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Fetal Surgery for Prenatally Diagnosed Malformations

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Disclaimer

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Each benefit plan, summary plan description or contract defines which services are covered, which services are excluded, and which services are subject to dollar caps or other limitations, conditions or exclusions. Members and their providers have the responsibility for consulting the member's benefit plan, summary plan description or contract to determine if there are any exclusions or other benefit limitations applicable to this service or supply. **If there is a discrepancy between a Medical Policy and a member's benefit plan, summary plan description or contract, the benefit plan, summary plan description or contract will govern.**

Coverage

Fetal surgery **may be considered medically necessary** for ANY of the following indications:

- Vesico-amniotic shunting as a treatment of bilateral fetal urinary tract obstruction when:
  - Evidence of hydronephrosis due to bilateral urinary tract obstruction; AND
  - Progressive oligohydramnios; AND
  - Adequate renal function; AND
  - No other lethal abnormalities or chromosomal defects.
- Open in-utero resection of malformed pulmonary tissue or placement of a thoraco-amniotic shunt when:
  - Congenital cystic adenomatoid malformation or bronchopulmonary sequestration is identified; AND
  - The fetus is at 34 weeks' gestation or less; AND
  - There is evidence of fetal hydrops, placentomegaly, and/or the beginnings of severe pre-eclampsia (i.e., the maternal mirror syndrome) in the mother.
- In-utero removal of sacrococcygeal teratoma when:
  - The fetus is at 32 weeks' gestation or less; AND

    - There is evidence of fetal hydrops, placentomegaly, and/or the beginnings of severe pre-eclampsia (i.e., maternal mirror syndrome) in the mother.
- In-utero occlusion of anastomotic vessels (e.g., laser photocoagulation, radiofrequency ablation, and ligation) for twin reversed arterial perfusion (TRAP) sequence**.**
- In-utero laser coagulation of anastomotic vessels in early (<26 weeks gestation), severe twin to twin transfusion syndrome (TTTS).
- Serial amnioreduction for TTTS after 26 weeks gestation.
- In-utero repair of myelomeningocele when:
    - The fetus is at less than 26 weeks' gestation; AND
    - Myelomeningocele is present with an upper boundary located between T1 and S1 with evidence of hindbrain herniation; AND
    - Singleton pregnancy; AND
    - Normal fetal karyotype; AND
    - Absence of **ALL** the following:
        a. Fetal anomaly unrelated to the myelomeningocele; and
        b. Severe fetal kyphosis; and
        c. Maternal 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/m^2; and
        g. Contraindications to surgery, including but not limited to previous hysterotomy in the active (upper) uterine segment.

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

- Singleton pregnancy; AND
- Gestational age less than 29 weeks and 6 days; AND
- Congenital diaphragmatic hernia (lung-to-head ratio [LHR] <1 and liver herniated into the chest) on the left side with no other major structural or chromosomal defects; AND
- Severe hypoplasia, defined as a 25% increase in the lung area to the head circumference (with or without liver herniation); AND
- Absence of **ALL** of the following:
    - Short cervix (less than or equal to 15 mm); and
    - Maternal contraindications to fetoscopic surgery or general anesthesia.

Other applications of fetal surgery **are considered experimental, investigational and/or unproven,** including but not limited to the treatment of congenital heart defects.

Policy Guidelines

None.

Description

Fetal surgery is used for specific congenital abnormalities that are associated with a poor postnatal prognosis. Prenatal surgery typically involves opening the gravid uterus (with a Cesarean surgical incision), surgically correcting the abnormality, and returning the fetus to the uterus and restoring uterine closure. Minimally invasive procedures through single or multiple fetoscopic port incisions are also being developed.

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 natural history and physiologic outcomes of certain congenital anomalies. Fetal surgery is the logical extension of these diagnostic advances, related in part to technical advancement in anesthesia, tocolysis, and hysterotomy.

