Pancreatitis

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Acute pancreatitis in children is not uncommon and can be secondary to a wide range of etiologies that includes genetic disorders, infections, trauma, and congenital malformations, among others. While interventions are hardly ever indicated during the acute phase, some of the potential causes and potential complications of acute pancreatitis (*pancreas divisum*, pseudocysts) may require endoscopic, percutaneous, or surgical intervention. When the cause that leads to acute pancreatitis persists, patients can develop repeated episodes over time. Most patients with *acute recurrent pancreatitis* eventually develop chronic pancreatitis, which is defined as the combination of signs and symptoms and the presence of certain irreversible anatomical changes within the pancreas detected on imaging studies. Surgical interventions certainly play a key a role in the management of chronic pancreatitis, but patients must be taken care of by a multidisciplinary team of expert radiologists, gastroenterologists, endoscopists, interventional radiologists, pain management practitioners, and surgeons.

Acute Pancreatitis

About a third of cases of acute pancreatitis in children and adolescents are due to gallstones, the prevalence of which has increased significantly in the last 2–3 decades. Cholesterol-based stones are by far the most common, followed by those that result from bilirubin breakdown in patients with sickle cell disease and other hemolytic disorders. Stones that have accumulated in the gallbladder migrate towards the common bile duct and can get temporarily lodged at the ampulla of Vater, causing pancreatic fluid hypertension and backflow. This, in turn, provokes pancreatic cell damage and triggers a profound inflammatory process. A similar phenomenon is thought to be the explanation for the post-endoscopic retrograde cholangiopancreatography (ERCP) pancreatitis, in which the manipulation of the ampulla of Vater results in transient swelling and temporary obstruction of the pancreatic fluid, even in cases where a sphincterotomy is done. The injection of contrast at high pressure and the insertion of guide wires in the pancreatic duct (PD) are also sources of potential pancreatic damage during ERCP.

Pancreatitis can occur as a side effect of several medications used in children. The most common are valproic acid, azathioprine, mercaptopurine, and L-asparaginase. The mechanisms that lead to pancreatic damage are not fully known but likely involve a combination of direct toxicity and a focal immunologic reaction. Interestingly, patients can develop drug-induced pancreatitis weeks or even months after receiving the medication for the first time. Trauma is a well-known cause of pancreatitis in children. The traumatic impact, however, does not necessarily need to be major, as minor contusions without ductal disruption (grade-1 and -2 injuries) can lead to pancreatic inflammation.

Congenital anomalies of the pancreaticobiliary system are infrequent but well-known causes of acute recurrent pancreatitis, and typically lead to chronic pancreatitis if left untreated. The most common is pancreaticobiliary malunion (PBM), also known as *common channel*, in which the common bile duct (CBD) and the pancreatic duct (PD) merge before the ampulla of Vater/sphincter of Oddi, allowing bile to reflux into the PD and pancreatic fluid to reflux into the CBD. The chronic exposure of the CBD to pancreatic fluid is thought to be one of the mechanisms that lead to the development of a choledochal cyst and potentially to biliary mucosal metaplasia. The reflux of bile into the PD, plus the inefficient drainage of the pancreatic fluid, are the likely sources of pancreatic damage and recurrent pancreatitis in patients with PBM. Less common anomalies that can lead to pancreatitis are *pancreas divisum* (separate dorsal and ventral pancreatic ducts) and the very rare *ansa pancreatica* (extreme redundancy of the duct of Santorini).

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Acute pancreatitis in children can also result from infection: viruses (mumps, coxsackievirus, hepatitis B, herpes zoster, CMV, HIV), bacteria (leptospira, salmonella), fungi (aspergillus), and parasites (cryptosporidium, ascaris). Metabolic diseases such as hypertriglyceridemia and hypercalcemia are also potential causes of acute pancreatitis. A very rare form of pancreatitis in children is autoimmune pancreatitis (AIP). This newly described entity is quite complex and thought to be a component of a systemic autoimmune disease. The diagnosis is mostly based on the detection of elevated serum IgG4. AIP tends to be a chronic process, can lead to strictures in the PD, usually resolves with pulse steroids, and can be accompanied by an autoimmune process of the biliary tree, *IgG4-related cholangitis*.

Lastly, genetic disorders that affect the trypsinogen pathways, the cystic fibrosis transmembrane regulator (CFTR), and a number of other genes can lead to pancreatitis, which in most cases is recurrent and will evolve towards chronic pancreatitis. The PRSS1 gene encodes the cationic trypsinogen, a precursor of the protease trypsin. Gain-of-function mutations that lead to premature activation of trypsinogen within the pancreas lead to pancreatitis. A small amount of trypsin activates naturally within the pancreas, but is self-controlled by auto-cleavage. Certain mutations in the PRSS1 gene can render the trypsin resistant to auto-cleavage and therefore cause pancreatitis. The SPINK1 gene encodes the serine protease inhibitor Kazal-type 1, which cleaves prematurely activated trypsinogen within the pancreas. Any loss-of-function in the SPINK1 gene will lead to pancreatitis. In the case of the CFTR gene, certain mutations can cause alterations in the composition and thickness of the pancreatic fluid, rendering it more viscous. This, in turn, causes protein sedimentation and slow flow within the lumen of the pancreatic duct, leading to obstruction and cell damage.

