

Case Report

Primary repair of rectourinary fistula in a patient with associated esophageal atresia

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PRENATAL DIAGNOSIS: polyhydramnios, suspicion of esophageal atresia

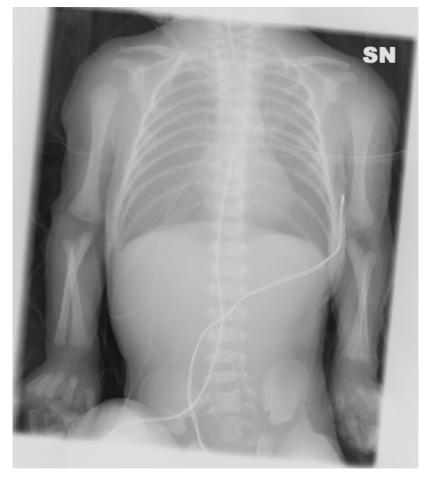
38 weeks

Birth weight: 2070 g

At birth:

- Esophageal atresia
 with no distal fistula
- Anorectal malformation

 (evidence of meconium in urine)



SCREENING FOR ASSOCIATED ANOMALIES:

- Supernumerary lumbar vertebra
- Sacral ratio: AP 1, LL 0.95
- Tethered cord
- Right kidney agenesis
- No cardiac abnormalities

What would you do as a first operation?

- 1. Tracheoscopy, gastrostomy, and colostomy
- 2. Tracheoscopy, esophageal atresia repair, and colostomy
- 3. Tracheoscopy, gastrostomy, and anorectal malformation repair
- 4. Tracheoscopy, esophageal atresia repair, and anorectal malformation repair
- 5. Manage the esophageal atresia only
- 6. Manage the ARM only

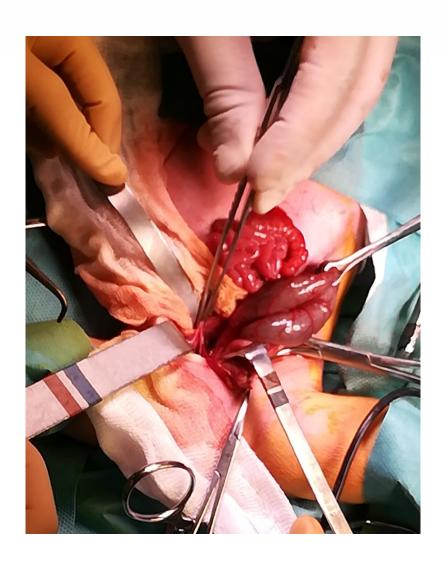
Treatment of Esophageal Atresia and ARM

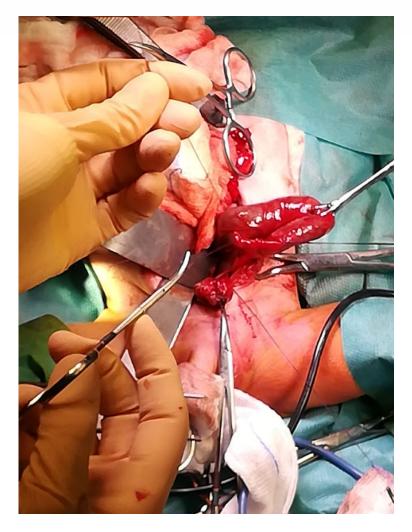
SURGERY: 1° day of life

✓ Tracheoscopy: proximal tracheo-esophageal fistula

✓ Laparotomy for gastrostomy

At laparotomy a recto-prostatic fistula was found and a primary repair of the anorectal malformation--combining abdominal and perineal approach--was performed







POST-OP:

- Minimal dehiscence of the posterior perineal body, managed conservatively
- Mild rectal prolapse (right > left)

He underwent a dilation program reaching Hegar size 12

1 month of life:

SURGERY

Thoracoscopic ligation of the proximal tracheo-esophageal fistula

The esophageal stumps were too distal to achieve anastomosis, so a cervical esophagostomy was opened

The patient was discharged

He currently is 5 months old

- Tolerating feedings via gastrostomy
- Oral stimulation
- 2-3 bowel movements/day
- The prolapse improved, currently minimal right hemiprolapse

We are planning to perform an endoscopic evaluation of the gap between the two esophageal stumps to plan the next surgical step.

How do you manage long-gap esophageal atresia in patients with ARM?

- 1. Gastric pull-up
- 2. Jejunal interposition
- 3. Colonic interposition
- 4. Other

The three-stage surgical correction of ARM with recto-urinary fistula (colostomy, PSARP, and colostomy closure) has been known to be the most effective approach in preventing complications

The colostomy allows to perform the distal colostogram, thus helping to define the anatomy of the malformation

However primary repair represents an attractive choice:

- -reduces the number of operations (from 3 to 1)
- -avoids colostomy; thus, it reduces the familiar discomfort in terms of management, psychological and economic burdens
- -allows the early passage of stools, with a theoretical early establishment of the brain-defecation reflex
- -avoids continued urinary tract contamination through the fistula

In addition, in patients with associated long-gap esophageal atresia....

..the primary repair of the ARM leaves an intact colon that might be considered as an option for esophageal replacement

Primary repair of recto-urinary fistula should be performed only by expert surgeons with adequate skills

In primary repair, the combination of abdominal and perineal approach seems to be more effective to prevent complications, because it allows the bowel to decompress with enterotomy and to follow the rectum to the fistula

Further studies are needed to evaluate if the incidence of surgical and functional complications is similar to the traditional staged management



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ORIGINAL ARTICLE

Single-Stage Surgical Correction of Anorectal Malformation Associated with Rectourinary Fistula in Male Neonates

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ABSTRACT

Introduction: The treatment of children affected by ano-rectal malformations (ARM) is characterized by some unsolved problems. The three-stage surgical correction has been known to be most effective in preventing complications, but recently new approaches have been proposed. We describe our experience with the newer approaches.

Methods: Twenty three male newborns, affected by ARM and recto-urinary fistula, were treated in 2 different centers in 8 years. Nineteen neonates (birth weight 2.4 - 3.5 kg) received a primary posterior sagittal anorectoplasty (PSARP) at the Department of Pediatric Surgery of the Chittagong Medical College Hospital (group 1). Four term neonates (birth weight 2.9 - 3.4 kg) received a primary pull-through with combined abdomino-perineal approach at the Pediatric Surgery Department of Fondazione Cà Granda of Milan (group 2).

Results: Among patients of Group 1, 11 patients had a recto-bulbar fistula and 8 a recto-prostatic fistula. Among the Group 2, 2 had a recto-bulbar fistula and 2 a recto-prostatic fistula. The site of fistula was decided at the time of surgery. In Group 1, 5 post-surgical complications were recorded (26%); 1 child died of sepsis, 3 had dehiscence and 1 stenosis, which resolved with dilatation. In Group 2, the only post-operative complication of small rectal prolapse resolved spontaneously after a few months on follow-up. Group 2 patients were followed-up in a dedicated multidisciplinary colorectal center.

Conclusions: Primary repair of ARMs with recto-urinary fistula is a feasible, safe and effective technique in the neonatal period. A combined abdominal and perineal approach seems to guarantee better results. A dedicated team is mandatory, both for the surgical correction and for a long-term follow-up.

Key words: Anorectal malformation, Primary repair, Recto-urinary fistula

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Thank you