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Therapeutic step-up strategy for management of hereditary pancreatitis in children

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ABSTRACT

Background/Purpose: Various different regimes exist for the treatment of hereditary pancreatitis in childhood. Here, we propose a therapeutic pathway with emphasis on endoscopic and surgical procedures.
Methods: From 2006 to 2013, 12 patients with a diagnosis of hereditary pancreatitis were prospectively included in a therapeutic step-up schema. The treatment outcome was evaluated and correlated to aetiological factors and pathoanatomic findings.
Results: After diagnostic work-up (laboratory data, ultrasound examination, magnetic resonance cholangiopancreatography and genetic testing), all 12 patients underwent early endoscopic retrograde cholangiopancreatography (ERCP), which was successfully performed in ten children. Obstructive pancreatitis was found in eight children, and required sphincterotomy, dilation and stenting for 12 months. In two children with unsuccessful ERCP, open surgical drainage procedures were performed. After a mean follow-up of 32 months all children are free of recurrence of pancreatitis without any impairment of everyday activities.
Conclusions: For children with hereditary pancreatitis, a therapeutic step plan with early ERCP and open surgical drainage procedures in case of impossible or insufficient endoscopic treatment prevents recurring pancreatitis and offers a normal quality of life without any major complications.
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Pancreatitis in children is rare and the high rate of idiopathic pancreatitis reported in children shows that the aetiology of childhood pancreatitis has been poorly understood in many cases. Recently, it has been shown that genetic factors play a far more important role in the development of chronic pancreatitis in infancy and childhood than previously thought.

Hereditary pancreatitis is defined as two first-degree relatives or at least three second-degree relatives in two or more generations with chronic pancreatitis for which there is no other aetiology [1]. Since the initial identification of a causative mutation in the cationic trypsinogen gene (PRSS1) by Whitcomb et al. [2], a few other mutations associated with chronic pancreatitis have been described. In addition, pancreatitis has been linked to mutations in the Serine Protease Inhibitor Kazal Type 1 (SPINK1), carboxypeptidase A1 (CPA1) and cystic fibrosis transmembrane regulator (CFTR) genes [3–6].

There is increasing understanding concerning the aetiology of hereditary pancreatitis and therapeutic options have recently been evaluated for adults with this disease [7,8]. Nevertheless, guidelines for the treatment of hereditary pancreatitis in childhood are still lacking.

Here, we evaluate therapeutic options in children with hereditary pancreatitis. Based on our experience, we propose a therapeutic pathway with emphasis on endoscopic and surgical procedures.

1. Patients and methods

From 2006 to 2013, we treated 12 patients with a diagnosis of hereditary pancreatitis at our Department of Pediatric surgery. All 12 patients were prospectively included in a therapeutic step-up schema including conservative management, endoscopic retrograde cholangiopancreatography (ERCP) and surgery (Fig. 1). We evaluated the success of treatment and correlated the initial findings and pathoanatomic variations with the available treatment options.

2. Results

From 2006 to 2013, 12 patients with hereditary pancreatitis were treated at our Department of Pediatric Surgery (Table 1). All children were younger than 16 years. Seven out of 12 patients had a family history of pancreatitis or pancreatic adenocarcinoma. In addition, our series included three siblings.

All patients presented with a history of diffuse abdominal complaints, until elevated amylase and lipase (at least three times over the limit) led to the diagnosis of pancreatitis. The average age at diagnosis was 8 years (range 4–15), but age at presentation did not

