### Coverage Policy

Cigna covers ANY of the following fetal surgeries as medically necessary for the associated indication(s):

- serial amnio-reduction for twin-to-twin transfusion syndrome (TTTS)
- fetoscopic occlusion of anastomotic vessels (e.g., laser photocoagulation, radiofrequency ablation, ligation) for twin reversed arterial perfusion (TRAP sequence)
- fetal vesico-amniotic shunt procedures for bilateral fetal urinary-tract obstruction
- in-utero needle access and open resection of sacrococcygeal teratoma
- fetal thoraco-amniotic shunt placement for ANY of the following indications:
  - congenital cystic adenomatoid formation (CCAM)
  - extralobar pulmonary sequestration (EPS)
  - fetal pleural effusion
- myelomeningocele repair when ALL of the following criteria are met:
  - singleton pregnancy
  - myelomeningocele with the upper boundary of the lesion located between T1 and S1
  - evidence of hindbrain herniation
  - gestational age ≥19.0 and < 26 weeks
  - normal fetal karyotype
- nonselective or selective fetoscopic laser coagulation for severe twin-to-twin transfusion syndrome (TTTS) when ALL of the following criteria are met:
  - fetal gestational age of less than 26 weeks
  - evidence of polyhydramnios in the recipient fetus
  - donor fetus is oligohydramniotic

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### Hyperlink to Related Coverage Policies

- Ultrasound in Pregnancy (Including 3D and 4D Ultrasound)
- evidence of abnormal blood flow documented by Doppler studies in one or both fetuses
- fetal lobectomy for congenital cystic adenomatoid formation (CCAM)/congenital pulmonary airway malformation (CPAM) when BOTH of the following criteria are met:
  - evidence of fetal hydrops
  - presence of large and multicystic or predominantly solid lesions

Cigna does not cover ANY of the following because each is considered experimental, investigational or unproven (this list may not be all-inclusive):

- laser, thermocoagulation or radiofrequency ablation techniques for the treatment of sacrococcygeal teratoma
- endoscopic approach (i.e., fetoscopic cystoscopy) for the treatment of lower urinary tract obstruction
- amniotic band syndrome
- aqueductal stenosis (i.e., hydrocephalus)
- cleft lip and/or cleft palate
- congenital diaphragmatic hernia
- congenital heart defects
- in-utero gene therapy
- in-utero hematopoietic stem-cell transplantation for stem-cell-related diseases

General Background

In-utero fetal surgery involves a highly technical, multidisciplinary approach to correct malformations of the fetus that interfere with organ development and that have potentially fatal outcomes if left untreated. The procedure involves opening the gravid uterus through the less-invasive laparoscopic technique or through an open caesarian surgical incision; surgically correcting the fetal abnormality; and closing the uterus to allow gestational development to complete. Fetal surgery should be performed by highly trained physicians, in advanced centers equipped to provide extracorporeal membrane oxygenation (ECMO) in Level III newborn intensive care units. The multidisciplinary approach employs pediatric surgeons, intensive care specialists, geneticists, ethicists, perinatologists, gynecological specialists, maternal/fetal specialists, pathologists and utilizes highly specialized radiology.

Fetal endoscopic surgery, a recently developed method of treating congenital conditions, can lessen maternal morbidity and additional stress to the fetus when the latter is removed from the amniotic fluid environment. Combined with the use of tocolytic drugs, this procedure may also decrease the occurrence of postoperative preterm labor.

Fetal intervention is recommended when preterm delivery is contraindicated and the condition can be corrected allowing for normal development. Experts generally recommend early surgical intervention after a confirmed diagnosis of fetal decompensation. In general, surgery is performed prior to 32 weeks of gestation; after that time, standard treatment consists of early delivery and medically necessary interventions.

There are several contraindications to in-utero surgery, including severe congenital anomalies, chromosomal anomalies that jeopardize fetal survival, and maternal mirror syndrome. Patients with maternal mirror syndrome are not considered candidates for prenatal intervention, as this condition may warrant immediate delivery. Maternal mirror syndrome is a maternal illness where the mother’s condition mimics that of the sick fetus, as a result of severe fetal hydrops. Fetal hydrops is a condition where there is accumulation of fluid in two or more fetal compartments (e.g., abdomen, pleural space, pericardial space) due to multiple factors. With maternal mirror syndrome related to a hyperdynamic cardiovascular state, the mother develops symptoms that are similar to pre-eclampsia and may include vomiting, hypertension, peripheral edema, proteinuria and pulmonary edema. For cases of severe fetal hydrops where the cause is unknown and unable to be corrected, immediate delivery is indicated (Vidaeff, et al., 2002).

