Clinical Policy Title: Fetal surgery in utero

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Related policies:

None.

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Coverage policy

Arbor Health Plan considers the use of fetal surgery in utero for the following conditions and/or diagnoses to be clinically proven and therefore, medically necessary, when the following criteria are met:

A. Myelomeningocele (MMC) repair:
   • MMC with upper boundary of the lesion located between T1 and S1.
   • Evidence of hindbrain herniation.
   • Singleton pregnancy.
   • Gestational age 19 to 26 weeks.
   • Normal fetal karyotype NS.

B. Serial amnioreduction for twin-to-twin transfusion syndrome (TTTS):
   • Women after 26 weeks of gestation.
- Evidence of abnormal blood flow documented by Doppler studies in one or both fetuses. Evidence of polyhydramnios in the recipient fetus.
- Donor fetus is oligohydramniotic.

C. Vesico-amniotic shunting as a treatment of urinary tract obstruction:
- Bladder distension.
- Bilateral hydronephrosis due to urinary tract obstruction.
- Evidence of progressive oligohydramnios or anyhydramnios.
- Adequate renal function.
- No other lethal abnormalities or lethal chromosomal abnormalities.

D. Fetoscopic or open laser ablation of anastomotic vessels, with or without amnioreduction, as treatment for TTTS when both of the following criteria are met:
- Diagnosis of severe TTTS is confirmed clinically and by ultrasound between 15 and 26 weeks gestation.
- The benefits of laser surgery outweigh the risks to a previable fetus that is not a candidate for delivery, and has a high mortality rate otherwise.

E. Resection of malformed pulmonary tissue, or placement of a thoracoamniotic shunt as a treatment for any of the following:
- Congenital cystic adenomatoid malformation (CCAM) when both of the following criteria are met:
  - Evidence of fetal hydrops.
  - Presence of one lesion, consisting of a single large cyst.
- Extralobar pulmonary sequestration (EPS), when there is evidence of tension hydrothorax and/or fetal hydrops.
- Fetal pleural effusion (FPE) 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 hypoplasia.

F. In utero needle access and open resection of sacrococcygeal teratoma (SCT) as medically necessary for a previable fetus with or without early maternal pre-eclampsia, when there is evidence of fetal hydrops, placentomegaly or polyhydramnios.

G. Optimal outcomes of fetal surgery in utero are found at facilities with the expertise, multidisciplinary teams, services, and highly credentialed surgeons who specialize in fetal surgery in utero, thus providing a high level of intensive care. (Rigorous patient care selection that offers the best outcomes with the least risk is important.)
H. Documentation is provided from the requesting physician stating that the anticipated outcome from proposed in utero surgery has a greater likelihood of a positive outcome than surgery performed post-delivery. This documentation must contain all necessary test results and studies that lead to a positive outcome.

I. Twin reversed arterial perfusion (TRAP): ablation or occlusion of anastomotic vessels (e.g., laser coagulation or radiofrequency ablation).

Limitations:

Arbor Health Plan considers the use of the following in fetal surgery in utero to be investigational and therefore, not medically necessary for any of the following conditions:

- Fetal tracheal occlusion for congenital diaphragmatic hernia (CDH).
- Amnioexchange procedure for gastrochisis.
- Treatment of cleft lip and/or palate.
- Treatment for aqueductal stenosis (i.e., hydrocephalus).
- Amniotic band syndrome.
- Treatment of congenital heart defects.
- In utero gene therapy.
- In utero hematopoietic stem cell transplantation for stem cell diseases.

All other uses of fetal surgery in utero are not medically necessary.

Note: The following CPT/HCPCS codes are not listed in the Nebraska Medicaid fee schedule:

S2401 - Repair, urinary tract obstruction in the fetus, procedure performed in utero
S2402 - Repair, congenital cystic adenomatoid malformation in the fetus, procedure performed in utero
S2403 - Repair, extralobar pulmonary sequestration in the fetus, procedure performed in utero
S2404 - Repair, myelomeningocele of the fetus, procedure performed in utero
S2405 - Repair of sacrococcygeal teratoma in the fetus, procedure performed in utero
S2409 - Repair, congenital malformation of fetus, procedure performed in utero not otherwise classified
S2411 - Fetoscopic laser therapy for treatment of twin-to-twin transfusion syndrome
Alternative covered services:

Post-delivery and obstetrical management and treatment.

