

# Annular Pancreas in an Adult Presenting with Acute Pancreatitis

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## ABSTRACT

Annular pancreas is a rare congenital condition in which tissue from the head of the pancreas partially or completely surrounds the second part of the duodenum. This condition has a variable clinical presentation in adults and children. Radiologists should be aware of the imaging findings in adults as an annular pancreas can be overlooked or misdiagnosed in this patient population. In this case report we describe an adult patient with pancreatitis associated with the computed tomography findings of a complete annular pancreas and discuss additional imaging techniques and findings for differential diagnoses of this condition.

## CASE REPORT

### CASE REPORT

A 58-year-old male patient with a past medical history of hypertension and hyperlipidemia and notably no history of alcohol abuse presented to the emergency department (ED) with epigastric pain and blood per rectum. The patient came to the ED three years prior with similar symptoms. At that time, he was diagnosed with gastroenteritis. His past surgical history was only significant for a prior appendectomy. He reported taking some leftover ciprofloxacin, which he was prescribed after the previous episode of abdominal pain. Due to the presenting symptoms, pancreatitis was considered in the differential diagnosis; however, initial laboratory studies showed lipase levels within normal limits (209 U/l, normal 73-393 U/l) and no other clinically significant laboratory values.

A contrast-enhanced computed tomography (CT) scan of the abdomen demonstrated the findings of congenital annular pancreas with pancreatic tissue surrounding the duodenum (Fig. 1). Inflammatory fat stranding was seen around the pancreatic head (Fig. 1). CT images also revealed edema involving the distal duodenum and hepatic flexure of the colon

(Fig. 1, Fig. 2). However, there was narrowing of the duodenum secondary to the aberrant pancreatic tissue (Fig. 2). Three-dimensional (3-D) reconstructed images were created and the surrounding soft tissue subtracted. These images demonstrate a complete ring of pancreatic tissue through which the second portion of the duodenum traverses (Fig. 3). There was no evidence of cholelithiasis, bile or main pancreatic duct obstruction, or pancreatic parenchymal lesion (Fig. 4). Gallbladder function was normal based on nuclear medicine hydroxy-iminodiacetic acid (HIDA) imaging with gallbladder ejection fraction of nearly 100%.

The patient was diagnosed with acute pancreatitis secondary to congenital annular pancreas, as he had no cholelithiasis, no history of alcohol abuse, and no other risk factors for pancreatitis such as viral infections, hypertriglyceridemia or neoplasm. The patient's symptoms improved after intravenous fluids and pain medicine and he was discharged home from the ED. No follow-up plans for further pancreatic imaging or work up of congenital annular pancreas were made. The patient has not presented to our hospital system for 2 years since the last episode.

## DISCUSSION

### Embryology:

Pancreatic development is an intricate process. Errors occurring during the fusion of the dorsal and ventral pancreatic buds can result in fusion, migration or duplication anomalies [1]. Annular pancreas, like ectopic pancreas, is defined as a migration anomaly. During the fourth to fifth weeks of gestation, the pancreas begins to develop. Two ventral buds and one dorsal bud appear as outgrowths from the primitive foregut. The two (right and left) ventral buds will fuse and by the seventh gestational week the duodenum will begin to expand. This expansion results in rotation of the ventral bud behind the duodenum, from right to left (clockwise if viewed from the top), where the ventral and dorsal buds fuse.

Fusion of the ventral and dorsal ducts results in formation of the main pancreatic duct [1,2]. The ventral bud gives rise to the uncinate process and head of the pancreas, while the dorsal bud develops into the body and tail of the pancreas. It is failure of this complete rotation of the ventral pancreatic bud around the duodenum during gestation that results in a ring of pancreatic tissue around the second part of the duodenum.

Several theories have been proposed to explain the formation of annular pancreas. These include: adhesion of the right ventral bud to the duodenal wall (Lecco's theory), persistence of the left ventral bud (Baldwin's theory), and a third theory that proposes adherence of a small portion of the left ventral bud to the duodenal wall with concomitant pancreatic tissue enveloping the duodenum during rotation [1,3]. This third theory allows for differing degrees of duodenal envelopment depending on location of the left ventral bud adherence [1,3].

