1. Introduction

The incidence of anorectal malformations (ARMs) is 1 in 4000–5000 live births [1,2]. ARM is commonly associated with other congenital anomalies [3]. The classification of female ARM is complex and still evolving because of the difficult anatomy of female ARM and presence of various rare and regional variants [4]. 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 institute.

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.
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 meningomyelocele (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.

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 [13]. 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. [2] 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

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Conflicts of interest

The authors declare that they have no conflict of interest.

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