CASE REPORT

A Case of Intraductal Papillary Mucinous Carcinoma in the Head of the Pancreas Associated With Absence of the Duct of Wirsung

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ABSTRACT

A Seventy-one-year-old woman received CT scan for a checkup of elevated serum amylase. It revealed a solid tumor associated with cystic portion in the head of the pancreas. The duct of Wirsung was not detected by endoscopic retrograde cholangio-pancreatography form Vater's papilla. The cystic lesion and dilated pancreatic duct of the body of the pancreas was detected by pancreatography form the accessory papilla. We performed pancreaticoduodenectomy and pathological diagnosis was intraductal papillary mucinous carcinoma. Fluorography from Vater's papilla of the resected specimen revealed the absence of duct of Wirsung connecting to the Vater's papilla. Fluorography from main pancreatic duct in the stump of the resected specimen revealed communication between the cystic portion of the tumor and the duct of Santorini. Histopathological study with serial section also revealed absence of duct of Wirsung connecting to Vater's papilla. However, the posterior area of the head of the pancreas was positive for anti-pancreatic polypeptide antibody that is a specific marker for ventral pancreas on immuno-histopathological analysis, which explained the presence of ventral pancreas. Comparing the resected specimen with preoperative pancreatography, we confirmed that the pancreatic duct existed in the ventral pancreas connecting to the accessory papilla. The pathophysiology of absence of duct of Wirsung may not a hypoplasia or deficiency of ventral pancreas itself, but a congenital anomalous connection of the duct of ventral pancreas.

INTRODUCTION

Pancreas divisum is a most common variation of pancreatic ductal configuration during organogenesis. Its pathophysiology is complete or incomplete deficiency of communication between dorsal and ventral pancreatic ducts, and some patients show the absence of the ventral pancreatic duct of Wirsung (WD) [1, 2]. Pancreas divisum is frequently associated with pancreatitis because of outflow block of pancreatic juice into duodenum through a relatively small orifice of the accessory papilla [3]. Recently, pancreas divisum is thought to have significant correlation with prevalence of pancreatic cancer [4, 5, 6, 7] and possibly with intraductal papillary mucinous neoplasm (IPMN) of the pancreas [8, 9, 10]. We herein report a case of intraductal papillary mucinous carcinoma (IPMC) associated with absence of WD. We examined the resected specimen with careful attention to the existence of ventral pancreas, and considered about the essential pathophysiology of absence of WD.

Received January 03rd, 2017 - Accepted January 28th, 2018
Keywords Pancreas; Pancreatic Ducts
Abbreviations IPMN intraductal papillary mucinous neoplasm; MRCP magnetic resonance cholangiopancreatography; SD duct of Santorini; SLE systemic lupus erythematosus; WD duct of Wirsung
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CASE REPORT

A seventy-one-year-old woman had received medical treatment for systemic lupus erythematosus (SLE). On routine blood examination during the treatment for SLE, elevated serum amylase was pointed out. CT scan revealed a well enhanced solid tumor with central cystic portion in the head of the pancreas and dilated pancreatic duct of distal side from the tumor **(Figure 1)**. Magnetic resonance cholangiopancreatography (MRCP) detected cystic portion of the tumor in the head of the pancreas and obvious dilatation of pancreatic duct of distal side from the tumor **(Figure 2)**. WD was not detected by endoscopic retrograde cholangiopancreatography (ERCP) form Vater's papilla **(Figure 3a, b)**. The orifice of the accessory papilla was slightly dilated by projection of mucin **(Figure 3c)**. The cystic portion of the tumor in the head of the pancreas and dilated pancreatic duct of distal side from the tumor was detected by pancreatography form the accessory papilla **(Figure 3d)**. We obtained pathological finding of adenocarcinoma by endoscopic ultrasonography (EUS) guided fine needle aspiration biopsy. We diagnosed the tumor as IPMC, preoperatively. We performed pylorus-preserving pancreaticoduodenectomy. Pathological diagnosis for the tumor was IPMC derived from the duct of Santorini (SD). Fluorography from Vater's papilla of the resected specimen revealed the absence of WD connecting to the Vater's papilla **(Figure 4a)**.

