



# CIGNA MEDICAL COVERAGE POLICY

The following Coverage Policy applies to all health benefit plans administered by CIGNA Companies including plans formerly administered by Great-West Healthcare, which is now a part of CIGNA.

Effective Date ..... 7/15/2011  
Next Review Date ..... 7/15/2012  
Coverage Policy Number ..... 0175

Subject **Fetal Surgery**

## Table of Contents

Coverage Policy .....	1
General Background .....	3
Coding/Billing Information .....	10
References .....	13
Policy History.....	19

## Hyperlink to Related Coverage Policies

Ultrasound in Pregnancy (Including 3D and 4D Ultrasound)

### INSTRUCTIONS FOR USE

Coverage Policies are intended to provide guidance in interpreting certain **standard** CIGNA HealthCare benefit plans. Please note, the terms of a customer's particular benefit plan document [Group Service Agreement (GSA), Evidence of Coverage, Certificate of Coverage, Summary Plan Description (SPD) or similar plan document] may differ significantly from the standard benefit plans upon which these Coverage Policies are based. For example, a customer's benefit plan document may contain a specific exclusion related to a topic addressed in a Coverage Policy. In the event of a conflict, a customer's benefit plan document **always supercedes** the information in the Coverage Policies. In the absence of a controlling federal or state coverage mandate, benefits are ultimately determined by the terms of the applicable benefit plan document. Coverage determinations in each specific instance require consideration of 1) the terms of the applicable benefit plan document in effect on the date of service; 2) any applicable laws/regulations; 3) any relevant collateral source materials including Coverage Policies and; 4) the specific facts of the particular situation. Coverage Policies relate exclusively to the administration of health benefit plans. Coverage Policies are not recommendations for treatment and should never be used as treatment guidelines. In certain markets, delegated vendor guidelines may be used to support medical necessity and other coverage determinations. Proprietary information of CIGNA. Copyright ©2011 CIGNA

## Coverage Policy

**CIGNA covers fetal surgery for myelomeningocele repair as medically necessary 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  $\geq 19.0$  and  $< 26$  weeks
- normal fetal karyotype
- absence of ALL of the following:
  - fetal anomaly unrelated to the myelomeningocele
  - severe fetal kyphosis
  - short cervix (less than or equal to 15 mm)
  - previous pre-term birth
  - placental abruption
  - maternal Body Mass Index (BMI) greater than or equal to 35
  - contraindications to surgery, including but not limited to previous hysterotomy in the active (upper) uterine segment

**CIGNA covers fetal surgery using nonselective or selective fetoscopic laser coagulation as medically necessary for the treatment of 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
- evidence of abnormal blood flow documented by Doppler studies in one or both fetuses

**CIGNA covers serial amnioreduction for TTTS as medically necessary when ALL of the following criteria are met:**

- evidence of polyhydramnios in the recipient fetus
- donor fetus is oligohydramniotic
- evidence of abnormal blood flow documented by Doppler studies in one or both fetuses

**CIGNA covers fetoscopic occlusion of anastomotic vessels (e.g., laser photocoagulation, radiofrequency ablation, ligation) for twin reversed arterial perfusion (TRAP sequence) as medically necessary when BOTH of the following criteria are met:**

- acardiac to pump twin weight ratio is greater than 50%.
- evaluation is performed using umbilical cord Doppler velocimetry and fetal echocardiography to assess strain on the pump twin's heart and to document reversed flow.

**CIGNA covers fetal vesicoamniotic shunt procedures as medically necessary for the treatment of bilateral fetal urinary-tract obstruction when BOTH of the following criteria are met:**

- evidence of early-onset oligohydramnios
- evidence of adequate renal function as indicated by fetal urine studies and ultrasound

**CIGNA covers fetal lobectomy as medically necessary for the treatment of 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 covers fetal thoracoamniotic shunt placement as medically necessary for ANY of the following conditions:**

- treatment of CCAM when BOTH of the following criteria are met:
  - evidence of fetal hydrops
  - presence of one lesion, consisting of a single large cyst.
- treatment of extralobar pulmonary sequestration (EPS) when there is evidence of tension hydrothorax and/or fetal hydrops.
- treatment of fetal pleural effusion when ALL of the following criteria are met:
  - fetal gestational age is 32 weeks or less.
  - failure, contraindication or intolerance to fetal thoracentesis
  - evidence of fetal hydrops and/or pulmonary hypoplasia

**CIGNA covers in-utero needle access and open resection of sacrococcygeal teratoma as medically necessary for a previable fetus with or without early maternal pre-eclampsia, when there is evidence of fetal hydrops, placentomegaly or polyhydramnios.**

**CIGNA does not cover EITHER of the following in-utero surgical procedures because they are 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., cystoscopy) for the treatment of lower urinary tract obstruction

**CIGNA does not cover in-utero fetal surgery as a treatment for the following medical conditions because it is considered experimental, investigational or unproven for these indications (this list may not be all-inclusive):**

- 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 and chromosomal anomalies that may jeopardize fetal survival in addition to maternal mirror syndrome. Fetal hydrops is a condition in which the fetus develops subcutaneous-tissue edema, accompanied by serous effusions into one or more body cavities. It may present as serous fluid build-up in the skin, pleura, pericardium, placenta, peritoneum, or amniotic fluid. The condition frequently results in death in-utero or shortly after birth.

