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A Case Series on Vaginoplasty Using Amnion Graft in Women with Mrkh Syndrome – Our Experience

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ABSTRACT:

Vaginal agenesis seen in Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome is a condition with significant psychological and social consequences where there is embryonic underdevelopment of müllerian duct. Mayer-Kuster- Hauser (MRKH) syndrome is an uncommon congenital condition characterized by agenesis of mullerian structures, uterus and upper 2/3rd of vagina with fully developed secondary sexual characteristics and is one of a cause for primary amennorhea.

Normalizing sexual life with vaginal reconstruction to adequate length is usually the treatment of choice. Among many non-surgical and surgical options available, **McIndoe vaginoplasty** is the commonest surgical management performed with the use of amnion as a graft. Here we present a case series of 3 women with MRKH syndrome with vaginal agenesis treated with a successful amnion graft vaginoplasty.

1. Introduction

Vaginal agenesis is a significant distress in a woman's sexual and reproductive life. Vaginal agenesis is a rare congenital anomaly of female genital tract. Such malformations of vagina are uncommon. Vaginal agenesis is estimated to occur in 1 in 4000-5000 live birth[1]. Primary amenorrhea the most common clinical presentation in unmarried, pubertal age females and dyspareunia or primary infertility in newly married women. Most commonly seen with Mayor-rokitansky –kuster hauser syndrome and Androgen insensitivity syndrome.
Women with MRKH Syndrome have a female chromosome pattern (46 XX) and normally functioning ovaries. There is no known exact cause but MRKH syndrome is associated with the (Wnt4) mutation, LIM homeobox 1 (LHX1), HNF1 homeobox B (HNF1B) and T-box 6 (TBX6) mutations.[A]
There are 2 types of MRKH syndrome: **TYPE 1**- having only uterovaginal agenesis and **TYPE 2-** uterovaginal agenesis with anomalies in fallopian tube, kidney, spine and heart. They also have normal female external genitalia and normal breast and pubic hair development. Although women with this condition are usually unable to carry a pregnancy, they may be able to have children through assisted reproduction.
Vaginal reconstruction has become a well established method to permit or restore sexual function and a variety of procedures have been described.
Historical background: Surgical correction of vaginal agenesis has long history of evolution as summarised below:
1907 – Baldwin used double loop of ileum for covering the neovagina.
1910 – Popaw used the rectum mobilised anteriorly to cover the raw area of newly created vagina.
1911 – Schubert modified the Popaw's method.
1938 – Wharton, in Johns Hopkins used a balsa form covered with a thin rubber sheath to dilate the newly created vagina.
1950-1989 – Mc Indoe used skin graft in Johns Hopkins Hospital in 94 patients with 83% success rate. He used foam rubber form as mould. [C]
In 1910 Davis was first to report the use of foetal membranes as surgical material in skin implantation, since then use of amniotic membrane in surgery has been expanded. In 1934 , Brindeau used human amnion to construct the vagina for a patient with mullerian agenesis.

The most popular method involves lining a surgically
created space either with partial thickness skin graft or with amnion. The **modified Abbe-McIndoe method** is the preferred and fruitful procedure.
[Amniotic membrane is considered a useful material for grafting the artificially created vagina for the following reasons: i) It facilitates migration of epithelial cells.
ii) It reinforces basal cells adhesion.
iii) It promotes epithelial differentiation.
iv) It prevents epithelial apoptosis.
v) It Promotes epithelialisation in healing of the wounds.
vi) Good permeability of basement membrane of amnion facilitates good oxygenation of epithelial cells [B]]

Amnion membrane graft is the optimal material for vaginal reconstruction because it is readily accessible, and affordable; furthermore, the graft is safe and simple with physiological benefits, and excellent functional outcome.
In our case it is a patient of MRKH syndrome (type 2 or type B) managed surgically with vaginoplasty.