Johnston classified annular pancreas into extramural and intramural based on the presence or absence of pancreatic tissue in the adjacent wall of the duodenum [4]. These two types may only be distinguished with invasive procedures such as endoscopic ultrasound or surgery. With the extramural type, the ventral pancreatic duct encircles the duodenum and communicates with the main pancreatic duct. In the intramural type, pancreatic tissue and duodenal muscular fibers are interwoven with small ducts draining directly into the duodenum [4,5].

### Clinical presentation:

Annular pancreas is a rare congenital anomaly where the clinical presentation differs in children and adults. In the pediatric population, annular pancreas is frequently found associated with other congenital anomalies such as duodenal stenosis, duodenal atresia and Down syndrome [6]. In these patients the presentation most commonly consists of symptoms related to duodenal obstruction, such as vomiting, bloating, and feeding intolerance [6–8]. When associated with atresia, most cases are diagnosed in the postpartum period with bilious vomiting and intolerance to feeding. Alternatively, cases of less severe duodenal stenosis where annular pancreas might also be present, may present months later with failure to thrive and frequent vomiting.

Adults in the third through sixth decade of life, when symptomatic, tend to present with abdominal pain, postprandial fullness, vomiting, acute or chronic pancreatitis, upper gastrointestinal bleeding from peptic ulcer disease, and less commonly biliary obstruction [2,8]. Adults often present with intermittent or mild symptoms, which may be secondary to duodenal stenosis from the annular pancreas constricting the duodenum. The adult prevalence of annular pancreas is believed to be between 15 to 400 cases in 100,000 adults [2]. However, the true prevalence is unknown as this condition seldom presents itself with symptoms in the adult population [9,10]. There is no clear gender predilection seen upon review of the literature [2,8,10,11].

### Imaging findings:

CT or magnetic resonance (MR) examinations reveal enlargement of the pancreatic head with encasement of the second portion of the duodenum. While annular pancreas is often thought of as pancreatic tissue completely encircling the duodenum, a complete ring of pancreatic tissue around the duodenum is not required for the diagnosis of annular pancreas. Sandrasegaran et al. describe different configurations of pancreatic tissue around the second part of the duodenum [2]. If imaging demonstrates pancreatic tissue extending posterior to the second part of the duodenum, and appropriate clinical symptoms are present such as chronic biliary or gastric outlet obstruction, then suspicion for annular pancreas is warranted [2]. A crocodile jaw configuration, where the pancreatic head is found anterior and posterior to the second part of the duodenum, or the presence of pancreatic tissue posterolateral to the duodenum, is thought to be highly suggestive of annular pancreas [2]. Zyromski et al. showed a strong association between annular pancreas and pancreas divisum in adults [8]. This suggests that patients undergoing magnetic resonance cholangiopancreatography (MRCP) for delineation of biliary ductal anatomy may benefit from complementary image analysis of the pancreatic parenchyma for other anomalies.

In the pediatric population, annular pancreas may present with the classic double bubble sign on abdominal radiographs or sonography [12]. The double bubble sign is a non-specific finding for duodenal obstruction, in which the larger, proximal bubble represents gastric distention and the smaller distal bubble represents distention of the proximal duodenum [5]. In neonates, this obstruction may be due to congenital duodenal atresia, duodenal stenosis or duodenal web. Transabdominal sonography may demonstrate associated anomalous pancreatic tissue encircling the duodenum, consistent with an annular pancreas [12]. The portability of ultrasound and lack of ionizing radiation makes this the ideal test for evaluating the neonate with obstructive symptoms.

