I. POLICY/Criteria

Fetal surgery may be considered medically necessary when evaluated and performed at fetal surgery centers which offer comprehensive evaluation, surgery, support services and follow-up. The following conditions may be considered for fetal surgery when meeting the below associated criteria and the criteria of the fetal surgery center:

A. Vesico-amniotic shunting as a treatment of urinary tract obstruction for fetuses with hydronephrosis due to bilateral urinary tract obstruction when ALL of the following criteria are met:

- evidence of progressive oligohydramnios
- evidence of adequate renal function
- no other lethal abnormalities or chromosomal defects
- male fetus
- no other maternal complications

B. Either open in utero resection of malformed pulmonary tissue or placement of a thoraco-amniotic shunt as a treatment of congenital cystic adenomatoid malformation, extralobar pulmonary sequestration or pleural effusion when ALL of the following criteria are met:

- fetuses of 32 weeks' gestation or less
- evidence of fetal hydrops or high risk of high cardiac output
- no other lethal abnormalities or chromosomal defects
- no other maternal complications

C. In utero removal of sacrococcygeal teratoma (SCT) when ALL of the following criteria are met:

- fetuses of 32 weeks' gestation or less
- high risk of high cardiac output
- no other lethal abnormalities or chromosomal defects
D. Fetoscopic laser coagulation as a treatment for severe twin-to-twin transfusion syndrome (TTTS) as medically necessary when ALL of the following criteria are met:

- The gestational age of the fetus is less than 26 weeks
- There is evidence of polyhydramnios in the recipient fetus
- The donor fetus is oligohydramniotic
- There is an absence of other fetal abnormalities
- No other maternal complications

E. Fetal surgery is considered medically necessary for in utero repair of myelomeningocele when the following conditions are met:

- Singleton pregnancy; AND
- Myelomeningocele with the upper boundary of the lesion located between T1 and S1; AND
- Evidence of hindbrain herniation; AND
- Gestational age of 19.0 to 25.9 weeks; AND
- Normal fetal karyotype; AND
- Absence of ALL of the following:
  i. Fetal anomaly unrelated to the myelomeningocele; AND
  ii. Severe fetal kyphosis; AND
  iii. Short cervix (less than or equal to 15 mm); AND
  iv. Previous pre-term birth; AND
  v. Placental abruption; AND
  vi. Maternal Body Mass Index (BMI) greater than or equal to 35; AND
  vii. Contraindications to surgery, including but not limited to previous hysterotomy in the active (upper) uterine segment.

F. Other applications of fetal surgery are considered investigational, including but not limited to temporary tracheal occlusion as a treatment of congenital diaphragmatic hernia, fetal surgery for aqueductal stenosis (hydrocephalus), heart block, pulmonary valve or aortic obstruction, tracheal atresia or stenosis, and cleft lip and palate. In utero gene therapy and in utero stem cell transplantation are also considered to be experimental.

II. MEDICAL NECESSITY REVIEW

☑ Required  ☐ Not Required  ☐ Not Applicable
III. APPLICATION TO PRODUCTS

Coverage is subject to member’s specific benefits. Group specific policy will supersede this policy when applicable.

- **HMO/EPO:** This policy applies to insured HMO/EPO plans.
- **POS:** This policy applies to insured POS plans.
- **PPO:** This policy applies to insured PPO plans. Consult individual plan documents as state mandated benefits may apply. If there is a conflict between this policy and a plan document, the provisions of the plan document will govern.
- **ASO:** For self-funded plans, consult individual plan documents. If there is a conflict between this policy and a self-funded plan document, the provisions of the plan document will govern.
- **INDIVIDUAL:** For individual policies, consult the individual insurance policy. If there is a conflict between this medical policy and the individual insurance policy document, the provisions of the individual insurance policy will govern.
- **MEDICARE:** Coverage is determined by the Centers for Medicare and Medicaid Services (CMS); if a coverage determination has not been adopted by CMS, this policy applies.
- **MEDICAID/HEALTHY MICHIGAN PLAN:** For Medicaid/Healthy Michigan Plan members, this policy will apply. Coverage is based on medical necessity criteria being met and the appropriate code(s) from the coding section of this policy being included on the Michigan Medicaid Fee Schedule located at: [http://www.michigan.gov/mdch/0,1607,7-132-2945_42542_42543_42546_42551-159815--.00.html](http://www.michigan.gov/mdch/0,1607,7-132-2945_42542_42543_42546_42551-159815--.00.html). If there is a discrepancy between this policy and the Michigan Medicaid Provider Manual located at: [http://www.michigan.gov/mdch/0,1607,7-132-2945_5100-87572--.00.html](http://www.michigan.gov/mdch/0,1607,7-132-2945_5100-87572--.00.html), the Michigan Medicaid Provider Manual will govern. If there is a discrepancy or lack of guidance in the Michigan Medicaid Provider Manual, the Priority Health contract with Michigan Medicaid will govern. For Medical Supplies/DME/Prosthetics and Orthotics, please refer to the Michigan Medicaid Fee Schedule to verify coverage.

