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| POLICY NUMBER | EFFECTIVE DATE | APPROVED BY |
|---------------|----------------|--------------------------------|
| MG.MM.ME.63 | 2/14/2025 | MPC (Medical Policy Committee) |

IMPORTANT NOTE ABOUT THIS MEDICAL POLICY:

Property of ConnectiCare, Inc. All rights reserved. The treating physician or primary care provider must submit to ConnectiCare, Inc. the clinical evidence that the patient meets the criteria for the treatment or surgical procedure. Without this documentation and information, ConnectiCare will not be able to properly review the request for prior authorization. This clinical policy is not intended to pre-empt the judgment of the reviewing medical director or dictate to health care providers how to practice medicine. Health care providers are expected to exercise their medical judgment in rendering appropriate care. The clinical review criteria expressed below reflects how ConnectiCare determines whether certain services or supplies are medically necessary. ConnectiCare established the clinical review criteria based upon a review of currently available clinical information (including clinical outcome studies in the peer-reviewed published medical literature, regulatory status of the technology, evidence-based guidelines of public health and health research agencies, evidence-based guidelines and positions of leading national health professional organizations, views of physicians practicing in relevant clinical areas, and other relevant factors). ConnectiCare, Inc. expressly reserves the right to revise these conclusions as clinical information changes, and welcomes further relevant information. Identification of selected brand names of devices, tests and procedures in a medical coverage policy is for reference only and is not an endorsement of any one device, test or procedure over another. Each benefit plan defines which services are covered. The conclusion that a particular service or supply is medically necessary does not constitute a representation or warranty that this service or supply is covered and/or paid for by ConnectiCare, as some plans exclude coverage for services or supplies that ConnectiCare considers medically necessary. If there is a discrepancy between this guideline and a member's benefits plan, the benefits plan will govern. In addition, coverage may be mandated by applicable legal requirements of the State of CT and/or the Federal Government. Coverage may also differ for our Medicare members based on any applicable Centers for Medicare & Medicaid Services (CMS) coverage statements including including National Coverage Determinations (NCD), Local Coverage Determinations (LCD) and/or Local Medical Review Policies (LMRP). All coding and web site links are accurate at time of publication.

Background

Fetal surgery (also referred to as in-utero or prenatal surgery) is a complex surgical intervention performed on the developing fetus in-utero, using open or minimally invasive techniques, to correct fetal abnormalities that interfere with organ development and fetal survival.

Guideline

Fetal surgery is considered medically necessary for any of the following:

- Amniotic band syndrome (ABS)
- Bladder outlet obstruction (BOO)
- Congenital diaphragmatic hernia (CDH)
- Congenital high airway obstruction syndrome (CHAOS)
- Congenital lung masses/malformations, e.g.:
 - Bronchial atresia
 - Bronchogenic cysts
 - Bronchopulmonary sequestration (BPS) (aka lung or pulmonary sequestration)
 - Congenital pulmonary airway malformation (CPAM) (previously known as congenital cystic adenomatoid malformation [CCAM])
- Extralobar pulmonary sequestration (EPS)

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- Fetal cystic hygroma
- Fetal renal failure (FRF)
- Hydronephrosis
- Mediastinal teratoma
- Myelomeningocele (spina bifida)
- Pleural Effusion
- Sacrococcygeal Teratoma (SCT)
- Twin anemia-polycythemia sequence (TAPS)
- Twin reversed arterial perfusion (TRAP)
- Twin-Twin Transfusion Syndrome (TTTS)
- Urinary Tract Obstruction (UTO) (aka obstructive uropathy, e.g., congenital posterior urethral valves)

Limitations/Exclusions

The following indications, considered experimental, investigational or unproven will be reviewed on a case-by-case basis upon request (list not all-inclusive):

- Aqueductal stenosis (i.e., hydrocephalus)
- Cleft lip and/or cleft palate
- Congenital heart defects/disease (e.g., aortic stenosis, mitral valve dysplasia/regurgitation, pericardial teratoma)
- Gastroschisis
- Hydronephrosis

The following utero interventions, considered experimental, investigational or unproven, will be reviewed on a case-by-case basis upon request (list not all-inclusive):

