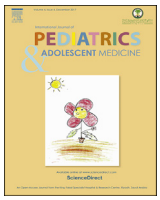
International Journal of Pediatrics and Adolescent Medicine 4 (2017) 138–140

HOSTED BY

Contents lists available at ScienceDirect

International Journal of Pediatrics and Adolescent Medicine

journal homepage: http://www.elsevier.com/locate/ijpam



Full length article

# Congenital rectovaginal fistula with anorectal agenesis: A rare anorectal malformation

Charu Tiwari, Hemanshi Shah*, Jyoti Bothra, Gursev Sandlas

*Department of Paediatric Surgery, TNMC & BYL Nair Hospital, Mumbai, Maharashtra, 400008, India*

ARTICLE INFO

*Article history:*
Received 23 June 2017
Received in revised form
13 August 2017
Accepted 22 August 2017
Available online 16 December 2017

*Keywords:*
Rectovaginal fistula
Congenital
Anorectal malformation

ABSTRACT

*Background:* Rectovaginal fistula is a rare type of anorectal malformation; the incidence being less than 1%. We describe five cases of rectovaginal fistula managed at our institution.
*Materials and methods:* Case records of five female neonates with rectovaginal fistula managed at our institute between 2010 and 2016 were reviewed and analysed with respect to age at presentation, clinical presentations, physical findings, investigations, management and outcome.
*Results:* The age at presentation varied from 1 day to 2 years of age. Three of them presented in the neonatal period, one presented at 1 month of age and one at two years of age with sigmoid loop colostomy done elsewhere. All had absent anal opening; two neonates passed small amounts of stools through vagina, but little in amounts. The one-month old patient had history of passing stools through vaginal orifice, but had presented to us with obstruction. All patients underwent high sigmoid loop colostomy followed by definitive procedure at a later date – Posterior Sagittal Anorectoplasty. One patient is awaiting definitive repair.
*Conclusion:* Rectovaginal fistula is a rare anorectal malformation and needs thorough investigation and appropriate management for good outcome.



## 1. Introduction

The incidence of anorectal malformations (ARMs) is 1 in 4000–5000 live births [1,2]. ARMs are commonly associated with other congenital anomalies [1]. The classification of female ARM is very complex and still evolving because of the difficult anatomy of female ARM and presence of various rare and regional variants [3]. Moreover, there is no clear demarcation between high and low ARM in females [3]. Rectovaginal fistula is included as one of the rare/regional variants in the Krickenbeck classification (2005) [1,3,4]. Rectovaginal fistula, along with congenital pouch colon (CPC) and H-type fistula are more common in India and some Asian countries [5–8].

The incidence of anorectal agenesis with rectovaginal fistula is less than 1% [3]. However, the reported incidence varies widely from 0% to 84% literature [9]. These false high reported incidences have been attributed to the indiscriminate labelling of rectovestibular fistula and cloaca as rectovaginal fistula [10]. In fact, Pena states that an isolated rectovaginal fistula is almost non-existent and higher reported incidence is due to the misdiagnosed cases of rectovestibular fistula and persistent cloaca [10,11].

We present the analysis of rectovaginal fistula managed at our institution.

## 2. Materials and methods

Case records of female patients with rectovaginal fistula managed at our institute between 2010 and 2016 were reviewed and analysed with respect to age at presentation, clinical presentations, physical findings, investigations, management and outcome.

All patients with absent anal opening were admitted. A careful examination was done for the number of openings in the introitus. The perineum was examined to look for any fistulous opening. A general and systemic examination was done to rule out other anomalies.

* Corresponding author.
*E-mail address:* hemanshisshah@gmail.com (H. Shah).
Peer review under responsibility of King Faisal Specialist Hospital & Research Centre (General Organization), Saudi Arabia.

https://doi.org/10.1016/j.ijpam.2017.08.003

Patients with single opening in the introitus suggesting cloaca underwent colostomy in the neonatal period followed by definitive procedure. Patients with three openings in the introitus suggestive of rectovestibular fistula were started on rectal washouts through the fistulous opening before definitive repair.

Neonates with two openings in the introitus underwent colostomy in the neonatal period.

## 3. Results

A total of 5 patients with rectovaginal fistula were managed during 2010 and 2016. The age at presentation varied from 1 day to 2 years of age. Three of them presented in the neonatal period, one presented at one month of age and one at 2 years of age.

All had an absent anal opening at birth (Fig. 1). One neonate had abdominal distention and had not passed any meconium. Two neonates were passing meconium through the vaginal orifice, but little in amounts and had abdominal distention. The one-month-old girl had history of passing stools per vaginally but presented with abdominal distention and decreased amounts of stools vagina. All patients underwent a high sigmoid colostomy. The two-year old girl had sigmoid loop colostomy done elsewhere for absent anal opening and had presented to us for further management.

