

Medical Policy

Subject:	Fetal Surgery for Prenatally Diagnosed Malformations	Publish Date:	12/29/2021 <u>2/24/2022</u>
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Description/Scope

This document addresses the use of surgical techniques to correct or treat fetal malformations in utero. This document does not address surgery to correct placental or uterine abnormalities including, but not limited to, amnioreduction or laser coagulation therapy to address interfetal transfusion syndrome.

Position Statement

Medically Necessary:

- A. Fetal surgery is considered **medically necessary** for vesico-amniotic shunting as a treatment of urinary tract obstruction in fetuses when **all** the following conditions are met:
1. Bilateral obstruction; **and**
 2. Evidence of progressive oligohydramnios; **and**
 3. Adequate renal function reserves; **and**
 4. No other lethal or chromosomal abnormalities.
- B. Fetal surgery is considered **medically necessary** for fetuses at 32 weeks gestation, or less, with evidence of fetal hydrops, placentomegaly, or the beginnings of severe pre-eclampsia (for example, the maternal mirror syndrome) in the mother, for the following procedures:
1. Either open or in-utero resection of malformed pulmonary tissue or placement of a thoraco-amniotic shunt as a treatment of either congenital cystic adenomatoid malformation or extralobar pulmonary sequestration;
or
 2. In-utero removal of sacrococcygeal teratoma.

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C. Fetal surgery is considered **medically necessary** for repair of myelomeningocele when **all** the following conditions are met:

1. Singleton pregnancy; **and**
2. Myelomeningocele with the upper boundary of the lesion located between T1 and S1; **and**
3. Evidence of hindbrain herniation; **and**
4. Gestational age of 19.0 to 25.9 weeks; **and**
5. Normal fetal karyotype; **and**
6. Absence of **all** of the following:
 - a. Fetal anomaly unrelated to the myelomeningocele; **and**
 - b. Severe fetal kyphosis; **and**
 - c. Short cervix (less than or equal to 15 mm); **and**
 - d. Previous pre-term birth; **and**
 - e. Placental abruption; **and**
 - f. Maternal Body Mass Index (BMI) greater than or equal to 35 kg/m²; **and**
 - g. Contraindications to surgery, including but not limited to previous hysterotomy in the active (upper) uterine segment.

D. Fetal surgery to perform fetoscopic endoluminal tracheal occlusion (FETO) is considered **medically necessary** in fetuses with pulmonary hypoplasia due to severe isolated congenital diaphragmatic hernia (CDH) when **all** the following conditions are met:

1. Singleton pregnancy; **and**
2. Gestational age less than 29 weeks and 6 days; **and**
3. Congenital diaphragmatic hernia on the left side with no other major structural or chromosomal defects; **and**
4. Severe hypoplasia, defined as a quotient of the observed-to-expected lung-to-head ratios of less than 25.0%, irrespective of liver position; **and**
5. Absence of **all** of the following:
 - a. Maternal contraindications to fetoscopic surgery or severe medical conditions that would make fetal intervention risk full; **and**
 - b. Technical limitations precluding fetoscopic surgery including, but not limited to, severe obesity (maternal BMI greater than or equal to 35 kg/m²), or uterine fibroids; **and**
 - c. Short cervix (less than or equal to 15 mm); **and**
 - d. Müllerian anomalies; **and**

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e. Placenta previa.**Investigational and Not Medically Necessary:**

~~A. Fetal surgery to induce temporary tracheal occlusion as a treatment of congenital diaphragmatic hernia (CDH) is considered **investigational and not medically necessary**.~~

~~B.A.~~ Fetal surgery is considered **investigational and not medically necessary** for the conditions indicated above when medically necessary criteria are not met.

~~C.B.~~ All other applications of fetal surgery including, but not limited to, aqueductal stenosis are considered **investigational and not medically necessary**.

Rationale*Fetal Urinary Tract Obstruction*

Few cases of prenatally diagnosed urinary tract obstruction require prenatal intervention. However, bilateral obstruction is often associated with serious disease such as pulmonary hypoplasia secondary to oligohydramnios. Therefore, fetuses with bilateral obstruction, oligohydramnios, adequate renal function reserve, and no other lethal or chromosomal abnormalities may be candidates for fetal surgery. The most common surgical approach is vesico-amniotic shunting by means of shunt or stent placement. The shunting procedure bypasses the obstructed urinary tract, permitting fetal urine to flow into the amniotic space. Small case series have shown improved survival associated with fetal surgery (Freedman, 1999; Sutton, 1999). Long-term follow-up shows that significant proportions of survivors (50%–60%) do not have normal renal function (Biard, 2005; McLorie, 2001). A randomized controlled trial published in 2013 by Morris involved 31 subjects who were assigned to either surgical intervention (n=16) or standard care (n=15). The study was closed prematurely due to poor enrollment. Intrauterine death occurred in 1 fetus in each group and pregnancy termination occurred in 3 surgical and 2 control subjects. Survival to 28 days was reported in 8 surgical and 4 control subjects. In the as-treated analysis, this difference was statistically significant (p=0.03), however, in the intent-to-treat analysis this difference was not statistically significant (relative risk [RR] 1.88, p=0.27). The authors reported substantial short-term and long-term morbidity in both groups, with only 2 babies, both from the surgical group, surviving to 2 years with normal renal function. The authors concluded that, survival appeared higher in the surgical group, but the magnitude and direction of the

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benefit is unclear and remains to be elucidated, and the likelihood of newborn survival with normal renal function is very low regardless of treatment choice.

