Rectal atresia



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Girl, 36+2 weeks of gestation

- Birth weight 2.6kg, length 46cm, head circumference 34cm
- 33 weeks of gestation
 - Prenatal US findings:
 - Bi-carotid aortic trunk
 - Polyhydramnion and double bubble sign
- 35 weeks of gestation
 - Amniocentesis and drainage of 2000ml
 - 46 XX, arr 5q33.3q34 (6.2Mb)



Clinical findings

- Facial dysmorphism (low set ears, lateral downward deviation of the lid axis)
- Bilateral partial cutaneous syndactly of the toes II/III
- Nurses report difficulties to pass the rectal temperature probe farther than 1cm
- Clinical exam with a normal looking anus





What diagnostic workup would you do?

- a) ECHO
- b) US Head
- c) Babygram
- d) US spinal cord
- e) All of the above

Workup

- X-ray pelvis: Abnormal sacral segmentation S2/S3
- ECHO: Bicarotid aortic trunk and ASD
- US head: normal
- US abdomen: normal kidneys, fluid filled stomach and duodenum, collapsed small bowel
- US spine: normal spine and cord, no signs of tethering, no presacral mass





What kind of ARM do you suspect?

- a) Anal atresia without fistula
- b) Anal atresia with fistula
- c) Rectal atresia
- d) Posterior cloaca

Age of 3 days

- Intraoperative examination
 - Anus normal with good circumferential sphincter contractions
 - Anus can be easily intubated with a Hegar of 11 but not farther introduced than 2cm
 - Distal rectum blind ending 2 cm above the pectinate line



Age of 3 days

Diagnostic laparoscopy with conversion to midline laparotomy

- Duodenoduodenostomy
- Descending colostomy



What diagnostic workup would you do?

a) Contrast enema and distal colostogram

b) Distal colostogram while having a Hegar inserted through

the anus

c) MRI pelvis

d) US of the perineum





How would you surgically approach it?

a) Transanal resection and end-to-end rectorectal anastomosis

b) PSARP

c) Laparoscopic assisted transanal resection and end-to-end rectorectal anastomosis
d) I don't know

Surgical treatment at age of 6 months

- The patient underwent a laparoscopic-assisted transanal resection
- Intraoperatively the distal rectum was in continuum to the proximal rectum separated by a septum
- Starting rectal dilations 2 weeks postoperatively



A.Pena, A.Bischoff, Surgical treatment of Colorectal Problems in Children, DOI 10.1007/978-3-319-14989-9_14