LIVER AND PANCREAS DISORDERS IN PEDIATRIC SURGERY

Department of Pediatric Surgery
CM UMK Bydgoszcz, Poland
NESIDIOLASTOSIS/CONGENITAL HYPERINSULINISM
Preferred term is congenital hyperinsulinism

Incidence 1:50,000 live births

Unregulated secretion of insulin in pancreatic \( \beta \)-cell hyperplasia due to alterations of the \( K^+ \)-ATP channel (mutations of four genes, for the following: Kir6.2 and sulfonylurea, glucokinase, glutamate dehydrogenase receptor)

Persistent hypoglycemia is often resistant to therapy
### Classification of Congenital Hyperinsulinism

<table>
<thead>
<tr>
<th>Pathology</th>
<th>Pathological correlate</th>
<th>Cases</th>
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<tr>
<td>Diffuse pancreas involvement</td>
<td>β-cells with abnormal large nuclei and abundant cytoplasm</td>
<td>60%</td>
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<tr>
<td>Focal adenomatous hyperplasia</td>
<td>Apparently normal β-cells</td>
<td>40%</td>
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Signs are related to severe persistent hypoglycemia in the neonatal period
Blood glucose of <40 mg/dl in premature and term babies
Bradycardia
Irritability, fatigability, convulsions
Jitteriness, tremulousness, tachycardia
Poor feeding
Inappropriate sweating, coma
Glycemic profile

Selective transhepatic catheterization for sampling of blood glucose and insulin levels in the pancreatic veins

Ultrasonography (not always contributory)

CT (not always contributory)

MRI (not always contributory)
## Criteria for Diagnosis of Congenital Hyperinsulinism

| Insulin                                                                 | Inappropriate plasma levels of insulin in the presence of hypoglycemia  
Plasma glucose <40 mg/dl simultaneous with elevated plasma insulin >13 μU/ml (glucose:insulin ratio <3:1) |
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<td>Glucose substitution</td>
<td>High glucose requirement to maintain normoglycemia &gt;10 mg/kg/min</td>
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<td>Fat</td>
<td>Low plasma β-hydroxybutyrate and free fatty acids &lt;1.0 mmol/l</td>
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<tr>
<td>Glucagon</td>
<td>Inappropriate glycemic response to intravenous glucagon (rise of &gt;30 mg/dl in serum glucose level)</td>
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CONSERVATIVE THERAPY

- Maintenance of normal glycemia by glucose infusion (up to 15-20 mg/kg/min) and/or high-calorie enteral feeding
- Diazoxide, insulin antagonist (up to 20 mg/kg/day) (however, many side-effects)
- Followed by octreotide or glucagon for diazoxide-resistant cases
- Hydrochlorothiazide (synergistic action with diazoxide)
Purpose is to reduce the mass of insulin-producing β-cells.

Control of hyperinsulinism is achieved through the extent of pancreas resection.

For diffuse form, usually 95%.

For focal form, less radical excision (after pancreatic venous sampling and with the help of frozen-section biopsies during surgery).

Resection of more than 98% may result in endocrine and exocrine insufficiency.

Resection of less than 95% may result in failure to cure or recurrence.
Make an upper transverse abdominal incision
Open the omentum major
Expose the pancreas after mobilization of the duodenum (Kocher maneuver)
Perform the resection: options include the tail, body, uncinate process and the majority of the head, sparing the spleen
Dissect the pancreas and ligate all small pancreatic arterial and venous branches
Leave a sliver of pancreatic tissue to the left of the common bile duct and on the surface of the duodenum
Ligate the pancreatic duct with a nonabsorbable stitch
Seal the pancreatic parenchyma with collagen glue or equivalent
Ensure peritoneal drainage
POSTOPERATIVE CARE

