Anorectal Malformations in Adolescent Females: A Retrospective Study
Jileedar Rawat, Sudhir Singh, Nitin Pant

Context: While most of the females with anorectal malformations (ARMs) present in either neonatal period or early infancy, a small percentage presents in their adolescence.

Aim: The aim is to study the causes of delay in presentation, management, and postoperative outcome in these cases.

Settings and Design: This is a retrospective observational study conducted in the Department of Paediatric Surgery, King George’s Medical University, Lucknow.

Subjects and Methods: Records of females with ARM managed in one unit of the department from 2010 to 2015 were scrutinized. Of these, record of females who primarily presented in adolescence was reviewed. Data regarding the demographics, clinicoradiological presentation, management, postoperative stay, and follow-up were analyzed. Causes for delay in presentation were looked for.

Statistical Analysis Used: Statistical analysis used was not required.

Results: Out of 627 cases of ARM managed over 5 years, ten girls (5.3%) presented between 12 and 18 years (average 14.4 years). Main reasons for the delay in presentation were misinformation, illiteracy, and poverty. These comprised of anovestibular fistula (n = 5), anterior ectopic anus (n = 3), perineal canal (n = 1), and rectovestibular fistula (n = 1). Three (30%) girls had no problem apart from an abnormal anal orifice. Five (50%) girls had constipation as a major additional symptom. Four cases had a normal to mildly enlarged rectum on contrast study, whereas the rectum was moderately to hugely dilated in the remaining six cases. Six cases were managed with posterior sagittal anorectoplasty while an anterior sagittal anorectoplasty was done in four. Minor wound dehiscence developed in two cases. There was no mortality. In a mean follow-up of 16 months (8–26), constipation and soiling were seen in 5 (50%) girls. The overall outcome in these girls was satisfactory.

Conclusion: Presentation of females with ARM in adolescence is not uncommon in the third world. A primary pull through is possible in these girls with mild-to-moderate rectal dilatations. A diverting colostomy before pull through is always a safe option in cases with severely dilated rectum and also otherwise. Moreover, a greater awareness regarding these malformations in the general public is required.

Keywords: Adolescent female, anorectal malformation, anterior sagittal anorectoplasty, posterior sagittal anorectoplasty

Address for correspondence: Dr. Sudhir Singh, Department of Paediatric Surgery, King George’s Medical University, Lucknow - 226 003, Uttar Pradesh, India. E-mail: drsudhir_singh25@yahoo.in

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age. In one study, adolescent ARMs comprised around 15%–20% ARM cases. Management of these cases is challenging as they are uncommon, sparingly reported in literature, and there is a lack of information in available literature regarding their management and outcome. In this study, we describe the presentation, management, and outcome of adolescent females presenting with ARM.

Subjects and Methods

Records of all females with ARM managed in the department from 2010 to 2015 were scrutinized. Of these, record of females who presented for the first time in adolescence was reviewed. Data regarding the demographics, clinicoradiological presentation, management, postoperative stay, and follow-up was analyzed. Causes for delay in presentation were looked for. A contrast barium enema was performed in all along with an ultrasound kidney, ureter and bladder, echocardiogram, and an X-ray spine. Parameters studied were cause of delay, type of ARM, main clinical problems, surgical procedure and intraoperative problems, and outcomes. The choice of approach, whether anterior sagittal or posterior sagittal, was based on the rectal dilatation as seen on barium enema. Cases with no constipation, a normal caliber, or mildly dilated rectum on a contrast enema were managed with a primary pull through by an anterior sagittal approach [Figure 1d-f]. Those with moderately dilated rectum on contrast enema were treated by primary posterior sagittal approach [Figure 1a-c]. These patients were admitted at least 3 days in advance, and a preoperative bowel preparation with polyethylene glycol solution was started. Postoperatively, these cases were kept nil orally for at least 5 days.

