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Case Report

Single stage repair of anorectal malformation with rectovestibular fistula in adult

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ABSTRACT

Anorectal malformations are a wide spectrum of disorders, affecting both genders and rare adult presentation as with increased knowledge and advancements, the majority of cases are diagnosed and rectified at birth. This case is a classic example of delayed presentation caused by illiteracy and a lack of adequate health care in rural locations and highlights the uniqueness of this disease, presentation in adult age group and its management and effect on patient psychology and mental health. Because all low anorectal abnormalities can be treated in a single stage, neonatal assessment at birth and early referral to advanced health care centres are critical, with anterior or posterior sagittal anorectoplasty performed by the age of six months. Concerning the anomaly, parent education is an important aspect of patient care.

1. Introduction and importance

The distal anus and rectum, as well as the genito-urinary tract, are involved in anorectal malformations, which afflict both genders and have a rare adult appearance. Females are more likely to be affected than males, with an incidence of 1 in 5000 live births. With greater awareness and visible cosmetic distress, the majority of cases are recognised and rectified at birth [1]. Delay in diagnosis is linked to the development of chronic constipation or social and psychological morbidity as a result of issues with defecation control, flatus passage, and sexual activity and must usually leave paediatric surgical institutions around late adolescence (15–21 years) [2,3]. During the 1980s, the PSARP Posterior Sagittal Ano- Rectoplasty was introduced by Pena and soon became the preferred way to surgically correct these malformations [1,4]. Despite such advances, many female children do not undergo surgery at the appropriate time, perhaps due to parental neglect and also because most of them keep on passing stools through a perineal opening. Even after treatment, frequent hospital visits are needed during childhood because of constipation in 21%–67% and soiling in 10%–73% [5]. The few long-term postoperative follow-ups following PSARP during adolescence and adulthood conclude that half patients still experience symptoms such as incontinence and severe constipation after childhood [1,6,7]. When adolescents with ARM reach adulthood, they may need to seek adult medical help because ARM is a rather uncommon diagnosis. This case report has been reported in line with the SCARE Criteria [8].

2. Case presentation

The patient, a 27-year-old married woman, complained of faecal urgency and partial incontinence to watery stools. She stated that she had experienced uterine prolapse ten years ago, which was treated by Manchester repair. She then had a spontaneous abortion 5 years later, during which she was diagnosed with Low ARM with rectovestibular fistula. She had been married for six years and had given birth to her first child via vaginal delivery three years prior. She had a faint recollection of undergoing ARM surgery when she was about 5 years old, but no details were accessible and due to the absence of anal opening at normal site during presentation and failure of any documentation that surgical procedure seems to be unrelated to ARM. Incontinence had begun following the birth of the child. Her sexual function was also severely hampered, and she was separated from her husbandwith no other congenital abnormalities were found. Local examination revealed absence of anal opening at the normal site, and presence of a wide and

; PSARP, Posterior Sagittal Ano- Rectoplasty; ARM, Anorectal Malformation; MRI, Magnetic Resonance Imaging.