Congenital *lower* urinary tract obstruction (LUTO) is typically secondary to a number of other possibility conditions including posterior urethral valves (PUV) and urethral atresia. LUTO is the leading cause of pediatric end-stage kidney disease and reportedly has been associated with a mortality rate as high as 45%. The most commonly performed antenatal treatments include serial ultrasound-directed vesicocentesis, vesico-amniotic shunting, fetal cystoscopy and valve ablation. Sacconne and colleagues (2020) conducted a meta-analysis to evaluate the effectiveness of antenatal intervention for the treatment of LUTO in improving perinatal survival and postnatal renal function by evaluating 10 articles with a total of 355 fetuses. Overall survival was higher in the vesico-amniotic shunt group compared to the conservative group (Odds Ratio [OR]=2.54, 95% confidence interval [CI], 1.14 to 5.67). 64/112 fetuses (57.1%) survived in the vesico-amniotic shunt group compared to 52/134 (38.8%) in the control group. A total of 5 out of the 10 studies reported on postnatal renal function between 6 months and 2 years of life. Postnatal renal function was higher in the vesico-amniotic shunt group compared to the conservative group (OR=2.09, 95% CI, 0.74 to 5.9). Data from 2 studies reported results of 45 fetuses who underwent fetal cystoscopy; perinatal survival was higher in the cystoscopy group compared to the conservative management group (OR=2.63, 95% CI, 1.07 to 6.47). Normal renal function was noted in 13/34 fetuses in the cystoscopy group versus 12/61 in the conservative management group at 6 month follow-up (OR=1.75, 95% CI 1.05 to 2.92). From this meta-analysis, antenatal bladder drainage appears to improve perinatal survival and other important clinical outcomes in cases of LUTO. The study authors conclude, “Further randomized trials with long-term follow-up are required to determine the role of antenatal treatment in clinical setting.”

Congenital Cystic Adenomatoid Malformation (CCAM) and Extralobar Pulmonary Sequestration (EPS)

CCAM and EPS are the two most common congenital cystic lung lesions. When associated with fetal hydrops before 32 weeks gestation, the survival is poor. These individuals may be candidates for prenatal surgical resection of a large mass or placement of a thoraco-amniotic shunt for a large unilocular cystic lesion. Small case series report that prenatal intervention has resulted in a greater than 50% survival rate (Adzick, 1998; Adzick, 2003a; Adzick, 2003b).

Sacroccygeal Teratoma (SCT)

SCT is both a neoplasm with the power of autonomous growth and a malformation made up of multiple tissues foreign to the region of origin and lacking organ specificity. It is the most common tumor of the newborn. Postnatal

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SCT carries a good prognosis with morbidity and mortality determined largely by extent of local disease and malignant potential. However, in utero fetal mortality has approached 100% when SCT is associated with fetal hydrops, which is related to high output heart failure secondary to arteriovenous shunting through the tumor. While the published literature is minimal, given the rarity of the condition, small case series have reported that in utero surgery may result in prenatal resolution of hydrops, healthy long-term survival, and normal development (Adzick, 2003b; Hedrick, 2004; Kamata, 2001). These results are impressive given the near-certain fetal mortality if fetal hydrops is left untreated.

Congenital Diaphragmatic Hernia (CDH)

CDH is a defect that permits abdominal viscera to enter the chest cavity, frequently resulting in hypoplasia of the lungs. CDH can vary widely in severity, depending on the size of the hernia and the timing of herniation. For example, late herniation after 25 weeks of gestation may be adequately managed postnatally. In contrast, liver herniation into the chest prior to 25 weeks of gestation is associated with a poor prognosis; fetuses in which this condition occurs have been considered candidates for fetal surgery. Temporary tracheal occlusion has been investigated as a technique to prevent the normal efflux of fetal lung fluid, which in turn enhances positive pressure in the growing lungs, promoting lung growth and ultimately reducing abdominal viscera back into the abdominal cavity. However, a randomized study comparing tracheal occlusion with conventional postoperative care was terminated early due to lack of improvement in survival or morbidity rates (Harrison, 2003). Jani and colleagues (2009) reported on a case series study involving 210 fetuses with CDH treated with fetoscopic endoluminal tracheal occlusion (FETO). This study reported a high number of prelabor rupture of membranes (47.1%) and preterm deliveries (30.9%). Out of 210 cases, 204 (97.1%) babies were live births, and 98 (48.0%) were discharged from the hospital alive. A total of 10 deaths were reported being directly related to removal of the occlusal device. There was no comparison group used in this study.

[Deprest and colleagues \(2021b\) published the results of a randomized open-label trial comparing the effects on postnatal survival of FETO performed at 27 to 29 weeks of gestation to expectant care in a group of participants carrying singleton fetuses with severe isolated CDH on the left side. The study inclusion criteria were a maternal age of 18 years or more, singleton pregnancy, gestational age at randomization of less than 29 weeks 6 days, congenital diaphragmatic hernia on the left side with no other major structural or chromosomal defects, and severe pulmonary hypoplasia, defined as a quotient of the observed-to-expected lung-to-head ratios of less than 25%, irrespective of liver position. The exclusion criteria were maternal conditions that would make fetal surgery risky, technical limitations precluding fetal surgery \(including those caused by severe maternal obesity or uterine fibroids\), and an elevated risk of preterm birth \(cervical length < 15 mm, müllerian anomalies, or placenta previa\).](#)

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The primary outcome was infant survival to discharge from the neonatal intensive care unit (NICU). The investigators used a group-sequential design with five prespecified interim analyses for superiority. Fetoscopic placement of a tracheal balloon was carried out at 27 weeks 0 days to 29 weeks 6 days gestation. Reversal of occlusion, either by fetoscopy or by ultrasound-guided puncture of the balloon, was scheduled at 34 weeks 0 days to 34 weeks 6 days gestation. Participants assigned to FETO agreed to live near the FETO center for the duration of the tracheal occlusion.

