

# Annular pancreas: case report and review of the literature

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## SUMMARY

Annular pancreas is defined as a pancreatic ring which encircles the descending portion of the duodenum. Since its first description in 1818, attention has been paid to this variation due to its clinical manifestations, which may appear in the first days of life or even in adulthood, and usually require surgical treatment. Although an infrequent variation, more and more reports have been published due to new and improved diagnostic techniques, which include magnetic resonance cholangiopancreatography, endoscopic ultrasonography, and endoscopic retrograde cholangiopancreatography. A case of annular pancreas found during routine abdominal dissection of an adult female cadaver is reported here. It consisted of a complete pancreatic ring which surrounded the descending part of the duodenum. The duodenum was clearly distended proximally. After careful dissection, the ring presented a duct with a circumferential direction emptying into the pancreatic duct. The presence of this variation is associated with the embryology of the ventral pancreatic anlage. Clinical, diagnostic and therapeutic implications of this variation are discussed in this article.

**Key words:** Anatomy – Pancreas – Congenital – Abnormality – Annular

## INTRODUCTION

Annular pancreas (AP) is defined by the presence of a pancreatic ring that encircles the descending part of the duodenum (D2). Its first description is acknowledged to be Tiedemann

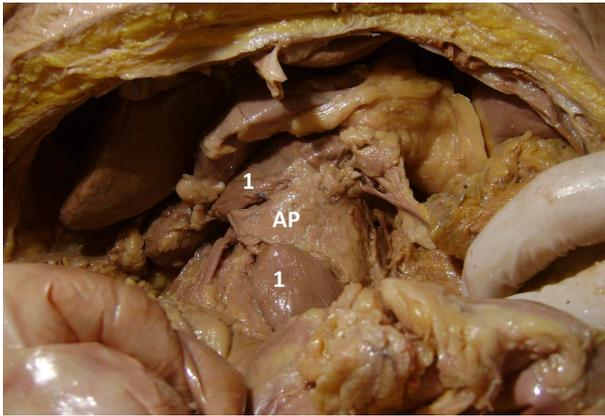
(1818), but it was first named “annular pancreas” by Ecker (1862). The first successful treatment case was reported by Vidal (1905), and consisted in a gastroenterostomy. Since its first reports, attention has been paid to this variation due to its clinical manifestations, which may appear during the first days of life or in adulthood (Zyromski et al., 2008). With the improvement of imaging techniques, more and more reports of AP are found by magnetic resonance cholangiopancreatography (MRCP), endoscopic ultrasonography (EUS) and endoscopic retrograde cholangiopancreatography (ERCP) (Fu et al., 2005). For these reasons, the presence of the AP may not be omitted by both kinds of surgeons (general and pediatric), or by radiologists. A cadaveric case of AP is reported here. Embryological, clinical, diagnostic and therapeutic implications of this variation are discussed.

## CASE REPORT

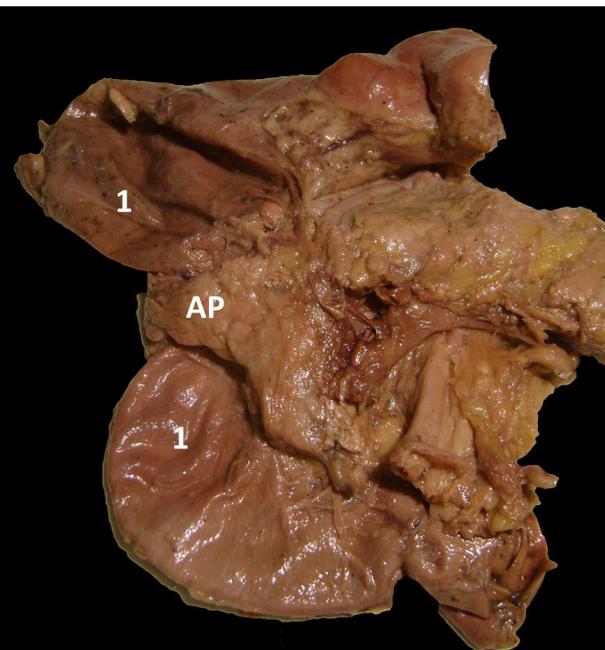
During a routine abdominal dissection of a female formalin-fixed cadaver aged 65, with no record of abdominal pain or vomiting, an anatomical variation was found in the pancreatic-duodenal area. The dissection was performed in the Anatomy Department, Facultad de Medicina, Universidad de la República, Montevideo, Uruguay. At first sight, it appeared as a pancreatic band that rose from its head and covered D2 (Fig. 1). After performing a duodeno-pancreatectomy with splenectomy, it was found that the pancreatic tissue completely encircled D2, corresponding with an AP (Fig. 2). It consisted of a 360 degree pancreatic ring, with a minimal width of 2cm on its lateral aspect (Fig. 3) and a maximum of 5,5cm on its posterior aspect (Fig. 4). The duodenum proximal to the AP was clearly distended (Figs. 2-3). Macroscopically, no bridging fibers were found that joined the AP to the duodenum. In fact, a plane for dissecting