Fetal Urinary Tract Obstruction
Although few cases of prenatally diagnosed urinary tract obstruction require prenatal intervention, bilateral obstruction can lead to distention of the urinary bladder and 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 decompression through percutaneous placement of a shunt or stent. Vesicoamniotic shunting bypasses the obstructed urinary tract, permitting fetal urine to flow into the amniotic space. The goals of shunting are to protect the kidneys from increased pressure in the collecting system and to assure adequate amniotic fluid volume for lung development.

Congenital Diaphragmatic Hernia
Congenital diaphragmatic hernia (CDH) results from abnormal development of the diaphragm, which permits abdominal viscera to enter the chest, frequently resulting in hypoplasia of the lungs. CDH can vary widely in severity, depending on the size and timing of hernia. For example, late herniation after 25 weeks of gestation may be adequately managed postnatally. In contrast, liver herniation into the chest before 25 weeks of gestation is associated with a poor prognosis, and these fetuses have been considered candidates for fetal surgery. Temporary tracheal occlusion using a balloon is being evaluated for the treatment of CDH. Occluding the trachea of a fetus prevents the normal efflux of fetal lung fluid, which results in a build-up of secretions in the pulmonary tree and increases the size of the lungs, gradually pushing abdominal viscera out of the chest cavity and back into the abdominal cavity. It is believed that this, in turn, will promote better lung maturation. Advances in imaging have resulted in the ability to detect less severe lesions, which has resulted in a decrease in mortality rates for defects detected during pregnancy.

Some fetuses with severe isolated CDH may be appropriate candidates for in-utero

fetoscopic endoscopic tracheal occlusion (FETO) although it should only be considered in select fetuses with a poor CDH prognosis. The goal of in-utero treatment in fetuses with severe CDH is to prevent or reverse pulmonary hypoplasia and restore adequate lung growth to improve neonatal survival. Overall, survival has improved with advances in antenatal diagnosis and neonatal care, but affected infants remain at significant risk of morbidity and mortality. (1)

Congenital Cystic Adenomatoid Malformation or Congenital Diaphragmatic Hernia Sequestration

Congenital cystic adenomatoid malformation (CCAM) also referred to as congenital pulmonary airway malformations, and bronchopulmonary sequestration (BPS) are the 2 most common congenital cystic lung lesions and share the characteristic of a segment of lung being replaced by abnormally developing tissue. CCAMs can have connections to the pulmonary tree and contain air, while BPS does not connect to the airway and has blood flow from the aorta rather than the pulmonary circulation. In more severe cases, the malformations can compress adjacent normal lung tissue and distort thoracic structure. CCAM lesions typically increase in size in mid-trimester and then in the third trimester either involute or compress the fetal thorax, resulting in hydrops in the infant and sometimes mirror syndrome (a severe form of preeclampsia) in the mother. Mortality is close to 100% when lesions are associated with fetal hydrops (abnormal accumulation of fluid in 2 or more fetal compartments). Fetuses with developing CCAMs or BPS may be candidates for prenatal surgical resection of a large mass or placement of a thoracoamniotic shunt to decompress the lesion.

Sacrococcygeal Teratoma

Sacrococcygeal Teratoma (SCT) is both a neoplasm with the power of autonomous growth and a malformation comprised of multiple tissues foreign to the region of origin and lacking organ specificity. It is the most common tumor of the newborn and generally carries a good prognosis in infants born at term. However, in-utero fetal mortality approaches 100% with large or vascular tumors, which may become larger than the rest of the fetus. In this small subset, SCT is associated with fetal hydrops, which is related to high output heart failure secondary to arteriovenous shunting. In some cases, mothers of fetuses with hydrops can develop mirror syndrome.

