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Original article

Surgery for pancreas divisum

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We present our experience of open surgical treatment in 5 patients with symptomatic pancreas divisum (PD). Choice of therapy was based on allocation of patients to one of five clinical presentation groups: (i) with minor symptoms (no operation); (ii) with recurrent acute pancreatitis or upper abdominal pain (RAP/RUAP) – 3 patients; (iii) with radiological evidence of chronic pancreatitis (CP) – 1 patient; (iv) chronic .pancreatic pain without radiological evidence of chronic pancreatitis (CPP); and (v) other pancreatic complications – 1 patient. This classification helps to decide management and predict possible outcome. Various types of operation were performed as indicated (open surgical accessory sphincteroplasty [2 also had distal pancreatectomy], n = 3; Puestow's operation, n = 1; or Beger's pancreatectomy, n = 1). All patients improved significantly and are now leading normal personal, professional, and social lives. We conclude that, with careful selection of patients and appropriate therapy, the response to surgical treatment is good.

Key words: Pancreas divisum – Surgery – Minor sphincteroplasty – Beger's operation – Puestow's procedure

Pancreas divisum (PD) is the commonest congenital anomaly of the pancreas with an incidence of up to 10%, but only about 5% of these individuals develop symptoms.[1] The presence of another factor in addition to PD is probably required for symptoms to occur. The symptoms are related to the nature of the pancreatic duct abnormality and to the secondary effects on the pancreas.

There are five main clinical presentations of PD: (i) minor symptoms; (ii) recurrent acute pancreatitis or upper abdominal pain (RAP/RUAP)' (iii) chronic pancreatitis with radiological evidence (CP); (iv) chronic pancreatic pain without radiological evidence of CP (CPP); or (v) with other pancreatic complications. This clinical classification helps to decide management and predict possible outcome.

We present our limited experience of open surgical treatment for symptomatic PD with good results.

Patients and Methods

Between October 1989 and July 1997, we operated upon 5 patients with incapacitating symptoms due to PD. Clinical, radiological and operative details of the patients were reviewed. Follow-up details were critically analysed to assess outcome after operation. Data recorded included the number of attacks of pain, age of onset of symptoms, time from start of symptoms to diagnosis, time from diagnosis to operation, associated and past medical conditions, alcohol consumption, use of analgesics, pancreatic enzymes or other drugs. All patients had routine haematological (full blood count and coagulation profile) and biochemical (renal and liver function tests) investigations. Patients were categorised into five main clinical presentation groups:

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Group 1 – with minor symptoms

Group 2 – with recurrent upper abdominal pain (RUAP) requiring admission to hospital or administration of opioid analgesics, with or without evidence of recurrent acute pancreatitis (RAP; elevated serum amylase concentration in association with abdominal pain)

Group 3 – with radiological evidence of chronic pancreatitis (CP)

Group 4 – with pain suggestive of chronic pancreatitis without radiological evidence of chronic pancreatitis (CPP)[2]

Group 5 – with other complications (*e.g.* pseudocyst, calculi, haemorrhage and abscess)

All patients had abdominal ultrasonography (US) either previously or during admission for operation to rule out any other pathology. All patients had endoscopic retrograde cholangiopancreaticography (ERCP), always trying to examine both major and minor pancreatic papillae and ducts. Computed tomography (CT) scan of the abdomen was performed in 3 patients in whom US and ERCP were not able to exclude other pancreatic pathology.

The decision to operate was based on clinical criteria, which included severity of pain, recurrent attacks of acute pancreatitis and development of complications.

Five patients underwent operation of various types (Table 1). Pancreaticojejunostomy was performed if the pancreatic duct was grossly dilated (> 8 mm); enlargement of the pancreatic head by an inflammatory mass was dealt with by resection of the head of the pancreas with preservation of the duodenum (Beger's operation); other patients were treated by accessory sphincteroplasty. If any specimen was resected, it was sent for histological examination. Operative details and postoperative complications were recorded. All patients were followed up in the surgeon's (CDJ) out-patient clinic for varying periods.

All data and proformas were carefully assessed and analysed for the result of the operation and effect on personal, social, and professional lives of the patients.

