CASE REPORT

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Vaginal Reconstruction with Native Rectovestibular Fistula in an Infant with Vaginal Agenesis and Imperforate Anus

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ABSTRACT Vaginal agenesis is a congenital anomaly which occurs as isolated developmental defect or as a part of complex anomalies. Vaginal anomalies associated with anorectal malformations are usually diagnosed and repaired in time of surgical correction of anorectal malformations during the infancy period. Herein a case with vaginal agenesis and recto-vestibular fistula, who underwent a neo-vaginal reconstruction with distal segment of fistula during the posterior sagittal anorectoplasty was presented. The use of recto-vestibular fistula as a neo-vagina in cases with vaginal agenesis combined imperforate anus seems to be a feasible and effective approach that has satisfactory anatomical and functional outcomes for vaginal reconstructions.

Keywords: Absence of vagina; anorectal malformation; imperforate anus

Vaginal agenesis, absence of vagina is a congenital anomaly which occurs as isolated developmental defect or as a part of complex anomalies. It is estimated to occur in 1 of 4.000-5.000 live female births.[1] Majority of these patients form part of Mayer Rokitansky Küster Hauser Syndrome, with absence or remnants of the uterus. Vaginal anomalies occur as a result of congenital malformations of Mullerian ducts, and/or can be associated with anorectal or cloacal malformations.[2-5] Imperforate anus with rectovestibular fistula is the most common type of anorectal malformation in girls, but in combination with vaginal agenesis it appears to be rare.[2]

To make a diagnosis of vaginal anomalies is relatively rare in neonatal period. Absence of vagina can easily be overlooked in newborns with anorectal malformation especially in imperforate anus with rectovestibular fistula if perineal examination wasn't done carefully. Vaginal anomalies associated with anorectal malformations are usually diagnosed during the repairing of anorectoplasty in infancy period.[4-6]

As well as delays in timing of the treatment, appropriate method for vaginal reconstruction in girls with vaginal agenesis still remains controversial still. The purpose of this study was to evaluate the outcomes of using native rectovestibular fistula for vaginal reconstruction in a girl with imperforate anus with vaginal agenesis during the ano-recto-vagino-plasty, and to show the feasibility of one-stage operation in infancy period.

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A one year old girl was admitted to pediatric surgery department due to imperforate anus with recto-vestibular fistula; the fistula had been dilated for comfortable defecation per day. Physical examination revealed the presence of an imperforate anus with a rectovestibular fistula without vaginal orifice (Figure 1). In the perineal inspection vaginal orifice was absent, and there was a midline pigmented area as anal dimple. Chromosomal analysis result was 46 XX. X-ray revealed mild scoliosis and agenesis of 4^{th}-5^{th} sacrum. Unilateral renal agenesis was determined by urinary ultrasound; and bifid uterus, both ovaries and tubes were determined by ultrasound and magnetic resonance imaging.

The parents were informed about the surgical methods; and informed consent was obtained for this surgery. During the operation, muscular contraction of this pigmented area was seen by the nerve/muscle stimulator. Cystoscopy revealed any abnormality of the urethra, bladder neck, or bladder and both ureteral orifices. Bifid uterus, both ovaries and tubes were seen by laparoscopy, and the diverting sigmoid colostomy was created in between descending and sigmoid colon.

Three months later, definitive surgical correction was done. The first step was performed in prone position by posterior sagittal incision between the coccyx and posterior line of the fistula; and fistula and distal rectum left in situ and a posterior plane of cleavage established to the level of peritoneal reflections. Penrose drain was placed through the pelvis as a guide, and posterosagittal incision was covered with sterile drape. As the second step, laparotomy was performed in decubitus dorsalis position by Phannenstiel incision; and bifid uterus, both ovaries and tubes were seen. Rectosigmoid colon was transected at the level of peritoneal reflection preserving mesenteric vessels of distal part of the rectum, thus native recto-vestibular fistula was left as a neo-vagina. Anastomosis was completed between the upper edge of distal part and bottom edge of blind double vaginal remnants. The peritoneum of pelvic floor was reconstituted, and abdominal wound was closed without drain. As the third step, perineal body was constructed, and neo-anus was created within the muscle complex to the anal dimple in prone position by rectosigmoid colon pull-through procedure. Native rectovestibular fistula had been left as a neo-vagina, and final length of the neo-vagina was approximately 8 cm. The operation was completed in approximately four hours without complication. Operative diagram was demonstrated with a drawing in Figure 2.

