Reviewing the wide spectrum of congenital variants and anomalies from the Pancreas and Pancreatic Ducts - from the asymptomatic to the clinically relevant

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Learning objectives

• Start with a brief review of pancreatic embryology;
• Proceed with a comprehensive approach to the most common congenital variants and anomalies of the pancreas and its duct system, with emphasis on magnetic resonance cholangiopancreaticography and multidetector computed tomography.
Background

Pancreas and pancreatic ducts have a wide spectrum of anatomical variations. In one hand, some of them may not be detected until adulthood as they are often discovered as an incidental finding in asymptomatic patients, doing exams for unrelated causes[1,2]. On the other hand, some of these anomalies may lead to clinically relevant signs and symptoms, such as relapsing abdominal pain, nausea and vomiting, resulting from recurrent pancreatitis or gastric outlet obstruction [2].

Radiologists also need to be familiar with these entities, since some might mimic other pathologic conditions, like various neoplastic, inflammatory, and posttraumatic conditions [3].

In this poster we will review the following morphologic variants and developmental abnormalities of the pancreas and its ducts:

- Variations in the pancreatic duct's course
- Direction and shape;
- Configuration and duplication anomalies;
  - Pancreas divisum;
  - Annular pancreas;
  - Ectopic pancreas;
  - Other variations of the pancreatic contour or constitution.
EMBRIOLOGY

Understanding the embryologic development of the pancreas and its duct system will help in the comprehension of this group of disorders.

The pancreas develops from two focal outgrowths of the primitive foregut, one dorsal and one ventral bud, both first appearing in the fourth-fifth week of gestation [1,3]. The gallbladder, major bile ducts, ventral pancreas and respective duct system are derived from a ventral (caudal) outpouching, and will rotate clockwise, posterior to the duodenum, to join the dorsal bud derivates, which are the precursors of the anterior part of the pancreatic head, body, tail of the pancreas and respective ductal system [1,3]. This common embryologic origin of ventral pancreatic duct and main bile ducts is therefore responsible for the most common adult pancreatic ductal system configuration, with both sharing a common entrance into the duodenum at the major duodenal papilla [3].

Approximately at the 7th gestational week, usually the dorsal and ventral pancreatic ducts fuse by the isthmus region. There are variations about the territory drained by each system. Most commonly the dorsal component of the ductal system drains the tail, body, and anterior portion of the pancreatic head, whereas the ventral duct system drains the posterior aspect of the pancreatic head [3]. Drainage of the uncinate process is quite variable and it might be either through dorsal or ventral aspects of the duct system[1,3].

Hence, we call duct of Wirsung to the portion of the ventral duct between the dorsal-ventral fusion point and the major papilla[1,3], and we say there is a duct of Santorini (or accessory duct) if a segment of the dorsal duct persists distal to the dorsal-ventral fusion point. Santorini’s duct is not constant. Thirty per cent of individuals with a Santorini’s duct don’t have a communication between this duct and the minor duodenal papilla[3]. The term main pancreatic duct is reserved for the ductal system upstream from the dorsal-ventral fusion point, inside the body and tail of the pancreas and derived from the dorsal precursor[1,3]. In most cases (80%-90%), the common bile duct and the Wirsung’s duct unite within a sphincteric complex (the sphincteric complex of Oddi which drain into the major papilla). This common channel usually measure about 10-15 mm in length. [3]

APPROACHING TO THE MOST COMMON CONGENITAL VARIANTS AND ANOMALIES OF THE PANCREAS AND ITS DUCT SYSTEM
Although the majority of these findings are asymptomatic, recognizing the wide spectrum of anatomical variants of pancreas and its duct system is important because these anomalies may be a surgically correctable if symptomatic[2,3]. Imagers should also be aware for these anomalies, since reporting it may guide surgical planning and prevent inadvertent structural injury [2].

Variations of the Pancreatic Duct course

The most common directions and shapes[1] of pancreatic duct course are:

- a descending course(50%);
- sigmoid shaped course;
- vertical course;
- loop shaped course.

The most common ductal configurations are:

- a bifid configuration (60%) with a dominant duct of Wirsung;
- an absent duct of Santorini (30%);
- a dominant duct of Santorini without divisum (1%);
- an 'ansa pancreatica', in which the duct of Santorini connects with a Wirsung side branch, forming a reversed-S shape

Wirsungocele and Santorinicele might also be seen in association with other conditions, and represent cystic dilatations of terminal portions of each duct.

