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Portal Annular Pancreas - An Underreported and Unexpected Intraop Catastrophe - Review of Anatomy, Classification and Management

Rohith Mudadla[1], Jeswanth Satyanesan[2], Satish Devakumar K[3], Amudhan A[4], Harshit Kamal[5], Selvaraj T[6]

[1]Resident, Institute of Surgical Gastroenterology and Liver Transplantation, Govt. Stanley Medical College, Chennai, India
Email: *rohithm9696[at]gmail.com*

[2]Professor and Director, Institute of Surgical Gastroenterology and Liver Transplantation, Govt. Stanley Medical College, Chennai, India
Email: *dr_jeswanth[at]yahoo.com*

[3]Assistant Professor, Institute of Surgical Gastroenterology and Liver Transplantation, Govt. Stanley Medical College, Chennai, India
Email: *dr. satishdevakumar[at]gmail.com*

[4]Assistant Professor, Institute of Surgical Gastroenterology and Liver Transplantation, Govt. Stanley Medical College, Chennai, India
Email: *amudhanmch[at]yahoo.co.in*

[5] Resident, Institute of Surgical Gastroenterology and Liver Transplantation, Govt. Stanley Medical College, Chennai, India
Email: doctorharshitkamal[at]gmail.com

[6]Associate Professor, Institute of Surgical Gastroenterology and Liver Transplantation, Govt. Stanley Medical College, Chennai, India

Abstract: ***Portal annular pancreas (PAP) is an anatomic variation due to aberrant fusion of uncinate process of the pancreas which arises from the ventral bud and extends to fuse with thedorsal pancreatic bud by encircling the portal vein or superior mesenteric vein. In this article, we present a case report in a 59 year old male who presented with obstructive jaundice and features of G. O. O and diagnosed as having moderately differentiated adenocarcinoma on endoscopic biopsy from second part of duodenum. He was planned for an elective whipples procedure, during pancreatic transection tuft of pancreatic tissue encircling the portal vein and SMV axis and joining the body of pancreas posteriorly was present. Portal annular pancreatic anomaly was identified intraoperatively. The retroportal tissue was soft in consistency and suture ligated with 3-0 prolene. Post operatively there was no POPF but developed ascites which was resolved spontaneously and discharged on post op day 13.***

Keywords: PAP-portal annular pancreas, G. O. O – Gastric outlet obstruction

1. Introduction

There are three types of pancreatic fusion anomalies:
Annular pancreas, pancreas divisum and portal annular pancreas.

Portal annular pancreas also known as circumportal pancreas is the rarest of these and is mostly asymptomatic with a varying incidence reported in literature ranging from 0.8 to 2.5%.1-4. One of the earliest reports of this fusion anomaly was reported bySuguira et al. in 1987. Portal annular pancreas may develop during aberrant embryogenesis, with the ventral and dorsal pancreatic primordium fusing over the portal vein (PV) /superior mesenteric vein (SMV) due to which a rind of pancreatic parenchyma, encircles the portal vein (PV) or the superior mesenteric vein (SMV) and fuses with the body of the pancreas. It is an asymptomatic condition and is usually an incidental finding on abdominal imaging. This unusual anomaly is underreported in radiological studies due its rarity and can even be misinterpreted or overreported as locally advanced pancreatic cancer with surrounding SMV in a normal anatomical variant. It is important for surgeonsbecause the postoperative pancreatic fistula (POPF) rates following pancreatic surgeries are higher in cases with portal annular pancreas than those with normal pancreas. The reported POPF rates was vs 16.5 % in normal anatomic variant after whipples procedure (2). Pancreatic surgeons and GI radiologists should be aware of this rare anomaly even with low index of suspicion to prevent complications because POPF in pancreaticoduodenectomy is responsible for most of the morbidity and mortality in post operative period. The complexity associated with surgery in PAP is due to the varying MPD anatomy and two pancreatic resection surfaces. Adequate clinical knowledge about this anomaly during preoperative assessment will avoid any intraoperative surprise and catastrophe

2. Case Report

A 59 year old male presented with painless progressive jaundice and pruritis with history of vomiting after food intake, he has loss of appetite and has lost 12 kgs inthe last three months. On examination he was icteric and palpable gall bladder was present. He has earlier consulted a gastroenterologist for the same for which endoscopy was done and showed ulcerative growth in second part of duodenum biopsy was done from the lesion and scope could not be passed beyond. Nasojejunal tube was placed for feeding in view of outlet obstruction features.

