

## CASE REPORT

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# Role of Endoscopic Ultrasound in the Diagnosis of Agenesis of the Dorsal Pancreas

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### ABSTRACT

**Context** Complete agenesis of the dorsal pancreas is an unusual congenital anomaly. Nowadays, the diagnosis is based on four imaging studies: transabdominal ultrasonography, computed tomography, magnetic resonance cholangiopancreatography and, the gold-standard, endoscopic retrograde cholangiopancreatography. Sometimes the ability of these studies are limited to distinguishing agenesis of the dorsal pancreas from other congenital abnormalities. Endoscopic ultrasound is a minimally invasive technique which permits us to obtain high resolution images of the pancreatic parenchyma and ductal system.

**Case report** We report the case of a 23-year-old woman with recurrent episodes of acute pancreatitis and non-conclusive classic imaging studies. Complete agenesis of the dorsal pancreas was demonstrated by endoscopic ultrasound.

**Conclusion** Endoscopic ultrasound may be useful in the diagnosis of agenesis of the dorsal pancreas.

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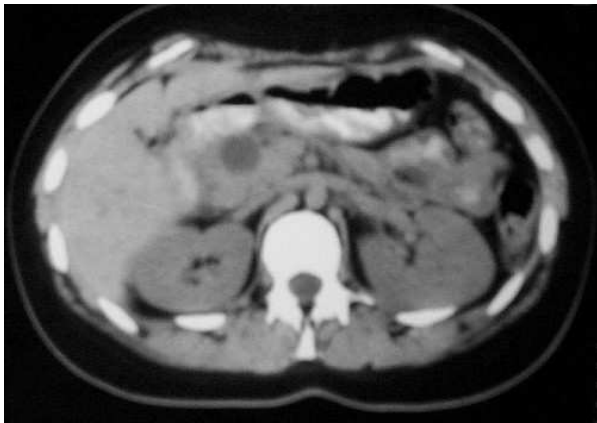
### INTRODUCTION

Complete agenesis of the dorsal pancreas is an unusual congenital anomaly. Nowadays

the diagnosis is based on four imaging studies: transabdominal ultrasonography (US), computed tomography (CT), magnetic resonance cholangiopancreatography (MRCP) and, the gold-standard, endoscopic retrograde cholangiopancreatography (ERCP) [1]. Sometimes the ability of these studies is limited to distinguishing agenesis of the dorsal pancreas from another congenital abnormalities [2]. Endoscopic ultrasound (EUS) is a new technique which permits us to obtain high resolution images of the pancreatic parenchyma and ductal system [3, 4]. We present a case of complete agenesis of the dorsal pancreas, with non-conclusive classic imaging studies, in which EUS played an important role in the diagnosis.

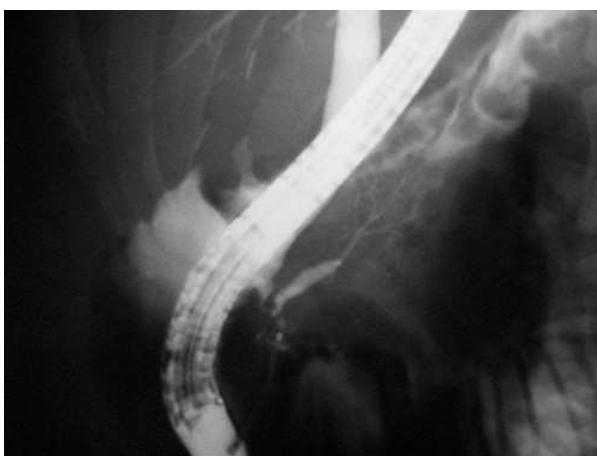
### CASE REPORT

A 23-year-old woman, without any history of diabetes, was admitted to our hospital to assess recurrent episodes of acute pancreatitis. An episode of severe biliary acute pancreatitis with a complicated pancreatic pseudocyst at the head of the pancreas had been reported 4 years earlier. Cholecystectomy, cystogastrostomy and cystoduodenostomy were performed after the episode. One year later, the patient developed a new episode of acute pancreatitis. An abdominal CT revealed a pancreatic pseudocyst (2.3 cm) at the head of the pancreas (Figure 1). An ERCP showed the main pancreatic duct with a length of 30 mm,

