

# Anomalies, Anatomic Variants, and Sources of Diagnostic Pitfalls in Pancreatic Imaging<sup>1</sup>

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In this review, a brief discussion of the important events of pancreatic embryology is followed by presentation of congenital anomalies and normal variants. For each variant, the appearance at different radiologic modalities including computed tomography, magnetic resonance (MR) imaging, endoscopic retrograde cholangiopancreatography, MR cholangiopancreatography, and fluoroscopy will be demonstrated.

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**F**or the resident physician, differentiation of normal variants of the pancreas from disease can be challenging. The goal of this review is to familiarize radiology residents and other practitioners with the basic knowledge of pancreatic imaging and to demonstrate normal conditions that, if not correctly diagnosed, might lead to unnecessary follow-up examinations or other diagnostic procedures.

### Pancreatic Development

By the 4th week of embryologic development, the pancreatic duct develops from separate ventral and dorsal buds originating from the endodermal lining of the duodenum (1). The gallbladder, extrahepatic bile ducts, central intrahepatic bile ducts, and ventral pancreas with its ductal network are derived from the ventral bud or outpouching. The dorsal bud is the precursor to dorsal pancreas and its ductal system (Fig 1a) (2). The ventral pancreas rotates clockwise posterior to the duodenum and comes into contact

with the dorsal pancreas in the 7th gestational week to develop into the future pancreatic neck. The dorsal and ventral pancreatic buds grow into a pair of branching, arborizing ductal systems, each with its own central or main duct. The two anlagen fuse with each other and, together with the duodenum, fuse with the abdominal wall. After fusion, a new duct connects the distal portion of the dorsal pancreatic duct with the shorter duct of the ventral pancreas to form the main pancreatic duct, also known as the duct of Wirsung, which empties into the major papilla. The remnant of the dorsal duct forms the duct of Santorini, which drains into minor papilla (Fig 1b) (3). The process of pancreatic fusion is complicated, and a wide spectrum of anatomic variants may occur during the course of pancreatic development.

### Congenital Anomalies

Anatomic anomalies of the pancreas are classified as either a fusion anomaly (pancreas divisum), migration anomaly (annular pancreas, ectopic pancreas), or duplication anomaly (number or form variation) (4). Pancreatic fusion or migration anomalies may result in anatomic variants that predispose to specific pancreatic or peripancreatic diseases.

### Pancreas Divisum

Pancreas divisum results from a failure of ventral and dorsal bud fusion. The ventral (Wirsung) duct drains only the ventral pancreatic anlage, whereas the majority of the gland empties into the minor papilla through the dorsal (Santorini) duct (3). It is the most common congenital anomaly, occurring in 4%–14% of the population, as evidenced by autopsy results; 3%–8% are seen during endoscopic retrograde cholangiopancreatography (ERCP) and approximately 9% are depicted at magnetic resonance (MR) cholangiopancreatography (MRCP) (5–7). In most cases, there is no communication between the ventral and dorsal duct; however, in some individuals, there is a filamentous communication remaining and in others the ventral duct is totally absent (8). Although still debatable, pancreas divisum is associated with acute

and recurrent pancreatitis. The reported frequency of complete pancreas divisum in patients with acute pancreatitis ranges from 25% to 38% (9–11).

In the past, ERCP was the modality of choice for diagnosing pancreas divisum; however, MRCP allows noninvasive multiplanar visualization of the biliary tree and pancreatic duct without injection of contrast material and avoids risks of ERCP-induced pancreatitis or those associated with sedation required for the procedure. Advances in therapeutic endoscopic procedures, such as minor papillotomy or insertion of stents into the minor papilla for treatment of patients with pancreas divisum, make recognition of this variant important (12). The main anatomic feature of pancreas divisum, continuity of the dorsal pancreatic duct with the duct of Santorini draining into the minor papilla, is readily identified at both ERCP and MRCP. The ventral duct drains into the major papilla without communication with the dorsal pancreatic duct (13). On ERCP images, the pancreas divisum is optimally identified with the injection of contrast material into both the minor and major papillae (Fig 2). On MRCP images, the normal ventral system may be so small in caliber that the ducts are not visible (Fig 3); however, it is the predominant pancreatic duct drainage into the minor papilla at a level superior to the level of the bile duct emptying into the major papilla that indicates the presence of divisum. The administration of secretin during MRCP increases the visibility of the main pancreatic duct and its side branches and also increases the sensitivity and specificity for detection of anatomic variants such as pancreas divisum (14–16). With the advent of multidetector computer tomographic (CT) scanners and high-spatial-resolution thin-section imaging, pancreas divisum may be routinely seen with use of CT as well (6).

