Portal Annular Pancreas
A Systematic Review of a Clinical Challenge

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Abstract: Portal annular pancreas (PAP) is an asymptomatic congenital pancreatic anomaly, in which portal and/or mesenteric veins are encased by pancreas tissue. The aim of the study was to determine the role of PAP in pancreatic surgery as well as its management and potential complication, specifically, postoperative pancreatic fistula (POPF). On the basis of a case report, the MEDLINE and ISI Web of Science databases were systematically reviewed up to September 2012. All articles describing a case of PAP were considered. In summary, 21 studies with 59 cases were included. The overall prevalence of PAP was 2.4% and the patients’ mean (SD) age was 55.9 (16.2) years. The POPF rate in patients with PAP (12 pancreaticoduodenectomies and 3 distal pancreatectomies) was 46.7% (in accordance with the definition of the International Study Group of Pancreatic Surgery). Portal annular pancreas is a quite unattended pancreatic variant with high prevalence and therefore still remains a clinical challenge to avoid postoperative complications. To decrease the risk for POPF, attentive preoperative diagnostics should also focus on PAP. In pancreaticoduodenectomy, a shift of the resection plane to the pancreas tail should be considered; in extensive pancreatectomy, coverage of the pancreatic remnant by the falciform ligament could be a treatment option.

Key Words: portal annular pancreas, circumportal pancreas, complete pancreatic encasement, pancreas anomaly

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Portal annular pancreas (PAP)1–3 or the so-called circumportal pancreas4–9/complete pancreatic encasement10–12 is an asymptomatic, pancreatic anomaly, in which the uncinate process of the pancreas encircles the portal vein and/or its influx, the superior mesenteric and splenic vein, and extends to the dorsal surface of the pancreas body. During embryogenesis, the pancreas is formed by 2 anlagen originating from the endoderm in the primitive duodenum. The caudal part of the head and the uncinate process of the pancreas are derived from the ventral bud, whereas the cranial part of the head, body, and tail of the pancreas is derived from the dorsal. After the duodenum rotation, the ventral pancreatic primordium moves dorsally below and behind the dorsal. Toward the end of the sixth week, the 2 primordia fuse and the ducts anastomose, forming the main pancreatic duct (MPD). The uncinate remains its own pancreatic duct, the inferior branch of the pancreatic duct. Two different hypotheses are discussed regarding the development of PAP: either it is a malformation of the portal venous system5,9 or the pancreas caused by hypertrophy of the ventral10,11 or dorsal anlagen7 and subsequent fusion to the left of the mesenteric or portal vein. This anatomic variant is well known from porcine pancreas12 but described as rare in human, with a prevalence from 1.1% to 3.4%5,8,13 in computed tomographic (CT) image studies of a healthy population13 or of patients with hepatoma, adrenal or colon adenoma, uterine carcinoma, or pancreatic cyst, respectively.1

Portal annular pancreas is classified depending on the topography of the MPD14 or in accordance with its relation to the portal confluence.1 Joseph et al15 proposed the following: type 1 is the fusion of the ventral bud of the pancreas with the body and retroportal MPD (RMPD); type 2 is type 1 associated with pancreas divisum; and type 3 is the portal vein encasement by the uncinate process with a normal anteportal MPD (AMPD). Following Karasaki et al,1 each type can be subdivided (A, B, and C) depending on the relation to the portal confluence (suprasplenic, infrasplenic, and mixed type; Fig. 1).

On the basis of a case report, we carried out a systematic review of the literature on PAP. The main purpose of this study was to determine the role of PAP in pancreatic surgery as well as its management and potential complication, specifically, postoperative pancreatic fistula (POPF).

MATERIALS AND METHODS

Search Strategy
A systematic literature search was conducted independently by 2 authors (J.M.H. and J.C.H.). The authors searched the National Library of Medicine (MEDLINE [PubMed, available at: http://www.pubmed.com/]) and the ISI Web of Science (available at: http://wokinfo.com) from 1987 to week 1 of October 2012. The primary search strategy was set up using a combination of text words combined with a Medical Subject Headings (MeSH) database search (circumportal [all fields] and “pancreas” [MeSH terms] or “pancreas” [all fields]) or (periportal [all fields] and “pancreas” [MeSH terms] or “pancreas” [all fields])). The reference lists of the retrieved manuscripts were manually cross-searched for additional publications. All types of articles including abstracts, case reports, editorials, letters to the editor, and reviews were considered. No language restrictions were applied.

Eligibility Criteria and Data Collection
All reports on PAP and its management were included, which described at least 1 case with full pancreatic encasement of the.
Two authors (J.M.H. and J.C.H.) screened the search findings for potentially eligible publications. Full-text articles were obtained to clarify potential eligibility and the included studies were evaluated in accordance with the Preferred Reporting Items for Systematic Reviews and Meta-Analysis (PRISMA) statement. Two authors (J.M.H. and J.C.H.) independently assessed the selected studies and extracted data on PAP’s management, surgery, and complications.