**Background**

For decades, experimental fetal surgery proved essential in studying normal fetal physiology and development, and pathophysiology of congenital defects. Clinical fetal surgery started in the 1960s with intrauterine transfusions. In the 1970s, the advent of ultrasonography revolutionized fetal diagnosis and created a therapeutic vacuum. Fetal treatment, medical and surgical, is slowly trying to fill the gap. Most detected defects are best treated after birth and require a modification in the time, mode, and place of delivery for optimal obstetrical and neonatal care. Surgical intervention in utero should be considered for the following conditions:

- Malformations that may cause progressive damage to the fetus and lead to death or severe morbidity.
- Malformations that can be corrected or palliated in utero, with a reasonable expectation of normal postnatal development.
- Malformations that cannot wait to be corrected after birth (regardless of preterm delivery), and are not accompanied by chromosomal or other major anomalies.

At present, congenital hydronephrosis is the most common indication for fetal surgery, followed by obstructive hydrocephalus. CDH also fulfills the criteria, but its correction poses more problems, and no clinical attempts have been reported thus far. In the future, many other malformations or diseases may be best treated in utero. The ethical and moral issues are complex and need to be discussed as clinical and experimental progress is made.

Although most prenatally diagnosed malformations are best managed by appropriate medical and surgical therapy, after maternal transport and planned delivery at a tertiary care center, an expanding number of simple anatomical abnormalities with predictable, lethal consequences have been successfully corrected before birth. A malformation amenable to prenatal surgical intervention must fulfill a number of conditions. It must be severe enough to warrant the risks associated with in utero treatment and must be reliably detectable before birth. Additionally, the pathophysiology must be reversible by fetal surgery, significantly improving the prognosis over post-natal treatment. Many technical intricacies of open fetal surgery have been solved, but preterm labor and premature rupture of membranes remain an omnipresent risk to both the mother and the fetus. To reduce maternal morbidity and the risk of prematurity, minimally invasive techniques to treat the fetus prenatally have been developed. Current indications of fetal surgery include the treatment of congenital diaphragmatic hernia, cystic adenomatoid malformation of the lung, SCT, obstructive uropathy, TTTS and MMC. Minimally invasive surgical techniques (e.g., Fetendo) have significantly lessened the incidence of preterm labor and promise to extend the indications for fetal surgical intervention.
In utero fetal surgery involves a highly technical, multidisciplinary approach to correct malformations of the fetus that interfere with organ development and 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, which are 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.

Myelomeningocele (MMC)

Also referred to as spina bifida, MMC is the most commonly observed malformation of the central nervous system, affecting more than 1,000 fetuses annually in the United States. MMC is characterized by protrusion of the meninges and spinal cord through a defect in the vertebral arches, muscle, and skin of a developing fetus. Although the condition is rarely fatal, MMC patients are often limited by a range of disabilities, including paraplegia, hydrocephalus, skeletal deformities, bowel and bladder incontinence, and cognitive impairment. The standard treatment for MMC is to close the defect postnatally; however, this approach does not address damage associated with exposure of the meninges and spinal cord to cerebrospinal fluid (CSF), while in utero. As a consequence, researchers have proposed the feasibility of intervening in utero to prevent this damage by establishing a normal CSF drainage pathway. In utero fetal surgery (IUFS) for MMC remains highly controversial because the MMC defect is not fatal, and postnatal treatment is much simpler and not traumatic to the mother. The present report evaluates IUFS for MMC relative to standard postnatal treatment.

In a Hayes report, on fetal inutero surgery evaluated evidence was published between 2002 and August 2012 and included 11 studies, which compared IUFS with standard care (postnatal surgery) for MMC. The majority of the studies were published within the past five years and sample size ranged from 20 to 293 fetuses. Eight of the studies were retrospective and three were prospective, including a randomized
controlled trial (RCT). In utero surgery was compared with postnatal surgery in all studies. None of the studies included blinding of patients to the surgical procedure. Surgeries were completed at five sites: (1) the Center for Fetal Diagnosis and Treatment at the Children’s Hospital of Philadelphia (five studies); (2) Division of Pediatric Surgery and Fetal Treatment Center at the University of California, San Francisco (two studies); (3) Division of Pediatric Urology, Vanderbilt University (five studies); (4) University Medical Center Groningen in the Netherlands (one study); and (5) University of North Carolina School of Medicine in Chapel Hill (one study). The gestational age at the time of IUFS was between 20 to 25 weeks for all studies except for two: one included a cohort that was operated on between 20 to 30 weeks, and another had a mean operation gestational age of 37 weeks. Timing of follow up across studies varied considerably. Four studies included testing only during postnatal stay at the hospital, one study included a one year follow-up, two included variable follow-up that was ≥ 1 year; one study included both a one year and 30 month follow-up, and two studies included longer-term follow-up of three years and approximately 9 to 10 years.