Even when bifid in configuration an anomalous pancreaticobiliary ductal junction might be of clinical relevance. Hence, variations like a long common channel (>15mm), characterized by a precocious fusion of the pancreatic and common biliary duct (CBD) duct, might be prone for choledochal cyst formation and biliary tract carcinoma[1].

### Pancreas Divisum

This is the commonest congenital pancreatic duct variation and is reported in about 4-14% of the cadaver studies[1,2]. Pancreas Divisum results from a fusion failure of the dorsal and ventral pancreatic primordium, during the sixth to eighth gestation weeks. [1,2] The clinical relevance of this condition remains controversial, since most patients remain asymptomatic. However, some patients have several episodes of pancreatitis, representing about 12-26% of the presumed idiopathic recurrent pancreatitis [1].
The key role of ERCP in the diagnosis of pancreas divisum has been challenged by MRCP, since this imaging method doesn’t need the contrast material and is noninvasive, avoiding the risk of ERCP-induced pancreatitis [1,2]. The administration of secretin has been used to increase the sensitivity of MRCP in the diagnosis of pancreatic divisum.

With the advent of MDCT scanners, pancreas divisum may be seen using CT as well [1] and the use of minIPs have also been advocated.

The majority of the cases of pancreas divisum fits within one of these types:

- Type 1, also called classical divisum: The dorsal duct does not communicate with the ventral duct[1,2] - representing the majority of cases.

- Type 2 where the ventral duct is absent [2]: in these cases all pancreas is drained through the minor papilla, and the major papilla only drains the bile ducts.

- Type 3, also known as functional, where there is a small filamentous or inadequate communication between the two ducts[2];

Generally speaking the dorsal pancreatic duct continues through the duct of Santorini, converging into the minor ampulla through which most pancreatic secretions are expelled. The ventral duct may or may not be present, and if present it only drains the posterior segment of the pancreatic head and uncinate process through the major ampulla, where bile secretions are also expelled [1,2].

### Annular Pancreas

Annular pancreas occurs in 1/20000 of the population [1,2] and is an anomaly where a band of pancreatic tissue, which in continuity with the pancreatic head, encircles completely or incompletely the descending duodenum, sometimes assuming a 'crocodile jaw' configuration. An widely accepted theory for its etiopathogenesis is that the ventral pancreatic primordium fails to rotate [2] and suffers an early division into two segments[1]. This anomaly may be asymptomatic for life, as in adults patients, where an incidental discovery represent approximately 50% of the cases, but in may also be symptomatic. In fact, this condition may associated with duodenal stenosis, often in neonates[2], or typically may present in the third to sixth decades with abdominal pain, postprandial fullness and vomiting resulting from gastric outlet obstruction, upper gastrointestinal bleeding from peptic ulceration, pancreatitis either acute or chronic, and in rare instances jaundice due to biliary obstruction[2]. Some authors recommend surgical intervention for symptomatic cases[1].
Oral contrast exams often reveal characteristic images, with narrowing of the second portion of the duodenum. CT or MR images may reveal normal pancreatic tissue encircling the duodenum, with or without a small pancreatic duct. [1] If a duct extending to the right side is discernible by ERCP, simple or secretin-enhanced MRCP, the diagnosis of Annular Pancreas established [2].

### Ectopic Pancreatic Tissue

Ectopic rests of pancreas, without anatomic or vascular continuity with the normal pancreas[2], may be located either in the submucosa of the gastric antrum (70%)[2], in the proximal portion of the duodenum[1] or in the jejenum [2]. Rarely it might be located in the mesentery, gallbladder, ileum, Meckel diverticulum, appendix or colon [2]. This ectopic tissue usually measures between 0.5 and 2.0 cm in its larger diameter, but in rare cases it might measure up to 5 cm.