Girls with a severely dilated rectum on contrast enema were initially managed with a diverting loop sigmoid colostomy, followed by distal rectal washouts in the postoperative period. Definitive pull through was done 2–3 months later with posterior sagittal approach. Stoma closure was done after 2–3 months of pull through. Postoperative anal dilatation (with a St. Marks Rectal Dilator No. 4) was started after 2 weeks. Follow-up was as recorded at the last outpatient visit.

Results

A total of 627 cases of ARMs were managed at our center over 5 years. Of these, 439 were male and 188 were female. Ten girls (5.3%) presented between 12 and 18 years (average 14.4 years). Main reasons for the delay in presentation were misinformation, illiteracy, and poverty as all patients were from the lower socioeconomic strata. The primary concern for which all the parents came for consultation was the abnormal anal opening and its impact on her prospective marriage. In seven cases, the parents were aware of the condition of their child since birth. Five patients had an anovestibular fistula, three had an anterior ectopic anus, one had a perineal canal, and one had a rectovestibular fistula. Five (50%) girls had constipation as a major symptom at presentation, one presented with features of a distal colonic obstruction while four were decompressing well at the time of presentation. Details regarding the clinical presentation and management are given in Table 1. Four cases had a normal to mildly enlarged rectum on contrast study, whereas the rectum was moderately to hugely dilated in the remaining six cases.

One girl who with perineal canal also had constipation at presentation. The parents knew of the girl’s condition since early infancy and she did not have constipation to begin with. The constipation was of 5-year duration. Per-rectal examination revealed fecoliths. Most likely, it was functional constipation with pseudocontinence since soiling and foul smell were present. However, then (again), the soiling could also be attributed to the fistula.

In six cases, a PSARP was performed while ASARP was done in four. Distal rectal wall tear occurred in two cases while doing PSARP. Both were managed by incorporation of tear in the anoplasty. No vaginal wall tear occurred in any of our cases. Wound infections with minor wound dehiscence developed in two cases post-ASARP. These two cases later presented with anal stenosis. There was no mortality. Colostomy closure was done at average interval of 10 weeks following the perineal procedure. At follow-up, assessment of size of anal opening, anal tone, and bowel habits was done. In a mean follow-up of 16 months (8–26), constipation and soiling were seen in 5 (50%) girls. Of these, two had anal stenosis requiring anoplasty. The rest were managed
with laxatives (polyethylene glycol or lactulose). The overall outcome in these girls was satisfactory.

**Discussion**

ARMs have an incidence of 1 in 5000 live births with a male preponderance. In females, vestibular fistula and ectopic anus are the most common varieties. Whereas most females present in neonatal period, some have been reported to present at adolescent age. In our study, 5.3% of cases presented in adolescence age. Despite the fact that in 7/10 cases the parents knew of the patient condition since early infancy, there was a substantial delay to seek treatment. The primary concern for which all the parents came for consultation was the abnormal anal opening and its impact on her prospective marriage. Five cases had constipation as an additional presenting symptom. Major reasons for the delay in presentation in our understanding were illiteracy, misinformation, and poverty since all these patients belonged to villages and had a poor socioeconomic condition. Reported reasons for a delayed presentation in one series are either delayed awareness on the part of parents or poor access to the appropriate facilities. Associated congenital anomalies were found only in one patient (10%), as opposed to the figure of 40%–50% reported in low and intermediate types of ARM.

There is a paucity of literature on the management and outcome of female ARM cases operated in adolescent or adult age group and no standard surgical protocol is available. These girls offer their own surgical challenges. A longer common channel, higher placed rectum, and tougher tissue for dissection make the surgery somewhat more difficult and time-consuming with greater amount of bleeding as compared to pull through in infants.