Fetal surgery has been researched for many different fetal abnormalities; however, when compared to traditional post-natal therapy, it has been shown to improve outcomes for only a few conditions that include myelomeningocele repair, twin-to-twin transfusion syndrome, twin reversed arterial perfusion syndrome,
urinary-tract obstruction, congenital cystic adenomatoid malformation, extralobar pulmonary sequestration, and sacrococcygeal teratoma. Few published studies have evaluated the safety and efficacy of fetal surgery for other conditions such as congenital heart defects, stem cell research and cleft lip and palate.

**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. Reduction in hindbrain herniation has been reported by some authors (Adzick, et al., 2011; Sutton, et al., 1999) as well as reduction in shunt-dependent hydrocephalus (Adzick, et al., 2011; Tulipan, et al., 2003; Bruner, et al., 1999).

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.

Many maternal complications associated with myelomeningocele repair have been reported. They include uterine rupture, placental abruption, and maternal bowel obstruction, which may occur as a result of post-hysterotomy adhesions. There is also increased risk of oligohydramnios, pre-term uterine contractions, and delivery at earlier estimated gestation and smaller birth weight. Infants born after in-utero repair may suffer not only from the myelomeningocele itself, but from prematurity as well.

Data evaluating in-utero repair of myelomeningocele is limited. However there is some evidence to support improved clinical outcomes. Johnson, et al., 2003 (n=50) reported overall perinatal survival of 94% with reversal of hindbrain herniation in all fetuses. Venticuloperitoneal shunting was required in 43% of the fetuses, compared to 68–100% in historical controls. Better-than-predicted leg function was demonstrated in 57% of thoracic- and lumbar-level patients. In 2006 Johnson et al. reported the neurodevelopmental and cognitive outcomes in children two years of age who underwent myelomeningocele repair in utero. Neurodevelopmental deficits were noted but did not appear to be worsened by fetal surgery; the deficits were considered characteristic of children with spina bifida.

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 (See Table 1):

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<th>TABLE 1:</th>
<th>Inclusion Criteria MOMS Trial</th>
<th>Exclusion Criteria MOMS Trial</th>
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<tr>
<td>Singleton pregnancy</td>
<td>Myelomeningocele with upper boundary located between T1 and S1</td>
<td>Unrelated fetal anomaly</td>
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<tr>
<td>Evidence of hindbrain herniation</td>
<td>Gestational age of 19.0 to 25.9 weeks at randomization</td>
<td>Severe kyphosis</td>
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<td>Normal karyotype</td>
<td>U.S. residency</td>
<td>Risk of preterm birth (including short cervix and previous preterm birth)</td>
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<td>Maternal age of at least 18 years</td>
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<td>Placental abruption</td>
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<td>Body-mass index of 35 or more</td>
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<td>Contraindication to surgery (e.g., including previous hysterotomy in the active uterine segment)</td>
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The primary outcomes measured included fetal death or the need for cerebrospinal fluid shunt by the age of 12 months, and at 30 months, a composite score of the Mental Development Index of the Bayley Scales of Infant Development II and the child’s motor function, with adjustment for lesion level. Secondary outcome measures included maternal, fetal, and neonatal surgical and pregnancy complications and neonatal morbidity and mortality as well as several other secondary outcomes. The authors reported the following results:

- The first primary outcome, fetal death or the need for cerebrospinal fluid shunt by the age of 12 months was significantly better in the prenatal surgery group (68%) compared to the postnatal surgery group (98%) (P < 0.001).
- The rates of actual shunt placement were 40% for the prenatal surgery group compared to 82% in the postnatal surgery group.
- At 12 months of age, the number of infants who had no evidence of hindbrain herniation was higher in the prenatal surgery group compared to the postnatal surgery group (36% versus 4%, respectively).
- At 12 months the prenatal surgery group also demonstrated lower rates of brainstem kinking, abnormal fourth ventricle location and syringomyelia.