### Essentials

- Pancreatic fusion or migration anomalies may result in anatomic variants that predispose patients to specific pancreatic or peripancreatic diseases such as recurrent acute pancreatitis, cystic dystrophy of the duodenum, duodenal obstruction, cholangiocarcinoma, and gall bladder carcinoma.
- Pancreatic fusion or migration anomalies and normal variants such as fat deposition or accessory splenic tissue may give rise to imaging appearances that mimic more serious pancreatic conditions.
- Knowledge of pancreatic embryology and of normal anatomic variants is essential to identify these entities and to help differentiate them from pathologic conditions, thus preventing potential unnecessary imaging investigation or more invasive procedures such as biopsy or surgery.

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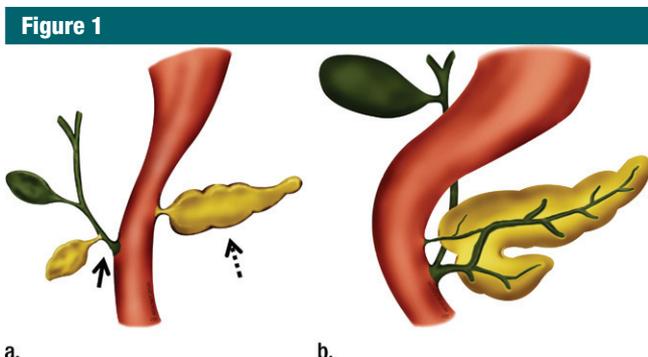
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#### Abbreviations:

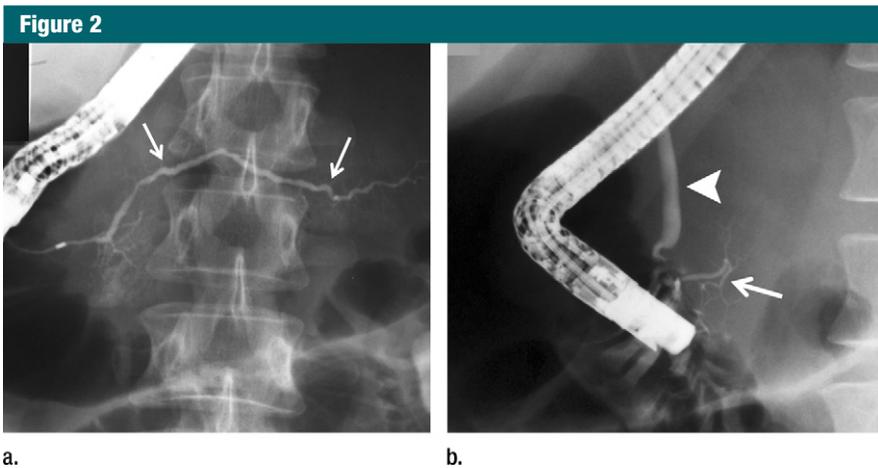
ERCP = endoscopic retrograde cholangiopancreatography

MRCP = MR cholangiopancreatography

Conflicts of interest are listed at the end of this article.



**Figure 1:** Schematic of pancreatic development. **(a)** Early development of the ventral (solid arrow) and dorsal buds (dashed arrow) prior to rotation. **(b)** Final configuration of the pancreas after periduodenal rotation of the ventral bud and approximation of the ventral and dorsal pancreas. The dorsal pancreatic duct merges with the ventral (Wirsung) duct and drains into major papilla. The rudimentary distal dorsal duct (Santorini) drains into minor papilla.



**Figure 2:** ERCP images of pancreas divisum in 26-year-old woman with abdominal pain. **(a)** The main pancreatic duct (arrows) drains through the minor papilla, consistent with pancreas divisum. **(b)** The short, terminally arborizing duct of Wirsung (arrow) is depicted by means of contrast material injection of the major papilla, with the common bile duct (arrowhead) also filling.