Diagnosis

The diagnosis of acute pancreatitis should be suspected when elevated serum pancreatic enzymes are detected in a patient with acute epigastric abdominal pain. According to the Atlanta classification, the diagnosis of acute pancreatitis is established when 2 of 3 criteria are present: (1) abdominal pain, (2) elevated amylase or lipase, and (3) typical findings on imaging studies. Imaging studies help confirm the diagnosis but are never required to initiate medical management. Plain radiographs of the chest and abdomen can help rule out other sources of acute epigastric pain and hyperamylasemia such as intestinal perforation and intestinal obstruction, and can also detect potential complications of acute pancreatitis such as pleural effusions. Abdominal US may confirm the diagnosis by showing pancreatic swelling and peri-pancreatic fluid. Additionally, US can detect cholelithiasis or choledocholithiasis. Advanced imaging studies (CT, MRCP) are not needed to confirm the diagnosis but can help in the detection of complications (necrosis, fluid collection), and can help identifying some of the potential underlying causes (pancreatic duct anomalies).

Serum amylase and lipase have classically been measured in the workup of patients with abdominal pain, and values three or more times higher than the upper normal limit are typically regarded as diagnostic for acute pancreatitis. While the sensitivity and specificity of amylase and lipase are not equal, for clinical purposes either can be used. If both are available, lipase is preferred. Amylase returns to normal values 5–7 days after its peak, whereas lipase takes up to 14 days. It is important to remember this because clinical improvement may occur much earlier than the normalization of serum enzyme values. In other words, the clinical condition should guide the management, not the serum amylase or lipase values. Other pancreas-specific biochemical markers such as urinary trypsinogen-2 and urine amylase are highly sensitive, but rarely used in children.

Clinical Classification

According to the North American Society for Pediatric Gastroenterology, Hepatology and Nutrition (NASPGHAN)–Pancreas Committee, acute pancreatitis in children is classified as mild (no organ failure, no local or systemic complications), moderately severe (organ failure that resolves within 48 h, local or systemic complications), and severe (organ failure that persists for more than 48 h). While organ failure tends to present early, it can still develop several days after the onset of pain. For this classification to be valid, one must follow the definitions of organ failure established for children by the International Pediatric Sepsis Consensus. Similarly, the only valid definitions of local and systemic complications are the ones established for children by the NASPGHAN: local complications include peri- or pancreatic complications including fluid collections or necrosis; systemic complications include exacerbation of previously diagnosed medical co-morbidities such as lung disease or kidney disease. From an imaging standpoint, acute pancreatitis can be characterized as edematous (the pancreas is swollen but homogeneously perfused) or necrotizing (when a segment of the pancreas does not take up the intravenous contrast) (Fig. 97.1).

Acute pancreatitis has a wide range of potential clinical severity that extends from short-lived mild symptoms to multiorgan failure and death. Clinicians have tried for decades to develop clinical severity scores able to predict, at the time of presentation or shortly after, in what clinical category of acute pancreatitis each patient will fall. Having such predictive ability could allow early determination of what level of care is appropriate for each patient at the time of admission, and could allow centers with limited resources to transfer patients early, before

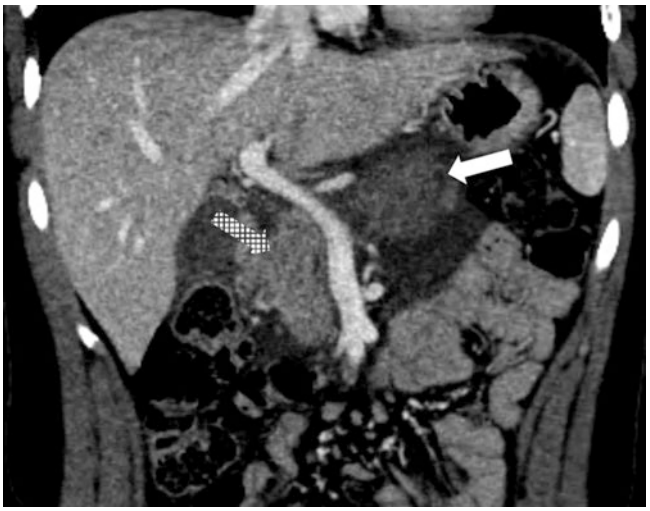

Fig. 97.1 The body of the pancreas (white arrow) is hypodense (less contrast uptake) compared to the pancreatic head (gray arrow)