All patients underwent abdominal and spinal ultrasound (USG), distal colostogram and 2-D Echo. One patient had associated large lumbosacral meningomyelocoele (MMC) with bilateral paraplegia along with hydrocephalus and patent foramen ovale. She underwent ventriculo-peritoneal shunt and MMC repair before definitive surgery. At examination under general anaesthesia (EUGA) and cystogenitoscopy, a rectovaginal fistula was confirmed.

Four patients underwent definitive repair by posterior sagittal anorectoplasty (PSARP) approach with division of fistula and repair of vagina. They were kept on postoperative anal dilatation. Colostomy closure was done after 6 weeks of definitive surgery. One patient is awaiting definitive repair.

All four patients are continent and have no complaints of constipation at follow up.

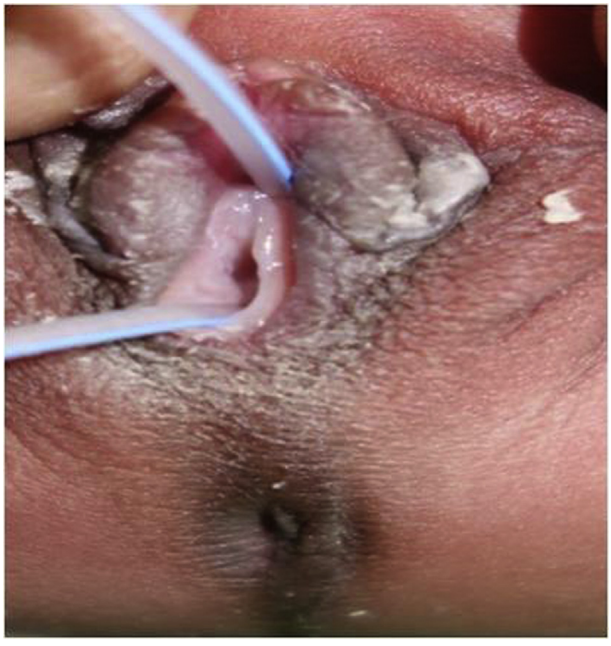


**Fig. 1.** Perineal examination of a neonate showing absent anal opening and two openings in the introitus suggesting a congenital rectovaginal fistula.

## 4. Discussion

Rectovaginal fistula is a rare type of female ARM. Recently, the better understanding of true anatomy of the different types of ARMs has led to a decrease in the incidence of rectovaginal fistula [12]. Also, cloacas are now being less commonly misdiagnosed as rectovaginal fistulas [12]. The close proximity of genital organs makes the anatomy of female ARM very complex and difficult to understand [3]. In a female with absent anal opening at the normal anal site, a careful local physical examination with respect to the number of openings present in the introitus will help in determining the type of ARM. The management will then vary accordingly. However, such examination may at times be difficult in a neonate [3].

A single perineal opening with shorter appearing introitus suggests cloaca. Three openings in the introitus with rectal opening appears as a fistula in the posterior vestibule, outside of the hymen suggests the diagnosis of a rectovestibular fistula. If two openings are seen in the introitus with an absent anus, then the differential diagnosis becomes imperforate anus with no fistula (commonly seen in patients with trisomy 21), anorectal agenesis with rectovaginal fistula and rectovestibular fistula with vaginal agenesis (Mayer Rokitansky Kauser Hauser Syndrome). A majority of patients with malformations present at neonatal period [13]. But, some cases with wide fistula may present late as they decompress well [3]. This is especially true in a developing country like India where there are still home deliveries in remote villages [13].

Thorough radiological investigations are required to rule out associated anomalies. An examination under general anaesthesia and cystogenitoscopy will help to delineate the anatomy. A Magnetic Resonance Imaging will also assist in delineating the anatomy. There is no universal protocol for management because of the varied rare and regional variants [3]. The surgical management of these anomalies is confusing and not standardized [3]. Definitive interval surgical reconstruction should be carefully planned and done in a single stage whenever possible [10]. A well-planned and coordinated approach usually results in a satisfactory outcome [10].

The neonates mostly require a preliminary colostomy for decompression. However, patients with wide fistula who are deflating well can be kept on washouts and managed by a single-stage approach at a later date. For the definitive surgery, the posterior sagittal approach provides good anatomical exposure and accurate and precise placement of distal rectum within the sphincter muscle complex [1,3]. However, some patients with high ARMs may eventually need bowel management after PSARP [1].

Recently, there has been more emphasis on single-stage correction of ARM [3,14,15]. The laparoscopically assisted technique was first introduced in 2000 by Georgeson et al. [16]. This approach has gained interest because of minimal abdominal and perineal wounds [3].