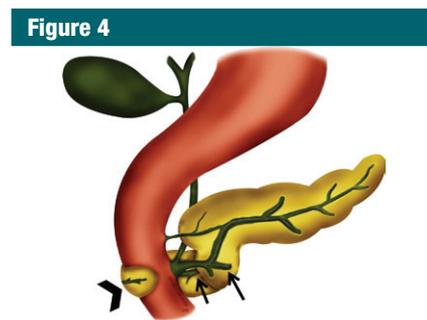
**Annular Pancreas**

Annular pancreas is a rare congenital anomaly in which incomplete rotation of the ventral anlage leads to a segment of the pancreas encircling the second part of duodenum (Fig 4). There are two types of annular pancreas: extramural and intramural. In the extramural type, the ventral pancreatic duct encircles the duodenum to join the main pancreatic duct. In the intramural type, the pancreatic tissue is intermingled with muscle fibers in the duodenal wall, and small ducts drain directly into the duodenum. In patients with extramural annular pan-

creas, the presenting symptoms are those of high gastrointestinal obstruction. For patients with the intramural type, symptoms are those of duodenal ulceration. Intervention for extramural obstructing annular pancreas is surgical and usually a bypass procedure; for intramural annular pancreas with duodenal ulceration, subtotal gastrectomy with or without vagotomy is the procedure of choice (17). Pediatric patients with annular pancreas may have diagnostic findings on conventional abdominal radiographs, the classic double-bubble sign. The larger proximal bubble is caused by gastric distention and



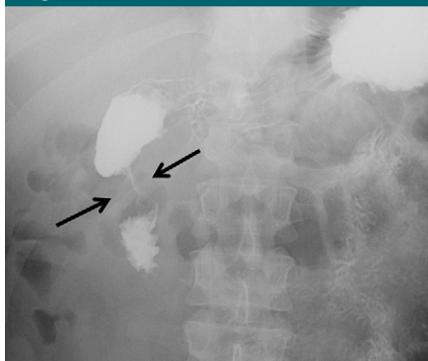
**Figure 3:** MRCP image of pancreas divisum in 46-year-old man. Angled coronal fat-suppressed half-Fourier acquisition single-shot turbo spin-echo (repetition time msec/echo time msec, 9450/740; 40-mm section thickness; 30-cm field of view) MR image shows the main pancreatic duct (dorsal Santorini duct, straight solid arrow) draining separately into the minor papilla (dashed arrow). The common bile duct (arrowheads) joins the smaller ventral pancreatic duct (curved arrow) at a more inferior level and drains into the duodenum through the major papilla.



**Figure 4:** Schematic of annular pancreas. Ventral pancreas (arrowhead) encircles the second portion of the duodenum. The duct of the ventral pancreas (arrows) makes a turn around the duodenum and merges with the main dorsal duct in a normal fashion to form the Wirsung duct and drain to the major papilla.

the smaller distal bubble is caused by a dilated duodenal bulb (18,19). Barium examination reveals focal stenosis of the periamпуляр region, with an extrinsic eccentric defect of the second portion of the duodenum (Fig 5). Findings on CT scans include enlargement of the pancreatic head, which encircles the second portion of the duodenum; this may be

Figure 5



**Figure 5:** Upper gastrointestinal radiograph of annular pancreas in a young female patient with vague intermittent abdominal discomfort and fullness shows slit-like smooth narrowing of the second portion of duodenum (arrows). No gastric distention or mechanical obstruction is present. Contrast material flows freely into an otherwise normal duodenum and proximal jejunum.

Figure 6



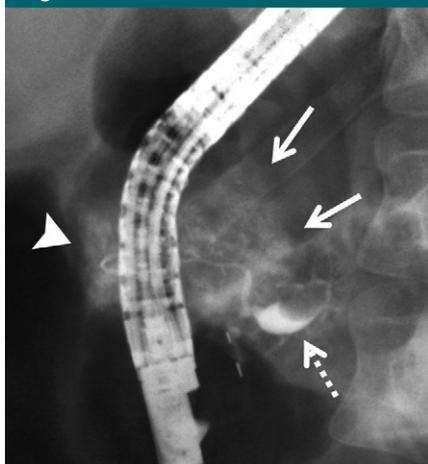
**Figure 6:** Unenhanced axial CT scan of annular pancreas in 43-year-old man with history of resected glucagonoma in the pancreatic tail. The pancreatic tissue encircles the positive oral contrast material-filled second portion of the duodenum (arrows), making the anomaly easy to recognize even without intravenous contrast material.