### Differential Diagnosis:

The imaging findings of pancreatic tissue encircling the second portion of the duodenum (and the variations described above) are diagnostic of annular pancreas. However, findings that raise concern for duodenal obstruction, such as the double bubble sign in infants (with or without lack of distal bowel gas), are nonspecific. The differential diagnosis of duodenal obstruction can be divided into intrinsic and extrinsic causes

[12]. Important intrinsic causes of obstruction include duodenal atresia, stenosis and webs. Extrinsic causes to be considered include annular pancreas and malrotation of the gut with Ladd bands or midgut volvulus [12]. Midgut volvulus is a condition in which the intestine twists upon itself, usually during fetal development, which on ultrasound and CT imaging may create the appearance of a whirlpool with the superior mesenteric artery at the center [13]. These conditions tend to present early in life.

Adults with symptoms of acute or chronic biliary colic will often undergo imaging with CT or MR imaging. Because symptoms may be nonspecific or mild in many cases, adults diagnosed with annular pancreas may only learn of their condition once the symptoms are persistent or severe enough to trigger an evaluation with imaging. Duodenal or pancreatic carcinoma may mimic annular pancreas although carcinoma tends to present late in the disease with obvious metastatic disease in about half of the cases [14]. Soft tissue prominence and thickening of the bowel wall may make differentiation from annular pancreas difficult, especially in cases without obvious invasive features at imaging or endoscopy. Pancreatic adenocarcinoma often presents as a hypoenhancing parenchymal mass with biliary and/or pancreatic ductal dilatation. These tumors also demonstrate a propensity for venous and arterial encasement, which leads to difficult resections or inoperable disease [15]. As with primary duodenal carcinomas, pancreatic adenocarcinoma will frequently present with metastatic disease to the liver, lymph nodes or peritoneum.

Treatment options:

There are no specific guidelines for the treatment of annular pancreas [16]. Symptomatic adult patients presenting with acute pancreatitis can be managed with supportive care. Surgical intervention is typically reserved for patients presenting with refractory duodenal and gastric outlet obstruction. When surgical intervention is necessary, duodenal bypass is the procedure of choice in both adults and children [8]. This can be achieved with duodenoduodenostomy or gastrojejunostomy [6,8,11,17,18]. More complex procedures disrupting the annular pancreas through significant pancreatic or biliary operation, such as pancreaticoduodenectomy and hepaticojejunostomy are less common.

TEACHING POINT

Annular pancreas is a congenital condition that in infants is often associated with other congenital anomalies, sometimes causing duodenal obstruction. When gone undetected, it can manifest later in adult life with chronic or acute biliary colic symptoms and acute pancreatitis due to annulus compression of the duodenum. In the absence of risk factors for pancreatitis, such as cholelithiasis, alcoholism, viral infections, hypertriglyceridemia or neoplasm, annular pancreas, whether complete or incomplete, should be considered in the differential diagnosis of acute pancreatitis.

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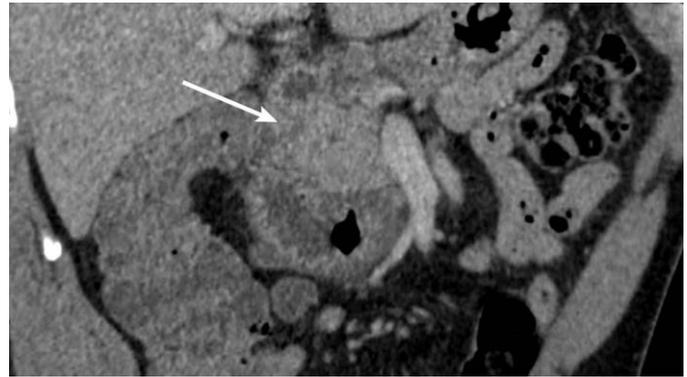
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**Figure 2:** 58-year-old male with annular pancreas presenting with acute pancreatitis.

Findings: Coronal contrast enhanced magnified oblique coronal CT in the venous phase demonstrate narrowing of the second portion of the duodenum (arrow) secondary to annular pancreatic tissue.

Technique: Coronal contrast enhanced CT, 202mA, 120kV, 1.25 mm slice thickness, intravenous contrast: 100mL of ISOVUE 370, exam DLP 619.90. GE LightSpeed VCT.

