



Hereditary Pancreatic Cancer: What You Need to Know

Are most cases of pancreatic cancer hereditary?

No, only 5-10 out of 100 (5-10%) cases of pancreatic cancers are thought to be due to an inherited risk. There are some known hereditary cancer syndromes that increase the risk for pancreatic cancer. Many families with these syndromes could have other types of cancer as well.

What genes are linked to hereditary pancreatic cancer?

Cancer genetics is an active area of research. Below is a list of genes that are currently known to be linked to hereditary pancreatic cancer. If genetic testing is pursued, you may ask your genetic counselor for a list of the specific genes included in your testing.

Gene(s)	Cancer(s)	Syndrome Name	Estimated Lifetime Risk of Pancreatic Cancer
<i>APC</i>	Colorectal, adenomatous polyps, pancreas	Familial adenomatous polyposis	2%
<i>ATM</i>	Breast, pancreas		3-5%
<i>BRCA1</i> and <i>BRCA2</i>	Breast, ovary, prostate, pancreas	Hereditary breast and ovarian cancer	5-7%
<i>CDKN2A (p16)</i>	Melanoma, pancreas	Familial atypical multiple mole melanoma	17%
<i>MLH1, MSH2, MSH6, PMS2</i> and <i>EPCAM</i>	Colorectal, endometrial (uterus), ovarian, stomach, pancreas	Lynch syndrome or Hereditary Non-Polyposis Colorectal cancer	9-11%
<i>PALB2</i>	Breast, pancreas		Unknown
<i>STK11</i>	Small bowel, breast, gastric, ovary, pancreas	Peutz-Jeghers syndrome	11-36%
<i>TP53 (p53)</i>	Breast, bone, brain, adrenal cortical, lung, colon and others	Li-Fraumeni syndrome	Unknown

What does it mean to test positive for a mutation in one of the genes listed above?

If you test positive for a mutation in one of these genes, your doctors may recommend a specialized medical plan to manage the increased cancer risks. If no mutation is identified, we may still recommend a specialized medical plan to manage your cancer risk based on your personal and/or family history. In addition, there is a 50% chance that a person with a mutation will pass it on to each of his/her children. In most cases, brothers and sisters also have a 50% chance to have the mutation. Other family members may also be at risk to have the mutation.

What is Familial Pancreatic Cancer (FPC)?

FPC is a term used for families that do not have an identifiable mutation in one of the genes above but who have ≥ 2 first degree family members with pancreatic cancer. If your genetic testing does not identify a mutation and you have multiple relatives with pancreatic cancer, your family may have FPC.

Number of Relatives with Pancreatic Cancer	Estimated Lifetime Risk of Pancreatic Cancer
None	1-2% (general population risk)
1 or 2 first degree relatives (sibling, parent, child)	4-7%
3 or more first degree relatives	17-32%

Adapted from Syngal et al. (2015)