Myelomeningocele

Myelomeningocele is a neural tube defect in which the spinal cord forms abnormally and is left open, exposing the meninges and neural tube to the intrauterine environment. Myelomeningocele is the most common cause of spina bifida, and depending on the location, results in varying degrees of neurologic impairment to the legs, bowel and bladder function, brain malformation (i.e., hindbrain herniation), cognitive impairment, and disorders of cerebrospinal fluid circulation, i.e., hydrocephalus requiring placement of a ventriculoperitoneal shunt. Traditional treatment consists of surgical repair after term delivery, with ventriculoperitoneal shunting to prevent infection and further neurologic dysfunction. Fetal surgical repair to cover the exposed spinal canal has been proposed as a means of preventing the deleterious exposure to the intrauterine environment with the hope of improving neurologic function and decreasing the incidence of other problems related to the condition.

Due to the risks involved related to fetal surgery, it should only be performed at facilities with the special expertise, multidisciplinary teams, and facilities to provide the intensive care required for these individuals. (2)

Cardiac Malformations

In-utero interventions are being investigated for several potentially lethal congenital heart disorders, including critical aortic stenosis with evolving hypoplastic left heart syndrome (HLHS), HLHS with intact atrial septum, critical pulmonary stenosis or pulmonary atresia. (3) Critical pulmonary stenosis or atresia with intact ventricular septum is characterized by a very narrow pulmonary valve without a connection between the right and left ventricles. Pulmonary atresia with intact ventricular septum can evolve into right ventricular hypoplasia; fetal pulmonary valvuloplasty may result in biventricular circulation. Critical aortic stenosis with impending HLHS is a very narrow aortic valve that develops early during gestation and may result in HLHS, a complex spectrum of cardiac anomalies characterized by hypoplasia of the left ventricle and aorta, with atretic, stenotic, or hypoplastic atrial and mitral valves. In-utero aortic balloon valvuloplasty relieves aortic stenosis with the goal of preserving left ventricular growth and halting the progression to HLHS. HLHS with intact trial septum is a variant of HLHS that occurs in about 22% of all HLHS cases in which blood flow across the foramen ovale is restricted, leading to left atrial hypertension and damage to the pulmonary vasculature, parenchyma, and lymphatics. For HLHS with intact atrial septum, fetal balloon atrial septostomy is designed to reduce the left atrial restriction.

Twin Reversed Arterial Perfusion (TRAP) Sequence

TRAP sequence refers to a rare complication of monochorionic twin pregnancy in which one fetus has an absent or a rudimentary nonfunctioning heart (acardiac twin) and is entirely perfused in a retrograde direction by its co-twin (pump twin) via placental arterial anastomoses. The acardiac twin usually has other severe congenital anomalies such as a poorly developed or absent upper body and head. The four types include acephalus, anceps, acormus, amorphus to reflect the degree of abnormal development; none can survive ex utero. Due to the circulatory burden of supporting its acardiac co-twin, the pump twin is at risk of heart failure and problems related to preterm birth. In addition, TRAP may also occur in monochorionic triplets and other higher-order multiple gestations. (4)

Twin to Twin Transfusion Syndrome (TTTS)

TTTS is a rare but serious condition that occurs during a twin pregnancy when blood moves from one twin (the “donor twin”) to the other (the “recipient twin”) while in the womb. It is a complication that specifically occurs in identical (monozygotic) twin pregnancies that share the same egg sac (monochorionic) although they may or may not share the same amniotic sac (monoamniotic). Since the fetal blood supply is disproportionate, it can result in each fetus growing at different rates resulting in the donor twin being smaller, pale, with possible anemia and dehydration. In contrast, the recipient twin may be larger, with redness, elevated blood volume, increased blood pressure causing increased risk of heart failure. (5)

The Society for Maternal-Fetal Medicine (SMFM) states the Quintero staging system (see Table 1) is used to stage TTTS. (15)

Table 1: Quintero staging of twin-twin transfusion syndrome

Stage	Ultrasound Assessment	Criteria
I	Amniotic fluid	Maximal vertical pocket <2 cm in donor sac and maximal vertical pocket >8 cm in recipient sac.
II	Fetal bladder	Non-visualization of fetal bladder in donor twin over 60 minutes of observation.
III	Doppler studies	Absent or reversed umbilical artery end-diastolic velocity, reversed ductus venosus a-wave flow, pulsatile umbilical vein flow.
IV	Fetal ascites or hydrops	Ascites or hydrops in 1 or both twins.
V	Fetal cardiac activity	Fetal demise in 1 or both twins.

