

Review Article

Annular pancreas in adults: a tertiary care institute experience and review of literature

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ABSTRACT

Annular pancreas (AP) in adults is a rare congenital anomaly characterized by the presence of ectopic pancreatic tissue surrounding the second part of the duodenum. This rare embryologic abnormality is usually detected after development of complications or as incidental finding in most cases. Diagnosis and treatment strategies for symptomatic adult AP remain controversial. No clearly defined specific guidelines and treatment protocols exist about management of adult AP, therefore, treatment and operative approaches must be individualized for each patient. It is always prudent to consider the possible post-operative complications, surgical treatment should be reserved only for cases where there is complete obstruction.

Keywords: Annular pancreas, Duodenojejunostomy

INTRODUCTION

Annular pancreas, more often described as a congenital anomaly of pancreas, was first described by Tiedemann in 1818.¹ It is a rare congenital anomaly which most commonly manifests itself in children but some may remain asymptomatic and present in late adulthood due to associated complications.² Nearly 400 reports of annular pancreas have been published in literature till date.³

Normally pancreas develops at 5th week of gestation, from one dorsal and two ventral buds which appear as evaginations of the primitive foregut.⁴ The two ventral buds fuse early to become one. Due to selective expansion of the duodenum by about seventh week, the ventral bud rotate along with the gut, thereby passing behind the duodenum from right to left. The unified ventral bud eventually fuses with dorsal bud. Failure of the rotation of ventral bud with the duodenum, results in annular rim of pancreatic tissue enveloping the second

part of duodenum. Not always this encirclement is complete, on occasions this may be incomplete leaving the anterior portion of the duodenum unconstricted. 25% cases form a complete pancreatic ring, and 75% have a partial ring.⁵ The pancreatic band is usually interspersed with the duodenal muscularis, although it can also be free from duodenum.

The annular pancreas is a rare congenital anomaly that affects males more commonly than females and occurs at a frequency of 1 in 20,000 births. In up to 50% of the cases, it is associated with other congenital anomalies including down's syndrome, tracheoesophageal fistula, esophageal atresia, imperforate anus and hirschsprung disease.⁶ Other pancreatic anomalies such as pancreaticobiliary malrotation, and pancreas divisum, are also associated with the annular pancreas.⁷ Almost half of patients present in the pediatric age group and about 90% of these are in the neonatal period. In adults, annular pancreas usually presents between third to fifth decade.

However, it has been estimated that only about one third are symptomatic and is most commonly associated with abdominal pain and gastric outlet obstruction, secondary to duodenal narrowing.

The symptoms of adult annular pancreas include abdominal pain, vomiting, weight loss, abdominal fullness and sometimes jaundice. When obstructive jaundice occurs, carcinoma of the ampulla of Vater should be considered as a possibility.⁸

The symptoms in adults are most often due to complications of associated peptic ulcer, pancreatitis, duodenal obstruction and biliary obstruction.⁹ The annular pancreas can present itself in a wide range of clinical severities depending upon the degree of duodenal obstruction. Even with complete annular pancreas, the food usually can pass through the duodenum without any difficulty. But later in the course, secondary to chronic pancreatitis and peptic ulcer disease, duodenal narrowing manifests and patient presents with features of gastric outlet obstruction.

Preoperative diagnosis is often difficult and imaging is of paramount importance to establish correct diagnosis. In adult patients, upper GI series have been considered the initial study of choice.¹⁰ A 'double bubble' sign on the simple radiograph can help to make a diagnosis of the annular pancreas, but this is neither specific nor sensitive.¹¹ Typical findings include stenosis or annular defect of the second portion of the duodenum, dilatation of the proximal stomach and D1 and retrograde intestinal movement from the proximal part to obstruction.⁷

Computed tomography scan illustrates the pancreatic tissue completely or partially encircling the duodenum. Abdominal CT and endoscopic ultrasonography are helpful to diagnose the annular pancreas which is usually made with computed tomography scanning and confirmed with upper gastrointestinal contrast fluoroscopy.⁹

An endoscopic retrograde cholangiopancreatography (ERCP) can make a specific diagnosis when the pancreatic duct is outlined. Around 85% of the pancreatic annular duct join the main pancreatic duct.¹² The typical appearance of an annular pancreatic duct is the complete encirclement of the second portion of the duodenum. However, it is difficult to use ERCP to diagnose patients with annular pancreatic ducts joining the accessory pancreatic duct and its indication is limited in patients with cicatrizing duodenal stenosis proximal to ampulla of vater. Other drawbacks of ERCP is that it is invasive and can cause iatrogenic pancreatitis. MRCP is a non invasive method of visualising the pancreatic duct encircling the duodenum, but sometimes the undilated annular pancreatic duct is not visible on routine MRCP.¹³ So, surgery is still ultimate for confirming the diagnosis in symptomatic cases.

