Original Article

Understanding H-type anorectal malformation in females for a suitable surgical approach – A single center experience from central India

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ABSTRACT

Background: H-type anorectovestibular fistula is a rare anorectal malformation (ARM) in females where the child passes stool through a vestibular opening in addition to the normal anus. The diagnosis is easy and largely clinical but controversies exist regarding the most suitable surgical approach and technique to minimize complications. **Objectives:** This study aims to discuss the different possible presentations of the anomaly and the most suitable surgical approach to it. **Materials and Methods:** We analyzed a total of ten patients with this condition treated over a period of 3 years from January 2017 to December 2019. All underwent definitive surgery by the anterior perineal approach without any protective colostomy. They were analyzed with respect to age, presentation, coanomalies, fistula location, internal opening to anus distance, wound related complications or recurrence, need for colostomy later, and presence of constipation over 8 months of follow-up. **Results:** Incidence was 8.77% (10/114) of all ARMs in females. Mean age at presentation was 47.1 days and 3 (30%) had coanomalies. Vestibular staining was the most common manifestation. Mean internal opening to anus distance was 2.04 cm. Two (20%) had wound breakdown postoperatively and one (10%) had recurrence in long-term follow-up. These patients underwent colostomy. Two had constipation but all allowed adequate dilatation during mean follow-up of 8 months. **Conclusion:** H-type ARM in females can be suitably managed with the anterior approach. However, a proximal colostomy appears to significantly decrease the complication rate of the definitive surgery.

Key words: Anorectoplasty, Anorectovestibular fistula, Colostomy, Dehiscence, H-type fistula

An H-type anorectovestibular fistula is an entity where there is an orifice in the vestibule discharging stool in addition to a normally placed anal opening. It is an extremely rare anorectal malformation (ARM) and its exact incidence has not been cited. The first case was reported by Bryndorf and Madsen in 1960 [1]. Similar case reports were later published by Sitkovskii and Pegum in early 1960s [2,3]. Varied opinions exist regarding the most suitable definitive procedure – whether fistulotomy, anterior anorectoplasty, posterior sagittal anorectoplasty, or a vestibuloanorectal pull through [4-9]. Furthermore, there are arguments if the definitive procedure should be preceded by a protective colostomy to avoid wound breakdown [1,10,11]. In spite of different techniques mentioned so far, there is no clear outline regarding the optimum management of this condition. This study revisits this condition through its multiple facets and attempts to throw light on the most effective management.

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MATERIALS AND METHODS

It was a prospective observational study conducted in the Department of Pediatric Surgery in a tertiary care center of Central India. Informed consent was taken from all and ethical clearance was taken from the institutional ethics committee. Details of a total number of female ARMs admitted over 3 years extending from January 2017 to December 2019 were collected and incidence calculated for H-type fistula. VACTER-L association was assessed by thorough clinical examination, ultrasound (US) of kidney ureter bladder region, echocardiography, and US spine for neonates and magnetic resonance imaging spine for those beyond neonatal age. The most common complaint was noted and local examination done. We did not do any contrast

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fistulogram. The labias were retracted upward and outward to visualize any apparent third orifice in the vestibule. In addition, per rectal examination was done with the little finger smeared with lignocaine jelly and methylene blue and vestibule examined for any dye.

A primary anterior perineal approach without prior colostomy was undertaken in all. Preoperatively, the patients were kept nil-per-os (NPO) for 24 h. Intravenous (IV) fluids and antibiotics were started. We did not give any rectal washes. In lithotomy position, a probe was gently inserted through the fistula which extruded from the normal anus. Incision was made from the fistula above to the anterior margin of the normal anus below. All tissues were divided by electrocautery exactly in the midline staying on the probe. This was continued till the fistula tract lay open and the internal orifice noted. The distance between internal orifice and normal anus was measured. The mucosa was completely excised and the defect closed with Vicryl 4-0 sutures in interrupted fashion. Stay sutures were placed at the edge of the anterior half of detached rectum and with due traction on it, the rectum was separated from the vagina. Optimum mobilization of the rectum was done till it reaches the skin surface without tension. Anoplasty was done with Vicryl 4-0. All tissues were opposed in the midline with Vicryl 3-0 to restore the vestibule to its normal appearance (Figs. 1-3).

