

Corporate Medical Policy

Fetal Surgery for Malformations

File Name: fetal_surgery_for_malformations
Origination: 3/2001
Last CAP Review: 2/2012
Next CAP Review: 2/2013
Last Review: 2/2012

Description of Procedure or Service

Fetal surgery is being investigated 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 or through single or multiple fetoscopic port incisions), surgically correcting the abnormality, and returning the fetus to the uterus and restoring uterine closure. Fetal surgery is a specialized technique that requires a multidisciplinary approach.

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.

This policy will pertain to fetal surgery performed for the following clinical conditions:

1. Fetal Urinary Tract Obstruction
2. Congenital Diaphragmatic Hernia (CDH)
3. Congenital Cystic Adenomatoid Malformation (CCAM) or Bronchopulmonary Sequestration (BPS)
4. Sacrococcygeal Teratoma
5. Myelomeningocele
6. Cardiac Malformations

*****Note: This Medical Policy is complex and technical. For questions concerning the technical language and/or specific clinical indications for its use, please consult your physician.**

Policy

BCBSNC will cover Fetal Surgery for Malformations when it is determined to be medically necessary because the medical criteria and guidelines shown below are met.

Benefits Application

Please refer to Certificate for availability of benefits. This policy relates only to the services or supplies described herein. Benefits may vary according to benefit design, therefore certificate language should be reviewed before applying the terms of the policy.

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When Fetal Surgery for Malformations is covered

Vesico-amniotic shunting as a treatment of urinary tract obstruction may be considered medically necessary in fetuses under the following conditions:

- 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 may be considered medically necessary under the following conditions:

- Congenital cystic adenomatoid malformation or bronchopulmonary sequestration is identified; AND
- 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., the maternal mirror syndrome) in the mother.

In utero removal of sacrococcygeal teratoma may be considered medically necessary under the following conditions:

- 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 repair of myelomeningocele may be considered medically necessary under the following conditions:

- 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.

When Fetal Surgery for Malformations is not covered

In utero repair of myelomeningocele is considered investigational in the following situations:

- Fetal anomaly unrelated to myelomeningocele; OR
- Severe kyphosis; OR
- Risk of preterm birth (e.g., short cervix or previous preterm birth); OR
- Maternal body mass index of 35 or more.

Other applications of fetal surgery are **investigational**, including but not limited to, temporary tracheal occlusion as a treatment of congenital diaphragmatic hernia or treatment of congenital heart defects.

Policy Guidelines

Amnioreduction and fetoscopic laser therapy as a treatment of twin-twin transfusion are not addressed in this policy. (See Evidence Based Guideline titled, "Twin-Twin Transfusion Syndrome, Treatment")

After 32 weeks' gestation, fetal lung maturity is adequate to permit Cesarean section and management of congenital cystic adenomatoid malformation, bronchopulmonary sequestration, or sacrococcygeal teratoma postnatally.

In utero surgery should be restricted to centers experienced in treating these conditions and staffed by surgeons adequately trained in fetal surgery techniques.

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Because of the differing benefits and risks of in utero versus postnatal surgeries, parents should make an informed choice between the procedures.

Due to a number of factors, including the rarity of the conditions and the small number of centers specializing in fetal interventions, the evidence on fetal surgery remains limited. Fetal surgery for many congenital conditions, including diaphragmatic hernia and heart defects, has not been shown to improve health outcomes in comparison with postnatal treatment. The available evidence is insufficient to demonstrate that fetal tracheal occlusion and aortic valvuloplasty provides improved health outcomes. For these and other applications of fetal surgery that are currently considered investigational, additional studies are needed to identify appropriate candidates and to evaluate longer term outcomes compared with postnatal management.

For conditions leading to fetal hydrops (certain cases of congenital cystic adenomatoid malformation, bronchopulmonary sequestration, or sacrococcygeal teratoma), for which mortality approaches 100%, fetal surgery may be considered medically necessary. Vesico-amniotic shunting for bilateral urinary tract obstruction may also be considered medically necessary to minimize the effects of this condition on kidney and lung development. Additional studies for these surgeries are needed to better define the appropriate surgical candidates, the most effective timing of the interventions, and the long-term health outcomes in surviving children.

Data from the MOMS trial show that prenatal repair of myelomeningocele reduces the need for shunting in the first 12 months after delivery and improves a composite measure of mental and motor function, with adjustment for lesion level, at 30 months of age. Prenatal surgery also improves the degree of hindbrain herniation and the likelihood of being able to walk independently when compared with postnatal surgery. The long-term impact on function needs to be evaluated, and benefits must be balanced against risks to mother and child. Thus, fetal surgery may be considered medically necessary following informed decision making for cases of prenatal myelomeningocele that meet the criteria of the MOM study.

Billing/Coding/Physician Documentation Information

This policy may apply to the following codes. Inclusion of a code in this section does not guarantee that it will be reimbursed. For further information on reimbursement guidelines, please see Administrative Policies on the Blue Cross Blue Shield of North Carolina web site at www.bcbsnc.com. They are listed in the Category Search on the Medical Policy search page.

