

## MP.116.MPC – Genetic Testing for Cystic Fibrosis

Maryland Physician sCare considers **Genetic Testing for Cystic Fibrosis (CF)** medically necessary for the following indications:

Genetic testing for CF is considered medically necessary when any of the following criteria are met:

- Couples planning a pregnancy or seeking prenatal care
- Persons with a family history of CF
- Persons with a first degree relative identified as a CF carrier
- Reproductive partners of persons with CF or are carriers of CF

Or

- Diagnostic purposes
  1. A clinical presentation of CF and a negative/equivocal sweat test
  2. Infant with meconium ileus or other symptoms indicative of CF who are too young to produce adequate amounts of sweat for a sweat chloride test
  3. Infant with elevated immunoreactive trypsinogen (IRT) value on newborn screening with no clinical suspicion of CF
  4. Male infertility in the father from either congenital bilateral absence of vas deferens (CBAVD) or azoospermia or severe oligospermia (i.e. <5 million sperm/milliliter) with palpable vas deferens

Or

- Prenatal testing of fetus may be indicated for any of the following:
  1. Embryo at risk when either parent has a diagnosis of CF, is a known carrier of a CFTR mutation or has a family history of CF
  2. Fetus with fetal echogenic bowel per ultrasound during second trimester
- Prenatal screening in individuals of Eastern European Jewish descent before conception or during early pregnancy

### Limitations

Genetic testing for CF is considered not medically necessary and therefore not covered for any of the following:

- Complete analysis of the CFTR gene by DNA sequencing is not appropriate for routine carrier screening in the general population
- If a patient has been previously tested, results should be documented and the test should not be repeated

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- Complete gene sequencing (beyond the ACMG-23 standard mutation panel) is not appropriate for carrier screening because it may yield results that are difficult to interpret, hence it is not covered and requests for exceptions will be reviewed on a case by case basis.

### See Also:

PA.097.MH Molecular-Genetic Testing

### Background

Cystic Fibrosis (CF) is a life-threatening autosomal recessive condition that affects several systems, including pulmonary, pancreatic, and gastrointestinal systems. CF affects an estimated 30,000 Americans, with 1,000 new cases diagnosed each year. The prevalence rates vary by ethnicity, with nearly 96% of those affected individuals are Caucasian.

The American College of Obstetricians and Gynecologists (ACOG) reports that the median survival is 37 years, resulting in respiratory failure as the most common cause of death. Common symptoms of CF include: coughing, wheezing, loose stools, abdominal pain, failure to thrive and infertility in men. CF is caused by mutations in the CF transmembrane regulatory (CFTR) gene on chromosome 7 and screening for the 23 most common mutations of this gene is available. These mutations can be detected through DNA sequencing.

### Codes:

CPT Codes / HCPCS Codes / ICD-10 Codes	
Code	Description
CPT Codes	
81220	CFTR gene analysis, common variants
81221	CFTR gene analysis; common variants, known familial variants
81222	CFTR gene analysis; common variants, duplication/deletion variants
81224	CFTR gene analysis; common variants, intron 8 poly-T analysis (male infertility)
ICD-10 codes covered if selection criteria are met:	
D25.9	Leiomyoma of uterus, unspecified
E84.0	Cystic fibrosis with pulmonary manifestations
E84.1	Cystic fibrosis with intestinal manifestations
E84.11	Meconium ileus in cystic fibrosis

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E84.19	Cystic fibrosis with other intestinal manifestations
E84.8	Cystic fibrosis with other manifestations
E84.9	Cystic fibrosis, unspecified
G43.909	Migraine, unsp, not intractable, without status migrainosus
I10	Essential (primary) hypertension
O09.519	Supervision of elderly primigravida, unspecified trimester
O34.21	Maternal care for scar from previous cesarean delivery
O35.8XX0	Maternal care for oth fetal abnormality and damage, unsp
O44.00	Placenta previa specified as w/o hemorrhage, unsp trimester
O99.280	Endo, nutritional and metab diseases comp preg, unsp tri
Z01.89	Encounter for other specified special examinations
Q55.8	Other specified congenital malformations of male genital organs
Z13.228	Encounter for screening for other metabolic disorders
Z13.71	Encounter for nonprocreative screening for genetic disease carrier status
Z33.1	Pregnant state, incidental
Z31.438	Encounter for oth genetic testing of female for pro mgmt
Z31.430	Encounter of female for testing for genetic disease carrier status for procreative management
Z31.440	Encounter of male for testing for genetic disease carrier status for procreative management
Z31.69	Encounter for oth general cnsl and advice on procreation
Z31.9	Encounter for procreative management, unspecified
Z32.00	Encounter for pregnancy test, result unknown
Z32.01	Encounter for pregnancy test, result positive
Z34.0	Encounter for supervision of normal first pregnancy
Z34.00	Encounter for supervision of normal first pregnancy, unspecified trimester
Z34.01	Encounter for supervision of normal first pregnancy, first trimester
Z34.02	Encounter for supervision of normal first pregnancy, second trimester
Z34.03	Encounter for supervision of normal first pregnancy, third trimester
Z34.80	Encounter for suprvsn of normal pregnancy, unsp trimester
Z34.81	Encounter for suprvsn of normal pregnancy, first trimester

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Z34.83	Encounter for supervn of normal pregnancy, first trimester
Z34.90	Encounter for supervn of normal pregnancy, first trimester
Z36	Encounter for antenatal screening of mother
Z78.9	Other specified health status
Z84.81	Family history of carrier of genetic disease

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