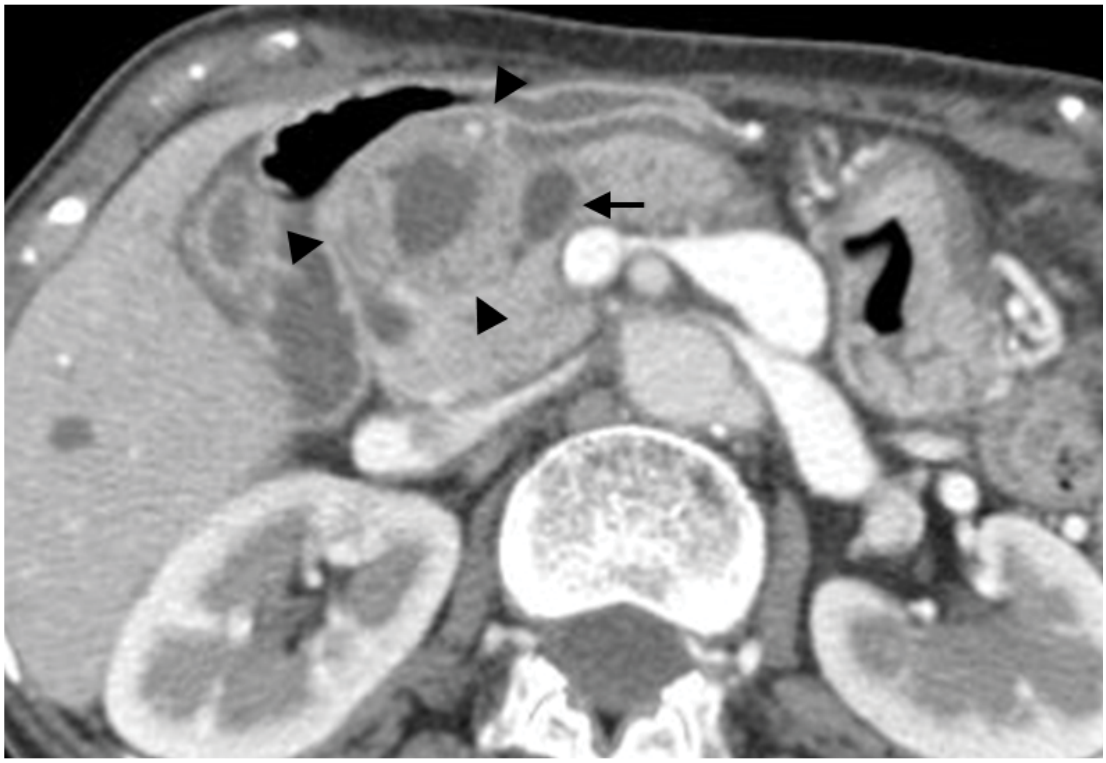

Figure 1. CT scan revealed a well enhanced solid tumor with central cystic portion (arrowhead) in the head of the pancreas and dilated pancreatic duct of distal side from the tumor (arrow).