- Gastric tube on suction
- Intravenous nutrition for 5-7 days
- Frequent serum glucose tests
- Administration of insulin as required
- Antibiotics for 5 days
- Peritoneal drain to be removed 3-4 days postoperatively
- Follow adaptation for 4-6 months
50% cure after <95% resection, 19% cure after >95% resection
Better results in focal disease assuming complete excision and normal remaining pancreas
Diabetes mellitus in 15%
Possible exocrine insufficiency
General considerations

Incidence: 1:10,000 live births

End result of a destructive inflammatory process, unknown etiology (viral, toxic)
- Intrahepatic hypoplasia (Alagille’s syndrome)
- Alpha-1-antitrypsin deficiency
- Thick bile syndrome (inspissated bile syndrome) after hemolysis
- Neonatal hepatitis (intrauterine viral infection), giant cell hepatitis
- Sepsis with jaundice
- Cystic fibrosis
- PFIC (progressive familial intrahepatic cholestasis or Byler disease)
<table>
<thead>
<tr>
<th>Type</th>
<th>Description</th>
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<tbody>
<tr>
<td>Type 1</td>
<td>Atresia of the choledochal bile duct with patent proximal ducts</td>
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<tr>
<td>Type 2</td>
<td>Atresia of the common hepatic duct, residual patency of proximal ducts</td>
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<tr>
<td>Type 3</td>
<td>Atresia of the entire extrahepatic duct system, involving right and left hepatic ducts towards the porta hepatis</td>
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Atresia of ductus choledochus,
Atresia of ductus hepaticus communis,
Atresia of ductus hepaticus communis, ductus hepaticus dexter and sinister
Early jaundice (first 36 h after birth)
Jaundice may appear at 3 weeks
Total bilirubin >12 µmol/l in term infants, >15 µmol/l in premature infants
Prolonged jaundice >8 days in term infants and >14 days in premature infants
Conjugated bilirubin >15% of total bilirubin
Nonpigmented stools
Dark urine
Increased size of liver and spleen
Later on ascites
Associated malformations (malrotation, situs inversus, polysplenia, preduodenal portal vein, absent inferior vena cava, cardiac defects)
PREOPERATIVE WORK-UP

- Exclude infections, metabolic, endocrine disease or genetic disorder
- Blood sample: bilirubin, transaminases, alkaline phosphatase,
  \( \gamma \)-glutamyl transferase (\( \gamma \)-GT), coagulation factors
- Cytomegalovirus (CMV) and hepatitis virus
- Urine: bilirubin, urobilinogen (lacking)
- Ultrasound (not very specific), exclude choledochal cyst
PREOPERATIVE WORK-UP

- Liver biopsy (in 10% there is difficulty of interpretation)
  Transcutaneously as a needle biopsy (beware of Bleeding!; small amount of material obtained)
  Laparoscopically (bleeding control, adequate amount of material obtained). Can also be combined with a cholangiography
- In special cases (not mandatory)
  Hepatobiliary excretion scans (technetium-labeled agents)
  Percutaneous cholangiography (easier laparoscopically)
  Endoscopic retrograde cholangio-pancreatography (ERCP)
- Vitamin K, i.v. (1 mg · day-1) for 4 days preoperatively
- Blood typing, cross-match
- Perioperative antibiotics
Perform the operation early, before liver fibrosis or cirrhosis
Place the patient in a prone position on the operating table to permit cholangiography
Make an incision usable for possible future liver transplantation (transverse upper abdominal subcostal incision)
Be prepared for possible Roux-en-Y anastomosis
Inspect the abdominal cavity (search for malrotation, situs inversus, preduodenal portal vein). Confirm diagnosis of biliary atresia (gallbladder may be hidden between segments 5 and 4)

- Dissect the hepatoduodenal ligament
- Mobilize the liver
- Mobilize the gallbladder from its liver bed; used as a guide to the fibrous remnant of the hepatic duct. This will lead to the porta hepatis
- Prepare the hepatic artery, right and left branches
- Ligate all lymphatic vessels
Prepare the portal vein; follow the right and left branches as far as possible and expose the porta hepatis behind the bifurcation of the portal vein (for extended hepatoporoportoenterostomy; not performed in the original Kasai procedure)

When necessary, exteriorize the liver

Ligate all small portal branches to the caudate lobe

Section Arantius’ ligament (or ligamentum venosum), which helps mobilization of the left portal branch

Widely expose the porta hepatis

Cholecystectomy: excise remaining tract and tissue of porta hepatis flush with liver capsule
Extensive numbers of bile ducts present posteriorly and laterally.

