Hereditary Pancreatic Cancer

Genetic testing with the Pancreatic Cancer Panel may be appropriate if your personal and/or family history is suggestive of a hereditary predisposition to cancer. This includes:
- Pancreatic cancer diagnosed at any age
- A first degree relative diagnosed with pancreatic cancer
- Multiple relatives diagnosed with pancreatic cancer and/or related cancers (including breast, colon, or melanoma)

Your healthcare provider will determine if genetic testing is medically necessary for you.

Genes Included on the Hereditary Pancreatic Cancer Panel are Listed in the Table Below

### High-Risk Genes

**Well-studied**
- Greater than 4-fold risk of developing one or more cancers
- Can cause a moderate risk for other cancers
- National or expert opinion guidelines for screening and prevention are established

### Moderate-Risk Genes

**Well-studied**
- Approximately 2- to 4-fold risk of developing one or more cancers
- May increase risk for other cancers
- Limited guidelines for screening and prevention

### Newer Genes

**Not as well-studied**
- Precise lifetime risks and tumor spectrum not yet determined
- Guidelines for screening and prevention are limited or not available

<table>
<thead>
<tr>
<th>Gene</th>
<th>Lifetime Cancer and/or Tumor Risks*</th>
</tr>
</thead>
<tbody>
<tr>
<td>APC</td>
<td>Colorectal (up to 93%), Duodenal or periampullary (4-12%), Gastric, Thyroid (up to 3%), Pancreatic, Brain-medulloblastoma, Liver-hepatoblastoma, Desmoid tumors, Gastrointestinal polyps</td>
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<tr>
<td>BRCA1</td>
<td>Female breast (55-87%), Ovarian (39-59%), Prostate, Male breast, Pancreatic, Fallopian tube, Primary peritoneal, Endometrial-serous</td>
</tr>
<tr>
<td>BRCA2</td>
<td>Female breast (33-84%), Prostate (up to 34%), Ovarian (11-27%), Pancreatic (up to 7%), Male breast ( up to 7%), Melanoma, Fallopian tube, Primary peritoneal, Endometrial-serous</td>
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<tr>
<td>CDKN2A</td>
<td>Melanoma (28-67%), Pancreatic (17%), Brain-astrocytoma</td>
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<tr>
<td>EPCAM**</td>
<td>Colorectal (69-75%), Endometrial (12-55%), Ovarian, Gastric, Pancreatic, Biliary tract, Urinary tract-transitional cell, Small bowel, Brain, Sebaceous neoplasms, Prostate</td>
</tr>
<tr>
<td>MLH1</td>
<td>Colorectal (34-46%), Endometrial (18-54%), Ovarian (10-20%), Gastric (6-20%), Urinary tract-transitional cell (1-4%), Pancreatic (1-4%), Biliary tract (2-3%), Small bowel (4-12%), Brain, Sebaceous neoplasms, Prostate</td>
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<tr>
<td>MSH2</td>
<td>Colorectal (37-48%), Endometrial (21-57%), Ovarian (10-24%), Urinary tract-transitional cell (8-20%), Gastric (&lt;1-9%), Pancreatic (1-4%), Biliary tract, Small bowel (1%), Brain, Sebaceous neoplasms, Prostate</td>
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<tr>
<td>MSH6**</td>
<td>Colorectal (20-44%), Endometrial (16-71%), Ovarian (1-13%), Gastric, Pancreatic, Biliary tract, Urinary tract-transitional cell, Small bowel, Brain, Sebaceous neoplasms, Prostate</td>
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<tr>
<td>PALB2</td>
<td>Female breast (up to 58%), Male breast, Pancreatic, Ovarian, Prostate</td>
</tr>
<tr>
<td>PMS2**</td>
<td>Colorectal (11-20%), Endometrial (12-26%), Ovarian, Gastric, Biliary tract, Urinary tract-transitional cell, Small bowel, Brain, Sebaceous neoplasms, Prostate</td>
</tr>
<tr>
<td>STK11</td>
<td>Female breast (up to 54%), Colorectal (39%), Pancreatic (11-36%), Gastric (29%), Ovarian tumors (21%), Lung (7-17%), Small bowel (13%), Cervical (10%), Testicular tumors (9%), Endometrial (9%), Gastrointestinal polyps</td>
</tr>
<tr>
<td>TP53</td>
<td>Female breast (85%), Soft tissue sarcoma, Osteosarcoma, Brain, Hematologic malignancies-Acute leukemias among others, Adrenocortical carcinoma, among others. Overall risk for cancer:up to 95% in females, 88% in males</td>
</tr>
<tr>
<td>VHL</td>
<td>Renal-clear cell (up to 69%), Hemangioblastomas-retinal and central nervous system (50-80%), Pheochromocytomas (11-19%), Pancreatic neuroendocrine tumors (8-17%), Endolymphatic sac tumors (up to 10%)</td>
</tr>
</tbody>
</table>
Medical Management Based on Genetic Test Results

Clinical guidelines may be available which provide options and recommendations for patients who have a positive (pathogenic or likely pathogenic variant) test result indicating an increased risk for cancer and/or tumors. Guidelines and recommendations for early detection and/or risk reduction are specific to the gene in which the pathogenic variant was found.

Recommendations may include:
- Clinical exams, such as skin and/or eye exams
- Imaging exams, such as a mammogram, MRI, CT and/or ultrasound
- Screening procedures, such as pancreatic surveillance, colonoscopy and/or endoscopy
- Risk-reducing medications and/or surgery

In some cases, guidelines for screening and prevention are limited or not available for a positive result. Once your test results are available, a discussion with your healthcare provider is recommended to determine the most appropriate medical management options for you and your family.

### Resources

**General**
- American Cancer Society
  - [www.cancer.org](http://www.cancer.org)
- GeneDx
  - [www.genedx.com/oncology](http://www.genedx.com/oncology)
- National Cancer Institute
  - [www.cancer.gov](http://www.cancer.gov)

**Pancreatic Cancer**
- Pancreatic Cancer Action Network
  - [www.pancan.org](http://www.pancan.org)
- Pancreatic Cancer Alliance
  - [www.pancreaticalliance.org](http://www.pancreaticalliance.org)

**Find a Genetic Counselor**
- Canadian Association of Genetic Counsellors
  - [www.cagc-accg.ca](http://www.cagc-accg.ca)
- National Society of Genetic Counselors
  - [www.nsgc.org](http://www.nsgc.org)