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Management of H-type rectovestibular and rectovaginal fistulas

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Abstract

Introduction: H-type rectovestibular or rectovaginal fistulas are rare entities in the spectrum of anorectal malformations seen in North America. Management options described in the literature have included perineal repair, anterior perineal anorectoplasty, vestibuloanal pull-through, and limited or formal posterior sagittal anorectoplasty, with a reported recurrence rate of 5% to 30%. We describe our approach and outcome in the management of these patients.

Methods: In a series of 1170 females with anorectal malformation, we cared for 8 patients who had an H-type rectovestibular or rectovaginal fistula and reviewed their clinical presentation, diagnosis, operative technique, and postoperative course.

Results: The patients' presenting symptoms included passage of stool per vagina (6), constipation (3), labial abscess (1), and recurrent urinary tract infection (1). There was associated anorectal stenosis in 3 patients. The remaining 5 patients had normal anal openings. Endoscopy was not helpful in locating the fistulas, but the fistulas were all demonstrated on direct inspection under anesthesia. The fistula was located in the vestibule (4), vagina (3), or labia (1). One patient had an associated presacral mass. Two patients had been operated on twice previously using a perineal repair and a protective colostomy and presented with third recurrences. In 5 cases, a posterior sagittal approach was used, placing sutures circumferentially around the fistulous opening on the rectal side, ligating the fistula, and pulling down a normal segment of rectum to be placed in front of the repaired vaginal wall. In our last 3 cases, we performed a transanal mobilization of the anterior rectal wall, leaving the perineal body intact. After our repairs, the patients have been followed up for 3 months to 15 years with a median of 15 months, and we have seen no recurrences.

Conclusions: In addition to vaginal passage of stool, an H-type fistula should be suspected when there is a labial abscess in an infant, and an associated anal stenosis or presacral mass must be checked for. Direct inspection is the key, with a careful look in the vestibule, because endoscopy may miss the fistula. The essential technical point for repair is to get healthy anterior rectal wall to cover the area of fistula on the posterior vagina. A transanal approach, leaving the perineal body intact, is an excellent option for this repair.

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H-type rectovestibular or rectovaginal fistulas are rare entities in the spectrum of anorectal malformations (ARMs) seen in North America, with few cases reported in the

literature [1-5]. The incidence in studies from Asia, for unknown reasons, is greater and has been reported to be as high as 7% to 14% of all ARMs in females [6,7]. Management options described in the literature have included perineal repair, vestibuloanal pull-through, anterior perineal anorectoplasty, and a limited or formal posterior sagittal anorectoplasty (PSARP), with a reported recurrence rate of 5% to 30% [1,5,6,8,9]. We describe our technique of managing this rarely encountered problem, reviewed the patients we treated, and analyzed their outcomes.

1. Methods

With institutional review board approval (no. 2008-1317), a retrospective review was conducted of all female patients managed in our center between 1990 and 2010 for ARMs. Of 1170 females with ARM, 8 had an H-type rectovestibular or rectovaginal fistula. We reviewed the clinical presentation, diagnosis, operative technique, and postoperative course for each patient.

2. Results

The age at presentation ranged from 1 day to 10 years with a median of 10 months. Six of the patients were 1 year or younger, 1 was 3 years old, and 1 was 10 years old. Passage of stool per vagina was the most common presenting symptom seen in 7 patients (88%). One patient presented with a labial abscess, whereas the others had no history suggestive of a preceding perineal inflammation. Recurrent constipation (3 patients) and urinary tract infections (1 patient) were the other symptoms noted. Two patients had been operated on twice previously using a perineal repair and a protective colostomy and presented with a third recurrence. There was associated anorectal stenosis in 3 patients; the remaining 5 had normal anal openings. Contrast enema, endoscopy, and examination under anesthesia were performed to evaluate the fistulas. Endoscopy was not helpful in locating the fistulas, but the fistulas were all demonstrated on direct inspection under anesthesia.

The fistula was located in the vestibule (4) (Fig. 1), vagina (3), and labia (1). Among the 6 patients who presented primarily, 5 fistulas were located above the dentate line, and 1 opened into the anal crypts at the dentate line. Anorectal stenosis (caliber smaller than a size 9 Hegar dilator) was confirmed in 3 patients at examination under anesthesia.