- Ensure hemostasis; replace liver; create Roux-en-Y loop.
- Level of jejunal section approximately 10 cm from the Treitz ligament.
- Length of jejunal loop at least 50 cm, placed in retrocolic position.
- Make a 3-cm-long incision on the antimesenteric border.
- Make an anastomosis of jejunal loop to tissue at the porta hepatis, going far on both lateral sides, posteriorly on the caudate lobe and anteriorly on the quadrate lobe.
- Any cystic dilatation not containing bile is considered not to be communicating and should be excised.
Occasionally, gallbladder and distal bile duct are not affected by the atretic process and the gallbladder can be used for the anastomosis at the porta hepatis
If the residual segment of the proximal bile duct is long enough, a hepaticojejunostomy is feasible (rare type I cystic lesion)
OPERATION: KASAI PORTOENTEROSTOMY /EXTENDED HEPATOPORTOENTEROSTOMY
OPERATION: KASAI PORTOENTEROSTOMY /EXTENDED HEPATOPORTOENTEROSTOMY
OPERATION: KASAI PORTOENTEROSTOMY/EXTENDED Hepatopportoenterostomy
The main problem is ascending cholangitis (best prevention: long loop of jejunum)

Gastric tube on suction

Initially total parenteral nutrition (TPN), but enteral nutrition as soon as possible

Antibiotics for 5-7 days, discuss continuous (for 2-3 months) prophylactic oral antibiotics (cephalosporin)

Prednisolone, although there is no general agreement on steroids, we recommend

- days 1,2: 5 mg/kg
- day 3: 2 mg/kg
- days 4-14: 1 mg/kg

- Cholestyramine (if bile flow)
- Choleretics (ursodeoxycholine)
- Vitamins A, D, E, K
Depends on grade of liver fibrosis or cirrhosis (late operation, more liver destruction)

When operation is performed in first 6-8 weeks of life, chances of obtaining bile flow is approximately 70%–90%; beyond 12 weeks of age chances decrease to 35%

5-year survival rate with native liver is approximately 60%

Portoenterostomy and transplantation are now complementary procedures and give good quality of life and a 80%-90% survival rate
CHOLEDOCHAL CYST
Incidence: 1:100,000 live births
Female:male ratio 3:1 to 4:1
60% of cases are diagnosed before 10 years of age
Most frequent etiology
  Common pancreaticobiliary channel
  Pressure in pancreatic duct higher than in the bile duct
  Reflux of pancreatic juice in the common bile duct damages endothelium, causing cystic dilatation
Other etiologies are also possible, such as obstruction of the distal common bile duct and genetic reasons
## Classification of Choledochal Cyst

<table>
<thead>
<tr>
<th>Type 1</th>
<th>Cystic or fusiform dilatation of choledochus (most frequent)</th>
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<tbody>
<tr>
<td>Type 2</td>
<td>Choledochus diverticulum</td>
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<tr>
<td>Type 3</td>
<td>Choledochocele</td>
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<tr>
<td>Type 4</td>
<td>Combination of intrahepatic and extrahepatic cysts (second most frequent)</td>
</tr>
<tr>
<td>Type 5</td>
<td>Isolated intrahepatic duct cysts, single or multiple (Caroli’s disease)</td>
</tr>
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</table>
Types of choledochal cysts.

