

Genetic Testing for Hereditary Pancreatitis

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Medical Guideline Disclaimer

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Definitions

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| Acute pancreatitis | Sudden inflammation of the pancreas caused by trypsin activation within the pancreas that usually resolves after a few days with treatment. |
| Chronic pancreatitis | Ongoing inflammation of the pancreas caused by trypsin activation within the pancreas that progressively leads to permanent damage exocrine and endocrine pancreatic insufficiency and increased risk for pancreatic cancer. |
| Hereditary pancreatitis (subset of chronic pancreatitis) | An autosomal dominant disease, primarily caused by sequence variants in the protease, serine, 1 (trypsin 1) gene (PRSS1) and characterized by recurrent episodes of epigastric pain, nausea and vomiting. Symptoms of hereditary pancreatitis can start in childhood and evolve into chronic pancreatitis by 20 years of age, however disease onset is variable and some people may not exhibit symptoms until adulthood. |

Related Medical Guidelines

[Carrier Screening for Parents or Prospective Parents](#)

[Genetic Counseling and Testing](#)

Guideline

Genetic testing for hereditary pancreatitis (PRSS1, SPIK1 and CFTR mutations) is considered medically necessary in children aged 18 years and under, with a confirmed diagnosis of acute or chronic pancreatitis, and for whom additional invasive diagnostic tests would be deemed unnecessary if genetic test result is positive.

Limitations/Exclusions

Genetic testing for hereditary pancreatitis is considered investigational for all for all other indications not meeting the above criteria.

Applicable Procedure Codes

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| 81220 | CFTR (cystic fibrosis transmembrane conductance regulator) (eg, cystic fibrosis) gene analysis; common variants (eg, ACMG/ACOG guidelines) |
| 81221 | CFTR (cystic fibrosis transmembrane conductance regulator) (eg, cystic fibrosis) gene analysis; known familial variants |
| 81222 | CFTR (cystic fibrosis transmembrane conductance regulator) (eg, cystic fibrosis) gene analysis; duplication/deletion variants |
| 81223 | CFTR (cystic fibrosis transmembrane conductance regulator) (eg, cystic fibrosis) gene analysis; full gene sequence |
| 81224 | CFTR (cystic fibrosis transmembrane conductance regulator) (eg, cystic fibrosis) gene analysis; intron 8 poly-T analysis (e.g., male infertility) |
| 81401 | Molecular pathology procedure, Level 2 (eg, 2-10 SNPs, 1 methylated variant, or 1 somatic variant [typically using nonsequencing target variant analysis], or detection of a dynamic mutation disorder/triplet repeat) LINC00518 (long intergenic non-protein coding RNA 518) (eg, melanoma), expression analysis PRAME (preferentially expressed antigen in melanoma) (eg, melanoma), expression analysis (Revision eff. 01/01/2018) |
| 81404 | Molecular pathology procedure, Level 5 (eg, analysis of 2-5 exons by DNA sequence analysis, mutation scanning or duplication/deletion variants of 6-10 exons, or characterization of a dynamic mutation disorder/triplet repeat by Southern blot analysis) (Revision eff. 01/01/2018) |
| 81479 | Unlisted molecular pathology procedure |

Applicable ICD-10 Diagnosis Codes

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| K85.00 | Idiopathic acute pancreatitis without necrosis or infection |
| K85.01 | Idiopathic acute pancreatitis with uninfected necrosis |
| K85.02 | Idiopathic acute pancreatitis with infected necrosis |
| K85.80 | Other acute pancreatitis without necrosis or infection |
| K85.81 | Other acute pancreatitis with uninfected necrosis |
| K85.82 | Other acute pancreatitis with infected necrosis |
| K85.90 | Acute pancreatitis without necrosis or infection, unspecified |
| K85.91 | Acute pancreatitis with uninfected necrosis, unspecified |
| K85.92 | Acute pancreatitis with infected necrosis, unspecified |
| K86.1 | Other chronic pancreatitis |

References

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