He was later imaged with contrast enhanced CT scan which showed 3* 2.8 cm ill defined hypodense mass lesion in Pancreaticoduodenal groove region – possibility of Groove pancreatitis with Infiltration in to medial duodenal wall at level of D1 and D2 causing significant narrowing. The mass partially encases the SMV in the region of uncinate process of pancreas and the MPD diameter was 4mm. He was planned for an elective whipples procedure. Pancreatic neck was transacted and during uncinate dissection tuft of pancreatic tissue was found encircling the portal vein simulating residual malignant tissue in the uncinate process and was an R2 resection. This tissue was extending posteriorly and joining the body of pancreas, but different in consistency with rest of malignant tissue in the head of pancreas. Anomaly of circumportal pancreas was interpreted at this stage of operation and then reviewed with the pre op images (Fig.1) confirming portal annular pancreas with absent duct in the retroportal tissue and a single anteportal main pancreatic duct. This has led to increased intra operative time and blood loss during dissection near PV-SMV junction. Portal vein is hooked with vascular loop rotated to left and the retroportal tissue is dissected from portal venous attachments, there was no duct in this tissue with single MPD identified in the anteportal portion of pancreas. Pancreatic stump was mobilised for about 4cm, the retroportal tissue was suture ligated with 3-0 prolene, by continuing suture technique. Pancreatic reconstruction was done by dunking technique of pancreaticogastrostomy and the annular portion dissected was not included in the anastomosis. Antecolic H-J (hepaticojejunostomy) done and antecolic G-J (gastrojejunostomy) done. Two abdominal drains were placed one in morisson's pouch and other near PG site. Post operatively he had delayed PPH (Post Pancreatectomy haemorrhage) on POD-3 with intraluminal bleed in ryles tube aspirate which was treated conservatively with double dose of I. V Pantoprazole and resolved by post operative day 5. There was increased drain output which was serous in nature 350 ml, 200 ml and 100ml on post operative day one, two and three respectively which was gradually resolved. Drain fluid amylase on Post operative day 3, 5, 7 was 87, 56, 52 IU respectively and within normal limits without any POPF, (post operative pancreatic fistula). He had grade A DGE-delayed gastric emptying, oral liquids were allowed on POD-6and he was discharged on POD-13. HPE was moderate to poorly differentiated pancreatic ductal adenocarcinoma with PDAC and pTNM staging – T3N2Mx (5/8 LN)