Many reports have been published in literature describing rectovaginal fistula. Chatterjee et al. [17] have reported 22 cases of rectovaginal fistula - three were high anomalies opening in the posterior fornix of vagina, and rest were intermediate anomalies opening in the lower vagina. Upadhyaya et al. [14] have reported the incidence of rectovaginal fistula to be 6% (15 out of 244 female ARM) in their series. Hashmi and Hashmi [8] have reported twenty cases out of 130 female ARM patients. Choudhary et al. [3] have recently reported 6% incidence of this malformation at a tertiary centre.

To summarize, rectovaginal fistula with anorectal agenesis is rare, needs thorough investigations to rule out associated anomalies and planned definitive procedure, possibly in a single stage to achieve a good outcome.

## Ethical clearance

Not applicable in this because this is a retrospective observational study. All patients were managed according to the existing protocols. No new investigations and intervention were done on these patients.

## Financial disclosure

None.

## Conflicts of interest

The authors declare that they have no conflict of interest.

## Acknowledgement

Nil.

## References

[1] Bailez MM, Cuenca ES, Benedetto VD, Solana J. Laparoscopic treatment of rectovaginal fistulas. feasibility, technical details and functional results of a rare anorectal malformation. J Pediatr Surg 2010;45:1837–42.
[2] Levitt M, Peña A. Outcomes from the correction of anorectal malformations. Curr Opin Pediatr 2005;17:394–401.
[3] Choudhury SR, Khan NA, Debnath PR, Yadav PS, Shah S, Chadha R. Anorectal agenesis with rectovaginal fistula: a rare/regional variant. J Indian Assoc Pediatr Surg 2017;22:79–82.
[4] Holschneider A, Hutson J, Peña A, Beket E, Chatterjee S, Coran A, et al. Preliminary report on the international conference for the development of standards for the treatment of anorectal malformations. J Pediatr Surg 2005;40:1521–6.
[5] Sarin YK, Sinha A. Two orifices in the perineum of a girl with imperforate anus: possibility of uterovaginal agenesis associated with rectovestibular stula. J Pediatr Surg 2002;37:1217–9.
[6] Chadha R, Gupta S, Mahajan JK, Bagga D, Kumar A. Congenital pouch colon in females. Pediatr Surg Int 1999;15:336–42.
[7] Abnormal anus and anorectum with a stula. In: Chatterjee SK, editor. Surgery of pediatric anorectal malformations. New Delhi: Viva Books Private Limited; 2005. p. 164–74.
[8] Hashmi MA, Hashmi S. Anorectal malformations in female children–10 years' experience. J R Coll Surg Edinb 2000;45:153–8.
[9] Galifer RB, Chavrier Y, Viala JS. Principles of the surgical treatment of cloacal malformation. Apropos of 3 cases. Chir Pediatr 1985;26:95–103.
[10] Patankar JZ, Mali V, Prabhakaran K, Bo L, Loh DL. Urogenital sinus, rectovaginal fistula and an anterior stenosed anus-another cloacal variant. Pediatr Surg Int 2004;20:556–8.
[11] Hendren WH. Repair of cloacal anomalies: current techniques. J Pediatr Surg 1986;21:1159.
[12] Rosen NG, Hong AR, Soffer SZ, Rodriguez G, Pena A. Rectoaginal fistula: a common diagnostic error with significant consequences in girls with anorectal malformations. J Pediatr Surg 2002;37(7):961–5.
[13] Kharnaior A, Lokendra SK, Devi AR, Moirangthem GS. Congenital high type anorectal malformation (Recto-Vaginal Fistula) in a 7 months old girl: a case report. J Evol Med Dent Sci 2015;4(34):5937–41. https://doi.org/10.14260/jemds/2015/868. April 27.
[14] Upadhyaya VD, Gangopadhyay AN, Pandey A, Kumar V, Sharma SP, Gopal SC, et al. Single-stage repair for rectovestibular stula without opening the fourchette. J Pediatr Surg 2008;43:775–9.
[15] Menon P, Rao KL. Primary anorectoplasty in females with common anorectal malformations without colostomy. J Pediatr Surg 2007;42:1103–6.
[16] Georgeson KE, Inge TH, Albanese CT. Laparoscopically assisted anorectal pull-through for high imperforate anus—a new technique. J Pediatr Surg 2000;35:927–31.
[17] Chatterjee SK. Rare/regional variants. In: Holschneider AM, Hutson JM, editors. Anorectal malformations in children embryology, diagnosis, surgical treatment, follow-up. Berlin, Heidelberg: Springer; 2006. p. 251–62.