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Introduction:

Anorectal malformation is a relatively common congenital anomaly, with an incidence of approximately 1 in 4,000 live births in sub-Saharan Africa. Early management typically involves the creation of a **divided sigmoid colostomy with mucous fistula**, particularly in cases where definitive surgical correction is not immediately achievable or when there is a concern regarding fecal contamination of the urinary tract, as seen in high or complex malformations.

In the best-case scenario, children with anorectal malformation are initially evaluated by a specialist pediatric surgeon. In some cases (perineal or vestibular fistula) the best treatment will be immediate repair without diverting colostomy. This chapter is intended for surgeons caring for children who do not have access to a specialist pediatric surgeon.

A divided proximal sigmoid colostomy with mucous fistula is performed in the neonatal period for multiple reasons:

- It diverts stool to relieve obstruction and allow feeding.
- The location preserves adequate length of distal colon for an eventual anoplasty.
- The stoma is not likely to prolapse because the descending colon is fixed in a retroperitoneal position.
- Stool diversion reduces contamination of the urinary tract when a urinary fistula is present.
- The mucous fistula can be used later for a distal pressure colostogram (contrast study) to assess the level of the malformation and identify any non-apparent connection to the genitourinary system.

The diagnosis of anorectal malformation is frequently established in the neonatal period. Typical presentation includes failure to pass meconium within the first 24 to 48 hours of life, and an absent or abnormal anal opening on routine physical examination. In some cases when there is a large fistulous opening, anorectal malformation may not be diagnosed until later in infancy or early childhood, when the stool becomes more solid and of larger

volume leading to obstruction or progressive fecal retention.

Upon identification of an absent anal opening, a comprehensive, systematic physical examination is essential to diagnose anorectal malformations and assess for associated "VACTERL" anomalies (Vertebral, Anorectal, Cardiac, Tracheoesophageal, Renal, and Limb defects). Typically the first 24 hours of care is dedicated to this crucial workup.

Key Examination Steps:

- 1. General Assessment: Evaluate the child's overall health, abdominal distension, and signs of distress.
- 2. Cardiac Evaluation: Auscultate for murmurs and consider echocardiography for congenital heart defects if available.
- 3. Tracheoesophageal Fistula: Insert a nasogastric tube to assess for failure to pass or coiling, which may indicate esophageal atresia or tracheaesophageal fistula.
- 4. Renal and Urinary Tract: use pelvic ultrasound and voiding cysto-urethrogram for further evaluation.
- 5. Perineal Examination: Inspect for absent anal opening, genital abnormalities, and possible fistulas (rectoperineal, rectovestibular).
- 6. Limb and Spinal Evaluation: Look for sacral dimples or hair tufts indicating possible spinal issues. Consider sacral X-rays and spinal ultrasound.

Adjunctive Investigations:

- Echocardiography for cardiac anomalies.
- Anteroposterior whole trunk x-ray ("baby-gram") for assessing bowel obstruction and to look for coiling nasogastric tube.
- Sacral X-rays and spinal ultrasound for spinal and sacral anomalies.

Steps:

1. After diagnosis of anorectal malformation, screening for associated congenital anomalies, and adequate resuscitation, the patient may be prepared for divided sigmoid colostomy and mucous fistula.





Suhail Sherman, Tabitha Nyakerario, Jason Axt

- 2. General anesthesia is induced, and a caudal block performed. Local anesthesia can also be given if there is not expertise for a caudal block.
- 3. The patient is positioned supine and prepared from the subcostal margin to the mid-thigh using warm agent and overhead heaters to prevent hypothermia. If a plastic cord clamp is in place, it should be removed.
- 4. The incision site is planned; a line is drawn from the left anterior superior iliac spine to the umbilicus. The incision will be made at a right angle to this line ½ from the umbilicus.



Marking for planned incision. Upper brackets mark end descending colostomy, lower brackets mark mucous fistula

5. The superior incision is made using a #15 blade.



Incision for proximal stoma

6. The superficial subcutaneous facias are lifted and carefully divided between forceps. The same

technique is used at the level the rectus sheath and peritoneum to ensure safe entry into the peritoneal cavity.



Toothed forceps are used to grasp and lift Scarpa's fascia. This technique will be repeated at every level from here to the peritoneum.

7. On peritoneal entry, small bowel is usually encountered and can be packed away using a malleable retractor and moist gauze. The gauze is introduced under the peritoneum towards the left para-colic gutter and is then retracted medially to allow visualization of the sigmoid colon.



