



A STUDY ON DIAGNOSTIC MODALITIES, MANAGEMENT AND OUTCOME OF ANORECTAL MALFORMATIONS

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ABSTRACT

AIM & OBJECTIVES - Anorectal malformation (ARM) is a common congenital condition that has a significant impact on the quality of life for children. It is more frequently observed in females. The primary posterior sagittal anorectoplasty (PSARP) procedure has shown excellent outcomes, particularly for males with intermediate and high ARM, as well as females with vestibular fistula. The objective of the study was to identify the specific type of ARM and associated anomalies in female patients. **METHODS**- This observational study was carried out at the Department of Surgery, Index Medical College and Hospital, involving patients with ARM between November 2019 and October 2021. The initial procedure involved performing colostomy, followed by PSARP/ anterior sagittal anorectoplasty technique (ASARP) as a second-stage procedure, and subsequently closing the colostomy. Patients were typically discharged between the 4th and 6th postoperative days and followed up with regular anal dilatation and assessments every six months. **RESULTS**- The present study findings indicate that the most common defect observed in females with ARM is the rectovestibular fistula. Evaluation of surgical outcomes can be performed following the completion of the final procedure, namely colostomy closure. Among the patients assessed, approximately 83.3% demonstrated favorable post-operative outcomes. **CONCLUSION**- Females have a higher incidence of intermediate ARM compared to other varieties. Low anomalies generally exhibit more favorable outcomes following surgery than other types. The utilization of the PSARP approach in treating patients with these conditions provides a more precise correction of the lesion.

KEYWORDS : Fistula, malformations, rectovaginal, rectovestibular

INTRODUCTION

Anorectal malformation (ARM) is a prevalent congenital condition that significantly impairs the quality of life for children, with a prevalence rate of 6.5%.¹ There is a higher incidence of ARM in females, with a sex ratio of 2:1, indicating that females are more commonly affected by this condition compared to males.² The presentation of ARM can manifest in various anatomical configurations, primarily influenced by the presence and position of a fistula. The location or specific characteristics of the fistula, typically situated between the gastrointestinal tract and the genitourinary tract or perineum, often play a crucial role in determining the appropriate type and timing of surgical intervention.³

Early detection of congenital ARM is vital for timely and imperative treatment of infants affected by these conditions. Patients diagnosed with this condition lack a typical anal opening and instead have a fistulous tract that opens either anteriorly to the anal muscle complex onto the perineum or into neighboring anatomical structures. In males, the fistulous tract may connect with the urinary system, while in females, it may connect with gynecologic structures.⁴ In the field of surgery, there are two approaches commonly used by surgeons to address ARM; the primary definitive repair performed in a single stage, or the traditional three-stage repair. While the single stage repair offers convenience with only one surgical intervention, it is associated with higher rates of wound dehiscence and potential loss of sphincter function due to fibrosis, as documented in previous studies.⁵⁻⁷ However, recent scientific literature presents compelling evidence that the primary posterior sagittal anorectoplasty (PSARP) procedure, although requiring a more skilled surgeon and anesthesiologist, yields excellent outcomes specifically for males with intermediate and high ARM, as well as females with vestibular fistula.⁸

General condition, duration of presentation and number of openings in the vestibule decide the management. Gross abdominal distension, sepsis, or single opening (persistent cloaca) warrants a diversion colostomy. The most common anomaly in females is a rectovestibular and shows a normal urethra, normal vagina, and another orifice, which is the rectal

fistula in the vestibule. Such cases can be managed by a diversion colostomy and delayed definitive repair by PSARP. In selected cases, a primary PSARP or a primary anterior sagittal anorectoplasty can also be performed. Definitive repair of persistent cloaca is performed through PSARP. In cases with common channels longer than 3 cm, it is difficult to mobilize the vagina through PSARP, and an abdominoperineal approach is required. In cases with a common channel of <3 cm, total urogenital mobilization is possible, in which both the vagina and urethra are mobilized as a unit, without separation. If the distance from the vagina to the perineum is long, a bowel segment can be used to bridge the gap, preferably a segment of the colon or a vaginal switch procedure can be done in cases with bicornuate uterus.

ARM poses not only a surgical complexity but also a distressing experience for parents and the entire family. Despite advancements in the field of surgery, it continues to test the knowledge and skills of surgeons, presenting significant challenges that require wisdom and expertise to address effectively.⁹ In the light of the above context, the present study aimed at diagnosing the type and assessing the management of ARM in female patients.

MATERIAL AND METHODS

This was an observational study conducted at Department of Surgery, Index medical college and Hospital, involving patients with ARM, from November 2019 to October 2021.

The female patients with age group of 2 months to 2 years with ARM were included.

The routine blood and urine examination were performed in all eligible patients. Other investigations like Invertogram, cross-prone lateral view, distal colostogram, ultrasonography, and 2D-ECHO were done. CT and MRI were performed to assess the anatomical status of sphincter, type of ARM, type of fistula, the developmental state of the SMC and developmental anomalies.