Deprest and colleagues (2021b) published the results of a randomized open label trial comparing the effects on postnatal survival of FETO performed at 27 to 29 weeks of gestation to expectant care in a group of women carrying singleton fetuses with severe isolated CDH on the left side. The primary outcome was infant survival to discharge from the neonatal intensive care unit (NICU). The investigators used a group sequential design with five prespecified interim analyses for superiority. The trial was stopped early for efficacy after the third interim analysis. The results included 80 participants, 40 in each group. A total of 16 of 40 infants (40%) in the FETO group and 6 of 40 infants (15%) in the expectant care group survived to discharge from the NICU (relative risk, 2.67; 95% CI, 1.22 to 6.11; p=0.009). In the intention to treat analysis, one neonatal death occurred after emergency delivery because of fetoscopic placental laceration from balloon removal, and one neonatal death occurred due to problems associated with balloon removal. Survival to 6 months of age was identical to the survival to discharge from the NICU (relative risk, 2.67; 95% CI, 1.22 to 6.11). Preterm, prelabor rupture of membranes occurred in 19 of 40 women (47%) in the FETO group and in 4 of 38 (11%) in the expectant care group. Preterm birth occurred in 30 of 40 women (75%) in the FETO group and in 11 of 38 women (29%) in the expectant care group. There was 1 case of placental abruption in each group. Tracheomalacia was diagnosed at 10 months of age in 1 infant. The same child, still dependent on oxygen at 3 years of age, had two cardiac operations for a ventricular septal defect that was not detected before birth, and assisted ventilation for 240 days. Additionally, the investigators identified five spontaneous balloon deflations which could potentially compromise any intended therapeutic effect. The authors note that because the trial involved experienced fetal surgery units, the findings should not be generalized to centers without extensive experience in fetoscopy and FETO or to centers that cannot ensure availability of a team that can perform safe and effective balloon retrieval. Further study is needed to assess the longer term outcomes after FETO as compared with expectant care for severe congenital diaphragmatic hernia.

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30 of 40 women (75%) in the FETO group and in 11 of 38 women (29%) in the expectant care group (relative risk, 2.59; 95% CI, 1.59 to 4.52). The median gestational age at delivery was 34 weeks 4 days and 38 weeks 3 days in the FETO and expectant care group, respectively, and the median birth weight in the FETO group was 481 g lower than that in the expectant care group.

There were no obvious between-group differences in the incidence of adverse neonatal outcomes (Deprest, 2021b). No maternal complications occurred during FETO. There was 1 case of placental abruption in each group. In the FETO group, there was one case of procedure-related fetoscopic placental laceration from balloon removal that resulted in neonatal death during resuscitation. One participant, who moved away from the FETO center, presented to her local unit at 33 weeks 6 days gestation in preterm labor and with intact membranes; postnatal puncture was unsuccessful and resulted in neonatal death. Additionally, the investigators identified five spontaneous balloon deflations which could potentially compromise any intended therapeutic effect. Tracheomalacia was diagnosed at 10 months of age in 1 infant. The same child, still dependent on oxygen at 3 years of age, had two cardiac operations for a ventricular septal defect that was not detected before birth, and assisted ventilation for 240 days. An adverse event of significant concern is an inability to remove the balloon, leading to rapid neonatal death. Deprest and colleagues (2021b) note that this is more likely to occur if the balloon removal becomes an emergency, rather than a planned procedure. Balloons were removed in nine fetuses at a non-FETO center by an inexperienced team and the removal was problematic in three of the fetuses. In the trial, participants assigned to FETO agreed to live near the FETO center for the duration of tracheal occlusion. If preterm birth was imminent, emergency balloon retrieval was performed in utero, at the time of delivery while the umbilical cord still connected the infant to the placenta, or by direct puncture immediately after delivery. The authors further emphasize that because the trial involved experienced fetal surgery units, the findings should not be generalized to centers without extensive experience in fetoscopy and FETO or to centers that cannot ensure availability of a team that can perform safe and effective balloon retrieval. Ideally, FETO should be performed at a center with a dedicated, multidisciplinary Fetal Care Team that is equipped to provide support in the medical, social, and ethical considerations of undergoing FETO procedures. The results of this study are promising. Further study is needed to evaluate the impact of FETO on longer-term health outcomes as compared with expectant care for severe congenital diaphragmatic hernia.

In another randomized open-label trial, Deprest and colleagues (2021a) compared the effects of FETO performed at 30 to 32 weeks of gestation to expectant care in women carrying singleton fetuses with moderate isolated congenital diaphragmatic hernia on the left side. The primary outcomes were infant survival to discharge from a NICU and survival without oxygen supplementation at 6 months of age. The trial was not stopped early for superiority. In an intention-to-treat analysis, 62 of 98 infants in the FETO group (63%) and 49 of 98 infants in the expectant care group (50%) survived to discharge (relative risk, 1.27; 95% CI, 0.99 to 1.63; two-sided $p=0.06$). The

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percentages of infants who survived without oxygen supplementation at 6 months of age were 54% (53 of 98 infants) for the FETO group and 44% (43 of 98 infants) for the expectant care group. Two unexplained fetal deaths occurred in the FETO group and one in the expectant care group. The incidence of preterm, prelabor rupture of membranes was 44% in the FETO group and 12% in the expectant care group. The incidence of preterm birth was 64% for the FETO group and 22% for the expectant care group. There were two problematic balloon removals, one of which result in death of the infant. The results did not show a significant increase in survival of infants to NICU discharge or a reduction in the need for oxygen supplementation at 6 months of age among infants assigned to FETO. Further study is needed to assess potential strategies to reduce FETO-associated complications, treatment criteria, and longer-term outcomes [in individuals with moderate isolated CDH on the left side](#).

In a retrospective case series study, 28 fetuses with CDH and intrafetal fluid effusions and severe pulmonary hypoplasia had fetoscopic endoluminal tracheal occlusion (FETO) attempted (Van Mieghem, 2012). A total of 21 subjects had CDH as their only complication, while the remaining 7 cases had additional congenital anomalies. A total of 3 pregnancies were terminated early and 19 cases underwent technically successful FETO, which was the sole intervention in 13 cases. No FETO was done in 5 cases. Thoracic drainage procedures were performed in 6 subjects. Postoperatively, 16 cases had stable effusions, and 3 had progressive ascites, hydrothorax and subcutaneous edema. No cases of fetal death were reported. Neonatal survival in the 25 remaining cases was 36% (n=9). Survival was similar in the FETO group (6/15) and no FETO group (2/5) (p=1.0).

