Megacolon congenitum

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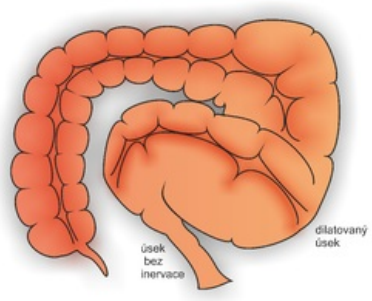
Congenital aganglionic megacolon (Hirschsprung's disease) is caused by abnormal innervation of the bowel, affecting the internal anal sphincter and the adjacent proximal segment of variable length.[1]



Megacolon congenitum

Epidemiology

- the most common cause of obstruction of the lower GIT in newborns;
- incidence 1:5000 live births;
- boys affected 4x more often than girls;
- may be associated with other birth defects, trisomy 21 or medullary carcinoma of the thyroid gland.[1]

Etiopathogenesis and pathophysiology

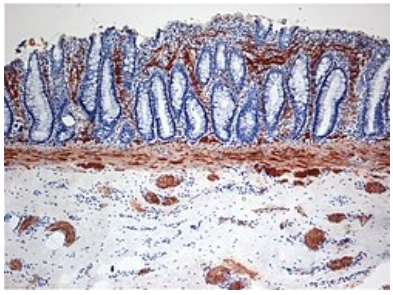
- disorder of neuroblast migration from the proximal to the distal intestine with subsequent absence of ganglion cells in the intestinal wall;
- histologically: absence of Meissner's and Auerbach's plexus with hypertrophic nerve endings and high concentration of acetylcholinesterase;
- classic form (75%): aganglionosis in the rectosigmoid; in 10% the entire colon is affected; ultrashort form: aganglionic section in the rectum 1-3 cm long;
- the aganglionic section is permanently contracted (missing inhibitory neurons) and causes functional obstruction → the healthy intestine above it dilates and hypertrophies to form a megacolon.[1]

Clinical picture

- in 90% of cases, manifestation immediately after birth: late departure of the pitchfork;
- milder forms: chronic constipation, failure to thrive, gradually developing abdominal distension with overgrowth of pathogens and symptoms of enterocolitis to sepsis;
- the onset of difficulties in infancy is typical (often after the introduction of non-dairy foods): increased tone of the internal sphincter, small volume of stool during defecation, smearing does not occur;
- for ultra-short English section, stool accumulates in the rectum, the sphincters gradually weaken, soiling + paradoxical diarrhea (dirty laundry).[1]

Diagnosis

- irrigography – after previous emptying of the intestine with enemas, delayed evacuation, transition zone between the narrow distal aganglionic segment and the proximally dilated section of the intestine; defecogram;
- anorectal manometry – measurement of anal pressure when a balloon is inflated in the rectum (anal pressure does not decrease, or paradoxically increases);
- rectal biopsy (may be false negative for ultrashort segment).[1]


Histopathological section of Hirschsprung's disease showing fibers containing abnormal ACHE (brown) in the lamina propria mucosae.

Therapy

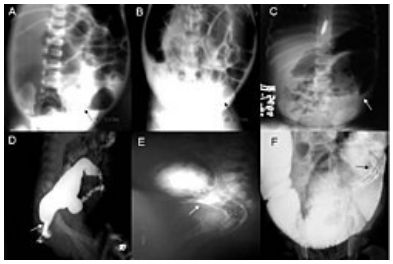
- surgical solution; prognosis is good, most patients maintain continence.[1]

Complications

- toxic megacolon with a septic course with the risk of secondary meningitis or bowel perforation.[2]

Notes

- *Megacolon idiopathicum*: disorder of autonomic innervation - imbalance between sympathetic and parasympathetic.
- *Megacolon symptomaticum*: dilatation over a stenotic site - e.g. congenital stenosis, scar after surgery...[3]



Hirschsprung's disease
A–C Abdominal plain X-ray
D–E Contrast-enhanced X-ray

Links

Related Articles

- Congenital atresias and stenoses of the gastrointestinal tract
  - pylorostenosis congenita
  - arteriae mesentericae superioris syndrome
  - intestinal malrotation and volvulus
  - small bowel obstruction
  - meconium ileus

References

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3. BENEŠ, Jiří. *Study materials* [online]. ©2007. [cit. 2010-04]. <http://www.jirben.wz.cz/>.