In 5 cases, a posterior approach in prone position was used to repair the malformation. This involved incising the perineal body between the fistula and anus, placing sutures circumferentially around the fistula opening on the rectal side mobilizing the anterior rectal wall, ligating the fistula, and then pulling down a normal segment of rectum to be placed in front of the repaired vaginal wall (Fig. 2). In our last 3 cases, we repaired the defect transanally, leaving the perineal body untouched. In a prone position, and with a Lone star retractor (CooperSurgical, Trumbull, CT) to improve visualization, we performed a transanal mobilization of the anterior rectal wall (Fig. 3). The fistula was ligated, and the anterior rectal wall was pulled down and sutured to the anal verge. If there is no colostomy, we have fasted the patient for 7 to 10 days and on intravenous nutrition.

Three patients had a protective colostomy for the repair: 2 patients presented with colostomies done after previous failed repairs elsewhere, and 1 patient with anal stenosis and a presacral mass presented with severe constipation and recurrent episodes of fecal impaction that was unresponsive to laxatives. Five patients had the repair done without a protective colostomy.

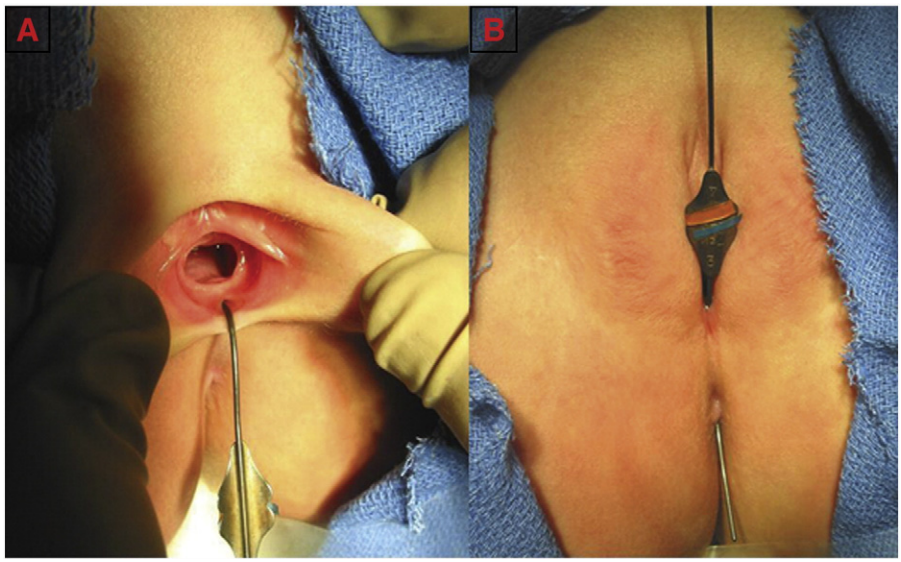

Fig. 1 A probe is inserted into the fistula, located at the vestibule (A) and seen to be exiting the anal opening (B).

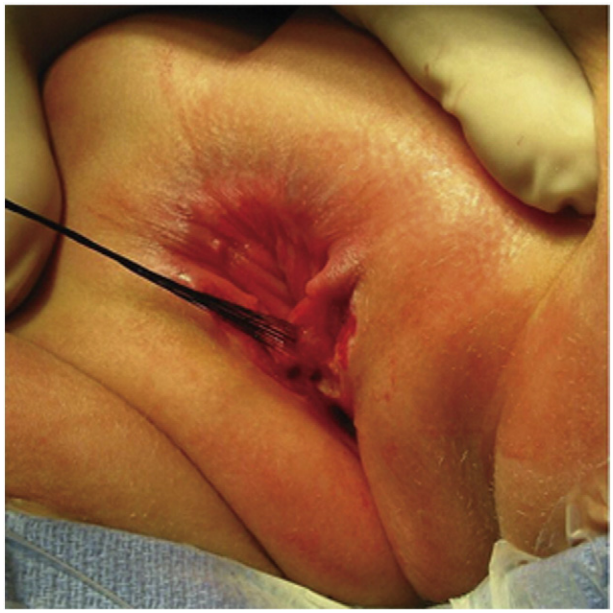

Fig. 2 The posterior sagittal approach was used in the first 5 cases. Silk sutures were placed at the rectal end of the fistula to provide traction.

There was a postoperative perineal body dehiscence in 1 patient, using our earlier approach, which necessitated resuturing and diversion. The patient subsequently did well. The patients have been followed up for 3 months to 15 years with a median of 15 months. We have seen no recurrences (Fig. 4). All the patients older than 3 years (6 patients) have normal bowel control, and the remaining 2 are yet to reach the age of toilet training.

