Hereditary pancreatic cancer continues to pose challenges to providers, as well as to patients and their families. Pancreatic cancer has a poor prognosis, and individuals with family histories of pancreatic cancer are often motivated to pursue genetic testing. This article reviews various hereditary pancreatic cancer syndromes, as well as pancreatic cancer screening recommendations, options, and limitations.

AT A GLANCE

- Individuals at risk of hereditary pancreatic cancer should be referred to a genetics professional for formal genetic cancer risk assessment and genetic testing, when appropriate.
- Nurses should understand the cancer risks associated with various hereditary pancreatic cancer syndromes, inform patients that recommendations are continually evolving, and provide patient support to ensure that cancer prevention and early detection recommendations are implemented.
- Individuals and family members at increased risk of pancreatic cancer should be encouraged to adopt positive health behaviors and find ways to cope with emotional distress and adapt.

KEYWORDS
hereditary pancreatic cancer; gene mutation; genetic testing; risk assessment

DIGITAL OBJECT IDENTIFIER
10.1188/19.CJON.579-582

Hereditary Pancreatic Cancer Syndromes

Providing care to at-risk families

Elisabeth King, MSN, RN, FNP, AOCNP®, AGN, and Bita M. Nehoray, MS, LCGC

The American Cancer Society ([ACS], 2019) estimates that 56,770 new cases of pancreatic cancer will be diagnosed in the United States in 2019. Pancreatic cancer accounts for 3% of all cancers and 7% of all cancer deaths (ACS, 2019). Most pancreatic cancers are adenocarcinomas.

Symptoms of pancreatic cancer can include jaundice, abdominal or back pain, anorexia and weight loss, nausea, and vomiting (ACS, 2019). Blood clots, diabetes, or gallbladder or liver enlargement might also suggest pancreatic cancer. Because of the location of the pancreas within the body, pancreatic cancer is notoriously difficult to detect at earlier stages. Often by the time that a patient is symptomatic, his or her cancer has progressed to a more advanced stage. Individuals diagnosed with pancreatic cancer often have a poor prognosis, which is linked to the challenges associated with early detection; the five-year survival rate among those diagnosed with any stage of pancreatic cancer is just 9% (ACS, 2019).

The primary modifiable risk factors for pancreatic cancer include using tobacco and being overweight (ACS, 2019). Others include alcohol abuse and diets high in sugar and meat (Midha, Chawla, & Garg, 2016), as well as increasing age, gender (more common in men), race (more common in African Americans), diabetes, chronic pancreatitis, family history, and inherited genetic syndromes (Midha et al., 2016).

Hereditary Pancreatic Cancer Syndromes

Cancers are generally caused by acquired gene mutations that occur over time because of various exposures or random events, and pancreatic cancer is no different. However, some individuals inherit gene mutations that increase their risk of pancreatic or other cancers. About 10% of all individuals diagnosed with pancreatic cancer have a first- or second-degree relative with pancreatic cancer, suggesting an inherited risk (Syngal et al., 2015).

The most common gene mutations found in familial pancreatic cancer testing are in the *BRCA1*, *BRCA2*, *CDKN2A*, *PALB2*, and *ATM* genes (Syngal et al., 2015). Mutations in these genes have also been associated with other types of cancer, such as breast cancer, prostate cancer, and melanoma (see Table 1). Of the more commonly identified familial pancreatic cancer gene mutations, *CDKN2A* confers the highest lifetime risk of pancreatic cancer, and individuals with a *CDKN2A* mutation have as much as a 39-fold increased risk of developing pancreatic cancer within their lifetime (Syngal et al., 2015). Although relatively uncommon, *STK11* mutations confer the highest known risk of pancreatic cancer (132-fold) (Syngal et al., 2015). Mutations in *STK11* cause a condition called Peutz–Jeghers syndrome in which