Postoperatively, patients were kept NPO for 3 days and orals allowed from post-operative day 4 onward. IV antibiotics (third-generation cephalosporin, metronidazole, and amikacin) were continued for 5 days. Gentle cleansing wash of the wound with 20% betadine saline was undertaken in the post-operative period and nursing done in prone or lateral position. The wound was examined and classified as "healthy" or "unhealthy." Healthy wounds were those with intact anoplasty sutures even if there was skin level break down. Unhealthy wounds were those in which anoplasty sutures had given away with retraction of the anal mucosa deep in to the margins of the incision. These underwent diversion sigmoid colostomy.

Anal dilatation was started after 21 days. Colostomies were closed 6–8 weeks later. Patients were followed up for a mean duration of 8 months. Outcome was assessed functionally in terms of the presence of constipation or not. Soiling was not taken as a parameter as our follow-up was relatively shorter till the completion of which many children would not get toilet trained.

Quantitative data were extracted in to an electronic spreadsheet and statistical analysis performed using SPSS Statistics base 22.0 software (IBM Inc., New York, USA). Results were presented as range, mean, and standard deviation (SD).

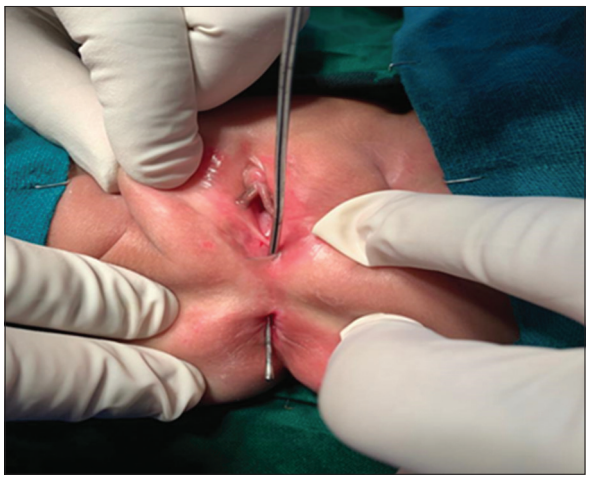

Figure 1: Probe demonstrating the H-type fistula

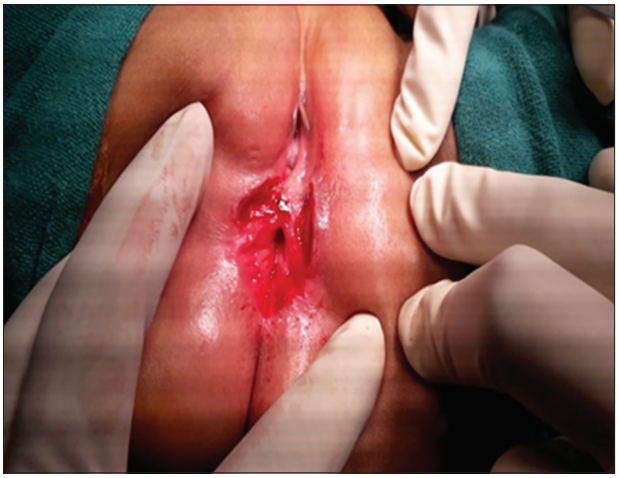

Figure 2: Fistula tract laid open exposing the lining mucosa which was excised

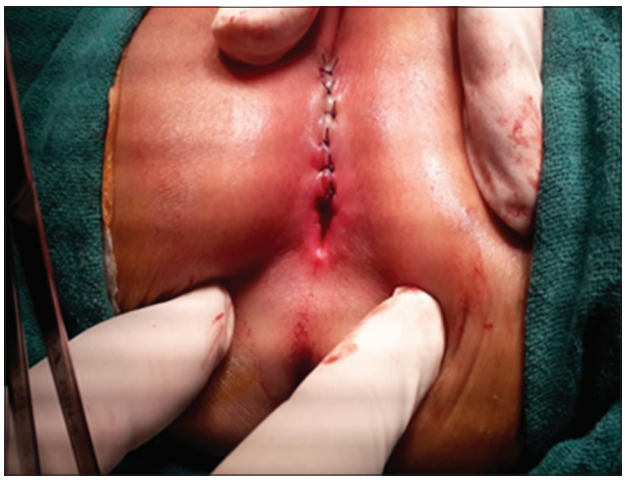

Figure 3: Midline apposing of tissues from vestibule to normal anus

RESULTS

A total of 114 female ARM were admitted. H-type fistula was diagnosed in 10 (8.77%) of them. Age varied between 3 days and 217 days with mean of 47.1 days and SD of 82.37. Coanomalies were found in 3 (30%) which were cardiac septal defect in 2 (20%) and hydronephrosis in 1 (10%). The most common complaint by the mother was stool emanating from the vestibule in addition to the child passing normally per anum. This was seen in 8 (80%). In all of them, we could clinically appreciate an ectopic anal opening in the vestibule below the vagina in midline. Methylene blue staining of this orifice was seen on per rectal examination. Other 2 (20%) were referred from pediatrics department with sudden onset of watery stool from the vestibule during an attack

of acute gastroenteritis. These were comparatively older children, their age being 188 and 217 days, respectively. In them, the fistula was clinically not very obvious and could be demonstrated quite laterally below the labia after retracting them. These two patients underwent surgery after 2 weeks of antibiotics and resolution of diarrhea. Intraoperatively, the internal opening to normal anus distance varied between 1.5 cm and 3 cm, mean being 2.04 cm and SD 0.56. Three (30%) developed complications.

Two had immediate post-operative complication in the form unhealthy wound with fecal contamination. One had delayed complication at 5 months after surgery as fistula recurrence during an acute diarrheal illness. Fecal diversion was accomplished by a colostomy in them. In 8 (80%), the functional outcome was satisfactory as they continued to pass well-formed stool without any need of washes or enemas and no fecaliths were palpable per abdomen. Hard stools requiring washes were seen in 2 (20%). Patient profile, per operative findings, and outcomes are mentioned in Table 1. There were no stenosis and all neo anuses allowed an age standardized Hegar's dilator during follow-up.