Regulatory Status

Fetal surgery is a surgical procedure and, as such, is not subject to regulation by the U.S. Food and Drug Administration.

Rationale

This policy is based on a review of relevant professional guidelines and position statements.

PROFESSIONAL GUIDELINES AND POSITION STATEMENTS

American College of Obstetricians and Gynecologists (ACOG) and American Academy of Pediatrics (AAP)

ACOG's Committee on Ethics and AAP's Committee on Bioethics issued a committee opinion on maternal-fetal intervention and fetal care centers in 2011 and reaffirmed in 2020. (6) The opinion recommended that:

- Fetal intervention cannot be performed without the explicit informed consent of the pregnant woman.
- Distinctions should be made to prospective parents between which protocols are standard or evidence-based therapies and which are innovative or experimental interventions.
- The informed consent process should involve thorough discussion of the risks and benefits for both the fetus and the pregnant woman.
- Safeguards should be in place to protect women considering fetal research.
- Access to support services such as social services, palliative care and perinatal hospice services, genetic counseling, and ethics consultation should be provided, when appropriate.

- The organization and governance of centers providing fetal interventions should involve a diverse group of professionals, Maternal fetal medicine and neonatologists should be included in this group. including members without direct ties to the center involved.
- Cooperation between fetal care centers should be encouraged to establish collaborative research networks and to support multicenter trials to accumulate more robust short- and long-term maternal and fetal outcome data on all categories of fetal intervention. In addition, the establishment of centers of excellence for those procedures that are particularly challenging and rare may help to optimize fetal and maternal outcomes.

Society of Fetal Medicine (SFM)

In 2024, the SFM published fetal therapy practice guidelines for fetal shunts. (7) A fetal shunt is placed in order to drain fluid from a fluid-filled fetal area into the amniotic cavity. The main indications are hydrothorax, congenital pulmonary airway malformation (CPAM) with dominant cyst, and lower urinary tract obstruction. SFM offers the following guidance:

- A fetal shunt is indicated if the hydrothorax is primary or hypertensive. In case of reaccumulation after a diagnostic tap or if the primary hydrothorax has led to a secondary nonimmune hydrops provided the primary nature is clearly established and in cases where gestational age is less than 34 weeks.
- A fetal shunt in CPAM is indicated in case of CPAM with a dominant cyst where the CPAM volume ratio (CVR) is greater than 1.6, where macrocysts are visible within the lesion, where the diameter of the dominant cyst is more than one-third of the largest diameter of the lesion and where presence of hydrops is not a contraindication for shunting.
- A fetal shunt is indicated in individuals with lower urinary tract obstruction (LUTO) when the etiology of LUTO is posterior urethral valve with oligohydramnios and fetal urinalysis is favorable.

ERKNet CAKUT-Obstructive Uropathy Work Group

ERKNet CAKUT-Obstructive Uropathy Work Group is a specialized team of physicians and specialty nephrology centers within the European reference network for rare kidney diseases (ERKNet) that focuses on congenital anomalies of the kidney and urinary tract (CAKUT). This multidisciplinary group of European experts develops clinical guidance, definitions, and research strategies for complex birth defects, aiming to improve diagnosis, management, and outcomes for patients. (8)

In 2022, the ERKNet CAKUT-Obstructive Uropathy Work Group published a consensus statement related to fetal LUTO which was supported by the ERKNet-CAKUT – Obstructive Uropathy Workgroup states that the diagnosis involves prenatal ultrasound (e.g., enlarged bladder, kidney changes, oligohydramnios) and fetal urine analysis for kidney function, with male fetuses often needing chromosome checks (karyotyping). Management focuses on multidisciplinary care, deciding between watchful waiting or fetal intervention (like bladder shunting) using prognostic indicators (e.g., renal pelvis diameter, urine sodium) to improve survival and prevent severe kidney failure, emphasizing referral to tertiary centers. (9)