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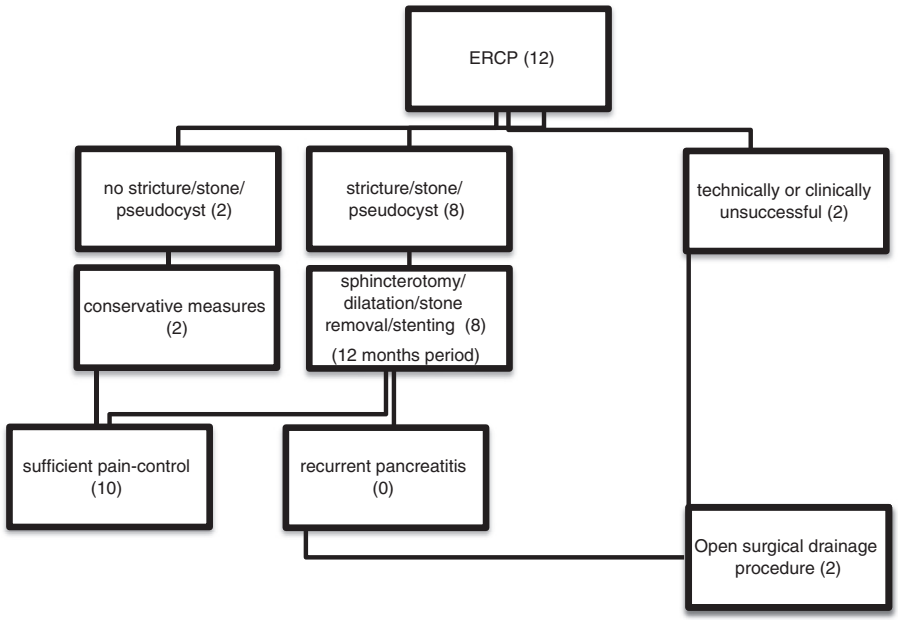

Fig. 1. Decision tree for therapeutic strategy in hereditary childhood pancreatitis in cases of insufficient medical management. Numbers of patients from this study are included in brackets.

correlate with either severity of the disease or with a specific cause. Eight girls and four boys were affected. In the absence of a causative agent, testing for genetic mutations in the PRSS1, SPINK1 and CFTR genes was performed. Different mutations and their frequencies are shown in Table 1. In one pair of siblings with chronic pancreatitis, no genetic variation had been found in these genes until now. Abdominal ultrasound and magnetic resonance cholangiopancreatography (MRCP) were performed in all patients to depict the distinct anatomy

Table 1
Patient characteristics of 12 children with hereditary pancreatitis.

Patient	Genetics	Age at diagnosis (years)	Age at first endoscopic procedure (years)	Complications	ERCP	Surgical treatment
1 male	SPINK1 heterozygosity	8	8		Terminated: impossibility of cannulating the papilla of Vateri	Longitudinal Pancreatico-jejunostomy
2 female	SPINK1 heterozygosity, heterozygous mutation in the ABCB 4 gene	4	4	Pancreatic pseudocyst	Sphincterotomy, stenting, stone removal, balloon dilation	
3 female	CFTR compound heterozygosity	9	9		Sphincterotomy, stenting	
4 male	No mutation found	4	4	Pancreticopleural fistula, intrathoracic pseudocyst	Sphincterotomy, stenting, balloon dilation	
5 female	No mutation found	6	11	Pancreatic pseudocyst	Sphincterotomy, stenting, stone removal	
6 female	PRSS1 heterozygosity	4	5		Terminated: duodenal stenosis	Longitudinal pancreatico-jejunostomy
7 male	PRSS1 heterozygosity	7	7		Sphincterotomy, stenting, stone removal, balloon dilation	
8 male	SPINK1 homozygosity	9	9		Diagnostic	
9 female	SPINK1 homozygosity, CFTR heterozygosity	12	12		Diagnostic	
10 female	PRSS1 heterozygosity	12	12		Sphincterotomy, stenting, stone removal, balloon dilation	
11 female	PRSS1 heterozygosity	15	16		Sphincterotomy, stenting, stone removal, balloon dilation	
12 female	PRSS1 heterozygosity	15	15		Sphincterotomy, stenting	

Patients 1 + 2, 4 + 5 and 8 + 9 are siblings.

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