Figure 7



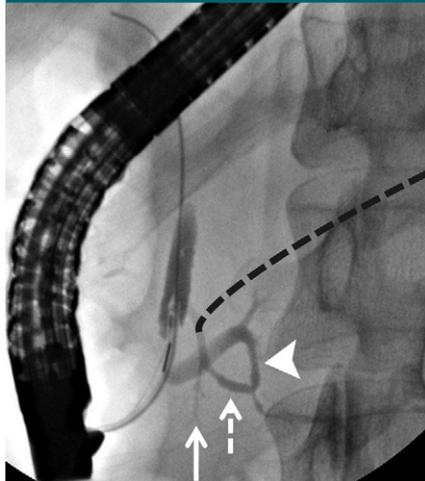
**Figure 7:** Intravenous contrast-enhanced axial CT scan of annular pancreas in 51-year-old man reveals normal enhancing pancreatic tissue (arrows) surrounding the collapsed duodenum. The patient received water as a negative oral contrast agent. Wirsung duct of the ventral pancreas (arrowhead) is draining into duodenum.

Figure 8



**Figure 8:** ERCP spot image of annular pancreas in 43-year-old man shows mild diffuse acinarization of the pancreas (solid arrows). The pancreatic tissue and pancreatic duct (arrowhead) are to the right of the second portion of the duodenum, consistent with annular pancreas. Focal dilatation of the main pancreatic duct (dashed arrow) with pooling of contrast material in the dependent portion is also present.

Figure 9



**Figure 9:** ERCP image of ansa pancreatica in 37-year-old man. Dashed line represents dorsal main pancreatic duct (technically underfilled). There is obliteration of the proximal dorsal pancreatic duct (solid arrow). The proximal dorsal duct connects with an inferior branch of the ventral duct (arrowhead) through S-shaped collateral (dashed arrow).

identified either with or without oral contrast material (Figs 6, 7). MR imaging demonstrates pancreatic tissue and occasionally the small annular duct encircling the descending duodenum. ERCP demonstrates normal main pancreatic duct

configuration in the body and tail along with the aberrantly oriented duct of the pancreatic head; this duct passes posteriorly around the duodenum (Fig 8) and enters the main pancreatic or common bile duct near the ampulla.

### Variation in Pancreatic Duct Course

The pancreatic ductal system may show variations based on the spatial relationship of the ducts of Wirsung and Santorini and their relation to the common bile duct and to the duodenum. The known ansa variant of the pancreatic duct (ansa pancreatica) is seen when there is obliteration of the dorsal pancreatic duct at the junction with ventral duct, and instead, the proximal portion of the dorsal duct connects with an inferior side branch of the ventral duct through an S-shaped collateral duct (Fig 9) (20).

Anomalous union of the pancreatic and bile ducts is seen when the junction of the pancreatic and common bile duct is outside of the duodenal wall, resulting in a combined channel leading to the major papilla that measures more than 1.5 cm in length. Therefore, the contraction of the sphincter of Oddi does not affect the pancreaticobiliary junction and causes pancreaticobiliary reflux, which may result in various clinical presentations such as pancreatitis (21). The anomalous union may be caused by abnormal insertion of the pancreatic duct into the bile duct (Fig 10) or of the bile duct into the pancreatic duct. These variants are important

Figure 10



**Figure 10:** ERCP spot image in 54-year-old woman with anomalous pancreaticobiliary duct union shows abnormal proximal insertion of the pancreatic duct to the normal-appearing common bile duct, resulting in a long combined pancreatic-common bile duct channel more than 1.5 cm in length (arrow).

to recognize as there is an increased association with choledochal cysts (22) and gallbladder carcinoma (23).