the development of complications. The first score reported in the literature was by Ranson et al. in 1974. Multiple other scores followed: Accuracy of Acute Physiology and Chronic Health Evaluation (APACHE), APACHE II, Bedside Index of Severity in Acute Pancreatitis (BISAP), the Atlanta classification, and more recently the imaging-based scores such as the Balthazar CT score and the Computed Tomography Severity Index (CTSI). The clinical value of these scores is quite limited for several reasons. Some cannot be finalized until 48 hours post admission, which is too late for rapidly evolving severe cases. Others are remarkably cumbersome, like the APACHE II, which has 9 possible values for 10 different clinical parameters. In addition, none of the scores takes into consideration the etiology of the pancreatitis, which can play a critical role in determining the appropriate management. Lastly, none of the scores are purely based on pediatric criteria.

With the goal of identifying an easy pediatric-oriented score, Vitale et al. identified and Farrell et al. subsequently validated that measurement of blood urea nitrogen (BUN) on admission correlates remarkably with the chances of developing severe acute pancreatitis (SAP). Specifically, a BUN >20 mg/dL has a 98% specificity for the development of SAP. Additionally, the percentage decrease in the BUN value 24–48 h post admission (as a surrogate for response to resuscitation) also proved to be an accurate predictor of the likelihood of developing SAP. In clinical practice, children with AP undergo a whole array of blood tests. While tests may be important for what they represent individually, the only one that seems to be a useful predictor of the overall outcome is the BUN on admission.

Medical Management

As there are no pancreatitis-specific drugs available, the management of acute pancreatitis is purely supportive. Surgical intervention is hardly ever needed during the acute phase of the disease. There are three key aspects in the care of patients with acute pancreatitis: fluid resuscitation, nutrition, and pain control. The profound systemic inflammatory response triggered by pancreatic inflammation results in capillary leak, third spacing, and intravascular volume depletion. Frequent vomiting adds a true volume loss. These factors lead to organ hypoperfusion and eventually to organ failure. There is a well-known direct relationship between the degree of intravascular volume depletion (manifested by a rise in BUN) and the odds ratio for mortality. The ideal fluid for resuscitation is Ringer's lactate solution, which should be started as soon as the diagnosis is suspected, at a 1.5 × maintenance rate, titrated to vital signs, urine output, and BUN response.

There is overwhelming evidence confirming that enteral nutrition is associated with a lower rate of multiorgan failure and mortality when compared to parenteral nutrition. The benefits of enteral feedings over parenteral feedings are even clearer in cases of severe acute pancreatitis. In the absence of any form of mechanical or functional intestinal obstruction, oral feedings should be started within 24 h of hospital admission. A low-fat, low-residue diet is preferred. Patients who cannot take feedings by mouth should receive nasogastric feedings. Nasojejunal feedings are only indicated in patients who have a mechanical duodenal obstruction due to swelling or external compression. The old dogma of providing jejunal feedings to decrease pancreatic stimulation has been abandoned. Patients who have severe ileus or any form of distal intestinal obstruction need parenteral nutrition.

Pain management is critical in patients with acute pancreatitis, as severe pain can contribute to organ hypoperfusion. The mainstay therapy is intravenous opioids, followed by enteral opioids as oral tolerance is ensured. Morphine has historically been avoided due to the thought that it could induce spasm of the sphincter of Oddi, but there has been found no differences between morphine, hydromorphone, meperidine, and fentanyl.

Prophylactic antibiotics have no role in acute pancreatitis, but they become critical in patients with infected necrotizing pancreatitis or an infected fluid collection. Somatostatin analogs have been used in the past, but no benefit was found in multiple randomized trials and are not recommended. Nonsteroidal anti-inflammatory drugs have also been studied as a way to decrease pancreatic inflammation, but no benefit has been proven.

Complications

Systemic complications of acute pancreatitis are defined as the exacerbations of any pre-existing medical co-morbidities and are treated medically. The list of potential local complications that can occur in patients with acute pancreatitis is

remarkably long and includes ascites, fluid collections, pseudocysts, necrotizing pancreatitis, pancreatic fistulas, vein thrombosis, arterial pseudoaneurysms, and gastric outlet obstruction.

From a surgical standpoint, it is important to highlight that there is hardly ever a need for surgery in the acute phase of acute pancreatitis. The complications of acute pancreatitis that are not pancreatitis-specific must be managed according to their particular gold standards, regardless of the context of acute pancreatitis. Splenic-vein thrombosis, which can occur in many different clinical settings, is relatively common in patients with acute pancreatitis (20–25%). As such, it must always be searched for and carefully surveilled when identified. Cases that do not resolve spontaneously must be treated due to the risk of gastric varices and bleeding. Additionally, cases that extend into the portal vein or the superior mesenteric vein should also be treated. Arterial pseudoaneurysms occur in about 3% of patients with acute pancreatitis, according to reports from adult patients. Symptomatic pseudoaneurysms that do not spontaneously thrombose must be treated according to the resources of each center, regardless of the background of acute pancreatitis. Ultimately, specific complications are treated according to established institutional standards regardless of the fact that the patient has underlying acute pancreatitis.