A single-center randomized controlled trial (RCT) conducted in Brazil by Ruano and colleagues (2012) involved 38 subjects with CDH. Fetuses had no other detectable anomalies, fetal lung-to-head ratio < 1.0 and sonographic evidence of at least one-third of the fetal liver herniated into the thoracic cavity. Subjects were randomized to undergo either FETO (n=20) or standard postnatal care (n=21). One subject in the FETO group and 2 in the control declined the assigned treatment; all 3 fetuses subsequently died before delivery. Balloon rupture was reported in two FETO cases, but a second balloon was placed with no subsequent related adverse events reported. Premature rupture of membranes at both < 32 and < 37 weeks was not different between groups. While gestational age at delivery was significantly earlier in the FETO group (p<0.01), overall, there were no differences in rates of prematurity between groups. In the received-treatment analysis, 10/19 (52.5%) of the infants in the FETO group and 1/19 (5.3%) of the infants in the control group survived (p<0.01). In the intent-to-treat analysis, 50% (10/20) in the FETO group survived to 6 months (primary outcome measure) while only 4.8% (1/21) in the control group survived to at least 6 months (p<0.01). Severe pulmonary hypertension was noted in 50% (10/20) of the FETO group and in 85.7% (18/21) of the control group (p=0.02). In the received-treatment analysis, 12 FETO subjects and 4 controls received postnatal surgical repair (81% received a prosthetic patch). While these results are promising, additional multi-center studies with larger sample sizes are needed.

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A report by AHRQ (2011) similarly concluded that the bulk of CDH literature is comprised of case reports, and long-term outcomes are not well reported regarding the survival rates of fetuses treated with tracheal occlusion relative to infants treated at birth. [Despite the scarcity of literature, there is growing support in the form of RCTS for the use of FETO demonstrating a positive impact on survival in a carefully selected group of individuals with severe isolated CDH.](#)

Myelomeningocele

Myelomeningocele is a neural tube defect in which the spinal cord and its coverings protrude through the skin in the lower back. Children with this disorder have varying degrees of neurologic impairment to the legs and bowel and bladder function, brain malformation, and disorders of cerebrospinal fluid circulation. Traditional treatment of the condition consists of surgical repair after term delivery. Surgical repair to the fetus has been proposed as a means of improving neurologic function and decreasing the incidence of other problems related to the condition. Two case series reporting short-term outcomes up to 6 months have shown improvement in anatomic hindbrain herniation and a lower incidence of hydrocephalus requiring ventriculoperitoneal shunt (Adzick, 2003b; Tulipan, 1999). A study reporting leg function at longer follow-up showed no difference between individuals treated with fetal surgery versus traditional surgery (Tulipan, 1999). Both case series show that the incidence of premature delivery is increased in the fetal surgery groups (Bruner, 1999; Sutton, 1999). A systematic review of 11 studies comprised of 518 women who received fetal myelomeningocele repair, reported an average overall rate of maternal and obstetric complications of 78.6%, the majority being obstetric complications, including chorioamniotic membrane separation (65.6%), oligohydramnios (13.0%), placental abruption (5.0%), spontaneous or preterm premature membrane rupture (42.0%), and early preterm delivery (11.3%). The most common medical complications were pulmonary edema (2.8%), gestational diabetes (3.7%), preeclampsia (3.7%), and need for blood transfusions (3.2%). Authors emphasize that overall lack of published data on maternal and obstetric complications related to myelomeningocele repair and highlight that “maternal health hazard will continue to be an issue of crucial importance and further studies are required.” (Licci, 2019).

Adzik and colleagues in the Management of Myelomeningocele Study (MOMS) group published the results of a large RCT that included 138 subjects who completed up to 30 months follow-up (2011). This study randomized subjects to receive myelomeningocele repair either in utero (fetal surgery group) or following delivery (perinatal group). The first primary outcome, fetal death or the need for cerebrospinal fluid shunt by the age of 12 months was significantly better in the fetal surgery group with 68% vs. 98% of subjects requiring shunts, respectively (p<0.001). The rate of actual shunt placement was 40% for the in utero group vs. 82% in the perinatal group.

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Fetal Surgery for Prenatally Diagnosed Malformations

Additionally, at 12 months of age, the fetal surgery group demonstrated significantly fewer infants with any evidence of hindbrain herniation (64% vs. 96%, $p<0.001$), brain stem kinking (20% vs. 46%, $p<0.001$), abnormal fourth ventricle location (46% vs. 72%, $p<0.002$), and syringomyelia (39% vs. 58%, $p<0.03$). The main secondary outcome was a composite score 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 fetal surgery group (mean 148.6 vs. mean 122.6, $p<0.007$). In the post hoc analysis, the authors reported that subjects in the fetal 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 also more likely to ambulate without orthotics or other devices (42% vs. 21%, $p<0.01$). Interestingly, the subjects in the fetal surgery group had significantly better motor function scores on the Bayley and Peabody motor scales, despite having more severe anatomical lesion levels at baseline. There were no significant differences in cognitive function between groups. The report also stated that the fetal surgery group had significantly higher rates of pre-term birth (79% vs. 15%), spontaneous membrane rupture (46% vs. 8%, $p<0.001$), oligohydramnios (21% vs. 4%, $p=0.001$), and maternal transfusion (9% vs. 1%, $p=0.03$). Evaluating this data, it is clear that there appears to be potentially significant benefits to the fetus with fetal surgery for myelomeningocele. However, these benefits must be balanced with the possible risks of maternal and pregnancy complications. Prompted by publication of the MOMS trial, the fetal myelomeningocele Maternal-Fetal Management Task Force (convened by the Eunice Kennedy Shriver National Institute of Child Health and Human Development) recently published a position statement on optimal practice criteria for facilities and practitioners performing this procedure (Cohen, 2014). A post-hoc analysis of 30-month cohort data from the MOMS trial, validated that cognitive and motor function outcomes favored in-utero repair of myelomeningocele over postnatal procedures (Farmer, 2018). Based on the MOM's study, the American College of Obstetricians and Gynecologists (ACOG)'s practice bulletin on Neural Tube Defects includes a recommendation for fetal surgery for myelomeningocele in eligible women and fetuses (ACOG, 2017).