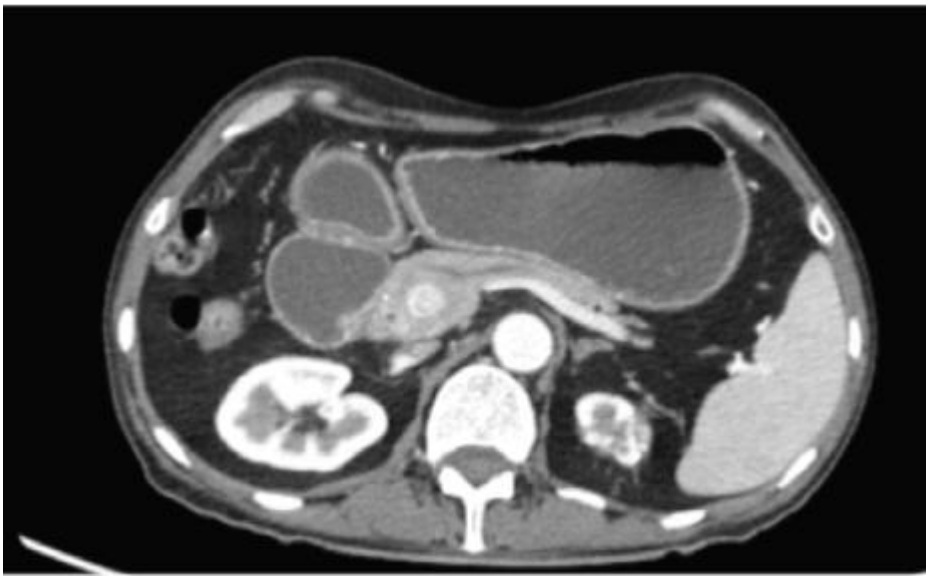


Figure 1: PRE OP-CECT showing MPD ante portal to portal vein with PAP – Type 3A-ANTEPORTAL MAIN PANCREATICDUCT

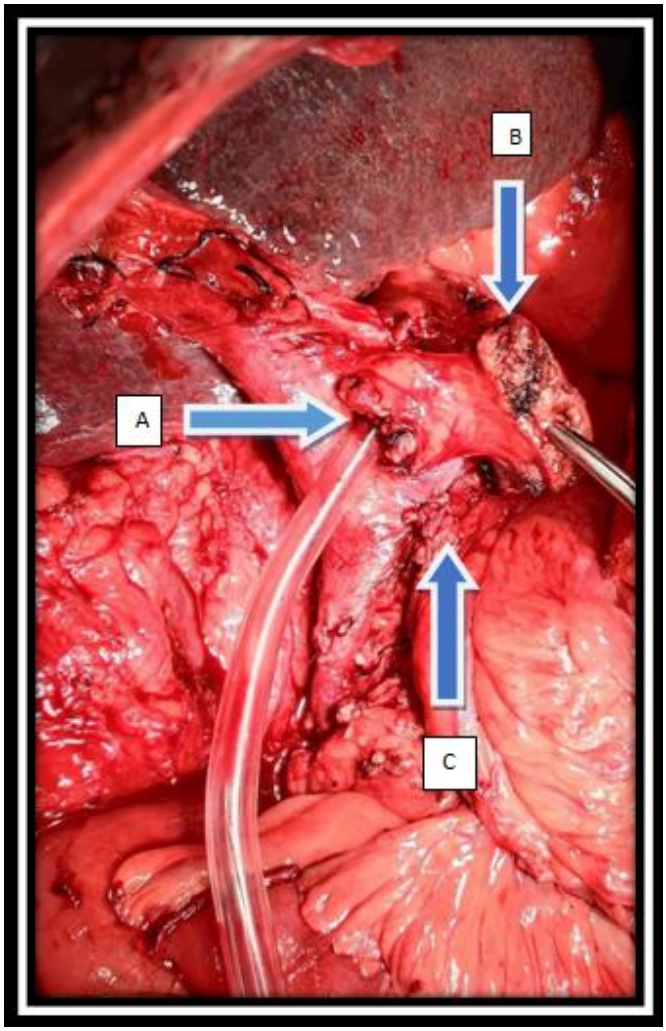


Figure 2: Intra operative image with annular pancreatic tissue above the SMV-SV confluence
a) PAP – portal annular pamcreas Type A (karasakietal)
b) MPD-main pancreatic duct
c) SMV-PV confluence

Embryogenesis of fusion anomalies of pancreas

There are three types of pancreatic fusion anomalies:
1) Annular pancreas-Incomplete and Complete
2) Pancreas divisum
3) Portal annular pancreas.

