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CASE REPORT

H-type Rectourethral Fistula with Anal Stenosis in a Male Patient: A Case Report with Literature Review

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ABSTRACT
Background: H-type rectourethral fistula is a rare variant of anorectal malformation that affects male neonates. It is usually associated with anal stenosis but with a normal anal position and shape, so it is difficult to be diagnosed early after birth. We present this case to evaluate the functional outcome of this rare anomaly in a male patient and compare it with other reported cases with different ages at diagnosis.
Case Presentation: A nine-month-old infant presented with recurrent attacks of urinary tract infection and failure to thrive. After proper evaluation, the diagnosis of an H-type rectourethral fistula with anal stenosis was confirmed, and a diverting double-barreled colostomy was performed as an initial procedure. Posterior sagittal anorectoplasty with ligation and division of fistula was successfully done, followed by colostomy closure. Now the patient is five years old and is continent for urine and stool.
Conclusions: The diagnosis of H-type rectourethral fistula with anal stenosis is challenging to physicians, and a high index of suspicion is required to reach the diagnosis as the anal opening had a normal shape and location. If this condition is diagnosed and treated early, these patients will have an excellent functional prognosis, as the muscle complex and anal sphincter are well developed.

KEYWORDS: Congenital defect, Rectourethral fistula, Anorectal malformation.

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INTRODUCTION

Anorectal malformations (ARMs) are congenital defects involving the anorectum and urogenital system in boys and girls with variable severity. The malformations range from simple defects at the skin level, such as rectoperineal fistulas, to complex ones, such as persistent cloaca. According to recent studies, the overall incidence of ARMs is 3.09 per 10,000 births [1]. The exact etiology has not been fully determined, but it is likely multifactorial, including genetic and environmental factors [1,2]. The prognosis of ARM depends on the complexity of this malformation [3]. The H-type rectourethral fistula is a rare variant of anorectal malformation that occurs in male patients, and it is usually associated with delayed diagnosis due to the presence of a normal anus [4]. This deformity is more commonly reported in female patients and is usually an acquired problem secondary to inflammatory events affecting the genitourinary system [4-6]. The diagnosis of this anomaly is confirmed by contrast study (barium enema and voiding cystourethrography), MRI, and endoscopy, revealing the fistulous connection between the rectum and urethra [5,7]. Many surgical procedures can be used to correct this anomaly, including limited posterior sagittal anorectoplasty (PSARP), anterior rectal wall pull-through, and endorectal pull-through operations [4,8]. The functional prognosis is good because the gluteal region and muscle complex are usually well developed [9,10]. We present a case of H-type rectourethral fistula in a male infant to assess the functional outcome of this rare anomaly and compare it with other reported cases with different ages at diagnosis. A brief literature review was conducted on male patients with similar pathology to evaluate the clinical aspects and functional outcomes and compare them with those in our case (Table 1).

CASE PRESENTATION

A nine-month-old male infant presented with failure to thrive, recurrent febrile attacks, and constipation starting at the fourth month of life. The family noticed a decrease in bowel motion frequency with the introduction of weaning foods (at the fourth month of life), and the baby started to pass a ribbon-like stool. The mother noticed a greenish stool material mixed with urine in the diaper, which was confirmed by a urine test (Figure 1).