Surgeons are most often called upon to assist a patient when a fluid collection is noted. The first step in the management of pancreatic fluid collections (PFC) is to understand their definitions, as established by the revised Atlanta classification of 2012: (1) acute fluid collection (AFC): a collection of simple, peri-pancreatic fluid, occurring in the context of edematous pancreatitis (no necrosis), within 4 weeks of the onset of the pancreatitis; (2) acute necrotic collection (ANC): a collection of necrotic pancreatic or peri-pancreatic tissue that by definition makes the diagnosis of *necrotizing pancreatitis*, heterogeneous on imaging studies that occurs within the first 4 weeks after the onset of the pancreatitis; (2) pancreatic pseudocyst (PC): a well-circumscribed collection of simple homogeneous fluid that has a defined pseudocapsule, occurring at least 4 weeks after the onset of the disease; and (4) walled-off necrosis (WON): a collection of well-circumscribed heterogeneous liquefied tissue that has a defined pseudocapsule and occurs more than 4 weeks after the onset of the disease. The second step in the management of PFC is to know that an asymptomatic PFC does not require drainage and should simply be followed every 3–6 months by US. The vast majority resolve spontaneously with time. The third step is to recognize that the best treatment option for a symptomatic collection is the one with which the treating team has the highest degree of expertise. There are three potential approaches for symptomatic PFC—surgical, endoscopic, and percutaneous—and each has different techniques. There has been a marked increase in the use of percutaneous and endoscopic techniques in the last decade, with surgical intervention having become much less frequent, even in the era of laparoscopy. The problems that PFC can lead to are divided in three main groups: (1) mass effect(gastric outlet obstruction, biliary obstruction, intestinal obstruction, vascular compression); (2) pain; and (3) infection. The specific management of each PFC is as follows:

AFC. Acute fluid collections that develop in the context of edematous pancreatitis rarely need an intervention. They do not usually cause mass effect, since they can freely expand throughout the abdominal cavity. Extreme cases can cause abdominal compartment syndrome, for which a percutaneous drain would be the optimal approach. Endoscopy and surgery play no role in those cases. An AFC with a high suspicion of being infected should be sampled by fine-needle aspiration, cultured, drained, and treated with the appropriate antibiotics. Most AFC resolve quickly and spontaneously, but some evolve towards a PC.

ANC. Acute necrotic collections rarely cause mass effect complications, and if they cause pain, it is indistinguishable from the pain caused by the acute pancreatitis itself. As a rule, ANC should be left alone. If there is no suspicion of infection (sterile necrosis), there is no need to intervene. If the ANC is suspected of being infected (air bubbles on the CT scan), FNA is recommended, with cultures and appropriate antibiotic treatment (although antibiotics should be started empirically right away). One of the most challenging situations that intensivists and surgeons face when managing a patient with necrotizing pancreatitis is what to do with an infected ANC that does not respond to antibiotics and leads to further clinical deterioration, since an infected ANC is one of the most common causes of death in acute pancreatitis. Patients who continue to deteriorate should undergo CT-guided percutaneous placement of drains. Endoscopy has no role in this context, since a mature wall is required for any type of endoscopic procedure. When patients continue to deteriorate despite having functioning drains, the option of a surgical debridement for source control should be considered. The operation, however, is associated with a high mortality, need for re-exploration, and damage to surrounding organs, and should therefore be done only as a last measure, if not avoided completely.

PC. Most pseudocysts resolve spontaneously with time. Size itself used to be an indication for drainage (>15 cm) but that is no longer the case. Asymptomatic PC should be simply followed by US every 3–6 months. Symptomatic PC can be drained surgically, endoscopically, or percutaneously, and the best option, again, is the one with which the treating team has the most experience. Surgical drainage involves creating a cyst-gastrostomy or cysto-jejunostomy, usually with a stapler, done open or laparoscopically. Percutaneous drainage is simply the placement of a drain that stays until the output is

near-zero. Contrary to what commonly happens after the percutaneous drainage of an acute collection, the percutaneous drainage of a PC rarely results in a pancreatic fistula. Endoscopic drainage is a commonly used option for PC, particularly in adults. There are multiple techniques, but all of them involve the creation of a cyst-gastrostomy by deploying a self-retaining stent. In recent years, endoscopic US guidance has been added to the armamentarium of the endoscopist, which helps to avoid vascular structures that could potentially be present between the cavity of the PC and the gastric lumen, among other benefits. Large studies in adults have shown similar efficacy and safety between the three treatment options for symptomatic PC.