FIGURES



**Figure 1:** 58-year-old male with annular pancreas presenting with acute pancreatitis.

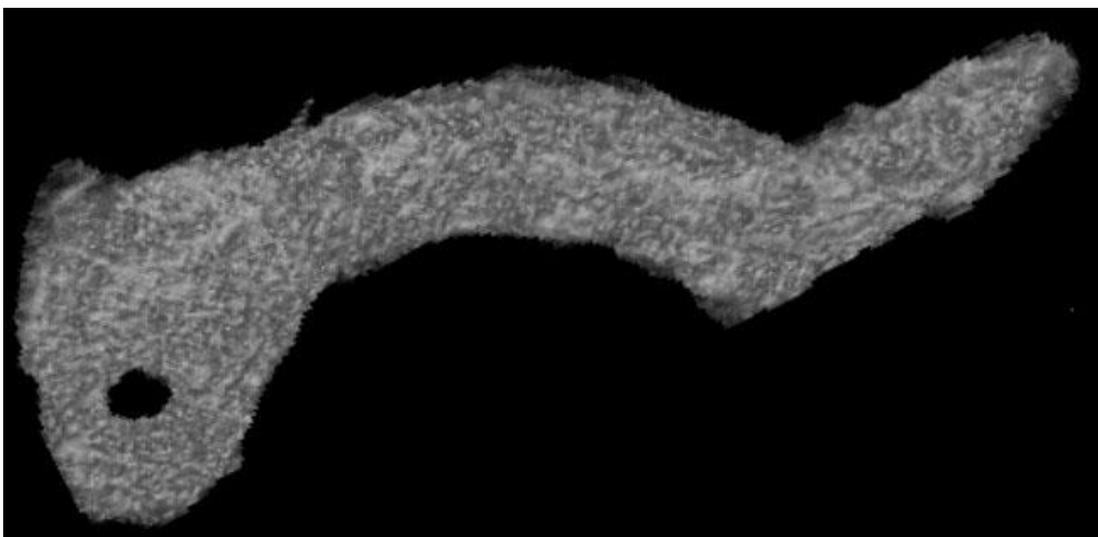
Findings: Axial contrast enhanced CT of the abdomen in the venous phase reveals pancreatic tissue surrounding the second portion of the duodenum (thin long arrow), peripancreatic fat stranding (thick long arrow), and inflammatory bowel wall thickening involving the adjacent colon (short arrow).

Technique: Axial contrast enhanced CT, 202mA, 120kV, 1.25 mm slice thickness, intravenous contrast: 100mL of ISOVUE 370, exam DLP 619.90. GE LightSpeed VCT.

**Figure 3 (bottom):** 58-year-old male with annular pancreas presenting with acute pancreatitis.

Findings: 3-D reconstruction from the source data with surrounding soft tissue subtracted, demonstrates a complete ring of pancreatic tissue through which the second portion of the duodenum traverses.

Technique: Contrast enhanced CT, 202mA, 120kV, 1.25 mm slice thickness, intravenous contrast: 100mL of ISOVUE 370, exam DLP 619.90. GE LightSpeed VCT.





**Figure 4 (left):** 58-year-old male with annular pancreas presenting with acute pancreatitis.

Findings: Axial contrast enhanced CT in the venous phase demonstrates normal appearance of the gallbladder (arrow) without cholelithiasis, obstruction of bile or main pancreatic ducts, or pancreatic parenchymal lesion.

Technique: Axial contrast enhanced CT, 202mA, 120kV, 1.25 mm slice thickness, intravenous contrast: 100mL of ISOVUE 370, exam DLP 619.90. GE LightSpeed VCT.