Portal annular pancreas is the rarest among all fusion anomalies

Prevalence and literature
PAP or circumportal pancreas the possibility of this anomaly during pancreatic head resection was first described by Sugiura et al. Complete fusion of the uncinate process with the body of the pancreas was described by Hamanaka et al. during a resection of the pancreas [1]. Karasakietal. [2] reported the identification rate of portal annular pancreas to be 1.14%. . Yilmaz and Celik [3] has reported prevalence of 0.8 % and Ishigami et al [4] reported as 2.4% among general population. The prevalence of portal annular pancreas in general population was 25 out of 1000 patients i. e 2.5% with a slight female predominance in a retrospective study conducted Rettujohn and Simon etal [4]

They reviewed all the CT abdominal images conducted in a 13 month period most of which are non hepatobiliarypancreatic pathologies there was an increased frequency of this anomaly in females; female: male ratio was 1: 2.1. karasakietal reported prevalence among female to male was 1.3: 1, whereas Harnossetal reported equal prevalence among males and females. This condition is clinically asymptomatic and undetected in most cases or can be incidentally detected when cross sectional imaging like CT /MRI done for some other intraabdominal condition.

Classifications and Subclassification
Joseph etal [3] has classified portal annular pancreas in to three subtypes

Type I-Is the fusion of the ventral bud of the pancreas with the body and retroportal MPD (RMPD)
Type II-when type I is associated with pancreas divisumwith both RMPD+AMPD (ante portal main pancreatic duct)
Type III-when the uncinate process alone is involved in theencasement of the vessels and fusion with only AMPD (ante portal main pancreatic duct)

Each of the type is further divided in to a, b and c (suprasplenic, infra-splenic and mixed) depending on its relationship to the
splenic vein)

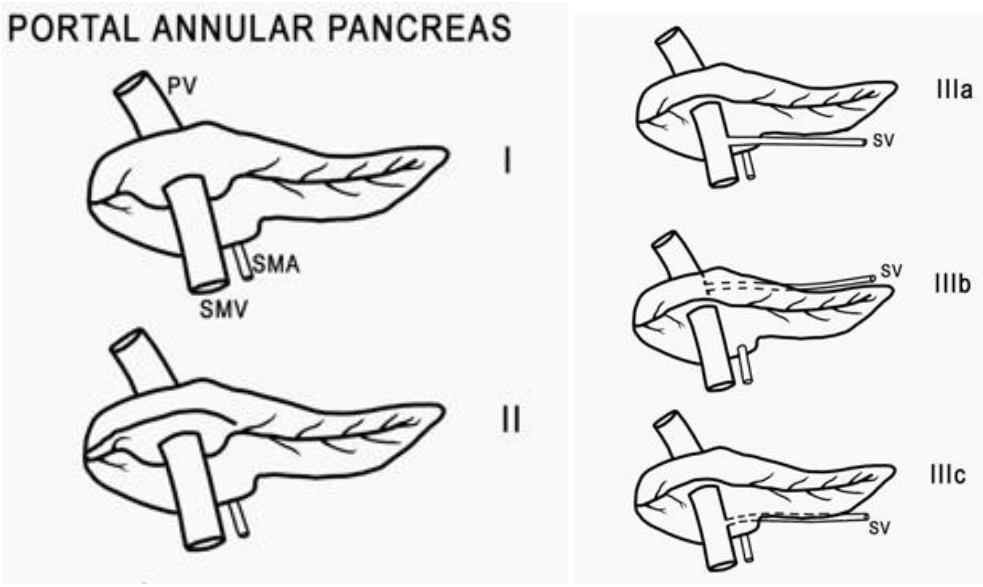
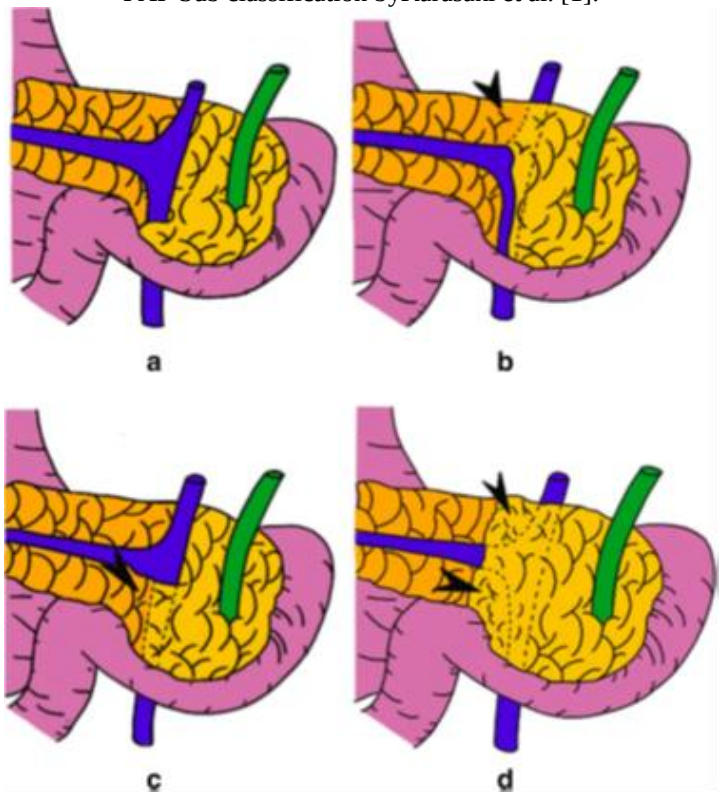