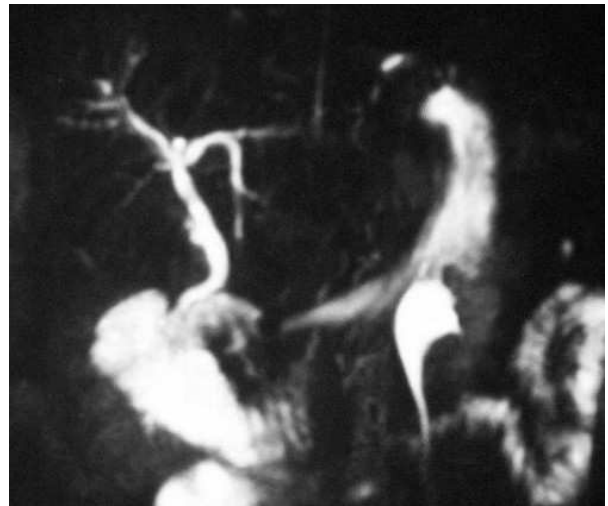


**Figure 1.** Abdominal CT showing pancreatic pseudocysts at the head of the pancreas.

sharp-ended and with multiple normal side branches (Figure 2). With these findings the original diagnosis was pancreas divisum. In a subsequent MRCP, the head of the pancreas was prominent having a short main pancreatic duct without any evidence of a dorsal pancreas (Figure 3). Different etiologies such as toxins and metabolic or autoimmune disorders were excluded. Intermittent epigastric pain with normal values of serum amylase and lipase had been present for more than 2 years. A transabdominal US showed a cyst (2 cm) at the head of the pancreas. A secretin-stimulated MRCP demonstrated a pseudocyst at the head of the pancreas and a short, irregular main pancreatic duct. The accessory pancreatic duct and the pancreatic dorsal parenchyma were not visible. A new ERCP showed a normal common bile duct



**Figure 2.** Endoscopic retrograde cholangiopancreatography showing a short main pancreatic duct with sharp-ended and multiple normal side branches.



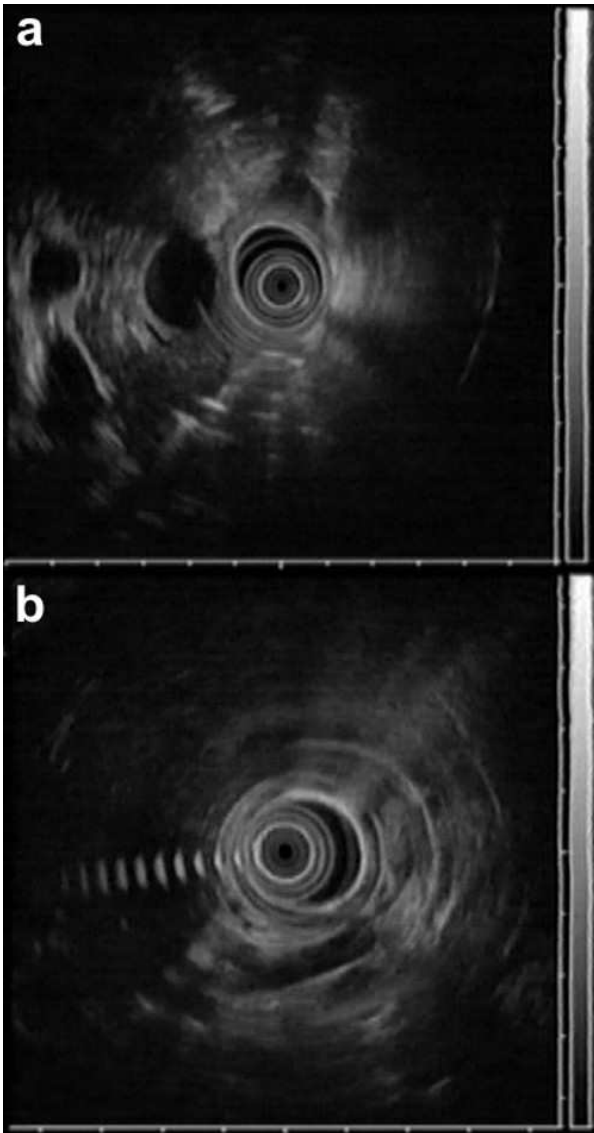
**Figure 3.** Magnetic resonance cholangiopancreatography showing a prominent short main pancreatic duct.

and a short main pancreatic duct with multiple side branches. A small prominence on the mucosa was detected in the descending part of the duodenum, but cannulation of this structure was impossible. An oral glucose tolerance test and Van de Kamer test (5.9 g/24h; reference range: 1-7 g/24h) were within normal limits but the fecal elastase concentration was low (103 U/g; reference range: 0-200 U/g). Finally, an EUS was performed. It showed the ventral pancreas and a main pancreatic duct of normal caliber, increased duct wall echogenicity and cystic lesion (1.6x2.2 cm). EUS did not show the accessory pancreatic duct and the dorsal pancreas. The splenic vessels were in contact with the posterior wall of the stomach (Figure 4).

