Case #4

International Colorectal Web Meetings

Lea Wehrli MD

Children's Hospital Colorado

Medical History



- Born full term, meconium within first 48 hours of life
- Bloody stools at 1 month, formula was switched and diagnosed with food protein proctocolitis with no further bloody stools
- Worsening constipation since one year of age

Medical History



- 1x/week, strains a lot, explosive, watery stool, sometimes clay-like. Pale gray or yellow. Never formed stools
- Tried laxatives with no improvement
- Normal growth and development

Clinical exam



- Abdomen distended but soft
- Decreased bowel sounds
- Anus orthotopic located
- No signs for spinal dysraphism







What is your differential diagnosis?

- 1) Chronic idiopathic constipation
- 2) Hirschsprung Disease
- 3) ARM with perineal fistula
- 4) Anal stenosis



- 1) Exam under anesthesia, full thickness biopsy with general anesthesia
- 2) Contrast enema
- 3) Exam and suction rectal biopsy in outpatient clinic without anesthesia
- 4) 1+2
- 5) 1+3





IONAL CENTER FOR RECTAL AND NITAL CARE





NAL CENTER FOR CTAL AND ITAL CARE





ATIONAL CENTER FOR DRECTAL AND ENITAL CARE



Histopathology

Rectum, biopsy:

- No ganglion cells identified
- Negative for calretinin
- No hypertrophic nerve fibers seen



.



What is your diagnosis?

- 1) Hirschsprung Disease
- 2) Chronic idiopathic constipation
- 3) Anorectal malformation with a perineal fistula



Significance



- No calretinin-positive fibers in the lamina propria
 - means; that there are no fibers seen from the ganglion cells
- No hypertrophic nerve fibers seen
 - means; that it might be a longer segment of aganglionosis
 - Cave; hypertrophic nerve fibers >40um is only valid in patients younger than 1 year of age, there are no norm values for older patients



Medical treatment

- 1) Rectal enemas
- 2) Rectal irrigations
- 3) Botox injection into the sphincter area



Surgical treatment



Transanal Swenson technique

- Incision made 1cm above the pectinate line
- Biopsy taken at 5cm and 25cm (30cm of colon resected)
- Proximal donut with ganglion cells in all of the circumference of the submucosal and myenteric plexus
- Colorectal anastomosis performed







Thank You!