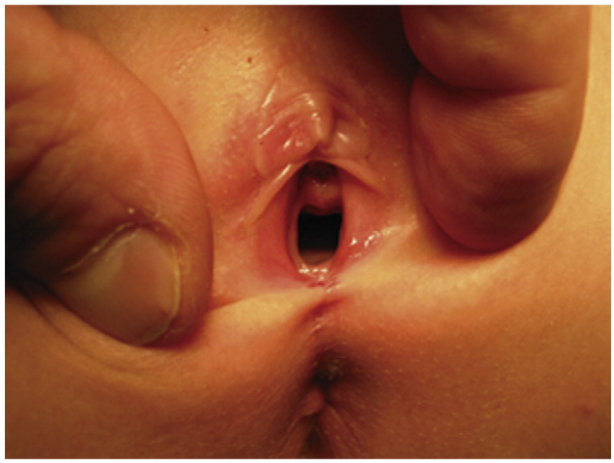

Fig. 4 Picture taken at 3 weeks of follow-up showing a satisfactory healing with no recurrence.

3. Discussion

The first report in the literature that we identified on H-type rectovestibular or rectovaginal fistulas was from 1960 [10]. These malformations are consistently noted to be rare in Western countries [1-5]. Much larger series are reported from Asia [6,7,9]. The reason for the geographical variation remains unexplained but has been suspected to be related to acquired fistulas related to an inflammatory process such as a perianal abscess [1,2,6,9]. H-type fistulas represent 14% of the ARMs seen in females in India and Japan [6,7] compared with 3.2% of 629 patients (males and females) with ARMs in a report from Finland [1]. Eight patients out of a total of 1170 females with ARM in our series have an H-type

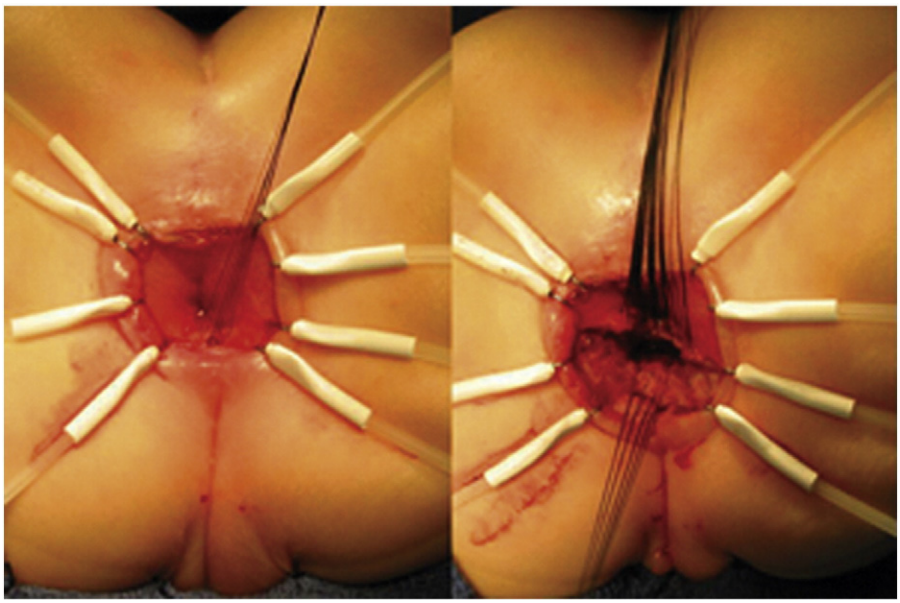

Fig. 3 The transanal approach in prone position for the mobilization of the anterior rectal wall (180°), ligation of the fistula, and repair. The anterior wall of the rectum is mobilized and pulled distally to cover the area of the fistula with healthy rectal wall.

rectovestibular or rectovaginal fistula. This is an incidence of 0.7%, which further confirms the rarity in the United States.

The etiology of H-type fistulas is unclear, and different theories have been proposed [11,12]. It is generally thought to be congenital in cases associated with ARMs and where a preceding perineal inflammation is absent [1]. The fistula is possibly acquired in nature if there is a preceding perineal inflammation or the opening is into one of the anal crypts at the dentate line. A history of prior perineal sepsis was absent in our patients, and in 5 of the 6 patients who presented without a previous operation, the fistula opened above the dentate line suggesting a congenital etiology. Its association with anal stenosis also connotes a congenital origin.

The presentation in most patients is incidentally noted as a small quantity of stool in the vestibule. In older children, the passage of feces through the vagina is worsened by episodes of diarrhea. The presence of a recurring labial abscess in infancy was the "red flag" in one of our patients, which has been reported by others [1,4]. In a review of 182 patients with H-type fistulas from China, the incidence of a prior perineal inflammation or vulval abscess was quite high with 86% having a history of vulval inflammation [9]. The fistulas in that series were probably all acquired in contrast to what we and others have noted [1,2,6,8]. For such patients, a high index of suspicion for a fistula is required, and the anus should be checked with Hegar dilators to evaluate for an anal stenosis.

