

Disconnected Pancreatic Duct Syndrome: Management Outcome Analysis and Literature Review

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Abstract: ***Background and objectives:** Disconnected pancreatic duct syndrome is a known complication post necrosectomy and is easily missed. There is little literature available to guide its management. This study aimed to investigate the demographics, etiology, clinical features, radiological features, and outcome of endoscopic and surgical management of patients with disconnected duct syndrome following surgery for severe acute pancreatitis and to compare it with management outcomes across the globe. **Method:** A retrospective analysis of patients diagnosed with disconnected duct syndrome following necrosectomy between 2008 and 2013 was conducted. **Results:** A total of 18 patients with DPDS were identified. The median patient age of presentation was 27 years, and 94% of the patients were men. The most common etiology was acute necrotizing pancreatitis. The median interval between diagnosis of acute necrotizing pancreatitis and surgery was two months (range 0 - 7 months). About 45% of patients with severe acute pancreatitis developed DPDS. Nine patients had an initial failed endoscopic intervention and required distal pancreatectomy. Only one patient underwent a successful endoscopy. The remaining 17 patients underwent surgery. The mortality after surgery was 0%, and 30% of the patients developed diabetes mellitus. **Discussion:** Disconnected pancreatic duct syndrome should be considered if external pancreatic fistula persists beyond two months. Endoscopic treatment has a high failure rate, and surgery is almost always required to treat disconnected pancreatic duct syndrome.*

Keywords: Disconnected pancreatic duct, ERCP, Fistulojejunostomy, Necrosectomy, Necrotising pancreatitis

1. Background and Objectives

Disconnected pancreatic duct syndrome is characterized by complete disruption of part of the pancreatic duct with evidence of viable pancreatic tissue distal to the cut-off [1,2]. DPDS most commonly occurs in the pancreatic neck or body following an episode of acute pancreatitis when there is necrosis of a portion of the pancreas or following pancreatic debridement. In addition to pancreatic fistula, DPDS can present as intrapancreatic and peripancreatic fluid collections and lead to chronic pancreatitis and diabetes mellitus [3, 4].

DPDS is diagnosed based on ERCP evidence of complete cut-off of the main pancreatic duct with an inability to cannulate the upstream duct. An accurate preoperative diagnosis of DPDS requires cross-sectional imaging (CT/MRI) and pancreatography. Treatment of DPDS involves both endoscopic and surgical intervention. As there is no clear consensus on the management of DPDS, this study aims to review our management outcomes and compare them with the literature published so far.

2. Material & Methods

Study design: The study is a single-center retrospective review of patients with DPDS conducted at a high-volume tertiary hospital. All patients admitted with acute necrotizing pancreatitis who underwent necrosectomy between 2008-2013 were reviewed. This study analyzed the demographics, etiology, clinical features, and outcome of endoscopic and surgical management of patients who developed DPDS following necrosectomy.

Inclusion criteria:

Patients following necrosectomy performed for acute necrotising pancreatitis -

- 1) Demonstrating complete cut off the main pancreatic duct on ERCP with an inability to cannulate the upstream duct.
- 2) With viable pancreatic tissue upstream on CECT/MRCP.
- 3) And/or having persistent pancreatic fistula, pseudocyst, or fluid collection .
- 4) Or showing necrosis of at least 2 cm of the pancreas on MRCP with viable upstream pancreatic tissue.

Exclusion criteria:

- 1) Spontaneous healing of pancreatic fistula on conservative management.

Data collection: Data were obtained from their discharge summaries and the review of their imaging features. This study does not involve any human or animal subjects.

3. Results

The age group of patients admitted with acute necrotizing pancreatitis ranged from 8 to 50 years with a median of 27 years.

Two hundred ten patients diagnosed with acute necrotizing pancreatitis were admitted during the study period. The most common cause of acute necrotizing pancreatitis was gall stones (61%) followed by blunt abdominal trauma (38.8%). They were initially managed conservatively: percutaneous drainage, octreotide, and fat-free diet. They were followed up conservatively for at least four months. Of the 210 patients, 91 underwent necrosectomy to drain the infected pancreatic necrotic fluid collection.

Out of the 91 patients treated with necrosectomy, 44 developed persistent pancreatic fistula, and 18 ended up with DPDS. DPDS was diagnosed based on CECT and MRCP findings. The median duration of the interval between

necrosectomy and DPDS development was five months with a range of 4-7 months.

CT features suggestive of DPDS were observed in 40%, 30 - 50% had a large intrapancreatic collection, 20% had pancreatic necrosis, 30% had active extravasation from the main pancreatic duct on ERCP. About 45% of patients with severe acute pancreatitis developed DPDS.

