Fact Sheet - *Hereditary Pancreatitis*

Hereditary Pancreatitis (HP) is a rare genetic condition characterized by recurrent episodes of pancreatic attacks, which can progress to chronic pancreatitis. Symptoms include abdominal pain, nausea, and vomiting. Onset of attacks typically occurs between within the first two decades of life, but can begin at any age. In the United States, it is estimated that at least 1,000 individuals are affected with hereditary pancreatitis.

HP has also been linked to an increased lifetime risk of pancreatic cancer. Pancreatic cancer is the 4th leading cause of cancer deaths among Americans. Individuals with hereditary pancreatitis appear to have a 40% lifetime risk of developing pancreatic cancer. This increased risk is heavily dependent upon the duration of chronic pancreatitis and environmental exposures to alcohol and smoking. One recent study suggested that individuals with chronic pancreatitis for more than 25 years had a higher rate of pancreatic cancer when compared to individuals in the general population. This increased rate appears to be due to the prolonged chronic pancreatitis rather than having a gene mutation (all cationic trypsinogen mutations). It is important to note that these risk values may be higher than expected because these studies on pancreatic cancer use a highly selective population rather than a randomly selected population.

At this time, there is no cure for HP. Treating the symptoms associated with HP is the choice method of medical management. Patients may be prescribed pancreatic enzyme supplements to treat maldigestion, insulin to treat diabetes, analgesics and narcotics to control pain, and lifestyle changes to reduce the risk of pancreatic cancer (for example, NO SMOKING!).

Dietary recommendations to help control pain with digestion include the consumption of small meals throughout the day that are high in carbohydrates and low in protein and fat. Pancreatic enzymes such as Creon, Pancrease, and Viokase are helpful in providing improved digestion and a reduction in diarrhea and pain for some patients with more advanced disease.

Exposure to smoking and alcohol are known to dramatically increase the risk for pancreatic attacks among individuals with HP. Smoking is strongly discouraged as it doubles the risk for pancreatic cancer. Similarly, alcohol consumption is not recommended for these patients because alcohol is a known risk factor for both acute and chronic pancreatitis. Therefore it is recommended that all HP patients avoid smoking and alcohol consumption.

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