<b>Etiology</b>	Failure of normal rotation of the ventral pancreatic bud around the duodenum during gestation
<b>Prevalence</b>	Estimates range from 15 to 400 in 100,000 adults True prevalence unknown
<b>Gender Ratio</b>	Appears to affect males and females equally. No clear gender predilection
<b>Age Predilection</b>	Congenital anomaly
<b>Risk Factors</b>	Associated with Down Syndrome
<b>Treatment</b>	Surgery for refractory obstruction Supportive care for pancreatitis
<b>Prognosis</b>	Good
<b>Imaging findings</b>	X-ray/sonography (pediatric population): double bubble sign (nonspecific)  CT/MRI: pancreatic tissue encircling the second part of the duodenum. Crocodile jaw configuration with pancreatic tissue anterior and posterior or posterolateral to the second part of the duodenum.

**Table 1:** Summary table for annular pancreas.

Differential Diagnosis	Radiograph/Fluoroscopy	Ultrasound	CT/MRI
<b>Annular Pancreas</b>	<ul style="list-style-type: none"> <li>Double bubble sign: gaseous distention of the stomach and proximal duodenum (nonspecific, neonates)</li> </ul>	<ul style="list-style-type: none"> <li>Double bubble sign/duodenal obstruction (neonates)</li> <li>Anomalous pancreatic tissue encircling the duodenum (neonates)</li> </ul>	<ul style="list-style-type: none"> <li>Pancreatic tissue encircling the duodenum forming a complete or an incomplete ring around the duodenum.</li> <li>Partial annular pancreas can appear in a crocodile jaw configuration with pancreatic tissue anterior and posterior to the duodenum or with pancreatic tissue posterolateral to the second part of the duodenum.</li> </ul>
<b>Duodenal Web</b>	<ul style="list-style-type: none"> <li>Double bubble sign</li> </ul>	<ul style="list-style-type: none"> <li>Dilated stomach and proximal duodenum, fluid may outline thin web</li> </ul>	--
<b>Duodenal Atresia</b>	<ul style="list-style-type: none"> <li>Double bubble sign</li> <li>Absent distal bowel gas</li> </ul>	<ul style="list-style-type: none"> <li>Double bubble sign: dilated stomach and proximal duodenum</li> </ul>	--
<b>Midgut Volvulus</b>	<ul style="list-style-type: none"> <li>Double bubble sign: distended stomach and proximal duodenum with mild distal bowel gas</li> </ul>	<ul style="list-style-type: none"> <li>Double bubble sign: dilated stomach and proximal duodenum</li> <li>Whirlpool sign</li> </ul>	<ul style="list-style-type: none"> <li>Abnormal rotation of the intestine</li> <li>Whirlpool sign</li> <li>Bowel obstruction</li> </ul>
<b>Duodenal Carcinoma</b>	<ul style="list-style-type: none"> <li>Proximal bowel obstruction if severe narrowing</li> </ul>	<ul style="list-style-type: none"> <li>Hypoechoic, ill-defined mass</li> <li>Double bubble (late finding)</li> </ul>	<ul style="list-style-type: none"> <li>Intrinsic mass with irregular thickening of duodenal wall</li> <li>Concentric narrowing of duodenal lumen</li> <li>Liver, nodal or peritoneal metastases</li> </ul>
<b>Pancreatic Adenocarcinoma</b>	<ul style="list-style-type: none"> <li>Gastric or proximal small bowel obstruction</li> </ul>	<ul style="list-style-type: none"> <li>Hepatic or pancreatic ductal dilatation</li> <li>Hypoechoic, ill-defined mass</li> </ul>	<ul style="list-style-type: none"> <li>Hypoenhancing mass</li> <li>Biliary and/or ductal dilatation</li> <li>Vessel encasement</li> <li>Liver, nodal or peritoneal metastases</li> </ul>

**Table 2:** Differential diagnoses table for annular pancreas.

#### ABBREVIATIONS

CT = Computed Tomography  
 ED = Emergency Department  
 MR = Magnetic Resonance  
 MRCP = Magnetic Resonance Cholangiopancreatography  
 MRI = Magnetic Resonance Imaging

#### KEYWORDS

case report; tomography, spiral computed; CT; annular pancreas; abnormality, congenital; disease, gastrointestinal pancreatitis; crocodile jaw configuration; double bubble sign

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