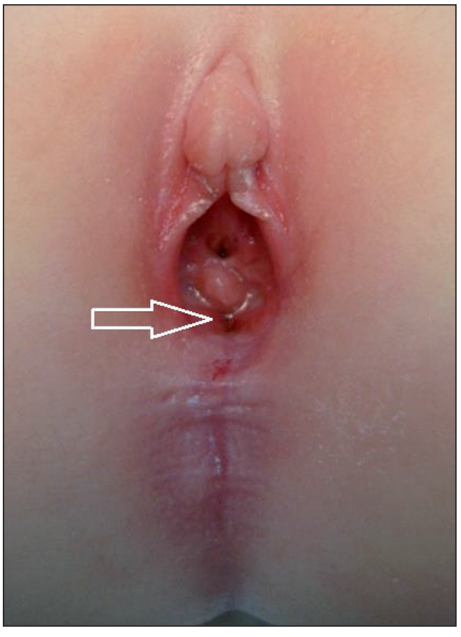

FIGURE 1: No vaginal introitus was seen in the detailed physical examination when she was one year old (restibular fistula was signed with white arrow).

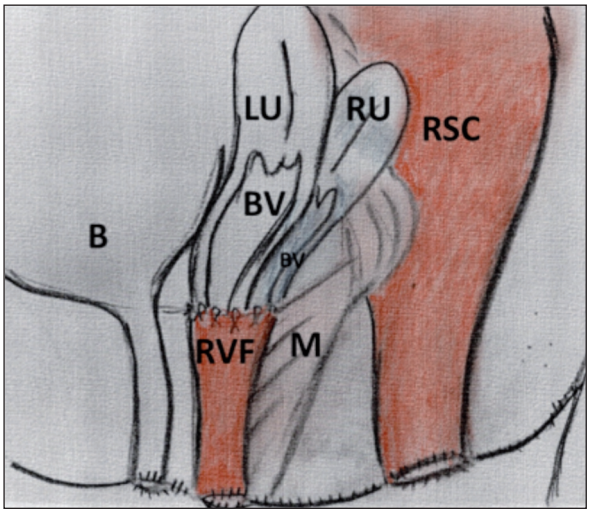

FIGURE 2: A drawing of the operation technique in vertical plan. Native Rectovestibular fistula vith 8 cm's proximal rectum was cerated as neovagina during the posterosagittal ano-recto-vagina plasty. (B: Bladder, LU: Uterus, RU: Right Uterus, BV: Blind Vagina, RSC: Rectosigmoid Colon, M: Mesenterium, RFV: Rectovestibular Fistula as native).

In the early postoperative period, perineum had a very natural appearance with orifices of neo-vagina and neo-anus. Dilatation of the neo-vagina was not required because opening of neo-vagina was natural without any sutures. Daily anal dilatation program for neo-anus was launched in the postoperative 15th day, and it was gradually increased until size number 15 Hegar dilator; and colostomy was closed 3 months later. Potty training was given at 3 years old. Urinary continence in day and night was very good; but voluntary bowel management was not good in early periods. There was no soiling with daily bowel management programming described by Alberto Peña.[5] Nineth years' postoperative follow-ups were uneventful.