WON. Walled-off necrosis collections can certainly cause mass effect problems and pain, and can also become infected. They can carry on an infection from the acute phase (a suboptimally treated infected ANC), or they can become infected afterwards. Infected WON have more subtle signs and symptoms than infected ANC (low-grade fever, anorexia, general malaise). Symptomatic WON can be drained percutaneously, but the thick nature of the liquefied necrotic tissue requires large-bore drains and frequent irrigations. Endoscopy is the most common approach. Just like the endoscopic drainage of a PC, a connection is created between the lumen of the stomach or duodenum and the cavity of the WON. If the content of the WON is thick, endoscopic necrosectomy should be considered. For this, the necrotic tissue is debrided mechanically with a variety of endoscopic instruments through a wide opening in the common wall between the stomach and the WON. If this treatment is not available, open or laparoscopic necrosectomy is the next step.

In summary, the management of acute pancreatitis in children is mostly medical. Surgery only plays a role in the management of some of the complications, and that role is gradually being replaced by nonoperative interventions. Where surgery provides a critical role is in the management of some of the potential sources of pancreatitis, such as biliary lithiasis and the anomalies of the pancreaticobiliary anatomy.

Chronic Pancreatitis

There is a continuum between the first episode of acute pancreatitis and full-blown chronic pancreatitis, and defining each stage has historically been difficult. According to the International Study Group of Pediatric Pancreatitis: In Search for a Cure (INSPPIRE), acute recurrent pancreatitis (ARP) is defined as 2 or more distinct episodes of acute pancreatitis with intervening return to normal pancreatic function and anatomy. Chronic pancreatitis (CP), on the other hand, is diagnosed when the following features are present: (1) typical abdominal pain plus characteristic imaging findings, or (2) exocrine pancreatic insufficiency plus imaging findings, or (3) diabetes plus imaging findings. By far, the most common etiology of ARP and CP in children is a genetic mutation, and as more genetic mutations are identified, the incidence of the idiopathic ARP and CP will continue to decrease. The diagnosis of hereditary pancreatitis can be confirmed when a patient without an identified genetic mutation has 1 or more first-degree relatives or 2 or more second-degree relatives in 2 or more generations with recurrent acute pancreatitis or chronic pancreatitis of unknown etiology.

The potential causes of CP are many, and are divided in four groups: (1) Obstructive: an obstruction to the outflow of the pancreatic duct (PD) develops at one or more places. The list includes pancreas divisum with obstructive papillae, annular pancreas, pancreaticobiliary malunion, and trauma-related obstructions; (2) Toxic: medications and, in adults, ethanol; (3) Systemic diseases: hypertriglyceridemia, lupus erythematosus, cystic fibrosis, and autoimmune pancreatitis, among others; and (4) Hereditary: caused by mutations in a variety of genes that codify different components of the trypsinogen pathway and other components of the pancreatic metabolism. The most commonly involved genes are SPINK1, PRSS1, and CFRT, but there are currently at least 14 genes known to cause CP, including chymotrypsin C (CTRC), carboxypeptidase A1 (CPA1), claudin 2 (CLDN2), and carboxylesterlipase (CEL), among others.

The typical features of CP on imaging studies are calcifications, atrophy, fibrosis, ductal tortuosity, and ductal dilatation (Fig. 97.2). US, CT, and MRI are all useful for the initial diagnosis of CP, but MRCP is the non-invasive gold-standard. MRCP is key for the follow-up of chronic pancreatitis and its 3D renderings are very helpful for surgical planning purposes. ERCP used to be widely used for the diagnosis of CP but has gradually been replaced by MRCP. ERCP still holds, however, an important role in the treatment of CP.

Left untreated, CP invariably progresses towards pancreatic endocrine and exocrine insufficiency. The goals of all forms of therapy for CP are (1) eliminate the cause, if possible, (2) eliminate or ameliorate chronic pain, (3) if there is an obstruction, provide a low-resistance outlet to the pancreatic fluid, (4) arrest the progression to pancreatic insufficiency, and (5) optimize the patient's nutrition. Patients with CP are complex and are best managed by a multidisciplinary team.

From a surgical/interventional perspective, patients with CP are divided into two groups: those with ductal dilatation and those without ductal dilatation. Their management is quite different.