The secondary outcome, made up of data from the Bayley Mental Developmental Index and the difference between the functional and anatomical lesion was calculated at 30 months and was significantly better in the prenatal surgery group (mean 148.6 vs. mean 122.6, P < 0.007). In the post hoc analysis, the authors reported that subjects in the prenatal surgery group were more likely to have a level of function two or more levels better than their anatomical level (32% vs. 12%, P < 0.005), and were more likely to ambulate without orthotics or other devices (42% vs. 21%, P < 0.01). The authors noted the prenatal surgery group had significantly better motor function scores on the Bayley and Peabody motor scales, although this same group had more severe anatomical lesion levels at baseline. Between groups cognitive scores were not significantly different. The authors acknowledged the prenatal surgery group had significantly higher rates of pre-term birth and uterine dehiscence at delivery; early intervention was associated with both maternal and fetal morbidity. Nonetheless, prenatal surgery for myelomeningocele reduced the need for shunting and improved motor outcomes at 30 months follow-up. When considering prenatal myelomeningocele repair, the potential benefits of prenatal surgery must be balanced against the risks of prematurity and maternal morbidity. The authors agreed additional follow-up is necessary to assess long-term outcomes, and to evaluate the effect of prenatal intervention on bowel and bladder continence, sexual function and mental capacity (Adzick, et al., 2011).

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.

**Twin-to-Twin Transfusion Syndrome**

Twin-to-twin transfusion syndrome (TTTS) is a condition in which abnormal chorionic vessels in the placenta connect the circulatory systems of two fetuses. As a result, the placenta does not correctly supply oxygen and nutrients to the fetuses’ circulation and causes an uneven blood flow to the twins. One twin (the recipient) receives excess blood, and the other (the donor) receives insufficient blood. Increased blood flow to the recipient results in hypervolemia, polyuria and polyhydramnios and, subsequently, in cardiac overload and congestive heart failure. The decreased blood flow to the donor results in hypovolemia, oliguria and oligohydramnios and, subsequently, in anemia and growth retardation. Although it occurs most frequently in twin pregnancies, it may occur in triplet or higher order multiple gestations provided that at least two of the fetuses are monochorionic (Quintero, 2003).

Standard interventions include selective termination, amnioreduction and fetoscopic laser surgery performed percutaneously or through open surgery.

The most severe cases are those diagnosed prior to 25 weeks of gestation. If TTTS is diagnosed in the second trimester and left untreated, the mortality rate rises to 80–90%. By 28 weeks of gestation, chances for survival improve, although the surviving fetus is prone to neurological damage and developmental impairment.
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).

The laser ablation, which is followed by amnioreduction, may be nonselective or selective. In nonselective laser treatment, all anastomosed vessels that cross the inter-twin septum are ablated, thereby creating a dichorionic placenta. In the selective approach, the ablation is limited to the participating vessels. Fetal and neonatal survival rates following selective ablation are higher than those following nonselective ablation, with a lower rate of spontaneous abortion.

While some research indicates that serial amniocenteses of the polyhydramniotic sac may stabilize the pathophysiological balance, other studies have shown that methods of interrupting the abnormal vascular connections may improve outcomes. In the peer-reviewed, published literature, no single therapy is associated with superior outcomes. Both amnioreduction and laser surgery have resulted in perinatal survival rates of 60–65% (Fisk and Galea, 2004).

Published studies evaluating treatment for twin-to-twin transfusion syndrome consist of prospective, retrospective and randomized trials (Kowitt, et al., 2012; Salomon, et al., 2010; Cincotta, et al., 2009; Crombleholme, et al., 2008; Rossi, et al., 2008; Graef, et al., 2006; Bussey, et al., 2004; Senat, et al., 2004; Quintero, et al., 2000). Several studies lend support to improved health outcomes, including perinatal survival and survival without neurological complications. The World Association of Perinatal Medicine (WAPM) consensus group published recommendations and guidelines for perinatal evaluation and treatment of TTTS (WAPM, 2011) supporting fetoscopic laser coagulation as the treatment of choice and amnioreduction as a palliative treatment that may prolong pregnancy by reducing the risk of polyhydramnios and relieve maternal discomfort. In 2013 the Society for Maternal-Fetal Medicine (SMFM) published clinical guidelines for TTTS. Within the guidelines the SMFM notes perinatal survival for Stage I cases is approximately 86% and that more than three quarters of cases regress or remain stable without invasive intervention. As a result many of Stage I cases can be managed conservatively. The natural history of advanced Stage (>III) is bleak. When invasive intervention is warranted fetoscopic laser photocoagulation is considered by most experts as the best available approach for Stage II, III and IV TTTS in pregnancies continuing at 26 weeks. However the meta-analysis did not support significant survival benefit and long term neurologic outcomes in the Eurofetus trial were no different than in non-laser treated controls. Despite laser treatment TTTS is associated with perinatal mortality of 30-50% and long-term neurologic handicap of 5-20%. The SMFM recommends extensive counseling to couples with pregnancies complicated by TTTS.