DISCUSSION

In the childhood, especially in small children diagnosis of isolated vaginal anomalies is very difficult, they are generally diagnosed in the adolescent period.[1,2,5,7]

Vaginal anomalies associated with anorectal malformations are a broad spectrum group of diseases. These anomalies are usually diagnosed in infancy, especially during the surgical correction of the anorectal malformations.[2-5] Vaginal replacement or reconstruction are required during the definitive surgery of the anorectal malformation in cases with vaginal abnormalities.[2-5] Early repair of anorectal malformation and postponed vaginal reconstruction seem to be a viable option for cases with anorectal malformation concomitant vaginal agenesis; but timing of the vaginal reconstruction is still controversial, especially in cases with isolated vaginal agenesis without any anorectal malformation.[1-9] In the adolescent period, local conditions and anatomic orientation are excellent; so it is easy to adapt the anatomy to physical development in adolescence; therefore, in cases with isolated vaginal agenesis, vaginal reconstructions should be planned after adolescense shortly prior to sexual course or marriage.[1,2,4-6]

Many methods for vaginal reconstruction had been described. Historically, reconstruction has involved the use of skin grafts and non-operative methods with less than ideal results.[1,5] These cases without anorectal malformations have been corrected surgically with Mc Indoe's vaginoplasty method using split thickness skin graft obtained from thigh or buttocks.[1,2] Autologous grafts like buccal mucosa have also been used with success.[1] The use of a split-thickness skin graft to line the vagina, the greatest postoperative problem was the tendency to stricture and collapse of the newly-formed cavity.[2]

Bowel makes a very satisfactory vaginal replacement, with little tendency to collapse, and unlike split-thickness skin, it has a strong supporting muscular wall. The earliest attempts at vaginal reconstruction utilized bowel as the replacement lining in 1892 as Snegurieff's procedure, and vaginal replacement with transplanting ileum was described in 1942.[2] Snegurieff's procedure was subsequently modified by Popow and Shubert.[2] Generally sigmoid colon is selected for vaginal reconstruction because of its large lumen, thick wall resistant to trauma, adequate secretion allows lubrication and acceptable complication rates.[2,5,10,11] The ileum loop is a good alternative when the sigmoid colon is not available.[2,9] Postoperative complications after vaginal reconstruction by intestinal segments are vaginal orifices stenosis, mucosal prolapse, perineal wound hematoma, superficial wound infection, and intestinal obstruction.[3,5] Vaginal reconstruction with segments of sigmoid colon was found safer and more effective method according to the ileum.[3,5,9,11,12]

There are two surgical alternatives to repair vaginal anomalies in cases with anorectal malformations. In the first option, the recto-vestibular fistula and the rectum can be separated from the urethra and reconstructed as a neo-anus within the confines of the sphincter mechanism. In this case, the new vagina is either created with the new anus at the time of initial repair, or it is created by other vaginal replacement techniques later in life. The second option is to leave the distal part of the rectum in place with the recto-vestibular fistula in place without detaching it from the urethra. Thus, this recto-vestibular fistula and distal rectum are used as neo-vagina, and the upper rectosigmoid colon is pulled down to be placed as a neo-anus within the boundaries of the sphincter mechanism.[3] It was first described by Cohn that both rectal and vaginal anomalies can be corrected together during abdomino-perineal dissection of the

rectum; and the last part of the intestine with fistula has been shown to be used as neo-vagina and the colon can be used as neo-anus.[2] This procedure was first utilized in a 7 years old girl with imperforate anus with vaginal agenesis in 1954. Until this date a neo-vagina was fashioned using the anteriorly-placed recto-vestibular fistula in a few girls with imperporate anus with vaginal agenesis, along with the usual abdominoperineal pull-through procedure to correct the imperforate anus.[2-10] Neovagina, which is formed by vaginal reconstruction from rectovestibular fistula meets all the necessary criteria for a vagina. It provides a vagina with an adequate diameter opening in the perineum and a sufficient length; self-moisturizes, easily adapts to the uterus, cervix and rudimentary atretic vaginal segments, and most importantly does not require routine dilatation after the operation. In appropriate patients, the fistula can be used to create the neo-vagina obviating the need for a bowel anastomosis.[2,3] On the other hand, performing the necessary bowel anastomoses and laparotomy in the other vaginal reconstruction techniques may cause serious complications and morbidity.[2,3,10]

