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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)

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.

Summary and Analysis of Evidence:

Lower Urinary Tract Obstruction

In a retrospective review Jeong et al 2018 reviewed 32 fetuses with LUTO who underwent vesicoamniotic shunting using a double-basket catheter between 1998 and 2013 to evaluate the perinatal survival and renal function of fetuses with lower urinary tract obstruction (LUTO). Among the 32 fetuses examined, 5 died because of termination of pregnancy, and 2 died in utero. The median gestational age at diagnosis was 15.5 (range, 10.0-27.3) weeks, and that at initial shunting was 17.1 (range, 12.3-32.2) weeks. Shunt dislocation or occlusion occurred in 18 of 42 procedures (42.8%). The median gestational age at delivery for the 25 live births was 35.5 (range, 28-40) weeks. Postnatal diagnosis revealed posterior urethral valves in 15 fetuses, a cloacal anomaly in 7, and urethral stenosis in 3. Three neonatal deaths occurred, resulting in an overall perinatal survival rate of 68.8% (22 of 32). The rates of normal renal function were 40.6% (13 of 32) at 28 days and 40% (10 of 25) at 2 years after birth. The absence of oligohydramnios after shunting was the only prognostic factor for normal renal function at 2 years ($P < .05$). The authors concluded that vesicoamniotic shunting may be helpful for fetuses with LUTO.

Kohl et al 2022 in a retrospective cohort study, the authprs compared the outcome of human fetuses with isolated severe lower urinary tract obstructions (LUTO) that were first treated before the completion of 16 weeks of gestation to fetuses first treated later in gestation. Vesicoamniotic shunt insertion (VAS) was performed in 63 subsequent fetuses with LUTO between 12 + 5 and 30 + 3 weeks. The fetuses were analyzed in three groups: Group-I-fetuses underwent their first intervention until the completion of 16 weeks, Group-II-fetuses were first treated between 16 + 1 and 24 + 0 weeks and Group-III-fetuses beyond 24 + 1 weeks. Renal and pulmonary outcome parameters and complicating factors were assessed. Vesicoamniotic shunt insertion (VAS) was performed in 63 subsequent fetuses with LUTO between 12 + 5 and 30 + 3 weeks. The fetuses were analyzed in three groups: Group-I-fetuses underwent their first intervention until the completion of 16 weeks, Group-II-fetuses were first treated between 16 + 1 and 24 + 0 weeks and Group-III-fetuses beyond 24 + 1 weeks. Renal and pulmonary outcome parameters and complicating factors were assessed. All mothers tolerated the procedures well. Overall fetal survival was 47 of 63 (75%). The mean age at delivery of survivors was 35 weeks. 68% of Group-I-fetuses, 77% of group-II-fetuses, and 100% of group-III-fetuses survived beyond postnatal hospital discharge. Amongst the survivors the chance for normal renal function was higher for group I with 79% (15/19) compared to first fetal intervention after the completion of 16 weeks with 32% (9/28, $p = 0.003$, OR = 7.9 [2.0, 30.8] 95% CI). Clinically relevant pulmonary hypoplasia was observed in 11% of Group-I-, 27% of Group-II-, and 20% of Group-III-fetuses. The authors concluded that early intervention in fetal LUTO before the completion of 16 weeks may achieve a higher rate of normal renal and pulmonary function in survivors than treatment beyond that point in time. This observation is important for the future management of this challenging patient population.

Congenital lung lesions

The evidence for fetal surgery for congenital lung lesions suggest that fetal thoracoamniotic shunt placement or fetal surgical resection is safe and feasible (Adzick 2003, Kunisaki 2021)

Sacrococcygeal teratoma

After more than 2 decades of experimental and clinical work, fetal surgery is an accepted treatment option for highly selected fetuses with life-threatening anomalies. Fetal lung masses associated with hydrops are nearly 100% fatal. These lesions can be resected in utero if they are predominantly solid or multicystic. Thoracoamniotic shunt placement may be effective in the setting of a single large cyst. Fetuses diagnosed with left congenital diaphragmatic hernia before 26 weeks' gestation with associated liver herniation and a low right lung to head circumference ratio have a relatively poor prognosis with conventional therapy after birth, but in utero therapeutic approaches have yet to show a comparative

survival benefit. A prospective randomized trial is required to critically evaluate the efficacy of fetal tracheal occlusion for severe diaphragmatic hernia. Fetal sacrococcygeal teratoma complicated with progressive high output cardiac failure may benefit from in utero resection of the tumor (Adzick, Kitano, 2003).

Myelomeningocele

Adzick et al 2011 randomly assigned eligible women to undergo either prenatal surgery before 26 weeks of gestation or standard postnatal repair. One primary outcome was a composite of fetal or neonatal death or the need for placement of a cerebrospinal fluid shunt by the age of 12 months. Another primary outcome at 30 months was a composite of mental development and motor function. The trial was stopped for efficacy of prenatal surgery after the recruitment of 183 of a planned 200 patients. This report is based on results in 158 patients whose children were evaluated at 12 months. The first primary outcome occurred in 68% of the infants in the prenatal-surgery group and in 98% of those in the postnatal-surgery group (relative risk, 0.70; 97.7% confidence interval [CI], 0.58 to 0.84; P<0.001). Actual rates of shunt placement were 40% in the prenatal-surgery group and 82% in the postnatal-surgery group (relative risk, 0.48; 97.7% CI, 0.36 to 0.64; P<0.001). Prenatal surgery also resulted in improvement in the composite score for mental development and motor function at 30 months (P=0.007) and in improvement in several secondary outcomes, including hindbrain herniation by 12 months and ambulation by 30 months. However, prenatal surgery was associated with an increased risk of preterm delivery and uterine dehiscence at delivery. The authors concluded that prenatal surgery for myelomeningocele reduced the need for shunting and improved motor outcomes at 30 months but was associated with maternal and fetal risks.