IV. DESCRIPTION

The objective of **fetal surgery** is to correct malformations of the fetus that interfere with organ development and fetal survival and have potentially fatal consequences. The specific techniques used for fetal surgery are determined by the diagnosis and specific circumstances of each individual case.

**Intrauterine or in utero fetal surgery (IUFS):**

**Open fetal surgery** involves making an incision through the abdomen of the mother and partial removal of the uterus from the body. Amniotic fluid is drained from the uterus and kept in a warmer for replacement after completion of the surgery. An incision is made in the uterus. Surgery is then performed on the fetus through the opening in the uterus to locate the abnormality and remove or fix it. Because of the nature of open fetal surgery, delivery for this child and all subsequent children of this mother will have to be performed by **cesarean section**.

**Fetoscopic surgery:** This type of surgery, which employs minimally invasive techniques, is used more often than open surgery. Surgeons can use fiberoptic...
telescopes and specially designed instruments to enter the uterus through small surgical openings to correct congenital malformations without major incisions or removing the fetus from the womb. This alternative is less traumatic and reduces the chances of preterm labor.

**Fetoscopic laser surgery** is a procedure for ablating abnormal vascular connections between monozygotic twins who share a single placenta and are affected by twin-twin transfusion syndrome (TTTS). TTTS is a serious and potentially fatal complication resulting from an imbalance in net blood flow from one twin, the donor, to the other, the recipient. Laser energy is used to ablate the placental connecting vessels, thereby interrupting fetofetal blood flow and restoring circulatory balance. The goal of this procedure is to decrease the risk of intrauterine fetal and neonatal death and improve the outcomes of the surviving infants.

**Fetal urinary-tract obstruction:** Lower urinary obstruction in the fetus is an obstruction to the flow of urine out of the bladder, causing backup of urine and damage to the kidneys. The most common cause of bladder obstruction is posterior urethral valves in males although the condition may be linked to a genetic abnormality. The patient selection criteria for intervention are based upon fetal-urine electrolyte studies, beta²-microglobulin levels and the use of ultrasound. The severity of damage at birth depends on the type, degree and duration of the obstruction. Conditions of minimal renal dysfunction and normal pulmonary development can be treated after delivery. Unilateral obstruction does not lead to oligohydramnios (decrease in amniotic fluid). However, bilateral urinary obstruction in the fetus is often associated with serious adverse outcomes, such as pulmonary hypoplasia secondary to oligohydramnios.

The most common surgical approach to repair the obstruction is vesicoamniotic shunting by means of a shunt or a stent inserted into the urinary tract above the obstruction and then passed through the abdominal wall to drain into the amniotic sac. This method of treatment restores amniotic fluid, preventing pulmonary hypoplasia.

**Congenital cystic adenomatoid malformation (CCAM)/congenital pulmonary airway malformation (CPAM):** Congenital cystic adenomatoid malformation, recently termed Congenital Pulmonary Airway Malformation (CPAM) is a benign cystic pulmonary mass that may lead to fetal hydrops and pulmonary hypoplasia. The CCAM/CPAM is typically unilateral and unilobular and receives blood supply from the pulmonary vasculature. The condition may result in air trapping and progressive respiratory compromise. Large lesions may cause mediastinal shift and fetal hydrops, pulmonary hypoplasia and persistent pulmonary hypertension. The mortality rate approaches 100% for cases in which both CCAM/CPAM and fetal hydrops are present, fortunately fetal hydrops occurs in fewer than 10% of cases. Most lesions can be successfully treated after birth, and some may resolve prior to birth; it is rare, however, that resolution of hydrops occurs in conjunction with
regression of the lesion (Adzick, 1996). When large lesions are identified prior to 26 weeks of gestation, the disease progresses rapidly, ultimately resulting in fetal demise.

**Sacrococcygeal teratoma (SCT):** A sacrococcygeal teratoma is a tumor derived from more than one embryonic germ layer. Most tumors are benign, but the odds of malignancy increase with increasing age. In many cases, the abnormal size of the uterus (from either the tumor or polyhydramnios) leads to diagnosis by ultrasound. Less commonly, presentation may include maternal pre-eclampsia.

The standard treatment is complete excision after birth if not detected prenatally. When SCT is detected prenatally, early surgical intervention may be performed to prevent the development of fetal hydrops. These are extremely vascular tumors. Fetal hydrops develops as a result of vascular shunting between low-pressure vessels within the tumor, leading to cardiovascular collapse in cases of large lesions. Left uncorrected, SCT, when it occurs in conjunction with high output failure that is associated with placentomegaly or hydrops, results in 100% fetal mortality.