Preoperative procedures (Figure 1)

A detailed case history was recorded as per the proforma. All

the eligible patients underwent detailed local examination of the perineum; presence or absence of anal opening, fistula, external genitalia and spine examination. A routine USG was performed to assess any genitourinary abnormalities. The routine preoperative procedures like NIL orally, nasogastric tube aspiration, colostomy wash, intravenous fluids and antibiotic injections were administered to all eligible patients indicated for surgery.

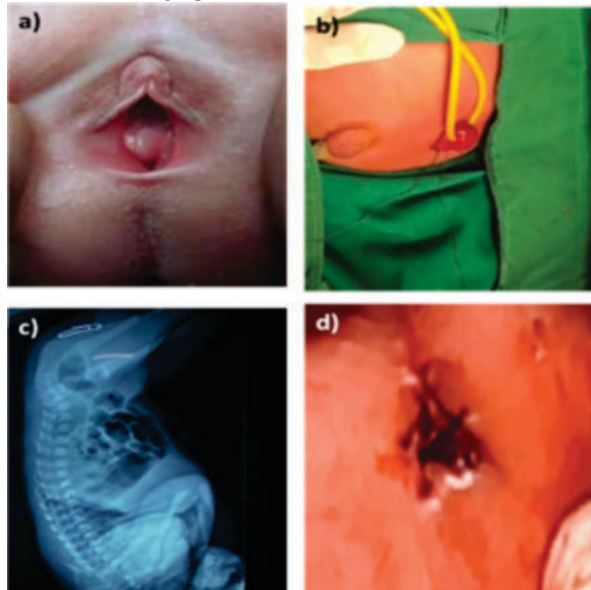


Figure – 1 Representative PREOP Images Of ARM (a) Preop picture of ARM, (b) Colostomy for ARM, (c) Invertogram and (d) Anoplasty

Operative Procedures

In patients with low anomalies, anoplasty, ASARP, and cruciate incision of anal membrane was performed. In patients with intermediate anomalies like recto-vestibular fistula and anal genesis, were treated with colostomy followed by PSARP or ASARP and colostomy closure. In patients with high anomalies, a 3-stage repair with colostomy was performed followed by definitive repair and colostomy closure upon indication.

Postoperative Procedures

After achieving the desired anus size through dilation, the closure of the colostomy was performed through a limited resection and anastomosis. The patient was kept nil by mouth until they were able to have bowel movements through the anus. The patient was discharged from the hospital once they were able to pass stools either through the rectum or the colostomy site, and the surgical area showed no signs of complications. All the patients were discharged between 4th to 6th post-operative delirium, followed by regular anal dilatations with six monthly assessments.

The primary endpoint of the study was to diagnose the type and management of ARM in female patients.

The secondary endpoint of the study was to assess the type of ARM and associated anomalies, diagnostic modalities, management, postoperative outcome and management of complications.

The follow-up period extended up to one year after the definitive procedure. The child's physical and mental developmental milestones were evaluated. The evaluation of sphincter control involved assessing the tone and squeezing ability of the sphincter by examining the finger. Routine dilatation of the anal canal was recommended, and continence assessment was conducted every six months initially, followed by annual assessments up to three years

after the definitive procedure. The evaluation of outcomes was based on the Kelly's score.

RESULTS

A total of six patients were included in this study. Among six patients, four (66.7%) patients presented with intermediate anomalies (rectovestibular fistula, rectovaginal fistula) whereas two (33.3%) patients presented with low anomalies (anal stenosis, anterior perineal anus). Four patients (66.7%) presented with passing of meconium through vagina or vestibule. Three patients (50.0%) of patients presented with rectovestibular fistula and one patient (16.7%) presented with rectovaginal fistula. These baseline characteristics are summarized in Table 1.

Table – 1 Baseline Characteristics In Patients

Parameter	No. of patients (N=6)
Types of anorectal malformations	
High	0
Intermediate	4 (66.7)
Low	2 (33.3)
Modes of presentation	
Passing of meconium through vagina/ vestibule	4 (66.7)
Narrow anal opening	2 (33.3)
Type of fistula	
Rectovestibular	3 (50.0)
Rectovaginal	1 (16.7)
Data shown as n (%).	

The majority of patients (50.0%) with intermediate ARM were managed with sigmoid colostomy and PSARP. Around 16.7% of patients with intermediate ARM were managed with transverse colostomy and ASARP. In patients with low ARM, 16.7% of patients were managed with cut anoplasty and ASARP each (Table 2).

