

## Rectal Atresia with Congenital Rectovaginal Fistula: A Rare Variant of Anorectal Malformations

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### ABSTRACT

**Background:** rectal atresia is a rare variant of anorectal malformations. The presence of fistulous communication between the rectum and the urogenital system in case of rectal atresia is even rarer to occur. **Aim of the work:** this article aimed to describe a case of rectal atresia in a female infant associated with proximal and distal congenital rectovaginal fistula. **Results:** the clinical, radiological findings and the surgical management were discussed. **Conclusion:** the case demonstrated the possibility to find a rectovaginal fistula in rectal atresia, and the possibility of recurrence after surgical intervention.

**Keywords:** anorectal malformations, rectal atresia, rectovaginal fistula.

### INTRODUCTION

Anorectal malformations are wide spectrum of birth defects mean abnormal development of the anus and rectum with an incidence rate of 2.0-2.5 per 10,000 live births<sup>(1)</sup>. Rectal atresia is one of the rare variant of anorectal malformations according to Wingspread classification; its incidence rate is 1-2% of all anorectal anomalies with male predominance. Blind ended rectum without any fistulous communication and normally developed anal canal and sphincter were usually found in such variety<sup>(2,3)</sup>. So, we studied a case of rectal atresia associated with proximal and distal congenital rectovaginal fistula.

### CASE REPORT

A 10-months-old baby girl of primigravida mother with type I diabetes mellitus born full term with weight of 2.8 kilograms referred to our hospital at age of three weeks with history of passing stool from vaginal opening and double outlet left ventricle. Examination showed soft, lax abdomen and normally placed anal opening. Calibrating the anal canal with a catheter revealed resistance about 2 cm from the anal

opening with no stool coming out. Lower gastrointestinal (GI) study showed short, blind ended anal canal, which was not connected to the rectum, urinary tract or vagina [Figure 1]. Computed tomography (CT) scan with contrast showed dilated, blind-ended rectum and leak of the contrast material to the vagina, which was suggestive of the presence of rectovaginal fistula [Figure 2].

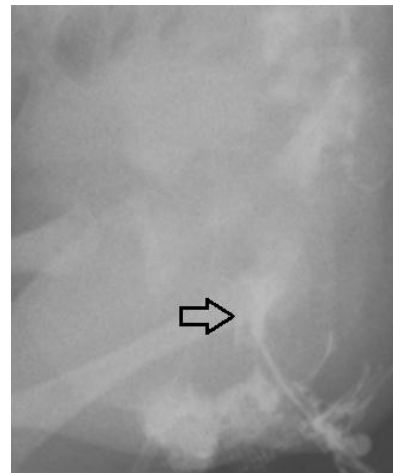
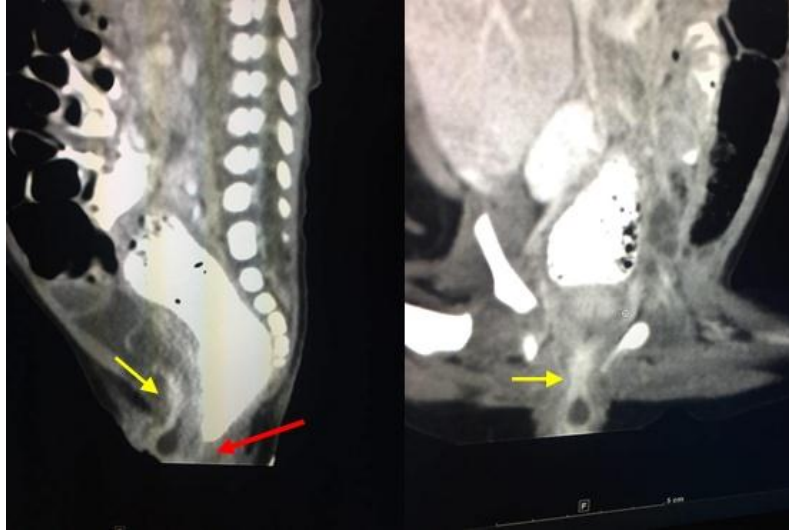


