

Annular Pancreas

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Abstract

Annular pancreas is a rare condition often presenting in neonates where pancreatic tissue encircles the duodenum, causing obstruction, vomiting, and abdominal distention. The causal mechanism of annular pancreas is unknown, but it is hypothesized to be a result of malformations in embryological development. Imaging is part of the diagnostic workup in neonates with bilious emesis. However, in these patients, imaging is used to identify other, more serious conditions. Older children and adults typically have less severe symptoms. In these patients, the diagnosis may be made incidentally via imaging. Once diagnosed, patients are treated surgically.

Keywords: Abdomen, Retroperitoneal, Congenital

Case Summary

An adolescent with a history of gastric polyps, status post-polypectomy 1 week prior presented with inability to tolerate food and drink.

Imaging Findings

An upper gastrointestinal (GI) examination (Figure 1) was performed, showing a dilated proximal duodenum. A transition point was present in the second portion of the duodenum. CT (Figure 2) and MRI (Figure 3) were then performed. Both show the pancreatic head encircling the second portion of the duodenum.

Diagnosis

Annular pancreas.

The presentation of annular pancreas (AnP) differs depending on the age of discovery. When presenting in neonates, it

often presents as a high-grade obstruction of the duodenum, causing a double-bubble sign with a differential diagnosis, including duodenal atresia, duodenal stenosis, duodenal web, malrotation, and AnP.¹ In an older child or adult patient, it may present with inability to tolerate meals, raising the possibility of peptic ulcer disease, pancreatic divisum, allergy, obstruction, infection, and primary duodenal and pancreatic malignancies, which should be additionally considered rather than only congenital pathologies.¹

Discussion

AnP is a rare congenital malformation in which the pancreatic tissue encircles the duodenum.² In embryonic pancreatic development, the dorsal and ventral endodermal buds arise by the 4th embryological week and fuse together by the 7th embryological week to form the pancreas.³ While the exact mechanism is

unknown, AnP is thought to arise from faulty rotation of the ventral pancreatic bud around the posterior duodenal anlage.² This absence of rotation results in the ventral bud enveloping the duodenum, constricting its lumen and causing obstruction. This condition can present as complete encircling or incomplete/partial encircling, and, depending on the severity of the partial variant of AnP, both can present symptomatically and radiologically equivalent.⁴

Patients with AnP may present during the neonatal period with bilious emesis and abdominal distention due to duodenal obstruction.⁵ However, some patients are not diagnosed until adulthood.⁵ Approximately 60% of adults with AnP are asymptomatic. In these patients, the diagnosis may be made as an incidental finding on imaging.⁶ Symptomatic adult patients may have acute or chronic pancreatitis, gastritis, duodenitis, jaundice, or GI outlet obstruction.⁵ Patients with congenital abnormalities such as Down syndrome, esophageal atresia, duodenal atresia, tracheoesophageal fistula, and Meckel diverticulum have higher rates of AnP compared with the general population.⁵

AnP may be diagnosed on prenatal imaging. The double-bubble sign of 2 fluid-filled structures, representing the

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Figure 1. Upper gastrointestinal examination showing a dilated first portion of the duodenum with a focal transition point (arrow) at the mid-second portion of the duodenum. Bowel distal to this is decompressed.

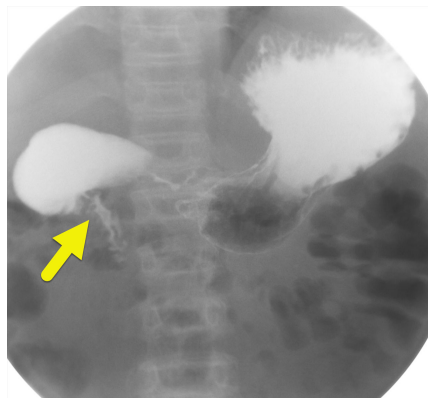


Figure 2. (A) Axial and (B) coronal contrast-enhanced CT images showing the duodenum (arrow) encircled by the head of the pancreas consistent with annular pancreas.

