# A two-year old boy with recurrent bouts of acute abdominal pain

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## Abstract

**In a small number of patients with pancreas divisum (with stenotic minor papilla) a relative obstruction to pancreatic exocrine secretory flow results in pancreatitis.**

**We report a 2-year-old boy presenting with recurrent bouts of abdominal pain. The diagnosis of acute pancreatitis was made based on blood biochemistry results. Ultrasound, computed tomography and magnetic resonance imaging showed several abdominal pseudocysts, peritoneal exsudate and confirmed pancreatitis but initially failed to reveal the aetiology. Ascites and cysts contained pancreatic enzymes. After weeks of combined conservative and surgical treatment, a magnetic resonance cholangiopancreaticography with secretin, showed a pancreas divisum with a cyst between the ducts of Santorini and Wirsung. Based on these findings, two endoscopic papillotomies (minor and major papilla) were performed. Three years follow-up was uneventful. In a child with recurrent pancreatitis or pancreatitis with chronic recurrent abdominal pain it is crucial to search aggressively for congenital abnormalities, including pancreas divisum. Secretin-enhanced magnetic resonance cholangio-pancreaticography or diffusion-weighted magnetic resonance imaging is a valuable diagnostic tool for visualizing pancreatic duct anatomy.** (Acta gastroenterol. belg., **2010**, 73, **517-520**).

**Key words :** pancreatitis, pancreas divisum, paediatric, s-MRCP, ERCP.

### Abbreviations

CT : computed tomography
DW-MRI : diffusion-weighted magnetic resonance imaging
ERCP : endoscopic retrograde cholangiopancreaticography
MRCP : magnetic resonance cholangiopancreaticography
PD : pancreas divisum
s-MRCP : secretin-enhanced MRCP

## Introduction

Pancreas divisum (PD), occurs in approximately 10% of the population (1-3) and is the most common congenital pancreatic abnormality (4,5). The phenomenon occurs when the ductal systems of the ventral and dorsal pancreatic ducts fail to fuse. As a result of non-union of the ducts, a major portion of pancreatic exocrine secretions enter the duodenum via the dorsal duct and minor papilla (1). Acute and chronic pancreatitis can be associated with PD (6-8). Diagnosis is usually made by magnetic resonance cholangiopancreaticography (MRCP), but can also be missed (9-11). However the diagnosis is crucial for the correct treatment.

## Case report

A previously healthy 2-year-old boy presented with recurrent bouts of acute abdominal pain since one month. The sole finding on physical examination was abdominal tenderness. Amylase and lipase serum levels were 500 U/L (normal : 30-110) and 6,150 U/L (normal : 23-300). Infectious parameters were all normal. An ultrasound of the abdomen showed a nodular hyperechogenic cystic structure surrounded by liquid, localized near the processus uncinatus of the pancreas. Computed tomography (CT) scan confirmed the presence of pancreatitis with a pancreatic cyst, surrounded by fluid, in the head of the pancreas. The CT-scan also revealed a small collection underneath the diaphragm and near the head of the pancreas. Based on biochemical results and imaging, acute pancreatitis with pseudocysts was diagnosed and the patient was referred to our hospital.

Parents mentioned a car accident five months prior to the first symptoms. Physical examination on admission revealed no peritoneal signs. The boy was mostly complaining of night time pain. An extensive search for aetiology was undertaken. There were no indications for infectious, malignant, metabolic or genetic aetiologies. A traumatic or obstructive cause was therefore suspected. MRCP, performed initially without secretin, showed a single diffusely oedematous cyst of 12 mm in the pancreas head and a normal Wirsung duct without stenosis, dilatation, calcifications or other abnormalities.

The patient was treated conservatively with nasogastric feedings using a amino-acid formula (Neocate®). His clinical condition deteriorated at day 10 with increased abdominal pain, fever and tachycardia, necessitating transfer to the paediatric intensive care unit. C-reactive protein level rose to 31.4 mg/dL (normal : < 1.0), white blood cell count was 11,800/mm$^3$ (normal : 6,000-17,000). Physical examination now showed peritoneal signs with shifting dullness. Ultrasonography demonstrated several pseudocysts around the pancreas and