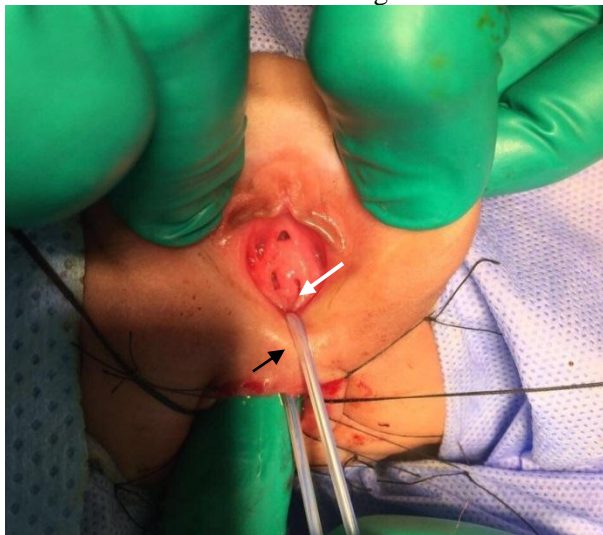
Figure 1: contrast material in the anal canal does not pass to the rectum



**Figure 2: yellow arrow points at the contrast material in the vagina ; red arrow points at the atretic part of the rectum**

Further investigations were done to identify any of vacterl association which revealed congenital heart diseases in form of atrial septal defect, ventral septal defect and double outlet left ventricle. Vertebral anomalies in form of decreased space and angulation of the lumbosacral joint between L5 and S1 vertebra. After proper investigation and consultations, the patient was taken to the operating room, fistula in the posterior vaginal wall was closed and end to end anastomosis was done. Unfortunately, the patient started to pass stool again from the vagina on the 5<sup>th</sup> post-operative day. Lower GI study revealed leak of contrast material to the vagina again. Patient moved to the operating room for examination under

anesthesia and that revealed a fistula to the distal pouch which was closed in layers. Post operation, stool was still coming through the anus as well as the vagina. So, loop colostomy at the distal one third of transverse colon was done. One month later, strictureplasty of rectal anastomotic stricture was done; fistula which was found to be rectovestibular fistula distal to the anastomosis site was ligated, double vaginal septum was divided [Figure 3]. Patient discharged home in good general condition. With follow up, patient showed evidence of stricture at site of anastomosis with recurrence of rectovaginal fistula. Colostomy closure, fistula ligation and transanal pull-through was done.



**Figure 3: white arrow points at the vaginal septum; black arrow shows catheter passing through the rectovaginal fistula**

## DISCUSSION

Wingspread classification of the anorectal malformations classified rectal atresia under the group of rarities<sup>(2)</sup>. Rectal atresia is different from imperforated anus and anal atresia because the anus and anal canal are essentially normal, however, a segment of the rectum is atretic only<sup>(4)</sup>. Studies showed that the incidence rate of this variant of anorectal malformation was 1-2% with male to female ratio of 7:3. Furthermore, rectal atresia in females associated with congenital rectovaginal fistula is even rarer to occur. Very few cases of rectal atresia associated with fistulous communication were reported in the literatures. Although, the exact etiology and pathogenesis of rectal atresia is unknown, but it was assumed that the underlying cause is of a vascular insult during embryogenesis<sup>(5)</sup>. History of failure to pass meconium, refuse feeding, recurrent vomiting and abdominal distention are suggestive for the presence of intestinal obstruction and the need for further evaluation. Fecaluria or passing stool from the vaginal opening are suggestive of fistulous communication between the rectum and the bladder or vagina, respectively, as it is the situation in our case with the later.

Physical examination in case of rectal atresia will show normally placed anus, with resistance upon administration of a rubber catheter or thermometer about 2 cm from the anal verge during rectal examination<sup>(6)</sup>. Lower GI study and CT scan give more details about the anatomy of the present anorectal malformation and they detect the existence of any fistulous communication between the rectum and the urogenital system. Multiple surgical approaches

have been suggested by authors including transanal end-to-end rectorectal anastomosis, transanal approach, and posterior sagittal approach<sup>(5,6)</sup>.

## CONCLUSION

Rectal atresia in females with rectovaginal fistula is an extremely rare variant of anorectal malformation. One should keep in mind the possibility of finding such fistulous communication when a history of passing stool from vaginal opening is given. Moreover, the chance for fistula to recur after repair is always there and this high lights the importance of following up the patient after discharge.

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