National Institute of Child Health and Human Development

Myelomeningocele

In 2014, the National Institute of Child Health and Human Development convened the fetal myelomeningocele Maternal-Fetal Management Task Force with representatives from the AAP, ACOG, American Institute of Ultrasound in Medicine, American Pediatric Surgical Association, American Society of Anesthesiologists, American Society of Pediatric Neurosurgeons, IFMSS, American Association of Neurological Surgeons/Congress of Neurological Surgeons, North American Fetal Therapy Network, Society for Maternal-Fetal Medicine, Society of Pediatric Anesthesia, and Spina Bifida Association. The Task Force provided recommendations about optimal practice criteria for maternal-fetal surgery for myelomeningocele repair. (10)

Recommendations are related to 6 key considerations for teams providing in-utero myelomeningocele repair:

1. Defining a fetal therapy center;
2. Perioperative management for fetal myelomeningocele repair;
3. Long-term care;
4. Counseling;
5. Reporting and monitoring;
6. Access and regionalization.

In general, the authors emphasized the need for access to multidisciplinary teams for prenatal, perinatal, and follow-up care, and recommended that in-utero myelomeningocele repair be performed under strict adherence to the MOMS trial protocol in terms of preoperative evaluation, intraoperative procedure, and immediate postoperative care.

<u>American College of Obstetricians and Gynecologists (ACOG)</u>

In 2013, (reaffirmed 2022) ACOG issued a committee opinion on maternal-fetal surgery for myelomeningocele. (11) This opinion states, “Open maternal–fetal surgery for myelomeningocele repair is a major procedure for the woman and her affected fetus. Although there is demonstrated potential for fetal and pediatric benefit, there are significant maternal implications and complications that may occur acutely, postoperatively, for the duration of the pregnancy, and in subsequent pregnancies. It is a highly technical procedure with potential for significant morbidity and possibly mortality, even with the best and most experienced surgeons. Maternal–fetal surgery for myelomeningocele repair should only be offered to carefully selected patients at facilities with an appropriate level of personnel and resources.”

ACOG acknowledged the strict surgical inclusion criteria to include requiring a singleton gestation, myelomeningocele with an upper boundary located between T1 and S1, evidence of hindbrain herniation on fetal magnetic resonance imaging, gestational age between 19 0/7 weeks and 25 6/7 weeks at randomization, and a normal karyotype. Major trial exclusion criteria included anomalies unrelated to the myelomeningocele, severe kyphosis, risk of preterm birth (such as short cervix or prior preterm birth), placental abruption, contraindication to surgery (such as previous hysterotomy in the active uterine segment), and a maternal body mass index of 35 or more.

In 2017 (reaffirmed 2025), ACOG (12) provided a practice bulletin on neural tube defects which provide the following recommendations:

- An individual with a fetus with neural tube defects should be offered the management options of pregnancy termination, expectant management with neonatal surgical repair and in-utero fetal repair for appropriate candidates (level B - recommendation is based on limited or inconsistent scientific evidence).
- Despite the maternal and obstetric risks, in-utero repair is an option for women who meet appropriate criteria. Counseling should be nondirective and include all options, with full disclosure of all potential benefits and risks for the fetus and woman, including implications for future pregnancies (Level C- recommendations are based primarily on consensus and expert opinion).

Society of Obstetricians and Gynaecologists of Canada

In 2021, the Society of Obstetricians and Gynaecologists of Canada (13) 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)."

Society for Maternal-Fetal Medicine (SMFM)

Nonimmune hydrops is the presence of ≥2 abnormal fetal fluid collections in the absence of red cell alloimmunization. The most common etiologies include cardiovascular, chromosomal, and hematologic abnormalities, followed by structural fetal anomalies, complications of monochorionic twinning, infection, and placental abnormalities. A 2015 SMFM guideline (14) includes the following interventions for select etiologies for nonimmune hydrops.