In 2020, Houtrow and colleagues published a long-term study of 161 school-aged (5.9-10.3 years) children from the original MOMS trial who were evaluated by blinded examiners to determine neuropsychological and physical difference. Long-term benefits of fetal surgery identified included improved mobility and independent functioning in addition to fewer surgeries for shunt placement and revision. There was no difference detected in cognitive functioning between the two cohorts. Similarly, Brock and colleagues (2019) evaluated long-term outcomes of 156 children (mean age 7.4 years) from the MOMS trial cohorts to determine differences in urological outcomes. Overall, 62% vs 87% in the prenatal and postnatal surgery groups, respectively, were placed on clean intermittent catheterization (RR=0.71, 95% CI 0.58-0.86, $p<0.001$). There was a significant difference between the groups in voiding status as 24% in the prenatal group vs 4% in the postnatal group were reported to be voiding without catheterization (RR=5.8, 95% CI 1.8-18.7; $p<0.001$). Despite more favorable urologic outcomes in the prenatal

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surgery group, the study authors emphasize that “urological outcomes alone should not be the sole impetus to perform in utero closure in children with spina bifida.”

In 2021, the Society of Obstetricians and Gynaecologists of Canada (Wilson, 2021) issued guidance on the pregnancy management for fetal neural tube defects. They included the following recommendation which they rated as strong with a high quality of evidence:

Once an isolated open or closed neural tube defect is detected, and diagnostic and genetic testing results (if applicable) are available, families should be offered a choice of 3 obstetrical care management options. In the absence of specific contraindications, families should be given information about the following options: prenatal surgical repair of myelomeningocele and prognosis, postnatal surgical repair of myelomeningocele and prognosis, and pregnancy termination with autopsy (strong, high).

Aortic Stenosis

Fetal aortic stenosis is a major prenatal complication that has been known to lead to hydrops, hypoplastic left heart syndrome (HLHS) and other conditions that lead to considerable morbidity and mortality. Additionally, this condition is often seen in conjunction with other congenital abnormalities such as mitral valve dysfunction. In an attempt to address these problems, intrauterine fetal interventions have been developed and investigated to try and address them early in their natural history. The available evidence is limited to small non-randomized studies.

In a retrospective study, Selamet Tierney and colleagues (2007) reported the effects of mid-gestation fetal balloon aortic valvuloplasty on subsequent fetal left ventricular function. A total of 42 fetuses had attempted aortic valvuloplasty, 12 were excluded from analysis due to inadequate follow-up data, pregnancy termination or fetal demise. Study fetuses (n=30) had pre-procedure echocardiography at a median gestational age of 23 weeks and were followed for a median of 66 ± 23 days post-intervention. In 26 of the 30 study fetuses, aortic valvuloplasty was reported to be technically successful. A control group was used which included the 4 fetuses that underwent technically unsuccessful aortic valvuloplasty plus 14 control fetuses that did not undergo the intervention. Left ventricular ejection fraction increased from $19 \pm 10\%$ pre-intervention to $39 \pm 14\%$ post-intervention ($p < 0.001$). Other improvements in Doppler cardiac characteristics were also noted in the study group, but not in the control group. The authors caution that further study is necessary to determine if changes in left heart physiology after in-utero aortic valvuloplasty can be used to predict postnatal outcome.

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The largest published case series to date describes the outcomes of aortic valvuloplasty in 70 fetuses with severe aortic stenosis and evolving HLHS (McElhinney, 2009). The procedure was reported to be successful in 52 (74%) cases. In 20 subjects with significant aortic regurgitation following valvuloplasty, all but 1 had resolution. The remaining subject had persistent regurgitation through the newborn evaluation. Within the study group, 1 pregnancy was terminated and 8 (13%) did not reach a viable term or preterm birth. Another 6 pregnancies ended in fetal death or preterm stillbirth, another 2 were live births, but were nonviable due to extreme prematurity. Of the remaining 61 subjects, 5 were delivered preterm, 54 were delivered at term, and 2 were still in utero at time of publication. Overall, 67% of subjects had a technically successful valvuloplasty and were born viable. In the group of 14 subjects with unsuccessful procedures but born at a viable age, 11 (78.6%) were alive at time of publication. At the time of birth, 15 subjects with successful interventions had biventricular circulation (30%). The remaining 30 subjects underwent further procedures postnatally, and 6 died. Only 2 of the remaining 24 subjects converted to biventricular circulation. Larger left heart structures and higher left ventricular pressure at the time of the aortic valvuloplasty were associated with a biventricular outcome postpartum.

Artz and colleagues provided data from a small case series in Austria of 23 fetal subjects with critical aortic stenosis (2011). Aortic valvuloplasty was attempted in all subjects, with success reported in 16 (69.6%); 15 of these procedures were judged to be technically successful with 1 intrauterine death in this group. In 8 fetuses the intervention was not successful, with intrauterine fetal death in 2 of these cases. In 10 of the 15 successfully-treated and live born fetuses, a biventricular circulation was achieved postnatally. Complications included severe sustained bradycardia requiring intracardiac treatment with epinephrine. Other complications included hemopericardium, left ventricular thrombus, and balloon tear off. At median follow-up of 27 months, 40% of the 10 newborns that achieved biventricular circulation, required only AV balloon dilatation in the first prenatal week and no further surgery. Although in this small series, fetal aortic valvuloplasty was performed successfully in two-thirds of selected fetuses with critical aortic stenosis, further study of the long-term outcome is needed to judge the permanent function of the left ventricle after this intrauterine intervention.