- a Cystic dilatation,
- b fusiform dilatation,
- c without dilatation,
- d cystic diverticulum,
- e choledochocele,
- f intrahepatic bile duct dilatation
Usually during the first decade of life
Some are asymptomatic (prenatal diagnosis)
This classic triad only present in 6% of cases
  Abdominal mass
  Intermittent episodes of jaundice
  Intermittent episodes of abdominal pain
Recurrent cholangitis
Pancreatitis
Biliary calculi
Pancreatic duct calculi
Pancreatitis
Cyst rupture, biliary peritonitis
Portal hypertension
Liver fibrosis or cirrhosis
Cholangiocarcinoma
Occasionally diagnosed antenatally
- Ultrasonography
- CT or magnetic resonance cholangiopancreatography (MRCP)
- ERCP
- Percutaneous transhepatic cholangiography
- Hepatobiliary scintigraphy

Not all investigations are necessary, decision according to infrastructure
Early operation prevents complications
Occasional acute pancreatitis that is resistant to conservative treatment with a gallbladder under tension requires percutaneous drainage of the gallbladder, in order to achieve resolution of pancreatitis before surgery
Aim of surgery is complete removal of the cyst
Cystenterostomy should not be done, because of potential complications (cholangitis, cholelithiasis, pancreatolithiasis, biliary cirrhosis, cholangiocarcinoma)
Perioperative antibiotics
Make a high transverse incision
Check appearance of liver and spleen
Perform liver biopsy
Sample bile aspirated from the cyst for culture and pancreatic enzyme concentration
Cholangiography to delineate precise anatomy, intrahepatic ducts and pancreaticobiliary junction
Mobilize the gallbladder and cystic duct
Care must be taken to avoid damage to an aberrant right hepatic artery, usually very adherent to the cystic wall
Lift the cyst and gallbladder with a tape placed under the bile duct (pay attention to the portal vein, often adherent to the posterior wall of the cyst)
Dissect the common hepatic duct at its bifurcation
Further dissect the bile duct to within the head of the pancreas
Remove the entire cyst; oversew the distal duct end
In difficult cases, open the cyst and remove mucosa from the bottom of the cyst
Possible calculi and debris of intrahepatic and pancreatic ducts should be cleared (intraoperative endoscopy)
Make a roux-en-Y loop anastomosis to the hepatic duct bifurcation (wide hilar anastomosis)
Occasionally, a transduodenal sphincteroplasty is necessary (difficulty in removal calculi from long common channel)
Postoperative care
As in biliary atresia, without prednisone and choleretics

Prognosis
Low mortality
10% complications: cholangitis, pancreatitis, anastomotic stricture (even late), calculi, cholangiocarcinoma
Operation before 5 years of age limits complications
Not as frequent as in adults. More frequently discovered within the last three decades, probably because of improvements in diagnostic techniques (ultrasonography)

Two peaks of appearance: first in infancy and second in early adolescence with a steady increase thereafter

Female:male ratio 1:1 in infancy, 2-4:1 in prepuberty
<table>
<thead>
<tr>
<th>Pathogenic mechanisms</th>
<th>Risk factors</th>
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<tbody>
<tr>
<td>Increased bilirubin secretion</td>
<td>Hemolytic disorders</td>
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<td>Excess bilirubin pigment</td>
<td>Sickle cell anemia</td>
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<td></td>
<td>Thalassemia</td>
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<td>Spherocytosis</td>
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<td></td>
<td>Syndromes associated with hyperbilirubinemia</td>
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<tr>
<td>Perturbation of enterohepatic circulation</td>
<td>Short bowel syndrome or ileal resection</td>
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<tr>
<td>Biliary stasis</td>
<td>Medication (ceftriaxone, somatostatin, ciclosporin)</td>
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<tr>
<td>Lithogenic bile</td>
<td>Cystic fibrosis</td>
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<td></td>
<td>Total parenteral nutrition</td>
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<td>Wilson’s disease</td>
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<tr>
<td>Malformations of biliary tree</td>
<td>Biliary stricture or cysts</td>
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<td>Post-cholangitis stenosis</td>
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May be asymptomatic