DISCUSSION

H-Type anovestibular fistula is an unusual form of female ARM. Literature shows a relatively higher incidence of 4–14% in Asian continent [9,11,12]. Incidence in our series (8.77%) was in the same bracket. This higher incidence is probably due to some geographical predisposition that has not yet been proved conclusively. Among Asian countries, majority of the cases have been reported from India [6,10-12] and Japan [9,13,14]. Most of the international classifications include this anomaly as "low" type but the term "H-type" was first recognized in Krickenbeck classification (2005) that grouped it as rare or regional variants. The term "double termination of the alimentary tract" was used synonymously but confusion with rectal duplication has led to its discontinuation. The relatively less common acquired form usually arises secondary to an inflammatory process of the anovestibular [15].

The presentation is that of a normal female baby with fecal stains in the vestibule apart from passing stool per anum. On examination, a third orifice in addition to the urethra and the vagina can be seen which at times may be minute. It may not necessarily be in midline and a lateral opening underneath the labia minora is not very uncommon that becomes obvious only when inflamed with or without labial abscess [4,16]. This abscess can rupture spontaneously causing fecal discharge, more so when the gut is inflamed as in gastroenteritis. Such clinically obscure fistulae present late beyond the neonatal period as seen in two of our patients. While examining, a probe can almost always be introduced through the normal anus and the fistula track up to the vestibule delineated. A fistulogram is not required and is not expected to give additional information [17]. When in doubt, per rectal examination can be done with finger smeared with dye and pressed anteriorly toward the vestibule. Any occult fistula is then likely to open up and get stained. Demonstration of the fistula with such maneuver was possible in all of our cases.

Initial poor understanding of the anatomy due to its rarity led to this condition being explained in various ways. Stephens and Smith coined the term "perineal canal" [18]. This was due to the fact that in their series, they had found the fistula opening low down in the anal canal. Practically, however, a more complex morphology have often been seen with the fistula going high above the levator ani. Chatterjee proposed a classification which was more relevant from management point of view [6]. Those where the fistula terminated in the rectum above levator ani were called high H-type fistula or rectovestibular fistula and those terminating in the anal canal below the levator ani were called low H-type fistula or anovestibular fistula. The low type is found to be more common with the fistula opening in to the anal canal approximately 2–2.5 cm above the anal verge [1-3,19]. This finding corroborated well in our study.

