Transfistula anorectoplasty on adult female anorectal malformation: A rare case report

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ABSTRACT
INTRODUCTION: Female anorectal malformation (ARM) is correctable congenital defects. Delayed presentation of patients with anorectal malformation is uncommon. However, presentation beyond teenage years is not commonplace. We describe a case of ARM with rectovestibular fistula and anal atresia.

PRESENTATION OF CASE: A 31 years old woman with chief complaint of small vaginal introitus. Gynecology examination showed urethra, labia majora, labia minora, and small vaginal introitus. Vaginal length was 6 cm. There was no anal canal. The patient underwent transfistula anorectoplasty and modified Fenton procedure.

DISCUSSION: The important step of transfistula anorectoplasty (TFARP) procedure is the placement of neoanus in the center of external sphincter to provide continence. The absence of skin incision in anterior and posterior to the neoanus provide good cosmetic result. Compared to PSARP procedure, TFARP procedure has advantage which is the absence of separation of the skin in the midline buttock, the levator muscle, and the external sphincter complex. This lead to optimal fecal continence. In our experience, TFARP is a safe and effective procedure. Daily vaginal dilation in postoperative period is unnecessary. This technique also give good cosmetic result with optimal fecal continence and sexual function.

CONCLUSION: Transfistula anorectoplasty procedure for ARM give good cosmetic result and optimal fecal continence and sexual function.

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1. Introduction

Female anorectal malformations (ARM) are one of the most prevalent congenital defect with incidence of 1 in 5000 live births [1–3]. Holschneider A reported that number of anorectal malformation in females reached 151,000,000 live births [4]. There are several anomalies that is grouped into one term of ARM. According to Wing spread, spectrum of anomalies found in ARM consists of low, intermediate, high, and miscellaneous anomalies [5]. Anorectal malformation type that is most common diagnosed are vestibular fistula. According to Pandley A, ARM can be corrected with good prognosis. Clinical examination in newborns can help doctors to find these anomalies. However, women in developing countries were often diagnosed late due to low education and awareness about ARM and also due to poverty and lack of care intended for girl children [6]. Adult presentation of this malformation is rare.

Diagnosis of ARM require several modalities started from clinical examination, rectal examination, and electrical stimulation of anal spincther, vaginoscopy, and cystoscopy. Diagnosis require several specialties such as gynecology, colorectal surgery, and urology. Evaluation of anal spincther with electrical stimulation of the anal spincther include investigation of anal mislocation relative to spinchter complex, anal stricture, rectal prolapse, and perineal body. Cystoscopy is conducted to assess defect in urethra and bladder or ectopic ureters. Vaginoscopy is used to evaluate remnants of rectovaginal or rectovestibular fistulae, residual vaginal longitudinal septum, evaluation of vaginal introitus, assessment for stricture and requirement for introitoplasty/dilatation and evaluation of cervix or cervices [7].

Anorectal malformations can be treated by reconstructive operation to repair the location of rectum to the center of spinchter complex. Main surgical procedure commonly performed for ARM were posterior sagittal anorectoplasty (PSARP) introduced in the 1980s [8]. ARM subtype determine the clinical outcome of the procedure. The concerns about the PSARP procedure are the occurrence of fecal incontinence and lack of voluntary bowel control [7]. In addition, posterior sagittal anorectoplasty associated with several complications such as small perineal body, persistent urogenital sinus, urethrovaginal fistula, acquired vaginal atresia or stricture, and rectovaginal fistula [7]. There are several technique to correct ARM other than PSARP including transfistula anorectoplasty.
Fig. 1. Anovestibular fistula with openings of urethra and vagina. There was no anal opening.

(TFARP). In this case report, we present the clinical outcome of two cases of ARM corrected by performing TFARP procedure. Because there was no published information of TFARP procedure in adulthood, this case became interesting.

This case report has been reported in line with the SCARE criteria [9].

2. Patient information

We present a case of anorectal malformation. A 31 years old woman with chief complain of small vaginal introitus. The patient complained difficulty in sexual intercourse. Patient admit that husband did not able to penetrate properly. The patient has been married for 4 years with sexual intercourse through one common channel. Menstruation and defecation was came out from the same channel. She had no complaint of micturation and defecation. Patient did not have any family history with similar problem. There was no history of medication consumption of her mother during pregnancy.

Physical examination (Fig. 1), revealed that there were urethra, labia majora, labia minora with small vaginal introitus. The size of introitus vagina was 1 cm. Portio was palpable with vaginal length was 6 cm.

Fig. 2. Ultrasonography.