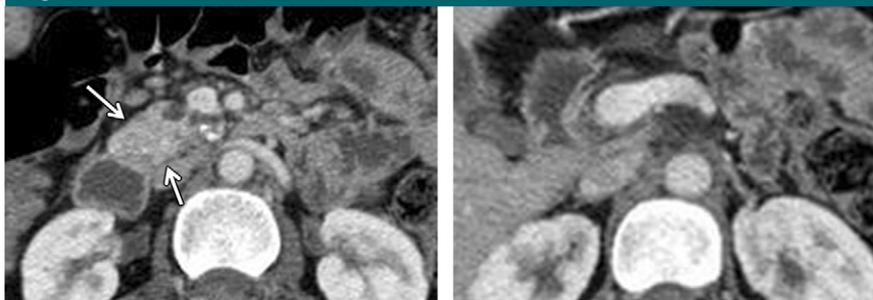
### Agensis and Hypoplasia of the Pancreas

Agensis of the pancreas is rare and generally incompatible with life. Hypoplasia results from absence of the ventral or dorsal anlagen. Partial agensis of the dorsal pancreas is more common than the agensis of the ventral portion, but complete agensis of the dorsal pancreas is extremely rare (3). At imaging, dorsal pancreatic hypoplasia manifests as a short, rounded pancreatic head adjacent to the duodenum, with absence of the pancreatic neck, body, and tail (Fig 11). Pancreas hypoplasia may be associated with an increased risk of pancreatitis and polysplenia syndrome (24).

### Ectopic Pancreas

Ectopic pancreatic tissue is usually seen in the submucosa of the gastric antrum (30%), the proximal portion of the duodenum (30%), the remaining duodenum (20%), or other regions of the small bowel (20%) (25). The most widely accepted theory of the embryologic origin of this lesion is its development from residual cells of the primitive ventral or dorsal

Figure 11



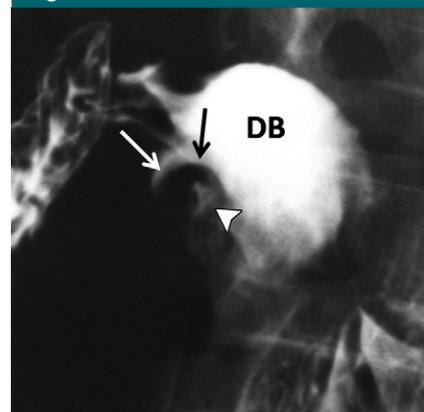
a.

b.

**Figure 11:** Contrast-enhanced axial multidetector CT scans of congenitally short pancreas (hypoplasia) in 28-year-old man show (a) the head and uncinete process of the pancreas (arrows) with (b) absence of the body and tail.

bud within the bowel lumen (26). The lesions are composed of normal pancreatic tissue, often including islet cells, and usually have a small pancreatic duct. Ectopic pancreatic tissue may be identified at barium upper gastrointestinal examination, where features of a small collection of barium located within a central niche or umbilication on a small rounded mass are diagnostic (Fig 12). The umbilication represents the orifice of the rudimentary duct through which the ectopic pancreas empties (27). If this feature is absent, the lesion cannot be reliably differentiated radiographically from other submucosal tumors such as Brunner gland adenoma, leiomyoma, or lymphoma. Ectopic pancreatic tissue is functional and subject to the same inflammatory and neoplastic disorders that afflict the normal pancreas; however, the majority of cases are asymptomatic and found incidentally (28,29). Cystic dystrophy, a serious but uncommon complication, represents dilatation of the ectopic pancreatic ducts within the heterotopic pancreatic tissue and occurs most often in the second part of the duodenum (30). It is thought to result from obstruction of the small ducts leading to repeated pancreatitis. Chronic alcohol consumption has been reported to trigger cystic dystrophy in ectopic pancreatic tissue (31). Imaging features include thickened duodenal wall containing multiple cysts, with moderate to strong enhancement. Adjacent inflammatory changes with or without enlarged lymph nodes can be seen in half of the patients (32).

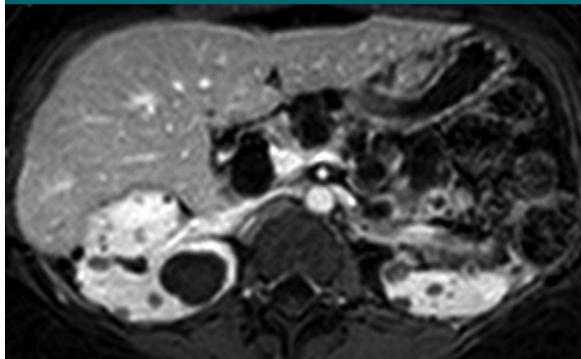
Figure 12



**Figure 12:** Spot radiograph from upper gastrointestinal examination in 33-year-old woman reveals a focal, smooth, round filling defect (arrows) on the inferior wall of duodenal bulb (DB), with tiny umbilication (arrowhead) characteristic of ectopic pancreas. At endoscopy, the lesion was raised and epithelialized, looking like a thickened papilla.