The main problem is that which of these two techniques has better functional results, especially in the long term, in cases with combined vaginal agenesis of the imperforate anus. The advantage of the first technique, namely the use of the fistula with the distal rectum as a neorectum, may be considered to be an advantage when using a natural tissue in its natural site; because the rectovestibular fistula, having a pectinate line, may mean that it has the necessary functional properties for intestinal control, such as sensory and reservoir function. Therefore, some authors have suggested that this recent technique should not be used because of the high risk for anal incontinence. The main disadvantage of this method is that if the rectovestibular fistula is used as a neorectum, a new vagina is needed using one of the techniques described for vaginal reconstruction.[5] Sacral bone structure of cases is also important in the decision of surgical method for vaginal reconstruction as well as the type of vaginal abnormalities and anorectal malformations. In cases with anorectal malformation and sacral bone agenesis, as we experienced in our patient, after the definitive operation, there is a high possibility of urinary and anal continence problems independently from the operating technique. Sacral bone agenesis is an indicator of poor prognosis for fecal continance in patients with anal atresia; these patients are less likely to be continent. Therefore, in these cases with sacral bone agenesis, using the distal rectum and rectovaginal fistula as a neo-vagina is seen as the best option cosmetic appearance and comfort of the patient.[5] Of course, in our case, eight years of follow up is no long enough to evaluate the long term risks of urinary and anal incontinence. Although 4^{th} and 5^{th} sacrum agenesis were available, she had quite in good condition according to early functional results including anal and urinary continence, results are promising. In cases where faecal continence cannot be achieved with a good toilet training, these cases are kept clean with "intestinal management programing" described by Alberto Peña.[5]

The technique of using recto-vestibular fistula for creating a neo-vagina in patients with anorectal malformations is less complex than other vaginal replacement techniques in the cases with anorectal malformations and is easy to learn and perform. In addition, no special surgical equipment is needed differently from the posterior sagittal anorectoplasty; cosmetic and functional results are very satisfying. A significant advantage of this technique is no need to vaginal dilatation. Hospitalization duration is not different from posterior sagittal anorectoplasty.[5] The creation of neo-vagina according to this technique from recto-vestibular fistula might provide a satisfactory alternative for the surgical management of vaginal atresia in patients with recto-vestibular fistula and anal atresia concomitant with vaginal atresia.[2,3,5-9] It can be performed with posterior sagittal approach with or without laparotomy during the childhood period. Neo-vagina should be anastomosed to probable proximal vaginal remnant to restore the continuity internal genitalia. This step could be made during laparotomy after posterior sagittal anorectoplasty on the same session or during colostomy closure or adolescent period.[5]

As in other vaginal replacement operations, problems such as hematocolpos or uterine atrophy in uterine didelphys during adolescence and post-adolescent period can be seen. Therefore, they should be

kept in close follow up until puberty. Laparotomy may be necessary especially in adolescent period in these cases, not only for therapeutic, but diagnostic reasons; even so, complex anomalies, such as vaginal duplication with unilateral atresia and a septate uterus, could not be suspected.[12]

In conclusion, the technique of using recto-vestibular fistula for creating a neo-vagina in patient with anorectal malformations is a simple, safe, and effective surgical option compared with other therapeutic options. Recto-vestibular fistula is a useful alternative for vaginal replacement especially in cases with sacral bone agenesis; in spite of the possible morbidity, it is valuable because of the inherent advantages of preserving native rectal fistula tissue for using as a vagina instead of neo-rectum.

Source of Finance

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Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

Idea/Concept: *Ayşenur Celayir;* ***Design:*** *Ayşenur Celayir, Osman Zeki Pektaş;* ***Control/Supervision:****Ayşenur Celayir;* ***Data Collection and/or Processing:*** *Serdar Moralıoğlu, Oktav Bosnalı;* ***Analysis and/or Interpretation:*** *Ayşenur Celayir, Serdar Moralıoğlu;* ***Literature Review:*** *Serdar Moralıoğlu, Ayşenur Celayir;* ***Writing the Article:*** *Ayşenur Celayir, Serdar Moralıoğlu;* ***Critical Review:*** *Ayşenur Celayir;* ***References and Fundings:*** *Ayşenur Celayir, Serdar Moralıoğlu, Oktav Bosnalı, Osman Zeki Pektaş;* ***Materials:*** *Ayşenur Celayir, Osman Zeki Pektaş.*

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