Table 2: Therapy for selected etiologies of nonimmune hydrops

Etiology	Therapy	Recommendation
Cardiac tachyarrhythmia, supraventricular tachycardia, atrial flutter, or atrial fibrillation	Maternal transplacental administration of antiarrhythmic medication(s)	Treatment with antiarrhythmic medication unless gestational age is close to term or there is maternal or obstetrical contraindication to therapy
Fetal anemia secondary to parvovirus infection or fetomaternal hemorrhage	Fetal blood sampling followed by intrauterine transfusion	Fetal intrauterine transfusion if anemia is confirmed, unless pregnancy is at an advanced gestational age and risks associated with delivery are considered to be less than

		those associated with procedure
Fetal hydrothorax, chylothorax, or large pleural effusion associated with bronchopulmonary sequestration	Fetal needle drainage of effusion or placement of thoracoamniotic shunt; if gestational age is advanced, needle drainage prior to delivery in selected cases	Consider drainage of large unilateral pleural effusion(s) resulting in NIHF, or, if gestational age is advanced, consideration of needle drainage prior to delivery
Fetal CPAM	Macrocystic type: fetal needle drainage of effusion or placement of thoracoamniotic shunt; microcystic type: maternal administration of corticosteroids, betamethasone 12.5 mg IM q24 h 2 doses or dexamethasone 6.25 mg IM q12 h 4 doses	Consider drainage of large macrocystic CPAM that has resulted in NIHF; if large microcystic CPAM has resulted in NIHF, we suggest that management options include maternal corticosteroid administration
TTTS or TAPS	Laser ablation of placental anastomoses or selective termination	Consideration of fetoscopic laser photocoagulation of placental anastomoses for TTTS or TAPS that has resulted in NIHF <26 weeks
Twin-reversed arterial perfusion sequence	Percutaneous radiofrequency ablation	Referral for consideration of percutaneous radiofrequency ablation that has resulted in NIHF

For each of these etiologies, it is recommended that treatment be performed at tertiary care center or center with expertise in relevant therapy; CPAM, congenital pulmonary airway malformation; IM, intramuscular; NIHF, nonimmune hydrops fetalis; TAPS, twin-anemia polycythemia sequence; TTTS, twin-twin transfusion sequence.

In 2024, the SMFM published evidence based clinical guidelines for twin-twin transfusion and twin anemia-polycythemia sequence. (15) The SMFM makes the following recommendations:

- Routine first-trimester sonographic determination of chorionicity and amnionicity (GRADE 1B);
- Ultrasound surveillance for TTTS beginning at 16 weeks of gestation for all monochorionic-diamniotic twin pregnancies and continue at least every 2 weeks until delivery, with more frequent monitoring indicated with clinical concern (GRADE 1C);
- Routine sonographic surveillance for TTTS minimally include assessment of amniotic fluid volumes on both sides of the intertwin membrane and evaluation for the presence or absence of urine-filled fetal bladders, and ideally incorporate Doppler study of the umbilical arteries (GRADE 1C);

