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CREATING PATHWAY FOR A DEAD END-CREATSAS MODIFICATION OF WILLIAM'S VAGINOPLASTY IN MRKH SYNDROME - A CASE REPORT

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ABSTRACT Vaginal agenesis is a congenital anomaly of the female reproductive tract. In this case a unmarried female presented to our OPD with primary amenorrhoea and difficulty in coitus. She was diagnosed to be a case of MRKH. She underwent Creatsas modification of Williams vaginoplasty . The post-operative events were uneventful and she was discharged with the advice of regular vaginal dilation with gradual hegar dilator . The procedure turned out to be a good success with patient satisfaction.

KEYWORDS : MRKH, Creatsas modification of Williams vaginoplasty, vaginal aplasia, primary amenorrhoea.

INTRODUCTION:

Mayer–Rokitansky–Küster–Hauser syndrome has a prevalence of one in 4000 females and is the most common cause of vaginal aplasia. Management of Mayer–Rokitansky–Küster–Hauser syndrome patients includes, along with psychological support, the creation of a neovagina. Creatsas vaginoplasty is a fast and simple technique in which a perineal skin flap is used to create a perineal pouch. Mayer–Rokitansky–Küster–Hauser (MRKH) syndrome (also referred to as 'Müllerian aplasia') is a rare congenital anomaly that represents the most common cause of vaginal aplasia. It is characterized by the absence or hypoplasia of the Müllerian duct derivatives (the uterus and the upper two-thirds of the vagina). The incidence of the syndrome ranges from one per 4000 to one per 10,000 female newborns [1]. Mayer–Rokitansky–Küster–Hauser syndrome presents two major consequences in a patient's life. First, owing to the absence of the vagina, normal sexual intercourse is not possible. Pregnancy is also impossible, owing to the absence of the uterus, despite the fact that the ovaries are present and have normal function. Taking into account the consequences that the diagnosis of MRKH syndrome has on a woman's self-image and sexuality, it is easy to understand that creation of a neovagina is necessary in order to provide the young girl the prospect of a normal sexual life. To date, numerous surgical and nonsurgical techniques have been proposed for the correction of vaginal aplasia. Nowadays, the most popular techniques are the McIndoe operation [2,3] (also known as the McIndoe–Bannister or Abbé–McIndoe operation), the Vecchietti procedure [4], the Williams vaginoplasty [5] and its modification by Creatsas [6–8]. The Davydov technique [9,10] and bowel colpoplasty [11–13] are also used for vaginal reconstruction, but they are not commonly used.

Case Report

A 22 years old young unmarried female presented to us with chief complain of primary amenorrhoea. There is no significant medical, surgical or drug history related to the disease.

General physical examination:
Height 161cm Weight 55kg BMI 21.2 kg/msq

pubic and axillary hairs growth is normal adult pattern , breast development is upto tanner stage 5.

on local perineal examination:
external genitalia appears normal , a shallow dimple was present at introitus area with blind vaginal pouch .

Ultrasound whole abdomen and pelvis was done which revealed absent uterus and left kidney, both ovaries were present. Her LH, FSH and TSH were normal. After complete evaluation a diagnosis of MRKH syndrome is considered. patient was counselled about the diseases and further treatment options were discussed. As patient is planning to marriage she was concern about her sexual function, so creation of neovagina was considered. till now, numerus surgical and nonsurgical procedures have been proposed to correct the vaginal aplasia.

For this we considered the William's vaginoplasty. It involves the use of a skin flap from the patient's perineum in order to create a perineal pouch. Procedure started under subarachnoid block and foley's catheter inserted into the urethra and four allis clamps are used to put the vulva under stretch(figure 1). Subsequently a symmetrical U shaped incision is made in the vulva starting 4 cm lateral to the external urethral sphincter at the medial side of labia majora(figure 2), extending down to the perineum and up to the other side of vulva, it is followed by mobilization of perineal subcutaneous tissue. A layer of interrupted 2-0 absorbable sutures is put between the inner margins with the knot lying on the inside of the neovagina being created in order to create the pouch. A neovagina of 6x6 cm was created(figure 3,5,6).The perineal muscles and subcutaneous tissue are then approximated using the same suture material. Finally absorbable suture are used in order to stich the external skin(figure 4). a 2 meter roller gauge packing was inserted in the neovagina for better hemostasis and better creation of neovagina which was removed after 24 hours. Patient was educated for serial dilatation by hegar dilatator with the help of lignocaine jelly from post operative day 2. Serial dilatation by hegar dilatator started from number 10 to number 15 till discharge on day 6. At the time of discharge 6x6 cm size of neovagina was maintained. At the time of discharge patient was advised to continue dilaltation for next three months. Patient was called for follow up after 2 weeks and 6 cm size of neovagina was confirmed(figure 7) and compliance regarding dilatation was checked. patient was counselled about their method of placement, removal and washing of dilator to facilitate the compliance and aseptic environment.

Careful hemostssis and asepsis was maintained during the whole procedure. Typical wound hygiene and administration of wide spectrum antibiotics postoperatively reduce the risk of wound infection.

Williams vaginoplasty is safe technique with a mean surgical time of 30 minutes and mean hospitalization of 6 days.