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. Because of 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) and reduction in shunt-dependent hydrocephalus has also been reported (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.

In 2003 Johnson and associates performed a retrospective review to compare the short-term clinical outcomes of 50 fetuses that had undergone open fetal surgery for myelomeningocele repair. Overall perinatal survival was 94% (47/50). Reversal of hindbrain herniation occurred in all fetuses. Ventriculoperitoneal 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.

Johnson et al (2006) studied 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.

Until recently, fetal surgery for myelomeningocele repair was available only through a clinical trial. The Management of Myelomeningocele Study (MOMS) was a randomized research study, funded by The National Institute of Child Health and Human Development (NICHD), part of the National Institutes of Health (NIH). The study, which began recruiting in 2003, sought to compare the results of prenatal treatment of spina bifida versus postnatal 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. The trial 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 criteria consisted of singleton pregnancy, myelomeningocele with upper boundary located between T1 and S1, evidence of hindbrain herniation, a gestational age of 19.0 to 25.9 weeks at randomization, a normal karyotype, U.S. residency, and maternal age of at least 18 years. Exclusion criteria included unrelated fetal anomaly, severe kyphosis, risk of preterm birth (including short cervix and previous preterm birth), placental abruption, a body-mass index of 35 or more, and contraindication to surgery, including previous hysterotomy in the active uterine segment. 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).

### **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, oligouria 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 (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.

### **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.

Published studies in the medical literature evaluating treatment for TRAP sequence is limited and consists mostly of case series involving small populations (Ville, et al., 1994; Weisz, et al., 2004, Quintero, et al., 2006). 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 patient selection criteria for intervention are based upon fetal-urine electrolyte studies, beta<sup>2</sup>-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. However, bilateral urinary obstruction in the fetus is often associated with serious adverse outcomes, such as pulmonary hypoplasia secondary to oligohydramnios (decrease in amniotic fluid). Pulmonary hypoplasia is fatal, and infants with this condition usually die shortly after birth (Harrison, 1996). Some authors have investigated endoscopic surgery (i.e., cystoscopy) to visualize and treat the urinary obstruction; 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). Surgery is not curative and further evaluation and surgical treatment are necessary following delivery (Wu and Johnson, 2009).

### **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).

### **Sacroccygeal Teratoma (SCT)**

A sacroccygeal 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 pre-viable 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 (Warner, 2004).

Evidence in the published literature evaluating these approaches is lacking and does not lend 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 (Flake, et al., 2000; Harrison, et al., 2003) and lack of proven effectiveness (Sydorak and Harrison, 2003) have been reported in the medical literature. 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, pseudosyndactylism, 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; Nicolaidis 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-atresia 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.

### Summary

Fetal surgery has become a viable option for some specific congenital anomalies. In some cases, early detection may allow prevention or reversal of detrimental outcomes. However, further well-designed and controlled clinical trials evaluating in-utero intervention compared to postnatal intervention are required to support long-term outcomes and improved survival rates.

## Coding/Billing Information

**Note:** This list of codes may not be all-inclusive.

**Covered when medically necessary:**

### Myelomeningocele Repair

CPT <sup>®</sup> * Codes	Description
59897	Unlisted fetal invasive procedure, including ultrasound guidance, when performed

HCPCS Codes	Description
S2404	Repair, myelomeningocele in the fetus, procedure performed in utero

ICD-9-CM Diagnosis Codes	Description
653.70- 653.73	Other fetal abnormality causing disproportion
655.03	Central nervous system malformation in fetus, unspecified as to episode of care in pregnancy
741.00- 741.03	Spina bifida with hydrocephalus
741.90- 741.93	Spina bifida without mention of hydrocephalus

### Twin to Twin Transfusion Syndrome

<b>CPT<sup>®*</sup> Codes</b>	<b>Description</b>
59001	Amniocentesis; therapeutic amniotic fluid reduction (includes ultrasound guidance)
59897 <sup>†</sup>	Unlisted fetal invasive procedure, including ultrasound guidance, when performed

<sup>†</sup>**Note:** Covered when used to report fetoscopic laser therapy for the treatment of twin-to-twin syndrome.