Twin Reversed Arterial Perfusion (TRAP) Syndrome
Twin reversed arterial perfusion (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 by 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.

If this condition is left untreated, mortality is 50–75% and occurs frequently, especially when the size of the acardiac/acephalic twin is greater than half that of the pump twin. Twin death occurs in 64% of cases in which the ratio of acardiac to pump twin weight is greater than 50%. The mortality for the pump twin increases to 90% for weight ratios greater than 75. Evaluation consists of three parts: umbilical-cord Doppler velocimetry and echocardiography to determine reversed flow; determination of twin weight ratios; and determination of mono- or diamniotic gestation.

To interrupt the vascular connection between the twins and promote survival of the pump twin, various treatment methods have been used, including hysterotomy and selective removal of the anomalous twin. Fetoscopic
occlusion of the anastomotic vessels using ultrasound-guided embolization, ligation of the umbilical cord (e.g., laser photocoagulation) or radiofrequency cord ablation, have been described in the literature. Selective removal and embolization have been associated with high morbidity and unreliable outcomes. Radiofrequency ablation, a newer technique, is less invasive compared to photocoagulation and some fetal specialty surgery centers have had promising results using this technique (Lee, et al., 2007; Livingston, et al., 2007). The results of some studies indicate outcomes are improved with umbilical-cord laser photocoagulation. During this procedure, the umbilical cord root to the affected fetus is coagulated. Quintero and colleagues (2006) reported, and other authors agree, that the surgical approach and technique for treatment of TRAP sequence should be tailored to the specific clinical presentation.

Evidence in the medical literature evaluating treatment for TRAP sequence is limited and consists mostly of retrospective cohort, reviews, case reports, case series involving small populations, and registry data (Ville, et al., 1994; Weisz, et al., 2004, Quintero, et al., 2006, Cabassa, et al., 2012; Lee, et al., 2013; Pagani, et al., 2013). The results of these studies however do support improved perinatal survival and favorable clinical outcomes.

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. In as many as 90% of all fetuses diagnosed with urinary-tract dilatation, intervention is not required (Harrison, 1996). 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. Pulmonary hypoplasia is fatal, and infants with this condition usually die shortly after birth (Harrison, 1996). Some authors have investigated endoscopic surgery (i.e., fetoscopic cystoscopy with laser) to visualize the posterior urethral valves, however, the data is limited, and further studies are needed to support safety and efficacy. 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. In the event that the shunt becomes displaced, or if it cannot be inserted, and if the fetus is at less than 22 weeks of gestation, the authors recommend creating a surgical opening in the bladder (vesicostomy). Fetuses with severe renal damage are not considered candidates for this procedure as it is not clear whether decompression can reverse the renal damage.

Evidence demonstrating that early surgical intervention results in improved survival is mainly in the form of small case series (Freedman, et. al., 1999; Johnson, et al., 1994) with few randomized trials. Early surgical intervention is not curative, further evaluation and surgical treatment following delivery are necessary (Wu and Johnson, 2009). Morris et al. (2013) published the results of a RCT (n=31) involving pregnancies complicated by isolated fetal lower urinary tract obstruction with treatment by either conservative management (n=15) or vesicoamniotic shunt placement (n=16) to assess the effect of treatment on survival. Improved survival was associated with renal morbidity. The primary outcome of the trial was survival of the baby to 28 days postnatal. Twelve live births occurred in each group with eight of the shunt group babies and four of the conservative group babies surviving to 28 days, the difference was not statistically significant. At one year follow-up one baby subsequently died in each subgroup; overall the outcomes were poor with only two babies, both from the shunt group, surviving with normal renal function. In the authors opinion by the time of diagnosis renal damage may have already taken place and may have been be irreversible.

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.

Resection of CCAM reverses hydrops and improves survival (Adzick, 2009; Adzick, et al., 2003, Adzick, et al., 1998). Treatment for a fetus with fetal hydrops and a large multicystic lesion involves resecting the large, cystic pulmonary lobe (lobectomy). A single large cyst may be treated by means of a thoracoamniotic shunt. Thoracoamniotic shunting appears to be beneficial in preventing lung hypoplasia in affected fetuses with CCAM (Morikawa, et. al., 2003). Fetal thoracentesis alone is minimally effective for treatment because cystic fluid reaccumulates; nonetheless, the procedure is often performed prior to resection or shunting. Catheter shunt placement has improved neonatal outcomes in some clinical studies. Other treatment options are to terminate the pregnancy or to continue observation.