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prolapsing anal opening in the vestibulewith no sphincteric contractions were felt. The urethral opening was normal, the vagina was very roomy and short with low lying uterus and cervix (Fig. 1). Furthermore, a pelvic magnetic resonance imaging (MRI) scan was performed to examine the pelvic floor musculature and sphincter complex, which demonstrated perineal body dystrophy(Fig. 2). The sphincteric muscle complex was poorly developed and lied behind the vestibular bowel opening. She was counselled for surgery and insisted on surgery without colostomy for fear of becoming complete outcast from society. The patient was in the Kraske's(prone-jack-knife) position after urinary catheterization, with routine preparation/draping performed by our team. A mid sagittal incision was made, from the coccyx to the vestibule circumventing the vestibular anus. The incision was deepened in the sagittal plane identifying the parasagittal muscle fibres on each side and the vertical sphincter muscle complex corresponding to the anal dimple. The thin elevator was incised in midline above the sphincter complex and endopelvic fascia over the posterior rectum was identified. Putting multiple stay sutures, the anorectic was circumferentially mobilised and carefully departed from the vagina anteriorly. The rectum dissection, especially in its anterior portion, where the rectum and vagina share a single wall, was the most essential portion of the operation. The dissection continued up to the cervix, where the rectum and vagina are separated by full-thickness walls. The most common mistake made during this procedure is inadequate rectum mobilisation from the vagina, which results in a poor perineal body and suboptimal Neo anus under stress. The perineal body was restored and the vaginal Introitus was constricted. The anterior boundaxry of the muscle complex was used to limit the perineal closure. The Levator was approximated behind the rectum and the rectum was placed in the centre of the muscle complex. The posterior edges of vertical muscle complex were approximated, thus ensuring rectum is placed in the centre of muscle complex. Mucocutaneous anastomosis was performed to create the neo-anus giving appearance of a natural dimple with continence.(Operative

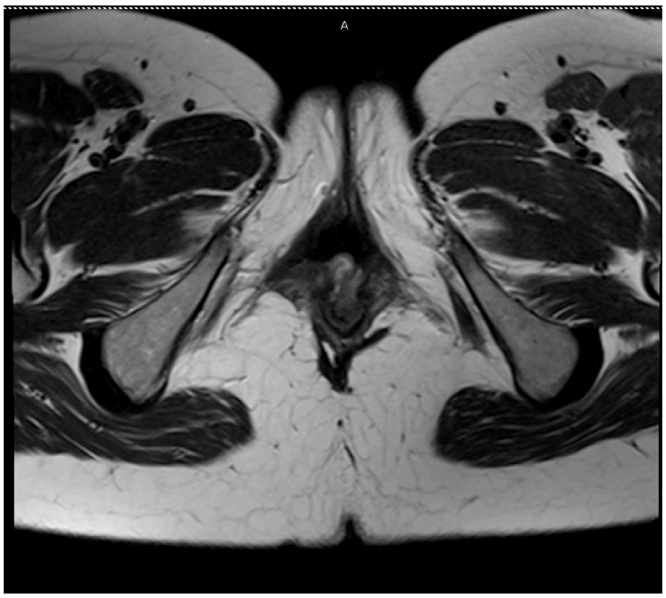

Fig. 2. Preoperative MRI Pelvis showing perineal body dystrophy with sphincteric muscle complex poorly developed and atrophic.

