Operative Management of Chronic Pancreatitis in Children

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Hypothesis: Chronic pancreatitis in childhood is a rare but potentially debilitating disorder, and failure of conservative therapy with chronic pain medication use is common. We hypothesize that aggressive surgical therapy may hold promise for long-term remission.

Design: Retrospective analysis of data collected prospectively for 12 years.

Setting: Tertiary care children’s hospital.

Patients: Eighteen children (aged 3-13 years, 11 girls) underwent surgical treatment of chronic pancreatitis after 1 to 6 years of various medical therapies (parenteral nutrition, somatostatin, or pain medication). These patients required a mean±SD 6±0 hospitalizations before operation. Pancreatitis was familial in 9 patients, idiopathic in 5, and secondary to trauma and medication use in 2 each. Preoperative endoscopic retrograde cholangiopancreatography showed pancreatic duct dilatation in 7, strictures in 5, ductal stones in 4, and normal findings in 2. The operative therapy consisted of longitudinal pancreaticojejunostomy in 2 children (both children failed pancreaticojejunostomy but improved after undergoing pancreatectomy) and distal pancreatectomy with Roux-en-Y pancreaticojejunostomy in 16 children.

Outcome Measures: Survival, need for rehospitalization or reoperation, and chronic pain medication requirements.

Results: All patients survived. Follow-up ranged from 1 to 15 years. Thirteen (72%) of 18 patients have required no further hospitalizations or medications. Two patients required a second operation to convert their longitudinal pancreaticojejunostomy to distal pancreatectomy, and 3 patients have required 2 to 5 additional hospitalizations for recurrent pancreatitis. Endoscopic retrograde cholangiopancreatography on 5 patients 2 to 4 years postoperatively showed patent distal pancreaticojejunostomy.

Conclusions: This series suggests that distal pancreatectomy and pancreaticojejunostomy are effective treatments for this difficult group of patients, while longitudinal pancreaticojejunostomy was ineffective. Long-term relief of pain and reduced need for rehospitalization are the usual results after this procedure.


Although chronic pancreatitis is unusual in children, it nonetheless can be as debilitating and unremitting as it is in the adult. The causes of pancreatitis in children tend to be more related to heredity, medication use, trauma, or idiopathic origins, as opposed to lifestyle choices (such as alcohol abuse) in the adult. Chronic pancreatitis in children is progressive and poorly responsive to medication in most cases, and addiction to pain medication, excessive number of missed school days, and restriction in activities (such as sports) are common.1 Several reports of the operative therapy of pancreatitis in children have emphasized correction of anatomic defects (pancreas divisum) or have presented small series of decompression of a dilated pancreatic duct, but there are few reports of a relatively large group of children who have been treated with an aggressive resection and diversion approach. This report details the management of 18 children with debilitating chronic pancreatitis for 12 years.

RESULTS

All children survived, with a follow-up of 1 to 15 years (mean, 7½ years). There were no operative complications, and patients were discharged 5 to 11 days postoperatively.

The histologic examination of resected specimens showed combinations of intense fibrosis, inspissated and lami-
PATIENTS AND METHODS

Eighteen children underwent surgical therapy for chronic pancreatitis at Cardinal Glennon Children’s Hospital, St Louis University Health Sciences Center, St Louis, Mo, from January 1986 to December 1998. Their ages at the time of operation ranged from 3 to 13 years (mean, 8 years), and 11 (61%) of the patients were girls. This cohort of patients is from a series of more than 100 children seen during the same period with acute or acute relapsing pancreatitis who were not prone to constant pain of chronic pancreatitis, or whose pancreatitis resolved completely without operative therapy. In addition, no children with pancreatic pseudocyst were included in the present series.