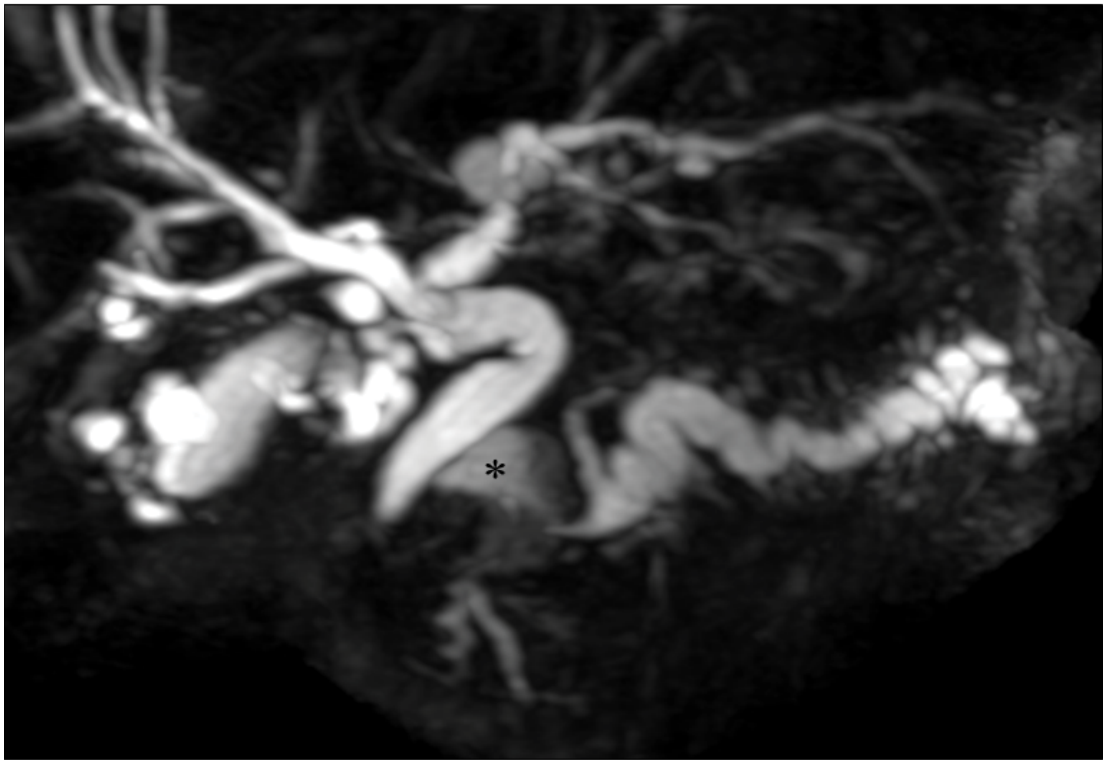

Figure 2. Magnetic resonance cholangiopancreatography detected cystic portion of the tumor (*) in the head of the pancreas and obvious dilatation of pancreatic duct of distal side from the tumor.

Fluorography from main pancreatic duct in the stump of the resected specimen revealed communication between the cystic portion of the tumor and SD **(Figure 4b)**. Histopathological study with serial section also revealed absence of WD connecting to Vater's papilla. However, the posterior area of the head of the pancreas was positive for anti-pancreatic polypeptide antibody that is a specific marker for ventral pancreas on immuno-histopathological analysis, which explained the presence of ventral pancreas **(Figure 5a, b)**.

DISCUSSION

Pancreas divisum is a common congenital anomaly of the pancreas, which results from an abnormal fusion between ventral and dorsal pancreatic ducts during fetal development. The concept of this disorder was