Out of 18 patients identified with DPDS, 16 were male, and 2 were females. The majority of them presented with persistent external pancreatic fistula (seven patients); five had a persistent pancreatic collection, four presented with constant pain, and one had pancreatic ascites as a sequela of DPDS.

All 18 were initially managed endoscopically. 1 out of 18 patients was successfully treated with ERCP stenting. Thirteen patients had persistent drainage, two had a recurrent pancreatic fluid collection, and two developed pancreatic stricture. Out of 13 patients with persistent external drainage, 11 had reattempt of ERCP because of anatomy suitable for endoscopic intervention, and the remaining six were subjected to distal pancreatectomy with or without splenectomy. Of these 11 patients, 4 underwent successful cannulation of the upstream pancreas. Two patients were successfully stented. After a follow-up period of 4 - 6 weeks, all 11 patients had persistent symptoms and were therefore referred for definitive surgical intervention. Following figure 1 depicts the algorithmic approach applied to patients diagnosed with DPDS.

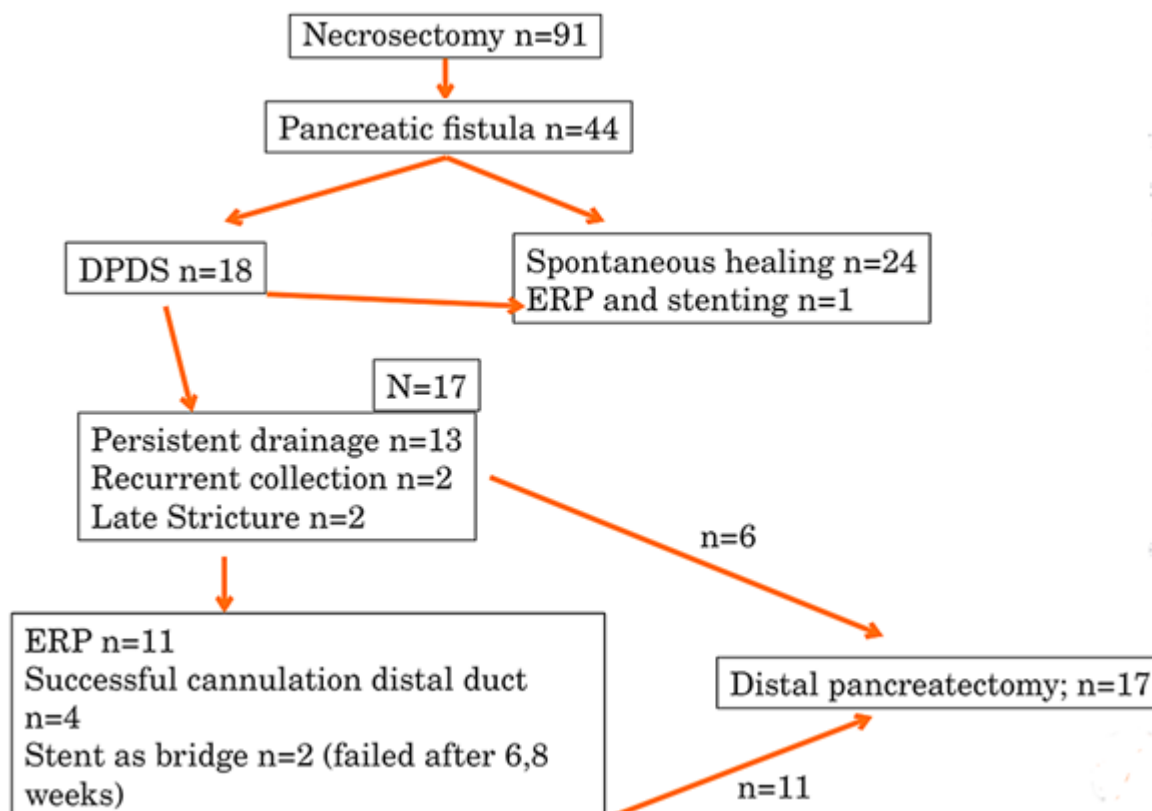


Figure 1: Algorithmic approach to patients diagnosed with DPDS

17 patients underwent distal pancreatectomy with or without splenectomy. They were followed up for 1.5 years following definitive surgery. The mortality after surgery was 0%, and 30% of the patients developed diabetes mellitus. In addition, three patients developed pancreatic fistula, and of them, one of the three needed reoperation for persistent fistula.

4. Discussion

DPDS commonly develops in 50 - 75% of patients following surgical necrosectomy performed secondary to acute pancreatitis or walled-off pancreatic necrosis (WOPN) [5]. The incidence of disconnected pancreatic duct syndrome was 45% among patients admitted with acute necrotizing pancreatitis in this study.

In a similar study conducted by Howard et al., 70% presented with EPF, and 30% had an intra-abdominal fluid

collection [6]. In this study, 44.44% of patients diagnosed with DPDS showed pancreatic fistula, 33.33% had a persistent pancreatic fluid collection, and 22.22% had constant pain.

