Intrauterine Fetal Surgery

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APPLICATION STATEMENT

The application of the Clinical Coverage Guideline is subject to the benefit determinations set forth by the Centers for Medicare and Medicaid Services (CMS) National and Local Coverage Determinations and state-specific Medicaid mandates, if any.

DISCLAIMER

The Clinical Coverage Guideline (CCG) is intended to supplement certain standard WellCare benefit plans and aid in administering benefits. Federal and state law, contract language, etc. take precedence over the CCG (e.g., Centers for Medicare and Medicaid Services [CMS] National Coverage Determinations [NCDs], Local Coverage Determinations [LCDs] or other published documents). The terms of a member’s particular Benefit Plan, Evidence of Coverage, Certificate of Coverage, etc., may differ significantly from this Coverage Position. For example, a member’s benefit plan may contain specific exclusions related to the topic addressed in this CCG. Additionally, CCGs relate exclusively to the administration of health benefit plans and are NOT recommendations for treatment, nor should they be used as treatment guidelines. Providers are responsible for the treatment and recommendations provided to the member. The application of the CCG is subject to the benefit determinations set forth by the Centers for Medicare and Medicaid Services (CMS) National and Local Coverage Determinations and state-specific Medicaid mandates, if any. All links are current at time of approval by the Medical Policy Committee (MPC) and are subject to change prior to the annual review date. Lines of business (LOB) are subject to change without notice; current LOBs can be found at www.wellcare.com. All guidelines can be found at this site as well but selecting the Provider tab, then “Tools” and “Clinical Guidelines”.

BACKGROUND

In utero fetal surgery is surgical treatment of a fetus with certain life-threatening conditions. During this type of surgery, a hysterotomy is performed and the fetus is partially removed to correct the malformation. Fetoscopic surgery uses minimally invasive techniques and devices to correct malformations using small incisions. The key conditions that have been researched include congenital cystic adenomatoid malformation, extralobar pulmonary sequestration, sacrococcygeal teratoma, urinary tract obstruction, twin-twin transfusion syndrome, twin reversed arterial perfusion syndrome, myelomeningocele repair, congenital diaphragmatic hernia and congenital heart disease. The information below highlights common indications for surgery.1
**Thoracic Lesions.** Congenital cystic adenomatoid malformation (CCAM) and bronchopulmonary sequestration (BPS) are congenital anomalies of the lung that share the characteristic of a segment of lung being replaced by abnormally developing tissue. A small percentage of those with congenital pulmonary airway malformations are candidates for in utero treatment. Within this group, the mass is large enough and in such an anatomically critical position that the fetal mediastinum is compressed, leading to an impaired venous return with resulting fetal hydrops secondary to cardiac failure.¹

**Sacrococcygeal Teratoma.** Fetuses with large, vascular sacrococcygeal teratomas (SCT) have a high incidence of prenatal mortality from high-output cardiac failure or spontaneous hemorrhage into or rupture of the growing tumor. Fetal surgical procedures for SCT have focused on the small subgroup of fetuses with SCT and hydrops because untreated cases are expected to die in utero or at birth. In severe cases, SCT with hydrops is associated with maternal risk of developing mirror syndrome, a severe form of preeclampsia.¹

**Urinary Tract Obstruction.** Fetal urinary tract obstruction (UTO) interferes with normal development of the kidneys and lungs, particularly when involving the lower urinary tract. Goals of fetal surgery have emphasized decompression rather than repair of the specific lesion. The goal of decompression of the distended portion of the urinary tract is to protect remaining renal function and to promote lung development. The trend in decompression is towards percutaneous shunting procedures.¹

**Twin-twin Transfusion Syndrome.** In twin-twin transfusion syndrome (TTTS), twins share a single chorionic membrane and therefore a single placenta, but have separate amniotic sacs. Most pregnancies presenting with severe TTTS prior to 26 weeks and not undergoing some sort of therapy will end with dual fetal demise. When both twins do survive, there is often severe neurologic compromise in survivors as well as other organ failure, including cardiac impairment. Treatment options include amnioreduction to relieve pressure and uterine size, termination of the sicker twin or fetoscopic laser ablation of the communicating vessels. In nonselective ablation, all vessels crossing the dividing membrane are ablated, whereas selective ablation is limited to vessels shown to be communicating between the two fetuses.¹

