CHAPTER 84
Annular Pancreas

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Introduction

Annular pancreas (from the Latin annularis, meaning ring-shaped) is a rare congenital disorder of the pancreas first recognised by Tiedeman in 1818. This abnormality, although at times clinically silent, may be the cause of a broad spectrum of disease. Complications range from neonatal intestinal obstruction to more complex pathologies in the adult. In cases of neonatal obstruction, annular pancreas is an important structural and anatomical cause that must be identified and treated appropriately. Currently, the majority of cases are diagnosed early in life, and prenatal diagnosis is becoming increasingly important.

Demographics

The incidence of this congenital anomaly is reported as 1–3 in 20,000,[1,2] and some studies have shown that it is more common in males. Detection of the condition is variable, as it may be asymptomatic and therefore detected only incidentally or at postmortem. However, the vast majority of cases are diagnosed either prenatally or in the first few days of life. If the condition is not diagnosed prenatally or does not present with complications in early life, it may be undetected until adulthood. The detection of an annular pancreas may occur at any time during adulthood (presentation age shows a bell-shaped distribution[1]), and may be discovered either incidentally or after presentation due to a complication such as pancreatitis.

There is a strong association between annular pancreas and other congenital abnormalities—up to 71% of cases have coexisting congenital anomalies.[1] The most common association is with Down syndrome. However, there may be a wide range of associated cardiac and gastrointestinal anomalies (including Hirschsprung's disease and imperforate anus), as well as tracheo-oesophageal fistula and oesophageal atresia.[3]

Aetiology and Pathophysiology

Annular pancreas is an embryological defect of the foregut. Development of the pancreas begins during week 5 of gestation. One dorsal and two ventral buds develop from the primitive foregut. By week 7, the ventral bud rotates with the gut to fuse with the dorsal bud, after passing behind the duodenum. The dorsal bud forms the body and tail of the pancreas, and the ventral bud forms the inferior part of the pancreatic head and the uncinate process. Fusion of the buds forms the main pancreatic duct.

Several theories have been proposed to explain the development of annular pancreas. One theory suggests that the tip of the ventral bud fuses abnormally to the duodenum, therefore rotating incorrectly around the duodenum and resulting in a band of fibrous or pancreatic parenchymal tissue around the second part of the duodenum.[1] An alternative theory posits that hypertrophy of the dorsal and ventral buds results in a complete band of pancreatic tissue around the duodenum (a complete ring is found in approximately 25% of the cases[4]).

Clinical Presentation

History

The diagnosis of annular pancreas may be made prenatally, upon emergency presentation, or incidentally after imaging, at operation, or postmortem. If the band of pancreatic tissue causes a duodenal obstruction, symptoms may appear within the first few hours of life. Signs of neonatal intestinal obstruction may initially be nonspecific, including poor feeding, vomiting, and irritability. If proximal to the ampulla of Vater, vomiting may be nonbilious, and therefore could be confused with less severe, non–life-threatening conditions.

The nature of presentation differs according to patient age, although most presentation occurs most commonly in infancy or early childhood. Children frequently present with gastrointestinal (GI) symptoms, including poor feeding, vomiting, and abdominal distention. In Africa, most patients present late, with malnutrition, failure to thrive, bile-stained vomiting, and—less frequently—abdominal cramps.

Of note is the fact that adults are much more likely to describe upper abdominal pain or present acutely with pancreatitis.[1] Complications arising due to the presence of an annular pancreas are a rare cause of neonatal or childhood presentation, but must be kept in mind, as cure requires identification and treatment of the abnormality.

Physical

Clinical examination findings will vary according to the age at presentation and the extent of systemic upset. For some patients, access to a hospital may be difficult—this includes patients whose presentation is delayed by parents and those who have to travel long distances to reach the hospital. These patients will likely be more unwell at presentation and may even die before they reach the hospital.

When presenting as intestinal obstruction, the abdomen will be distended (before decompression), and there may be palpable peristalsis. Bowel sounds are variable. If presentation is delayed, there may be clinical signs of hypovolaemic shock, including pallor, poor capillary refill, and drowsiness or lethargy.

Investigations

Prenatal diagnosis

It is becoming increasingly possible to detect the presence of annular pancreas in foetal life (2nd trimester). A nonspecific diagnosis of duodenal obstruction can be made prenatally with identification of simultaneous dilatation of the stomach and duodenum—the "double bubble" sign.[4] This does not predict the specific cause of the duodenal obstruction, and certainly is not specific to the diagnosis of annular pancreas. Indeed, presence of an annular pancreas is the cause in only 1% of cases of neonatal duodenal obstruction.[5] Therefore, there is a search for more specific prenatal markers of annular pancreas. Several groups have shown that the presence of hyperechogenic bands around the duodenum specifically indicates the presence of an annular pancreas.[4] This finding can now be used to specifically detect the presence of annular pancreas during foetal life.

Imaging modalities

Abdominal radiography

The plain abdominal radiograph will not provide diagnostic structural information about the pancreas, in part due to the lack of tissue contrast. Cross-sectional imaging modalities are much more useful in delineating the precise anatomy of this area. However, if obstruction is complete, duodenal obstruction classically appears as a double bubble on abdominal radiograph. This represents dilatation of the stomach (seen in the left upper quadrant) and proximal duodenum (seen in the right upper quadrant). A double bubble on abdominal radiograph is sufficient to diagnose a complete duodenal obstruction. However, an upper gastrointestinal contrast radiograph is helpful for diagnosing a partial duodenal obstruction.