Study (year)	Patient age	Clinical presentation	Perineal finding	Associated anomalies	Surgical technique	Functional outcome
Slater BJ (2014) [4]	2 months	Respiratory distress particulate matter in urine slit-like anus	Normal anus	Vertebral anomalies, grade II right VUR, VSD	PSARP	Could not be assessed (infant age)
Banu (2009) [8]	2 days	Passing meconium with urine	Ectopic anus	Cardiac anomaly, congenital cataract, sacral hypoplasia	Anterior sagittal anorectoplasty	Died on day 4 postoperatively
Banu (2009) [8]	2 days	Meconium with urine and through the anal pit	Ectopic anus	None	Anterior sagittal anorectoplasty	Normal urination & stooling
Banu (2009) [8]	3 days	Passing meconium with urine	Ectopic anus	None	Anterior sagittal anorectoplasty	Normal urination & stooling
Rintala (1996); 4 patients [6]	Neonate (1) Childhood (3)	Meconium in urine	Normal anus Ectopic anus	Cardiac, renal, vertebral, TEF, malrotation, limb anomalies	PSARP in 2 cases & anterior perineal in the other 2 cases	Unknown
Sharma (2002) [20]	3 years	Recurrent UTI, pass urine from anus	Normal anal site	Absent left kidney, spina bifida, sacral agenesis	anterior perineal	Normal urination & stooling (after 3 months)
Tiwari (2017) [21]	7 months	Passing fecal material with urine, ribbon-shaped stool through the stenosed anus	Normal anal site	Left talipes equinovarus, anal stenosis, Coloboma, ASD	PSARP	Normal urination & stooling
Al-Bassam (1998) [7]	5 years	Passing urine with stool, watery stool	ectopic	Pulmonary valve stenosis	Anterior perineal approach	Normal urination & stooling 8 months postop

Table1: Review of the literature for similar reported cases.

The patient was diagnosed with milk allergy (due to failure to thrive), and a medical formula was prescribed to him, but he remained malnourished and constipated. At the age of 9 months, which was the time of the first presentation to the pediatric surgery center, he was marasmic, and on examination, the following findings were observed:
- On perineal inspection, the shape and location of the anal orifice were normal (Figure 2).
- The gluteal region was well developed (Figure 2).
- On digital rectal examination, the anal canal was normal for about 1 cm, and above it, the rectum was severely stenosed, permitting the passage of 8 French dilators only.
A voiding cystourethrography (VCUG) was done, revealing reflux of contrast to the colon (Figure 3).

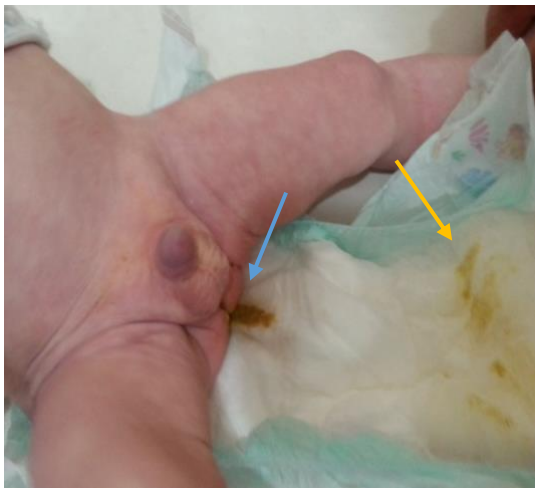

Figure 1: Evidence of fecal material mixed with urine in the diaper (marked by a yellow arrow) with a ribbon-like stool (blue arrow).

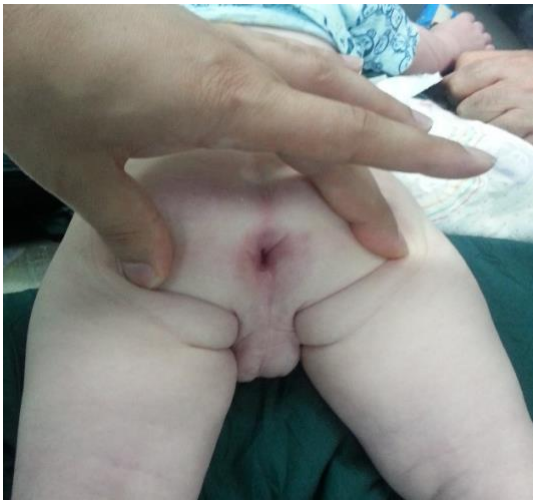

Figure 2: Normal shape and site of the anal orifice, with a well-developed gluteal region.

After preparation, a diverting double-barreled colostomy was done as an initial procedure. A distal colostogram was conducted to delineate the anatomy of the rectum and distal colon, which showed reflux of contrast to the urinary bladder (Figure 4). After one month, the definitive surgery (posterior sagittal anorectoplasty with ligation and division of fistula) was performed (Figure 5 (a,b)). The rectal dilatation program started two weeks postoperatively, and after completion of the dilatation program, the stoma was closed. Now, the patient is five years old and well thrived, and he is continent for urine and stool with a dramatic reduction in the frequency of urinary tract infections (UTIs).