CASE REPORT
A 48-year-old white woman presented with 4 weeks of left upper abdominal pain. Computed tomographic scan showed a large tumor mass in the left upper quadrant (Fig. 2). Intraoperatively, the 9 × 12 × 14-cm tumor mass presented retroperitoneally adjacent to the colon and pancreas. Therefore, an en bloc resection of the tumor mass with extensive distal pancreatectomy (DP), splenectomy, left nephroureterectomy, and hemicolectomy was performed. At extensive DP, the pancreas was transected over the portal vein using a stapler. Then, it was found that the uncinate process fused to the body of the pancreas, causing a PAP. The retroportal connection was transected using a stapler, resulting in 2 resection planes. In accordance with the PAP classifications, PAP was graded 3A. Histology revealed a suprarenal cancer. Postoperatively, the patient developed POPF grade B in accordance with the definition by the International Study Group of Pancreatic
Surgery (ISGPS),16 The patient was discharged on postoperative day 13. On 6-week follow-up, the patient presented completely recovered from surgery.

RESULTS

In total, 423 articles were retrieved from the primary database search and 450 articles were retrieved from the citation search using ISI Web of Science. Four additional articles were retrieved by manual cross-search of the reference lists. After removal of duplicates, 667 articles were assessed for further analysis; screening of the titles and abstracts enabled us to exclude 386 articles. The full-text articles of the remaining 281 studies were assessed for eligibility. Overall, 21 articles, including our own case, met the inclusion criteria and reported PAP in 59 patients in total1,4,13 and a published abstract, 8 as well as 16 case reports2,3,5,7,9,11,14,17,19-23 and 1 letter to the editor18 were included. All studies were published within the last 25 years (1987-2012). Three retrospective studies with a total number of patients of 1517 reported the general prevalence of PAP,1,8,13 which was 2.4% (36/1517 patients) overall. Almost all included studies,2,3,5-7,9,11,14,17,19-23 and 1 letter to the editor18 revealed information about patients’ sex and age (n = 42 patients) showing no significant sex but age prevalence (22 females/20 males; mean [SD] age, 55.9 [16.2] years; Table 1).

After the classification of Joseph et al,14 16 studies reported the MPD topography of 27 cases. Most (63%) had AMPD (type 3) and the rest had RMPD (type 1), of which 40% were associated with a pancreas divisum (type 2). According to Karasaki et al,17 studies with 30 cases revealed that PAP was suprasplenic in 70%, infrasplenic in 20%, and with mixed location in the rest, respectively (Table 2). Fourteen studies with 18 cases used both classifications: type 3A, as in our case report, was the most common (44.4%; Fig. 1C) and type 1A was the second most common (27.8%; Fig. 1D, Table 2).

Seventeen of the 59 patients underwent pancreatic resection, mainly for adenocarcinoma of the papilla of Vater2,12,14,21 but also for adenocarcinoma of the pancreas,13,19 insulinoma,12,23 or intraductal papillary mucinous neoplasm.13,19 Single cases were reported for gastric and gallbladder cancer,22 mucinous cyst adenoma,5 as well as chronic pancreatitis.11 In 52.9% of those cases, PAP was missed preoperatively in magnetic resonance imaging or CT scans (false negative) but incidentally discovered intraoperatively. In 15 cases reporting POPF (12 pancreaticoduodenectomies [PD] and 3 DPs), the incidence was 46.7% (2 grade A cases, 3 grade B cases, according to the definition by the ISGPS,16 and 2 cases were not further specified) (Table 1).

DISCUSSION

Portal annular pancreas is an incidental finding in half of the cases, which underwent surgery, suggesting that the prevalence of PAP is still not widely known or considered in preoperative diagnostics. However, PAP cannot only be overlooked (false-negative rate of 52.9%) but can also be misdiagnosed false positive as retroportal tumor mass.6 Therefore, more preoperative attention to this anomaly is required. Computed tomography and magnetic resonance imaging are both suitable to identify PAP3,4,13 (Fig. 2). If PAP is preoperatively identified, magnetic resonance pancreaticography (eventually with stimulation) should be considered to improve the depiction of the pancreatic duct system.

Reviewing a heterogeneous cohort of more than 1500 patients in 3 retrospective studies, the PAP prevalence of 2.4% is unexpectedly high. Although a significantly higher prevalence is reported in women,2 we could not confirm a sex specificity.

Because the literature is mainly based on case reports, the quality of the included studies is heterogeneous. In many of the 59 reported cases, the course of the MPD and the relation to the portal confluence are not indicated. Therefore, the present study is limited by the small number of 36 cases that were includable for the general prevalence of PAP as well as 27, 30, and 18 cases, respectively, which were includable for the description of the topography (Table 2).

Nevertheless, PAP is highly relevant in pancreatic surgery for the substantially increased risk for POPF associated with increased morbidity and mortality16, in PD, it is because of a small aberrant pancreatic fusion and pancreatic duct, which are not discovered intraoperatively; and in extensive DP, it is because of a large anteropetal and retroportal resection plane. A recent systematic review shows an overall incidence of POPF (in accordance with the ISGPS definition) after pancreatic resections of up to 28.6%.24

Because of the substantial risk for POPF secondary to pancreatic resections in PAP, a modification of PD and DP is suggested. Although, besides our own, only 2 other studies report DP in PAP (making the data less robust), it should be considered in extensive or subtotal DP with pancreatic resection at the level of the portal vein and superior mesenteric vessels.