In infancy nonspecific signs (poor feeding, vomiting, irritability)

Abdominal pain (right upper quadrant or epigastrium, nausea, vomiting)

Ileus

Biliary colic, acute cholecystitis, choledocholithiasis with obstructive jaundice, pancreatitis
PREOPERATIVE WORK-UP

- Detailed history, search for any general condition
- Ultrasonography
- X-ray of abdomen without preparation
- Magnetic resonance cholangiography
- ERCP, with possible papillotomy
Make a right subcostal incision
Dissect the hepatoduodenal ligament and gallbladder
Ligate and section the cystic artery
Dissect the cystic duct and perform a cholangiography in order to visualize concrements in the duct
Ligate and section the cystic duct
Visualize the choledochus and excise the gallbladder in an anterograde fashion
Ensure hemostasis of the gallbladder bed
Drainage not mandatory
T-tube drainage in cases of choledochus exploration
Has become the standard technique
If gallbladder is under tension, it may be punctured and its extremity grasped with instruments
Gallbladder dissection begins close to Hartmann’s pouch; window created above and behind the cystic duct and artery
Cholangiogram
Section the cystic artery and cystic duct between clips
Dissect the gallbladder in a retrograde fashion
Remove the gallbladder through the umbilical port (incision can be widened)
Remove the entire pneumoperitoneum
Choledocholithiasis must be treated (through laparoscopy, conversion to open exploration or postoperative ERCP and extraction)
OPERATIVE STEPS: CHOLECYSTECTOMY
POSTOPERATIVE CARE

- Rapid recovery after laparoscopy
- Standard postoperative care according to any intra-abdominal procedure
- Prognosis is good
The portal venous system drains blood from the stomach, pancreas, gallbladder, spleen, and intestines into the liver.

Portosystemic anastomoses exist in four main areas,
1. the gastro-esophageal veins via the cardiac vein and perforating esophageal veins,
2. the retroperitoneum via the pancreaticoduodenal veins and the retroperitoneal-paravertebral veins,
3. gastrorenal-splenorenal vein, and
4. the hemorrhoidal plexus.
Portal hypertension is defined as elevation of the portal venous-IVC pressure gradient above 10-12 mm Hg.

Portal hypertension in children can be divided into two major categories based upon the anatomic location of the increased portal resistance.

Extrahepatic portal hypertension (EHPH) is most commonly the result of portal vein obstruction due to thrombosis.

Intrahepatic portal hypertension (IHPH) is typically associated with congenital liver or biliary diseases in children. Biliary atresia is by far the most common cause of IHPH in children.
Children with IHPH usually present between several months to one year of life with severe hepatic dysfunction, manifested by jaundice, hepatic encephalopathy, and malnutrition complicated by poor growth and increased susceptibility to infections.

Extrahepatic portal hypertension most commonly presents in the first decade of life with gastrointestinal hemorrhage from esophageal varices.
DIAGNOSIS

- In a child presenting with an initial episode of gastrointestinal bleeding, abdominal ultrasonography is used to confirm the etiology. The presence of portal vein thrombosis, the extent of collateral formation, and the direction of portal vein flow is established by this noninvasive and relatively inexpensive diagnostic exam.
- Upper endoscopy is used to identify and quantitate esophageal varices.
- This procedure is possible therapeutic intervention (i.e., sclerotherapy, banding).
- Angiography is a much more rarely used diagnostic and potentially therapeutic modality used in certain cases of portal hypertension.
In the acute setting, massive hemorrhage is managed with intensive care monitoring, transfusion of red blood cells and fresh frozen plasma, and potentially intubation and sedation to minimize agitation that increases variceal pressure. Octreotide infusions effectively control acute hemorrhage in the vast majority of children with portal hypertension and variceal bleeding. Endoscopic variceal banding or sclerotherapy is occasionally used in cases where hemorrhage does not resolve with supportive care. Once the patient has stabilized, endoscopy with sclerotherapy or banding is employed to prevent repeat episodes of hemorrhage.
Surgical treatment of portal hypertension can be either direct, which involves ligation of the varices themselves, or indirect, in which the portal venous system is decompressed with a surgical shunt. Examples include portocaval, mesocaval and central splenorenal shunts. Selective portosystemic shunts shunt a portion of portal blood into the systemic circulation, with distal splenorenal shunt being the most common. Recently, selected children with EHPH due to portal vein thrombosis have been successfully treated by surgical creation of a mesenterico-portal venous bypass (Rex shunt).
Treatment of intrahepatic portal hypertension focuses on the primary liver disease.

