PICTORIAL ESSAY

Various Congenital Abnormalities and Anatomic Variants of the Pancreas: A Pictorial Review

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Numerous and various congenital abnormalities and anatomic variants of the pancreas (CAAVPs) have been reported. Some of them are not so uncommon. Recent advances and accessibility of various multiplanar imaging modalities today offer the increased capabilities of detection and full diagnosis of these CAAVPs. With a precise diagnosis, the symptomatic CAAVPs can not only be more specifically treated but even more their detection and exact description can modify the surgical or interventional strategy to avoid unexpected post-operative complications. This article aimed to review the embryogenesis of the pancreas and describe imaging findings of CAAVPs.

Keywords: Pancreas; Congenital abnormalities; Anatomic variation; Multidetector computed tomography; Magnetic resonance imaging

Introduction
Congenital abnormalities and anatomic variants of the pancreas (CAAVPs) are not uncommon and appear in a variety of ways [1]. Although many CAAVPs are incidentally detected due to the increasing accessibility of diagnostic imaging, some of them may present with symptoms, ranging from abdominal pain to pancreatitis [2]. It is important to recognize symptomatic CAAVPs because they can be corrected by surgery or interventional procedure [1, 3]. In addition, CAAVPs can affect surgical planning [4]. Therefore, radiologists must be familiar with CAAVPs to correctly diagnose them.

Imaging techniques
Computed tomography (CT) is relatively inexpensive and accessible, and preferentially used to evaluate the pancreas. Contrast-enhanced CT for the pancreas is performed 35–45 sec (pancreatic phase) and 60–70 sec (portal venous phase) after the start of IV injection of contrast material and uses thin sections for detailed characterization. The pancreatic parenchyma shows peak enhancement on the pancreatic phase [5]. Magnetic resonance imaging (MRI) reveals a clearer contrast to the soft tissue than CT. In particular, the pancreatic tissue shows a characteristic high signal intensity on the fat suppressed T1-weighted image [6]. In addition, MRI is more valuable for imaging children because of the absence of radiation exposure. Magnetic resonance cholangiopancreatography (MRCP) is useful for evaluation of the pancreatic duct.

Embryonic development of the pancreas
In the fourth week of the embryonic period, ventral and dorsal outpouchings arise at the junction of the foregut and midgut. The dorsal pancreatic bud arises from the dorsal outpouching, whereas the ventral pancreatic bud develops from the ventral one, which is also the primordium of the liver and biliary system (Figure 1A and 1B) [3, 7]. As the stomach rotates, the duodenum rotates to the right and becomes C-shaped. Then, the ventral pancreas moves backwards and lies beneath and behind the dorsal pancreas, eventually merging with the dorsal pancreas at 37 fetal days [2, 7]. The ventral pancreas becomes the uncinate process and the lower part of the pancreatic head, and the remainder of the pancreas is derived from the dorsal pancreas (Figure 1C). After fusion, ducts in the dorsal and ventral pancreas meet to form the major duct (duct of Wirsung) [2, 7]. The major duct opens to the duodenum via the major papilla with the common bile duct. The duct in the upper head portion arising from the dorsal pancreas becomes the duct of Santorini, opening through the minor papilla to the duodenum (Figure 1D) [3].

Congenital Abnormalities and Anatomic Variants of the Pancreas (CAAVPs)

Pancreas divisum
Pancreas divisum is the most common pancreatic congenital anomaly, and an autopsy series reported the prevalence as high as 14% [1]. This anomaly arises from failed fusion of the dorsal and ventral pancreatic ducts, resulting in two pancreatic ducts that are not joined together but separated and incompletely joined by very thin connections (Figure 2A). As a result, the dorsal pancreatic duct opens to the duodenum via the minor papilla. On MRCP,
the common bile duct and the main pancreatic duct are crossed, the so-called ‘crossing duct sign’ (Figure 2B) [8]. In patients with pancreas divisum, exocrine secretory flow may produce through the duct of Santorini, more frequently resulting in recurrent pancreatitis. Some of them may have a Santorinicele (Figure 3) [1, 2].

Annular pancreas

Annular pancreas refers to as an anomaly in which the pancreatic tissue encircles the second portion of the duodenum [1]. The pancreatic tissue around the duodenum continues with the pancreatic head (Figure 4A) [2]. Annular pancreas is the second most common congenital anomaly of the pancreas, occurring in 1/20000 persons of the general population [2, 9]. During embryonic development, the ventral pancreatic bud is composed of a right and left bud, and the left one normally disappeared. Annular pancreas is thought to be caused by adhesion of the right ventral bud to the duodenum or persistence of the left ventral bud [3, 7]. The clinical manifestations of annular pancreas...
pancreas vary from congenital anomaly to malignancy [7, 10]. Annular pancreas is easily detected on both CT and MRI (Figures 4 and 5) [1]. Surgical resection is needed in symptomatic patients [10].

