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ANORECTAL MALFORMATION IN PEDIATRIC PATIENT: A CASE REPORT

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ABSTRACT

Anorectal Malformations are the birth anomalies in which the rectum and anus are malformed, do not develop properly. They may be born with either anal agenesis (or) small opening in the wrong place (or) narrowed opening at the bottom. The main symptoms include new born do not pass stools even after 24-48 hours of birth, opening to the anus is either absent or is displaced to different location, stool may pass out, either through vagina, base of the penis, scrotum or urethra, presence of fistula between the rectum, reproductive organ and urethra is observed, presence of meconium in urine. A 1 year old female patient came with the known case of anorectal anomaly with colostomy insitu came for colostomy closure. She also had a past medical history of anorectal malformation stage-1 surgery at 1 month of age, cleft lip repair at 6 months of age, anorectal malformation stage-2 surgery at 7 month of age. The patient had no family history. The treatment option for imperforate anus is surgery, here in this patient Posterior Sagittal AnoRectoPlasty (PSARP) was done, it mainly involves 3 stages stage-1 is colostomy, stage-2 is PSARP, stage-3 is colostomy closure, following the patient was discharged upon recovery. This case underscores the importance of anorectal agenesis in pediatric patient and should be diagnosed and treated immediately after the baby is born. Early diagnosis and treatment are needful to prevent further complications like intestinal obstruction, bloating, not able to pass stools.

KEYWORDS: Anorectal malformations (ARMs), Anal agenesis, Imperforate anus, Fistula, Posterior Sagittal AnoRectoPlasty (PSARP), Colostomy closure, Congenital anomalies.

INTRODUCTION

Anorectal imperforations are a prevalent significant congenital anomaly that affect around 1out of every 5,000 live infants due to irregularity in the way the hindgut develops from the cloaca during embryonic development.[1,3] The clinical diagnosis can be established by presenting signs of imperforate anus, colonic obstruction, and failure of meconium passage.[1] Prenatal imaging may indicate ARM, but a postpartum physical examination usually confirms the diagnosis.[3] A rectovaginal fistula is the most typical form of ARM in female patients. It has also been documented that rectovaginal fistulas are used as a neovagina in conjunction with posterior sagittal anorectoplasty (PSARP).[5] PSARP was created by Dr.Alberto Pena, MD, Director, International Centre for Colorectal Care.[6,7] It include 3 operations, the first one is colostomy the surgeon cuts the colon into two and brings each end through the abdominal wall forming 2 openings this allows the stool to temporary pass into a bag outside of the body. A few weeks later surgeon use the PSARP or Pena procedure to place the anus in the correct location they start by removing the connection with the urinary or genital tract and pulling the rectum through the midline of the sphincter muscles to connected to the anus this forms a new path for stool to leave the body. Once the child is healed the final operation has to be done i.e. colostomy closure during this operation the colostomy bag is removed and the surgeon reconnects the two ends of the colon after this final surgery the child is able to have a bowel movement. We present a case of a 1-year-old female patient with Anorectal Malformation.

CASE PRESENTATION

A female infant was delivered at 40 weeks of gestation with good birth weight of 2.86 kg. The female baby was born via NVD (Normal Vaginal Delivery). The patient mother noticed that the child was passing stools from urogenital region and cleft lip and cleft palate was present at birth. On admission the heart rate and respiratory rate was found to be 140bpm & 51cpm respectively. Through the physical examination the patient was recognized with ARM with imperforate anus with rectovaginal fistula. The baby was started on IV antibiotics (Inj. Meropenem 60mg 1-1-1 and Inj. Metrogyl 6ml 1-0-1) and symptomatic treatment given. After thorough discussion, mother agreed to the operation and signed formal consent for the procedure. A surgeon performed laparotomy with sigmoid divided colostomy+ genitoscopy+ cystoscopy and catherization was done and she was transfused with 10% PCV after the procedure. The patient is haemodynamically stable and discharged and they were advised to review after 6 months in plastic surgery OPD.

At 6 months of age she was undergone cleft lip repair and after 1 month i.e., at 7 months of age anorectal malformation stage-II surgery[PSARP] was done. In stage-II the surgeon use the Pena procedure to place the anus in the correct location they start by removing the connection with the urinary or genital tract and pulling the rectum through the middle of the sphincter muscles to connected to the anus this forms a new path for stool to leave the body. She was discharged at 7 months of age.