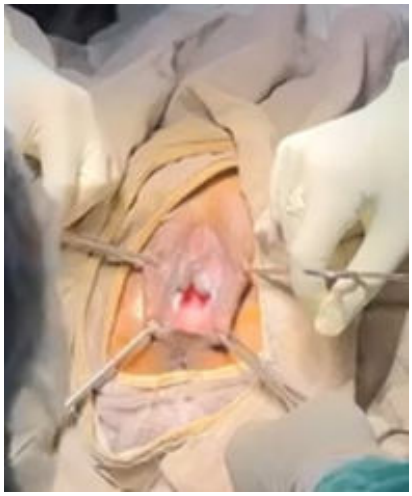

Figure 1- Placement of allis clamps

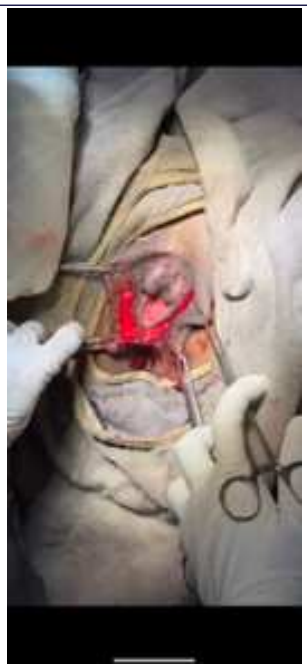

Figure 2- U shaped incision in the vulva

Figure 3- first layer of 2-0 absorbable sutures between the inner skin margins

Figure 4- Stiching the outer skin margins

Figure 5- immediate postoperative length of neovagina(6cm)

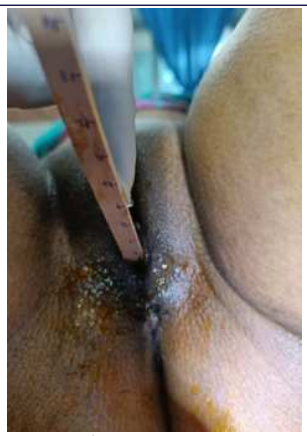

Figure 6- postoperative 15th day-neovagina length(6cm)

DISCUSSION-

According to the American College of Obstetricians and Gynecologists committee opinion on adolescent health care, the Abbe-McIndoe vaginoplasty is the most common surgical alternative among all available operative approaches (14).

Major complications of the method are the risk of injuries of the neighboring organs, such as the bladder or the rectum, during the dissection as well as dehiscences, fistulae, and retention cysts, mainly caused by local infections with bacteria from the rectal flora and/or prolonged continuous pressure exerted by the vaginal mould on the urethra and rectum (15). Graft shrinkage, due to the development of granulomatous tissue (15), may cause neovaginal stenosis and dyspareunia. Lack of lubrication is also a problem when the neovagina is constructed with skin (16).The Vecchetti operation and its laparoscopic version have been frequently performed in Europe over the last years (17).

Despite the fact that this procedure shows low perioperative morbidity and a quicker recovery period, laparoscopy may not be feasible or cannot be performed safely (18).The sigmoidal colpoplasty is considered as a major, complicated intraperitoneal operation that carries major intraoperative risks and complications (19, 20).

One of the major limitations of the procedure is the presence of a pelvic kidney that may hamper the procedure. The need for laparotomy and a high risk of bowel complications are considered to be the main disadvantages of this operation (21).The Creatsas modification of the Williams vaginoplasty is a simple, safe, and quick operative method resulting in a functioning vagina, similar to normal. The surgical time is usually of no more than 30 minutes, no specific instrumentation is needed, and postoperative complications are rare. The Creatsas modification prevents hemorrhage due to rupture of hymenal vessels during the first sexual intercourse. The use of absorbable suture material such as Dexon or Vicryl secures the stitching of both skin layers and protects the wound from reopening, especially in patients who do not follow the medical instructions. Patients with MRKH syndrome and existence of a pelvic, usually a solitary kidney, which is almost always associated with altered pelvic anatomy and higher perioperative risks (22), benefit even more, as no dissection of the vesicorectal space is required during this safe, extraperitoneal approach. A mean hospital stay of 6 days is required to prevent postoperative complications such as dehiscence during bathing at home to maximize patient compliance and allow immediate diagnosis and intervention in case of complications. In the vast majority of patients there is no need for postoperative vaginal dilators, which reduces the psychological impact of the treatment.

Macroscopically, differences between this neovagina and a normal vagina may be distinguished only after careful gynecologic examination by an experienced physician.

As previously reported, the functional axis of the neovagina is very similar to the anatomical axis of a normal vagina (23). In contrast to other grafting methods, the elasticity of the tissues formatting the lower part of the introitus of the neovagina permits pleasant and uncomplicated sexual intercourse. The latter may be attempted shortly after the operation to alleviate the patient's stress.

CONCLUSION

In conclusion, the aim of all methods for the management of MRKH is the creation of a vaginal channel of adequate functional depth and width, with axial deviation similar to natural vagina. In this case ,we have found that the Creatsas modification of Williams vaginoplasty is a simple, quick, and effective method that satisfies all the requirements.

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