The diagnosis of our patient was controversial because of the small prominence on the mucosa of the duodenum. An EUS and the review of the original imaging studies permitted us to conclude that the patient presented agenesis of the dorsal pancreas.

## DISCUSSION

The pancreas is formed by two separate anlagen, each from a ventral and a dorsal endodermal bud. The ventral bud forms the lower portion of the pancreatic head and the uncinata process while the dorsal bud forms



**Figure 4.** Endoscopic ultrasound. **a.** Ventral pancreas and main pancreatic duct having a normal caliber, increased duct wall echogenicity and a cystic lesion (1.6x2.2 cm). **b.** Absence of the accessory pancreatic duct and the dorsal pancreas. The splenic vessels are in contact with the posterior wall of the stomach.

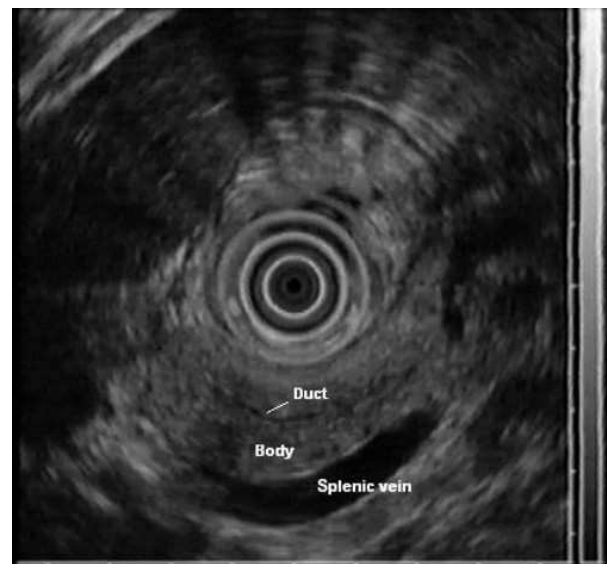
the upper head, neck, body and tail of the pancreas. Each anlagen possesses its own duct system. The pancreas presents a complicated embryogenesis between the 5<sup>th</sup> and the 7<sup>th</sup> week of gestation [5, 6]. At the 6-7<sup>th</sup> week of gestation, the ventral pancreas fuses with the dorsal pancreas. During the fusion, the ventral and the dorsal ducts form the main pancreatic duct. The accessory pancreatic duct is formed from the portion of the dorsal bud which gives rise to the upper pancreatic head. A disorder during the complicated embryologic-

al development of the pancreas can lead to congenital abnormalities.

Complete agenesis of the pancreas and agenesis of the ventral pancreas are unknown congenital abnormalities because complete agenesis of the pancreas is incompatible with life [7] and the agenesis of the ventral pancreas is extremely rare [1].

Agenesis of the dorsal pancreas is an uncommon congenital abnormality originating from a defect in early embryogenesis which may also be associated with other congenital abnormalities [8, 9]. Primary dysgenesis and ischemic injury to the development of the pancreas could play a role in the etiology [10]. Family cases have been reported in the literature, but the genetic transmission remains unclear [11]. Agenesis of the dorsal pancreas may be partial or complete; partial agenesis is more frequent than complete agenesis. In complete agenesis, the minor papilla, accessory pancreatic duct, body and tail of the pancreas are absent. In partial agenesis, the papilla minor with a remnant of the accessory pancreatic duct and the body of the pancreas are present [12].

We have reviewed 16 recent case reports of complete agenesis of the dorsal pancreas [1, 2, 11, 12, 13, 14, 15, 16, 17, 18, 19, 20, 21].



**Figure 5.** Endoscopic ultrasound in a healthy person: the body and the tail of the pancreas are visualized between the posterior wall of the stomach and the splenic vessels.

