Genetic aspects of tropical calcific pancreatitis.

Abstract:
Tropical calcific pancreatitis (TCP) is a subtype of chronic pancreatitis which is unique to tropical regions. Patients present at young age with recurrent abdominal pain, nutritional deficiencies, and insulin-requiring diabetes. For a long time, the aetiology of this disorder was poorly understood. Several environmental factors, such as malnutrition or the consumption of toxic food components such as cyanogenic glycosides, were proposed as pathogenic factors. In the last decade, a major impact on the understanding of the aetiology of TCP has come from genetic studies on hereditary and idiopathic chronic pancreatitis. Genetic alterations in at least five genetic loci are clearly associated with chronic pancreatitis in the Western world. These include alterations in genes coding for trypsinogens, the most abundant digestive enzymes (PRSS1 and PRSS2), the trypsin inhibitor (SPINK1) and the trypsin-degrading enzyme, chymotrypsinogen C (CTRC). In addition, alterations in the cystic fibrosis (CFTR) gene are associated with idiopathic pancreatitis. TCP clinically resembles non-alcoholic chronic pancreatitis of Western countries, suggesting that similar genetic defects might also be of importance in this disease entity. Indeed, alterations in at least two genes, SPINK1 and CTRC, are strongly associated with TCP. The current review focuses on the recent developments in the understanding of the genetic basis of inherited pancreatitis, with special emphasis on TCP.