Both ASARP and PSARP are standard approaches for management of female infants with ARM. We found both approaches equally good in terms of exposure and ease of dissection in adolescent females though we admit our bias in selection of approach in these patients. We opted for ASARP in normally functioning and mildly dilated rectum and PSARP for moderately to severely dilated rectum. This was due to the greater exposure said to be provided by posterior sagittal approach. Second, our reason that tapering proctoplasty...
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if required would be easier with a posterior approach in adolescent girls with deep pelvis. Moreover, the use of ASARP is not much described for adolescent age group in literature. Somehow, there was no requirement for tapering in cases of three girls with moderately dilated rectums and all were managed by a primary PSARP. In hindsight, one wonders whether they could also have been done by ASARP approach. We admit that preferentially selecting cases with dilated rectums for PSARP and those with normal rectum for ASARP is a surgical bias on our part, but then, this is not a comparative series. ASARP was also quite effective and provided good exposure in the four cases where it was used. Moreover, putting adolescent/adult females in lithotomy position is relatively easier than turning her over in the prone jackknife position.[7]

Two patients developed rectal wall tear intraoperatively at PSARP. The tear was incorporated in the anoplasty in both. Two patients developed wound infection with minor wound dehiscence following ASARP. These two patients went on to develop anal stenosis which later on required an anoplasty. Interestingly, they were cases of anovestibular fistula and were decompressing well before surgery. The position of anal orifice was the only complaint. These two cases make us rethink on our decision to operate on them. There was no incidence of vaginal tear.

Constipation was the main problem over long term, seen in five patients. Two developed it following anal stenosis. The other three patients were the ones where a tapering proctoplasty had been done. These patients had a colostomy to begin with. Still, rectum was dilated at the time of pull through. Despite the tapering, it did not function well. All the three patients presented with soiling. It could be that our tapering was inadequate in either caliber or length. Another possibility could be that some of the massively dilated rectosigmoids become redundant over time. Initially, they were managed with enemas and lactulose, while later, we switched over to polyethylene glycol which we have found a better choice in these cases.

The cosmetic outcome as assessed by the appearance of perineum and calibration of neoanus was near satisfactory in all. Although the cosmetic outcome is reported to be the superior in ASARP,[2,8,9] we did not find much difference between the two procedures in our series. The two girls with wound dehiscence had an acceptable perineum following anoplasty.

**Conclusion**

Presentation of females with ARM in adolescence is not uncommon in the third world. Whereas in experienced hands, a primary pull through is possible in these girls with mild-to-moderate rectal dilatations, a diverting colostomy is a safe option in cases with severely dilated rectum. Nevertheless, a clear treatment approach for these is required in cases as they pose their own surgical and postsurgical problems. Moreover, a greater awareness regarding these malformations in the general public, especially in the lower strata, is required to prevent late referral of these cases.

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**Conflicts of interest**

There are no conflicts of interest.

**References**

1. Zamir N, Mirza FM, Akhtar J, Ahmed S. Anterior sagittal approach for anorectal malformations in female children: Early results. J Coll Physicians Surg Pak 2008;18:763-7.
2. Sham M, Singh D, Phadke D. Anorectal malformations: Definitive management during and beyond adolescence. J Indian Assoc Pediatr Surg 2012;17:120-3.
3. Levitt MA, Peña A. Anorectal malformations. Orphanet J Rare Dis 2007;2:33.
4. Sinha SK, Kanojia RP, Wakhlu A, Rawat JD, Kureel SN, Tandon RK, et al. Delayed presentation of anorectal malformations. J Indian Assoc Pediatr Surg 2008;13:64-8.
5. Mittal A, Airon RK, Magu S, Rattan KN, Ratan SK. Associated anomalies with anorectal malformation (ARM). Indian J Pediatr 2004;71:509-14.
6. Wakhlu A, Kureel SN, Tandon RK, Wakhlu AK. Long-term results of anterior sagittal anorectoplasty for the treatment of vestibular fistula. J Pediatr Surg 2009;44:1913-9.
7. Aziz MA, Banu T, Prasad R, Khan AR. Primary anterior sagittal anorectoplasty for the treatment of vestibular fistula. Asian J Surg 2006;29:22-4.
8. Rasool N, Khan MA, Aslam M, Safdar A. Anterior sagittal anorectoplasty: the treatment of anorectal malformations in female children. Prof Med J 2014;21:845-50.
9. Harjai MM, Sethi N, Chandra N. Anterior sagittal anorectoplasty: An alternative to posterior approach in management of congenital vestibular fistula. Afr J Paediatr Surg 2013;10:78-82.