Myelomeningocele (MMC) is a congenital neural tube defect that occurs in approximately 1 in 2900 live births in the United States. It is a devastating disability with significant morbidity and mortality within the first few decades of life. MMC was the first nonlethal disease to be considered and studied for fetal surgery and is now the most common open fetal surgery performed. The recently completed MOMS randomized controlled trial has shown that fetal repair for MMC can improve hydrocephalus and hindbrain herniation, can reduce the need for vetriculoperitoneal shunting, and may improve distal neurologic function in some patients (Saadai, 2012).

Congenital Diaphragmatic Hernia (CDH)

Fetoscopic endoluminal tracheal occlusion (FETO) has been associated with increased postnatal survival among infants with severe pulmonary hypoplasia due to isolated congenital diaphragmatic hernia on the left side, but data are lacking to inform its effects in infants with moderate disease. In an open-label trial conducted at many centers with experience in FETO and other types of prenatal surgery, we randomly assigned, in a 1:1 ratio, women carrying singleton fetuses with a moderate isolated congenital diaphragmatic hernia on the left side to FETO at 30 to 32 weeks of gestation or expectant care. Both treatments were followed by standardized postnatal care. The primary outcomes were infant survival to discharge from a neonatal intensive care unit (NICU) and survival without oxygen supplementation at 6 months of age. In an intention-to-treat analysis involving 196 women, 62 of 98 infants in the FETO group (63%) and 49 of 98 infants in the expectant care group (50%) survived to discharge (relative risk , 1.27; 95% confidence interval [CI], 0.99 to 1.63; two-sided P = 0.06). At 6 months of age, 53 of 98 infants (54%) in the FETO group and 43 of 98 infants (44%) in the expectant care group were alive without oxygen supplementation (relative risk, 1.23; 95% CI, 0.93 to 1.65). The incidence of preterm, prelabor rupture of membranes was higher among women in the FETO group than among those in the expectant care group (44% vs. 12%; relative risk, 3.79; 95% CI, 2.13 to 6.91), as was the incidence of preterm birth (64% vs. 22%, respectively; relative risk, 2.86; 95% CI, 1.94 to 4.34), but FETO was not associated with any other serious maternal complications. There were two spontaneous fetal deaths (one in each group) without obvious cause and one neonatal death that was associated with balloon removal. The authors concluded that this trial involving fetuses with moderate congenital diaphragmatic hernia on the left side did not show a significant benefit of FETO performed at 30 to 32 weeks of gestation over expectant care with respect to survival to discharge or the need for oxygen supplementation at 6 months. FETO increased the risks of preterm, prelabor rupture of membranes and preterm birth (Deprest et al 2021).

Harrision et al 2003 performed a randomized controlled trial comparing fetal tracheal occlusion with standard postnatal care. Women carrying fetuses that were between 22 and 27 weeks of gestation and that had severe, left-sided congenital diaphragmatic hernia (liver herniation and a lung-to-head ratio below 1.4), with no other detectable anomalies, were randomly assigned to fetal endoscopic tracheal occlusion or standard care. The primary outcome was survival at the age of 90 days; the secondary outcomes were measures of maternal and neonatal morbidity. Of 28 women who met the entry criteria, 24 agreed to randomization. Enrollment was stopped after 24 patients had been enrolled because of the unexpectedly high survival rate with standard care and the conclusion of the data safety monitoring board that further recruitment would not result in significant differences between the groups. Eight of 11 fetuses (73 percent) in the tracheal-occlusion group and 10 of 13 (77 percent) in the group that received standard care survived to 90 days of age (P=1.00). The severity of the congenital diaphragmatic hernia at randomization, as measured by the lung-to-head ratio, was inversely related to survival in both groups. Premature rupture of the membranes and preterm delivery were more common in the group receiving the intervention than in the group receiving standard care (mean [+/-SD] gestational age at delivery, 30.8+/-2.0 weeks vs. 37.0+/-1.5 weeks; P<0.001). The rates of neonatal morbidity did not differ between the groups. The authors concluded that tracheal occlusion did not improve survival or morbidity rates in this cohort of fetuses with congenital diaphragmatic hernia.

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:

- Singleton pregnancy; **AND**
- 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 on fetal magnetic resonance imaging; **AND**
- Normal karyotype; **AND**
- Individuals who meet the conditions for in utero repair of myelomeningocele should be counseled in a nondirective fashion regarding all management options, including the possibility of open maternal–fetal surgery.

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

- Fetal anomaly unrelated to myelomeningocele
- Severe fetal 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.

If this Medical Coverage Guideline contains a step therapy requirement, in compliance with Florida law 627.42393, members or providers may request a step therapy protocol exemption to this requirement if based on medical necessity. The process for requesting a protocol exemption can be found at Coverage Protocol Exemption Request.

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 and Coverage Committee on 07/25/24.

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.
08/21/23	Update to Program Exceptions section.
08/15/24	Review; no change to position statement.Updated references.