- Fetoscopic laser surgery as the standard treatment for stage II through stage IV TTTS presenting between 16 and 26 weeks of gestation (GRADE 1A);
- Expectant management with at least weekly fetal surveillance for asymptomatic patients continuing pregnancies complicated by stage I TTTS, and consideration for fetoscopic laser surgery for stage I TTTS presentations between 16 and 26 weeks of gestation complicated by additional factors such as maternal polyhydramnios-associated symptomatology (GRADE 1B);
- An individualized approach to laser surgery for early- and late-presenting TTTS (GRADE 1C);
- All patients with TTTS qualifying for laser therapy be referred to a fetal intervention center for further evaluation, consultation, and care (Best Practice);
- After laser therapy, we suggest weekly surveillance for 6 weeks followed by resumption of every-other-week surveillance thereafter, unless concern exists for post-laser TTTS, post-laser twin anemia-polycythemia sequence (TAPS), or fetal growth restriction (GRADE 2C);
- Following the resolution of TTTS after fetoscopic laser surgery, and without other indications for earlier delivery, we recommend delivery of dual-surviving monochorionic-diamniotic twins at 34 to 36 weeks of gestation (GRADE 1C);
- In TTTS pregnancies complicated by posttreatment single fetal demise, SMFM recommends full-term delivery (39 weeks) of the surviving co-twin to avoid complications of prematurity unless indications for earlier delivery exist (GRADE 1C);
- SMFM recommends that fetoscopic laser surgery not influence the mode of delivery (Best Practice);
- Prenatal diagnosis of TAPS minimally requires either middle cerebral artery Doppler peak systolic velocity values >1.5 and <1.0 multiples of the median in donor and recipient twins, respectively, or an intertwin D middle cerebral artery peak systolic velocity >0.5 multiples of the median (GRADE 1C);
- Providers consider incorporating middle cerebral artery Doppler peak systolic velocity determinations into all monochorionic twin ultrasound surveillance beginning at 16 weeks of gestation (GRADE 1C);
- Consultation with a specialized fetal care center when TAPS progresses to a more advanced disease stage (stage ≥II) before 32 weeks of gestation or when concern arises for coexisting complications such as TTTS (Best Practice).

In 2024, the Society of Fetal Medicine (SFM) published guidelines for Fetoscopic Endoluminal Tracheal Occlusion (FETO). SFM stated that FETO was started as an experimental therapy in fetuses with congenital diaphragmatic hernia (CDH) who had a bad prognosis. (16) The Tracheal Occlusion to Accelerate Lung growth-trial (TOTAL) has proved its effectiveness. The intervention attempts to correct pulmonary hypoplasia before delivery to reduce perinatal and neonatal mortality and morbidity. SFM defines severe pulmonary hypoplasia as a 25% increase in the lung area to the head circumference (with or without liver herniation).

SFM offers guidance for appropriate candidates for FETO. Eligibility criteria offered includes:

- Fetuses with severe diaphragmatic hernia (LHR <1 and liver herniated into the chest) identified between 22 0/7 weeks and 29 6/7 weeks of gestation.

- The mother must be at least 18 years old.
- Singleton pregnancy.
- Left sided diaphragmatic hernia.
- A fetal echocardiography confirms that there are no severe cardiac anomalies.
- Normal chromosomes and no related aberrations (microarray exome sequencing recommended).
- Cervix that is longer than 15 mm in length.
- The absence of maternal contraindications to abdominal fetoscopic surgery or general anesthesia.
- No maternal latex allergy.
- Acceptance of responsibility for attending the FETO center for balloon removal.

National Institute of Health and Care Excellence (NICE)

The NICE has developed guidelines regarding performance of in-utero surgery to treat some fetal anomalies. According to these guidelines, the following recommendations were given:

- Percutaneous fetal balloon valvuloplasty for pulmonary atresia or aortic stenosis has not been proven safe and effective. (17, 18)
- Percutaneous laser therapy for fetal tumors (i.e., sacrococcygeal teratomas, cervical teratomas, cystic hygromas and congenital cystic adenomatoid malformations [CCAM]) has not been proven safe and effective. (19)

Other Conditions

In-utero fetal surgery is being considered for other fetal conditions including but not limited to amniotic band syndrome, decompression of the fetal trachea, complete heart block, treatment of hypoplastic left heart syndrome and cleft lip, in-utero hematopoietic stem cell transplantation (HSCT) for stem cell disorders (e.g., alpha thalassemia, sickle cell anemia, beta thalassemia) and for in-utero gene therapy for disorders that result in irreversible illness or death in the prenatal or neonatal period (e.g., Type 2 Gaucher's Disease, Krabbe's disease, Hurler's Disease). Current professional guidelines do not include the use of these as a standard treatment option. The lack of inclusion in society guidelines suggests that these treatment options do not meet the necessary criteria for standard of care (outside of clinical trials) due to insufficient clinical evidence.