The treatment of annular pancreas has been surgical in symptomatic cases. Surgical bypass of the narrowed duodenum is indicated in severe stenosis. The various procedures that has been described in literature includes gastro-enteral anastomosis and truncal vagotomy; duodenojejunostomy anastomosing first part of the duodenum with the jejunum; latero-lateral anastomosis of the pyloroduodenal portion with the jejunum; duodenoduodenal anastomosis; duodeno-jejunal anastomosis with Roux en Y loop and even resection of the annular pancreas. Bypass is always superior to local resection of the annular pancreas. The separation of the annulus is associated in 50% of the cases with serious complications.

Local resection of the annular segment is better avoided for the fear of development of pancreatic fistula. Resection is often difficult because of dense adhesions due to the local fibrosis, which can also be complicated by postoperative pancreatitis and recurrence of duodenal stenosis.¹⁴ Table 1 shows authors' experience with 6 cases of annular pancreas in the past two years.

Table 1: Annular pancreas, types and management; our institute experience.

Age	Sex	Diagnosis	Type	Management
55	Male	Annular pancreas with adhesive colic	Incomplete	Conservative
40	Female	Post bulbar ulcer with annular pancreas	Incomplete	Conservative
19	Female	Corrosive antral stricture with annular pancreas	Complete	Truncal vagotomy with gastrojejunostomy
63	Female	Bleeding gastric malignancy with annular pancreas	Complete	Total gastrectomy with esophagojejunostomy
34	Female	Goo with annular pancreas	Complete	Duodenojejunostomy
50	Male	Annular pancreas with pangastritis	Incomplete	Conservative

Duodenoduodenostomy or duodenojejunostomy are the surgical procedures of choice.¹⁵ Vidal in 1905, first

reported successful gastrojejunostomy in a 3-year-old patient with annular pancreas. Although

duodenoduodenostomy is routinely preferred in neonates and pediatric age groups, duodenojejunostomy or gastrojejunostomy especially minimally invasive is an attractive alternative option in adults with fibrotic duodenal C-loop.⁷

Zyromski et al, reported that duodenoduodenostomy or duodenojejunostomy caused less anastomotic site ulcers than gastrojejunostomy.³ Gastroenterostomy with truncal vagotomy is performed where a peptic ulcer is also present or as a prophylactic measure to avoid post anastomotic ulcer.

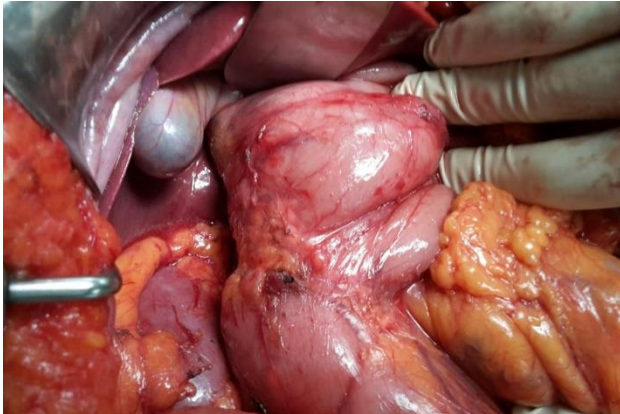


Figure 1: 34-year-old female with complete annular pancreas with gastric outlet obstruction.

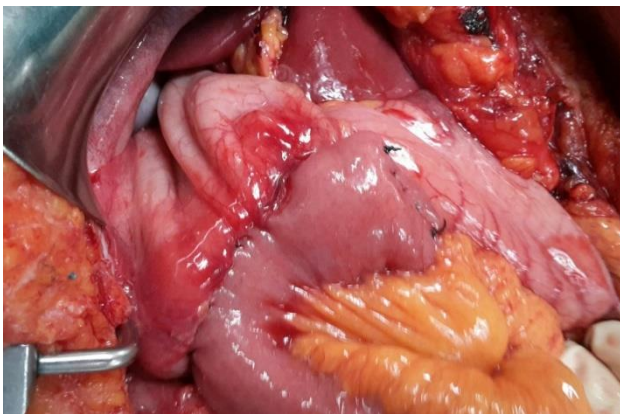


Figure 2: Side to side duodenojejunostomy done proximal to the duodenal narrowing without disturbing the annular pancreatic tissue.

Laparoscopic duodenoduodenostomy and laparoscopic gastrojejunostomy are successfully performed in pediatric patients and in adult patients, respectively. Laparoscopic gastrojejunostomy with vagotomy is a good treatment option for adult patients with annular pancreas.

CONCLUSION

Annular pancreas is a rare malformation that can present either in infancy or adulthood. Although the etiology of the annular pancreas has not been fully elucidated,

multiple developmental factors have been implicated in the pathogenesis. Most affected patients present with signs and symptoms of a proximal intestinal obstruction. After evaluation, these patients can be managed safely with surgical bypass of the annulus to restore intestinal continuity. Asymptomatic incidentally detected annular pancreas in adults can however be safely observed.

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