For advanced cirrhosis and other intrahepatic sources of portal hypertension, liver transplantation is the definitive treatment.
For patients with EHPH, sclerotherapy is effective in the treatment of acute variceal bleeding in up to 75% of patients. However, several follow-up sessions are necessary to obliterate the varices, and a rebleeding rate of 5-25% is expected.

Persistent variceal bleeding as well as hypersplenism may require a surgical shunt.

Selective shunts have proven successful for the control of bleeding, thrombocytopenia, and leukopenia, without creating great risk of encephalopathy.
DISORDERS OF THE PANCREAS
The pancreas develops in the 4th week of gestation and begins as two buds, dorsal and ventral, from the endoderm of the duodenum.

The growing dorsal portion of the developing pancreas spans across the hepatic diverticulum while the ventral portion lies below moving more distal. The dorsal and ventral portions fuse in week ten. The distal portion will create the duct of Wirsung while the proximal portion may obliterate or form the duct of Santorini. 10% of the population will have a double collecting system in the pancreas.

Fetal insulin production begins in the fifth gestational month and the exocrine function is present at birth.
The pancreas is a retroperitoneal organ located at the vertebral L1-L2 level.

The head of the pancreas lies to the right of the vertebral column and, along with the uncinate process, is intimately adherent to the duodenum. The body of the pancreas lies anterior to the superior mesenteric artery and vein, and the portal vein.

The arterial supply of the pancreas is derived from the gastroduodenal artery, superior mesenteric artery, and the splenic artery. The head of the pancreas receives arterial blood via the four pancreaticoduodenal arteries (i.e., anterior superior, anterior inferior, posterior superior, and posterior inferior). Venous drainage is via the splenic and portal vein.
The vast majority of cases of pancreatitis in children are from blunt abdominal injury. In the pediatric population, nearly 40% of cases of traumatic pancreatitis are attributable to bicycle-related injury.

After trauma, the most common causes of pancreatitis in children are drug therapy (corticosteroids, azathioprine, thiazides, furosemide, tetracyclines, and valproic acid), viral infection (Epstein-Barr, Coxsackie, enterovirus, and mumps), and bacterial infection. Cystic fibrosis, biliary disease, vasculitic diseases (systemic lupus, Henoch-Schonlein purpura), and type I and V hyperlipidemias are also associated with acute pancreatitis in the pediatric population.
Serum amylase, trypsinogen, and lipase levels are useful to establish the diagnosis of acute pancreatitis. An elevated serum amylase is the usual biochemical abnormality associated with acute pancreatitis. Because amylase production occurs from other nonpancreatic sources (i.e., salivary gland), elevated serum amylase is relatively nonspecific. Calculation of the amylase clearance may be helpful and is normally less than 5%. Trypsinogen and lipase are produced almost exclusively by the pancreas; elevated serum levels are more specific for pancreatitis.
Computed tomography (CT) is the best radiographic study to image the pancreas in cases of severe or complicated pancreatitis. Abdominal CT is often obtained as part of the trauma evaluation.

Ultrasound is sometimes useful, but often only provides limited visualization of the pancreas due to its retroperitoneal location and interposed bowel gas which further limits the study.