Nine (50%) of the 18 patients had a family history of pancreatitis (unrelated to alcohol abuse, gallstones, or other causes). Five had parents (4 mothers and 1 father) who had had pancreatitis diagnosed in childhood, 2 had an aunt or uncle with a similar history of pancreatitis, and 2 others had grandparents with such a history. There were no instances of siblings with known pancreatitis in this series. Five of the patients were believed to have idiopathic pancreatitis, although 2 of these had had pancreas divisum found on endoscopic retrograde cholangiopancreatography (ERCP). However, as explained herein, the pancreas divisum anatomy was not thought to be a major contributor to the episodes of pancreatitis in these patients. Two patients developed chronic pancreatitis 6 months to 2 years after blunt abdominal injury to the pancreas (both were bicycle handlebar injuries), and the final 2 patients had chronic pancreatitis related to medication use (long-term corticosteroid use for lupus erythematosus, and corticosteroid and aspiraginase used for treatment of lymphoma).

All of the patients in this series had chronic abdominal pain that interfered significantly with their activities almost daily. Only 2 of the 18 children had pain-free periods that exceeded 4 weeks, the remainder having essentially daily upper abdominal pain episodes that frequently radiated to the back. Nausea and anorexia were also common, and 12 (67%) of 18 had lost weight. In most children, the baseline chronic pain was exacerbated by eating.

Episodes of acute “flare-up” of pancreatitis that required hospitalization were common in this group of patients, as well as multiple less-severe episodes treated outside of the hospital. The number of hospitalizations for pancreatitis exacerbations in these children ranged from 4 to 13 (mean, 6), which were documented by physical examination, hyperamylasemia, increased serum lipase, or radiologic studies (ultrasonography and computed tomography).

The various therapies that were used for these patients before referral for surgical treatment included low-fat diets (18 patients), oral pancreatic enzymes (16 patients), parenteral nutrition within the hospital (14 patients) and at home (5 patients), intravenous somatostatin during hospitalization (13 patients) and for 1 to 3 months at home (3 patients), and intravenous and oral narcotic pain medication (18 patients). Eight of the 18 patients were dependent on narcotic use to maintain daily function.

The patients were evaluated by ultrasonography of the pancreas and biliary tree (18 patients), computed tomography (13 patients), and ERCP (18 patients). Endoscopic retrograde cholangiopancreatography for younger children (<14 years) was completed using general anesthesia, while older children tolerated ERCP with conscious sedation. Endoscopic sphincterotomy was performed in 10 children during ERCP, including 2 with pancreas divisum, and was unsuccessful in relieving pain in all instances.

All children were referred for surgical therapy when medical therapy proved to be ineffective, when narcotic medication could not be withdrawn, or when the episodes of pancreatitis and the chronic pain were interfering with daily activities, such as school, sports, and social contact. Five patients had had psychiatric consultation before referral, 3 had undergone attempted drug rehabilitation, and 1 had expressed suicidal ideation.

The operative approach was somewhat dictated by the results of computed tomography and ERCP. Two patients with a greatly dilated pancreatic duct seen on ERCP (Figure 1) initially underwent a longitudinal pancreaticojejunostomy (Puestow procedure), but continued episodes of pancreatitis necessitated reoperation and conversion to distal (distal to the superior mesenteric vessels, 50%-60%) pancreatectomy 6 months and 2 years after the initial operation. Two other patients with pancreas divisum underwent initial duodenotomy and contrast injection of both papillae, which demonstrated normal anatomy. Both of these patients had undergone previous endoscopic sphincterotomy. They then underwent distal pancreatectomy with Roux-en-Y pancreaticojejunostomy. The remaining 14 patients, many of whom had diffuse disease seen on ERCP (Figure 2), underwent distal (50%-90%) pancreatectomy with Roux-en-Y pancreaticojejunostomy (Duvall procedure). Splenectomy was not required in any of these patients. Careful dissection of the pancreas away from the splenic vessels, with preservation of the spleen, was possible in each patient. There were no concomitant biliary tract procedures required in these patients.