Fig. 3. Transfistula anorectoplasty technique (A). Circumference incision was performed in vestibule. Several fine silk traction sutures insertion around the fistula orifice. (B) Separation of the rectum from the posterior vaginal wall was conducted by sharp dissection. Meticulous dissection of anorectum about 4 to 5 cm length was performed to prevent vaginal damage or musculature surrounding the rectum. Spincter muscle was identified and by using the index finger as a marker, v-shaped incision is made through the anal canal. (C) Anoplasty was performed with apposition of vestibular wound using PGA 3.0. (D) Rectal tube number 11 was inserted, vagina and perineal body was reconstructed with modified Fenton procedure; vestibular wound was closed with PGA 2.0 interrupted stitches, bulbospongious muscle was sutured using PGA 2.0 and posterior vaginal mucose was sutured using PGA 2.0.
6. Discussion

ARMs are correctable congenital malformations. They can achieve reasonably good prognosis. Most of these anomalies can be easily detected on a clinical examination in newborns. This report highlights some important issues. Delayed presentation is common in developing countries; however, delaying the treatment up to the age of 20 years is uncommon. Delayed diagnosis may be attributed to lack of knowledge, awareness, poverty and poor. Anorectal malformation is repairable congenital defect [6]. The etiology of these abnormalities is not clearly understood. There are chromosome 7q39 which contains three essential loci associated with ARM : SHH, EN2, and HLXB9. Mutations of HLXB9 gene associated with sacral agenesis [10].

Wingspread classified ARM based on what level of rectal decent halt and sex. The female classification of ARM according to Wing spread was divided into four categories which were low, intermediate, high, and miscellaneous anomalies. Low anomalies comprise of anovestibular (perineal) fistula, anocutaneous, and anal stenosis. Intermediate anomalies include rectovestibular fistula, rectovaginal fistula, and anal agenesis without fistula. High anomalies encompass anorectal agenesis, rectovaginal fistula or without fistula and rectal atresia. Miscellaneous anomalies include anal stenosis, persistent cloacal anomaly, and rare malformations [5].

Traditionally, ARMs were reconstructed with a protective colostomy because of the fear of wound healing and subsequent loss of anal sphincter complex with the risk of impairment of future continence. Pena introduced the posterior sagittal approach (PSARP) followed by colostomy which is a gold standard for the procedure [11]. The belief that a protective colostomy may prevent wound infection is questionable. Colostomy related complications range from 28% to 74% [1]. PSARP involves many perirectal dissection. Hence, posterior sagittal anorectoplasty associated with change in external appearance of the anus, anal continence, and manometric parameter [12–14]. Gynecologic anatomic abnormalities may present following the procedure of posterior sagittal anorectoplasty (PSARP) indicated for anorectal malformation (ARM). Common abnormalities found were small perineal body, introital stenosis, the presence of vaginal septum, or the presence of rectovestibular fistula [7]. Other technique is anterior sagittal anorectoplasty (ASARP) where perineal skin, perineal body was cut to the midline perineal incision, which higher postoperative morbidity and poor aesthetic appearance of the perineum for the surgical treatment of vestibular fistula than transfistula anorectoplasty (TFARP) [15].

According to Mitul AR, transfistula anorectoplasty (TFARP) resulted in good functional and cosmetic outcome with good anal continence. There is no perineal or anterior/posterior sagittal incisions in TFARP hence there is no risk of disturbance in perineal body and neurovascular supply. The internal sphincter is preserved since there is no taping of rectal pouch. The TFARP surgery is performed with the patients lying in lithotomy position. The important step of this procedure is to place the neoanus through the center of external sphincter to prevent incontinence [1]. Good cosmetic in TFARP procedure is caused by the absence of incision of the skin anterior/posterior to the neoanus. In addition, the risk of wound dehiscence at that area is also reduced by using TFARP [15]. The TFARP procedure has advantage compared to PSARP due to the absence of division of skin in the midline buttock, the levator muscle, and the external sphincter complex. The lack of such division results in optimal fecal continence [2]. Several authors report excellent continence ranging from 90.5 to 100% were continent after TFARP. In other series, continent scores were excellent or good among 58.3–72% [1]. All of the data above was taken from literature which the procedure performed in neonates. There was
7. Conclusion

Transfistula anorectoplasty procedure for adult female ARM result in good functional and cosmetic outcome with good anal continence in adults.

Declaration of competing interest

This case report do not have any relationship with other people or organisations.

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Ethical approval

Because this is the case report and it is not a research, ethical approval was not required.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

Tyas Priyatini: concept, operator, data analysis, drafting and revising, final approval.
Roziana: data collection, data analysis, writing the paper.

Registration of research studies

N/A.