Coding

Procedure codes on Medical Policy documents are included **only** as a general reference tool for each policy. **They may not be all-inclusive.**

The presence or absence of procedure, service, supply, or device codes in a Medical Policy document has no relevance for determination of benefit coverage for members or reimbursement for providers. **Only the written coverage position in a Medical Policy should be used for such determinations.**

Benefit coverage determinations based on written Medical Policy coverage positions must include review of the member's benefit contract or Summary Plan Description (SPD) for defined coverage vs. non-coverage, benefit exclusions, and benefit limitations such as dollar or duration caps.

CPT Codes	59001, 59072, 59074, 59076, 59897

HCPCS Codes	S2400, S2401, S2402, S2403, S2404, S2405, S2409, S2411

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Centers for Medicare and Medicaid Services (CMS)

The information contained in this section is for informational purposes only. HCSC makes no representation as to the accuracy of this information. It is not to be used for claims adjudication for HCSC Plans.

The Centers for Medicare and Medicaid Services (CMS) does not have a national Medicare coverage position. Coverage may be subject to local carrier discretion.

A national coverage position for Medicare may have been developed since this medical policy document was written. See Medicare's National Coverage at <https://www.cms.hhs.gov>.

Policy History/Revision

Date	Description of Change
01/01/2026	Document updated. The following changes were made to Coverage: 1) Updated gestational age criteria from 32 to 34 weeks gestation for open in-utero resection of malformed pulmonary tissue or placement of a thoraco-amniotic shunt for congenital cystic adenomatoid malformation or bronchopulmonary sequestration; 2) Significantly revised criteria for fetal fetoscopic endoluminal tracheal occlusion (FETO); and 3) Removed in-utero stem cell transplantation and/or in-utero gene therapy. Added references 7-9, 15, 16. others updated and/or removed.
04/01/2024	Reviewed. No changes.

07/15/2023	Document updated with literature review. The following changes were made to Coverage: Expanded the existing experimental, investigational and/or unproven statement to include in-utero stem cell transplantation and/or in-utero gene therapy. Added references 2, 3, 13, 44, 56, 60-62, 77, 78; others updated and/or removed.
12/01/2022	Document updated with literature review. The following changes were made to Coverage: 1) Expanded the criteria for in-utero repair of myelomeningocele; 2) Changed fetal surgery to perform fetoscopic endoluminal tracheal occlusion (FETO) from "experimental, investigational and/or unproven" to "may be considered medical necessary" in fetuses with pulmonary hypoplasia due to severe isolated congenital diaphragmatic hernia (CDH) when specific criteria are met; 3) Removed congenital diaphragmatic hernia from the existing experimental, investigational, and/or unproven statement. Added references 1, 2, 18-23, 27, 28, 39-41, 47, 60-64, 67, 68; others updated and/or removed.
03/01/2021	Reviewed. No changes.
03/01/2020	Document updated with literature review. The following changes were made to Coverage: Added conditional coverage for treatment of twin-to-twin transfusion syndrome and twin reversed arterial perfusion sequence. The following references were added and/or updated: 1-2, 10-12, 14-22, 31-32, 37, 39-41, 44-47, and 50.
04/15/2016	Reviewed. No changes.
01/01/2015	Document updated with literature review. Coverage unchanged.
11/01/2012	Document updated with literature review. The entire policy was revised. The following two changes were made to Coverage: 1) In utero repair of myelomeningocele may be considered medically necessary when stated conditions are met; 2) Indications that previously may have been considered medically necessary (vesicoamniotic shunting for urinary tract obstruction, treatment of congenital adenomatoid malformation, extralobar pulmonary sequestration, and sacrococcygeal teratoma) now have specific conditions for medical necessity.
06/01/2008	Revised/updated entire document
03/15/2007	Revised/updated entire document
11/01/2000	New medical document