Overall, the current body of literature addressing the use of intrauterine fetal balloon valvuloplasty for the treatment of aortic stenosis is insufficient to allow a complete assessment of the safety and efficacy of this procedure. Significant questions still exist regarding optimal selection criteria as well as the actual benefit of the procedure itself.

Background/Overview

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Fetal Surgery for Prenatally Diagnosed Malformations

Description of Birth Defects

Birth defects, also referred to as congenital anomalies, are abnormalities of structure, function, or body metabolism that are present at the time of birth and frequently lead to mental or physical disabilities. Some birth defects are fatal. There are more than 4000 known birth defects ranging from minor to serious, and although many of them can be treated or cured, they are the leading cause of death in the first year of life. Birth defects can be caused by genetic, environmental, or unknown factors. According to the March of Dimes, about 150,000 babies are born with birth defects each year in the United States. The American College of Obstetricians and Gynecologists (ACOG) says that out of every 100 babies born in the United States, 3 have some kind of major birth defect.

Most fetal anatomic malformations are best managed after birth. However, advances in methods of prenatal diagnosis, particularly prenatal ultrasound, have led to a new understanding of the progression and outcomes of certain congenital anomalies. With these advances in diagnostic technology and understanding come additional improvements in surgical and medical procedures to treat birth defects earlier to improve outcomes. Perhaps one of the most significant advances in this field is fetal surgery, which allows doctors to address anatomical abnormalities early in the fetal development process with the goal of lessening significant problems due to the particular abnormality in question.

Description of Fetal Surgery

Fetal surgery involves opening a mother's uterus with either a traditional Cesarean surgical incision or through single or multiple incisions through which laparoscopic tools are inserted. When a Cesarean surgical approach is used the fetus is exposed in the maternal abdomen, the fetal abnormality is surgically corrected, and the fetus is returned to the uterus, which is then closed to allow the pregnancy to continue. When a laparoscopic approach is taken, the surgery takes place inside the uterus with the assistance of specialized tools, including a video camera. This is proposed to be a less traumatic method but can only be used under certain circumstances and requires a high degree of surgical skill and experience.

While fetal surgery has been researched as a treatment method for many different abnormalities, only a few conditions have been shown to have improved outcomes when compared to traditional postnatal therapy. Bilateral urinary tract obstruction is often associated with the development of serious health problems including lung disease and loss of amniotic fluid in the uterus. The most common surgical approach is called vesico-amniotic shunting. The shunting procedure bypasses the obstructed urinary tract, permitting fetal urine to flow into the amniotic space decreasing the potential for adverse outcomes. [SCT associated with fetal hydrops results in near-certain fetal](#)

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mortality if left untreated. In this case, fetal surgery to remove the tumor may result in resolution of hydrops and increased likelihood of long-term survival. In CDH, intrathoracic herniation of the abdominal viscera impairs normal airway and pulmonary vascular development. FETO has been associated with increased survival among infants with severe pulmonary hypoplasia due to isolated CDH on the left side. Myelomeningocele is a neural tube defect associated with varying degrees of neurological impairment. Surgical repair to the fetus has been proposed to prevent progressive damage during gestation, improve neurological function, and decrease the incidence of associated problems.

Definitions

Aqueductal stenosis: The most common cause of congenital hydrocephalus, a condition caused by a narrowing of the Aqueduct of Sylvius which is a space that connects the third and fourth ventricles of the brain and allows the flow of cerebrospinal fluid.

Body Mass Index (BMI): Weight in kilograms divided by the square of the height in meters.

Congenital cystic adenomatoid malformation: A benign (non-cancerous) mass of abnormal lung tissue, usually located in one lobe of the lung. This condition is caused by overgrowth of abnormal lung tissue that may form fluid filled cysts that do not function as normal lung tissue.

Congenital diaphragmatic hernia: A condition where there is a hole in the diaphragm, the muscle that separates the chest and abdominal cavities, through which the organs in the abdomen pass through and compress the lungs and heart.

Extralobar pulmonary sequestration: A condition where a portion of a normal lung lacks the usual connections to the rest of the lung, including air passages and blood flow. If uncorrected this portion of the lung cannot perform normal respiratory functions.

Fetal hydrops: A condition characterized by the presence of a generalized collection of extra fluid under the skin of a fetus and in one or more body cavities.

Fetal surgery: A surgical procedure performed on an unborn child while still in the mother's uterus, to correct congenital deformities that have significant health impact.

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Hindbrain herniation: A condition where a posterior portion of the brain inappropriately protrudes through the spinal cord opening in the base of the skull (the foramen magnum). This condition is referred to as a “Chiari Malformation” (CM) and may be classified as Type I, Type II or Type III. Type I involves the extension of the cerebellar tonsils (the lower part of the cerebellum) into the foramen magnum, without involving the brain stem. Type II, also called classic CM, involves the extension of both cerebellar and brain stem tissue into the foramen magnum. Type III is the most serious form of CM since the cerebellum and brain stem protrude through the foramen magnum and into the spinal cord.

Karyotype: The characterization of an individual's chromosomes, including number, type, shape, etc.

Kyphosis: An abnormal forward rounding of the upper portion of the spine. Individuals with this condition may appear to have a “hump.”

Maternal mirror syndrome: The occurrence of fetal and placental hydrops with maternal pre-eclampsia; it is called “mirror syndrome” because the edema in the mother mirrors that in the fetus (also known as Mirror Syndrome or Ballantyne’s Syndrome).

Myelomeningocele: A condition where the backbone and spinal canal of a fetus do not close before birth. This can result in the spinal cord and its covering membranes protruding out of the infant's back. This is the most common cause of spina bifida. Nearly all individuals with myelomeningocele have a Chiari II malformation.