The conclusion per Hayes report is: One RCT and ten nonrandomized controlled studies (NRSs) compared IUFS with standard postnatal surgery for MMC and provided moderate equality evidence that IUFS is associated with improved motor function, excretory function, and neuroanatomical outcomes, as well as reduced need for shunt placement. However, IUFS did not influence cognitive outcomes and did result in lower gestational age at birth (premature birth), relative to postnatal controls. Most of the studies were retrospective and did not follow up on the developmental outcomes of IUFS past three years of age. As a consequence, the relationship between many of the findings and longer-term clinical outcomes is not clear at this time. Therefore, based on this review of the available evidence, some published evidence suggests that safety and impact on health outcomes are at least comparable to standard treatment. However, substantial uncertainty remains about safety and/or impact on health outcomes, because of poor-quality studies, sparse data, conflicting study results, and other concerns (Hayes (2013)).

MOMS trial

In February 2003, the National Institutes of Health (NIH) began the Management of MMC Study (MOMS). Three centers (Vanderbilt University, Children’s Hospital of Philadelphia and the University of California at San Francisco) were chosen to participate in the study of 183 randomized fetuses: 91 for fetal repair and 92 for postnatal repair. The study took eight years to complete at a cost of $22.5 million. Of the 1,087 fetuses and mothers initially screened for the study, 183 met all the inclusion criteria. The Children’s Hospital of Philadelphia treated 77 patients, University of California at San Francisco treated 54 and Vanderbilt University treated 52.

Prenatal surgery was done at the assigned center between 19 and 25 weeks of pregnancy. Deliveries for both groups were performed by C-section at the assigned MOMS Center, at approximately 37 weeks of pregnancy. The infants in the postnatal surgery group had their spina bifida closed at the MOMS Center as soon as possible after delivery, usually within 48 hours. Medical information on the mothers and
babies were gathered throughout the study and follow up of their progress continued until the child reached at least two-and-a-half years of age. Two outcomes were considered: the first, at 12 months, was death or need for a ventricular shunt; the second, measured at 30 months, was a composite score of standardized tests for mental and motor development.

Outcome of the trial

An interim analysis conducted in December 2010, made public in February 2011, and released in the *New England Journal of Medicine* (NEJM) 2011, showed a statistically significant benefit to the surgery, and the trial was subsequently closed. The trial demonstrated that outcomes after prenatal spina bifida treatment are improved, to the degree that the benefits of the surgery outweigh the maternal risks. Results were reported in the *NEJM* by Adzick et al.

Specifically, the study found:

- Babies who underwent fetal repair of spina bifida were half as likely to need a ventricular shunt.
- Chiari malformation was less common in the fetal repair group.
- Standardized test scores for motor skills were superior in the fetal surgery group, and twice as many children were walking independently at 30 months, as compared to the postnatal surgery group.

Based on these outcomes, fetal repair of spina bifida is now considered a standard of care at some fetal centers. However, prenatal repair is a complex and challenging procedure, requiring the most expert, comprehensive care for both mother and fetus. The surgical team’s level of experience in all aspects of care surrounding the operation is of paramount importance.

Adzick (2010) stated that MMC is a common birth defect associated with significant lifelong morbidity. Little progress has been made in the postnatal surgical management of the child with spina bifida. Postnatal surgery is aimed at covering the exposed spinal cord, preventing infection, and treating hydrocephalus with a ventricular shunt. In utero repair of open spina bifida is now performed in selected patients and presents an additional therapeutic alternative for expectant mothers carrying a fetus with MMC. As of 2010, it is estimated that about 400 fetal operations have been performed for MMC worldwide. Despite these numbers, the technique remains of unproven benefit. Preliminary results suggest that fetal surgery results in reversal of hindbrain herniation (the Chiari II malformation), a decrease in shunt-dependent hydrocephalus, and possibly improvement in leg function, but these findings might be explained by selection bias and changing management indications. A prospective, RCT (the MOMS trial) is currently being conducted by three centers in the United States, and the estimated completion date is in 2010. The author stated that further research is needed to better understand the pathophysiology of MMC, the ideal timing and technique of repair, and the long-term impact of in utero intervention.

