

Neovaginal Prosthesis - Hallmark in the Management of Patients with MRKH Syndrome – A Case Report

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Abstract

Vagina is a part of the female genital tract leading from external genitals to the cervix of the uterus. It is a muscular and an elastic organ. There are various malformations that can occur in the female genital organ, one among them being Mullerian Agenesis. Mullerian agenesis/ MRKH syndrome is a congenital anomaly in females due to the developmental failure of vagina from the mullerian duct. This results in aplasia of the uterus/ vagina or presence of rudimentary uterus and upper part of the vagina.¹ It is the most common cause of primary amenorrhea. Women with vaginal agenesis usually suffer from mental depression and they are deprived of a normal social life. It is mandatory to educate and motivate the patients and their family about the various treatment options available and carry out the treatment which is best suited according to the patient's needs. The most common treatment modalities include non-surgical methods, surgical methods or combination of both. Surgical creation of neovagina along with the use of a neovaginal prosthesis can go a long way in improving the quality of life of the patient. This article presents a case report of a young woman with MRKH syndrome who was referred to the Department of Prosthodontics for fabrication of a neovaginal prosthesis which was to be used after surgery to maintain a patent passage. The neovaginal prosthesis was fabricated by a Maxillofacial Prosthodontist using autopolymerizing acrylic resin.

Keywords: Neovaginal prosthesis, vaginal dilator, vaginal stent, mullerian agenesis, MRKH Syndrome, vaginal agenesis.

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INTRODUCTION

Vagina is a part of the female genital tract which is muscular and an elastic organ leading from external genitals to the cervix of the uterus. It is located between the urethra and the anus. The wall of vagina is made of muscles and many elastic fibers covered by mucous membrane which makes it soft and flexible. The presence of several nerve endings attribute to its nature of high sensitivity.

There are various malformations that can occur in the female genital organ, one among them being Mullerian Agenesis. Mullerian Agenesis is also referred to as Vaginal Agenesis, Congenital Absence of Uterus and Vagina (CAUV), Mullerian Aplasia, Mayer-Rokitansky-Kuster-Hauser-Syndrome (MRKH SYNDROME), Genital Renal Ear Syndrome (GRES).

MRKH syndrome/mullerian agenesis is a congenital anomaly in female due to the developmental failure of vagina from the mullerian duct resulting in aplasia of the uterus or presence of rudimentary uterus and upper part of the vagina.¹ Women with vaginal agenesis have normal ovaries and external genitalia but possess only a vaginal dimple or blind ending vaginal pouch. They undergo puberty, develop breasts, underarm and pubic hair (normal secondary sexual characteristics) except menstrual bleeding.

The initial and most common sign for MRKH syndrome is primary amenorrhea. Primary amenorrhea is a condition in woman wherein there is absence of menarche up to 16 years of age. The other clinical manifestations of this syndrome include hearing defects, a possible spinal abnormality such as curvature of the spine and renal malformations [1].

The occurrence rate of this syndrome is 1 in 4500-5000 female births [2]. It is mainly sporadic; however, a subset of patients may have MRKH as an inherited disorder. It has autosomal dominant mode of inheritance with incomplete penetrance and variable expressivity. They also show a normal 46, XX karyotype [3]. The exact etiology of this syndrome remains obscure. The attributing factors include genetic or environmental. Hence an elaborate Family history should be obtained from the patient and her parents [2].

Women with vaginal agenesis commonly suffer from mental depression and they are deprived of normal social life. It is necessary to educate and motivate the patients and their family members about the various treatment options available and carry out the treatment which is best suited according to the patient's needs. The most common treatment modalities include non-surgical methods (use of neovaginal prosthesis) or surgical methods or combination of both. Surgical creation of neovagina along with the use of a neovaginal prosthesis / vaginal dilator can go a long way in improving the quality of life of the patient.

Investigations

The diagnostic tests that the patient must undergo are mainly determined based on the family history. The tests include Transabdominal ultrasonograph, urography and CT scan of abdomen, Magnetic resonance imaging (MRI), Celioscopy and Biologic status of the patient.

Transabdominal ultrasonograph is a simple and noninvasive method. It is usually the first investigation carried out in patients with suspected mullerian agenesis. This technique reveals an absence of the uterine structure between the bladder and the rectum [1].

Magnetic resonance imaging (MRI) is a non-invasive technique and provides more sensitive and specific means of diagnosis. It is performed when ultrasonographic findings are inconclusive or incomplete. MRI allows an accurate evaluation of the uterine aplasia, as well as a clear visualization of the rudimentary horns and ovaries [1].