Oligohydramnios: A condition characterized by too little amniotic fluid within the amniotic sack surrounding the fetus. Certain birth defects such as kidney and urinary tract problems and ruptured membranes are the most common cause for this condition.

Placental abruption: A condition where a placenta either partially or completely peels away from the inner wall of the uterus before delivery.

Placentomegaly: A condition where the mother’s placenta is abnormally enlarged.

Pre-eclampsia: Also referred to as toxemia, a condition characterized by maternal high blood pressure, swelling, high concentrations of protein in maternal urine; this condition may interfere with adequate blood supply to the fetus.

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Sacroccygeal teratoma: A type of tumor that develops in fetuses in the lower-most portion of the back, just above the buttocks. Most frequently these are benign tumors but the odds of malignancy increases with increasing age, necessitating removal at the earliest possible opportunity.

Tracheal occlusion: A condition where the main airway to the lungs is blocked, most frequently due to diaphragmatic hernia.

Thoraco-amniotic shunt: A tube that drains the fluid from the chest into the amniotic sac. Fluid accumulation in the chest may be due to congenital cystic adenomatoid malformation or extralobar pulmonary sequestration.

Urinary tract obstruction: A condition where the flow of urine through the urinary tract is blocked.

Vesico-amniotic shunting: A treatment for fetal urinary tract obstruction, where a tube is inserted into the urinary tract above the obstruction and passed through the abdominal wall to drain into the amniotic sac.

Coding

The following codes for treatments and procedures applicable to this document are included below for informational purposes. Inclusion or exclusion of a procedure, diagnosis or device code(s) does not constitute or imply member coverage or provider reimbursement policy. Please refer to the member's contract benefits in effect at the time of service to determine coverage or non-coverage of these services as it applies to an individual member.

When services may be Medically Necessary when criteria are met:

CPT

59076

Fetal shunt placement, including ultrasound guidance

59897

Unlisted fetal invasive procedure, including ultrasound guidance, when performed [when describing a procedure meeting medically necessary criteria]

HCPCS

S2400

Repair, congenital diaphragmatic hernia in the fetus using temporary tracheal occlusion, procedure performed in utero

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

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Fetal Surgery for Prenatally Diagnosed Malformations

S2403	Repair, extralobar pulmonary sequestration in the fetus, procedure performed in utero
S2404	Repair, myelomeningocele in the fetus, procedure performed in utero
S2405	Repair of sacrococcygeal teratoma in the fetus, procedure performed in utero
S2409	Repair, congenital malformation of fetus, procedure performed in utero, not otherwise classified [when describing a procedure meeting medically necessary criteria]

ICD-10 Procedure

10Q00YE-10Q08ZE	Repair nervous system in products of conception [includes codes 10Q00YE, 10Q00ZE, 10Q03YE, 10Q03ZE, 10Q04YE, 10Q04ZE, 10Q07YE, 10Q07ZE, 10Q08YE, 10Q08ZE based on device and approach]
10Q00YK-10Q08ZK	Repair respiratory system in products of conception [includes codes 10Q00YK, 10Q00ZK, 10Q03YK, 10Q03ZK, 10Q04YK, 10Q04ZK, 10Q07YK, 10Q07ZK, 10Q08YK, 10Q08ZK based on device and approach]
10Q00YR-10Q08ZR	Repair musculoskeletal system in products of conception [includes codes 10Q00YR, 10Q00ZR, 10Q03YR, 10Q03ZR, 10Q04YR, 10Q04ZR, 10Q07YR, 10Q07ZR, 10Q08YR, 10Q08ZR based on device and approach]
10Q00YS-10Q08ZS	Repair urinary system in products of conception [includes codes 10Q00YS, 10Q00ZS, 10Q03YS, 10Q03ZS, 10Q04YS, 10Q04ZS, 10Q07YS, 10Q07ZS, 10Q08YS, 10Q08ZS based on device and approach]

ICD-10 Diagnosis

All diagnoses

When services are Investigational and Not Medically Necessary:

For the procedure codes listed above when criteria are not met; or when the code describes a procedure indicated in the Position Statement section as investigational and not medically necessary.

When services are also Investigational and Not Medically Necessary:

HCPES

S2400	Repair, congenital diaphragmatic hernia in the fetus using temporary tracheal occlusion, procedure performed in utero
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ICD-10 Procedure

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Fetal Surgery for Prenatally Diagnosed Malformations