<b>HCPCS Codes</b>	<b>Description</b>
S2411	Fetoscopic laser therapy for treatment of twin-to-twin transfusion syndrome

<b>ICD-9-CM Diagnosis Codes</b>	<b>Description</b>
651.03	Twin pregnancy; antepartum condition or complication
657.03	Polyhydramnios, antepartum complication
658.03	Oligohydramnios, antepartum
678.03	Fetal hematologic conditions, antepartum condition or complication
762.3	Fetus or newborn affected by placental transfusion syndromes

### Twin Reversed Arterial Perfusion

<b>CPT<sup>®*</sup> Codes</b>	<b>Description</b>
59072	Fetal umbilical cord occlusion, including ultrasound guidance

<b>ICD-9-CM Diagnosis Codes</b>	<b>Description</b>
762.3	Fetus or newborn affected by placental transfusion syndromes

### Bilateral Fetal Urinary Tract Obstruction

<b>CPT<sup>®*</sup> Codes</b>	<b>Description</b>
59076	Fetal shunt placement, including ultrasound guidance

<b>HCPCS Codes</b>	<b>Description</b>
S2401	Repair, urinary tract obstruction in the fetus, procedure performed in utero

<b>ICD-9-CM Diagnosis Codes</b>	<b>Description</b>
658.03	Oligohydramnios, antepartum
753.6	Congenital atresia and stenosis of urethra and bladder neck

### Congenital Cystic Adenomatoid Formation (CCAM)/Congenital Pulmonary Airway Malformation (CPAM)

<b>CPT<sup>®*</sup></b>	<b>Description</b>
-------------------------	--------------------

<b>Codes</b>	
59897	Unlisted fetal invasive procedure, including ultrasound guidance, when performed

<b>HCPCS Codes</b>	<b>Description</b>
S2402	Repair, congenital cystic adenomatoid malformation in the fetus, procedure performed in utero
S2409 <sup>†</sup>	Repair, congenital malformation of fetus, procedure performed in utero, not otherwise classified

<sup>†</sup>**Note:** Covered as medically necessary when used to report fetal lobectomy.

<b>ICD-9-CM Diagnosis Codes</b>	<b>Description</b>
653.70-653.73	Other fetal abnormality causing disproportion
773.3	Hydrops fetalis due to isoimmunization
778.0	Hydrops fetalis not due to isoimmunization

### **Thoracoamniotic Shunt Placement**

<b>CPT<sup>®*</sup> Codes</b>	<b>Description</b>
59076	Fetal shunt placement, including ultrasound guidance

<b>HCPCS Codes</b>	<b>Description</b>
S2403	Repair, extralobar pulmonary sequestration in the fetus, procedure performed in utero

<b>ICD-9-CM Diagnosis Codes</b>	<b>Description</b>
653.70-653.73	Other fetal abnormality causing disproportion
748.5	Congenital agenesis, hypoplasia, and dysplasia of lung
773.3	Hydrops fetalis due to isoimmunization
778.0	Hydrops fetalis not due to isoimmunization

### **Sacroccygeal Teratoma**

<b>HCPCS Codes</b>	<b>Description</b>
S2405	Repair of sacroccygeal teratoma in the fetus, procedure performed in utero

<b>ICD-9-CM Diagnosis Codes</b>	<b>Description</b>
653.70-653.73	Other fetal abnormality causing disproportion
657.03	Polyhydramnios, antepartum complication
773.3	Hydrops fetalis due to isoimmunization
778.0	Hydrops fetalis not due to isoimmunization

**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:**

<b>CPT®*</b> <b>Codes</b>	<b>Description</b>
59897	Unlisted fetal invasive procedure, including ultrasound guidance, when performed

<b>HCPCS</b> <b>Codes</b>	<b>Description</b>
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

<b>ICD-9-CM</b> <b>Diagnosis</b> <b>Codes</b>	<b>Description</b>
742.3	Congenital hydrocephalus
745.0- 745.9	Bulbus cordis anomalies and anomalies of cardiac septal closure
746.00-746.9	Other congenital anomalies of heart
749.10- 749.14	Cleft lip
749.20- 749.25	Cleft palate with cleft lip
756.6	Congenital anomaly of diaphragm

**\*Current Procedural Terminology (CPT®) © 2010 American Medical Association: Chicago, IL.**

## References

1. Adzick NS, Harrison MR, Crombleholme TM, Flake AW, Howell LJ. Fetal lung lesions: management and outcome. *Am J Obstet Gynecol.* 1998 Oct;179(4):884-9.
2. Adzick NS. Management of fetal lung lesions. *Clin Perinatol.* 2003 Sep 1;30(3):481-92.
3. Adzick NS. Management of fetal lung lesions. *Clin Perinatol.* 2009 Jun;36(2):363-76, x.
4. Arca MJ, Teich S. Current controversies in perinatal care: fetal versus neonatal surgery. *Clin Perinatol.* 2004 Sep;31(3):629-48.
5. Adzick NS, Thom EA, Spong CY, Brock JW 3rd, Burrows PK, Johnson MP, Howell LJ, Farrell JA, Dabrowiak ME, Sutton LN, Gupta N, Tulipan NB, D'Alton ME, Farmer DL; MOMS Investigators. A randomized trial of prenatal versus postnatal repair of myelomeningocele. *N Engl J Med.* 2011 Mar 17;364(11):993-1004. Epub 2011 Feb 9.
6. Berhman: Nelson Textbook of Pediatrics, 17<sup>th</sup> ed. Fetal Hydrops. Clinical Manifestations. p. 602. ©Elsevier 2004.
7. Bruner JP. Maternal-fetal surgery. *Clin Perinatol.* 2003 Sep 1;30(3):xiii-xvi.
8. Bruner JP, Davis G, Tulipan N. Intrauterine shunt for obstructive hydrocephalus--still not ready. *Fetal Diagn Ther.* 2006;21(6):532-9. Epub 2006 Sep 12.