**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. No diagnostic landmarks have been found that can identify intralobar sequestration. 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).

**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.

Additional methods that have been proposed for treating SCT have involved the use of laser ablation, radiofrequency ablation, and thermocoagulation. In laser ablation, the vessels leading to the tumor are ablated with the use of a laser. Radiofrequency ablation employs radiofrequency energy for the same purpose; this technique may be performed under ultrasound guidance with minimal access. In thermocoagulation, another minimal-access method, an insulated wire is passed through a needle into the SCT, heating the vessels until blood flow diminishes. Authors propose coagulating the vessels decreases the blood supply to the tumor, decreases cardiovascular demand, and ultimately reverses the fetal hydrops. While minimal access techniques may reduce complications (e.g., preterm labor, premature rupture of membranes) that are often associated with more invasive techniques, these techniques do not support superior outcomes compared to those for percutaneous drainage and open resection (Hirose and Farmer, 2003).

Although there are few published clinical trials, it has been proposed that in-utero resection may reverse the physiologic effects of the tumor and improve fetal survival in a previable fetus. Surgical resection in cases with evidence of fetal hydrops, placentomegaly, and gestational age prior to 32 weeks has shown favorable outcomes compared to cases with untreated fetal hydrops (Hedrick, et al., 2004).

**Aqueductal Stenosis (Hydrocephalus)**
Stenosis of the aqueduct of Sylvius leads to congenital hydrocephalus. The aqueduct of Sylvius is a space that connects the third and fourth ventricles of the brain and allows for flow of cerebrospinal fluid. Obstruction of the flow dilates the ventricles and leads to compression of the brain, eventually compromising brain function. When hydrocephalus is diagnosed, the treatment options include termination or continuation of the pregnancy with monitoring for progression of the disease and detection of additional anomalies. Traditionally, the condition is detected and then treated after birth with a shunt procedure. Researchers suggest that decompressing the ventricles may prevent adverse effects on the developing brain, although in-utero treatment with ventriculoamniotic shunts has not led to improved perinatal outcomes.

If isolated hydrocephalus occurs, it is followed with serial ultrasounds because with increasing length of gestation, the outcome is variable and worsening developmental outcomes may result. Nonetheless, outcomes after early shunting and delivery have been poor; hence, such treatment is not recommended until 32 weeks of gestation.

A moratorium, initially implemented at the third annual meeting of the International Fetal Medicine and Surgery Society in 1985, still remains in effect on percutaneous shunting for fetal hydrocephalus.

**Congenital Diaphragmatic Hernia (CDH)**

Congenital diaphragmatic hernia (CDH) is a condition that results in abdominal viscera entering the chest cavity through an opening, or hernia, in the diaphragm. It frequently results in pulmonary hypoplasia and pulmonary hypertension; outcomes can vary widely, however, depending on the size of the hernia and the timing of herniation. Prognosis depends on the degree of liver herniation, the presence or absence of other anomalies, and the lung-to-head ratio. Although the condition is correctable after birth, most babies die because of underdeveloped lungs.

In cases without liver herniation, in-utero correction involves reduction of the viscera, reconstruction of the diaphragm, and enlargement of the abdomen to accept the herniated organs. The surgical correction performed on a fetus with liver herniation involves temporary occlusion of the fetal trachea to expand the lungs, thus displacing the viscera back into the abdomen and hastening fetal lung growth. At birth, the tracheal occlusion is then reversed, and the hernia is repaired.

The goal of fetal intervention for CDH is to prevent or reverse hypoplasia and restore adequate lung growth. Three surgical approaches have been attempted in the human fetus for CDH and include: open tracheal clipping, application of a tracheal clip using the fetal endoscopic approach (FETENDO clip), and tracheal balloon occlusion (Arca and Teich, 2004). Reported survival rates for CDH vary widely. Open fetal surgery has failed to demonstrate any advantage and is high risk to both mother and fetus. The use of balloons, sponges or clips generally results in larger but abnormal lungs (Chung, 2012).

Evidence in the published literature evaluating approaches for treatment of CDH is lacking and does not lend strong support to improved patient outcomes; the effectiveness of treating CDH has not been firmly established. Tracheal occlusion did not improve survival or decrease morbidity in a cohort of fetuses with CDH when compared to standard postnatal care (Harrison, et al., 2003). Lack of improved survival rates in some published studies (Flake, et al., 2000; Harrison, et al., 2003) and lack of proven effectiveness (Sydorak and Harrison, 2003) have also been reported in the medical literature.