Cystic dystrophy is one of several benign conditions that affect the duodenal wall in the region of the minor pancreatic papilla. Others include pancreatic hamartoma, paraduodenal wall cyst, myoadenomatosis, and groove pancreatitis, which has been collectively termed paraduodenal pancreatitis (33,34). In general, these conditions produce a thickened duodenal wall that may contain dilated ducts and pseudocystic changes, Brunner gland hyperplasia, or dense myxoid stromal proliferation with intervening rounded lobules of pancreatic tissue (35).

Figure 13



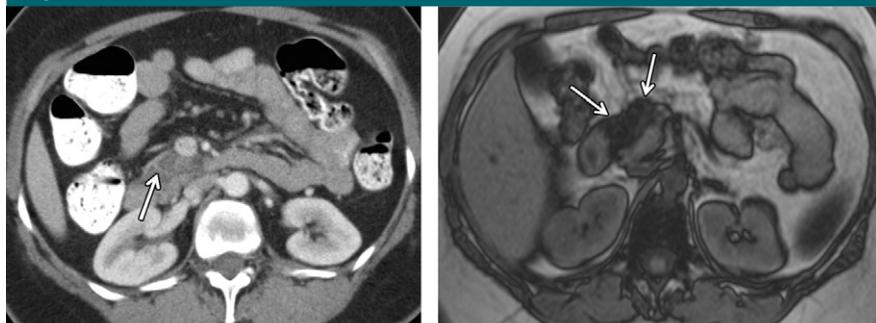
**Figure 13:** Axial fat-saturated T1-weighted MR image (4.0552/1.968, 5-mm section thickness, 31.5-cm field of view) in 29-year-old woman with Von Hippel-Lindau disease shows multiple well-defined hypointense cysts scattered throughout the pancreas. Multiple renal cysts are also present; the patient had undergone partial nephrectomy and radiofrequency ablation for renal carcinoma in the past and has additional neoplasm at other levels (not shown).

Figure 14



**Figure 14:** Diffuse fatty infiltration of pancreas in 84-year-old man receiving steroids for chronic obstructive pulmonary disease. Axial portal venous phase multidetector CT scan through the body region of the pancreas reveals low-attenuating fat replacing the body, neck, and tail, with strands of acinar tissue and ductal structures still evident.

Figure 15



a.

b.

**Figure 15:** Focal fatty infiltration in 43-year-old woman. **(a)** Axial contrast-enhanced CT scan shows ill-defined geographic hypodense region in the pancreatic head, consistent with focal fatty infiltration (type Ia). The small vessel (arrow) coursing through the “mass” is not displaced, an unusual finding in a true neoplastic lesion. **(b)** Axial out-of-phase T1-weighted MR image (200/2.303, 6-mm section thickness, 32-cm field of view) shows low signal intensity in the corresponding location of the head and uncinete process (arrows), consistent with fatty infiltration.

### Pancreatic Cysts

A congenital true pancreatic cyst is a very rare entity, mostly seen in children younger than 2 years of age (36). These cysts develop as a result of sequestration of primitive pancreatic ducts and are lined by cuboidal epithelium (37). Congenital pancreatic cysts are generally asymptomatic, although abdominal distention, vomiting, jaundice, or pancreatitis can be observed (38). At imaging, this condition manifests as a uniform thin-walled cyst usually in the region of the pancreatic body and tail. Congenital true pancreatic cysts may be idiopathic or may be observed in association with other systemic diseases such as Von Hippel-Lindau disease (Fig 13), Beckwith-

Wiedeman syndrome, or polycystic disease of the pancreas and kidneys (39,40).

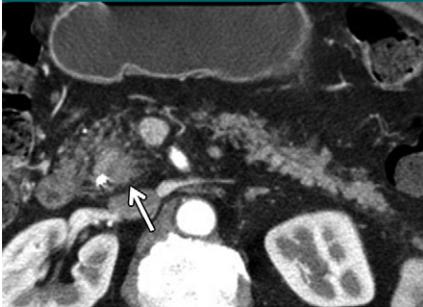
### Normal Variants and Pitfalls

#### Fatty Infiltration

Replacement of pancreas by adipose tissue can be focal or diffuse and in some instances can mimic a mass or a neoplasm. MR imaging, and to a lesser extent CT, are used to differentiate patterns of fatty infiltration from neoplasm. Complete replacement of the pancreas by fat is seen most commonly in patients with cystic fibrosis or occasionally in rare conditions such as Schwachman-Diamond or Johansen-Blizzard syndromes (41,42).