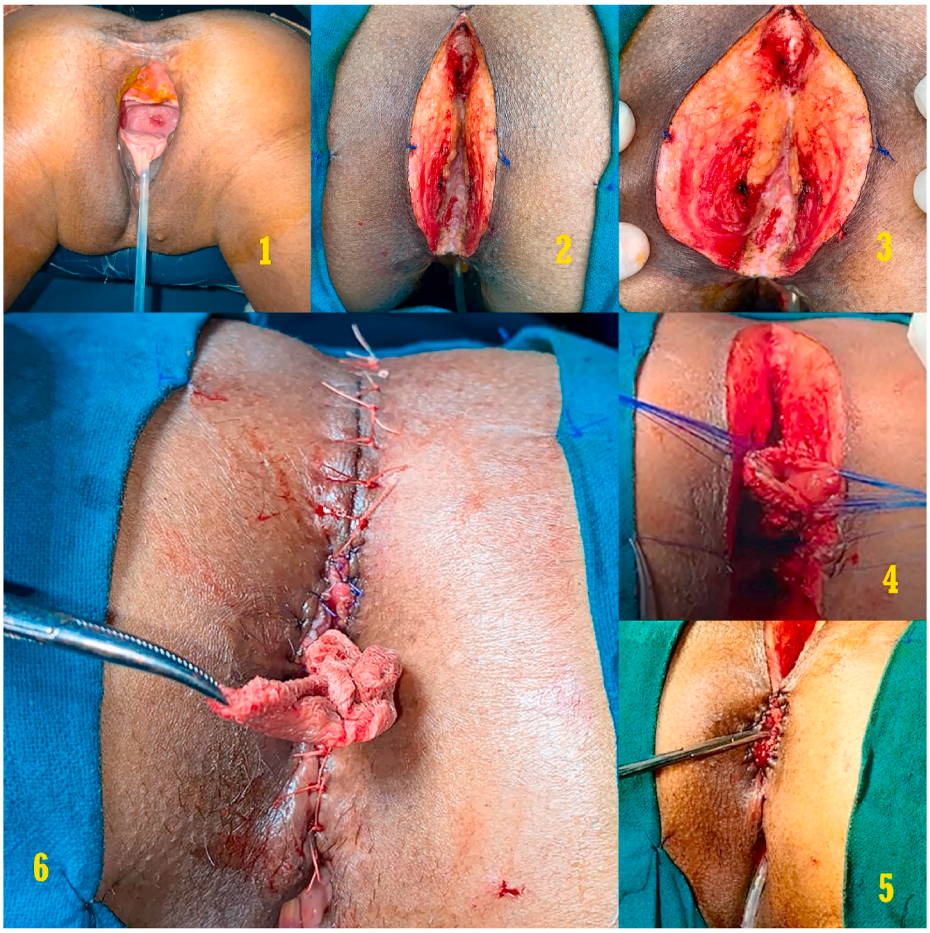

Fig. 1. Image showing Preoperative (1), Operative steps (2–5), Neoanus (6).

Steps and Neoanus: Fig. 1). No covering stoma was made. On the fifth post-operative day, oral feeding began. On the eighth day after surgery, the patient was discharged with a full oral diet regimen, a moderate laxative, and local wound care. Anal dilatation with Hegar dilators was initiated three weeks later followed by daily finger dilatation. At 6 months follow up, she is passing normal stools with good control and no leakage. She is pleased with her genital appearance and has resumed sexual activity.

3. Discussion

ARMs are managed differently depending on the severity of the defect. In addition, the majority of abnormalities are diagnosed and corrected during childhood. In females with rectal malformations do not usually present as a neonatal emergency as these babies continue to pass stools through the vestibular or perineal opening - which is the most common defect in females. Most parents seek surgical attention when the child develops constipation and anal stenosis. The surgery in such cases is fairly straightforward and involves a preliminary colostomy followed by posterior sagittal and rectoplasty about three months later. Colostomy is closed in the third operation once adequacy of the Neo anus has been confirmed [9]. At times the ARM in girls is characterised by a relatively wide anus that is situated much anterior to the site of muscle complex. Many of them have normal sexual life and successful marriages culminating in pregnancy also. They typically present with perineal tears subsequent to vaginal delivery - as was the case with our patient. In such cases when a pediatric surgeon operates during childhood, the parents are educated about the need for cesarean section delivery in such girls because the risk of perineal tears is very high if a vaginal delivery is attempted [10,11].Illiteracy, a lack of newborn care, and insufficient medical facilities can cause this condition to persist into adulthood or result in the neglect of a female child. This patient was not treated for any anorectal malformation at birth, due to lack of proper medical care and diagnosis and she continued to defecate through the vestibular fistula, which at birth was not probably very stenotic - thereby not leading to intractable constipation. For the treatment of high ARM, several abdomino-perineal and later sacro-abdomino-perineal methods were devised. Because the preceding methods did not allow for proper vision and identification of the levators and external sphincter complex, clinical outcomes suffered the most. The realisation of de Vries and Pena solved this difficulty, by dividing the external sphincter and levators in the midline to expose the bowel using a posterior sagittal approach [12]. A healthy sacrum, a normal gluteal cleft with normal gluteal muscle size, and undamaged nerves capable of executing an anal wink are all signs of a favourable outcome. When assessing a patient for PSARP, the anal wink must be in a different position than the existing anal location. The anus is usually found anterior to the sphincteric muscle complex. Patients with a flattened perineum, gluteal cleft with hypoplastic muscular development, sacral anomalies, and the inability to produce an anal wink, as well as those with a neurogenic bladder, are unlikely to achieve continence following a future repair [11]. Associated malformations occur in about 50% cases and involve vertebral, sacral, Genito urinary and cardiac malformations. Such patients should undergo echocardiography, USS KUB and sacral x rays routinely. Further testing by MCUG (Micturating cystourethrogram), and MRI of the spine and pelvis should be used in selected cases. A pelvic MRI was helpful in our instance in not only proving normal sacral anatomy but also understanding the architecture of the rectum in respect to the muscles, allowing us to better prognosticate her. Reconstruction should be undertaken utilising a multidisciplinary team approach as appropriate after the patient has been carefully evaluated, depending on the preoperative anomalies [11]. After reconstructive surgery, 60% have satisfactory continence, 24% have modest anal function issues such as occasional and/or modest smearing, and 13% have sexual function issues [13]. Table 1 summarizes the landmark Articles on this topic.