The most common type of ARM associated with H-type rectovestibular or rectovaginal fistulas is anorectal stenosis, which was present in 3 of our patients (37.5%). In general, this is a relatively rare type of ARM, most notably associated with a presacral mass [13]. This was the cause for recurrent constipation in these patients, a finding also noted by others [6,7]. In addition, H-type fistulas may be associated with other anomalies such as a presacral mass, which was noted in one of our patients. A thorough evaluation for these associations should be made in any female with an H-type fistula. Other congenital anomalies reported by others in association with H-type fistulas include vertebral, urologic, cardiac, and genital anomalies [1,5,14].

In the evaluation of patients with H-type fistulas, contrast enema examination, proctosigmoidoscopy, and examination under anesthesia are the most commonly used tools for diagnosis [2,6-8]. Although some authors have found contrast enemas to be diagnostic in most of their patients [7], we and others [6] have found contrast enemas and endoscopy unhelpful. The sensitivity of contrast enema in the identification of the fistula track is related to the size of the fistula because it is more likely to identify larger ones with diameters greater than 5 mm [9]. In our review, examination under anesthesia was the most helpful way of confirming the diagnosis and was also useful to check for anorectal stenosis using Hegar dilators.

Different surgical techniques have been described to approach H-type vaginal or vestibular fistulas. These include perineal repair, fistulectomy, abdominoperineal repair, limited or formal PSARP, vestibuloanal pull-through, anterior perineal anorectoplasty, and a transanal approach [1,2,4-9,14]. The major complications are fistula recurrence (5%-30%) and wound dehiscence (0%-25%). The rate of recurrence after perineal repair is difficult to compare because of the different techniques used by various authors, but the rate tends to be lower when there is some mobilization of the normal proximal anterior rectum, which is then sutured to the anal canal or anocutaneous junction

Table 1 The different surgical techniques used to repair H-type fistulas in the literature

Author, year of publication	No. of patients	Surgical technique used	Fistula recurrence
Chatterjee, 1969 [6]	7	Fistula laid open (1)	1/1
		Perineal (3)	1/3
		Vestibuloanal (3)	1/3
Tsuchida, 1984 [7]	12 (11 had surgery)	Abdominoperineal (1)	0/1
		Perineal (3)	2/3
		Perineal (no opposing suture line) (7)	0/7
Rintala, 1996 [1]	14 (1 died before surgery)	Perineal (8)	3/8
		Limited PSARP (4)	0/4
		PSARP (1)	0/1
Willems, 1996 [2]	1	Mini PSARP (1)	0/1
Tsugawa, 1999 [8]	19	Perineal (no opposing suture line) (19)	1/19
Meyer, 2009 [4]	2	Perineal (no opposing suture line)	0/2
Yu, 2009 [5]	1	Fistulectomy (perineal)	1/1
Mahajan, 2009 [14]	1	Fistulectomy (perineal)	0/1
Li, 2010 [9]	182	Vestibulorectal (98)	8/98
		Transanal (69)	11/69
		APARP (15)	2/15
Our series	8	PSARP (5)	0/8
		Transanal (3)	

APARP indicates anterior perineal anorectoplasty.

(Table 1). An added advantage when the normal rectum is pulled downward is that the suture line on the repaired posterior vaginal wall will not be adjacent to a suture line on the anterior rectal wall. Incorporating this principle, a posterior sagittal approach has been successfully used by us and others [1] without fistula recurrence. When using the posterior sagittal approach, it is essential to keep the lateral and, if possible, the posterior walls of the anal canal intact, to preserve the continence mechanism.

More recently, we have been approaching the fistula transanally. In addition to the advantages of a transanal approach, the perineal body is left intact, and a potential dehiscence is avoided. This is a reproducible technique that is familiar to pediatric surgeons who use this approach to Hirschsprung pull-throughs. When combined with the principle of not leaving suture lines behind the repaired posterior vagina (which helps avoid fistula recurrence) as well as avoiding an incision in the perineal body, we believe it is the preferred approach (Fig. 4).

4. Conclusions

In addition to vaginal passage of stool, an H-type fistula should be suspected when there is a labial abscess in an infant, and an associated anal stenosis or presacral mass must be checked for. Direct inspection is the key, with a careful look in the vestibule, because endoscopy and contrast studies may miss the fistula. The essential technical point for repair is to get the healthy anterior rectal wall to cover the area of fistula on the posterior vagina. A transanal approach, leaving the perineal body intact, is an excellent option for this repair.

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