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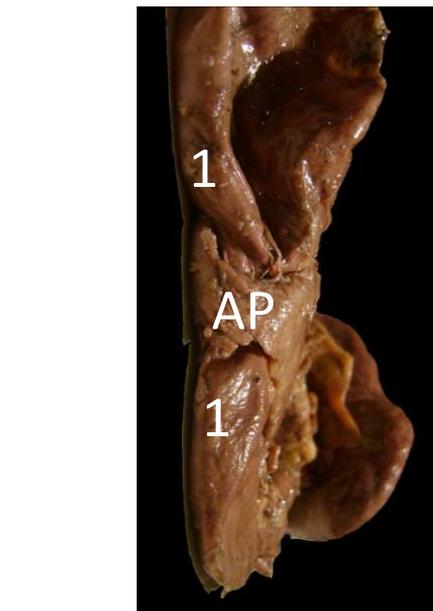
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**Fig. 1 (top).** *In situ* anterior abdominal view. Note the pancreatic band (AP) surrounding the descending duodenum (1).



**Fig. 2 (middle).** *Ex situ* anterior view. Note the pancreatic band (AP) completely surrounding the descending part of the duodenum (1). Proximally, the duodenum is clearly distended.



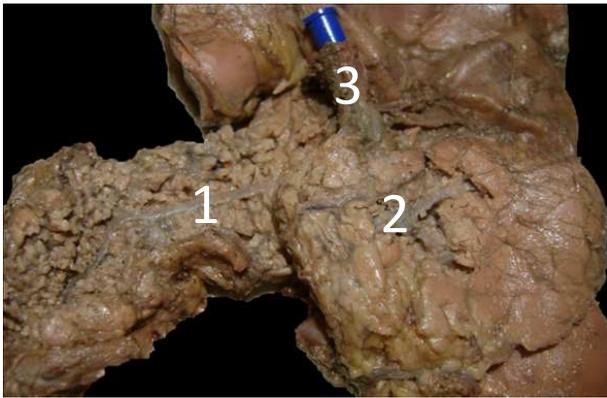
**Fig. 3.** *Ex situ* lateral view. AP encircling the lateral aspect of the descending part of the duodenum (1). Proximally, the duodenum is clearly distended.

the AP away the duodenum was found. Careful dissection of the pancreatic duct system showed a pancreatic duct with its usual pattern, which received near its termination the annular duct. This had a circumferential pattern around the AP (Fig. 4). Complete inspection of the abdominal cavity showed no other associated anomalies. For purposes of reviewing the literature, the PubMed database was queried using the key term “annular pancreas”, and limiting the search to references published in English. Additional reports were identified from the bibliography of selected articles.

## DISCUSSION

The incidence of AP is very low. To our knowledge, the biggest post-mortem series corresponds to Ravitch and Woods (1950), with 3 cases in 20000 specimens. Surgical series by Vasconcelos and Sadek (1949) found a single case in 22243 surgical cases. Incidence reported in patients undergoing ERCP has been 1:1000 (Fu et al., 2005). A recent EUS series (Papachristou et al., 2007), showed 5 cases in 9776 patients. Finally, after reviewing 7897 prenatal 2-D ultrasonographies, Dankovic et al. (2008) found 3 cases which were confirmed postnatally. Several theories have been proposed to explain the embryology of AP (Ikeda and Irving, 1984). Following Tieken (1901), AP is a consequence of hypertrophy of both ventral and dorsal primordia. Lecco (1910) postulated that fixation of the tip of the ventral anlage to the duodenal wall before gut rotation would result in the anlage being stretched around the duodenum to form the AP. In favor of this theory, the AP duct encircles D2, finally entering the pancreatic duct, as in the case here reported. Finally, Baldwin (1910) stated that the persistence and enlargement of the left ventral anlage is responsible of the presence of AP.