10Q00YF-10Q08ZF	Repair cardiovascular system in products of conception [includes codes 10Q00YF, 10Q00ZF, 10Q03YF, 10Q03ZF, 10Q04YF, 10Q04ZF, 10Q07YF, 10Q07ZF, 10Q08YF, 10Q08ZF based on device and approach]
10Q00YG-10Q08ZG	Repair lymphatics and hemic in products of conception [includes codes 10Q00YG, 10Q00ZG, 10Q03YG, 10Q03ZG, 10Q04YG, 10Q04ZG, 10Q07YG, 10Q07ZG, 10Q08YG, 10Q08ZG based on device and approach]
10Q00YH-10Q08ZH	Repair eye in products of conception [includes codes 10Q00YH, 10Q00ZH, 10Q03YH, 10Q03ZH, 10Q04YH, 10Q04ZH, 10Q07YH, 10Q07ZH, 10Q08YH, 10Q08ZH based on device and approach]
10Q00YJ-10Q08ZJ	Repair ear, nose and sinus in products of conception [includes codes 10Q00YJ, 10Q00ZJ, 10Q03YJ, 10Q03ZJ, 10Q04YJ, 10Q04ZJ, 10Q07YJ, 10Q07ZJ, 10Q08YJ, 10Q08ZJ based on device and approach]
10Q00YL-10Q08ZL	Repair mouth and throat in products of conception [includes codes 10Q00YL, 10Q00ZL, 10Q03YL, 10Q03ZL, 10Q04YL, 10Q04ZL, 10Q07YL, 10Q07ZL, 10Q08YL, 10Q08ZL based on device and approach]
10Q00YM-10Q08ZM	Repair gastrointestinal system in products of conception [includes codes 10Q00YM, 10Q00ZM, 10Q03YM, 10Q03ZM, 10Q04YM, 10Q04ZM, 10Q07YM, 10Q07ZM, 10Q08YM, 10Q08ZM based on device and approach]
10Q00YN-10Q08ZN	Repair hepatobiliary and pancreas in products of conception [includes codes 10Q00YN, 10Q00ZN, 10Q03YN, 10Q03ZN, 10Q04YN, 10Q04ZN, 10Q07YN, 10Q07ZN, 10Q08YN, 10Q08ZN based on device and approach]
10Q00YP-10Q08ZP	Repair endocrine system in products of conception [includes codes 10Q00YP, 10Q00ZP, 10Q03YP, 10Q03ZP, 10Q04YP, 10Q04ZP, 10Q07YP, 10Q07ZP, 10Q08YP, 10Q08ZP based on device and approach]
10Q00YQ-10Q08ZQ	Repair skin in products of conception [includes codes 10Q00YQ, 10Q00ZQ, 10Q03YQ, 10Q03ZQ, 10Q04YQ, 10Q04ZQ, 10Q07YQ, 10Q07ZQ, 10Q08YQ, 10Q08ZQ based on device and approach]
10Q00YT-10Q08ZT	Repair female reproductive system in products of conception [includes codes 10Q00YT, 10Q00ZT, 10Q03YT, 10Q03ZT, 10Q04YT, 10Q04ZT, 10Q07YT, 10Q07ZT, 10Q08YT, 10Q08ZT based on device and approach]
10Q00YV-10Q08ZV	Repair male reproductive system in products of conception [includes codes 10Q00YV, 10Q00ZV, 10Q03YV, 10Q03ZV, 10Q04YV, 10Q04ZV, 10Q07YV, 10Q07ZV, 10Q08YV, 10Q08ZV based on device and approach]

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Fetal Surgery for Prenatally Diagnosed Malformations

10Q00YY-10Q08ZY Repair other body system in products of conception [includes codes 10Q00YY, 10Q00ZY, 10Q03YY, 10Q03ZY, 10Q04YY, 10Q04ZY, 10Q07YY, 10Q07ZY, 10Q08YY, 10Q08ZY based on device and approach]

ICD-10 Diagnosis

All diagnoses

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Congenital Cystic Adenomatoid Malformation
 Congenital Diaphragmatic Hernia
 Extralobar Pulmonary Sequestration
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FETO

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Fetal Surgery for Prenatally Diagnosed Malformations

Temporary Tracheal Occlusion
Thoraco-Amniotic Shunt
Urinary Tract Obstruction
Vesico-Amniotic Shunting

Document History

Status	Date	Action
Revised	02/17/2022	Medical Policy & Technology Assessment Committee (MPTAC) review. Clarified unit of measurement for BMI in MN criteria. Added MN criteria for FETO. Updated Rationale, Background/Overview, Coding, References, Websites and Index sections.
Reviewed	11/11/2021	Medical Policy & Technology Assessment Committee (MPTAC) MPTAC review. Updated Rationale, References and Websites sections.
Reviewed	11/05/2020	MPTAC review. Updated References and Websites sections.
Reviewed	11/07/2019	MPTAC review. Updated References and Websites sections.
Reviewed	01/24/2019	MPTAC review. Updated Rationale, Definitions and References sections.
Reviewed	01/25/2018	MPTAC review. Updated References section.
Reviewed	02/02/2017	MPTAC review. Updated References section.
Reviewed	02/04/2016	MPTAC review. Updated Rationale and Reference sections. Removed ICD-9 codes from Coding section.
Reviewed	02/05/2015	MPTAC review. Updated Rationale and Reference sections.
Reviewed	02/13/2014	MPTAC. Updated Rationale and Reference sections.
Reviewed	02/14/2013	MPTAC review. Updated Rationale and Reference sections.
Reviewed	02/16/2012	MPTAC review.
Revised	02/17/2011	MPTAC review. Added the treatment of myelomeningocele as medically necessary with criteria. Updated Rationale, Definitions, Coding and Reference sections.
Reviewed	05/13/2010	MPTAC review.
Reviewed	05/21/2009	MPTAC review.
Reviewed	05/15/2008	MPTAC review.
	02/21/2008	The phrase "investigational/not medically necessary" was clarified to read "investigational and not medically necessary." This change was approved at the November 29, 2007 MPTAC meeting.

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[This Medical Policy provides assistance in understanding Healthy Blue's standard Medicaid benefit plan. When evaluating coverage for a specific member benefit, reference to federal and state law, as well as contractual requirements may be necessary, since these may differ from our standard benefit plan. In the event of a conflict with standard plan benefits, federal, state and/or contractual requirements will govern. Before using this policy, please check all federal, state and/or contractual requirements applicable to the specific benefit plan coverage. Healthy Blue reserves the right to modify its Policies and Guidelines as necessary and in accordance with legal and contractual requirements. This Medical Policy is provided for informational purposes. It does not constitute medical advice. Healthy Blue may also use tools and criteria developed by third parties, to assist us in administering health benefits. Healthy Blue's Policies and Guidelines are intended to be used in accordance with the independent professional medical judgment of a qualified health care provider and do not constitute the practice of medicine or medical advice.](#)

Fetal Surgery for Prenatally Diagnosed Malformations

Reviewed	05/17/2007	MPTAC review.
Reviewed	06/08/2006	MPTAC review. Updated references.
Revised	07/14/2005	MPTAC review. Revision based on Pre-merger Anthem and Pre-merger WellPoint Harmonization.

Pre-Merger Organizations	Last Review Date	Document Number	Title
Anthem, Inc.	04/27/2004	SURG.00036	Fetal Surgery for Prenatally Diagnosed Malformations
WellPoint Health Networks, Inc.	12/02/2004	3.09.07	Fetal Surgery

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