9. Bruner JP, Tulipan N, Reed G, Havis GH, Bennett K, Luker K, Dabrowiak ME. Intrauterine repair of spina-bifida: Preoperative predictors of shunt-dependent hydrocephalus. *Am J Obstet Gynecol.* 2004 May;190(5):1305-12.
10. Bussey JG, Luks F, Carr SR, Plevyak M, Tracy TF Jr. Minimal-access fetal surgery for twin-to-twin transfusion syndrome. *Surg Endosc.* 2004 Jan 1;18(1):93-6.
11. Carr MC. Prenatal management of urogenital disorders. *Urol Clin North Am.* 2004 Aug;31(3):389-97,vii
12. Childrens Hospital of Philadelphia (CHOP). Center for Fetal Diagnosis and Treatment. Accessed June 25, 2010. Available at URL address: <http://www.fetalsurgery.chop.edu/ftldiag7.shtml>
13. Cincotta RB, Gray PH, Gardener G, Soong B, Chan FY. Selective fetoscopic laser ablation in 100 consecutive pregnancies with severe twin-twin transfusion syndrome. *Aust N Z J Obstet Gynaecol.* 2009 Feb;49(1):22-7.
14. Crombleholme TM, Shera D, Lee H, Johnson M, D'Alton M, Porter F, et al. A prospective, randomized, multicenter trial of amnioreduction vs selective fetoscopic laser photocoagulation for the treatment of severe twin-twin transfusion syndrome. *Am J Obstet Gynecol.* 2007 Oct;197(4):396.e1-9.
15. Crombleholme TM, Harrison MR, Golbus MS, Longaker MT, Langer JC, Callen PW, Anderson RL, Goldstein RB, Filly RA. Fetal intervention in obstructive uropathy: prognostic indicators and efficacy of intervention. *Am J Obstet Gynecol.* 1990 May;162(5):1239-44.
16. Davis GH. Fetal hydrocephalus. *Clin Perinatol.* 2003 Sep 1;30(3):531-9.
17. Department of Health. Gene Therapy Advisory Committee. Report on potential uses of gene therapy in utero. London, UK: Department of Health; November 1998. Updated July 17, 2002. Accessed June 25, 2010. Available at: <http://www.advisorybodies.doh.gov.uk/genetics/gtac/inutero.htm>
18. Deprest J, Jani J, Gratacos E, Vandecruys H, Naulaers G, Delgado J, Greenough A, Nicolaidis K; FETO Task Group. Fetal intervention for congenital diaphragmatic hernia: the European experience. *Semin Perinatol.* 2005 Apr;29(2):94-103.
19. Fetal Care Center of Cincinnati. Cincinnati Children's Hospital Medical Center. Fetal surgery for pleural effusions and chest cysts. ©2004-2009, Fetal Care Center of Cincinnati. Accessed June 25, 2010 Available at URLA address: <http://www.fetalcarecenter.org/conditions/pleural-effusions/default.htm>
20. Findik H, Malkoc C, Uzunismail A. Long-term effects of amniotic bands not treated at an early age. *Plast Reconstr Surg.* 2006 Feb;117(2):713-4.
21. Fisk NM, Galea P. Twin-twin transfusion-as good as it gets? *N Engl J Med.* 2004 Jul 8;351(2):182-184.
22. Flake AW, Crombleholme TM, Johnson MP, Howell LJ, Adzick NS. Treatment of severe congenital diaphragmatic hernia by fetal tracheal occlusion: clinical experience with fifteen cases. *Am J Obstet Gynecol.* 2000 Nov;18(5):1059-66.
23. Freedman AL, Johnson MP, Smith CA, Gonzalez R, Evans MI. Long-term outcome in children after antenatal intervention for obstructive uropathies. *Lancet.* 1999 Jul 31;354(9176):374-7.
24. Gabbe: *Obstetrics-Normal and Problem Pregnancies*, 4<sup>th</sup> edition. ©2002 Churchill Livingstone, Inc. p. 300.
25. Graef C, Ellenrieder B, Hecher K, Hackeloer BJ, Huber A, Bartmann P. Long-term neurodevelopmental outcome of 167 children after intrauterine laser treatment for severe twin-twin transfusion syndrome. *Am J Obstet Gynecol.* 2006 Feb;194(2):303-8.