The patients were followed up by gastroenterologic and surgical services and assessed for survival, recurrent pain, need for rehospitalizations for pancreatitis episodes or pain medication adjustment, and quality of life, including social or athletic activity modification. Methadone was used as needed to aid in weaning from narcotic medication, and psychiatric consultation was helpful in the rehabilitation postoperatively in several patients.
Phoma has been off chemotherapy for 5 years. Finally, the 2 patients with posttraumatic strictures have had no further episodes of pain.

Three patients have had recurrent episodes of documented pancreatitis (abdominal pain and hyperamylasemia) 2 to 6 years postoperatively. All of these patients have documentation by ERCP of patent pancreaticojejunostomy (Figure 6), and their pancreatitis episodes are easily managed with low-fat diets, oral pancreatic enzymes, short-term intravenous pain medication, and somatostatin. None of the 3 patients are receiving long-term pain medication. The final 2 patients have intermittent abdominal pain, associated with nausea. However, the episodes are short-lived, have not required hospitalization, and have not been documented as episodes of pancreatitis. Both of these patients have also had difficulty withdrawing from oral narcotic medications, despite attempts at drug rehabilitation. All 5 of the patients with recurrent pain episodes had initially been diagnosed as having hereditary pancreatitis.

No patient has developed endocrine or exocrine pancreatic insufficiency as a result of pancreatic resection. However, the 5 patients with suspected or documented recurrent pancreatitis continue to receive oral pancreatic enzymes in an attempt to suppress pancreatic secretion to prevent pancreatitis, and therefore the exact status of their exocrine pancreatic function remains unknown. Follow-up studies for the presence of diabetes (insulin levels and glucose tolerance tests) have not been routinely performed.

**Comment**

Chronic pancreatitis is unusual in children and is much more common in adults, in whom it is usually associated with alcohol abuse. Most cases of acute pancreatitis in children (secondary to trauma, medication use, and biliary tract calculi) are short-lived and rarely progress to a chronic state. However, chronic pancreatitis in children can be as debilitating as it is in the adult and can...
lead to lifestyle modification, missed school, dietary restriction, and pain medication dependence.

Chronic pancreatitis in children can be broadly categorized by the presence or absence of hereditary factors, abnormalities in ductal anatomy, and the presence of associated metabolic disorders, such as cystic fibrosis, hyperparathyroidism, hyperlipidemia, and malnutrition. Regardless of the cause, chronic pancreatitis in children typically has an unrelenting progression that has been notoriously difficult to treat medically. Chronic pancreatitis in children, as in adults, is differentiated from acute relapsing pancreatitis, in which periods without pain or other symptoms occur between acute episodes. The periods between episodes can be as long as 1 to 2 years, and, in general, acute relapsing pancreatitis does not result in serious disability, lifestyle alteration, or chronic medication use. The episodes tend to cease in many patients. In addition, most cases of acute pancreatitis in children, usually related to trauma, infection, or medication use, are self-limited and rarely progress to chronic pancreatitis, unless a specific anatomic defect develops, such as pancreatic duct stenosis, which was present in 2 of our patients after traumatic injury.

Hereditary, or familial, pancreatitis is an autosomal dominant disorder with 80% penetrance. Several kindreds have been identified, and generation “skipping” is not unusual. A careful family history should be obtained for each patient with pancreatitis that cannot be explained by any other cause. Hereditary pancreatitis may be associated with a dilated pancreatic duct that is amenable to drainage procedures. Recently, a mutation in the cationic trypsinogen gene has been reported as a cause of hereditary pancreatitis. It is speculated that this defect results in a failure to inactivate trypsin, allowing autodigestion of the pancreatic gland in these patients.

The various ductal abnormalities that have been reported as causes of chronic pancreatitis in children include traumatic stricture, pancreas divisum, congenital sphincter of Oddi abnormality, choledochal cyst, duodenal duplication, and annular pancreas. Obviously, lesions that obstruct the pancreatic duct or ampulla of Vater can cause episodes of acute pancreatitis, but generally the lesions are found and repaired before a more chronic condition develops. Early ERCP can be helpful in this regard. Similarly, coexisting metabolic disorders that lead to pancreatitis are usually detected and treated, although the episodes of acute and chronic pancreatitis may continue while the primary disorder is being treated.

