Hereditary pancreatitis in children: surgical implications with special regard to genetic background.

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Résumé en anglais	**PURPOSE:** Hereditary pancreatitis (HP) is the primary etiology of chronic pancreatitis during childhood, progressing through recurrent episodes of acute pancreatitis and finally leading to pancreatic insufficiencies. Hereditary pancreatitis is because of mutations of the cationic trypsinogen (PRSS1) gene. Some other genes, such as SPINK1 or CFTR, have been associated with familial idiopathic chronic pancreatitis. The aim of our study was to clearly define diagnostic and therapeutic strategies for HP patients, through an analysis of our study group and a review of the literature. **METHODS:** All children admitted from 1995 to 2007 with a final diagnosis of hereditary pancreatitis were restrospectively included in the study. We analyzed all medical records with special attention given to cases involving genetic screening (PRSS1, SPINK1, and CFTR genes). **RESULTS:** Ten children were included. Eight had HP with PRSS1 mutation, 2 of them without a familial history of chronic pancreatitis. The 2 others patients had SPINK1 mutations. Three HP patients were operated on for acute complications of pancreatitis and are well with a mean follow-up of 5.5 years. No patient had pancreatic insufficiencies or weight loss. **CONCLUSIONS:** Hereditary pancreatitis is associated with severe pancreatitis, with a greater risk of developing pancreatic cancer. It must therefore be diagnosed correctly and treated to prevent its considerable complications.
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