Celioscopy is an invasive technique requiring hospitalization and anesthesia. It is performed in cases of doubtful diagnosis after ultrasonography and/or MRI. Celioscopy is nowadays mainly reserved for women in whom surgical construction of a neo-vagina is likely to be undertaken. It defines the precise anatomical location and abnormalities of the uterus, the possible tubar remnants, the vestigial lamina and the ovaries [1].

Biological status - Karyotyping can be carried out in patients with mullerian agenesis. It is used to examine the chromosomes in a sample of cells. Females

with MRKH syndrome have a normal 46,XX Karyotype with no visible chromosome modification.

Once MRKH syndrome is identified, a complete clinical, laboratory and radiographic investigations has to be done to rule out any other associated malformations. Since renal and skeletal abnormalities may not be symptomatic, it is necessary to perform transabdominal ultrasonography and spine radiography. In case of suspicion of hearing impairment and/or a cardiac anomaly, complementary audiogram and/or echocardiography must also be carried out.

Moreover, when diagnosing an MRKH syndrome in a patient it is important to consider the family history. Depending on the background, investigation of the patient's relatives may also be recommended for renal and skeletal malformations [1].

Differential Diagnosis

The differential diagnosis of MRKH syndrome include: the congenital absence of uterus and vagina (aplasia or agenesis), isolated vaginal atresia, WNT4 defects and androgen insensitivity syndrome (AIS) or Mullerian derivative aplasia [4].

Management

The common treatment modalities for vaginal agenesis include: non-surgical, surgical or combination of both.

Common Surgical Techniques

1. McIndoe's Technique
2. Davydou technique
3. Vecchietti laparoscopic technique
4. Creatsas vaginoplasty
5. Wharton-Sheares-George neovaginoplasty
6. Vaginal loosening using LASER

Non Surgical Techniques

1. Psychological counselling
2. Neovaginal prosthesis/Vaginal dilator/ stent
3. Franck's dilator [2].
4. Hormone treatments [5].
5. Vaginal steam bath [5].

Materials Used For Fabrication Of Neovaginal Prosthesis

The various materials used for fabrication of neovaginal prosthesis/ vaginal dilator as mentioned in the literature are: Silicon, foam rubber, wood, plastic, glass, teflon, dextran, vacuum expandable condom, simple syringe, polyethylene bag, heat polymerizing acrylic, autopolymerizing acrylic and PLA. PLA or Poly Lactic acid is a biodegradable polymer derived from lactic acid, designed by 3D technology (a material which is highly biocompatible and has the property to stimulate epithelial regeneration [6].

CLINICAL CASE REPORT

A 17 year old female patient was referred to the Department of Prosthodontics, Sree Mookambika Institute of Dental Sciences, Tamil Nadu, India, from the Department of Gynaecology, Sree Mookambika Institute of Medical Sciences, for the fabrication of neovaginal prosthesis. The girl was unmarried and was accompanied by her parents.

Clinical examination was carried out by a gynaecologist. On examination, patient had normally developing secondary sexual characteristics and showed the presence of vaginal agenesis. The patient was diagnosed with MRKH Syndrome, with normal secondary sexual characteristics as per the findings obtained from clinical examination, radiological and other investigations.

The diagnosis, treatment plan, duration of the treatment as well as the cost were explained and discussed with the patient and her attendee. After procuring a written consent from the patient, the treatment was commenced. Patient was informed that the treatment will provide a better quality of life.

Fabrication of the Neovaginal Prosthesis

The treatment plan was to fabricate a hollow vaginal prosthesis using autopolymerizing acrylic.

The stent was fabricated with a dimension of 8.5 cm × 2 cm, according to the dimensions acquired from the Department of Gynaecology.

1. Wax pattern was fabricated using modelling wax to the required dimension. The wax pattern was cylindrical in shape with rounded top and a small depression near the base for finger grip.
2. Then an index of the wax pattern was made with poly vinyl siloxane impression material (Dentsply, Aquasil, India).
3. The index was then split into half and autopolymerising acrylic was adapted to each half of the mold to a thickness of approximately 2mm.

4. The two halves of the stent were then joined together using autopolymerizing resin to obtain a hollow vaginal stent and the whole unit was immersed in hot water.
5. The final prosthesis was very well trimmed and polished to a high gloss to eliminate sharp areas in the prosthesis.
6. The prosthesis was stored in water for 21 days. After which it was delivered to the Department of Gynaecology and the patient was instructed about proper care and maintenance of the prosthesis.