**Extralobar pulmonary sequestration (EPS):** Bronchopulmonary sequestration is a condition characterized by the presence of nonfunctioning lung tissue which is not connected to the tracheal bronchial tree. It may be intralobar or extralobar. The ability to determine the actual type of sequestration is very limited unless extralobar pulmonary sequestration (EPS) is associated with pleural effusion or is located in the abdomen. If not corrected, bronchopulmonary sequestration results in abnormal respiratory functioning and ultimately in fetal hydrops. Large lesions may cause esophageal compression, which may interfere with fetal swallowing of amniotic fluid and eventually result in polyhydramnios. Fetal hydrops develops secondary to vena caval obstruction and cardiac compression. Bronchopulmonary sequestration may also result in a tension hydrothorax from associated fluid or lymph secretion. In-utero correction involves placement of a thoracoamniotic shunt and is supported mainly by evidence on the form of case reports and reviews (Adzick, et al, 1998; Adzick, et al, 2003).

**Pleural effusions:** Isolated fetal pleural effusions have an incidence rate of approximately 1:10,000 to 15,000 pregnancies and may be bilateral but are most commonly unilateral. There are a variety of causes which include congenital abnormalities and chromosomal abnormalities. The persistence of pleural effusion in early pregnancy interferes with normal lung development and often results in pulmonary hypoplasia. Treatment consists of draining the intrathoracic fluid by the insertion of pleuro-amniotic shunts or by thoracentesis, where liquid is drained after single or multiple transthoracic punctures.

**Twin-to-twin transfusion syndrome:**
Twin-twin transfusion syndrome (TTTS) results from unbalanced blood flow through vascular arteriovenous anastomoses, which are present in the placenta of the majority of monochorionic multiple pregnancies. With preferential blood flow in severe cases, one twin becomes the donor and the other is the recipient, which may result in wide discordance in fetal growth.

The recipient twin, the larger fetus in the amniotic sac with polyhydramnios, typically has a large umbilical cord, abdominal circumference, kidneys, and bladder. Excessive volume can lead to cardiovascular decompensation with cardiomegaly, tricuspid regurgitation, and ventricular hypertrophy; hydrops fetalis may also develop. Polycythemia in the recipient fetus can lead to thrombosis or hyperbilirubinemia after birth.

The donor twin is the smaller fetus and is in the oligohydramniotic sac. This fetus may have severe growth restriction with anemia, hypovolemia, and renal insufficiency (UptoDate, 2017)

Standard interventions include amnioreduction and fetoscopic laser surgery performed percutaneously or through open surgery. The most severe cases are those diagnosed prior to 25 weeks of gestation.

The most widely used therapy for TTTS, amnioreduction, seeks to equalize the volume of amniotic fluid between the twins. This treatment involves serial amniocentesis and is recommended for pregnancies of gestation later than 26 weeks if delivery is not an option. Amnioreduction does not correct the underlying vascular abnormality. Survival rates have been reported to be between 50–65% with this intervention (Children’s Hospital of Philadelphia [CHOP]).

Fetoscopic laser surgery corrects the underlying circulatory imbalance. The surgery may be performed through an open approach or percutaneously. Laser energy is used to ablate the placental anastomoses, thus interrupting fetal blood-flow transfusion and restoring the circulatory balance. The reported survival rates average 67%, with 80% of pregnancies having at least one survivor; laser photocoagulation is associated with reduced neurologic morbidity (CHOP).

**Myelomeningocele:** Myelomeningocele, commonly referred to as spina bifida, is a neural-tube defect in which the spinal cord forms but remains open, exposing the meninges and neural tube to the intrauterine environment. The defect may include abnormal positioning of the brain (Arnold-Chiari II malformation). A variety of medical problems may result from the open neural tube. These include, but are not limited to, physical and mental disabilities, deformity of the extremities, scoliosis, and urinary dysfunction or failure. Some researchers contend that the intrauterine exposure causes secondary trauma to the spinal cord.
Traditional treatment consists of surgical repair after delivery, with ventriculoperitoneal shunting. In-utero surgical repair to the fetus has been proposed as a way to improve neurological outcomes; however, the procedure’s long-term effects on brain function have not been determined.