Minimally invasive approaches using smaller instrumentation are now being investigated. A randomized controlled trial published by Ruano and colleagues (2012) evaluated a minimal access approach using fetal endoscopic tracheal occlusion (FETO) for fetuses with severe CDH; subjects were randomized to undergo a minimal approach (FETO) (n=20) or to undergo standard postnatal management (n=21). With the FETO technique a smaller diameter fetoscope was placed percutaneously under ultrasound guidance. The authors hypothesized a smaller diameter fetoscope would result in less complications and improved perinatal survival rates. Fetuses who underwent FETO were delivered by ex-utero intrapartum therapy (EXIT); 14 were planned at an average of 37-38 weeks, four were delivered at 34-36 weeks after premature rupture of membranes and five were emergent due to preterm contractions. Fetuses in the control group were delivered by Cesarean section; 15 were planned at 38 weeks gestation and four were emergent. Balloon removal occurred at the time of delivery. The authors noted current guidelines recommend removal 6 weeks following the initial placement to improve neonatal outcome. In the intention-to-treat analysis 10 of 20 infants in the FETO group survived, while 1 of 21 infants in the control group survived. The authors noted that although fetal survival was improved with
FETO and the use of a smaller fetoscope, the procedure was associated with risk of prematurity and that the occurrence of preterm premature rupture of membranes was high.

Fetal mortality and morbidity from CDH remain high; premature delivery, which is detrimental to the fetus, continues to be associated with intervention, regardless of whether open or endoscopic approaches are used (Arca and Teich, 2004).

**Amniotic Band Syndrome (ABS)**

Amniotic band syndrome (ABS) also referred to as amnion disruption sequence, constriction ring syndrome or annular constriction rings, is an abnormality that occurs in approximately 1:1200 to 15,000 live births. The exact cause is unknown; however, authors have proposed that early rupture of the amnion without damage to the chorion sac results in oligohydramnios and formation of amniotic bands; oligohydramnios results in abnormal pressure on the fetal distal extremities. Amniotic bands may result in ring constrictions, limb autoamputations, pseudosyndactylysm, and other fetal defects. In many cases, ABS is associated with congenital anomalies that are beyond surgical repair, although some cases may result in the isolated constriction of an extremity without amputation. Isolated extremity ABS is not a life-threatening condition (Keswani, et al., 2003). There is currently no effective treatment for ABS, and reconstructive surgery is typically performed in the postnatal period. According to the literature, bands may be snipped after birth, and Z-plasty may be performed on the affected limb. Surgical release of the bands in-utero has been proposed by some authors to avoid amputation or permanent damage to the extremity. Nevertheless, histologic changes, neurological paresis, contractures or hypoplasia persist despite surgical release. Attempts at identifying patient selection criteria for in-utero surgery are currently being investigated, however at present there is no prenatal classification available (Husler, et al, 2009).

The evidence in the peer-reviewed scientific literature consists mainly of case reports and is insufficient to support improved patient outcomes with in-utero surgical release of amniotic bands. The reported clinical outcomes vary and include salvage of an intact limb, a viable extremity with limited function, and a grossly deformed extremity requiring postnatal amputation (Keswani, et al., 2003). Ronderos-Dumit et al. (2006) reported on a case of constriction amniotic bands involving both legs of a fetus with compromising blood flow to the distal extremity. The constriction ring was successfully released in-utero, although the baby underwent Z-plasty of the compromised leg in the postnatal period. While successful lyses of amniotic bands have been reported, further clinical trials are warranted to support the benefit of in-utero surgical release and the avoidance of limb dysfunction.

**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. Congenital hydrothorax is a rare disorder and is defined by the accumulation of fluid in the pleural cavity. Congenital chylothorax, defined as accumulation of chyle in the thoracic cavity, is also a frequent cause of pleural effusions (Harrison, Adzick, 1991). The persistence of pleural effusion in early pregnancy interferes with normal lung development and often results in pulmonary hypoplasia. Mediastinal compression resulting from effusion can cause hemodynamic compromise leading to fetal hydrops and perinatal death. Prenatal intervention is dependent on the severity of fluid accumulation and the gestational age of the fetus at the time of diagnosis. In some cases, spontaneous resolution occurs and no intervention other than observation is indicated. Poor outcomes are generally associated with isolated hydrothorax, and neonatal death rates vary from 55% when diagnosis is made prior to 32 weeks’ gestation to 30% when the diagnose is made later (Prontera, et al., 2002). When the condition is associated with hydrops, mortality rates approach 100%. 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. Authors agree when diagnosed in early pregnancy (i.e., prior to 32 weeks) the initial treatment of choice is thoracentesis; however, most effusions reaccumulate and often cause fetal hydrops. When reaccumulation of fluid occurs, shunting is recommended. When hydrothorax is diagnosed later in pregnancy (close to term), the treatment is ultrasound-guided thoracentesis or transthoracic puncture immediately after birth.