Endoscopic retrograde cholangiopancreatography (ERCP) is an invasive test that can accurately delineate pancreatic ductal anatomy. ERCP causes pancreatitis in 5-10% of cases and is generally avoided during the early phases of acute pancreatitis.
Medical management is the mainstay of treatment for pancreatitis. Volume resuscitation is essential to counter retroperitoneal third space fluid losses.

Nasogastric decompression is recommended to avoid gastric distention and patients are initially maintained NPO with nasogastric decompression.

Pain management is essential. Meperidine is preferred because it does not cause sphincter of Oddi contraction like morphine does.

Hyper-alimentation may be necessary if the course of pancreatitis is prolonged. Enteral feeding distal to the ligament of Treitz via duodenal feeding tube is the preferred method of providing nutrition in refractory cases. The majority of cases of pancreatitis are self-limited and resolve spontaneously with supportive therapy.
In severe cases (i.e., necrotizing pancreatitis, infected pancreatic necrosis), surgical intervention may be necessary for irrigation and/or debridement of the pancreas.

The morality rate in this scenario approaches 15%.
Pancreatic cysts are broadly classified based on etiology. These are simply categorized as:

1. congenital,
2. retention,
3. pseudocysts,
4. neoplastic,
5. parasitic.
Congenital cysts of the pancreas are a rare finding in children. The cysts may be unilocular or multilocular and are most commonly found in the body or tail of the pancreas. These cysts are lined with true epithelium and most commonly contain nonenzymatic fluid. The majority of these lesions are asymptomatic unless they are large. Symptomatic cysts are excised.
Pancreatic retention cysts occasionally occur in children and are associated with chronic obstruction of the pancreatic ductal system.

These cysts are filled with enzyme containing fluid.

Surgical treatment is by excision or internal drainage.
Approximately 90% of pseudocysts occur secondary to trauma. This condition is more common in males (nearly 3:1). Patients present with symptoms (most common to least) of vomiting, abdominal pain, abdominal mass, fever, and anorexia.

Pseudocysts are usually located in the lesser sac and the cyst wall consists of granulation tissue. If the cyst communicates with the pancreatic ductal system, high amylase levels are measurable with the cyst fluid. Useful diagnostic tests include serum amylase, ultrasound, and CT.

Surgical treatment is indicated for large, persistent, or infected/symptomatic cysts. The surgical options include external drainage, cystgastrostomy, cystjejunostomy, or excision. Surgical therapy is associated with low mortality, minimal morbidity, and low recurrence.
CONGENITAL PANCREATIC ABNORMALITIES
Annular pancreas is a rare congenital anomaly that occurs due to abnormal rotation of the pancreatic ventral bud. It is the most common of the congenital pancreatic abnormalities. The annular pancreas usually completely encircles the second portion of the duodenum. This anomaly is associated with Down’s syndrome, abnormalities of rotation, duodenal atresia, and biliary atresia. Seventy percent of children with this lesion are symptomatic and will present with high intestinal obstruction. The emesis is most often bilious but can be nonbilious as well. Surgical therapy consists of bypass anastomosis: duodenoduodenostomy or duodenojejunostomy. Division of the pancreatic tissue is not recommended due to the high association of fistula formation. Gastrojejunostomy is not recommended due to associated growth problems and the risk of marginal ulcers.
Pancreas divisum is a congenital anomaly in which the dorsal and ventral pancreatic tissue fail to fuse in utero. Pancreas divisum is identifiable in 10-15% of the population. This condition is usually asymptomatic, however there may be an increased incidence of pancreatitis due to the inability of the accessory duct to adequately drain the pancreatic tissue. Diagnosis is made exclusively with ERCP.

Treatment, if necessary, is by endoscopic or surgical sphincterotomy of the accessory ampulla.
Ectopic pancreatic tissue is frequently identified in the duodenum, colon, pylorus, appendix, or a Meckel’s diverticulum.

Ectopic pancreas can cause local inflammation and bleeding and is noted in approximately 3% of postmortem examinations.