The infant was seen at the age of one year and got admitted to paediatric department who is a known case of anorectal abnormality with rectovaginal fistula came for colostomy closure. In general physical examination, blood pressure was found to be 100/70 mmhg, Pulse rate 110bpm, Respiratory rate 40cpm and temperature is afebrile. In the systemic examination, CVS- S1S2+ no murmur, CNS- conscious, alert, active, no neurological defect, P/A- normal, colostomy insitu 3x3 cm, no s/o infectious stools, RS- B/L AE+, B/L NVBS+, no added sounds. Through the relevant examinations the patient was detected with High ARM with Post PSARP. The patient was shifted to OT for laparotomy and colostomy closure which was the stage-III surgery of anorectal malformation during this operation the colostomy bag is removed and the surgeon reconnects the two ends of the colon after this final surgery the child is able to have a bowel movement. After the patient's symptoms improved, a thorough follow-up plan was provided upon discharge.

DISCUSSION

This case underscores the prominence of anorectal malformation in pediatric patient. The patient was ascertained with high congenital anorectal anomaly with rectovaginal fistula since the patient mother noticed that the child was passing stools from vagina and cleft lip palate at birth. The only treatment for congenital anorectal defect is surgery. Here, in this patient PSARP was done.

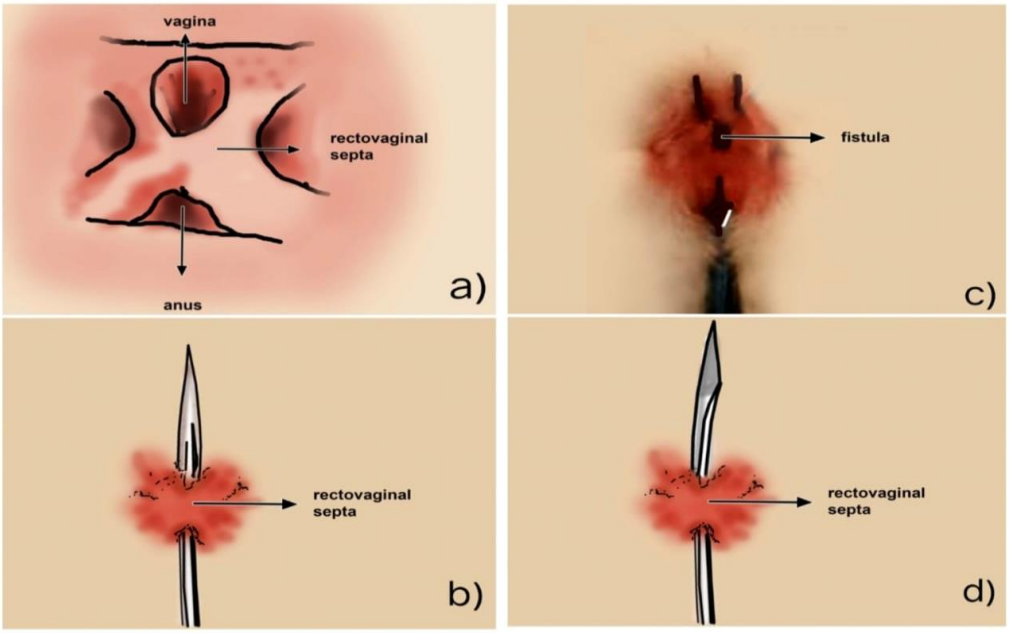

Figure 1: Step-by-step Illustration of PSARP for correcting Rectovaginal fistula.

Image (a): Identifying anatomy before surgery

- It represents the patient's preoperative anatomy with a rectovaginal fistula.
- The vagina is labeled, and the anus is absent in its normal position.
- The rectovaginal septum (the thin layer separating the rectum and vagina) is marked, showing the defective barrier.

Image (b): Dissection of the Rectovaginal Septum

- A scalpel is depicted dissecting the tissue between the rectum and vagina to separate them.
- The rectovaginal septum is reconstructed to create a clear anatomical distinction between the two structures.
- Dotted lines represent the area where precise dissection and separation occur.

Image (c): Identification of Fistula

- A fistula (abnormal connection between the rectum and vagina) is clearly marked.
- The arrow indicates the site where the rectum has abnormally fused with the vagina.

Image (d): Reconstruction of the Rectovaginal Septum

- After separating the fistula, the rectovaginal septum is reconstructed.

- The rectum is now anatomically separated from the vagina and aligned for its proper position.
- The surgical instrument demonstrates the controlled dissection and positioning of the rectum.
- The rectum is pulled down to its new position through the sphincter muscle complex to create a functioning anal opening.