Figure 3: JosephetalSchematic representation of the types of portal annular pancreas

PAP Sub classification byKarasaki et al. [1]:



a-Normal uncinate process
b-Suprasplenic fusion of pancreas around portal vein
c-Infrasplenic fusion
d-Mixed

Review of literature and meta analysis of portal annular pancreas in patients undergoing pancreatic resections by Manish S. Bhandare, Shailesh V. Shrikhande et al. . [7] reviewed 29 articles which were retrospectively studied and total reported cases in literature were53 and of which the type 3Awas the most common variant identified in 25 cases out of 53 reported cases among the different types. Our case in the present study was also type 3A anomaly and there was no POPF. Regarding POPF in PD, POPFs were reported for a mean overall fistula rate of 21.3%. According to a systematic review conducted by Harnoss et al which included 21 studies the pancreatic fistula rate in patients with PAP (12pancreaticoduodenectomies and 3 distal pancreatic tomies being 46.7%as per ISGPS classification. [8] Retrospective metanalysis conducted by Manish S. Bhandare, Shailesh V. Shrikhande et al [8] which included29 studies with 53 patients identified 42.55% rate of POPF and 34% CR-POPF [clinically relevant] which was double the rate of POPF in PD without circumportal pancreas, after pancreaticoduodenectomy with out PAP the reported rate of POPF is highly variable, ranging from 2%-20% [10, 11]

POPF-related mortality rate was 1% after PJ and 0.8% PG after PD in systematic review and analysis of the POPF-related mortality rate in 60, 739 patients retrieved from the English literature published between 1990 and 2015by Sergio Pedrazzoli [9]

If preoperative suspicion of portal annular pancreas during surgery for pancreas all these cases have to undergo MRI with MRCP to delineate proper pancreatic ductal anatomy, the threshold should be at the minimum to undergo an MRCP as pancreaticoduodenectomies or distal pancreatic resections with circumportal pancreas has high POPF rate compared to resections without this anomaly. There are 53 reported cases of portal annular pancreas in pancreatic odueodenectomy or in other pancreatic resections, the case from present study will be 54 th reported case. Because of the rarity of the anomaly there are no large number case series, and most of them are published case reports.

The non-dominantcutting plane, i. e., the plane without MPD can be either sutured or stapled. (12) It has also been suggested that PG withinvagination of the two resected pancreatic planes togetherinto the stomach after PD helps to minimise resected volumeof the pancreas as well as possibly reduce chance of POPF (13)

Portal annular pancreas seems to be associated with increased risk for POPF after pancreatic resection because of additional section 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 should be considered. After PD, pancreaticojejunostomy in types 1 and 2 is technically difficult because of the retroportal anastomosis; intype 3, ligation of a retroportal branch duct of the uncinate process is possible. In the suprasplenic and infrasplenic type, an additional resection is required to liberate portal or supra mesenteric vein, respectively

Preoperative or intraoperative identification of PAP is extremely essential so as to adapt to a different strategy during pancreatic resection as well as stump reconstruction to decrease the devasting complication of POPF

Surgical strategies to minimise the rate of CRPF during PD should involve

1) Extended resections to left of SMV to have single MPD
2) Suture closure of the retroportal tissue
3) Pancreaticogastrostomy or PJ by dunking depending on the type of PAP.