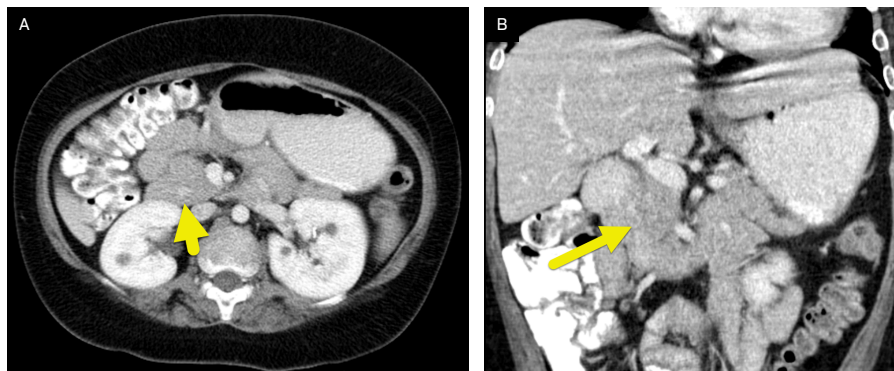


Figure 3. MRI of the (A) axial precontract T1-weighted image, (B) axial T1 gradient recalled echo image, and (C) coronal Fast Imaging Employing Steady-State Acquisition showing the second portion of the duodenum (arrow) encircled by the head of the pancreas.



stomach and proximal duodenum, may be identified on routine fetal US or MRI.^{7,8}

Abdominal radiography is often the first imaging study performed in neonates with bilious emesis. In these patients, a double bubble with a dilated air-filled stomach and proximal duodenum is noted. Distal bowel gas may be present. The finding of distal bowel gas can help exclude duodenal atresia from the differential diagnosis. An upper GI study may be performed next. In this study, the stomach and proximal duodenum are dilated and fill with contrast. There is often a delay of contrast passage to the decompressed mid and distal duodenum. In this setting, it is not possible to distinguish AnP from duodenal web or duodenal stenosis.

The diagnosis of AnP can be made via US. This modality shows a hyperechoic band of pancreatic tissue encircling the descending duodenum, causing an acute angle between the pre-stenotic and post-stenotic descending duodenum. This band of pancreatic tissue surrounding causes the duodenum to have a characteristic “S” shape starting from the pylorus and duodenal bulb.⁹ However, in patients with incomplete or partial AnP, these imaging findings are less specific.⁹ While these findings are helpful in some instances, upper GI is a more specific modality.¹

In older patients, endoscopic retrograde cholangiopancreatography is thought to be the gold standard to diagnose AnP.¹⁰ This modality allows the endoscopist to view the pancreatic duct along with the major

and minor papilla.¹⁰ In most patients with AnP, the ring of pancreatic tissue is proximal to the major and minor papillae.¹⁰

Magnetic resonance cholangiopancreatography (MRCP) and contrast-enhanced CT allow for direct visualization of the pancreatic tissue surrounding the duodenum.¹⁰ When dilated, the pancreatic ducts can be seen on MRCP encircling the duodenum. Occasionally, AnP is diagnosed via the upper GI later in life. In these instances, there is regional circumferential narrowing of the second portion of the duodenum.

Surgery is used to resect the annular portion of the pancreas in symptomatic children. In neonates, a diamond-shaped or side-to-side duodenoduodenostomy is

performed.⁶ Outcomes are similar between the 3 approaches.⁶ The distal duodenal diameter (with a smaller diameter favoring the longitudinal incision in a diamond-shaped duodeno-duodenostomy) and the surgeon's experience are the primary deciding factors in selecting an approach.⁶ Surgical complications are rare, but may include blind-loop syndrome (in which impaired absorption of nutrients and an overgrowth of bacteria occurs due to bypassing a segment of bowel), megaduodenum, gastritis, and esophageal reflux.⁶ Surgery is generally successful in alleviating duodenal obstruction. However, long-term complications may occur. These problems include cholestatic jaundice (53%), upper GI motility issues (47%), failure to thrive (40%), chronic diarrhea (33%), and steatorrhea occasionally.¹¹

Conclusion

AnP is a rare condition often presenting in neonates where pancreatic tissue encircles the duodenum causing obstruction, vomiting, and abdominal distention. The causal mechanism of AnP is unknown, but it is hypothesized to

be a result of malformations in embryological development. Imaging is part of the diagnostic workup in neonates with bilious emesis. However, in these patients, imaging is used to identify other, more serious conditions. Older children and adults typically have less severe symptoms. In these patients, the diagnosis may be made incidentally via imaging. Once diagnosed, patients are treated surgically.

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