Results

Between October 1989 and July 1997, we operated upon 5 patients with incapacitating symptoms due to PD. There was 1 man and 4 women aged 24–49 years (mean, 34 years). All patients had long-standing upper abdominal pain associated with vomiting during attacks of pain. Number of

Table 1 Summary of cases

	Case 1	Case 2	Case 3	Case 4	Case 5
Numbers of attacks pain or AP	4–5	4	4–5	18–20	Almost constant
Duration symptoms before diagnosis (months)	4	6	12	3	12
Time from diagnosis to treatment (months)	1	36	3	52	< 1
US finding	Pseudocyst	NAD	NAD	Dilated accessory duct	Bulky head pancreas
CT finding	Pseudocyst	NAD	NAD	Dilated accessory duct	Chronic pancreatitic mass head pancreas
ERCP finding	PD	PD	PD	PD, dilated accessory duct	PD. stricture lower CBD
Presentation type (group 1–5)	Group 5 (pseudocyst)	Group 2 (RAP/RUAP)	Group 2 (RAP/RUAP)	Group 2 (RAP/RUAP)	Group 3 (CP)
Previous relevant treatment	–	Endo. minor . papillotomy, Lap. chole	–	–	–
Operation	Open surgical, minor sphincteroplasty + distal pancreatectomy	Open surgical, minor sphincteroplasty	Major & minor sphincteroplasty + distal pancreatectomy	Puestow's operation	Beger's pancreatectomy
Histology if available	Pseudocyst and chronic pancreatitis	–	Normal pancreas	–	Chronic pancreatitis

In cases 1 and 3, distal pancreatectomy was added to minor sphincteroplasty in order to cannulate the pancreatic duct for intra-operative pancreatography[27] and to obtain material for histology.

attacks of pain, clinical presentation group, time to diagnosis, time from diagnosis to operation, radiological and endoscopic findings, type of operation and histology (if available) are shown in Table 1.

No patient had significant associated disease or past medical history. Only 1 patient was a smoker and 3 (60%) patients consumed alcohol in insignificant (< 20 units/week) quantity. Two patients were taking pancreatic enzymes, without being confirmed as having pancreatic exocrine insufficiency by any laboratory investigation. One of these patients had a normal *p*-amino-benzoic acid (PABA) test pre-operatively.

Three patients required regular analgesics (non-steroidal and/or opioids) while others required analgesics during the attacks of pain or pancreatitis. None had a family history of PD.

One patient with pseudocyst of the tail of the pancreas had raised amylase (5 times the normal), All patients had normal renal functions. One patient had deranged liver function (raised bilirubin, alkaline phosphatase and aspartate transaminase). She did not present clinically with jaundice. She had had a laparoscopic cholecystectomy but still had recurrent attacks of acute pancreatitis.

Our patients had various operations (open surgical accessory sphincteroplasty (2 of them also had distal pancreatectomy) $n = 3$; longitudinal pancreaticojejunostomy, $n = 1$; Beger's pancreatectomy, $n = 1$) depending on the indication (Table 1).

Only one patient, who had Beger's pancreatectomy required blood transfusion (2 units). Patients stayed in the hospital for a median of 10 days (range, 7–16 days). There were no deaths, re-operation or postoperative complications.

All patients were followed up in the surgeon's (CDJ) outpatient clinic for varying period (range, 1–3 years), as deemed necessary and were discharged when they were symptom-free. They were asked to return to the clinic or to inform the surgeon if they developed symptoms or complications again. None has done so.

All patients improved significantly. None developed endocrine or exocrine pancreatic insufficiency following operation. Two patients who were taking pancreatic enzymes pre-operatively, continue to take pancreatic enzymes post-operatively. All patients have now normal or near-normal personal, professional, and social lives. Only one patient requires occasional analgesics (NSAIDs).

Discussion

Most patients with PD are asymptomatic and only a few develop pancreatitis or pancreatic pain syndrome, probably from relative obstruction at the minor papilla,[3] leading to dorsal duct hypertension.[4,5]

In most patients, the presence of another factor in addition to PD is probably required for symptoms to occur. This would explain why patients with a relatively common anomaly (up to 10% of the population) rarely develop significant symptoms or complications (only about 5% of individuals with PD).