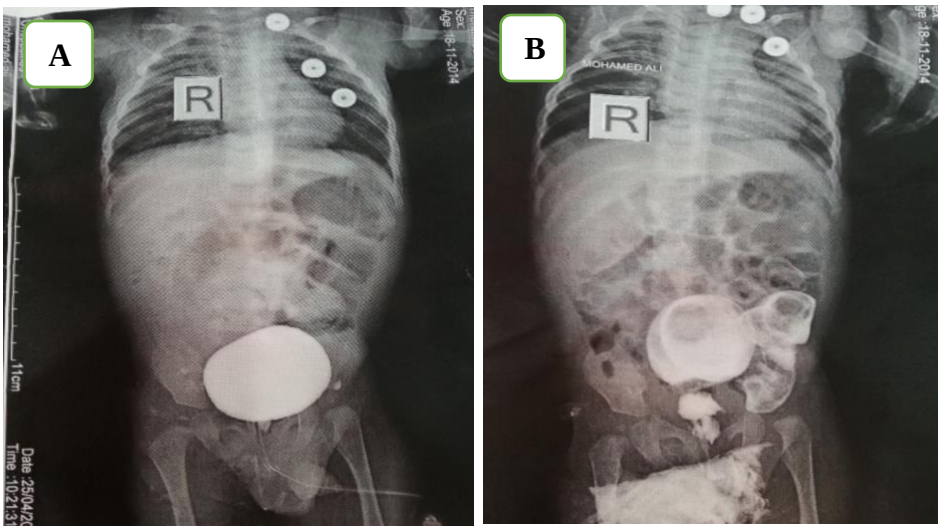

Figure 3: (A) VCUG during filling phase (normal). (B) VCUG during micturating phase shows reflux of contrast to the rectum.

DISCUSSION

H-type rectourethral fistula with anal stenosis is a rare variant of ARMs [4]. The incidence of this anomaly is higher in Asian than that in western countries with a female predominance [3-5,8]. DeVries and Friedland first described this anomaly in 1974 [11]. The embryological fact of H-type ARM in males is due to the misalignment of septal components between the anorectum and the lower urinary tract [12]. In females, this defect has been reported to follow some inflammatory process, and many authors have described this defect as a double termination of the alimentary canal, perineal canal, or H- or N-type fistula [3.13]. The associated renal or intestinal anomalies (TEF) and even VACTERL spectrum have been reported in male patients with an H-type ARM because of similar septation abnormalities [4]. Sharma et al. have summarized these associated anomalies in the acronym VACTERG: vertebral, anorectal, cardiac, tracheoesophageal, renal, and genitourinary [14]. Lang EK has reported a rare association with H-type tracheoesophageal fistula in only two cases [15]. The diagnosis of H-type ARM in males is challenging with the presence of a normal anal opening; however, the abnormal (ectopic) anus should raise the suspicion of this anomaly, especially when associated with recurrent UTIs [14].

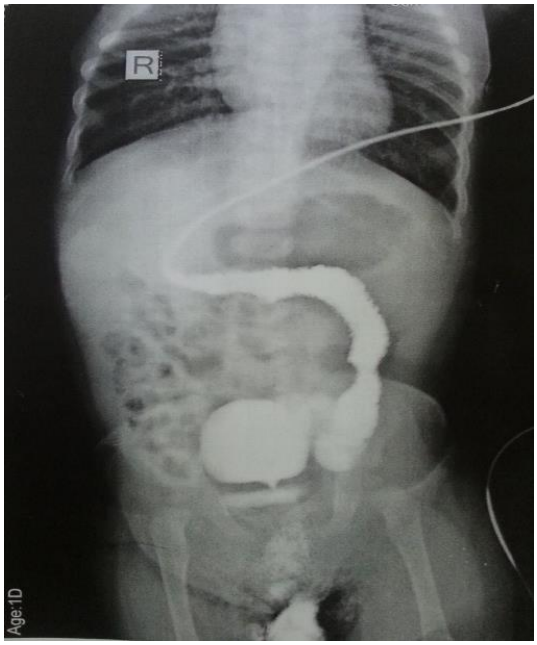

Figure 4: Distal colostogram showing the passage of contrast to the urinary bladder.