Lesser degrees of diffuse fatty infiltration are more common in patients with diabetes or obesity, as well as in the elderly (Fig 14). When the condition is severe, the pancreas will have the same signal intensity and density as the mesenteric fat and thus may not be identifiable. Differentiation between lipomatosis and pancreatic agenesis is important and is made on the basis of whether the ductal system is present (lipomatosis) or absent (agenesis) (1). Matsumoto et al (43) described four different types of pancreatic fatty infiltration: type Ia, focal infiltration of the pancreatic head with sparing of the uncinete process, peribiliary region, and body; type Ib, infiltration of head, neck, and body with sparing of the peribiliary region; type IIa, infiltration of uncinete process and head with sparing of the peribiliary region and body (Fig 15); and type IIb, complete infiltration of the pancreas, except for the peribiliary region. Interestingly, all types of pancreatic fatty infiltration share sparing of the peribiliary region. Other causes of focal low attenuation in the pancreatic parenchyma, such as focal acute pancreatitis, should be excluded on the basis of ancillary findings. When diffuse fatty replacement of the pancreas is present, masses can be readily appreciated (Fig 16), since the relative density of the mass compared with the low-attenuating

**Figure 16**



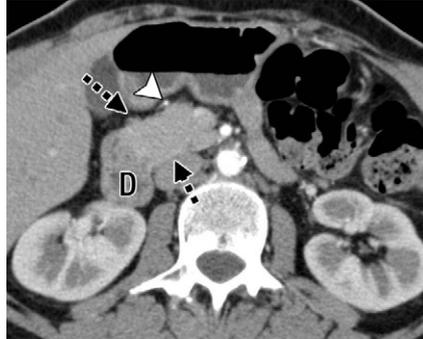
**Figure 16:** Pancreatic adenocarcinoma in 64-year-old man with fatty replaced gland. Axial contrast-enhanced multidetector CT scan shows slightly heterogeneous hypovascular adenocarcinoma (arrow) in the region of the head. The fatty replacement provides inherent tissue contrast and thus increases conspicuity of this lesion.

fat alters perception and potentially affects mass characterization.

**Pseudomass**

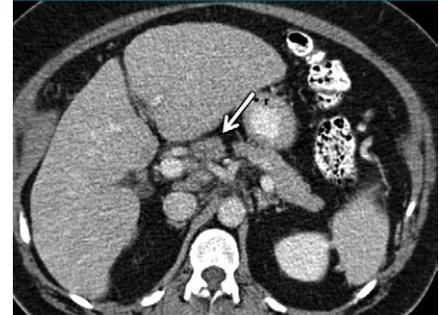
Normal alteration of the pancreatic contour is referred to as pseudomass and can mimic pancreatic neoplasm. Lobulation of the pancreatic head and neck is defined as extension of the parenchyma beyond 1 cm relative to the anterior superior pancreaticoduodenal artery, whether anteriorly (type I), posteriorly (type II) (Fig 17), or horizontally (type III) (44). Enlargement of lymph nodes adjacent to the pancreas can mimic a pancreatic mass (Fig 18) and may result in potential diagnostic challenges. Paraaortic and periportal lymphadenopathy should be differentiated from pancreatic neoplasm to avoid unnecessary additional diagnostic procedures. Occasionally, accessory splenic tissue may be located in the body-tail region of the pancreas, mimicking an infiltrative process such as autoimmune pancreatitis or neoplasm, especially a pancreatic endocrine tumor. This entity can generally be recognized at multiphase CT, since the tissue follows the enhancement pattern of the spleen (45). When CT is not definitive, MR imaging is very useful because the signal in the accessory splenic tissue is more readily defined (Fig 19), compared with the surrounding pancreatic parenchyma (46). Heat-damaged red blood cells labeled with

