decreasing the incidence of other problems related to the condition.

POSITION STATEMENT:

NOTE: Refer coverage requests for in utero fetal surgery to the Medical Director for approval.

In utero fetal surgery **meets the definition of medical necessity** for the following conditions:

Vesico-amniotic shunting as a treatment of urinary tract obstruction **meets the definition of medical necessity** in fetuses under the following conditions:

- Evidence of hydronephrosis due to bilateral urinary tract obstruction; **AND**
- Progressive oligohydramnios; **AND**
- Adequate renal function; **AND**
- No other lethal abnormalities or chromosomal defects.

Open in utero resection of malformed pulmonary tissue or placement of a thoracoamniotic shunt **meets the definition of medical necessity** under the following conditions:

- Congenital cystic adenomatoid malformation or bronchopulmonary sequestration is identified; **AND**
- The fetus is at 32 weeks' gestation or less; **AND**
- There is evidence of fetal hydrops, placentomegaly, and/or the beginnings of severe pre-eclampsia (i.e., the maternal mirror syndrome) in the mother.

In utero removal of sacrococcygeal teratoma **meets the definition of medical necessity** under the following conditions:

- The fetus is at 32 weeks' gestation or less; **AND**
- There is evidence of fetal hydrops, placentomegaly, and/or the beginnings of severe pre-eclampsia (i.e., maternal mirror syndrome) in the mother.

In utero repair of myelomeningocele may be considered **meets the definition of medical necessity** under the following conditions:

- The fetus is at less than 26 weeks of gestation; **AND**
- Myelomeningocele is present with an upper boundary located between T1 and S1 with evidence of hindbrain herniation.

In utero repair of myelomeningocele is considered **experimental or investigational** in the following:

- Fetal anomaly unrelated to myelomeningocele
- Severe kyphosis
- Risk of preterm birth (e.g., short cervix or previous preterm birth)
- Maternal body mass index of 35 kg/m^2 or more.

All other applications of fetal surgery are considered **experimental or investigational**, including but not limited to following:

- Temporary tracheal occlusion as a treatment of congenital diaphragmatic hernia
- Treatment of congenital heart defects.

BILLING/CODING INFORMATION:

HCPCS Coding:

S2400	Repair, congenital diaphragmatic hernia in the fetus using temporary tracheal occlusion, procedure performed in utero **(investigational)**
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

LOINC Codes:

The following information may be required documentation to support medical necessity: physician history and physical, physician progress notes, plan of treatment and reason for in utero fetal surgery.

Documentation Table	LOINC Codes	LOINC Time Frame Modifier Code	LOINC Time Frame Modifier Codes Narrative
Physician history and physical	28626-0	18805-2	Include all data of the selected type that represents observations made six months or fewer before starting date of service for the claim
Attending physician progress note	18741-9	18805-2	Include all data of the selected type that represents observations made six months or fewer before starting date of service for the claim
Plan of treatment	18776-5	18805-2	Include all data of the selected type that represents observations made six months or fewer before starting date of service for the claim

REIMBURSEMENT INFORMATION:

Refer to section entitled **POSITION STATEMENT**.

PROGRAM EXCEPTIONS:

Federal Employee Program (FEP): Follow FEP guidelines.

State Account Organization (SAO): Follow SAO guidelines.

Medicare Advantage products:

No National Coverage Determination (NCD) and/or Local Coverage Determination (LCD) were found at the time of the last guideline reviewed date.

DEFINITIONS:

Abdominal viscera: pertaining to the abdominal cavity contents.

Fetal hydrops: the clinical condition in infants or cardiac decompensation with hepatosplenomegaly, of the fetus before birth.

Gravid uterus: a pregnant uterus.

Oligohydramnios: an abnormal small amount of amniotic fluid; the presence of less than 300ml of amniotic fluid at term.

Pulmonary hypoplasia: underdevelopment of the lungs.

RELATED GUIDELINES:

None applicable.

OTHER:

Other names used to report fetal surgery for prenatal diagnosed malformations:

Congenital Cystic Adenomatoid Malformation, Fetal Surgery (CCAM)
Congenital Diaphragmatic Hernia, Fetal Surgery (CDH)
Extralobar Pulmonary Sequestration, Fetal Surgery (EPS)
Maternal-Fetal Surgery
Temporary Tracheal Occlusion
Thoraco-Amino Shunt
Urinary Tract Obstruction, Fetal Surgery (UTO)
Vesico-Aminotic Shunting

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COMMITTEE APPROVAL:

This Medical Coverage Guideline (MCG) was approved by the Florida Blue Medical Policy & Coverage Committee on 07/22/21.

GUIDELINE UPDATE INFORMATION:

08/15/02	New Medical Coverage Guideline.
09/15/03	Annual review. Added myelomeningocele to description section. Added the wording “either open” under when services are covered.
06/15/04	Scheduled review. Added investigational statement for tracheal occlusion as a treatment for congenital diaphragmatic hernia. Added cross-reference for Treatment of Twin-Twin Transfusion Syndrome (TTS) with Amino-reduction and/or Fetoscopic Laser Therapy. Updated references.
06/15/05	Scheduled review. No change in coverage statements. Added information to the description section regarding: vesico-amniotic shunting, fetal surgery for congenital cystic adenomatoid malformation, extralobar pulmonary sequestration and sacrococcygeal teratoma. Updated references.
05/15/06	Scheduled review. No change in coverage statements. Revised description section, Revised when services are covered. Deleted statement; refer coverage requests for in utero fetal surgery to Medical Director for approval. Revised when services are not covered. Updated references.
05/15/07	Scheduled review; no change in coverage statements; reformatted guideline; references updated.
04/15/08	Scheduled review. Added treatment of congenital heart defects as experimental or investigational. Deleted ICD-9 diagnoses. Added maternal-fetal surgery to other section. Updated references.
05/15/09	Annual review. No change in position statements. Updated references.
05/15/11	Revised position statements; added conditions for vesico-amniotic shunting, malformed pulmonary tissue and placement of a thoracoamniotic shunt and sacrococcygeal teratoma. Updated references.
10/01/11	Revision; formatting changes.
05/11/14	Revision: Program Exceptions section updated.
10/15/17	Review; added position statement for myelomeningocele. Updated references.

07/15/19	Review; no change to position statement, Updated references.
08/15/21	Review; no change to position statement.