After PD, the pancreaticojejunostomy in types 1 and 2 is technically difficult because of the retroportal anastomosis; in type 3, ligation of a retroportal branch duct of the uncinate process is possible5,14 but the 2 planes of resection will remain. In the suprasplenic and infrasplenic type, an additional resection is required to liberate portal or supr mesenteric vein, respectively.
In the mixed type, even 2 more resections are needed. However, dissecting the uncinate process results in 2 planes of resection; the retroportal includes the MPD (types 1 and 2) or a branch duct of the uncinate process and the anteportal includes the MPD (type 3) or the accessory pancreatic duct (type 2). Furthermore, in suprasplenic type, the lingual projection may include the celiac artery, common hepatic artery, and splenic artery and may complicate resection. Because the mean (SD) horizontal length of the aberrant pancreatic tissue ranges from 9.4 (3.3) mm to 10 (4) mm in CT images, a shift of the resection plane approximately 10 mm to the pancreatic tail seems to be suitable to get 1 resection plane and to decrease the risk for POPF. However, the residual pancreatic endocrine and exocrine function of the pancreas needs to be considered.

After DP, pancreatic stump leak remains a common surgical complication independent of the closure technique. The use of omentum or falciform ligament patch for coverage of the pancreatic remnant still remains controversial. Recently, a decrease in the leakage rate was reported, whereas the results of Tani et al refute this.

### TABLE 1. Review of the Literature

| Reference* | n | Age, y/Sex (M/F) | Classification by Joseph et al | Classification by Karasaki et al | Surgery | POPF/Grade^2 |
|------------|---|-----------------|-------------------------------|---------------------------------|--------|-------------|
| Sugiura et al^11 | 1 | 51/F | 3 | B | PD | No |
| Hamanaka et al^12 | 1 | 59/M | 3 | C | PD | — |
| Mizuma et al^22 | 1 | 64/M | — | — | PD | Yes |
| Savastano et al^1 | 1 | 63/M | 3 | A | — | — |
| Yamazaki et al^2 | 1 | 62/F | 2 | B | — | — |
| Marjanovic et al^7 | 1 | 65/F | 3 | A | PD | Yes/B |
| Leyendecker et al^6 | 4 | 78/F | 3 | A | — | — |
| Song et al^8 | 1 | 54/M | 1 | A | — | — |
| Hashimoto et al^5 | 1 | 52/M | 1 | A | — | — |
| Karasaki et al^1 | 9 | 78/M | 3 | A | — | — |
| | | 7 × F | 6 × A, 1 × B, 1 × C | — | — | — |
| | | 1 × M | — | — | — | — |
| Kin and Shapiro^20 | 1 | 55/F | A | — | — | — |
| Ishigami et al^13 | 10 | 31/M | 3 | — | — | — |
| Izuishi et al^18 | 1 | 50/M | 3 | — | — | — |
| Joseph et al^14 | 1 | 51/M | 2 | — | PD | No |
| Kin and Shapiro^10 | 1 | 38/M | A | — | — | — |
| Gonoi et al^4 | 2 | 70/M | 1 | A | — | — |
| Jang et al^19 | 2 | 78/M | 1 | A | — | — |
| | | 71/M | 3 | A | PD | Yes/B |
| | | 74/F | C | DP | Yes/A |
| Matsumoto et al^2 | 1 | 81/F | 2 | B | PD | No |
| Muto et al^23 | 1 | 45/F | 2 | — | PD | No |
| Kobayashi et al^21 | 1 | 61/F | 3 | A | PD | No |
| Harnoss et al (this study) | 1 | 48/F | 3 | A | DP | Yes/B |

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*Year of publication.
†Number of reported cases/patients.
‡Postoperative pancreatic fistula in accordance with the ISGPS definition.
— indicates not reported.
However, regarding the large anterportal and retroportal resection planes after extensive DP in PAP, we suggest this closure technique secondary to extensive DP.

The 2 main findings for the surgical management of PAP are as follows:

1. According to the classifications of Karasaki et al and Joseph et al, the most common type of PAP is type 3A, which is associated with AMPD and suprasplenic fusion (Fig. 1C).

2. Portal annular pancreas seems to be associated with increased risk for POPF after pancreatic resection because of additional resection plains and variable courses of the pancreatic ductal system. An intraoperative pancreatography might be useful for confirmation in selected cases. In PD, a shift of the resection plain to the left and coverage of the pancreatic remnant after extensive DP should be considered.

To our knowledge, this is the first systematic review of PAP and its management. The results indicate the clinical relevance of this pancreatic anomaly, which occurs with high prevalence. Portal annular pancreas is associated with increased risk for POPF and therefore increased morbidity and mortality requiring modification of pancreatic resection. Because of its low degree of familiarity, PAP remains a diagnostic and operative challenge for the radiologist and surgeons, respectively, to avoid preoperative misdiagnosis and postoperative complications.

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