CP with Ductal Dilatation

Dilatation of the pancreatic duct is always the result of an obstruction, which can be congenital (pancreas divisum), iat-

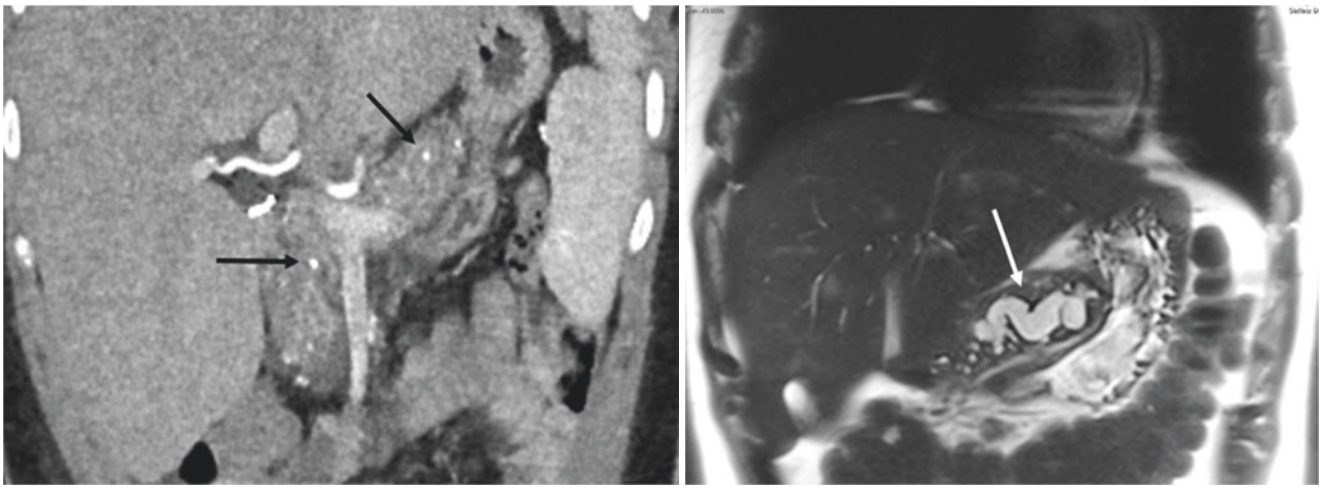

Fig. 97.2 Calcifications in the head and body (left image, clack arrows), and severe dilatation of the pancreatic duct (right image, white arrow)

rogenic (after duodenal atresia repair), or secondary to chronic inflammation and fibrosis in patients with genetic forms of CP, autoimmune pancreatitis, toxic pancreatitis, and other forms of CP. The goal of any intervention is to either correct or bypass the obstruction. Treatment options include sphincteroplasty (endoscopic or surgical) and a pancreatic drainage operation. The optimal option depends on the exact anatomy. Patients with an obstruction in the vicinity of the major papilla/ampulla of Vater are ideal candidates for an endoscopic sphincteroplasty and stent, as long as more obstructions in the head or tail have been ruled out. This endoscopic approach is the least invasive, but it requires great expertise, is not always feasible in small children, and it does not work if the obstructed area of the pancreatic duct is not reachable with the stent. A minor papilla sphincteroplasty is also the initial treatment for patients with CP secondary to pancreas divisum. Surgical transduodenal sphincteroplasty is rarely used in CP patients nowadays and is only indicated after failure of endoscopic sphincteroplasty.

Pancreatic drainage procedures have been used for many decades and are safe and effective. The decision of when to perform a pancreatic drainage procedure is not easy to make, but as a general rule it should be offered before the patient becomes opioid-dependent or when the episodes of recurrent pancreatitis become frequent enough to cause deterioration in the patient's quality of life. The most commonly performed operation in children is the Puestow procedure as modified by Partington and Rochelle. The abdomen is entered either via a transverse supraumbilical or a Chevron incision. The lesser sac is opened and the duodenum mobilized to expose the entire anterior aspect of the pancreas. The dilated main pancreatic duct, which needs to be at least 3 mm in diameter for the operation to be feasible, is identified by intraoperative US and needle aspiration is used to confirm its location. The dilated pancreatic duct is filleted open with electrocautery along its entire length. If intraductal stones are present, they are removed. A 20 to 30 cm-long Roux-en-Y jejunal limb is created, an end-to-side jejunojejunostomy is performed, and the Roux limb is brought to the pancreatic area through the transverse mesocolon. The Roux-en-Y is laid over the pancreas oriented with its free end on the pancreatic tail. The pancreaticojejunostomy is performed in two layers. First, a series of interrupted 3-0 silk sutures are placed between the seromuscular layer of the jejunum, just posterior to the antimesenteric edge, and the capsule of the pancreas 2–3 mm away from the inferior edge of the opened duct. Next, the antimesenteric border of the jejunum is opened matching the length of the opened pancreatic duct and a running suture of 3-0 polydioxanone is placed between the full-thickness jejunal wall and the edge of the opened duct including the ductal mucosa in order to obtain water-tight apposition between the jejunal and pancreatic ductal mucosa (Fig. 97.3). The pancreaticojejunostomy is completed with interrupted 3-0 silk sutures placed between the seromuscular layer of the jejunum and the pancreatic capsule, cephalad to the superior half of the previous running suture. Finally, the entire length of the pancreaticojejunostomy is covered with an omental flap. No drains are needed.