- Aortic or pulmonary balloon valvuloplasty
- Arial needle septoplasty
- Endoscopic approach (i.e., fetoscopic cystoscopy) for the treatment of lower UTO
- Fetoscopic laser ablation for type 2 vasa previa
- In-utero gene therapy
- In-utero hematopoietic stem-cell transplantation for stem-cell-related diseases
- Laser, thermocoagulation or radiofrequency ablation techniques for the treatment of sacrococcygeal teratoma
- Percutaneous sclerotherapy
- Shunting for the treatment of fetal cerebral ventriculomegaly

Procedure Codes

| 59001 | Amniocentesis; therapeutic amniotic fluid reduction (includes ultrasound guidance) | |
|-------|---|--|
| 59072 | Fetal umbilical cord occlusion, including ultrasound guidance | |
| 59074 | Fetal fluid drainage (eg, vesicocentesis, thoracocentesis, paracentesis), including ultrasound guidance | |



| 59076 | Fetal shunt placement, including ultrasound guidance |
|-------|--|
| 59897 | Unlisted fetal invasive procedure, including ultrasound guidance, when performed |
| S2401 | Repair, urinary tract obstruction in the fetus, procedure performed in utero |
| S2402 | Repair, congenital cystic adenomatoid malformation in the fetus, procedure performed in utero |
| S2403 | Repair, extralobar pulmonary sequestration in the fetus, procedure performed in utero |
| S2404 | Repair, myelomeningocele 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 |
| S2411 | Fetoscopic laser therapy for treatment of twin-to-twin transfusion syndrome |

Diagnosis Codes

| D18.1 | Lymphangioma, any site |
|----------|---|
| D48.0 | Neoplasm of uncertain behavior of bone and articular cartilage |
| 030.021 | Conjoined twin pregnancy, first trimester |
| 030.022 | Conjoined twin pregnancy, second trimester |
| 030.23 | Conjoined twin pregnancy, third trimester |
| 030.029 | Conjoined twin pregnancy, unspecified trimester |
| O33.7XX0 | Maternal care for disproportion due to other fetal deformities, not applicable or unspecified |
| 033.7XX1 | Maternal care for disproportion due to other fetal deformities, fetus 1 |
| 033.7XX2 | Maternal care for disproportion due to other fetal deformities, fetus 2 |
| O33.7XX3 | Maternal care for disproportion due to other fetal deformities, fetus 3 |
| 033.7XX4 | Maternal care for disproportion due to other fetal deformities, fetus 4 |
| O33.7XX5 | Maternal care for disproportion due to other fetal deformities, fetus 5 |
| O33.7XX9 | Maternal care for disproportion due to other fetal deformities, other fetus |
| 036.8910 | Maternal care for other specified fetal problems, first trimester, not applicable or unspecified |
| 036.8911 | Maternal care for other specified fetal problems, first trimester, fetus 1 |
| 036.8912 | Maternal care for other specified fetal problems, first trimester, fetus 2 |
| 036.8913 | Maternal care for other specified fetal problems, first trimester, fetus 3 |
| 036.8914 | Maternal care for other specified fetal problems, first trimester, fetus 4 |
| 036.8915 | Maternal care for other specified fetal problems, first trimester, fetus 5 |
| 036.8919 | Maternal care for other specified fetal problems, first trimester, other fetus |
| 036.8920 | Maternal care for other specified fetal problems, second trimester, not applicable or unspecified |
| 036.8921 | Maternal care for other specified fetal problems, second trimester, fetus 1 |