**Twin Reversed Arterial Perfusion** Twin reversed arterial perfusion (TRAP). A TRAP sequence is a condition in which an acardiac/acephalic twin receives all of its blood supply from a normal twin, the so-called "pump" twin. Blood enters the acardiac twin through a retrograde flow via the umbilical artery and exits via the umbilical vein. The extra work places an increased demand on the heart of the pump twin, resulting in cardiac failure. Twin death occurs more frequently when the size of the acardiac twin is greater than half that of the pump twin. The goal of fetal surgery is to interrupt blood supply to the non-viable twin.¹

**Myelomeningocele (MMC).** MMC (or spinal bifida) is a neural tube defect in which the spinal cord forms but remains open. Although MMC is rarely fatal, individuals affected with it have a range of disabilities, including paraplegia, hydrocephalus, skeletal deformities, bowel and bladder incontinence and cognitive impairment. Standard therapy is postnatal surgical closure of the MMC followed by shunting for hydrocephalus if needed.¹

**Congenital Diaphragmatic Hernia (CDH).** CDH results from abnormal development of the diaphragm which allows abdominal organs like the bowel, stomach and liver to protrude into the chest cavity. Fetuses diagnosed in utero as a result of maternal symptoms have a high mortality risk. Less invasive fetal procedures are being developed that focus on methods to accomplish tracheal occlusion.¹

**Congenital Heart Disease.** In utero procedures are performed for cardiac conditions such as pulmonary atresia with intact ventricular septum, critical aortic stenosis with impending hypoplastic left heart syndrome and hypoplastic left heart syndrome with intact atrial septum. All of these conditions, if untreated either in utero or soon after birth, are lethal.¹

**Professional Organizations**

The American College of Obstetricians and Gynecologists (ACOG) issued a bulletin to address the role of fetal surgery for treatment of neural tube defects however no detailed recommendations have been made.² The committee opinion of an ACOG committee opinion states that maternal-fetal surgery is a major procedure for the woman and her fetus, and it has significant implications and complications that occur acutely, postoperatively, for
the duration of the pregnancy and in subsequent pregnancies. It should be offered at facilities with the expertise, multidisciplinary teams, services and facilities to provide the intensive care required for these patients.3

A clinical guideline from the Society for Maternal-Fetal Medicine (SMFM)4 reviewed the history, diagnosis and treatment options for twin-twin transfusion syndrome (TTTS). The SMFM recommends that extensive counseling be provided to patients with pregnancies by TTTS. Further, SMFM states that more than 75% of stage I TTTS cases remain stable or regress without invasive intervention, with perinatal survival of about 86%. Many with stage I TTTS may often be managed expectantly. Fetoscopic laser photocoagulation of placental anastomoses is considered by most experts to be the best available approach for stages II, III and IV TTTS in continuing pregnancies at <26 weeks, but the metaanalysis data show no significant survival benefit, and the long-term neurologic outcomes in the Eurofetus trial were not different than in nonlaser-treated controls. Even laser-treated TTTS is associated with a perinatal mortality rate of 30-50%, and a 5-20% chance of long-term neurologic handicap. Steroids for fetal maturation should be considered at 24 0/7 to 33 6/7 weeks, particularly in pregnancies complicated by stage ≥III TTTS, and those undergoing invasive interventions.

POSITION STATEMENT

Applicable To:
☑ Medicaid
☑ Medicare

Exclusions

Due to a lack of evidence to support its efficacy and medical necessity, intrauterine fetal surgery is considered experimental and investigational for the treatment of:

- Amniotic band syndrome
- Aqueductal stenosis (i.e., hydrocephalus)
- Cleft lip and/or cleft palate
- Congenital heart disease (e.g. mitral valve dysplasia)
- Fetal hydronephrosis
- Gastrochisis
- Fetal tracheal occlusion for congenital diaphragmatic hernia (CDH)7
- Congenital heart disease (CHD)

In addition, the following are considered experimental and investigational for the following:

- In utero stem cell transplantation
- In utero gene therapy

Coverage

Intrauterine fetal surgery is considered medically necessary for the following:

- **Congenital cystic adenomatoid malformation (CCAM) and extralobar pulmonary sequestration (EPS):** Resection of malformed pulmonary tissue, or placement of a thoracoamniotic shunt as a treatment of Congenital cystic adenomatoid malformation **OR** Extralobar pulmonary sequestration.5

- **Sacroccygeal teratoma (SCT):** SCT resection

- **Urinary tract obstruction (UTO):** urinary decompression via vesicoamniotic shunt placement

- **Twin-twin transfusion syndrome:** fetoscopic laser surgery consisting of an ablation of anastomotic vessels in early, severe twin-twin transfusion syndrome. Serial amnioreduction for twin-to-twin transfusion syndrome medically necessary when criteria are met:
  - Women after 26 weeks of gestation; and
  - Evidence of abnormal blood flow documented by Doppler studies in one or both fetuses; and
  - Evidence of polyhydramnios in the recipient fetus; and
Donor fetus is oligohydramniotic.