A malleable retractor folded into a "L" shape and moist gauze can be used to retract small bowel medially until the sigmoid colon is seen.

8. The sigmoid colon can be identified by the proximal retro-peritoneal attachments. The distal





Suhail Sherman, Tabitha Nyakerario, Jason Axt

portion will extend into the pelvis. It is imperative that the sigmoid colon is correctly identified and oriented. Taenia coli, haustra and fatty appendages are not always obvious in newborns. If omentum is attached to the colon, the segment is the transverse colon and not the sigmoid. Twisting must also be avoided, and the most proximal part exteriorized as the stoma. This is to ensure adequate length is available for future anorectoplasty.



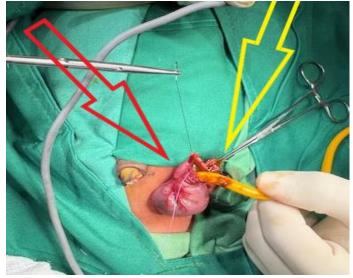
The sigmoid colon is labeled with arrows, proximal in black and distal in red

9. If there is difficulty visualizing the sigmoid colon or understanding its orientation, the incision can be extended to its inferior limit as shown below. The skin and fascia between the two ostomies is closed later.



The previously drawn line can be extended to its full length if the identification or orientation of the sigmoid colon is not clear.

- 10. The mesentery and bowel lumen are divided as proximally as possible.
- 11. A non-crushing clamp (Babcock) is placed on the proximal colon to keep it from slipping into the abdomen.



Babcock clamp is on the proximal sigmoid colon (Yellow arrow). The lumen of the distal colon (Red arrow) is narrowed to accommodate a 14 French irrigation catheter.

12. A 14 French tube of any type is introduced into the distal colon and 4-0 absorbable suture is used to reduce the lumen of the distal colon until the size of the catheter is achieved.



Suhail Sherman, Tabitha Nyakerario, Jason Axt



Black arrow shows a Babcock clamp grasping proximal stoma. The Yellow arrow shows a 14 Fr Foley catheter instilling saline into the distal lumen. Note that after narrowing of the distal lumen as described above, the final suture is left attached to a clamp and used as a stay suture, providing traction and guiding any spilled enteral contents into the kidney dish.

13. Irrigate and aspirate the distal colon and rectum. Stay sutures around the enterotomy can be used to guide the colon into a kidney dish where enteric contents will be irrigated. A 60cc syringe is coupled with the 14 French catheter and warm saline is instilled into the distal colon. It is then aspirated and expelled into the kidney dish to be suctioned. One operator can instill and aspirate saline as the assistant suctions the enteral contents from the kidney dish.



Saline is aspirated from the distal lumen, note the aspirated fluid becoming clearer.

- 14. Irrigation is continued until the saline aspirated from the distal colon is clear, this is imperative to prevent formation of hardened stool (concretion) which will complicate future anoplasty.
- 15. The proximal stoma is anchored to the fascia on four points circumferentially at the superior pole of the incision.



Fascia to seromuscular sutures are placed about 0.5 cm from the mucosal edge on four sides securing the bowel to the fascia

16. The distal mucous fistula is anchored to the fascia at the inferior pole of the incision, and the fascia between is reapproximated.

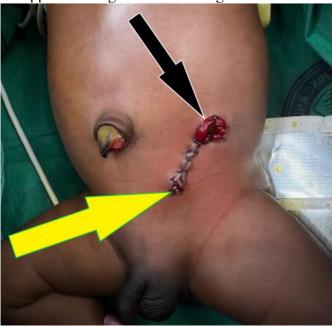


Proximal stoma and distal mucous fistula with fascia closed between



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17. The skin is then closed between the two ostomy sites; skin sutures are added circumferentially approximating the mucosal edge to the skin.



Skin sutured between proximal stoma (Black arrow) and distal mucous fistula (Yellow arrow).

Pitfalls

- The stoma must be placed in the most proximal sigmoid colon (at the junction between descending colon and sigmoid colon) or there may not be length to later perform anorectoplasty.
- The proximal sigmoid must be appropriately identified and matured as a stoma. If the proximal stoma and mucous fistula are reversed, intestinal obstruction will result.
- Failure to mature the mucous fistula well can lead to stricture and require revision.
- If the distal sigmoid colon is not adequately irrigated, a "fecaloma" will form, complicating the future anoplasty.

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