

Case Report

Rectal atresia with pouch colon without fistula in a female newborn– a rare association: A case report

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ABSTRACT

Background: Rectal atresia is a rare variant of anorectal malformation (ARM). Pouch colon is an abnormal dilation of the colon distally connected by the fistula. Pouch colon without fistula is extremely rare.

Case Presentation: We report a case of unusual association of rectal atresia with pouch colon without fistula in a female newborn. At surgery, a type IV pouch colon was found which was resected and colostomy was done as the initial procedure.

Conclusion: Rectal atresia with congenital pouch colon without genitourinary fistula in a female child is an extremely rare association.

Keywords: Atresia, Anorectal malformation, Congenital pouch colon, Rectal atresia.

INTRODUCTION

Rectal atresia is an unusual variant of anorectal malformation. These newborns have a normal appearance of the perineum and anal opening up to 1 to 3 cm. It forms 1-2% of cases of ARM. But, the reported incidence of rectal atresia in south India is 14%. [1] Congenital pouch colon (CPC) is the abnormal replacement of the colon, partial or complete, by a dilated segment of the bowel which is distally connected with the urogenital system. [2-6] According to international classification (Krickenbeck 2005) CPC is a rare high type of ARM. [7,8] Congenital pouch colon is generally ended by fistula in the genitourinary system. Congenital pouch colon without fistula is a very uncommon incidence. [9,10].

CASE REPORT

A one-day-old female child weighing 2.4 kg was admitted to our institute with a complaint of severe abdominal distension and no passage of meconium after birth. She was the second live birth of the second gravid mother. The firstborn child was female and apparently healthy without similar complaints. On clinical examination, the baby was crying, the abdomen markedly distended, the anal opening was present. There was no staining of meconium at the perineum or diaper. There was no history of meconuria present. Urethral and vaginal openings were present. An infant feeding tube was introduced in the anal opening but could not negotiate beyond one centimeter. X-ray abdomen and babygram were suggestive of

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congenital pouch colon (Fig. 1). We optimized the baby and bowel decompression was done by nasogastric tube.



Figure 1: X-ray abdomen and babygram were suggestive of congenital pouch colon



Figure 2: The pouch was separated and resected

On day 2 of life, laparotomy was done by hockey stick incision in the left lower abdomen. We found a type 4 pouch colon. The pouch was separated and resected (Fig. 2), sigmoid end colostomy was formed. There was no

fistulous communication of the pouch colon. The colostomy became functioning on the second postoperative day. Oral feed was started on the third postoperative day. The baby was discharged on postoperative day five. There was no complaint in two month follow-up period. The definitive plan in my case of rectal atresia with congenital pouch colon will be colo-rectal anastomosis after gaining weight 6-10 kilogram. Meanwhile, we do lengthening of the rectum by hegar anal dilator.

DISCUSSION

Rectal atresia is not a common variant of ARM. It is classified as type 4 according to the Ladd-Gross classification. It is common in males. Its male-female ratio is 7:3. [1] The embryology of rectal atresia is not clear yet, but a vascular accident during development is the most acceptable theory. [11] Rectal atresia is classified into four types on the basis of the proximal and distal rectal pouch: 1. Rectal atresia with a short gap 2. Long gap 3. Both pouches are separated by thin septa 4. Rectal stenosis. [1] A newly revised classification of rectal atresia is also being used: type I: rectal stenosis (rare); type II: rectal atresia with a septal defect; type III: rectal atresia with a fibrous cord between the two atretic ends (common); type IV: rectal atresia with a gap; type V: multiple rectal atresias with stenosis (A), and multiple atresias (B). [12] In the index case, the proximal end forms a pouch colon. It is a long gap rectal atresia.

Classification for Anorectal malformation (ARM) was given with time with modifications. In 1934 Ladd and Gross proposed the classification for ARM and grouped it into 4 types. Rectal atresia was kept as type 4. [1] Wingspread classification did not include pouch colon and other rare types of ARM. [13] In 1995 Penna proposed the new classification to include a new variant of ARM. Based on gender and need for colostomy for treatment. [14]. In 2005, the Krickenbeck conference proposed a newly modified classification, and congenital pouch colon was also included as ARM. [15]

Very few cases of congenital pouch colon without urogenital fistula were reported in the literature. [10] Our case also had CPC without a urogenital fistula. This suggests the need for modification in the definition of congenital pouch colon. Till now eight cases of rectal atresia with congenital pouch colon were reported in the literature (Table 1). There were six male and two female children in reported cases. The genitourinary fistula was present in six cases and the absent fistula in two cases [16-22].

In conclusion, the concurrence of rectal atresia with congenital pouch colon without a genitourinary fistula is a rare entity that warrants a modification in the definition of congenital pouch colon.