Early identification and diagnosis of this syndrome is the keystone to prevent complications, particularly fistula formation. The diagnosis of DPDS is fundamentally based on contrast-enhanced computed tomography (CECT), endoscopic retrograde cholangiopancreatography (ERCP), and magnetic resonance cholangiopancreatography (MRCP) features. Drain fluid amylase measurement also helps in the diagnosis [7].

Although CECT shows evidence of DPDS as early as within two weeks, ERCP is usually employed to confirm the diagnosis of DPDS. If ERCP fails to demonstrate the upstream pancreatic duct and cannot differentiate between

high-grade stenosis and a disconnected duct MRCP is used. Although the use of MRCP to diagnose DPDS is increasing, it is still not routinely available at many institutions, especially its secretin variant (sMRCP). Secretin-enhanced MRCP has been identified as the gold standard for diagnosing DPDS [8]. In this study, CECT was primarily employed to manage all acute necrotizing pancreatitis, and ERCP with MRCP confirmed the diagnosis of DPDS.

Conservative management for the resolution of DPDS has a high failure rate. CT-guided percutaneous fluid drainage is usually the first-line treatment unless the necrosis is infected, which indicates the need for surgical necrosectomy [9]. These drainage procedures are bound to be complicated by fistula formation and often require endoscopic intervention. Compared to other studies, the usual treatment strategy in this study was ERCP-guided insertion of an indwelling stent between the two disconnected ends of the main pancreatic duct [10].

An ERCP stenting in DPDS is associated with difficulty passing the stent between the two disconnected ends of the pancreatic duct. EUS is used to locate the fluid collection and the duct and place a prosthesis joining the two if ECP fails to cannulate [10]. Only one patient was treated successfully endoscopically in this study. Nine patients failed the endoscopic treatment and underwent surgery. The major limitation was the unavailability of EUS guided rescue procedures at this institute, leading to poor endoscopic success rates.

Surgery is considered to be the definitive solution if endoscopic techniques fail or as the first option. The surgical treatment of DPDS has a success rate of 80%. The two surgical options are Roux-en-Y fistulojejunostomy (FJ) and distal pancreatectomy with or without splenectomy. These surgical procedures are associated with high mortality and morbidity represented by infection, chronic pancreatic insufficiency, necrosis, multiorgan failure, and death [11]. In this study, the mortality after surgery was 0%, and 30% developed diabetes mellitus. Three patients developed pancreatic fistula, and one of them was reoperated.

Previously surgery was the preferred treatment of DPDS. Initially, endoscopic drainage of DPDS yielded low success rates. However, endoscopic management has become increasingly popular nowadays, especially after introducing EUS-guided transmural drainage and stenting [12]. The duration for which the stent is placed has a significant impact on the recurrence rates of PFC. Arvanitakis et al. has suggested that long-term stent placement is associated with lower recurrence rates of PFC [13]. The use of metal stents carries a lesser risk of stent migration and reduces the chance of PFC recurrence [14].

The two main surgical options are bypass - FJ and resection technique - distal pancreatectomy with or without splenectomy. Surgical bypass techniques have the following advantages: less risk of bleeding intraoperatively, transfusion, shorter operating time, lesser risk of postoperative complications (6%), hospital stay, and endocrine and exocrine function preservation with a lower risk of postoperative diabetes [15]. FJ has a success rate of

77%-100%. However, a well-formed fibrous tract is required to perform the FJ. Therefore it is essential to wait for a reasonable amount of time before undertaking the operation [16]. Murage et al. recommend using a duct-to-mucosa bypass as they believe this to be better than FJ, although it is difficult as it needs small pancreatic tissue resection [17].

Pancreatic resection - distal pancreatectomy with or without splenectomy is associated with loss of pancreatic tissue and exocrine and endocrine dysfunction. In addition, the incidence of intraoperative bleeding and morbidity is higher in comparison with the bypass techniques. Howard et al. recommended resection if there is thrombosis of the splenic vein or left-sided portal hypertension when there is suspicion of malignancy and the ductal remnant is small (<6 cm). The success rate of the resection techniques is approximately 75% [18]. In this study, we preferred distal pancreatectomy with or without splenectomy as the majority had left-sided portal hypertension with a small pancreatic remnant.

5. Conclusion

Pancreatic fistula is a common complication after necrosectomy. Disconnected pancreatic duct syndrome should be considered if external pancreatic fistula persists beyond two months. In the majority, the leak is seen at or distal to the neck of the pancreas. With recent technical advancements, endoscopic intervention for DPDS is the main cornerstone of treatment. Surgical intervention is a definite treatment option and has a high success rate.

There are no conflicts of interests.

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