Among the various ductal abnormalities reported in children, pancreas divisum has received the most attention. Pancreas divisum is found in 10% of the general population and is thought to occur as a result of failure of fusion of the dorsal and ventral anlage of the fetal pancreas. When pancreatitis occurs, it is believed to be due to stenosis of the dorsal pancreatic duct (Santorini duct) draining through the minor papilla. Endoscopic retrograde cholangiopancreatography with papilla sphincterotomy has been proposed as primary therapy, although this procedure has not been universally successful. Our series includes 2 patients with this anatomic variant who failed to improve with ERCP sphincterotomy, but who eventually improved markedly with distal pancreatectomy and pancreaticojejunostomy.

Although somewhat controversial, aggressive operative therapy for chronic pancreatitis in the adult has been shown to be effective in many patients who fail nonoperative treatment, but its application in children has been limited. The choice of operation for chronic pancreatitis in the child, as in the adult, is controversial. Attempts to spare pancreatic tissue by ductal diversion alone, without resection, have produced mixed results. Because chronic pancreatitis tends to be a diffuse glandular disorder, and because the source of chronic pain is

Figure 5. Photomicrograph of resected pancreas showing nerves (arrow) trapped within extensive fibrosis (S100 neuronal, original magnification ×25).

Figure 6. Endoscopic retrograde cholangiopancreatogram performed 3 years after distal pancreatectomy with pancreaticojejunostomy shows patency between the distal pancreatic duct and jejunum (arrow).
probably more related to neuropathy rather than ductal pressure, it is not surprising that simple ductal decompression may not provide significant long-lasting relief. Our series includes 2 children who failed simple longitudinal pancreaticojejunostomy but subsequently improved after distal pancreatectomy, somewhat confirming this observation. Similarly, simply resecting the distal pancreas without ductal diversion may not relieve pain as well.

For these reasons, we have chosen to resect the distal pancreas, which was the area with the most intense fibrosis in most patients in the present series, combined with Roux-en-Y drainage of the distal end of the remaining pancreas (Duval procedure). In our experience, this has provided long-term relief of pain, and follow-up ERCP has demonstrated patency of the anastomosis, even several years later (Figure 6). An alternative procedure, distal pancreatectomy combined with longitudinal pancreaticojejunostomy (modified Puestow procedure), has been used in this setting with success.

That a few patients continue to have episodes of pain after operation is not surprising and has also been emphasized in other published studies. Although some studies have suggested that successful ductal drainage might reverse the progression of the disease, other authors have reported a more unrelenting course despite successful drainage. We believe that these procedures provide significant pain relief, but we have not investigated their effects on the long-term endocrine and exocrine pancreatic function in these patients. However, no patients in our series have shown any signs of diabetes, steatorrhea, or other manifestations of pancreatic insufficiency.

The operative treatment of chronic pancreatitis in children will remain controversial, despite the generally good results in this and other published series. Attempts to preserve pancreatic tissue by performing pancreaticojejunostomy without distal resection may provide pain relief, but the surgeon should be prepared for a more aggressive approach if the conservative operation fails. We have found that combining a distal resection with ductal drainage can provide long-term relief of pain and will allow resumption of a more normal lifestyle and development in this group of children.