AP is described to have a 42% of incidence in models with homozygous inactivation of the *Indian hedgehog* gene (Hebrok et al., 2000). The latter, with the addition of a reported case of duodenal obstruction with AP in identical twins (Kumar et al., 2006), provides evidence for AP as a possible genetic disease. AP is one of the few embryological anomalies in which the symptoms may start later in life (Alexander, 1970). If the AP is very tight from the outset, it will cause obstruction in early childhood (Alexander, 1970). Now, why should signs of



**Fig. 4.** *Ex situ* posterior view. After careful dissection, the pancreatic duct (1) is visualized. Dissection of the AP showed its duct (2), with a circumferential pattern. For reference the common bile duct (3) is catheterized in blue.

duodenal obstruction develop in an elderly person or in a teenager? There are basically two main theories: on one hand, AP is associated with pancreatitis (incidence between 13-22%, Zyromski et al., 2008), insufficient drainage of pancreatic juice through the annular duct, and duodenal stagnation is a possible pathology (Urushihara et al., 2010), and this inflammatory process may compress and obstruct D2. On the other hand, the duodenal obstruction could cause a ptosis of the first portion of the duodenum that could lead to angulation at the annulus (Alexander, 1970), a morphological change that was found in the specimen here presented (Fig. 2). AP accounts for 5% of all duodenal obstructions (Jimenez et al., 2004). Symptomatic children usually present vomiting, bloating and feeding intolerance (Jimenez et al., 2004). In contrast, the majority of adults with AP present abdominal pain, and less frequently vomiting, nausea and other gastrointestinal symptoms (Zyromski et al., 2008).

The association of other congenital anomalies with AP in children is well recognized (Zyromski et al., 2008). The most common congenital chromosomal anomaly is Down syndrome (Zyromski et al., 2008). Other anomalies commonly associated with AP include cardiac defects, intestinal atresia, biliary anomalies, and tracheoesophageal fistula (Zyromski et al., 2008). Diagnostic techniques include prenatal ultrasonography, based on detection of the double-bubble sign, resulting from simultaneous dilatation of the stomach and duodenum (Dankovic et al., 2008). Dankovic et al. (2008) described in addition to the bubble sign a hyperechoic band which was highly suggestive of AP. The bubble sign is also described in plain radiographs for children, and less frequently in adults (Alexander 1970). Computed tomography can illustrate the pancreatic tissue encircling D2 (Singh et al., 2010). ERCP is specific in the diagnosis of AP when the pancreatic duct is outlined. Thus, when duodenal obstruction is present, ERCP may

be technically difficult or impossible (Fu et al., 2005). MRCP is a non-invasive method for visualizing the biliary tree and pancreatic duct (Mulugeta and Hilmes, 2010). When the aberrant pancreatic duct is identified by MRCP, the diagnosis of AP is made, as previously reported (Mulugeta and Hilmes, 2010). EUS assesses whether AP is complete or partial, and visualizes signs of pancreatitis with calcifications (Dankovic et al., 2008). It is reported that 40% of diagnoses required surgical confirmation (Uruyama et al., 1995) (Maker et al., 2003). AP ductal system has been the subject of multiple investigations (Cholet et al., 2004). One of the most popular is Lin's (1989), which by means of ERCP described three types of pancreatic ducts in the context of AP. Type 1 associates pancreas divisum and AP; in Type 2, the annular duct arises from the pancreatic duct as a branch; and in Type 3, the tail of the annular duct fuses with the dorsal duct to form the pancreatic duct. Taking this classification into account, the AP reported here corresponds to a Type 2. Most patients with symptomatic AP will require surgical treatment (Alexander, 1970). The operative procedures fall into two categories (Ladd and Madura, 2001). Duodenal bypass (duodenoduodenostomy or duodenojejunostomy) is the procedure of choice for treating both children and adults (Zyromski et al., 2008; Maker et al., 2003). And, on the other hand, resection or division of AP is described. This procedure is no longer used due to its risk of pancreatitis, pancreatic fistula and the incomplete relief of obstruction (Alexander, 1970). Nevertheless, not too long ago Ladd and Madura (2001) pointed out that it was essential to determine whether the AP was complete, partial, intramural or extramural. The authors stated that, if AP consists of a partial ring and the duodenum is not densely oppressed, partial excision on the AP could be performed with good result (Ladd and Madura, 2001). The AP reported here consisted of a complete ring with no macroscopic connection to the duodenal wall. In conclusion, a cadaveric case of AP is presented, as a product of an infrequent embryological anomaly; its presence must not be overlooked by surgeons and radiologists due to its clinical and radiological implications.

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