Successful placement of pleuro-amniotic shunts is supported in the published scientific literature (Roche, et al., 2006; Smith, et al., 2005; Wilson, et al., 2004; Nicolaides and Azar, 1990). Published evidence is however limited to case reports (Hamada, et al, 2001), small case series and retrospective reviews and the indications for pleuro-amniotic shunting are not well-defined. Nevertheless, authors agree the presence of fetal hydrothorax-
induced hydrops or polyhydramnios are indications for shunting. Some authors have recommended shunting for primary fetal hydrothorax with evidence of effusion under tension, even without hydrops.

Miscellaneous Conditions
In-utero fetal surgery has been performed for correction of other fetal abnormalities, such as complete heart block (open or percutaneous placement of pacemaker), treatment of hypoplastic left heart syndrome (laser atrial septotomy), pulmonary-aortic obstruction (percutaneous placement of a balloon catheter to open the stenotic heart valve [i.e., balloon valvuloplasty procedures]), tracheal-atriesia stenosis (fetal tracheostomy), cleft lip and palate (in-utero correction to avoid scarring), and fetal stem-cell transplantation for related stem-cell disease (to decrease fetal rejection and need for immuno-suppression). In addition, some authors have investigated in-utero gene therapy for disorders that result in irreversible illness or death in the pre- or neonatal period (e.g., Type 2 Gaucher's Disease, Krabbe's disease, Hurler's Disease). Several concerns exist with in-utero gene therapy regarding safety and efficacy, and further clinical investigation is necessary to support improved patient outcomes. Presently, in-utero gene therapy is not an established treatment modality. Evidence in the published, peer-reviewed scientific literature is inadequate to support improved perinatal outcomes with the use of in-utero fetal surgery to treat these conditions.

Use Outside of the US: In utero fetal surgery has been performed in countries outside the U.S. and is generally regulated by professional societies/organizations similar to those of the U.S. While the conditions for which this type of surgery is being performed vary, it is recommended the procedures be performed in centers specializing in invasive fetal medicine. The National Institute of Health and Care Excellence (NICE) has developed guidelines regarding performance of in utero surgery to treat some fetal anomalies, such as pulmonary atresia, aortic stenosis and twin to twin transfusion syndrome. According to these guidelines, percutaneous fetal balloon valvuloplasty for pulmonary atresia or aortic stenosis have not been proven safe and effective; intrauterine laser ablation of placental vessels for treatment of twin to twin transfusion syndrome has been proven safe and effective.

Summary
Fetal surgery has become an acceptable option for treatment of some congenital anomalies. Evidence in the published peer-reviewed scientific literature and professional society position support improved clinical outcomes and perinatal survival for conditions such as myelomeningocele repair, twin to twin transfusion, and bilateral urinary tract obstruction, to name a few. Early detection, treatment and monitoring involve a multidisciplinary approach typically available at experienced fetal surgery centers. Nonetheless there remains significant risk to both fetus and mother and rigorous patient selection is important. For many conditions data is lacking; well-designed and controlled clinical trials evaluating in-utero intervention for those conditions compared to postnatal intervention are required to support improved survival rates.

Coding/Billing Information

Note: 1) This list of codes may not be all-inclusive.
2) Deleted codes and codes which are not effective at the time the service is rendered may not be eligible for reimbursement.

Myelomeningocele Repair
Covered when medically necessary when used to report myelomeningocele repair:

<table>
<thead>
<tr>
<th>CPT® Codes</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>59897</td>
<td>Unlisted fetal invasive procedure, including ultrasound guidance, when performed</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>HCPCS Codes</th>
<th>Description</th>
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</thead>
<tbody>
<tr>
<td>S2404</td>
<td>Repair, myelomeningocele in the fetus, procedure performed in utero</td>
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</table>
Nonselective or Selective Fetoscopic Laser Coagulation for Twin to Twin Transfusion Syndrome

Covered when medically necessary:

<table>
<thead>
<tr>
<th>CPT® Codes</th>
<th>Description</th>
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<tbody>
<tr>
<td>59001</td>
<td>Amniocentesis; therapeutic amniotic fluid reduction (includes ultrasound guidance)</td>
</tr>
<tr>
<td>59897†</td>
<td>Unlisted fetal invasive procedure, including ultrasound guidance, when performed</td>
</tr>
</tbody>
</table>

†Note: Covered when used to report fetoscopic laser therapy for the treatment of twin-to-twin syndrome.