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REFERENCES


DISCUSSION

Karen W. West, MD, Indianapolis, Ind: [Read by Fabrizio Michiassi, MD, in Dr West's absence.] This report involves a very difficult group of children with chronic pancreatitis and offers a glimpse of the long-term problems encountered in the postoperative patient. Half of these patients had familial pancreatitis. The authors point out that symptoms were present 1 to 6 years prior to surgical intervention. Nine of the 18 patients experienced a drug dependence or psychological problems by the time of operation. Remarkably, no postoperative complications were encountered, regardless of the type of procedure employed. Dr Weber reminds us that patients with familial pancreatitis have the poorest long-term outcomes, with 5 of 9 experiencing much milder, yet ongoing, episodes of pain following surgical intervention. My questions are: (1) Ten patients underwent endoscopic sphincterotomy without improvement. What were the indications (proximal stones, strictures, elevated pressures), and do you recommend pursuing this procedure in light of these results? Were there any post-ERCP sphincterotomy complications encountered?

(2) Were you able to correlate duration of symptoms preoperatively to the problems with recurring pain or the need for conversion from a Puestow to a Duval procedure? Do you think earlier intervention before fibrosis and neuropathy developed could play a role in the subsequent course of patients with familial pancreatitis? How extensive is your distal pancreatectomy? How long was your longitudinal pancreaticojejunostomy anastomosis? We have not needed to convert any of our Puestow procedures, although 2 have had episodes of limited pain and transient hyperamylasemia in long-term follow-up. The Puestow procedure can be used successfully in these difficult pediatric patients.

(3) You indicated that none of the patients had developed symptoms of exocrine/endocrine dysfunction. Are these children being followed in a multidisciplinary setting to be able to deal with long-term problems, including narcotic dependence?
Stephen G. Jolley, MD, Anchorage, Alaska: As indicated by Dr West, this is a very difficult group of patients. I have 2 questions for Dr Weber. The first is, were you able to save the spleen in all of these pancreatic resections? The second, if I understood your presentation correctly, you operated on 2 patients who had normal ERCPs. Why was that done and what operations were done, and what was the follow-up in those patients?

Richard C. Thirlby, MD, Seattle, Wash: Dr Weber, the results in the familial group were not that good; 4 out of 9, or 44%, were pain-free. Did those patients have dilated ducts, and, if they didn’t, might they be better served with a head resection, a pancreas-preserving Whipple, which I think has been shown to be preferable in the adult population with small duct chronic pancreatitis?

Dr Weber: With regard to Dr West’s first question, the indications for sphincterotomy were those that were determined by the gastroenterologists. All of the sphincterotomies were performed during ERCP, and it was done before the patients were referred. We had no specific complications after sphincterotomy that I am aware of, other than a couple of occasions of mild pancreatitis.

There was no correlation between the pattern, severity, or the length of pain and the choice of operation. Two children underwent Puestow procedure, both of whom had hereditary pancreatitis, but neither of these patients improved significantly. At follow-up ERCP, both of the Puestow anastomoses were closed. The 2 Puestows were performed almost to the head of the pancreas, and so I believe they were quite extensive.

All of these patients are followed up by a combination of gastroenterologists and surgeons, but, as I mentioned, psychiatrists have been very helpful, particularly in the weaning of narcotic agents.

Dr Jolley, we performed all of the pancreatectomies without splenectomy. The 2 children with a normal ERCP are interesting. They both had hereditary pancreatitis and were relatively young children. They were referred fairly early, and I believe their normal ERCP was because they simply hadn’t had the years of repeat episodes of severe pancreatitis to cause their ERCP to become abnormal. Clearly, however, these 2 children had chronic pancreatitis and have benefited from their pancreatectomy.

Dr Thirlby, the hereditary pancreatitis group is a problem, both in our series as well as in the literature. It’s difficult to achieve complete pain remission in these children, and what we have been able to do in most of them is to decrease or eliminate their dependence on narcotics. There is no question in my mind that either I or one of my adult surgical colleagues is going to have to reoperate on some of these children and complete their pancreatectomy, perhaps even with a Whipple-type procedure.

Surgical Anatomy

The origin of the superior mesenteric artery courses over the left renal vein (LRV) and the third part of the duodenum; compression of the LRV may cause abdominal pain and hematuria.