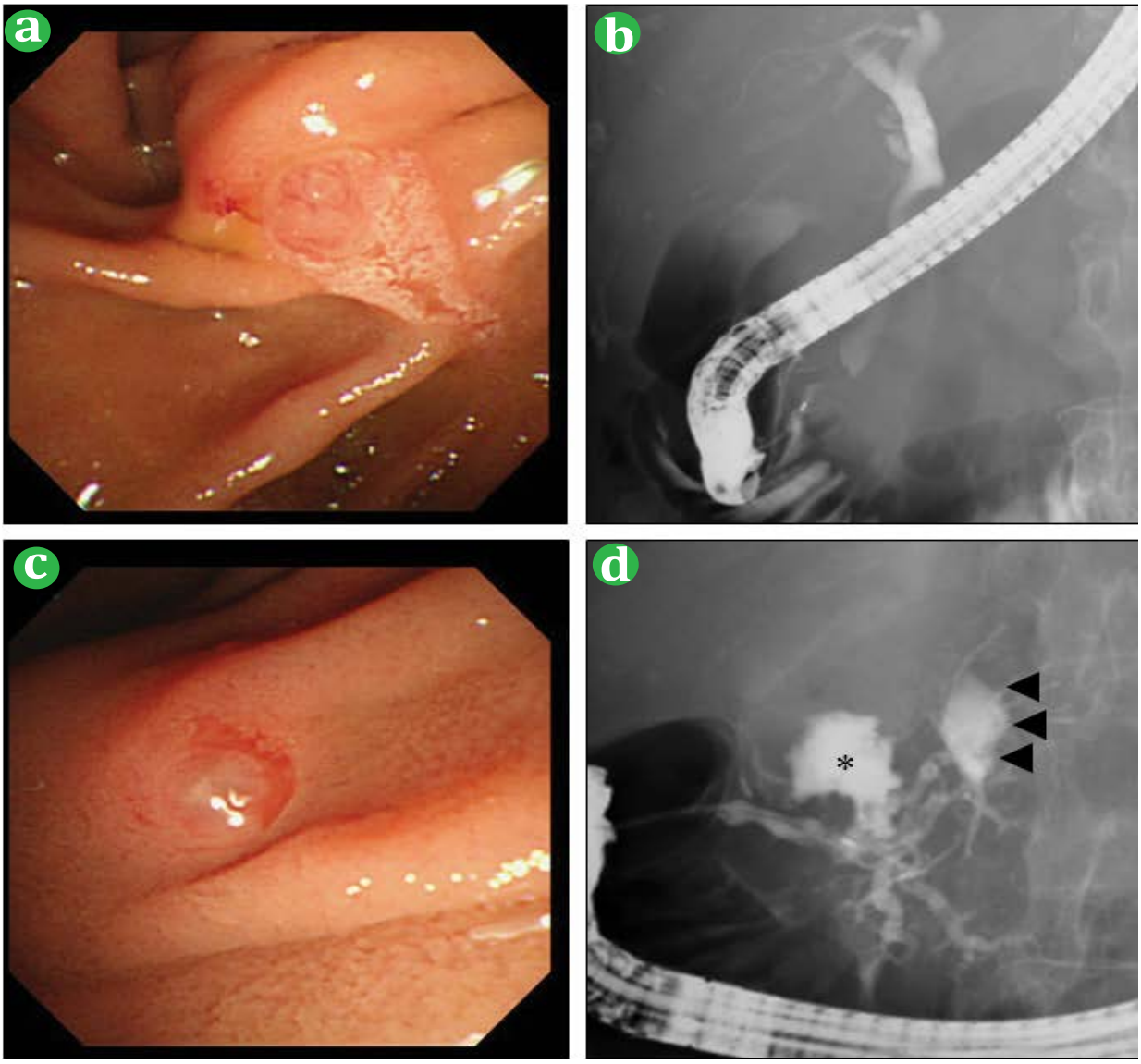

Figure 3. (a). Endoscopic view of Vater's papilla showed no abnormality. **(b).** The duct of Wirsung was not detected by endoscopic retrograde cholangiopancreatography form Vater's papilla. **(c).** The orifice of the accessory papilla was slightly dilated by projection of mucin. **(d).** The cystic portion of the tumor (*) in the head of the pancreas and dilated pancreatic duct of distal side from the tumor (arrowhead) was detected by pancreatography form the accessory papilla.