Twin-to-twin transfusion syndrome (TTTS)
TTTS is a complication unique to monochorionic twin pregnancies, in which twins share a common placenta, and unequal blood exchange from one twin (donor) to the co-twin occurs through placental arterovenous anastomoses. The diagnosis is posed with the sonographic detection of oligohydramnios in the donor’s sac, as a consequence of hypervolemia, and polyhydramnios in the recipient’s sac, resulting from hypervolemia. The severity of TTTS is assessed according to the Quintero staging system, which is based on five stages that range from visualization of the donor’s bladder to intrauterine demise of one or both twins. Because laser therapy (LT) has proven to be the optimal treatment for TTTS, management of twins affected with TTTS consists of preoperative and postoperative assessment of the twins by ultrasound examination.

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 percent. 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 is amnioreduction, which 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 are reported to be between 50 – 65 percent with this intervention (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 percent, with 80 percent 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.

Multiple pregnancies represent two percent of all pregnancies, but account for 20 percent of admissions to neonatal intensive care units. The outcome of multiple pregnancies is mainly dependent on chorionicity. Most perinatal complications are 3 to 12 times more prevalent in monochorionic pregnancies. The increased mortality and morbidity in this setting are mainly related to vascular anastomoses on the chorionic plate, joining the two fetal circulations. Intrauterine death (IUD) of a monochorionic twin leads to exsanguination of the survivor, with fatal outcome in 20 to 30 percent of cases and a similar rate of severe ischemic complications. The most severe and acute complication is the
TTTS. The main clinical manifestations of TTTS are the polyuric polyhydramnios — oliguric oligohydramnios sequence in the recipient and donor twin, respectively. TTTS is associated with a perinatal mortality rate of around 90 percent, and neurological sequelae are present in 20–40 percent of survivors born at around 25 weeks. Serial amnioreduction was the only fetal therapy for TTTS, improving the survival rate to around 50 percent for at least one twin delivered at around 28 weeks, and reducing the risk of sequelae to around 20 percent among survivors.

An intrauterine fetoscopic surgical treatment has been developed for TTTS. A 2-mm endoscope and a diode laser fiber are introduced percutaneously, under local anesthesia, through a single 3-mm trocard coagulation of feto-fetal anastomoses on the chorionic plate, which leads to the survival of at least one twin in around 80 percent of cases, at 33 weeks. Fewer than 10 percent of survivors have sequelae, mainly related to prematurity. We conducted an RCT that confirmed the superiority of LT over amnioreduction. The long-term outlook of these infants is good, with up to six years of follow up.

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 percent 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). Surgery is not curative and further evaluation and surgical treatment are necessary following delivery (Wu and Johnson, 2009).

Fetal pleural effusions (FPEs)

Isolated FPEs 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 that 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 percent when diagnosis is made prior to 32 weeks gestation, to 30 percent when the diagnosis is made later (Prontera et al., 2002). When the condition is associated with hydrops, mortality rates approach 100 percent. 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). However, published evidence is 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.

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 (IFMSS) in 1985, still remains in effect on percutaneous shunting for fetal hydrocephalus.

Sacrococcygeal teratoma (SCT)

An SCT 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 preeclampsia.

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 percent fetal mortality.

Additional methods that have been proposed for treating SCT involve 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).

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

Lee et al. (2013) reported the North American Fetal Therapy Network (NAFTNet) Registry data on the outcomes of using radiofrequency ablation to treat TRAP. This was a retrospective review of all patients who underwent percutaneous radiofrequency ablation of an acardiac twin, after referral to a NAFTNet institution. The primary outcome was neonatal survival to 30 days of age. Of the 98 patients identified, there were no maternal deaths. Mean gestational age at delivery was 33.4 weeks overall, and 36 weeks for survivors. Median gestational age at delivery was 37 weeks. Survival of the pump twin to 30 days was 80 percent in the In Utero Fetal Surgery: Medical Policy (effective 05/01/2014) overall cohort. The authors concluded this data suggests that radiofrequency ablation of the acardiac twin is an effective treatment for TRAP sequence.

Pagani et al. (2013) conducted a retrospective cohort study and meta-analysis of intrafetal laser treatment for TRAP sequence. Twenty-three cases of TRAP were identified during the study period. Six were managed conservatively and 17 were treated with laser therapy. All cases managed conservatively were complicated by IUD at a median gestational age of 14 weeks. Among the treated cases, 14 (82 percent) delivered a healthy twin at a median gestational age of 37 weeks. The overall neonatal survival was 80 percent. Adverse pregnancy outcome was significantly lower when the treatment was performed before 16 weeks gestation.

