Diseases of the gallbladder and pancreas in children

Diseases of the gallbladder and bile ducts

- **gallbladder anomalies** – accompany bile duct atresia or cystic fibrosis
- **acute gallbladder hydrops** – during generalized infections, sepsis or Kawasaki disease
  - manifestations – fever, jaundice, more palpable gallbladder
  - treatment – conservative, surgery is indicated only in gangrene of the gallbladder
- **cholecystitis** – G+ (streptococci) but also G- bacteria (salmonella, shigels), parasites (ascaris, giardia)
  - sometimes it is a complication of periarteriitis nodosa and other vasculitis or in Kawasaki disease
  - possibly also in Wilson's disease
  - therapy - ATB, good hydration
- **cholelithiasis** – is relatively rare in children
  - mainly in obese girls during puberty or after repeated hemolysis (pigment stones) - in our country most often after spherocytosis, in black people after sickle cell disease)
  - may also be in CF, Wilson, congenital bile abnormalities. pathways, in IgA deficiency
  - clinical picture – nausea, vomiting, jaundice, fever, often colic-like abdominal pain with radiation back and under the right scapula or under the right shoulder
  - therapy – surgical, for colic - hydrate well, strict diet and give short-term antispasmodics resp. analgesics (Algifen)

Extrahepatic atresia of the bile ducts

- absence of extrahepatic bile ducts
- incidence - 1:14 000 births
- It is thought to be the result of a sclerosing inflammatory process that begins fetally, at delivery or immediately after, etiology unknown
- Newborns are usually born out of a normal pregnancy and are fine immediately after birth
- atresia manifests at the age of several days, max. weeks - cholestatic syndrome , ie prolonged jaundice with a change in the color of urine (dark) and stool (light, acholic)
- percutaneous liver biopsy is important , but there are no typical histological findings that would distinguish it from other liver diseases
- obstruction can be verified isotopically or by ERCP
- surgery - in all infants in whom we have ruled out other liver diseases
  - hepatoportoenteroanastomosis is most often used (Kasai's operation) - anastomosis between the hepatic porta and the intestine is established

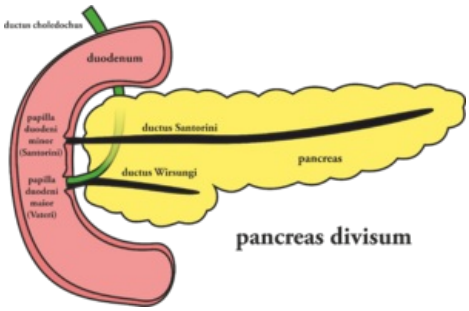
Diseases of the pancreas

- The most common pancreatic disease in children is cystic fibrosis
- otherrs are quite rare
- diagnosis - examination of pancreatic secretion - collection of duodenal juice after secretion stimulation (demanding and burdensome)
  - screening test for pancreatic lipase secretion disorder - examination of fat droplets in stool
  - PABA test - we administer a peptide from which chymotrypsin cleaves PABA (paraaminobenzoic acid) - it is absorbed and excreted in the urine

Congenital anomalies of the pancreas

- **annular pancreas**
  - it is formed by incomplete rotation of the ventral part, the pancreas revolves around the duodenum and jejunum and makes a typical picture of congenital GIT obstruction
  - more common in Down syndrome
  - symptoms - see stenosis

- **divided pancreas**
  - is one of the most common GIT anomalies, mostly it causes only minimal problems or is detected by chance
  - the incidence is estimated at 10% of the population
  - the result of the disorder is that part of the pancreas (cauda, corpus and part of the head) does not secrete into the ductus Wirsungi, but through the ductus Santorini
  - the anterior part may be compressed and chronic pancreatitis may develop
- **ectopic pancreatic tissue**
  - It can be localized in the small intestine or stomach, usually does not cause problems



- sometimes it can cause bleeding, partial obstruction or a **intussusception** (ie insertion of a part of the intestine into the adjacent part of the intestine)

Congenital disorders of pancreatic excretory function

- isolated deficits of pancreatic enzymes - rare
- **Johanson–Blizzad syndrome** – congenital pancreatic insufficiency, deafness, microcephaly, skull skin defects, hypothyroidism, dwarfism and nasal wing aplasia
- **Schwachman-Diamond Syndrome** – the most common congenital secretion disorder
  - AR, incidence 1:20 000, pancreatic tissue is infiltrated with fat
  - clinical picture – malabsorption syndrome, growth and development retardation
    - reminds of CF but without finding chlorides in sweat, there are usually more associated congenital diseases
    - neutropenia (neutrophils show loss of chemotaxis), metaphyseal dysplasia → growth disorders
    - growth disorders persists even with pancreatic replacement. enzymes (unlike CF)
    - neutrophilia is cyclical, leads to frequent bacterial infections and often to lethal sepsis
  - diagnosis - malabsorption, skeletal changes, disorders of pancreatic secretion, normal Cl concentration in sweat
    - on CT is a hypodense pancreas
  - therapy - enzyme substitution, consistent and early treatment of bacteria. infections, on neutropenia - G-CSF
  - prognosis - severe