26. Habli M, Lim FY, Crombleholme T. Twin-to-twin transfusion syndrome: a comprehensive update. *Clin Perinatol.* 2009 Jun;36(2):391-416, x.
27. Harrison MR, Adzick NS. The fetus as a patient. Surgical considerations. *Ann Surg.* 1991 Apr;213(4):279-91; discussion 277-8.
28. Harrison MR, Keller RL, Hawgood SB, Kitterman JA, Sandberg PL, Farmer DL, Lee H, Filly RA, Farrell JA, Albanese CT. A Randomized Trial of Fetal Endoscopic Tracheal Occlusion for Severe Fetal Congenital Diaphragmatic Hernia. *N Engl J Med.* 2003 Nov 13;349(20):1916-1924.
29. Harrison MR, Adzick NS, Bullard KM, Farrell JA, Howell LJ, Rosen MA, Sola A, Goldberg JD, Filly RA. Correction of congenital diaphragmatic hernia in utero VII: a prospective trial. *J Pediatr Surg.* 1997 Nov;32(11):1637-42.
30. Harrison MR, Adzick NS, Flake AW, VanderWall KJ, Bealer JF, Howell LJ, Farrell JA, Filly RA, Rosen MA, Sola A, Goldberg JD. Correction of congenital diaphragmatic hernia in utero VIII: Response of the hypoplastic lung to tracheal occlusion. *J Pediatr Surg.* 1996 Oct;31(10):1339-48.
31. Harrison MR, Mychaliska GB, Albanese CT, Jennings RW, Farrell JA, Hawgood S, Sandberg P, Levine AH, Lobo E, Filly RA. Correction of congenital diaphragmatic hernia in utero IX: fetuses with poor prognosis (liver herniation and low lung-to-head ratio) can be saved by fetoscopic temporary tracheal occlusion. *J Pediatr Surg.* 1998 Jul;33(7):1017-22;discussion 1022-3.
32. Harrison MR, Sydorak RM, Farrell JA, Kitterman JA, Filly RA, Albanese CT. Fetoscopic temporary tracheal occlusion for congenital diaphragmatic hernia: prelude to a randomized, controlled trial. *J Pediatr Surg.* 2003 Jul;38(7):1012-20.
33. Harrison MR. Fetal surgery. Clinical opinion. *Am J Obstet Gynecol.* 1996 Apr;174(4):1255-64.
34. Hedrick HL, Crombleholme TM. Current status of fetal surgery. *Contemporary OB/GYN® Archive.* 2001 Dec 3;12:42-70. Accessed August 15, 2005. Available at URL address: [http://www.contemporaryobgyn.net/be\\_core/content/journals/g/data/2001/1203/past\\_issues\\_show\\_article.jsp?filename=gcrombleholme1.html&title=Cover%40Story%3A%40Current%40status%40of%40fetal%40surgery&navtype=g&showPoll=yes&path=/be\\_core/content/journals/g/data/2001/1203](http://www.contemporaryobgyn.net/be_core/content/journals/g/data/2001/1203/past_issues_show_article.jsp?filename=gcrombleholme1.html&title=Cover%40Story%3A%40Current%40status%40of%40fetal%40surgery&navtype=g&showPoll=yes&path=/be_core/content/journals/g/data/2001/1203)
35. Hedrick HL, Flake AW, Crombleholme TM, Howell LJ, Johnson MP, Wilson RD, Adzick NS. Sacrococcygeal teratoma: prenatal assessment, fetal intervention, and outcome. *J Pediatr Surg.* 2004 Mar;39(3):430-8; discussion 430-8.
36. Hirose S, Farmer DL. Fetal surgery for sacrococcygeal teratoma. *Clin Perinatol.* 2003 Sep 1;30(3):493-506.
37. Hüsler MR, Wilson RD, Horii SC, Bebbington MW, Adzick NS, Johnson MP. When is fetoscopic release of amniotic bands indicated? Review of outcome of cases treated in utero and selection criteria for fetal surgery. *Prenat Diagn.* 2009 May;29(5):457-63.
38. Jani JC, Nicolaidis KH, Gratacos E, Vandercruys H, Deprest JA, FETO Task Group. Fetal lung-to-head ratio in the prediction of survival in severe left-sided diaphragmatic hernia treated by fetal endoscopic tracheal occlusion (FETO). *Am J Obstet Gynecol.* 2006 Dec;195(6):1646-50.
39. Johnson MP, Bukowski TP, Reitleman C, Isada NB, Pryde PG, Evans MI. In utero surgical treatment of fetal obstructive uropathy: a new comprehensive approach to identify appropriate candidates for vesicoamniotic shunt therapy. *Am J Obstet Gynecol.* 1994 Jun;170(6):1770-6; discussion 1776-9.
40. Johnson MP, Gerdes M, Rintoul N, Pasquariello P, Melchionni J, Sutton LN, Adzick NS. Maternal-fetal surgery for myelomeningocele: neurodevelopmental outcomes at 2 years of age. *Am J Obstet Gynecol.* 2006 Apr;194(4):1145-50; discussion 1150-2.

