

## Case Report

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

Aditya Pratap Singh,<sup>1</sup> Dinesh Kumar Barolia,<sup>2\*</sup> Harsha Vinod Bathia,<sup>3</sup> Vipal H Parmar,<sup>4</sup> Bhavana Asit Mehta,<sup>5</sup> Shraddha Mehta,<sup>6</sup>

1. Department of Paediatric Surgery, Bhagwan Mahavir Hospital, Mahavir Nagar, Rajasthan, India,

2. Department of Paediatric Surgery, J. L. N. Medical College, Ajmer, Rajasthan, India.

3. Department of Anaesthesia, Bhagwan Mahavir Hospital, Mahavir Nagar, Rajasthan, India.

4. Department of histopathology at Neuberg Supratech References Laboratories, Ahmedabad (NSRL), India.

5. Department of Histoanatomic Pathology, Neuberg Supratech Reference Laboratory, Ahmedabad, Gujarat, India,

6. Department of Pathology, Bhagwan Mahavir Hospital, Mahavir Nagar, Rajasthan, India.

**Cite as:** Singh AP, Barolia DK, Bathia HV, Parmar VH, Mehta BA, Mehta S. Rectal atresia with pouch colon without fistula in a female newborn– a rare association: A case report. J Pediatr Adolesc Surg. 2024; 2: 106-8.

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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 (Krackenbeck 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

Correspondence\*: Dr. Dinesh Kumar Barolia, Barolia Bhawan, Near Panchali Phatak, Srimadhapur, District – Sikar Rajasthan, India, Pin code – 332715.

Email: [dbaroliamt@gmail.com](mailto:dbaroliamt@gmail.com)  
Submitted on: 25-07-2021

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Accepted on: 01-03-2022

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 Krickbeck 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	Colobaladder 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

**Source of Support:** Nil

**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.

**Authors Contribution:** Author(s) declared to fulfill authorship criteria as devised by ICMJE and approved the final version. Authorship declaration form, submitted by the author(s), is available with the editorial office.

**Acknowledgements:** None

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