Table 1
Landmark Articles on Posterior Sagittal Anorectoplasty in Adults and Pediatric Age group regarding Anorectal malformations.

Author	Year	Paitent Population	Outcome	p value
R. Rintala et al.[7]	2001	•22 pubertal or postpubertal patients (median age 15; range, 13 to 25) with high or intermediate anorectal malformations repaired by PSARP	Constipation disappears & improved fecal continence outcome.	<0.05
J. Danielson et al.[14]	2015	•All patients born with ARM operated for Twenty AP pull-through procedures and 24 PSARP procedures	The outcomes of function and QoL in adulthood are superior after PSARP compared to those after AP-procedures.	0.07
K. Kyrklund et al.[15]	2017	•Results of institutional follow-up of 159 patients treated during the PSARP era	Outcomes following PSARP for are considerably better than those observed after classical pull-through operations in terms of both fecal control and sexual function	<0.05
A.G.Bedada et al.[16]	2018	•Two adult female patients born with rectovestibular fistulas.	Good functional outcome	0
M. M. M. Bayoumi et al.[17]	2020	•30 patients (19 female and 11 male) •Age at time of repair ranged from 3 to 78 months	Limited posterior sagittal anorectoplasty can have a role in treating constipation among cases of recto-perineal fistula	0.005

4. Conclusion

Constipation and the resulting large colon, rectum, are a major problem due to the late age of presentation. Inadequate weight gain and increased parental worry can result from a delayed presentation [18,19]. In terms of long-term anorectal function, anterior or posterior sagittal anorectoplasty has been the procedure of choice [20].The occurrence of ARM in the post-adolescent age group is quite uncommon. In the majority of cases, the illness goes unnoticed or is addressed insufficiently during childhood. It has received very little attention in the medical literature.

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Consent

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Author contribution

Ruchir Bhavsar: Research, Methodology, Review of Literature, Formal analysis, Writing - original draft. Samrat Ray: Conceptualization, Supervision, Writing - review & editing. Muni Verma: Writing - review & editing. Satish K Agarwal: Chief supervisor. Final correction and approval of the manuscript. Samiran Nundy: Chief supervisor. Final correction and approval of the manuscript.

Registration of Research Studies

1. Name of the registry: NOT APPLICABLE.
2. Unique Identifying number or registration ID: NOT APPLICABLE.
3. Hyperlink to your specific registration (must be publicly accessible and will be checked): NOT APPLICABLE.

Guarantor

Ruchir Bhavsar , Samrat Ray, Muni Verma , Satish K Agarwal , Samiran Nundy

Declaration of competing interest

No benefits in any form have been received or will be received from a commercial party related directly or indirectly to the subject of this article.

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