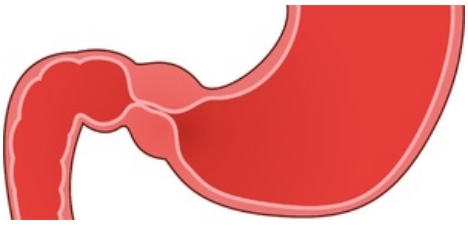
Congenital hypertrophic pyloric stenosis

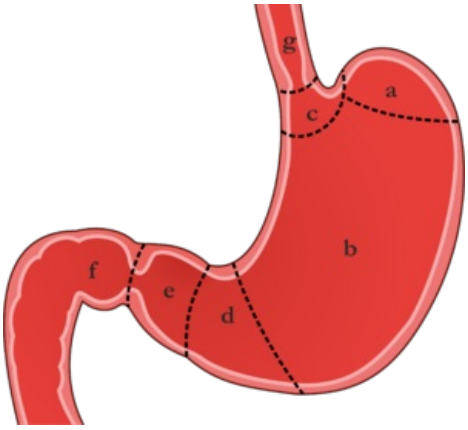
Hypertrophic pyloric stenosis is an acquired diffuse hypertrophy and hyperplasia of the smooth muscle of the pylorus and the entire stomach.



Pyloric stenosis.

Etiology

Etiology is unknown, polygenic inheritance and environment are assumed. Familial occurrence was demonstrated in 15%. It is sometimes associated with hiatal hernia, esophageal atresia or Turner syndrome. The incidence of the disease varies greatly geographically, with around 1:5000 live births in the Czech Republic, it is up to five times more often in first-born boys. Newborns and infants between 3-6 weeks of life are affected. Children older than 3 months are rarely affected.



Stomach a) fundus b) corpus c) cardia d) antrum pyloricum e) canalis pyloricus f) duodenum g) oesophagus.

Clinical picture

- Explosive **arc vomiting** dominates, projectile (up to 1 m);
- Vomit contains acidic gastric juices, usually the content is digested milk, vomit is **free of bile**;
- there is **dehydration**, the child has a large appetite, he/she drinks eagerly; he/she loses weight (dehydration, insufficient caloric intake)
- the child is lethargic, has constipated or hungry stools, has an olf-fashioned appearance;
- can lead to severe **hypochloremic alkalosis** and **hypokalemia** - severe condition, shallow breathing, loss of consciousness, convulsions - *coma pyloricum*;
- A peristaltic wave (from left to right epigastrium) can be observed on the abdomen immediately after drinking;
- with gentle palpation, about 70% of children have **palpable resistance** = olive, the size of a cherry in the epigastrium, to the right of the midline - **tumor pylori**;
- icterus may occur rarely

Laboratory examination

- typically **hypochloremic alkalosis** with **hypokalemia**, **hyponatremia** and **dehydration**;
- hypochloremia can reach extreme values (below 75 mmol / l), its degree better reflecting potassium loss than potassium;
- elevated gastrin levels

Hypertrophic pyloric stenosis

Diagnostics

- clinical picture
- abdominal ultrasound
  - the length (17 mm and more) and width (4 mm and more) of the pyloric channel are measured
  - sensitivity 97%
- X-ray contrast examination (GIT passage) is currently used for diagnostic doubts
  - stomach dilatation
  - elongated and narrow pyloric canal (so-called rail or shoelace image)
  - contrast medium must be aspirated with a nasogastric tube after examination (possible aspiration)
  - rapid passage of contrast through the stomach eliminates pyloric stenosis
  - this test may reveal other causes of vomiting without bile:
    - gastric atony
    - delayed gastric emptying,
    - gastroesophageal reflux

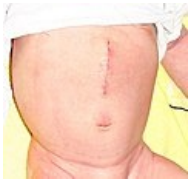
Differential diagnosis

- other causes of explosive vomiting:
  - intracranial hypertension,
  - pyloric atresia,
  - antral membrane,
  - stomach duplications,
  - gastric atony
  - delayed gastric emptying,
  - gastroesophageal reflux;

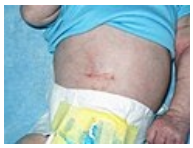
- other causes of similar metabolic breakdown:
  - acute adrenal insufficiency - bleeding or congenital hyperplasia - MAc,
  - hyperkalemia,
  - sodium loss in urine;
- hereditary metabolic disorders – Metabolic alkalosis can cause disorders of AMK metabolism (urea cycle disorder).

Therapy

- a conservative approach is not recommended
- the treatment is surgical
  - **longitudinal pyloromyotomy** of hypertrophic pyloric muscle is most often performed (Weber-Ramstedt operation):
    - it begins with a **transverse laparotomy** in the right part of the epigastrium
    - the straight abdominal muscles and the oblique abdominal muscles are intersected longitudinally
    - after opening the peritoneal cavity, a hypertrophic pylorus is luxated into the surgical wound
    - a sharp longitudinal incision is made of serosa and superficial muscle fibers (the incision starts 1-2 mm from the pyloroduodenal junction and ends in the area of the pyloric junction in the stomach)
    - the muscle fibers are separated (along the entire length of the incision) by blunt dissection
    - complication is perforation of the mucosa - this must be sutured with absorbable material and covered with omentum
  - patients with more than 5% weight loss, metabolic alkalosis and hypochloremia must parenterally rehydrate and correct the internal environment within 24 hours before surgery
- can also be performed by laparoscopic technique
- prognosis - good with timely operation.



Pyloromyotomy scar 30 hrs post-operation



Horizontal scar 10 days after pyloric stenosis surgery

References

Related articles

- Congenital atresia and stenosis of the gastrointestinal tract

- Esophageal atresia
- Atresia and stenosis of the small intestine
- Anal and rectal atresia
- Superior mesenteric artery syndrome

- Malrotation of volvulus and intestines
- Meconium ileus
- Congenital Megacolon

Source

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Literature

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- ŠAŠINKA, Miroslav, Tibor ŠAGÁT a László KOVÁCS, et al. *Pediatria.* 2. vydání. Bratislava : Herba, 2007. ISBN 978-80-89171-49-1.

- ŠNAJDAUF, Jiří a Richard ŠKÁBA. *Dětská chirurgie.* 1. vydání. Praha : Galén, 2005. ISBN 807262329X.

External references

- Pyloric stenosis– learning video (https://www.youtube.com/watch?v=GICHybVAIDs%7CPylorostenóza)
- Pyloric stenosis – Osmosis (https://www.youtube.com/watch?v=AFMtilrmLYk%7CPylorostenóza)