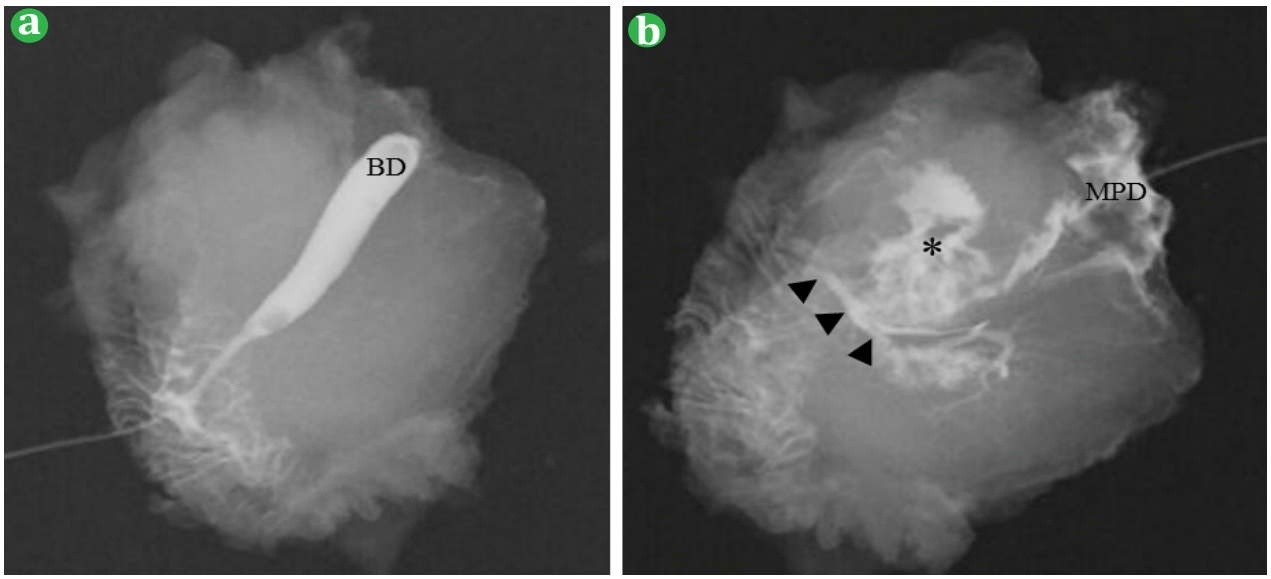

Figure 4. (a). Fluorography from Vater's papilla of the resected specimen revealed the absence of the duct of Wirsung connecting to the Vater's papilla. **(b).** Fluorography from main pancreatic duct in the stump of the resected specimen revealed communication between the cystic portion of tumor (*) and the duct of Santorini (arrowhead).

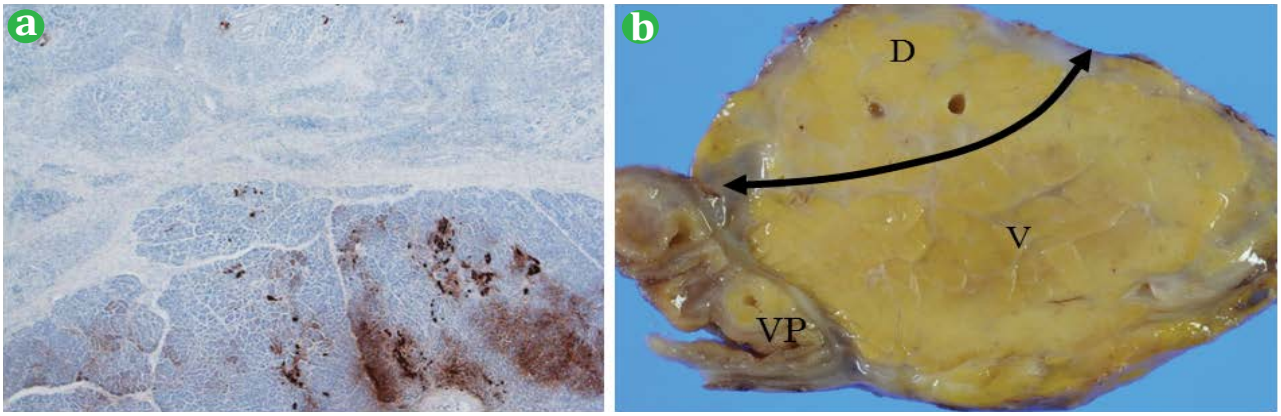

Figure 5. (a). The posterior area of the head of the pancreas was positive for anti-pancreatic polypeptide antibody which explained the presence of ventral pancreas. **(b).** Supposed borderline between dorsal and ventral pancreas by the result of immuno-histopathological analysis was drawn on the photography of the resected specimen. D dorsal pancreas; V ventral pancreas; VP Vater's papilla