- **Twin reversed arterial perfusion (TRAP):** ablation or occlusion of anastomotic vessels (e.g., laser coagulation or radiofrequency ablation)
- **Myelomeningocele (MMC) repair**
- Insertion of pleuro-amniotic shunt for fetal pleural effusion

**CODING**

**Covered CPT Codes**
- 59070  Transabdominal amnioinfusion, including ultrasound guidance
- 59072  Fetal umbilical cord occlusion, including ultrasound guidance
- 59074  Fetal fluid drainage (e.g., vesicocentesis, thoracocentesis, paracentesis), including ultrasound guidance
- 59076  Fetal shunt placement, including ultrasound guidance

**Covered HCPCS Codes**
- 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
- S2411  Fetoscopic laser therapy for treatment of twin-to-twin transfusion syndrome

**Covered ICD-10 CM Diagnosis Codes**
- D48.0  Neoplasm of uncertain behavior of bone and articular cartilage
- O33.7XX0-O37.7XX9  Maternal care for disproportion due to other fetal deformities  
  NOTE: Both in Covered and non-covered
- O35.8xx0  Maternal care for other (suspected) fetal abnormality and damage, not applicable or unspecified
- O35.8xx0-035.8XX9
- O36.8210  Fetal anemia and thrombocytopenia, first trimester, not applicable or unspecified
- O36.8211  Fetal anemia and thrombocytopenia, first trimester, fetus 1
- O36.8212  Fetal anemia and thrombocytopenia, first trimester, fetus 2
- O36.8213  Fetal anemia and thrombocytopenia, first trimester, fetus 3
- O36.8214  Fetal anemia and thrombocytopenia, first trimester, fetus 4
- O36.8215  Fetal anemia and thrombocytopenia, first trimester, fetus 5
- O36.8219  Fetal anemia and thrombocytopenia, first trimester, other fetus
- O36.8220  Fetal anemia and thrombocytopenia, second trimester, not applicable or unspecified
- O36.8221  Fetal anemia and thrombocytopenia, second trimester, fetus 1
- O36.8222  Fetal anemia and thrombocytopenia, second trimester, fetus 2
- O36.8223  Fetal anemia and thrombocytopenia, second trimester, fetus 3
- O36.8224  Fetal anemia and thrombocytopenia, second trimester, fetus 4
- O36.8225  Fetal anemia and thrombocytopenia, second trimester, fetus 5
- O36.8229  Fetal anemia and thrombocytopenia, second trimester, other fetus
- O36.8230  Fetal anemia and thrombocytopenia, third trimester, not applicable or unspecified
- O36.8231  Fetal anemia and thrombocytopenia, third trimester, fetus 1
- O36.8232  Fetal anemia and thrombocytopenia, third trimester, fetus 2
- O36.8233  Fetal anemia and thrombocytopenia, third trimester, fetus 3
- O36.8234  Fetal anemia and thrombocytopenia, third trimester, fetus 4
- O36.8235  Fetal anemia and thrombocytopenia, third trimester, fetus 5
- O36.8239  Fetal anemia and thrombocytopenia, third trimester, other fetus
- O36.8990  Maternal care for other specified fetal problems, unspecified trimester, not applicable or unspecified
- O30.021  Conjoined twin pregnancy, first trimester
- O30.022  Conjoined twin pregnancy, second trimester
- O30.023  Conjoined twin pregnancy, third trimester

Coding information is provided for informational purposes only. The inclusion or omission of a CPT, HCPCS, or ICD-10 code does not imply...
member coverage or provider reimbursement. Consult the member's benefits that are in place at time of service to determine coverage (or non-coverage) as well as applicable federal / state laws.

REFERENCES


MEDICAL POLICY COMMITTEE HISTORY AND REVISIONS

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