41. Johnson MP, Sutton LN, Rintoul N, Crombleholme TM, Flake AW, Howell LJ, Hedrick HL, Wilson RD, Adzick NS. Fetal myelomeningocele repair: short-term clinical outcomes. *Am J Obstet Gynecol.* 2003 Aug 1;189(2):482-7.
42. Keswani SG, Johnson MP, Adzick NS, Hori S, Howell LJ, Wilson RD, Hedrick H, Flake AW, Crombleholme TM. In utero limb salvage: fetoscopic release of amniotic bands for threatened limb amputation. *J Pediatr Surg.* 2003 Jun;38(6):848-51.
43. Koike T, Minakami H, Kosuge S, Izumi A, Shiraishi H, Sato I. Severe hypoproteinemia in a fetus after pleuro-amniotic shunts with double-basket catheters for treatment of chylothorax. *J Obstet Gynaecol Res.* 2000 Oct;26(5):373-6.
44. Lee H, Wagner AJ, Sy E, Ball R, Feldstein VA, Goldstein RB, Farmer DL. Efficacy of radiofrequency ablation for twin-reversed arterial perfusion sequence. *Am J Obstet Gynecol.* 2007 May;196(5):459.e1-4.
45. Lenclen R, Ciarlo G, Paupe A, Bussieres L, Ville Y. Neurodevelopmental outcome at 2 years in children born preterm treated by amnioreduction or fetoscopic laser surgery for twin-to-twin transfusion syndrome: comparison with dichorionic twins. *Am J Obstet Gynecol.* 2009 Sep;201(3):291.e1-5.
46. Livingston JC, Lim FY, Polzin W, Mason J, Crombleholme TM. Intrafetal radiofrequency ablation for twin reversed arterial perfusion (TRAP): a single-center experience. *Am J Obstet Gynecol.* 2007 Oct;197(4):399.e1-3.
47. Lylerly AD, Mahowald MB. Maternal-fetal surgery for treatment of myelomeningocele. *Clin Perinatol.* 2003 Mar 1;30(1):135-65.
48. Moise KJ JR. Neurodevelopmental outcome after laser therapy for twin-twin transfusion syndrome. *Am J Obstet Gynecol.* 2006 May;194(5):1208-10. Epub 2006 Mar 30.
49. Moise KJ Jr, Dorman K, Lamvu G, Saade GR, Fisk NM, Dickinson JE, Wilson RD, Gagnon A, Belfort MA, O'Shaughnessy RO, Chitkara U, Hassan SS, Johnson A, Sciscione A, Skupski D. A randomized trial of amnioreduction versus septostomy in the treatment of twin-twin transfusion syndrome. *Am J Obstet Gynecol.* 2005 Sep;193(3 Pt 1):701-7.
50. National Institute of Health (NIH). Management of myelomeningocele study (MOMS). Updated April 1, 2009. Accessed June 25, 2010. Available at URL address: <http://clinicaltrials.gov/ct2/results?term=myelomeningocele>
51. National Institute for Health and Clinical Excellence (NICE). Fetal cystoscopy for the diagnosis and treatment of lower urinary tract obstruction. Interventional procedure guidance IPG205. United Kingdom. January 24, 2007. Accessed June 25, 2010. Available at URL address: <http://guidance.nice.org.uk/IPG205/guidance/pdf/English>
52. National Institute for Health and Clinical Excellence (NICE). Fetal vesico-amniotic shunt for lower urinary tract outflow obstruction. Interventional procedure guidance IPG202. United Kingdom. December 13, 2006. Accessed June 25, 2010. Available at URL address: <http://www.nice.org.uk/guidance/index.jsp?action=byTopic&o=7261>
53. National Institute for Health and Clinical Excellence (NICE). Insertion of pleuro-amniotic shunt for fetal pleural effusion. Interventional procedure guidance IPG190. United Kingdom. September 27, 2006. Accessed June 25, 2010 Available at URL address: <http://www.nice.org.uk/guidance/index.jsp?action=byTopic&o=7261>
54. National Institute for Health and Clinical Excellence (NICE). Percutaneous fetal balloon valvuloplasty for aortic stenosis. Interventional procedural guidance IPG175. United Kingdom. May 24, 2006. Accessed June 25, 2010. Available at URL address:

<http://www.nice.org.uk/search/guidancesearchresults.jsp?keywords=balloon+valvuloplasty&searchType=guidance>