To assess the clinical presentation we excluded 2 cases with several concomitant pathologies (tumor in the head of the pancreas and diffuse peritoneal carcinosis of the adenocarcinoma) [19, 20]. In the other 14 cases, 6 men and 8 women with a mean age of 42 years, the most frequent clinical events were abdominal pain (13 cases, 92.9%) and the development of diabetes mellitus (6 cases, 42.9%). Acute pancreatitis episodes, present in our patient, were described in 7 cases (50.0%). Some authors suggest that abdominal pain is more common in patients with partial agenesis and diabetes mellitus is more common in patients with complete agenesis of dorsal pancreas [12, 15]. The relationship between agenesis of the dorsal pancreas and exocrine pancreatic insufficiency remains unclear; only one case has been reported [13]. The fecal elastase concentration of our patient was low, but there were no other findings at clinical presentation, function tests or diagnostic imaging to confirm the diagnosis of exocrine pancreatic insufficiency.

The diagnosis of agenesis of the dorsal pancreas requires a high index of suspicion in addition to imaging studies. Before the use of modern diagnostic imaging techniques, the diagnosis was made by autopsy or laparotomy [11]. Nowadays, transabdominal US, CT, MRCP and ERCP allow the diagnosis of agenesis of the dorsal pancreas. However, there are some patients, such as ours, in which the classical imaging techniques are not able to differentiate between agenesis of the dorsal pancreas and other congenital abnormalities. Transabdominal US in our patient did not reveal any diagnostically suspicious findings. Congenital abnormalities can only occasionally be identified by transabdominal US because the bowel gas obscures the pancreas. However, a hyperechoic line of demarcation between the head of the pancreas and the retroperitoneal fat with the presence of a small hypoechoic mass in the pancreas, just ventral to the portal confluence, has been reported in agenesis of the dorsal pancreas [14, 15]. Contrast-enhanced CT has frequently been used with ERCP to evaluate

pancreatic abnormalities [13]. In complete agenesis of the dorsal pancreas, only the pancreatic head is visible [13, 14]. A dorsal pancreatic parenchyma was not detected in our patient at either CT examination. In recent years, MRCP has been used as a non-invasive alternative to ERCP for the evaluation of the pancreatic ductal system and congenital abnormalities [22]. On MRCP, the presence of a short main pancreatic duct in the normal head of a pancreas with the absence of pancreatic dorsal tissue and an accessory pancreatic duct, such as in our patient, are the characteristics of complete agenesis of the dorsal pancreas [1, 15]. The inability of performing biopsies and interventional approaches are the disadvantages of MRCP. ERCP is an invasive technique with an important morphologic assessment of the pancreatic ductal system, and is considered the gold standard for agenesis of the dorsal pancreas [12]. Furthermore, brush cytology and therapeutic intervention are possible during ERCP. Invasivity, dependency on the operator and difficulty in the identification and cannulation of the papilla minor are the disadvantages of ERCP [1]. In our patient, the presence of multiple side branches in the main pancreatic duct and the small prominence in the mucosa detected in the second segment of the duodenum did not allow us to reach a definitive diagnosis.

EUS is a relatively new imaging technique which provides direct visualization of the total pancreatic parenchyma and the ductal system [3]. The role of EUS in the identification of agenesis of the dorsal pancreas has not been evaluated, but it may be as good as ERCP. The head of the pancreas, uncinate process, ampulla and distal pancreatic ducts (main and accessory ducts) can be examined with the EUS positioned in the first and second part of the duodenum [23, 24]. "Stripping" is necessary in order to visualize the body and the tail of the pancreas. "Stripping" is the retraction of the EUS device from the duodenum to the gastric antrum. With the EUS positioned in the gastric antrum, the body and the tail of the pancreas are visualized between the posterior

wall of the stomach and the splenic vessels [4] (Figure 5). Agenesis of the dorsal pancreas may be suspected, as in our patient, if the accessory pancreatic duct and the dorsal pancreas (without any structure between the stomach and the splenic vessels) are not visualized. The most crucial differential diagnosis in the agenesis of the dorsal pancreas is a pancreatic carcinoma with proximal atrophy [15]. Electronic linear array EUS has a significant impact on the diagnosis of pancreatic carcinoma, it provides excellent resolution images of the pancreatic parenchyma and the ductal system and the possibility of performing fine-needle aspiration [3, 4].

In summary, complete agenesis of the dorsal pancreas can be mimicked by other congenital abnormalities requiring different clinical management. EUS is a minimally invasive technique which may be useful in the diagnosis of agenesis of the dorsal pancreas.

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**Keywords** Digestive System Abnormalities; Endosonography; Pancreatitis

**Abbreviations** CT: computed tomography; ERCP: endoscopic retrograde cholangiopancreatography; EUS: endoscopic ultrasound; MRCP: magnetic resonance cholangiopancreatography; US: ultrasonography

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