The prosthesis was light weight as it was hollow from the inside. It was cylindrical and round with a narrowed top to allow easy insertion and had slight depression near the base for finger grip that would help with easy insertion and removal.

The patient was advised to use the prosthesis with the help of a lubricant jelly (K-Y lubricating jelly) for easy and painless insertion. She was also instructed to use the stent for a duration of 4-5 hrs per day after surgery for 2 weeks and then the whole night thereafter.

Pressure was applied and maintained with the help of tight underwear. Instructions about personal hygiene and maintenance of the prosthesis was given to the patient, that it should be held under running water and cleaned with mild vaginal wash to prevent fungal and bacterial growth, the prosthesis was to be dried with soft towel or paper towel and that it should be stored in normal tap water when not in use.

The vaginal stent was fabricated before the surgery which would be eventually inserted into the neo-vaginal cavity. It was stored in normal tap water for 21 days, for the residual monomer to get released and the water was changed everyday. The purpose of the prosthesis was to maintain the passage created during the surgery and it was a critical cornerstone of the treatment. The acrylized prosthesis was handed over to the surgeon pre-operatively. The same was to be used by the patient after the surgery for a period of 3-4 months.



Fig-1: Wax pattern for the neovaginal prosthesis



Fig-2: Neovaginal prosthesis fabricated using autopolymerizing acrylic

Follow up

The patient was reviewed 1 month after the surgery. The neovaginal prosthesis was inserted by applying lignocaine gel to prevent painful insertion. The prosthesis had good stability during functional movements. The patient was trained to insert the prosthesis by herself and advised for its maintenance and regular use.

The patient was satisfied with the current prosthesis and was comfortable using it. Minimal contracture of the neovaginal tissues was observed as

the tissues were healing and the patient did not report any signs of discomfort. Hence the patient was advised to continue using the same prosthesis and replacing with a larger one was not necessary.

DISCUSSION

Vagina is a muscular and elastic organ which is a part of the female genital tract leading from external genitals to the cervix of the uterus. There are various malformations that can occur in the female genital organ, one among them being Mullerian Agenesis. Mullerian Agenesis is also referred to as Vaginal

Agenesis, Congenital Absence of Uterus and Vagina (CAUV), Mullerian Aplasia, Mayer-Rokitansky-Kuster-Hauser-Syndrome (MRKH Syndrome), Genital Renal Ear Syndrome (GRES).

The Mullerian ducts (or paramesonephric ducts) are paired ducts of the embryo that runs down the lateral sides of the urogenital ridge and terminate at the Mullerian eminence in the primitive urogenital sinus. They are present in both sexes but in the females, they will develop to form the fallopian tubes, uterus, and the upper portion of the vagina and in males they degenerate.

Mullerian agenesis or congenital absence of vagina can occur as an isolated developmental defect or as a part of complex anomaly. It is a rare condition and is the cause for around 15% cases of primary amenorrhea. Amenorrhea is the condition where there is absence of menstrual bleeding. It is of two types; primary amenorrhea and secondary amenorrhea. Primary amenorrhea is the absence of menarche up to the age of 16 years and secondary amenorrhea refers to the absence of menses more than 3 menstrual cycles [2].

Vaginal agenesis commonly forms part of MRKH syndrome. These patients show normal development of secondary sexual characteristics and a normal 46,XX karyotype. They have normal external genitalia and ovaries but only a vaginal dimple or blind ending vaginal pouch. It is mainly sporadic; however, a subset of patients may have MRKH as an inherited disorder. It has autosomal dominant mode of inheritance with incomplete penetrance and variable expressivity. The etiology remains obscure. The attributing factors include genetic or environmental. Family history also plays an important role in the examination process [2].

The other organ abnormalities seen in patients with MRKH Syndrome are: Renal abnormalities seen in up to 40% of women affected, Skeletal abnormalities reported in 12–20% of women affected, auditory malformations are seen in 10–25% of cases, malformations of the ear and auditory canal and rarely cardiac malformations are also reported [7].

There are several treatment options to treat Mullerian agenesis. The two main treatment options are surgical and non-surgical methods. The treatment option should be tailored according to the patient's needs.