The majority of these cases represent an incidental finding [2], but there may be functional ectopic tissue, which is subject to the same disorders that affect the normal orthotopic pancreas[1,2] and may also cause complications such as ulceration, bleeding, intussusception or bowel obstruction. Oral contrast exams may show an extramucosal, smooth, broad-based lesion either at gastric antrum, along the greater curvature, or in the proximal duodenum[1,2], and some authors describe a typical small collection of contrast within 45% of the cases of ectopic pancreas, representing a central niche or umbilication, remnant of an orifice of a rudimentary pancreatic duct [1,2]. Laparoscopic wedge resection may be a treatment option for symptomatic patients[1].

### Other variations of the pancreatic contour or constitution

Sometimes even a healthy pancreatic tissue may assume unusually lobulated contours, especially in the lateral aspect of the head and neck, where it might mimic a pancreatic neoplasm, [1] a peripancreatic metastatic tumor deposit or a lymphadenopathy [2]. To avoid diagnostic errors we should look for attenuation, signal intensity and pattern of enhancement, which in this variations lacking pathology are identical to the healthy pancreatic tissue [1,2].

Fatty replacement of pancreas may either focal or diffuse [1]. MRI and CT might be useful for differentiating these conditions from neoplasms. Complete fatty replacement of the pancreatic tissue is reported in 56-93% of the patients with cystic fibrosis[1].

Pancreatic cysts might also be congenital, and are often incidentally detected. These cysts usually have a thin wall and are variable in size, where in extreme cases it may lead to cystic replacement of the gland. Some of these cysts might be associated with
von Hippel-Lindau disease [1]. The discussion of choledochal cysts is beyond the scope of this poster.
Fig. 1: Images courtesy of MD Koenraad J. Mortelé. Drawings illustrate the normal embryologic development of the pancreas and biliary tree. The ventral pancreatic bud (arrow in a and b) and biliary system arise from the hepatic diverticulum, and the dorsal pancreatic bud (arrowhead in a and b) arises from the dorsal mesogastrium. (c) After clockwise rotation of the ventral bud around the caudal part of the foregut, there is fusion of the dorsal pancreas (located anterior) and ventral pancreas (located posterior). (d) Finally, the ventral and dorsal pancreatic ducts fuse, and the pancreas is predominantly drained through the ventral duct, which joins the common bile duct (CBD) at the level of the major papilla. The dorsal duct empties at the level of the minor papilla.

Fig. 2: Axial T2-weighted MR image shows independent drainage sites of dorsal and ventral ducts, and a dominant dorsal pancreatic duct is already suggested (see Figure 3)

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**Fig. 3:** Coronal MR cholangiopancreatogram shows a dominant dorsal pancreatic duct with larger caliber. The ventral duct is short, curved and very narrow, almost atretic. A small filamentous communication between the two ducts can’t be excluded - Type 3/1 Pancreas Divisum?

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Fig. 4: Coronal reformatted CT image shows a dominant dorsal pancreatic duct, draining to the minor papilla - Pancreas Divisum (see image 5).

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Fig. 5: Coronal reformatted CT image shows a minor ventral pancreatic duct, draining to the major papilla - Pancreas Divisum (see image 4).

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Fig. 6: Axial contrast-enhanced CT scan shows pancreatic tissue encircling the descending portion of the duodenum - Annular pancreas.

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Fig. 7: Axial contrast-enhanced CT scan shows pancreatic tissue encircling the descending portion of the duodenum - Annular pancreas.

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**Fig. 8:** Images courtesy of MD Koenraad J. Mortelé. CT scan obtained in a patient with abdominal pain shows ectopic pancreatic tissue (arrow) within the small bowel mesentery - Ectopic pancreas.

Conclusion

Congenital anomalies of the pancreas and pancreatic duct are not uncommonly encountered at radiologic examinations.

Radiologists should be aware of these variable imaging features to distinguish these anomalies from other pancreatic conditions. MRCP is the imaging modality of choice for the work-up of suspected developmental anomalies of the pancreas and pancreatic duct. It may depict noninvasively the course and drainage pattern of the pancreatic duct and can easily diagnose developmental anomalies of the pancreas. Alternatively, multiplanar reconstructed MDCT images may also help.

Recognition of developmental anomalies of the pancreas and pancreatic duct is important because these anomalies may be a surgically correctable cause of clinical conditions, like recurrent pancreatitis or the cause of gastric outlet obstruction. An awareness of these anomalies may help in surgical planning and preventing inadvertent ductal injury [1,2].
References

