

HIRSCHSPRUNG'S DISEASE

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Abstract Hirschsprung's disease is a form of mega colon that occurs when part or all of the large intestine or antecedent parts of the gastrointestinal tract have no ganglion cells. A number of genes are associated with Hirschsprung's disease. Hirschsprung's disease, the nerves that control this movement are missing from a section at the end of the bowel. Signs and symptoms are Swollen belly, Chronic constipation, Gas, Failure to thrive, Fatigue. Treatment of Hirschsprung's disease consists of surgical removal (resection) of the abnormal section of the colon, followed by reanastomosis.

Key words: Megacolon, large intestine, gastrointestinal tract, ganglion cells, Signs and symptoms, constipation, reanastomosis.

1. INTRODUCTION

Hirschsprung (HERSH-sproong) disease affects the large intestine (colon) of newborns, babies, and toddlers. The condition which prevents bowel movements (stool) to pass through the intestines due to missing nerve cells in the lower part of the colon is caused by a birth defect. Most of the time, the problems with pooping start at birth, although in milder cases symptoms may appear months or years later. Hirschsprung disease can cause constipation, diarrhea, and vomiting and sometimes lead to serious colon complications, like enterocolitis and toxic mega colon, which can be life-threatening [1]. So it's important that Hirschsprung disease be diagnosed and treated as early as possible.

2. DEFINITION

Hirschsprung's (HIRSH-sproongz) disease is a condition that affects the large intestine (colon) and causes problems with passing stool.

3. CAUSES:

In Hirschsprung's disease, these ganglion cells are missing from a section at the end of bowel, extending up from the anus, the opening in the bottom that poo passes through. A number of genes are associated with Hirschsprung's disease, genetic condition such as Down's syndrome[2].

4. PATHOPHYSIOLOGY:

In Hirschsprung's disease, the nerves that control this movement are missing from a section at the end of the bowel, which means poo can build up and form a blockage. This can cause severe constipation, and

occasionally lead to a serious bowel infection called enterocolitis[4] if it's not identified and treated early on.

5. SIGNS AND SYMPTOMS:

5.1 In newborns

- failing to pass meconium within 48 hours – the dark, tar-like poo that healthy babies pass soon after being born
- a swollen belly
- vomiting green fluid (bile)
- diarrhea

5.2 In older children

- Swollen belly
- Chronic constipation
- Gas
- Failure to thrive
- Fatigue

6. DIAGNOSIS

- rectal examination
- X-ray
- rectal biopsy,
- Microscopic examination
- Measuring control of the muscles around the rectum.

7. TREATMENT

- "pull-through" operation - affected section of bowel is removed and the remaining healthy sections of bowel are joined together.
- Ileostomy, removes the entire colon and connects the small intestine to the stoma. Stool leaves the body through the stoma into a bag.
- Colostomy, leaves part of the colon intact and connects it to the stoma. Stool leaves the body through the end of the large intestine.

7.1 Risks of surgery

- bleeding during or after the operation
- the bowel becoming infected (enterocolitis)
- Bowel contents leaking into the body, which could lead to serious infection (peritonitis) if not treated quickly[3].
- The bowel becoming narrowed or blocked again, requiring further surgery.

CONCLUSIONS

Hirschsprung's (HIRSH-sproongz) disease is a condition that affects the large intestine (colon) and causes problems with passing stool. A number of genes are associated with Hirschsprung's disease. Hirschsprung's disease, the nerves that control this movement are missing from a section at the end of the bowel. Signs and symptoms are Swollen belly, Chronic constipation, Gas, Failure to thrive, Fatigue. Treatment of Hirschsprung's disease consists of surgical removal (resection) of the abnormal section of the colon, followed by reanastomosis.

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