Patients diagnosed with mullerian agenesis usually suffer from anxiety and psychological distress which affects their social well being. All these have an impact on their family members too. To overcome these issues it is necessary that the patient as well as their family undergo psychological counseling, to educate

and motivate them about the treatment before commencement of the procedure [6].

Surgical treatment for creation of neovagina is being widely practiced. Historically, the creation of a neovagina by Abbe-McIndoe's technique is the most commonly used surgical procedure. It involves dissection of the space between rectum and urinary bladder. Followed by the placement of a prosthesis like a neovaginal prosthesis/ stent/ vaginal dilator covered with a split thickness skin graft.

Laparoscopic Vecchietti procedure. is a technique frequently used in Europe which is a mixture of surgical and non surgical procedures. Other procedures for the creation of the neovagina include the Davydou technique, Creatsas vaginoplasty, and Wharton- Sheares-George neovaginoplasty [6].

The recent methods include vaginal loosening using LASER, vestibulectomy, Carbon dioxide LASER and flash lamp excited dye LASER technique. But most common and conservative treatment includes dilator therapy, which involves use of neovaginal prosthesis [5].

The non surgical creation of a neovagina with the help of prosthetic vaginal dilators is usually the first line of treatment if suitable. Dilators are often helpful in widening a narrowed vagina, without resorting to surgery. It is a preferred mode of treatment as it is a more affordable method. They are easily fabricated and can be modified if needed intraoperatively or postoperatively.

Surgical treatment should be considered only when the patient can participate in the decision making and is highly motivated to use a vaginal prosthesis for several months after surgery. The use of a vaginal stent/ vaginal dilator is highly recommended after surgery to prevent restenosis and also acts as a hemostat, to keep the tissues around the operation site healthy.

These dilators are very important for the success of the treatment and a maxillofacial Prosthodontist plays a pivotal role in the fabrication of the prosthesis. Failure to wear these stents is one of the major cause for treatment failure. If not properly constructed it can lead to graft maceration, sloughing and graft detachment [3].

Other Non-surgical procedures include hormone treatments to help enlarge the vagina. However, these treatments can be dangerous for women with a family history of certain types of cancer, such as breast cancer because hormone therapy is linked to the development of these cancers [5]. Another non surgical procedure is vaginal steam bath.

Some of the other non-surgical methods which are of historical importance for creation of neovagina include Frank's hand held dilator method & Ingram method.

Frank's hand held dilator method is self dilation of the vagina by using the patient's fingers; the limitation of this method is fatigue and uncomfortable position.

Ingram's variation of this method was the bicycle stool method which provided perineal pressure via the narrow and elevated anterior portion of the bicycle seat. In the year 2006 Mee-Hwa Lee modified Ingram's technique by using an ordinary chair.

The fabrication of neovaginal prosthesis using autopolymerizing acrylic as described in this case report has concluded a satisfactory result. The prosthesis was light weight as it was hollow from the inside and the patient was comfortable. This prosthesis was made user friendly so that the patient can remove and wear it by herself.

The advantage of the technique mentioned in this article was the use of tight underwear which applied firm, continuous and constant pressure to the vaginal tissue without the use of the patient's hands.

Vaginal prosthesis fabricated with acrylic resin helps to achieve better functional success as they are hard and hence minimizing chances of restenosis. The prosthesis was immersed in water for 21 days before handing it over to the surgeon before surgery. This was done to get rid of the residual excess monomer which if present can cause tissue irritation.

Prosthesis made with silicone material are soft and more flexible which may lead to failure in achieving the desired size of vagina. It can also deteriorate and tear in long term use. Acrylic material is economic when compared with silicone. It is less tedious and can be easily relined. It is also easy to keep it clean, preventing any deposition, allowing for better hygiene [4].

Silicone vaginal stents if not maintained properly can be more prone for fungal infections and deterioration with time. Taking into consideration all the above factors, the vaginal dilator was fabricated

with auto polymerizing acrylic resin which gave satisfactory results.

CONCLUSION

Neovaginal prosthesis/Vaginal dilator/vaginal stent plays a vital role in the post-operative success of the treatment for MRKH syndrome/ Mullerian agenesis. It is an ideal passive method which is carried out after psychological counseling and surgical creation of neovagina. As the vaginal tissue is elastic the dilator would help to maintain the patency of the neovagina. Fabrication of vaginal stent using autopolymerizing acrylic is very simple, relatively easy, cost effective technique and it offers a convenient removable treatment option for maintaining the patency of the neovagina which can go a long way in improving the quality of life of the patient.

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