Surgery for this condition has evolved over last three decades. Chatterjee treated the low H-type fistula by a simple perineal operation where the subcutaneous fistula lining was laid open [6]. This method was associated with high recurrence due to the left over mucosa of the fistula tract in and gave an ugly vestibular scar. For the high type, he described a technique where vestibulo-rectal pull-through was performed by a circumferential

Table 1: Patient profile, intraoperative finding, and outcome

Age (Days)	Coanomaly	Presentation	Fistula location in vestibule	Internal opening to anus distance (cm)	Surgical wound	Colostomy	Recurrence	Constipation
3	None	Stool in vestibule	Midline	1.8	Healthy	No	No	No
6	ASD	do	Midline	1.5	Healthy	No	No	Yes
4	HDN	do	Midline	2	Unhealthy	Yes	No	No
188	None	AGE f/b stool in vestibule	Lateral	2.7	Healthy	No	No	No
13	None	Stool in vestibule	Midline	1.5	Healthy	No	No	No
8	VSD	do	Midline	1.9	Healthy	No	No	No
21	None	do	Midline	2.2	Unhealthy	Yes	No	No
8	None	do	Midline	2.5	Healthy	No	No	Yes
217	None	AGE f/b stool in vestibule	Lateral	3	Healthy	Yes (Later)	Yes	No
3	None	Stool in vestibule	Midline	1.3	Healthy	No	No	No

ASD: Atrial septal defect, HDN: Hydronephrosis, VSD: Ventricular septal defect, AGE: Acute gastroenteritis

vestibular incision. However, two out of his nine patients had a wound dehiscence followed by recurrence of the fistula. Tsuchida described two types of perineal procedures [9]. Three infants were treated by a simple perineal operation but there were recurrence in two. The second operation performed in other seven patients was named "pull-through of the anterior wall of the rectum." Four of these seven patients were on colostomy. In this technique, anterior half circumference of the anorectum was excised along with the fistula. The proximal freed rectum was mobilized down and sutured at the anal verge. None of these seven patients had recurrence. Different techniques thus proposed by different surgeons and complications specific to each thus gave rise to two very pertinent questions. First, what is the best approach? Second, should the definitive surgery be preceded by a colostomy to avert the wound related complications?

The anterior perineal approach is a relatively simple technique for repairing this condition. Its suitability is justified by two particular reasons. First, by this technique, the mucosal lining of the fistula tract can be excised completely which is very important to prevent any recurrence. Second, separation of the anterior anorectum from vagina leads to a tension free anorectoplasty. However, we found that the common wall separating the vagina and the anal canal extended not only inferiorly but also superolaterally up to variable distance. Thus, we feel that an adequate surgery and a tension free anoplasty will only be possible by a near circumferential dissection of the anorectum and not only the anterior part. Adequate dissection will also reduce the probability of stenosis and constipation later. We followed this maneuver in all our patients. All of them had a low or intermediate type of anomaly and rectum could be brought to the verge without tension. However, wound dehiscence was seen in two patients. In these two patients, the lateral common wall between rectum and vagina was very thin which compromised the dissection due to fear of injuring the rectum. Recurrence was encountered in another one. This was the oldest child in our study group who had an acquired fistula and was also diagnosed of an immunodeficiency disorder.

The recurrence probably can be attributed to repeated attacks of gastroenteritis with a chronically inflamed rectum and ischemia of the previously closed internal opening. A prior diversion in the form of sigmoid colostomy therefore appears to be a judicious plan that would allow a complete dissection later. This shall decompress the distal rectum and also would not cause much worry if rectal serosa is injured during dissection later in the definitive surgery. Authors acknowledge the fact that the sample size is small due to rarity of the anomaly and an emphatic conclusion can only be drawn on further studies with bigger sample and follow-up beyond the toilet trained years.

CONCLUSION

H-type anorectovestibular fistula is a rare form of female ARM. The anatomy can be difficult to understand and surgery may be associated with significant morbidity in terms of wound dehiscence or recurrence. The anterior perineal approach as a definitive surgery is easy, reproducible and allows satisfactory dissection of the anorectum all around. An upfront surgery however may have complications depending on factors such as proximity of rectum and vagina limiting the local dissection. A protective sigmoid colostomy would negate this factor with better outcome.

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