Table – 2 Management Strategies In Different Anomalies

Parameter	Procedure	No. of patients (N=6)
Management of intermediate ARM		
Sigmoid colostomy	For 2 nd stage	3 (50.0)
PSARP	For 3 rd stage	3 (50.0)
Transverse colostomy	For 2 nd stage	1 (16.7)
ASARP	For 3 rd stage	1 (16.7)
Management of low ARM	-	
Cut anoplasty		1 (16.7)
ASARP		1 (16.7)
Data given as N (%).		
ARM, anorectal malformations; ASARP, anterior sagittal anorectoplasty technique; PSARP, posterior sagittal anorectoplasty.		

The primary method for evaluating functional outcomes has traditionally been clinical assessment, primarily based on patient history. These outcomes are typically categorized as good, fair, or poor. A patient with a good outcome is defined as being continent most of the time, experiencing only occasional soiling during episodes of diarrhea and physical stress. A fair outcome entails occasional soiling with normal stool consistency, but still maintaining acceptable social continence. On the other hand, a poor outcome is characterized by frank incontinence or the need for a permanent colostomy. To assess the results in present study, Kelly scoring system was utilized. Out of six patients, 83.3% (5 out of 6) achieved good outcomes. One case was lost to follow-up, however, there were no reported deaths. These operative results are summarized in Table 3. Some complications like excoriation and mild erythema were reported around the colostomy site.

Table – 3 Operative Results In Different Anomalies

Type of anomaly	Good	Fair	Poor	LOF
Intermediate	3	-	-	1
Low	2	-	-	-

DISCUSSION

Congenital ARM have been recognized since ancient times, representing the most frequent cause of neonatal intestinal obstruction.⁹ Although the majority of cases of ARMs are typically detected during the neonatal period or infancy, there are rare instances where cases are reported at a later age.¹⁰ Managing these cases poses a challenge due to their infrequent occurrence, limited reporting in the literature, and a lack of comprehensive information regarding their management and outcomes available in the existing literature.¹¹

A study by Rawat et al. observed that the majority of females with ARMs are typically diagnosed and present symptoms during the neonatal period or early infancy, however, a small percentage may experience the presentation of ARMs during their adolescence.¹¹ Considering this valuable insight, the present study included the female patients with age group of 2 months to 2 years with ARM. In the present study, the most common presentation seen was passing of meconium through vagina or vestibule (66.7%), and narrow anal opening (33.3%). These was consistent with another study by Singh et al. which also noted not passing meconium since birth (50.0%), and absent or abnormal anal opening.⁹ According to a study by Halawa et al. conducted in Egypt, the invertogram continues to be the primary diagnostic method employed before surgical intervention.¹² In the present study, invertogram was one of the diagnostic methods used with cross-prone lateral view, distal colostogram, and ultrasonography.

Among the study population in a study by Maqtadir et al., the most frequently utilized definitive operative treatment was PSARP, and the outcomes were deemed satisfactory.¹³ In the present study, the majority of patients (50.0%) with intermediate ARM were managed with sigmoid colostomy and PSARP. To assess the results in present study results, Kelly scoring system was utilized. Out of six patients, 83.3% (5 out of 6) achieved good outcomes. One case was lost to follow-up, however, there were no reported deaths.

The operative repair of ARMs is associated with various complications. Initial postoperative problems commonly include wound infection, dehiscence (wound opening), and strictures (narrowing). Many complications arise due to excessive tension or inadequate blood supply to mobilized structures. Misplacement of the anoplasty procedure outside the sphincter complex and injury to neighboring structures such as the vas deferens, seminal vesicles, ectopic ureters, or urethra can also occur. Conversely, patients with a poor prognosis, such as those with bladder neck fistula, typically exhibit a low incidence of constipation but a high rate of incontinence. However, in this present study, excoriation and mild erythema around colostomy site were noted. This was consistent with a study by Maqtadir et al., in which perineal excoriation was seen in 20.4% of patients.¹³

In this present study, various key observations were noted. In females, there was a higher prevalence of intermediate anomalies compared to other types. Low anomalies generally exhibited superior post-surgical outcomes compared to other varieties. Staged repair is recommended for intermediate and high anomalies, as it yields improved results. The utilization of the PSARP approach in treating these patients allows for a more precise correction of the lesion.

neonates with ARM and the impact of corrective surgery on their quality of life, future studies should be conducted with larger sample sizes and longer follow-up periods. These efforts will provide more comprehensive insights into the experiences of these individuals and the effectiveness of surgical interventions in improving their overall well-being.

The current study had certain limitations that should be acknowledged; the sample size was small, limiting the generalizability of the findings. In addition to this, the follow-up period was short, which may not have allowed for a comprehensive evaluation of long-term outcomes.

CONCLUSIONS

Females exhibit a higher occurrence of intermediate anomalies compared to other types. In contrast, low anomalies demonstrate superior post-surgical outcomes compared to other varieties. The utilization of the PSARP approach in treating these patients allows for a more precise correction of the lesion. It is essential to have a longer duration of follow-up to enable a more comprehensive assessment of post-operative results.

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To gain a deeper understanding of the challenges faced by