55. National Institute for Health and Clinical Excellence (NICE). Percutaneous fetal balloon valvuloplasty for pulmonary atresia with intact ventricular septum- information for the public. Interventional procedural guidance IPG176. United Kingdom. May 26, 2006. Accessed June 25, 2010. Available at URL address: <http://www.nice.org.uk/guidance/index.jsp?action=byTopic&o=7261>
56. National Institute for Health and Clinical Excellence (NICE). Percutaneous laser therapy for fetal tumours. Interventional procedure guidance IPG180. United Kingdom. June 28, 2006. Accessed June 25, 2010. Available at URL address: <http://www.nice.org.uk/guidance/index.jsp?action=byTopic&o=7261>
57. Nicolaidis KH, Azar GB. Thoraco-amniotic shunting. *Fetal Diagn Ther.* 1990;5(3-4):153-64.
58. Odibo AO, Biron-Shental T, Tomlinson TM, Shim TL, Wanat K. Discussion: 'Predictive value of lung-head ratio in congenital diaphragmatic hernia' by Yang et al. *Am J Obstet Gynecol.* 2007 Jul;197(1):e1-e5.
59. Prontera W, Jaeggi ET, Pfizenmaier M, Tassaux D, Pfister RE. Ex utero intrapartum treatment (EXIT) of severe fetal hydrothorax. *Arch Dis Child Fetal Neonatal Ed.* 2002 Jan;86(1):F58-60.
60. Quintero RA. Twin to twin transfusion syndrome. *Clin Perinatol.* 2003 Sep;30(3):591-600.
61. Quintero RA; Chmait RH; Murakoshi T; Pankrac Z; Swiatkowska M; Bornick PW; Allen MH. Surgical management of twin reversed arterial perfusion sequence. *Am J Obstet Gynecol.* 2006Apr;194(4):982-91.
62. Quintero RA, Comas C, Bornick PW, Allen MH, Kruger M. Selective versus non-selective laser photocoagulation of placental vessels in twin-to-twin transfusion syndrome. *Ultrasound Obstet Gynecol.* 2000 Sep;16(3):230-6.
63. Quintero RA, Huhta J, Suh E, Chmait R, Romero R, Angel J. In utero cardiac fetal surgery: laser atrial septotomy in the treatment of hypoplastic left heart syndrome with intact atrial septum. *Am J Obstet Gynecol.* 2005 Oct;193(4):1424-8.
64. Quintero RA, Martinez JM, Lopez J, Bermudez C, Beccera C, Morales W, Arroyo J. Individual placental territories after selective laser photocoagulation of communicating vessels in twin-twin transfusion syndrome. *Am J Obstet Gynecol.* 2005 Apr;192(4):1112-8.
65. Quintero RA, Shukla AR, Homsy YL, Bukkapatnam R. Successful in utero endoscopic ablation of posterior urethral valves: a new dimension in fetal urology. *Urology.* 2000 May 1;55(5):774.
66. Rocha G, Fernandes P, Rocha P, Quintas C, Martins T, Proenca E. Pleural effusions in the neonate. *Acta Paediatr.* 2006 Jul;95(7):791-8.
67. Ronderos-Dumit D, Briceno F, Navarro H, Sanchez N. Endoscopic release of limb constriction rings in utero. *Fetal Diagn Ther.* 2006;21(3):255-8.
68. Rossi AC, D'Addario V. Laser therapy and serial amnioreduction as treatment for twin-twin transfusion syndrome: a metaanalysis and review of literature. *Am J Obstet Gynecol.* 2008 Feb;198(2):147-52.
69. Rossi AC, D'Addario V. Umbilical cord occlusion for selective feticide in complicated monochorionic twins: a systematic review of literature. *Am J Obstet Gynecol.* 2009 Feb;200(2):123-9.
70. Ruano R. Fetal surgery for severe lower urinary tract obstruction. *Prenat Diagn.* 2011 Mar 17. doi: 10.1002/pd.2736.