Weisz et al. studied six cases. To describe our management of pregnancies complicated by TRAP sequence, three patients in whom there were no signs of deterioration in the status of the pump twin, and in whom the acardiac twin was smaller than the pump twin, were managed conservatively. However, one of these with monoamniotic twins ended in intrauterine fetal death of the pump twin. The other two cases presented with spontaneous cessation of blood flow in the umbilical artery of the acardiac twin. Both delivered at term normal neonates whose follow-up revealed no signs of neurological sequelae. One case of quadruplet pregnancy (with TRAP sequence and two dichorionic twins), was treated by selective termination of the monochorionic twins. Two cases with signs of impending cardiac failure were treated by in utero YAG laser occlusion of the vessels in the acardiac mass. Both interventions had a favorable outcome. Conservative treatment is suitable for milder cases of TRAP sequence, in which the pump twin is the larger one. Cases in which the acardiac twin is larger have a poorer prognosis and should be treated by invasive intervention and cord occlusion.

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).
**Searches**

Arbor Health Plan searched PubMed and the databases of:

- UK National Health Services Centre for Reviews and Dissemination.
- Agency for Healthcare Research and Quality’s National Guideline Clearinghouse and other evidence-based practice centers.
- The Centers for Medicare & Medicaid Services (CMS).

We conducted searches on Month Day, Year. Search terms were “fetal survival, surgery in utero using the terms prenatal malformations, congenital diagnosis.”

We included:

- **Systematic reviews**, which pool results from multiple studies to achieve larger sample sizes and greater precision of effect estimation than in smaller primary studies. Systematic reviews use predetermined transparent methods to minimize bias, effectively treating the review as a scientific endeavor, and are thus rated highest in evidence-grading hierarchies.
- **Guidelines based on systematic reviews**.
- **Economic analyses**, such as cost-effectiveness, and benefit or utility studies (but not simple cost studies), reporting both costs and outcomes — sometimes referred to as efficiency studies — which also rank near the top of evidence hierarchies.

**Findings**

Fetal surgery has become an acceptable option for the treatment of select 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, MMC repair, TTTS, 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 still lacking. Well-designed RCTs evaluating in utero intervention for these conditions, compared to postnatal intervention are required to support improved survival rates.

**Policy updates:**

Updated additional background information and references:

An up to date review on “Diagnosis and management of twin reversed arterial perfusion (TRAP) sequence” (Holland et al., 2014) states that “In utero therapy -- For continuing pregnancies with one or more poor prognostic criteria, antenatal intervention, delivery, and expectant management are options. As the acardiac twin is nonviable, treatment for TRAP sequence is focused on improving the outcome for the pump twin. Historically, intervention in pregnancies with TRAP sequence was limited to
amnioreduction to reduce hydramnios or relief for the pump twin by selective delivery of the acardiac twin via hysterotomy or administration of sclerosing agents (e.g., alcohol) into the umbilical cord of the acardiac twin. For pregnancies between 18 and 27 weeks of gestation, current treatment modalities target occlusion of the umbilical cord of the acardiac twin and include laser ablation, bipolar cord coagulation, and radiofrequency ablation (RFA), which are performed with local anesthesia and conscious sedation. Fetoscopic cord ligation is an alternative, but is less common.”

An up to date review on “Overview of antenatal hydronephrosis” (Baskin and Ozcan, 2014) states that “Fetal surgery -- Although there have been several prospective and retrospective studies of antenatal surgery in fetuses with sonographic findings consistent with lower urinary tract obstruction, there is no good evidence that this intervention improves renal outcome. These procedures increase the amount of amniotic fluid, thus potentially improving lung development and survival rate. However, there remains a high rate of chronic renal disease in the survivors, necessitating renal replacement therapy in almost two-thirds of the cases.”

Furthermore, an UpToDate review on “Congenital diaphragmatic hernia: Prenatal diagnosis and management” (Hedrick and Adzick, 2015) states that “In utero therapy is an investigational procedure with limited geographic availability.”