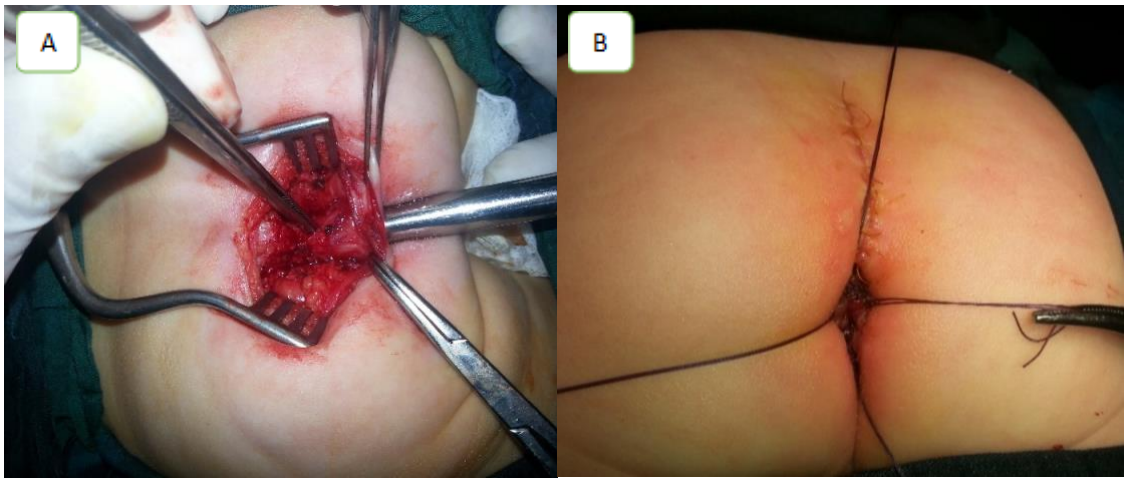

Figure 5: (A) The Hegar dilator passed inside the anus for about 1 cm only, and the pointed area is the site of stenosis. (B) The final result after PSARP.

In females, the passage of stool out of the vagina is the usual presenting feature. The diagnosis can be confirmed by contrast study (VCUG and barium enema), MRI, and endoscopy, revealing the fistulous connection between the rectum and urethra [5,7]. However, these cases are usually diagnosed late (as in H-type tracheoesophageal fistula) because of the lack of external findings [16,17]. Different surgical techniques are described for the management of H-type ARM, including posterior sagittal anorectoplasty (PSARP), anterior sagittal anoplasty (ASAP), anterior perineal (sagittal) anorectoplasty, and anterior perineal repair (APR) (Table 1).

The postoperative functional outcome was excellent as the patients achieved normal urine and stool continence because of the normal anatomy of anal sphincters, with a notable exception of patients with the associated multisystem congenital malformations [5,19-21].

CONCLUSION

Diagnosing an H-type rectourethral fistula with anal stenosis is challenging for pediatric physicians, and a high index of suspicion is required to reach the diagnosis as the anal opening has a normal shape and location. If this condition is diagnosed and treated early, these patients will have an excellent functional prognosis regarding fecal and urinary continence because the muscle complex and anal sphincter are well developed.

LIST OF ABBREVIATIONS

ARMs: Anorectal malformations**.**
PSARP: Posterior saggital anorectoplasty.
VCUG: Voiding cystourethrography.
TEF: Tracheoesophageal fistula.

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CONSENT FOR PUBLICATION

Written informed consent was obtained from the patient's parents to publish this case report with the accompanying images.

CONFLICTS OF INTEREST

The author declares that there are no conflicts of interest regarding the publication of this article.

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