PSARP is a new and widely accepted technique for repairing ARMs was developed by Pena.[7] In short it is also called as Pena procedure. Colostograms were ideally conducted on children with intermediate or high deformities in order to identify the kind and location of any fistulas before they were repaired. Peri-operative antibiotics were administered to all patients who experienced bowel washout through the stoma.[8] The surgical technique followed Pena et al. [Pena and deVries] with some modifications. Pinprick or digital stimulation was used to identify the region of maximal sphincter contraction prior to surgery while under a mild anaesthetic. Since there was no muscle stimulator accessible, the muscle fibres were identified during the procedure both visually and by low current diathermy. Sutures are used on both sides of the posterior sagittal incision to identify the muscle groups. The levator muscle and the sphincter complex are rebuilt around a size 24 or 26 Foley's catheter, placed as deeply into the pelvis as feasible, in cases where the rectal pouch is still too high for mobilization through the perineal wound. After that, the patient is put in the lithotomy posture. The previously implanted catheter is currently being employed to position the rectum towards the perineum.

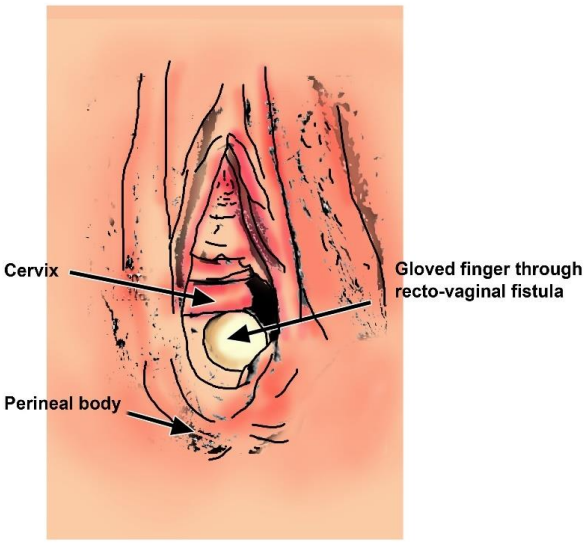

Figure 2: Visualization of a gloved figure through a Recto-Vaginal Fistula.

Where the neo-anus is formed, following a laparotomy with intra-abdominal mobilization of the rectal pouch.[7,8] Provided that wound healing had occured, postoperative dilatation was started 10–14 days after surgery and continued every day until the colostomy was closed. Once the wound is healed the patient is discharged after the anus can accept the maternal little finger.

CONCLUSION

The case highlights the critical need for early diagnosis and surgical intervention in paediatric patients with anorectal malformation(ARM) and associated conditions such as imperforate anus and rectovaginal fistula. The patient in this case underwent the Posterior Sagittal Anorectoplasty(PSARP), commonly known as the Pena procedure, which is a widely accepted surgical technique for repairing ARMs. Preoperative assessments, including colostograms, were essential for identifying fistulas. The surgery involved precise identification and reconstruction of muscle groups to create a functional neo-anus, followed by a carefully managed postoperative regimen including dilatation and eventual colostomy closure. The successful outcome of this surgery underscores the importance of meticulous surgical planning and postoperative care in managing complex anorectal malformations.

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CONFLICT OF INTEREST

The authors declare that there is no conflict of interest.

PATIENT CONSENT

The patient addressed in this case report has provided consent for publishing, acknowledging the report's nature and understanding that their identity will be kept confidential.

ABBREVIATIONS

ARM- AnoRectal Malformations; **PSARP**- Posterior Sagittal AnoRectoPlasty; **NVD**- Normal Vaginal Delivery; **PCV**- Packed Cell Volume; **CNS**- Central Nervous System; **CVS**- Cardiovascular System; **P/A**- Per Abdomen; **NVBS**- Normal Vescicular Breath Sound.

SUMMARY

Anorectal malformations (ARM) are congenital anomalies affecting about 1 in 5,000 infants due to abnormalities in hindgut development from the cloaca during embryonic stages. Diagnosis is typically confirmed postnatally through symptoms like imperforate anus, bowel obstruction, and absence of meconium passage. Prenatal imaging can sometimes indicate ARM, but a physical exam after birth is crucial for diagnosis. In females, recto vestibular fistula is a common type of ARM where the rectum connects to the vestibule rather than the anus. Treatment often involves the posterior sagittal anorectoplasty (PSARP) procedure developed by Dr. Alberto Pena, which includes stages like colostomy to divert stool temporarily, surgical repositioning of the rectum, and closure of the colostomy to restore normal bowel function.

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