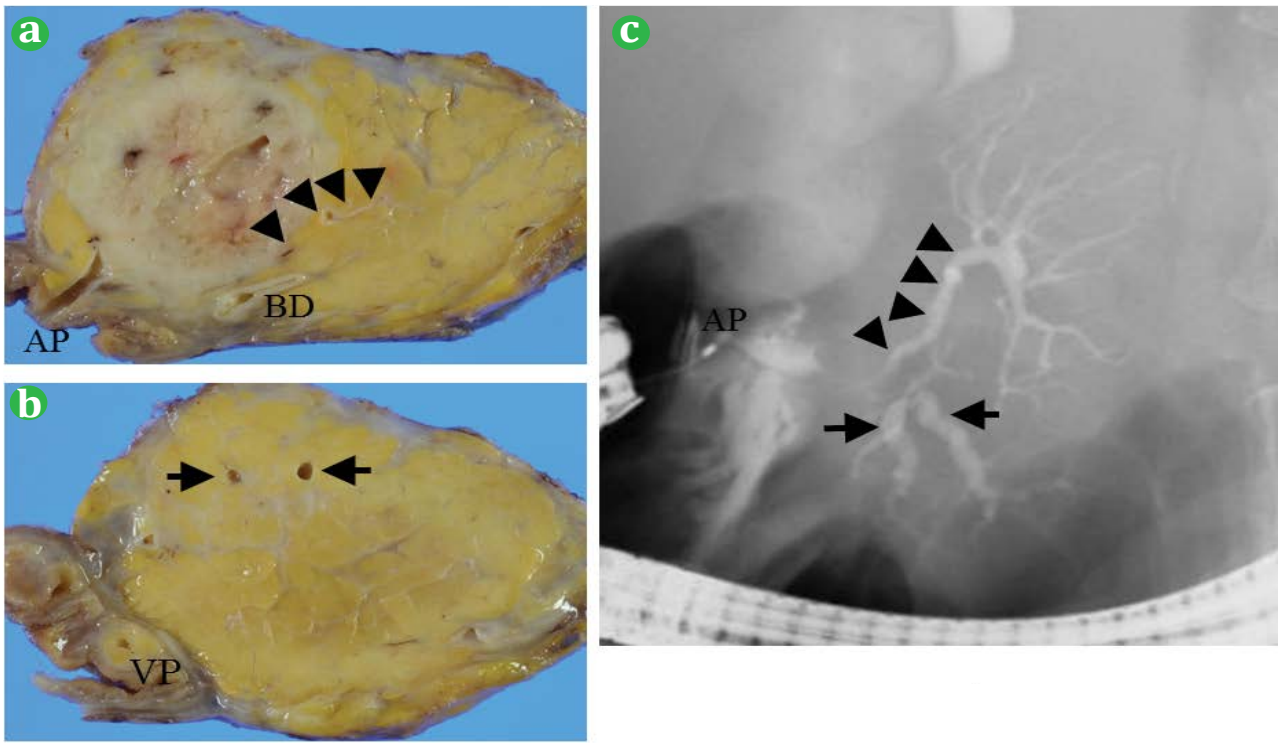

Figure 6. Comparing the resected specimen **(a, b).** with preoperative pancreatography, **(c).** we confirmed that the pancreatic duct existed in the ventral pancreas connecting to the accessory papilla.

Arrowhead: pancreatic duct in ventral pancreas connecting to the accessory papilla

Arrow: dilated pancreatic duct of descending branch in dorsal pancreas

AP accessory papilla; BD bile duct; VP Vater's papilla

redefined as dominant dorsal duct syndrome (DDDS) and its variation was classified into 3 types by Warshaw in 1990 [11]. According to this classification, type 1 is defined as separation of dorsal and ventral pancreatic ducts, and corresponded to classical pancreas divisum. Type 2 is defined as absence of WD, and type 3 is filamentous communication between dorsal and ventral pancreatic ducts. Among these variations, our case applied to type 2. In this situation, whether the ventral pancreas itself is absent as well as WD has not been clarified.

Ventral pancreas can be distinguished from dorsal pancreas by immunohistochemical technique because islets of ventral pancreas contain a large amount of pancreatic polypeptide compared with those of dorsal pancreas [12, 13]. In our case, by the immunohistochemical study with anti-pancreatic polypeptide antibody, we confirmed that ventral pancreas existed in the posterior area of the head of the pancreas in spite of absence of WD connecting to Vater's papilla.

Comparing the resected specimen with preoperative pancreatography, we confirmed that the pancreatic duct existed in the ventral pancreas connecting to the accessory papilla **(Figure 6a-c)**.

CONCLUSION

In other words, although WD connecting to Vater's papilla was absent, there was a ventral pancreas whose duct connecting to the accessory papilla. The pathophysiology of DDDS of Warshaw type 2 may not a hypoplasia or deficiency

of ventral pancreas itself, but a congenital anomalous connection of the duct of ventral pancreas.

Conflict of Interest

Authors are declared that there is no conflict of interest.

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