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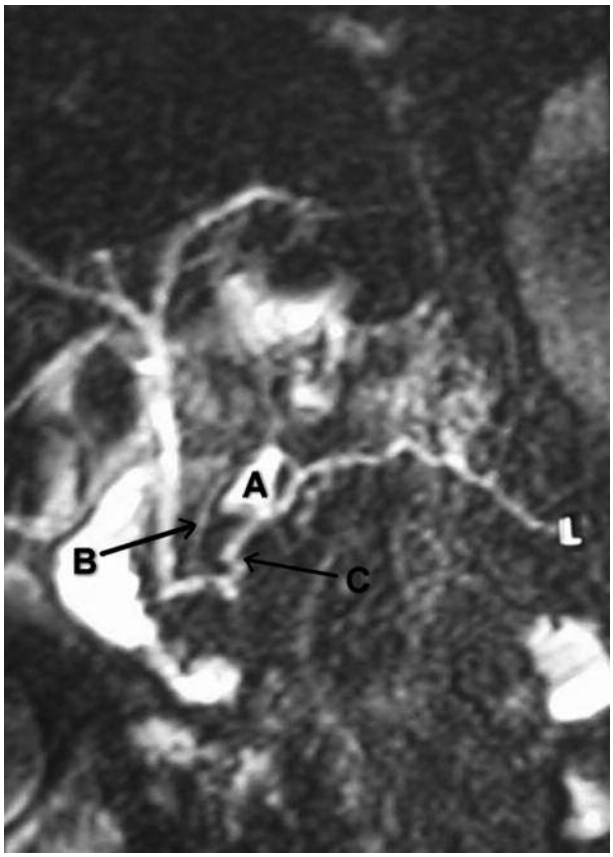
peritoneal exsudate in the pelvis. A CT-scan confirmed pancreatitis with pseudocysts and ascites. Pseudocysts were punctured, fluid was rich in amylase (10,718 U/L) and lipase (> 200,000 U/L) and infected with *E. coli*. The treatment was altered to total parenteral nutrition, morphine, antibiotics (amoxicillin, -clavulic acid, fluconazol) and somatostatin iv.

The patient's clinical condition remained unstable despite dropping serum amylase and lipase levels. The boy developed pleural effusion with respiratory distress and tachypnea. Diuretics were tried unsuccessfully. A follow-up ultrasound (day 14) showed the absence of resolution of pseudocysts and ascites. Surgery was required (laparotomy). The pseudocysts near the kidney were deroofed and drained, ascites was removed. The pancreas itself did not appear necrotic. Pleural fluid was aspirated during the procedure : it was an exsudate as it did not contain pancreatic enzymes.

After surgery the patient was ventilated mechanically for 4 days (days 14-17). Due to colonization of the ascites by *E. coli* and of the central catheter by coagulase-negative *staphylococci*, the antibiotics were altered to meropenem and vancomycin. C-reactive protein level normalized and finally his high temperature dropped to normal as well. A week after extubation his clinical condition improved. Enteral feedings, with amino-acid formula, were carefully reintroduced. Somatostatin was reduced progressively and stopped. On day 25 ultrasonography showed shrinkage of the pseudocysts. The cyst formed in the head of the pancreas however, remained unchanged. Amino-acid based was replaced by soy based enteral formula and although the patient was clinically stable, serum amylase and lipase levels increased again (292 U/L and 3,659 U/L respectively) (day 30). Evidence of relapse was not documented by ultrasonography and CT-scan. Neocate® and somatostatin were reintroduced. The pancreatic enzymes decreased rapidly. Still the aetiology was unclear, the car accident mentioned priorly seemed to have been uneventful and could hardly explain this severe clinical picture, CT-scan and MRCP were not diagnostic for congenital cyst (such as a choledochal cyst), a duplication of the pancreas or PD. There was no evidence for a tumourous mass (such as a pancreatoblastoma) nor were there any malignant cells in the punctured fluids. Prior to resuming oral fluids and feeds, the patient was transferred (day 36) in ordered to obtain a secretin-enhanced MRCP (s-MRCP). s-MRCP finally revealed a PD with a cyst between the Santorini and Wirsung ducts (Fig. 1). Based on this finding, endoscopic retrograde cholangiopancreaticography (ERCP) was subsequently performed. Stenting was impossible due to the twisting and curly aspect of the Wirsung duct (Fig. 2). Thus, endoscopic papillotomies (of the major and minor papilla) were performed to relieve the outflow obstruction. Subsequently the boy recovered fully, tolerating oral feeding and not needing any medication. After a follow-up of 3 years his course has remained uneventful.




**A** = cyst, **B** = Santorini duct, **C** = Wirsung duct.

Fig. 1. — Secretin-enhanced MRCP

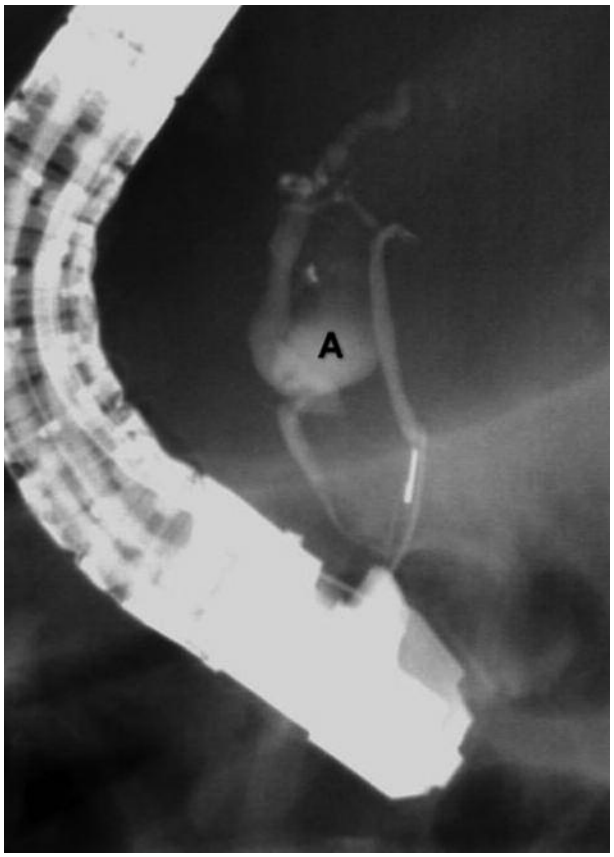
## Discussion

This case illustrates the usefulness of s-MRCP in revealing PD with a ductular cyst and a twisty, curly Wirsung associated with severe pancreatitis.