**Figure 17**



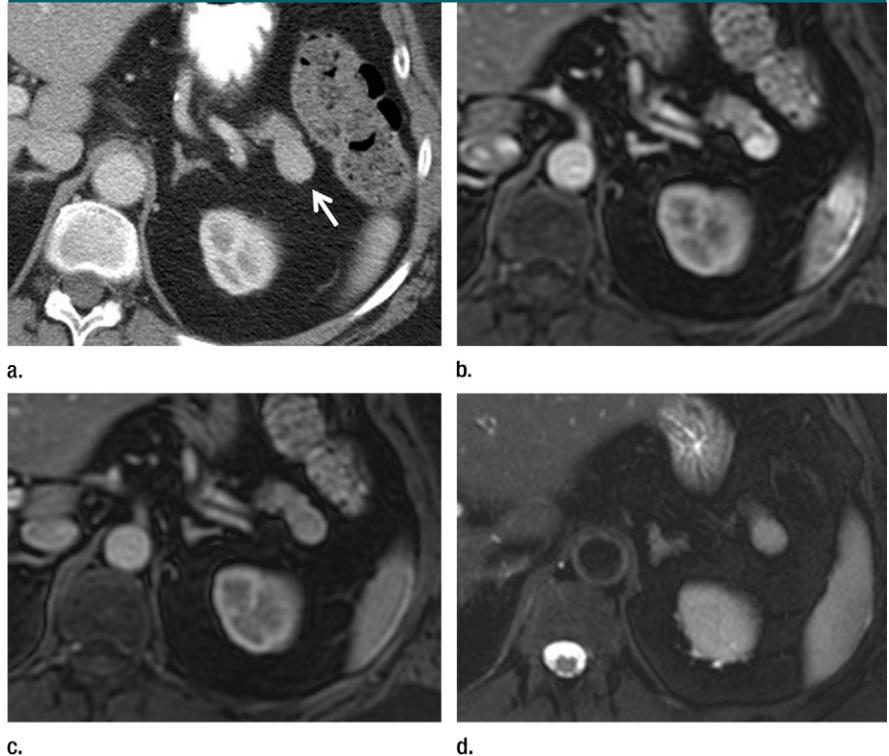
**Figure 17:** Prominent pancreatic head in 28-year-old woman. Note the lateral extension of the pancreatic head (arrows) in relation to anterior superior pancreaticoduodenal artery (arrowhead), representing type II (posterior) lobulated pancreas head variant. *D* = duodenum.

**Figure 18**



**Figure 18:** Peripancreatic and periportal lymph nodes mimicking pancreatic mass in 52-year-old woman. Portal venous phase multidetector CT scan obtained to evaluate generalized abdominal pain shows cirrhotic liver contour and prominent node anterior to the common hepatic artery (arrow), mimicking a partially exophytic pancreatic mass. Note the slight difference in attenuation of pancreatic parenchyma compared with the nodes, which helps in differentiation.

**Figure 19**



**Figure 19:** Intrapaneatic accessory spleen in 54-year-old man with an incidental pancreatic mass. (a) Contrast-enhanced axial multidetector CT scan shows a dense pancreatic body mass (arrow). (b) Contrast-enhanced T1-weighted fat-suppressed axial MR image (4.2011/2.069, 5-mm section thickness, 36-cm field of view) shows pancreatic body lesion that enhances slightly more than normal pancreatic parenchyma on (c) early postgadolinium-enhanced pancreatic phase MR image. (d) T2-weighted MR image (1800/70, 6-mm section thickness, 34-cm field of view) shows features also consistent with splenic tissue rather than a mass.

technetium 99m ( $^{99m}\text{Tc}$ ) or  $^{99m}\text{Tc}$ -labeled sulfur colloid scans can also be used to diagnose accessory splenic tissue, showing increased uptake in the region of interest (47). Focal autoimmune pancreatitis can also manifest as a pseudomass and focal parenchymal enlargement. One imaging feature that can help distinguish this entity from pancreatic adenocarcinoma is the pattern of enhancement. At the portal venous phase, focal autoimmune pancreatitis may appear hyperdense compared with the usual hypodense pancreatic adenocarcinoma (48).

In conclusion, congenital anomalies and normal variants of the pancreas can present a diagnostic challenge when encountered. Knowledge of pancreatic embryology and of normal anatomic variants is essential to identify these entities and help differentiate them from pathologic conditions, thus preventing potential unnecessary imaging investigation or more invasive procedures such as biopsy or surgery.

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