Table 1 – showing all reported cases of rectal atresia with congenital pouch colon in literature

S.no.	Reference year	Age	weight	Gender	Type of CPC	Genitourinary fistula
1	Mathur P et al. 2002 [16]	-	-	M	CPC type 2	Colovesical fistula
2	Kazez A et al. 2009 [17]	-	-	M	CPC type 4	Absent
3	Parelkar S et al. 2010 [18]	-	-	F	CPC type 4	Absent
4	Singh S et al. 2011 [19]	3 day		M	Intermediate CPC type 1 and 2	Proximal colovesical fistula
5	Mirza B et al. 2016 [20]	2 day	2.5 kg	M	CPC type 4	Colovesical fistula
6	Mathur P et al. 2017 [21]	4 day	2 kg	M	CPC type 4	Colovesical fistula
7	Mathur P et al. 2017 [21]	2 day	2.25 kg	M	CPC type 4	Colobadder neck fistula
8	Tiwari C et al. 2021 [22]	5 day	1.9 kg	F	CPC type 1	colovaginal fistula
9	Reporting case	1 day	2.4 kg	F	CPC type 4	Absent

Conflict of Interest: Nil

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Consent to Publication: Author(s) declared taking informed written consent for the publication of clinical photographs /material (if any used), from the legal guardian of the patient with an understanding that every effort will be made to conceal the identity of the patient, however it cannot be guaranteed.

REFERENCES

- Dorairajan T. Anorectal atresia. In: Stephens FD, Smith ED, Paul NW (eds) Anorectal Malformations in Children: Update 1988. Liss, New York, NY, pp 105–10.
- Sharma S, Gupta DK. Management options of congenital pouch colon – A rare variant of anorectal malformation. *Pediatr Surg Int.* 2015; 31:753–8.
- Gupta DK, Sharma S. Congenital pouch colon – Then and now. *J Indian Assoc Pediatr Surg.* 2007; 12:5–12.
- Singh A, Singh R, Singh A. Short colon malformation with imperforate anus. *Acta Paediatr Scand.* 1977; 66:589–94.
- Narsimha Rao KL, Yadav K, Mitra SK, Pathak IG. Congenital short colon with imperforate anus (CPC syndrome) *Ann Pediatr Surg.* 1984; 1:159.
- Wardhan H, Gangopadhyay AN, Singhal GD, Gopal SC. Imperforate anus with congenital short colon (pouch colon syndrome): Review of 18 cases. *Pediatr Surg Int.* 1990; 5:124–6.
- Maudar KK, Gandhi P, Varshney S, Budhwani KS, Ghritalaharey RK. Congenital pouch colon: Review of current Clinical And Molecular Studies; *Journal Of Paediatric And Neonatal Care* 2016; 5(9).
- Mathur P, Saxena AK, Bajaj M, Chandra T, Sharma NC, Simlot A, et al. Role of plain abdominal radiographs in predicting type of congenital pouch colon. *Pediatr Radiol* 2010; 40:1603–8.
- Pandey V, Panigrahi P, Kumar R. Congenital pouch colon without fistula:our experiences and lessons learned. *Asian Journal of Health and Medical Research* 2017; 3:1–2.
- Chaturvedi V, Gupta R, Tuteja N, Barolia DK, Bal R. Congenital Pouch Colon without Fistula: A Rare Variant. *Journal of Neonatal Surgery.* 2019; 8:4: 38.
- Hamrick M, Eradi B, Bischoff A, Louden E, Peña A, Levitt M. Rectal atresia and stenosis: unique anorectal malformations. *J Pediatr Surg.* 2012 Jun; 47(6):1280–4.
- Gupta DK, Sharma S (2006) Rectal atresia and rectal ectasia. In: Hutson J, Holschneider A (eds) Anorectal malformations in children, chap 12, 1st edn. Springer, Heidelberg, pp 223–30.
- Stephens FD, Durham-Smith E. Classification, identification, and assessment of surgical treatment of anorectal anomalies. *Pediatr Surg Int.* 1986; 1:200–5.
- Levitt MA, Peña A. Anorectal malformations. *Orphanet J Rare Dis.* 2007; 2:33.
- Gupta DK. Anorectal malformations – Wingspread To Krickenbeck. *J Indian Assoc of Pediatr Surg.* 2005; 10:75–7.
- Mathur P, Prabhu K, Jindal D. Unusual presentations of pouch colon. *J Pediatr Surg* 2002; 37(9):1351–3.
- Kazez A, Ozel SK, Bakal U, Sarac M. Abdominotransanal approach to pouch colon associated with rectal atresia. *J Pediatr Surg* 2009 Jun; 44(6):E19–21.
- Parelkar S, Oak S, Mishra PK, Agrawal A, Joshi M, Beejal S, et al. Congenital pouch colon with rectal atresia: A case report. *J Pediatr Surg* 2010 Mar; 45(3):639–41.
- Singh S, Ahmed I, Wakhlu A. A newer variant of congenital pouch colon with rectal agenesis: Management strategy and review of the literature. *BMJ Case Rep* 2011 Aug 31; 2011:bcr1220103635.
- Mirza B, Saleem M. Congenital pouch colon with rectal atresia: A rare association. *J Neonatal Surg* 2016; 5:66.
- Mathur P, Gupta R, Gupta PK. Congenital pouch colon with rectal atresia revisited. *J Indian Assoc Pediatr Surg* 2017; 22(2):119–121.
- Tiwari C, Shah H, Desale J, Waghmare M. Congenital pouch colon with colovaginal fistula and rectal atresia in a female: An unusual presentation and literature review. *Saudi Surg J* 2016; 4:125–8.