Ultrasonography

The use of ultrasonography (US) allows a real-time evaluation of the paediatric pancreas, without the use of ionising radiation and usually without the need for sedation. This modality provides a safe, relatively cheap, and often readily available technique for imaging the paediatric pancreas. The reduced body wall thickness of the child, in combination with the anatomical "window" provided by the liver, ensures that a better quality of pancreatic image is obtained in the child, as compared to the adult.[3]

Computed tomography

The lack of intraabdominal fat in children, combined with the often thin band of pancreatic tissue around the duodenum, may make identification of the annular pancreas with computed tomography (CT) difficult. Despite this, detection rates can be greatly improved with a specific, intravenous (IV)-contrast-enhanced, pancreatic study.[3,6] CT may also highlight extrinsic compression of the duodenum, which must be differentiated from thickening of the wall of the duodenum. CT may be used for further investigation of abnormalities detected with ultrasound imaging.

Magnetic resonance imaging

Magnetic resonance imaging (MRI) is an alternative to CT imaging for evaluation of the pancreas. Combined with magnetic resonance cholangiopancreatography (MRCP), it allows detailed imaging of the biliary system.[3]

Typically, the exact cause of the duodenal obstruction is not known until surgical exploration.

Management

In clinically silent cases of annular pancreas, no specific intervention is required. Specific management will depend upon the nature of the complication or symptoms caused by the annular pancreas.

Duodenal Obstruction

Ultimately, urgent surgical intervention is required in cases of duodenal obstruction, even if the precise cause of the obstruction is unclear. Initially, however, management must include decompression with a nasogastric tube and fluid resuscitation. It is appropriate for patients to be transferred to a special care/high dependency unit, if available, where appropriate monitoring and interventions can be instigated. Delayed presentation may result in significant systemic upset, and it is vital that adequate resuscitation be carried out before surgery. Prior to surgery, fluid and electrolyte resuscitation is an absolute requirement.

The management of duodenal obstruction in the presence of annular pancreas requires surgical bypass of the obstruction with duodenoduodenostomy, or duodenojejunostomy. Laparotomy may be required to determine the cause of obstruction if this is not specifically identified on preoperative imaging. Tapering of the bowel is not necessary, even if the proximal segment is grossly dilatated. Transanastamotic nasogastric tubes may be necessary in centers where parenteral nutrition is not available, due to the delay in enteral feeding as a result of the dilatated proximal segment.

Prognosis and Outcomes

Successful treatment of neonatal intestinal obstruction requires rapid identification, resuscitation, and definitive management. In addition to the technical skill required to bypass the obstruction caused by abberant pancreatic tissue, appropriate perioperative care is required to ensure a successful outcome after surgery. Surgical treatment of annular pancreas has an excellent prognosis,[7] as long as there is sufficient appropriate perioperative and postoperative care. This includes appropriately trained anaesthetic staff, ventilatory support, and the facility for parenteral nutrition.[8]

Ultimately, and despite appropriate surgical intervention to relieve duodenal obstruction if present, the outcome may be affected by the presence of severe associated congenital abnormalities.

Prevention

With the improvement in prenatal diagnosis—in particular, the identification of specific markers for annular pancreas—detection of this congenital abnormality can be optimised. Although it cannot be prevented, earlier identification of complications—in particular, intestinal obstruction—will help to ensure that appropriate treatment can be instigated as soon as possible.

Evidence-Based Research

Table 84.1 presents a retrospective review of cases of annular pancreas for a period of 10 years.

Table 84.1: Evidence-based research.

Title	Annular pancreas in children: a recent decade's experience
Authors	Jimenez JC, Emil S, Podnos Y, Nguyen N
Institution	Division of Pediatric Surgery, Irvine Medical Centre, Orange, California, USA; Department of Surgery, Miller Children's Hospital, Long Beach, California, USA
Reference	J Pediatr Surg 2004; 39(11):1654–1657
Problem	A 10-year review of clinical, radiological, and prognostic findings in patients with annular pancreas.
Intervention	Retrospective review of all cases of annular pancreas between 1993 and 2002
Outcome/effect	All patients required surgical intervention, and all patients survived to be discharged from hospital (mean stay, 24 days).
Historical significance/ comments	This paper highlights the rarity of this condition and its association with other congenital abnormalities. Despite the high percentage of concurrent abnormalities, all patients survived and were discharged from the hospital.

Key Summary Points

1. Annular pancreas is a rare congenital structural abnormality of the primitive foregut.
2. There is a strong association between annular pancreas and other congenital abnormalities, particularly Down syndrome.
3. The spectrum of disease that may be caused by annular pancreas is broad.
4. Diagnosis may be made prenatally with evidence of duodenal obstruction. More specific markers for prenatal detection of annular pancreas are currently under investigation.
5. A plain abdominal radiograph will show evidence of duodenal obstruction, but ultrasound or cross-sectional imaging techniques are required to specifically identify annular pancreas.
6. Urgent surgical intervention is required in cases of duodenal obstruction caused by annular pancreas. Surgery is not required to correct annular pancreas if there are no complications caused by its presence.
7. Prognosis after surgery for duodenal obstruction is good; however, overall outcome will also be affected by the severity of associated congenital anomalies.

References

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