Alternatives to the modified Puestow procedure are the Frey and the Beger procedures, which were designed for cases of CP in which the pancreatic head is fibrotic and causes compression of the biliary tree, the duodenum, or the retropancreatic vessels. In the Frey procedure the majority of the pancreatic head is cored out, leaving a thin posterior layer and a thin rim on the duodenum, and the pancreatic duct of the body and tail is filleted open as in the modified Puestow procedure (Fig. 97.4a). In the Beger procedure, the pancreatic head is partially cored out and the pancreas is transected at the neck, reconstructing the drainage with a lat-

eral pancreaticojejunostomy to the remaining head and an end-to-end pancreaticojejunostomy to the pancreatic body (Fig. 97.4b). All surgical pancreatic drainage procedures are effective in providing a low-resistance outlet for the pancreatic fluid. However, the long-term outcomes in terms of pain control depend largely on the etiology of the CP and whether or not the causative factor persists after the operation. Patients with obstructive CP are likely to have definitive pain relief after the surgery, as long as the pancreatic duct remains appropriately drained. Similarly, patients with toxic CP, particularly ethanol-induced, are likely to have definitive pain relief after the decompression of the pancreatic duct if they stop consuming ethanol. In contrast, in patients with chronic pancreatitis of genetic origin the intrinsic cause of the disease does not disappear after the operation and, while for most patients pain score improve, long-term results are variable. The same is true regarding the efficacy of the drainage procedures in arresting the progression of CP to pancreatic insufficiency—it varies according to the etiology.

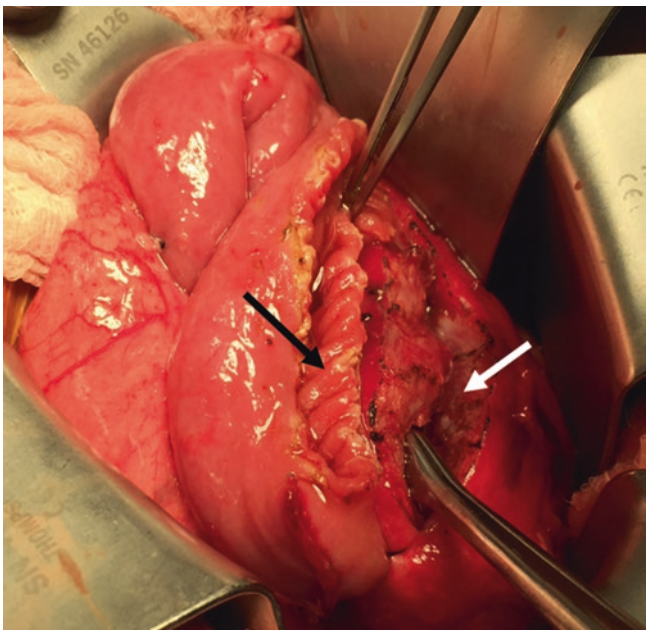

Fig. 97.3 The lumen of the jejunum (black arrow) is anastomosed to the lumen of the dilated pancreatic duct (white arrow)

CP Without Ductal Dilatation

For these patients there are no endoscopic therapies and the only surgical option to control the chronic pain is a total pancreatectomy. From a technical perspective, a total pancreatectomy is straightforward to perform, but such an operation renders the patient diabetic. This can be prevented by a simultaneous pancreatic islet autotransplantation (TPIAT). The principle of the technique is to harvest the islets contained in the pancreatectomy specimen and infuse them via the portal vein into the liver. The islets are expected to engraft within the

