Rectal Atresia an uncommon entity: Experience of three cases

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ABSTRACT
Background: Rectal atresia (RA) is a unique and rare congenital malformation contributing to about 1% of anorectal malformations. It may be associated with an abnormal sacrum and a presacral mass. The sphincter mechanism in these cases is well developed. Different surgical procedures have been described for correction of this anomaly, with variable outcomes.

Case Presentation: We present three cases of rectal atresia with their management. All the patients presented in early neonatal life with failure to pass meconium. All the patients underwent colostomy in neonatal life as initial management. Definitive surgery was performed later in all patients. All the patients are doing fine on follow-up with good continence.

Conclusion: The Posterior Sagittal Approach with the division of septum/tissue between the rectum and anal canal and end-end anastomosis gives excellent results.

INTRODUCTION
Rectal atresia is a rare anomaly with atresia or stenosis near the dentate line. The space is either separated by a septum (which may be perforated) or at times by thick tissue. The sphincter mechanism is normal in most cases signifying better postoperative bowel control.[1] The baby is born with a normal anal opening with failure to pass meconium and abdominal distension. An anal dilator can usually be negotiated up to 1.5 to 3cm from the anal verge. Initial management is a colostomy, followed by a definitive procedure and stoma closure. Herein, we report our experience with three cases of rectal atresia.

CASE SERIES
Case 1: Three days old male baby presented with abdominal distension, bilious vomiting, and non-passage of meconium. Initial examination revealed normal perineum, genitalia, and anus with abdominal distension. A blind-ending rectum was found on digital rectal examination (DRE). X-ray abdomen showed distal gas reaching up to the tip of the coccyx. Hegar dilator could be negotiated only up to 3 cms from the anal verge. Initial ultrasonography (USG) revealed no associated anomaly. A sigmoid colostomy was performed. A distal colostogram with a Hegar dilator in the anal canal was performed which confirmed the diagnosis of rectal atresia (Fig. 1). A definitive operation was performed at the age of three months. At five years old, the baby had normal continence.

Case 2: A 30-hour old, full-term male baby weighing 3 kg, presented to us with abdominal distension and failure to pass meconium. DRE revealed blind ending
anal canal 1.5 cms above the pectinate line. A sigmoid colostomy was performed. At 3 months of age, a colostogram delineated a blind-ending rectum. This patient was operated on by posterior sagittal approach with the division of septum and end-end anastomosis preserving the dentate line (Fig. 2). Anal dilatation was performed as per protocol. The colostomy was closed at 7 months of age. Postoperatively patient had good continence at 4 years of follow-up.

**Case 3:** A 20-hour female baby born via lower segment Cesarean section (LSCS) presented with abdominal distension and vomiting. Perineal examination revealed a normal anus. Upon probing the anal canal, the dilator could be negotiated only up to 2.5 cm. Prone cross-table lateral x-ray (PCTLX) revealed gas at the level of the coccyx. The distance between the metal probe and the gas was about 1 cm. The baby underwent a divided sigmoid colostomy. The posterior sagittal approach with the division of thick tissue and end-end recto anal anastomosis was performed. The baby was discharged on the 7th postoperative day. Anal dilatation was started 14 days after surgery. Colostomy was closed at 4 months of life. The baby had associated hydronephrosis for which she was in close observation. The baby had good continence on follow-up at 3 years. (Table 1).

### Table 1: Clinical characteristics of three patients

<table>
<thead>
<tr>
<th>Sr. #</th>
<th>Day of life</th>
<th>Weight (kg)</th>
<th>Presentation</th>
<th>Level of distal gas</th>
<th>Stoma type</th>
<th>Thickness of septum</th>
<th>Associated anomaly</th>
<th>Approach</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>3/M</td>
<td>2.5</td>
<td>Abdominal distention/Non-passage of meconium</td>
<td>Tip of coccyx</td>
<td>Loop</td>
<td>1 cm</td>
<td>PDA, HDN</td>
<td>Posterior sagittal</td>
<td>Good continence</td>
</tr>
<tr>
<td>2</td>
<td>2/M</td>
<td>3.0</td>
<td>Abdominal distention/Non-passage of meconium</td>
<td>Tip of coccyx</td>
<td>Divided</td>
<td>1.2 cm</td>
<td>ASD &amp; HDUN</td>
<td>Posterior sagittal</td>
<td>Good continence</td>
</tr>
<tr>
<td>3</td>
<td>1/F</td>
<td>3</td>
<td>Abdominal distention/vomiting</td>
<td>Tip of coccyx</td>
<td>Loop</td>
<td>2.5 cm</td>
<td>HDN</td>
<td>Posterior sagittal</td>
<td>Good continence with few episodes of encopresis</td>
</tr>
</tbody>
</table>

M=Male, F=Female, PDA=Patent ductus arteriosus, ASD=Atrial septal defect, HDN=Hydronephrosis, HDUN=Hydroureteronephrosis.

![Figure 2: Intraoperative photograph showing thick septum after opening rectum from posterior sagittal approach.](image)

**DISCUSSION**

Rectal Atresia is a rare variant of ARM and there is no standardized management in the literature.[2] They have a normal anus and sphincter mechanisms. The level of stenosis could vary but is usually at the junction of the anal canal and rectum.[3] It usually corresponds to the level just above the dentate line as demonstrated in our three cases. The thickness of the stenotic part may vary from just a septum to a thick tissue about 1 cm in thickness. In two of our cases, it was thick tissue separating the rectum and anal canal. All our cases presented with failure to pass meconium and were diagnosed when a Hegar dilator could not be negotiated beyond 2-3 cms from the anal verge.

Rectal atresia develops due to vascular accidents between 13 to 14 weeks of gestation, with a high incidence among consanguineous marriages, especially in South India. This supports the genetic association of the condition. An infective theory was supported by Magnus who believed that intrauterine infection causing thrombosis of the vessels could lead to acquired atresia of the rectum.[4]

A colostogram is very critical to rule out a fistula which changes the management in such cases. Some authors recommend surgery without a prior colostogram.[5] In our cases, although the diagnosis was made on probing the anal canal and on PCTL x-ray, we still performed colostomy as a safer approach for this variant of ARM, which also helps in ruling out any fistulous communication. Furthermore, placing a metallic probe in anus predicts the thickness of atresia. As demonstrated in our series, the ideal definitive procedure is via the posterior sagittal approach endorsed by many authors like Kisra M et al.[6] Another initial approach described for this anomaly is the division of the septum/atretic tissue with end-to-end anastomosis. This approach may be challenging in some cases because of the discrepancy...
in the size of the upper dilated rectum and lower narrow anal canal.[7] As demonstrated in our cases, if surgery is done within 3-4 months, the size discrepancy is not an issue as far as functional outcome is concerned.

We believe that the anal canal and dentate line, being very important for the overall continence mechanism, should not be dissected or stretched. We did a Heineke-Mikulicz type of anastomosis. Our approach ultimately makes an oblique dentate line without causing any mechanical/surgical trauma to it and is more or less similar to that described by Hamrick M TL et.al.[8] Some authors perform laparoscopic transanal pull-through.[9] Although this approach has the advantages of minimally invasive surgery like less pain and early recovery, we believe that it is unnecessary to do total circumferential rectal dissection and not warranted. Proper follow-up is necessary for the dilatation program and overall assessment of continence.

To conclude, the perineal examination and colostogram help determine the thickness of the septum. The Posterior Sagittal Approach with the division of septum/tissue between the rectum and anal canal and end-end anastomosis gives excellent results.

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**REFERENCES**