71. Salomon LJ, Orqvist L, Aegerter P, Bussieres L, Staracci S, Stirnemann JJ, Essaoui M, Bernard JP, Ville Y. Long-term developmental follow-up of infants who participated in a randomized clinical trial of amniocentesis vs laser photocoagulation for the treatment of twin-to-twin transfusion syndrome. *Am J Obstet Gynecol.* 2010 Nov;203(5):444.e1-7.
72. Senat MV, Deprest J, Boulvain M, Paupe A, Winer N, Ville Y. Endoscopic laser surgery versus serial amnioreduction for severe twin-to-twin transfusion syndrome. *N Engl J Med.* 2004 Jul 8;351(2):136-44.
73. Smeland E, Prydz H, Orstavik K, Froland S. Gene therapy: Status and potential in clinical medicine. Oslo, Norway: The Norwegian Knowledge Centre for the Health Services; 2000. Accessed June 25, 2010. Available at URL address: <http://www.crd.york.ac.uk/CRDWeb/ShowRecord.asp?View=Full&ID=32001000031>
74. Smith RP, Illanes S, Denbow ML, Soothill PW. Outcome of fetal pleural effusions treated by thoracoamniotic shunting. *Ultrasound Obstet Gynecol.* 2005 Jul;26(1):63-6.
75. Smith NP, Jesudason EC, Featherstone NC, Corbett HJ, Losty PD. Recent advances in congenital diaphragmatic hernia. *Arch Dis Child.* 2005 Apr;90(4):426-8.
76. Sutton LN. Fetal surgery for neural tube defects. *Best Pract Res Clin Obstet Gynaecol.* 2008 Feb;22(1):175-88. Epub 2007 Aug 22.
77. Sutton LN, Adzick NS, Bilaniuk LT, Johnson MP, Cromblehome TM, Flake AW. Improvement in hindbrain herniation demonstrated by serial fetal magnetic resonance imaging following fetal surgery for myelomeningocele. *JAMA.* 1999 Nov 17;282(19):1826-31.
78. Sydorak RM, Harrison MR. Congenital diaphragmatic hernia: advances in prenatal therapy. *Clin Perinatol.* 2003 Sep;30(3):465-79.
79. Tan TY, Sepulveda W. Acardiac twin: a systematic review of minimally invasive treatment modalities. *Ultrasound Obstet Gynecol.* 2003 Oct;22(4):409-19.
80. Tulipan N, Sutton LN, Bruner JP, Cohen BM, Johnson M, Adzick NS. The effect of intrauterine myelomeningocele repair on the incidence of shunt-dependent hydrocephalus. *Pediatr Neurosurg.* 2003 Jan 1;38(1):27-33.
81. Vidaeff AC, Pschirrer ER, Mastrobattista JM, Gilstrap LC III, Ramin SM. Mirror syndrome. A case report. *J Reprod Med.* 2002 Sep;47(9):770-4.
82. Ville Y, Hyett JA, Vandenbussche FP, Nicolaides KH. Endoscopic laser coagulation of umbilical cord vessels in twin reversed arterial perfusion sequence. *Ultrasound Obstet Gynecol.* 1994 Sep 1;4(5):396-8.
83. Waddington SN, Kramer MG, Hernandez-Alcoceba R, Buckley SM, Themis M, Coutelle C, Prieto J. In utero gene therapy: current challenges and perspectives. *Mol Ther.* 2005 May;11(5):661-76.
84. Walsh WF, Chescheir NC, Gillam-Krakauer M, McPheeters ML, McKoy JN, Jerome R, Fisher JA, Meints L, Hartmann KE. Maternal-Fetal Surgical Procedures [Internet]. Rockville (MD): Agency for Healthcare Research and Quality (US); 2011 Apr. Report No.: 10(11)-EHC059-EF. AHRQ Comparative Effectiveness Reviews.
85. Warner BW. Congenital Diaphragmatic Hernia. In: Townsend CM, Beauchamp RD, Evers BM, Mattox KL, editors. *Sabiston Textbook of Surgery*, 17<sup>th</sup> ed., Copyright © 2004 Elsevier. Chapter 70.
86. Weisz B, Peltz R, Chayen B, Oren M, Zalel Y, Achiron R, Lipitz S. Tailored management of twin reversed arterial perfusion (TRAP) sequence. *Ultrasound Obstet Gynecol.* 2004 May 1;23(5):451-5.

87. Wilson RD, Baxter JK, Johnson MP, King M, Kasperski S, Crombleholme TM, Flake AW, Hedrick HL, Howell LJ, Adzick NS. Thoracoamniotic shunts: fetal treatment of pleural effusions and congenital cystic adenomatoid malformations. *Fetal Diagn Ther.* 2004 Sep-Oct;19(5):413-20.
88. Winer N, Salomon LJ, Essaoui M, Nasr B, Bernard JP, Ville Y. Pseudoamniotic band syndrome: a rare complication of monochorionic twins with fetofetal transfusion syndrome treated by laser coagulation. *Am J Obstet Gynecol.* 2008 Apr;198(4):393.e1-5.
89. Wu S, Johnson MP. Fetal lower urinary tract obstruction. *Clin Perinatol.* 2009 Jun;36(2):377-90, x.
90. Yang SH, Nobuhara KK, Keller RL, Ball RH, Goldstein RB, Feldstein VA, Callen PW, Filly RA, Farmer DL, Harrison MR, Lee H. Reliability of the lung-to-head ratio as a predictor of outcome in fetuses with isolated left congenital diaphragmatic hernia at gestation outside 24-26 weeks. *Am J Obstet Gynecol.* 2007 Jul;197(1):30.e1-7.
91. Yinon Y, Kelly E, Ryan G. Fetal pleural effusions. *Best Pract Res Clin Obstet Gynaecol.* 2008 Feb;22(1):77

---

## Policy History

<b>Pre-Merger Organizations</b>	<b>Last Review Date</b>	<b>Policy Number</b>	<b>Title</b>
CIGNA HealthCare	8/15/2008	0175	Fetal Surgery

“CIGNA”, “CIGNA HealthCare” and the “Tree of Life” logo are registered service marks of CIGNA Intellectual Property, Inc., licensed for use by CIGNA Corporation and its operating subsidiaries. All products and services are provided by such operating subsidiaries and not by CIGNA Corporation. Such operating subsidiaries include Connecticut General Life Insurance Company, CIGNA Health and Life Insurance Company, CIGNA Behavioral Health, Inc., CIGNA Health Management, Inc., and HMO or service company subsidiaries of CIGNA Health Corporation and CIGNA Dental Health, Inc. In Arizona, HMO plans are offered by CIGNA HealthCare of Arizona, Inc. In California, HMO plans are offered by CIGNA HealthCare of California, Inc. In Connecticut, HMO plans are offered by CIGNA HealthCare of Connecticut, Inc. In North Carolina, HMO plans are offered by CIGNA HealthCare of North Carolina, Inc. In Virginia, HMO plans are offered by CIGNA HealthCare Mid-Atlantic, Inc. All other medical plans in these states are insured or administered by Connecticut General Life Insurance Company or CIGNA Health and Life Insurance Company.