**Dorsal pancreas agenesis**

Dorsal pancreas agenesis is an uncommon anomaly that manifests as the absence of the body and tail of pancreas. This anomaly results from the absence of the dorsal pancreatic bud. Because most of the islet cells are located in the dorsal pancreas, dorsal pancreas agenesis is linked with diabetes mellitus [7]. Partial dorsal pancreas agenesis, also called short pancreas, is occasionally part of heterotaxy syndrome (Figure 6) [7]. It may be difficult to differentiate dorsal pancreas agenesis from fatty replacement of the distal pancreas, but the presence of stomach or intestine in the distal pancreatic bed and absence of the dorsal pancreatic duct favor a diagnosis of dorsal pancreas agenesis (Figures 7 and 8) [11].

**Heterotopic pancreas**

Heterotopic pancreas occurs in 0.6%–13.7% of the general population and is defined as pancreatic tissue separated from the normal pancreas [12]. Although the aberration of the normal developmental process associated with heterotopic pancreas is unknown, the most widely accepted hypothesis
is misplacement of pancreatic tissue [13]. The most common location of heterotopic pancreas is the proximal gastrointestinal (GI) tract. The stomach is concerned in 26–38%, the duodenum in 28–36%, and the jejunum in 16% [7]. On CT images, heterotopic pancreas usually measures less than 2 cm, and it can be misdiagnosed as a small submucosal tumor of the GI tract. Kim et al. [14] reported that CT findings useful for differentiation of heterotopic pancreas from gastrointestinal stromal tumor or leiomyoma in the stomach or duodenum are an ill-defined border, prominent enhancement of overlying mucosa, endoluminal growth pattern, prepyloric antrum or duodenal location, and flat shape (Figure 9). Malignant neoplasm or inflammation can develop from heterotopic pancreatic tissue (Figures 10 and 11) [7, 12, 15].

**Circumportal pancreas**

Pancreatic tissue can encase the portal vein or superior mesenteric vein (instead of the duodenum as in annular pancreas), and this anomaly is termed circumportal pancreas [16]. It is not rare and has a prevalence of 1.1% to 2.5% (Figure 12). Although the exact developmental mechanism of circumportal pancreas has not been elucidated, it is thought to result from abnormal fusion of the dorsal and ventral pancreatic buds [17]. Patients with this anomaly are usually asymptomatic, but it is clinically important to recognize circumportal pancreas prior to pancreatic surgery because its presence can change surgical planning or cause unexpected complications such as fistula formation [4, 17]. Circumportal pancreas can be classified into the
**Figure 7:** A 22-year-old woman with dorsal pancreas agenesis. (A) Axial portal venous phase CT image shows absence of dorsal pancreas. Note stomach in the distal pancreatic bed. (B) Endoscopic retrograde cholangiopancreatography image shows absence of dorsal pancreatic duct.

**Figure 8:** A 49-year-old woman with fatty replacement of distal pancreas. Axial portal venous phase CT image shows almost totally fatty change of distal pancreas (arrow). Note the pancreatic duct (open arrow) within the fat density pancreas tissue.

**Figure 9:** A 79-year-old woman with heterotopic pancreas in stomach. Axial pancreatic phase CT image shows an enhancing mass (arrow) in gastric antrum. The mass reveals ill-defined border and endophytic growth.

**Figure 10:** A 68-year-old woman with adenocarcinoma arising from heterotopic pancreas. (A, B) Axial and coronal reformatted CT images show irregular mass (arrow) with heterogeneous enhancement in gastric pylorus. (C) Positron emission tomography-CT image shows intense uptake in the mass (arrow).
following four subtypes according to the course of the main pancreatic duct and the relationship between the splenic vein and the fused pancreas: 1) anteportal suprasplenic, 2) retroportal suprasplenic, 3) anteportal infrasplenic, and 4) retroportal infrasplenic [4].

Intrapancreatic accessory spleen
An accessory spleen is an anatomical variation frequently observed in daily practice [18]. They result from failed fusion of the splenic precursors in the dorsal mesogastrium during the fifth week of embryogenesis [19]. Although most accessory spleens are located around the splenic hilum, approximately 16% are found in or around the pancreas [20]. An accessory spleen shows a similar signal intensity and enhancement compared with the mother spleen on MRI (Figure 13A and 13B) [19]. Radio-uptake on Tc-99m heat-damaged red blood cell scintigraphy can diagnose an accessory spleen without pathologic confirmation (Figure 13C) [19]. Rarely, an epithelial or epidermoid cyst can develop from an intrapancreatic accessory spleen (Figure 14) [19, 21].

Conclusion
CAAVPs are not uncommon. Many affected patients remain asymptomatic. However, some patients could present with symptoms such as pancreatitis. In addition,
CAAVPs can lead to unnecessary surgeries or unexpected complications. Familiarity with the imaging findings of various CAAVPs is paramount for managing patients in daily practice.

**Abbreviations**

CAAVPs = Congenital abnormalities and anatomic variants of the pancreas

CT = Computed tomography
GI = Gastrointestinal
MRI = Magnetic resonance imaging
MRCP = Magnetic resonance cholangiopancreatography

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Competing Interests
The authors have no competing interests to declare.

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