3. Conclusion

PAP is frequently underreported by radiologists. It may also be overreported as locally advanced pancreatic mass which was actually a normal anatomic variant. Precise knowledge regarding this rare anomaly to HPB surgeons is of utmost importance during pancreatic resections in addition to the aberrant or replaced arterial vasculature to avoid intraop surprise

Table 1: Published cases with pancreatic resection in PAP (7)

Sr. no	Author	Patient	Primary tumour	Surgery	PAP type	Non MPD pancreas stump	PJ: PG	POPF
1	Suguiraetal.	51/F	Inflammatoryhead	PD	3/B	Interruptedsutures	PJ	No
2	Hiroshietal.	76/M	MetastaticRCC	Central pancreatectomy	3/A	Proximal: stapled, distal-PJ	PJ	
3	Kawamotoetal.	76/F	PDAC	PD	1/A	Mobilised->PJ[c]	PJ	B
4	Tousifetal.	81/F	IPMN	TP	3/C	Uncinatestapled	-	
5		76/M	Bileduct	PD	3/C	Sutured		No
6	Hamanakaetal.	59/M	Ampullary	PD	3/C	NA	-	NA
7	Hashimotoetal.	39/F	Mucinousneoplasm	DPS	2	Interruptedsutures	-	
8	Balilaetal.	72/M	DuodenalGIST	PD	3/A	NA	PJ	NA
9	Ishigamietal.	45/F	Insulinoma	PD	NA	NA	-	A
10		80/M	IPMN	PD-	NA			A
11		65/M	PDAC	PD	3/A			No
12	Jangetal.	71/M	IPMN	PD	3/A	Stapled	PJ	
13		74/M	IPMN	LapRAMPS	C	NA	-	
14	Josephetal.	51/M	Ampullary	PD	2/A	Interruptedsutures, sideto side PJ	PJ	B
15	Kobayashietal.	61/F	Ampullary	PD	3/A	Interruptedsutures	PJ	A
16	Kuriyamaetal.	47/F	Serouscysticeoplasm	LapDPSP	3/A	Stapled	-	No
17	Izuishietal.	50/M	Bileduct	PD	3/C	Extendedresection	PJ	
18	Marjanovicetal.	65/M	Castomach	Multivisceral	3/A	Stapled	PJ	No
19	Baskaranetal.	47/M	Ampullary	PD	3	Sutures	PJ	B
20	Karasakietal.	73/F	Bileduct	PD	3/C	NA	PG	
21	Zimmittietal.	71	Ampullary	LapPD	3/B	Extendedresection	PG	No
22	Mutoetal.	45/F	Insulinoma	PD	2	Extendedresection	PJ	B
23	Kiuchietal.	78/M	PDAC	PD	3/B	Cautery	PJ	No
24		76/M	Bileduct	PD	3/B	Stapled	PJ	B
25		55/M	Ampullary	PD	3/B	Cautery	PJ	No
26		74/M	Bileduct		3/B	Stapled	PJ	No
27		66/M	Duodenal		3/B	Stapled	PJ	No
28		65/M	IPMN		3/B	Stapled	PJ	BCBBB No
29		79/F	Ampullary		3/B	Stapled	PJ	