<table>
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<th>HCPCS Codes</th>
<th>Description</th>
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<tbody>
<tr>
<td>S2411</td>
<td>Fetoscopic laser therapy for treatment of twin-to-twin transfusion syndrome</td>
</tr>
</tbody>
</table>

Fetoscopic Occlusion of Anastomotic Vessels (e.g., laser photocoagulation, radiofrequency ablation, ligation) for Twin Reversed Arterial Perfusion

Covered when medically necessary:

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<tr>
<th>CPT® Codes</th>
<th>Description</th>
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<tbody>
<tr>
<td>59072</td>
<td>Fetal umbilical cord occlusion, including ultrasound guidance</td>
</tr>
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</table>

Fetal Vesicoamniotic Shunt Procedures for Bilateral Fetal Urinary Tract Obstruction

Covered when medically necessary:

<table>
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<tr>
<th>CPT® Codes</th>
<th>Description</th>
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<tbody>
<tr>
<td>59076</td>
<td>Fetal shunt placement, including ultrasound guidance</td>
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<table>
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<tr>
<th>HCPCS Codes</th>
<th>Description</th>
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</thead>
<tbody>
<tr>
<td>S2401</td>
<td>Repair, urinary tract obstruction in the fetus, procedure performed in utero</td>
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Fetal Lobectomy for Congenital Cystic Adenomatoid Formation (CCAM)/Congenital Pulmonary Airway Malformation (CPAM)

Covered when medically necessary when used to report fetal lobectomy:

<table>
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<tr>
<th>CPT® Codes</th>
<th>Description</th>
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<tbody>
<tr>
<td>59897</td>
<td>Unlisted fetal invasive procedure, including ultrasound guidance, when performed</td>
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<table>
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<tr>
<th>HCPCS Codes</th>
<th>Description</th>
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<tbody>
<tr>
<td>S2402</td>
<td>Repair, congenital cystic adenomatoid malformation in the fetus, procedure performed in utero</td>
</tr>
<tr>
<td>S2409*</td>
<td>Repair, congenital malformation of fetus, procedure performed in utero, not otherwise classified</td>
</tr>
</tbody>
</table>

*Note: Covered as medically necessary when used to report fetal lobectomy.
Fetal Thoracoamniotic Shunt Placement

Covered when medically necessary:

<table>
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<tbody>
<tr>
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<tbody>
<tr>
<td>S2403</td>
<td>Repair, extralobar pulmonary sequestration in the fetus, procedure performed in utero</td>
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</table>

In-utero Needle Access and Open Resection for Sacrococcygeal Teratoma

Covered when medically necessary:

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<th>HCPCS Codes</th>
<th>Description</th>
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<tbody>
<tr>
<td>S2405</td>
<td>Repair of sacrococcygeal teratoma in the fetus, procedure performed in utero</td>
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</tbody>
</table>

Experimental/Investigational/Unproven and Not Covered when used to report any procedure listed as such in this policy including, but not limited to: laser, thermocoagulation or radiofrequency ablation techniques for the treatment of sacrococcygeal teratoma or endoscopic approach (i.e., cystoscopy) for the treatment of lower urinary tract obstruction:

<table>
<thead>
<tr>
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<th>Description</th>
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</thead>
<tbody>
<tr>
<td>59897</td>
<td>Unlisted fetal invasive procedure, including ultrasound guidance, when performed</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>HCPCS Codes</th>
<th>Description</th>
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</thead>
<tbody>
<tr>
<td>S2400</td>
<td>Repair, congenital diaphragmatic hernia in the fetus using temporary tracheal occlusion, procedure performed in utero</td>
</tr>
<tr>
<td>S2409</td>
<td>Repair, congenital malformation of fetus, procedure performed in utero, not otherwise classified</td>
</tr>
</tbody>
</table>


References


73. Quintero RA; Chmait RH; Murakoshi T; Pankrac Z; Światkowska M; Bornick PW; Allen MH. Surgical management of twin reversed arterial perfusion sequence. Am J Obstet Gynecol. 2006 Apr;194(4):982-91.