In approximately 8 years, we operated on only 5 patients for PD. This supports the suggestion that this relatively common anomaly is rarely of clinical significance.

We had only 1 man, with a female:male ratio of 4:1, consistent with other series which suggest a female preponderance of about 3:1.[6–8]

All 5 patients had a long history of attacks of abdominal pain or pancreatitis associated with vomiting during the attacks. Four patients had recurrent attacks of pancreatitis or upper abdominal pain. Three of them belonged to group 2 and one to group 5, as he had a pseudocyst of the tail of the pancreas. One patient had almost constant pain and was found to have a chronic pancreatitic mass in the head of the pancreas (group 3).

Three of the five patients consumed alcohol, but in insignificant quantities (< 20 units/week). Two of these patients had chronic pancreatitis on histological examination of resected specimen. Alcohol intake may have contributed to symptoms of these patients. There is a suggestion that the anatomy of the minor papilla in PD, with a relatively small orifice, makes the pancreas more prone to injury from alcohol, drugs and during hypersecretion.[9,10]

It is suggested that patients with minimal symptoms (group 1) should receive medical therapy (low fat diet, analgesics, pancreatic enzymes, anticholinergics) at first. This therapy does not deal with the underlying pathology or anomaly.[11] However, only those with symptoms that disturb their life-style should be considered for operation.

ERCP is diagnostic, as in all of our patients. Incomplete visualisation of the main pancreatic duct after major papilla cannulation should raise the suspicion of PD. In these cases, the minor papilla should be cannulated and the dorsal duct should be properly evaluated. It is important that ventral ductography is normal before accepting PD as the cause of pancreatic pain or disease. ERCP and a satisfactory US may be sufficient to exclude complications affecting the pancreas, such as pseudocyst or pancreatic duct dilatation. For this reason, CT was not required in 2 of our 5 patients.

Attacks of pancreatitis are generally mild.[12] Other complications like pseudocyst,[13] calculi,[14] abscess,[15] or haemosuccus pancreaticus[16] occur occasionally. If complications occur, it is generally an indication for surgical intervention and is essentially treated as that complication would be treated normally. Some form of minor papilla therapy is advisable in these cases.

Repeated mild attacks of acute pancreatitis may lead to chronic pancreatitic mass in the head of the gland, drained

by the accessory duct. This has been confirmed by histological,[17] and endoscopic studies.[3] Two out of five patients have developed chronic pancreatitis as suggested by histological examination.

We have found it useful to categorise patients into 5 groups depending on their clinical presentation, In addition to the three main groups of symptomatic patients,[18] recurrent acute pancreatitis or upper abdominal pain (group 2); with radiological evidence of chronic pancreatitis (group 3); chronic pancreatic pain without evidence of chronic pancreatitis (group 4), we recognise those with minor/mild symptoms (group 1) and those with other complications, *e.g.* pseudocyst, calculi, abscess or haemorrhage (group 5). This system helps deciding the appropriate management.

For critically stenotic minor papilla, open surgical sphincteroplasty gives the best results.[19–22] Three of our patients had open surgical sphincteroplasty. In the literature, up to 85% of patients with RAP/RUAP (group 2) and 60% of patients with chronic pancreatic pain (groups 3 & 4) improve with open surgical sphincteroplasty. Minor papilla restenosis rate is lower (6–7%) with open surgical techniques[21] than with endoscopic techniques (10–20%).[1,20]

All our patients who underwent open surgical sphincteroplasty had good results.

Indications to other forms of pancreatic surgery are few. An occasional patient with dilated dorsal duct and pain will do well with adequate longitudinal pancreaticojejunostomy (Puestow's operation) using a Roux-en-Y limb of jejunum.[23,24] Occasionally, if chronic pancreatitis is restricted to head of the pancreas, Beger's pancreatectomy may be performed in these patients.[25]

All have resumed normal or near normal personal, professional, and social lives. We conclude that, with careful selection of patient and appropriate operation, the response to surgical therapy is good.

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