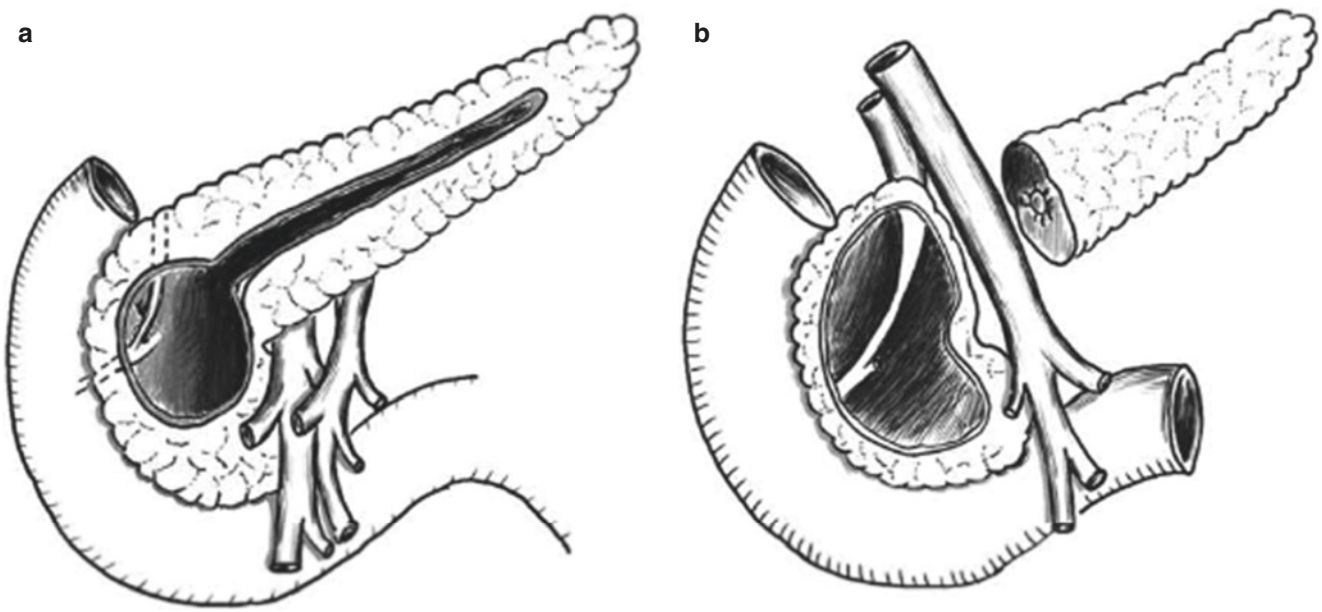

Fig. 97.4 (**a**) Frey procedure. The pancreatic head is cored out and the remaining pancreatic duct filleted open. The reconstruction is done with a lateral pancreaticojejunostomy. (Reprinted from Frey C, Smith G. Description and Rationale of a New Operation for Chronic Pancreatitis. Pancreas. 1987; 2(6): 701–7, with permission from Wolters Kluwer Health). (**b**) Beger procedure. The pancreatic head is cored out and the pancreas transected at the neck, reconstructing the drainage with a lateral pancreaticojejunostomy to the remaining head and an end-to-end pancreaticojejunostomy to the pancreatic body. (From Buchler MW. Duodenum-preserving pancreatic head resection: long-term results. J Gastrointest Surg. 1997; 1(1): 13–9, reprinted with kind permission from Springer Science + Business Media)

liver sinusoids and survive permanently. The success of the procedure greatly depends on the number of islets isolated from the pancreatectomy specimen. The islet yield is generally lower in patients with severe pancreatic fibrosis and in patients who underwent previous pancreatic drainage procedures. TPIAT can only be offered if there is a reasonable beta-cell mass, which is measured by c-peptide levels. An additional benefit of TPIAT in patients with hereditary pancreatitis is the complete elimination of the risk of pancreatic cancer. The optimal timing of TPIAT is still unknown. On one hand, early TPIAT may be beneficial in terms of islet yield (less fibrosis). On the other hand, even the largest series reveal a 10-year insulin independence rate of less than 15%, so even though children do better than adults in this regard, doing TPIAT before the age of 10 years currently leads to most patients becoming diabetic before the age of 20 years. Improvements and refinements in the islet isolation technique will hopefully result in much better TPIAT outcomes in the future.

Editor's Comments

Acute pancreatitis in children is usually idiopathic (most are likely due to exposure to toxins or medications), though the workup should include a search for gallstones, hyperlipidemia, specific toxins (alcohol, l-asparaginase), anatomic abnormalities, cysts, and a positive family history suggesting a genetic predisposition. Treatment is supportive and individualized, but the Ranson criteria are not very useful and imaging does not always correlate with clinical severity. It turns out that most of what we were taught (stop me if you've heard this before) was wrong: oral or gastric feedings are not only safe but beneficial, morphine does not make things worse, acute fluid collections can safely be observed, and none of the medications we often administered empirically—including octreotide, H2 blockers, nonsteroidal anti-inflammatory drugs (NSAIDs), and antibiotics—add any benefit.

Pancreatic necrosis is uncommon and infected pancreatic necrosis requiring intervention is especially rare. If the patient is stable, percutaneous drainage or laparoscopic debridement might be reasonable before embarking on a morbid and protracted course of serial necrosectomy. Pancreatic pseudocysts in children almost always eventually resolve spontaneously. Indications for intervention include persistent symptoms or a large cyst that persists for more than 6 weeks. Radiology-guided percutaneous drainage with placement of an internal stent is increasingly popular and seems to work in many cases. The laparoscopic approach is a reasonable consideration but is probably best done as an intragastric procedure, which even for experienced laparoscopists, can be tricky. The traditional open approach is safe and can usually be done through a relatively small incision and an anterior gastrotomy using an endo-stapler to create an effective cyst-gastrostomy.

The patient with chronic pancreatitis and a dilated pancreatic duct should be offered a Puestow procedure, though in many cases the surgeon should be prepared to include if necessary the head of the pancreas (Frey modification). The key is to offer the operation before the damage is too far gone for it to make a difference or after the patient has become addicted to opiates.

Further Reading

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