Ruano et al., (2013) evaluated the effect of early fetoscopic tracheal occlusion (FETO) (22 to 24 weeks gestation) on pulmonary response and neonatal survival in cases of extremely severe, isolated CDH. This was a multi-center study involving fetuses with extremely severe CDH (lung-to-head ratio less than 0.70, liver herniation into the thoracic cavity and no other detectable anomalies). Between August 2010 and December 2011, a total of eight fetuses underwent early FETO. Data was compared with nine fetuses that underwent standard FETO, and 10 without fetoscopic procedure, from January 2006 to July 2010. FETO was performed under maternal epidural anesthesia, supplemented with fetal intramuscular anesthesia. Fetal lung size and vascularity were evaluated by ultrasound, before and every two weeks after, FETO. Post-natal therapy was equivalent for both treated fetuses and controls. Primary outcome was infant survival to 180 days and secondary outcome was fetal pulmonary response. Maternal and fetal demographic characteristics and obstetric complications were similar in the 3 groups (p > 0.05). Infant survival rate was significantly higher in the early FETO group (62.5 percent) compared with the standard group (11.1 percent) and with controls (0 percent) (p < 0.01). Early FETO resulted in a significant improvement in fetal lung size and pulmonary vascularity, when compared with standard FETO (p < 0.01). The authors concluded that early FETO may improve infant survival by further increases of lung size, and pulmonary vascularity in cases with extremely severe pulmonary hypoplasia in isolated CDH. They stated that the findings of this study support formal testing of the hypothesis with an RCT.

Ruano et al., (2014) reviewed the indications, technical aspects, preliminary results, risks, and clinical implications of FETO for severe CDH performed outside the United States and its potential future directions in this country and globally. CDH occurs in approximately 1 in 2,500 live births and results in high neonatal morbidity and mortality, largely associated with the severity of pulmonary hypoplasia and pulmonary arterial hypertension. With the advent of prenatal imaging, CDH can be diagnosed before
birth, and in utero treatment is now available in some centers. The prognosis of CDH can be evaluated by assessing the fetal lung size, the degree of liver herniation, and the fetal pulmonary vasculature in isolated forms of CDH. These parameters help classify fetuses as having mild, moderate, severe, or extremely severe isolated CDH. Severe and extremely severe diaphragmatic hernias have poor outcomes and thus are candidates for innovative therapies such as FETO. FETO is usually performed between 26 and 30 weeks gestation. In utero, an endoscope is passed through the fetal mouth and down to the carina; the balloon is deployed just above the carina. After the procedure, ultrasound surveillance every two weeks ensures the balloon’s structural integrity and measures the fetal pulmonary response. At approximately 34 weeks gestation, the balloon is deflated and removed. FETO is thought to improve outcomes by decreasing mortality and allowing more rapid neonatal stabilization. Ultimately, the goal of FETO is to minimize pulmonary hypoplasia and pulmonary arterial hypertension. Following delivery, neonates still require diaphragm repair.

Summary of clinical evidence:

<table>
<thead>
<tr>
<th>Citation</th>
<th>Content, Methods, Recommendations</th>
</tr>
</thead>
<tbody>
<tr>
<td>Rossi and D'Addario (2008)</td>
<td>Key points:</td>
</tr>
<tr>
<td>Current controversy on LT versus serial amnioreduction (SA) performed for TTTS. A search in PubMed from 1997 to 2007 was performed</td>
<td>Inclusion criteria were diamniotic monochorionic pregnancy, TTTS diagnosed with standard parameters, and peri- and neo-natal outcomes well-defined.</td>
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<td></td>
<td>• Triplets and investigations on other topics of TTTS rather than perinatal outcomes were excluded. A meta-analysis was performed by fixed-effect model (heterogeneity less than 25%). A total of 10 articles provided 611 cases of TTTS (LT: 70%; SA: 30%) and included four studies comparing the two treatments (395 cases: LT, 58%; SA, 42%).</td>
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<tr>
<td></td>
<td>• Fetuses undergoing LT were more likely to survive than fetuses undergoing SA (overall survival rate: p &lt; 0.0001; odds ratio [OR], 2.04; 95% confidence interval [CI]: 1.52 to 2.76; neonatal death: p &lt; 0.0001; OR, 0.24; 95% CI: 0.15 to 0.40; neurologic morbidity: p &lt; 0.0001; OR, 0.20; 95% CI: 0.12 to 0.33). The authors concluded that this meta-analysis showed that LT is associated with better outcomes than SA.</td>
</tr>
<tr>
<td>Johnson et al., 2003. Reversal of hindbrain herniation. PubMed.</td>
<td>Key points:</td>
</tr>
<tr>
<td></td>
<td>• (n=50) reported overall perinatal survival of 94% with reversal of hindbrain herniation in all fetuses. Ventricleoperitoneal 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 MMC repair in utero.</td>
</tr>
</tbody>
</table>
|                           | • Neurodevelopmental deficits were noted but did not appear to be
worsened by fetal surgery; the deficits were considered characteristic of children with spina bifida.