| 036.8922 | Maternal care for other specified fetal problems, second trimester, fetus 2 |
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| 036.8923 | Maternal care for other specified fetal problems, second trimester, fetus 3 |
| 036.8924 | Maternal care for other specified fetal problems, second trimester, fetus 4 |
| 036.8925 | Maternal care for other specified fetal problems, second trimester, fetus 5 |
| 036.8929 | Maternal care for other specified fetal problems, second trimester, other fetus |
| 036.8930 | Maternal care for other specified fetal problems, third trimester, not applicable or unspecified |
| 036.8931 | Maternal care for other specified fetal problems, third trimester, fetus 1 |
| 036.8932 | Maternal care for other specified fetal problems, third trimester, fetus 2 |
| 036.8933 | Maternal care for other specified fetal problems, third trimester, fetus 3 |
| 036.8934 | Maternal care for other specified fetal problems, third trimester, fetus 4 |
| 036.8935 | Maternal care for other specified fetal problems, third trimester, fetus 5 |
| 036.8939 | Maternal care for other specified fetal problems, third trimester, other fetus |
| 036.8990 | Maternal care for other specified fetal problems, unspecified trimester, not applicable or unspecified |
| 036.8991 | Maternal care for other specified fetal problems, unspecified trimester, fetus 1 |
| 036.8992 | Maternal care for other specified fetal problems, unspecified trimester, fetus 2 |
| 036.8993 | Maternal care for other specified fetal problems, unspecified trimester, fetus 3 |
| 036.8994 | Maternal care for other specified fetal problems, unspecified trimester, fetus 4 |
| 036.8995 | Maternal care for other specified fetal problems, unspecified trimester, fetus 5 |
| 036.8999 | Maternal care for other specified fetal problems, unspecified trimester, other fetus |
| 043.021 | Fetus-to-fetus placental transfusion syndrome, first trimester |
| 043.022 | Fetus-to-fetus placental transfusion syndrome, second trimester |
| 043.023 | Fetus-to-fetus placental transfusion syndrome, third trimester |
| 043.029 | Fetus-to-fetus placental transfusion syndrome, unspecified trimester |
| P02.3 | Newborn affected by placental transfusion syndromes |
| P28.89 | Other specified respiratory conditions of newborn |
| Q05.0 | Cervical spina bifida with hydrocephalus |
| Q05.1 | Thoracic spina bifida with hydrocephalus |
| Q05.2 | Lumbar spina bifida with hydrocephalus |
| Q05.3 | Sacral spina bifida with hydrocephalus |
| Q05.4 | Unspecified spina bifida with hydrocephalus |
| Q05.5 | Cervical spina bifida without hydrocephalus |
| Q05.6 | Thoracic spina bifida without hydrocephalus |
| Q05.7 | Lumbar spina bifida without hydrocephalus |
| Q05.8 | Sacral spina bifida without hydrocephalus |



| Q05.9 | Spina bifida, unspecified |
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| Q07.00 | Arnold-Chiari syndrome without spina bifida or hydrocephalus |
| Q07.01 | Arnold-Chiari syndrome with spina bifida |
| Q07.02 | Arnold-Chiari syndrome with hydrocephalus |
| Q07.03 | Arnold-Chiari syndrome with spina bifida and hydrocephalus |
| Q07.8 | Other specified congenital malformations of nervous system |
| Q07.9 | Congenital malformation of nervous system, unspecified |
| Q18.8 | Other specified congenital malformations of face and neck |
| Q33.0 | Congenital cystic lung |
| Q33.2 | Sequestration of lung |
| Q33.3 | Agenesis of lung |
| Q33.6 | Congenital hypoplasia and dysplasia of lung |
| Q62.31 | Congenital ureterocele, orthotopic |
| Q62.32 | Cecoureterocele |
| Q62.39 | Other obstructive defects of renal pelvis and ureter |
| Q64.2 | Congenital posterior urethral valves |
| Q64.31 | Congenital bladder neck obstruction |
| Q64.32 | Congenital stricture of urethra |
| Q64.33 | Congenital stricture of urinary meatus |
| Q64.39 | Other atresia and stenosis of urethra and bladder neck |
| Q89.4 | Conjoined twins |
| Q89.8 | Other specified congenital malformations |
| R89.7 | Abnormal histological findings in specimens from other organs, systems and tissues |
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References

Holcomb & Ashcraft's Pediatric Surgery, 7th Ed., Elsevier Pub. 2020, Chapter on "Fetal Surgery."

Pediatric Surgery, 2nd Ed., Coppola et al Eds., Springer Pub. 2022, Chapter on "Fetal Surgery and Interventions," pp. 263-272.

Patel, et al: Procedural, pregnancy, and short-term outcomes after fetal aortic valvuloplasty, Catheterization and Cardiovascular Interventions, 96(3); 626-632, 2020.

Pickard, et al: Fetal Aortic Valvuloplasty for Evolving Hypoplastic Left Heart Syndrome: A Decision Analysis, Circulation, Cardiovascular Quality and Outcomes, 13(4): epub 2020.

Specialty matched clinical peer review.



Revision History

| Company | DATE | REVISION |
|------------------------------|---------------|--|
| ConnectiCare | Feb. 14, 2025 | Added fetal cystic hygroma as covered indication |
| EmblemHealth ConnectiCare | Dec. 8, 2023 | New policy |