Acute pancreatitis

- it can accompany or complicate many general childhood diseases
- **etiology**: infections - parotitis virus, VHA, coxsackie, pancreas is also sensitive to many toxins and drugs
  - in adolescents it can occur after alcohol and drugs, in all children after some immunosuppressants or antimicrobial drugs (tetracyclines, sulfonamides), diuretics, antiepileptics, Au preparations
  - occasionally occurs in congenital bile duct and pancreatic disorders, CF, Rey's syndrome and MODS
  - from systemic diseases - most often in - CF, DM, binder diffusion disorders, Rey's sy, Kawasaki
  - trauma and obstruction of the bile ducts can also be the cause
- **pathogenesis**: the effect of occlusion of the leads by inflammation, which is caused by a given etiological factor, is presumed, followed by activation of enzymes and escape into the stroma - the most important is the activation of trypsin and chymotrypsin
  - autoactivation is controlled by enterokinase, bile regurgitation
  - stasis destroys protease inhibitors, which otherwise prevent activation in the pancreas
  - necrosis with calcifications and saponification occurs, the process penetrates into the environment
  - in a very severe course, there is a risk of circulatory shock and ARDS
- **clinical picture**: very acute pain
  - sudden pain in the epigastrium, the patient locates it in depth, as a fickle, practically permanent pain, radiates to the back (when the cauda is affected, it may radiate to the left shoulder with irritation of the diaphragm)
  - vomiting, fever, tachycardia, tachypnoea, sweating and hypothesia prone to collapse
  - the pain is extremely intense, the child takes a crouched position, attracts the limbs to the abdomen, usually lies on his side, prevents any change of position and any examination
  - the abdomen is hard to the touch, dilated, arched above the level of the chest, sometimes resistance can be felt in the depths of the left epigastrium
  - the pain worsens, the peristalsis is weakened or absent
  - hypocalcaemia with latent or overt tetany may result from salt deposition
- the most serious form – **acute hemorrhagic pancreatitis** – is rare in childhood, may be in adolescents after drug or alcohol excess, jaundice, ascites, shock, acute renal insufficiency, DIC, gastrointestinal bleeding, respiratory disorders
- **laboratory**: vomiting leads to dehydration, hyponatremia, hypochloraemia and potassium depletion, hypocalcaemia is characteristic
  - in serum and urine the value of amylases increases within 2–12 hours (decreases in 24–48 hours) - predominance of pancreatic isoenzymes
  - increase in serum lipases - with a threefold increase, this value is more specific and sensitive than amylases
  - rising ALT , AST , LDH
  - leukocytosis and hyperglycemia
- **diagnosis**: hyperamylasemia is really important
  - imaging methods - especially valuable - abdominal sono and CT will provide the most data - pancreatic enlargement, hypoechogenic edematous pancreas, presence of pseudocysts filled with fluid or blood, duct dilatation, sometimes purulent abscesses
  - X-ray - non-specific findings - elevation of the left diaphragm arch, small pleural effusion on the left, smaller infiltrates at the base of the lungs or pulmonary atelectasis
    - dilated colon transverse, calcification in the pancreas
    - ERCP in recurrent pancreatitis
- **differential diagnosis**: other causes of pain - appendicitis, peptic ulcer, intestinal obstruction, cholecystitis
  - other causes of elevated amylases - parotid disease (mumps , EBV , CMV , HIV , sialoadenitis), eating disorders (anorexia, bulimia)
    - biliary tract diseases, some systemic conditions (burns, head injuries, renal insufficiency, MAc, transplant rejection)

    - Acute pancreatitis requires repeated surgical consultation and close cooperation with a pediatric surgeon
- **therapy**: no specific therapy
    - symptomatic: rehydration, pain relief, pancreatic secretion suppression and complication prevention and treatment
    - patients are hospitalized in the ICU and constantly monitored
    - pain relief - opiates are contraindicated because they increase the tone of Oddi's sphincter, we use tramadol the most
    - we stop the oral supply of anything to the child (this will reduce the secretion)
    - we pay attention to rehydration (we monitor CVP), we adjust the internal environment according to acute changes
    - we constantly aspirate gastric and duodenal contents with a probe
- **the prognosis of** acute hemorrhagic pancreatitis still has up to 50% mortality
    - serious prognostic indicators - high leukocytosis (above 16.10 9 / l), disproportionate glycemia, high value of LDH and aminotransferases, hypoclacemia below 2 mmol / l, hypoxia and resp. insufficiency
    - acute without hemorrhage usually resolves during conservative treatment within 2–4 days, but convalescence is long (0.5–2 years following a strict diet)

Chronic pancreatitis

- most often on a genetic basis - such as **familial chronic pancreatitis**
- **etiology**: AD, penetration is high (affects all offspring with a defective gene)
    - but the expressiveness (degree of disability) is very diverse
    - in addition, chronic relapsing pancreatitis can occur in other diseases - inherited hyperlipidemia I., IV. and V., in CF, hyperparathyroidism or as a result of pancreatic anomalies
- **clinical picture**: at first the symptoms are mild, often just dyspepsia
    - they often start at the beginning of school and are related to a change in diet (school canteen)
    - takes place in the form of short-term, several-hour seizures
    - seizures become more severe
    - chronic pancreatic insufficiency gradually develops - it suffers from fat digestion - first steatorrhea, then a picture of malabsorption syndrome
- **diagnosis**: careful family anamnesis
    - during the attack we can detect biochemical changes in pancreatitis
    - outside seizures we can detect pancreatic calcification and the development of pseudocysts
- **differential diagnosis**: we must determine chlorides in sweat, examine serum lipoproteins (exclude hyperlipoproteinemia), Ca and P concentrations, or tests for hyperparathyroidism, we examine the stool for ascaris eggs
- **therapy**: enzyme substitution, dietary measures (prohibition of alcohol and toxins, adequate protein intake, fat reduction to 35% intake, reduction of simple carbohydrates, fiber preferences (if the pancreas does not fail completely), eat smaller doses more often

Links

Source

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