### Key points:

- It was a multi-center study of singleton pregnancies with CDH treated by FETO.
- The entry criteria for FETO were severe CDH, on the basis of sonographic evidence of intra-thoracic herniation of the liver and low lung area to head circumference ratio (LHR), defined as the observed to the expected normal mean for gestation (o/e LHR) equivalent to an LHR of one or less.
- FETO was carried out in 210 cases, including 175 cases with left-sided, 34 right-sided and one with bilateral CDH.
- In 188 cases, the CDH was isolated and in 22 there was an associated defect. FETO was performed at a median gestational age of 27.1 (range of 23.0 to 33.3) weeks.
- The first eight cases were done under general anesthesia, but subsequently either regional or local anesthesia was used.
- The median duration of FETO was 10 (range of 3 to 93) mins. Successful placement of the balloon at the first procedure was achieved in 203 (96.7%) cases.
- Spontaneous preterm prelabor rupture of membranes (PPROM) occurred in 99 (47.1%) cases at 3 to 83 (median of 30) days after FETO and within three weeks of the procedure in 35 (16.7%) cases.
- Removal of the balloon was prenatal either by fetoscopy or ultrasound-guided puncture, intrapartum by ex-utero intrapartum treatment, or postnatal, either by tracheoscopy or percutaneous puncture.
- Delivery was at 25.7 to 41.0 (median of 35.3) weeks and before 34 weeks in 65 (30.9%) cases.
- In 204 (97.1%) cases, the babies were live born and 98 (48.0%) were discharged from the hospital alive.
- There were 10 deaths directly related to difficulties with removal of the balloon.
- Significant prediction of survival was provided by the o/e LHR and gestational age at delivery. On the basis of the relationship between survival and o/e LHR in expectantly managed fetuses with CDH, as reported in the antenatal CDH registry, these researchers estimated that in fetuses with left CDH treated with FETO, the survival rate increased from 24.1% to 49.1%, and in right CDH survival, increased from 0% to 35.3% (p < 0.001).
- The authors concluded that FETO in severe CDH is associated with a high incidence of PPROM and preterm delivery, but only a substantial improvement in survival. They also stated that these findings need to be tested in an RCT.

### Glossary
Amniocentesis — Procedure in which a small amount of fluid surrounding the baby is removed from the womb and studied for a variety of possible birth defects. Open spina bifida can be detected by abnormally high levels of alpha-fetoprotein in this fluid.

Anesthesiologist — A doctor who specializes in the use of medications that put people to sleep during surgery.

Body mass index (BMI) — Measurement of body fat content in the body found by dividing an individual’s weight in kilograms, by their height in meters squared (i.e., weight in kg/height2 in meters).

Chromosomes — Rod-shaped structures found inside every cell of the human body, which contain information that determines all of an individual’s characteristics. Normal human cells contain 46 chromosomes.

Folic Acid — A common B vitamin. A daily dose of 0.4mg taken for three to four months prior to the start of a pregnancy, and continued daily during the first three months of pregnancy, will decrease the chances of having a baby with spina bifida, or another neural tube defect.

General anesthesia — The use of medications to put someone completely to sleep during an operation.

Hydrocephalus — Overfilling of the fluid cavities (ventricles) of the brain. A shunting device is commonly needed to control hydrocephalus in individuals with spina bifida.

Karyotype — A test that looks at the number and appearance of the chromosomes of an individual. The karyotype of an unborn baby can be done on cells taken from the fluid removed from the womb, via amniocentesis.

Management of Myelomeningocele Study — MOMS.

Maternal serum alpha-fetoprotein (MSAFP) — Alpha-fetoprotein is a substance made by all babies while they are in the womb. If the baby has an open spina bifida defect or other neural tube defect, abnormally high levels of this protein are found in the fluid surrounding the baby and in the mother’s blood. Blood can be drawn from the mother and the amount of alpha-fetoprotein measured. If the level is suspiciously high, further testing is usually recommended. Usually a sonogram of the baby is done, along with an amniocentesis to directly measure the amount of alpha-fetoprotein in the fluid surrounding the baby.
**Myelomeningocele (MMC)** — Synonym for the most severe form of spina bifida, a congenital defect wherein the spinal cord has not completely developed and is open on the back of the baby. The overlying bones, muscle, and skin are also exposed.