Three types of fetal surgery are performed to treat myelomeningocele: fetoscopic myelomeningocele repair; maternal hysterotomy; and microsurgical, three-layered, fetal myelomeningocele repair (fetal patch repair). Myelomeningocele repair consists of closing the dura and skin over the exposed spinal cord. Data from the Management of Myelomeningocele Study (MOMS) compared the results of prenatal and postnatal myelomeningocele repair. After recruiting 183 of the planned 200 subjects the trial was stopped due to significantly improved clinical outcomes for the prenatal surgery group compared to the post-natal treatment group. In 2011, Adzick and colleagues published the results of this trial which included 158 subjects who completed up to 12 months follow-up; 134 of those subjects were also available for evaluation at 30 months. Individuals were randomized to receive myelomeningocele repair in-utero or repair following delivery. Inclusion and exclusion criteria were as follows:

<table>
<thead>
<tr>
<th>Inclusion Criteria MOMS Trial</th>
<th>Exclusion Criteria MOMS Trial</th>
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<tbody>
<tr>
<td>Singleton pregnancy</td>
<td>Unrelated fetal anomaly</td>
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<tr>
<td>Myelomeningocele with upper</td>
<td>Severe kyphosis</td>
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<tr>
<td>boundary located between T1</td>
<td>Risk of preterm birth</td>
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<tr>
<td>and S1</td>
<td>(including short cervix and</td>
</tr>
<tr>
<td>Evidence of hindbrain</td>
<td>previous preterm birth)</td>
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<tr>
<td>herniation</td>
<td>Placental abruption</td>
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<tr>
<td>Gestational age of 19.0 to</td>
<td>Body mass index of 35 or</td>
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<tr>
<td>25.9 weeks at birth</td>
<td>more</td>
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<tr>
<td>Normal karyotype</td>
<td>Contraindication to surgery</td>
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<tr>
<td>U.S. residency</td>
<td>(e.g., including previous</td>
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<tr>
<td>Maternal age of at least 18</td>
<td>hysterotomy in the active</td>
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<td>years</td>
<td>uterine segment)</td>
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ACOG published a committee opinion (ACOG, 2013) acknowledging publication of the MOMS trial and the rigorous requirements for the study. ACOG noted further that maternal fetal surgery has significant implications and complications that may occur acutely, postoperatively, for the duration of the pregnancy and in subsequent pregnancies. The Committee recommends that treatment is only offered at facilities with the expertise, multidisciplinary teams, services and facilities to provide the intensive care required for these patients.

V. CODING INFORMATION

**ICD-10 Codes:**
*Not specified - see criteria*

**HCPCS/CPT Codes:**
* PA rules do not apply
36460* Transfusion, intrauterine, fetal
76941* Ultrasonic guidance for intrauterine fetal transfusion or cordocentesis, imaging supervision and interpretation

59012* Cordocentesis (intrauterine), any method
59015* Chorionic villus sampling, any method
59030* Fetal scalp blood sampling

59070* Transabdominal amniinfusion, including ultrasound guidance
59072* Fetal umbilical cord occlusion, including ultrasound guidance
59074* Fetal fluid drainage (eg, vesicocentesis, thoracocentesis, paracentesis), including ultrasound guidance
59076* Fetal shunt placement, including ultrasound guidance
59897* Unlisted fetal invasive procedure, including ultrasound guidance (Use this code in lieu of S codes for Priority Medicare or Medicaid claims. Explanatory notes must accompany all claims)

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

**“S” codes not covered for Priority Medicare, Medicaid**

Not covered:
S2400 Repair, congenital diaphragmatic hernia in the fetus using temporary tracheal occlusion, procedure performed in utero
S2409 Repair, congenital malformation of fetus, procedure performed in utero, not otherwise classified

VI. REFERENCES


Fetal Surgery, Cigna HealthCare Coverage Position # 0175, September 15, 2005.  

Fetal Surgery for Prenatally Diagnosed Malformations, The Regence group of The Blue Cross and/or Blue Shield Plans, policy # 13, July 5, 2005.  
www.regence.com accessed April 6, 2006

Amnioreduction and/or Fetoscopic Laser Therapy for the Treatment of Twin-Twin Transfusion Syndrome, The Regence group of The Blue Cross and/or Blue Shield Plans, policy #12, November 5, 2002.  
www.regence.com accessed April 6, 2006

Endoscopic laser surgery versus serial amnioreduction for severe twin-to-twin transfusion syndrome.  


“Fetal Surgery for Prenatally Diagnosed Malformations” Anthem Blue Cross Medical Policy SURG.00036.  

“Fetal Surgery for Prenatally Diagnosed Malformations” Blue Cross Blue Shield Blue Care Network of Michigan Medical Policy.  

“Intrauterine Fetal Surgery” Humana Medical Coverage Policy HGO-0364-009.  

“Intrauterine Fetal Surgery” UnitedHealthcare Commercial Medical Policy 2017T0035P.  


AMA CPT Copyright Statement:
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Priority Health’s medical policies are intended to serve as a resource to the plan. They are not intended to limit the plan’s ability to interpret plan language as deemed appropriate. Physicians and other providers are solely responsible for all aspects of medical care and treatment, including the type, quality, and levels of care and treatment they choose to provide.

The name “Priority Health” and the term “plan” mean Priority Health, Priority Health Managed Benefits, Inc., Priority Health Insurance Company and Priority Health Government Programs, Inc.