30	Matsumotoetal.	81/F	Ampullary	PD	1	Extendedresection		
31	Matsumotoetal.	78/M	Duodenal	PD	3/A	Bothstumps-PG[b]	PG	
32	Pardiwalaetal.	81/F	Duodenal	PD	3/A	Ligated	PJ	
33	Yuan et al.	74/M	PDAC	RAMPS	3/B	NA	-	
34	Shonakaetal.	53/M	PDAC	PD	3/A	Bothstumps-PG[b]	PG	No
35	Zhangetal.	66/M	IPMN	PD	3/C	Continuoussutures	PJ	No
36	Ohtskaetal.	66/M	PDAC	PD	3/A	Stapled	-	No
37		64/M	IPMN	PD	3/A	Extendedresection		No
38		65/F	Bileduct	HPD	3/A	Extendedresection		A
39		63/M	NET	DP	3/A	stapled		No
40		61/F	PDAC	DP-CAR	3/A	Stapled		No
41		76/F	Bileduct	PD	3/A	Stapled		B
42		46/M	Bileduct	PD	3/A	Stapled		No
43		84/F	Bileduct	PD	3/A	Extendedresection		
44		77/F	PDAC	PD	3/A	Extendedresection		No
45	Harnossetal.	48/F	Suprarenal	Multivisceral	3/A	NA		B
46	Luuet al.	81/M	Ampullary	PD	3/A	Extendedresection		No
47		49/F	IPMN	PD	2/A	Extendedresection	PJ	B
48		60/M	Chronicpancreatitis	PD	3/A	Extendedresection	PJ	B
49		65/F	Ampullary	TP	2/A	NA	PJ	No
50		73/F	PDAC	PD	3/A	Extendedresection	-	B
51		55/F	Serouscystadenoma	TP	3/A	NA	PJ	No
52	Naritaetal.	72/F	PDAC	PD	1/A	Both stumps PG	PG	NA
53	Shailesh. v. shrinkandeetal	58/M	PDAC	PD	3/A	PJ	PJ	B
54	Case from Present study	59/M	PDAC	PD	3/A	Interrupted sutures	PG	No

Non MPD pancreas stump—pancreas stump in PAP without the main pancreatic duct and its management
PD- Pancreatico Duodenectomy
PG- Pancreatico Gastrostomy
PJ- Pancreatico Jejunostomy
TP- Total Pancreatectomy
Multivisceral 14-subtotal gastrectomy+ right hemicolectomy+PD,
DP+splenectomy+Lnephrouretrectomy+ hemicolectomy
RAMPS radical antergrade modular pancreaticosplenectomy
HPD hepato-pancreaticoduodnectomy
DP distal pancreatecomy
DPSP distal pancreatectomy spleen preserving
CAR coeliac axis resection
PAP type Joseph/Karasaki
PDAC- Pancreatic ductal adenocarcinoma
IPMN- Intraductal papillary mucinous neoplasm
NET- Neuroendocrine tumour
RCC- Renal cell carcinoma.

Conflict of interest
None of the authors have any conflicts of interest to disclose.

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Consent
Written consent in accordance with the ethics committee has been obtained.

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Author Profile

Rohith Mudadla is Resident, Institute of surgical gastroenterology and Liver transplantation, Govt. Stanley medical college, Chennai, India **rohithm9696[at]gmail.com**

Jeswanth Satynasean is Professor and director **dr_jeswanth[at]yahoo.com,** Institute of surgical gastroenterology and Liver transplantation, Govt. Stanley medical college, Chennai, India

Satish Devakumar K is Assistant Professor, Institute of surgical gastroenterology and Liver transplantation, Govt. Stanley medical college, Chennai, India **dr. satishdevakumar[at]gmail.com**

Harshit Kamal is Resident, Institute of surgical gastroenterology and Liver transplantation, Govt. Stanley medical college, Chennai, India doctorharshitkamal[at]gmail.com

Amudhan A is Assistant professor, Institute of surgical gastroenterology and Liver transplantation, Govt. Stanley medical college, Chennai, India **amudhanmch[at]yahoo. co. in**

Selvaraj. T is Associate professor, Institute of surgical gastroenterology and Liver transplantation, Govt. Stanley medical college, Chennai, India