PD, the most common congenital pancreatic abnormality, is often considered innocuous but can lead to recurrent attacks of acute pancreatitis or abdominal pain in children (4-8).

Early in the organogenesis (at the 6th or 7th week of gestation) the normal adult pancreas forms from fusion of the ventral and dorsal pancreatic buds. PD occurs when the ductal systems of the ventral and dorsal pancreatic ducts fail to fuse and to communicate (1). As a result, a major portion of pancreatic exocrine secretions enter the duodenum via the dorsal duct and minor papilla. A relative obstruction to pancreatic exocrine secretory flow through the minor duct and minor papilla can result in pancreatitis in a small number of patients with PD (with stenotic minor papilla).

The role of PD in causing acute and relapsing pancreatitis and bouts of abdominal pain is controversial (2,6). Most cases have been reported with onset of symptoms in adulthood, although the anatomical anomaly is present from birth. The reported experience of pancreatitis with




**A** = cyst.

Fig. 2. — ERCP

PD in childhood is limited and correlation between these disorders in children has not been well elucidated (8).

MRCP is increasingly used in children. Conventional MRCP is a useful, non-invasive tool that can demonstrate strictures, dilatations and irregularities of the main pancreatic ducts such as congenital malformations (12-16). However, as stated by many investigators, there are a high numbers of false negative results (9-11). The diagnostic accuracy of this examination can be increased with 3D-MRCP or dynamic MRCP with secretin stimulation (12,13,17). s-MRCP is a non-invasive diagnostic procedure that relies on the dynamic response of the main pancreatic after secretin stimulation. Secretin causes accumulation of fluid and bicarbonates in the ductal system and subsequent enlargement of the ducts. This increase in calibre improves the assessment of the morphology of pancreatic ducts and their abnormalities. The tail end and side branches of the main pancreatic duct are difficult to assess with conventional MRCP. The accuracy in assessing ductal abnormalities, improves after secretin administration. s-MRCP not only facilitates the depiction of anatomic variations or morphologic changes of the pancreatic duct in the normal and diseased pancreas but also helps assessing functional abnormalities of the exocrine pancreas (10,17,18).

Matos *et al*. studied the frequency of PD in 279 patients. Thirty patients (10.8%) had a PD shown with s-MRCP. Secretin stimulation improved the detection of PD in 23% (7/30) (2). A recently developed technique, secretin-stimulated diffusion-weighted magnetic resonance imaging (DW-MRI), is even more accurate. DW-MRI may help to detect mild or early pancreatitis and can evaluate the pancreatic exocrine function (12). Due to the advent of these less invasive and safer diagnostic modalities the role of ERCP has evolved from a largely diagnostic to a mostly therapeutic tool in the management of acute pancreatitis and its complications. Therapeutic (and diagnostic) ERCP results are similar in children and adults (12,19-22).

PD by itself should not necessarily require intervention but it is important to search aggressively for PD in children with recurrent pancreatitis or pancreatitis with a history of chronic recurrent abdominal pain (4,8). There is no consensus regarding the appropriate treatment for pancreatitis associated with PD. Surgical intervention is directed toward relief of ductal obstruction (8). Endoscopic pancreatic stent placement after minor papilla and dorsal duct dilatation provides safe and effective endoscopic treatment in patients with PD and acute recurrent pancreatitis. Endoscopic papillotomy of the minor papilla appears to yield improvement in most cases with acute recurrent pancreatitis, but long-term outcomes of minor papilla endotherapy in PD are limited (23,24). Patients with PD and bouts of pancreatitis benefited from minor papilla sphincterotomy more frequently than those without symptom-free intervals or those with abdominal pain but without elevated pancreatic enzymes (25,26). Minor papilla sphincterotomy offers advantages over chronic stent therapy in treating patients with PD. The complications, such as acute pancreatitis or stenosis of the minor papilla, seem to be less frequent after minor papilla sphincterotomy than after pancreatic stent insertion. These complications are mostly managed conservatively (27-29).

In conclusion, we recommend that in a child with recurrent acute or chronic pancreatitis, the search for congenital abnormalities including PD, is actively pursued. When no aetiology is demonstrated, s-MRCP or DW-MRI imaging should definitely be obtained.

In this case, PD with a cyst led to outflow obstruction. The young patient was successfully treated by endoscopic papillotomies, which resulted in a favourable mid-long term follow-up.

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