Applicable codes: S2400, S2401, S2402, S2403, S2404, S2405, S2409, 59076, 59897

BCBSNC may request medical records for determination of medical necessity. When medical records are requested, letters of support and/or explanation are often useful, but are not sufficient documentation unless all specific information needed to make a medical necessity determination is included.

Scientific Background and Reference Sources

BCBSA TEC Evaluation, February 1999, Volume 13, No. 22

BCBSA TEC Evaluation, February 2000, Volume 14, No. 23

BCBSA Medical Policy Reference Manual, 12/15/2000; 4.01.10

Specialty Matched Consultant Advisory Panel - 9/2001

BCBSA Medical Policy Reference Manual, 12/18/2002; 4.01.10

Specialty Matched Consultant Advisory Panel -8/2003

Neural Tube Defects. ACOG Practice Bulletin. Clinical Management Guidelines for Obstetrician-

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Gynecologists. Number 44, July 2003.

BCBSA Medical Policy Reference Manual [Electronic Version]. 4.01.10, 3/15/05

Specialty Matched Consultant Advisory Panel - 8/25/05

BCBSA Medical Policy Reference Manual [Electronic Version]. 4.01.10, 12/12/06

Specialty Matched Consultant Advisory Panel - 8/29/07

BCBSA Medical Policy Reference Manual [Electronic Version]. 4.01.10, 12/13/07

Specialty Matched Consultant Advisory Panel -2/2010

BCBSA Medical Policy Reference Manual [Electronic Version]. 4.01.10, 2/11/2010

BCBSA Medical Policy Reference Manual [Electronic Version]. 4.01.10, 3/10/2011

Medical Director review 5/2011

BCBSA Medical Policy Reference Manual [Electronic Version]. 4.01.10, 12/8/2011

Specialty Matched Consultant Advisory Panel -3/21/12

Policy Implementation/Update Information

3/01	Original policy issued.
9/01	Specialty Matched Consultant Advisory Panel review. No changes to policy.
12/02	Added codes S2400 - S2409 to billing and coding section. System coding changes.
8/03	Specialty Matched Consultant Advisory Panel review 8/4/03. Description and Benefits Application sections revised. No changes to criteria.
4/04	Billing/Coding section updated for consistency. Individual CPT codes listed for CPT code ranges S2400-S2409 under Billing/Coding section.
9/1/05	Temporary tracheal occlusion as a treatment of congenital diaphragmatic hernia is considered investigational-removed from "When covered" section to "When not covered" section with explanation. Explanation re: investigational status of fetal surgery for myelomeningocele or aqueductal stenosis added to "When not covered" section. Reference sources added. Specialty Matched Consultant Advisory Panel review -8/25/05. Notification given 9/1/05. Effective date 11/3/05.
6/4/07	CPT codes 59076 and 59897 added to Billing/Coding section. (pmo)
9/24/07	Reference sources added. Specialty Matched Consultant Advisory Panel review 8/29/07. Medical term definitions added. No changes to policy criteria. (pmo)
3/16/10	Specialty Matched Consultant Advisory Panel review 2/11/2010. "Description" section revised. No changes to policy statement. Added additional statement to the "When Not Covered" section for clarification to indicate "Treatment of congenital heart defects. Further research is needed to determine optimal patient selection, timing and technique for the different types of correctable heart defects. Studies on fetal surgery for congenital heart defects are lacking." References added. (btw)
6/22/10	Policy Number(s) removed (amw)
1/18/2011	Medical criteria in the "When It Is Covered" and "When It Is Not Covered" sections revised to read: Vesico-amniotic shunting as a treatment of urinary tract obstruction may be considered medically necessary in fetuses under the following conditions: 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 may be considered medically necessary under the following conditions: Congenital

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cystic adenomatoid malformation or bronchopulmonary sequestration is identified; AND 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., the maternal mirror syndrome) in the mother. In utero removal of sacrococcygeal teratoma may be considered **medically necessary** under the following conditions: 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. Other applications of fetal surgery are **investigational**, including but not limited to, temporary tracheal occlusion as a treatment of congenital diaphragmatic hernia, treatment of congenital heart defects, or fetal surgery for myelomeningocele. Policy Guidelines updated. Specialty Matched Consultant Advisory Panel review 12/16/2010. Policy accepted as drafted. (adn)

5/24/11 Description section updated. The following statement was added to the When Fetal surgery Is Covered section: In utero repair of myelomeningocele may be considered medically necessary under the following conditions: 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. The following was added to the When Fetal Surgery Is Not Covered section: In utero repair of myelomeningocele is considered investigational in the following situations: fetal anomaly unrelated to myelomeningocele; or severe kyphosis; or risk of preterm birth; or maternal body mass index of 35 or more. Policy Guidelines updated. (adn)

4/17/12 Added references. Specialty Matched Consultant Advisory Panel review 3/21/12. (sk)

Medical policy is not an authorization, certification, explanation of benefits or a contract. Benefits and eligibility are determined before medical guidelines and payment guidelines are applied. Benefits are determined by the group contract and subscriber certificate that is in effect at the time services are rendered. This document is solely provided for informational purposes only and is based on research of current medical literature and review of common medical practices in the treatment and diagnosis of disease. Medical practices and knowledge are constantly changing and BCBSNC reserves the right to review and revise its medical policies periodically.