**Obstetrician** — A doctor specializing in the management of pregnant women and the delivery of babies.

**Placenta previa** — The location of the placenta at or near the internal opening of the cervix. If the placenta is located near the opening of the cervix, severe bleeding can occur during delivery.

**Randomization** — The process of selection done in a way so that there is no predictable or specific pattern. In a research study, the assignment of volunteers to one test group or another is randomized to eliminate the possibility that the results will be influenced by factors not measured in the study. In the MOMS, the chances of being in either the prenatal or postnatal surgery groups are 50 percent, the same odds as getting a heads or tails when flipping a coin.

**Twin reversed arterial perfusion sequence (TRAP sequence)** — Occurs when one twin, lacking a functioning cardiac system, receives blood from the normally developing “pump twin” in a monochorionic pregnancy. This places an enormous demand on the heart, putting the pump twin at risk for cardiac failure. Left untreated, the pump twin will die in 50 to 75 percent of cases. TRAP sequence can be treated in utero using selective bipolar cord coagulation and radiofrequency ablation (RFA) procedures.

**Twin-to-twin transfusion syndrome (TTTS)** — In this syndrome, blood passes unequally between identical twins that share a placenta. Without intervention, the condition can be fatal.

**Short cervix** — The length of the cervix can be measured by sonogram. A measurement less than 20mm is considered short.

**Shunt** — A thin, flexible, straw-like device that is implanted in the fluid cavities of the brain to control the buildup of cerebrospinal fluid. Shunts most commonly run under the skin from the head to the abdominal cavity.

**Sonogram** — A test performed using sound waves that can show various internal structures. Sonograms are commonly used to examine unborn babies.

**Ventricles** — Fluid cavities within the brain. Enlargement of the ventricles with fluid is called hydrocephalus.

**References**

**Professional society guidelines/other:**


Fetal Medicine Unit, Department of Obstetrics and Gynecology and Department of Pediatric Surgery, University of São Paulo, Medical School Hospital, Brazil. Web site.


Peer-reviewed references:


Baskin LS, Ozcan T. Overview of antenatal hydronephrosis. UpToDate [online serial]. Waltham, MA: UpToDate; reviewed February 2014.


Hedrick HL, Adzick NS. Congenital diaphragmatic hernia: Prenatal diagnosis and management. UpToDate [online serial]. Waltham, MA: UpToDate; reviewed February 2015.

Holland MG, Mastrobattista JM, Lucas MJ. Diagnosis and management of twin reversed arterial perfusion (TRAP) sequence. UpToDate [online serial]. Waltham, MA: UpToDate; reviewed February 2014.


Clinical trials:

Searched clinicaltrials.gov on November 20, 2015 using terms fetal surgery in utero | Open Studies. 12 studies found, 2 relevant.


Additional resource information on clinical trials is at http://clinicaltrials.gov/info/resources.

CMS National Coverage Determinations (NCDs):

No NCDs identified as of the writing of this policy.

Local Coverage Determinations (LCDs):

No LCDs identified as of the writing of this policy.

Commonly submitted codes

Below are the most commonly submitted codes for the service(s)/item(s) subject to this policy. This is not an exhaustive list of codes. Providers are expected to consult the appropriate coding manuals and bill accordingly.

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<thead>
<tr>
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<th>Comments</th>
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21
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<th>ICD-10 Code</th>
<th>Description</th>
<th>Comments</th>
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<td>Amniocentesis; therapeutic amniotic fluid reduction (includes ultrasound guidance)</td>
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<td>59072</td>
<td>Fetal umbilical cord occlusion, including ultrasound guidance</td>
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<td>Fetal fluid drainage (eg, vesicocentesis, thoracocentesis, paracentesis), including ultrasound guidance</td>
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<td>59076</td>
<td>Fetal shunt placement, including ultrasound guidance</td>
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<td>59897</td>
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<td>Note: Covered when used to report fetoscopic laser therapy for the treatment of twin-to-twin syndrome</td>
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<td>Repair, extralobar pulmonary sequestration in the fetus, procedure performed in utero</td>
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<td>Fetoscopic laser therapy for treatment of twin-to-twin transfusion syndrome</td>
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