Case Presentation

Case 1:

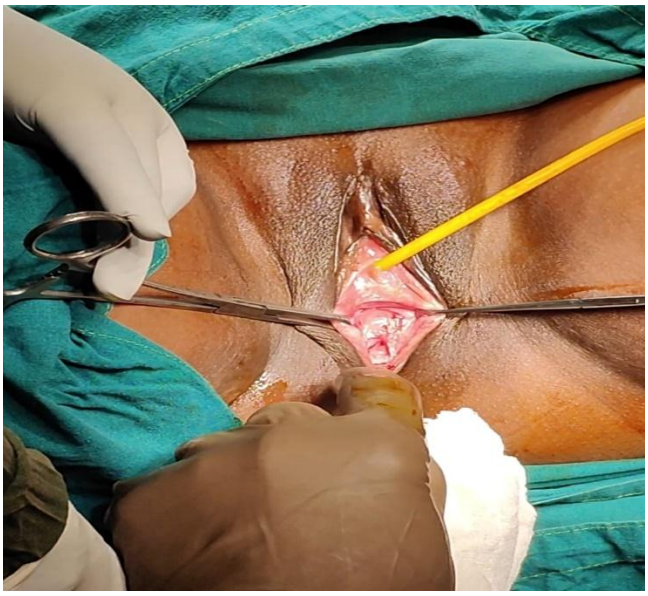
A 30 year old female, married since 2 years, nulligravida hailing from Pune, Maharashtra came to the OPD of Dr D Y Patil medical college and hospital with a history of failure to attain menarche and complaints of difficulty during intercourse since 2 years.

She had no cyclical abdominal pain, no history of amennorhoea in first and second degree relatives, no significant drug history, and no past surgical history. She had a history of laproscopic vaginoplasty 2 years back which was failed.

On physical examination her general condition was fair with stable vitals. Systemic examinations were normal.

On further examination, the patient indicated the development of secondary sexual characteristics including normal breast development of Tanner stage 5, normal stature, normal distribution of pubic hair, and normal external genitalia resembling her age.

On local examination 1.5cm vaginal depression noted with no clear vaginal canal.



(figure1)

She underwent preoperative investigations which included blood investigations ultrasound , MRI and Karyotyping.

On local examination, a 2 cm vaginal introitus was present. Ultrasonography showed an absent uterus, cervix, and upper two third of the vagina. Both ovaries were present. The patient was psychologically mature and was not sexually active. Her bowel and bladder habit

of pubic hair. Labia majora and minora well developed.

On Ultrasonography : A pear shaped small structure noted posterior to urinary bladder measuring 40x 7mm. Severely atretic or hypoplastic uterus and cervix and vaginal canal could not be delineated.

MRI findings:

A lentiform shaped tubular structure seen posterior to the urinary bladder and in the right paramedian suggestive of severly hypoplastic/ rudimentary uterus (nonfunctional myometrium)

Visialised lower 1/3rd of vagina appears to have a blind ending at cranial aspect and short in length.

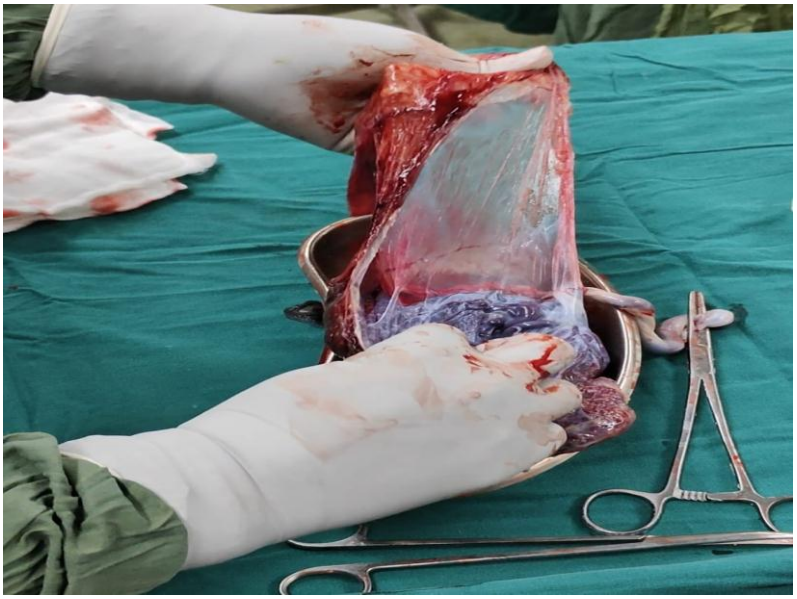
Uterts,cervix, bilateral fallopian tubes and upper 2/3rd of vagina are not visualized.
Ectopic malrotated left kidney seen.
Findings consistent with MRKH type B(figure 6)
All her blood parameters and labs were within normal limits.(table 1)
A diagnosis was made of MRKH syndrome was made and a decision was made to take her up for vaginoplasty.

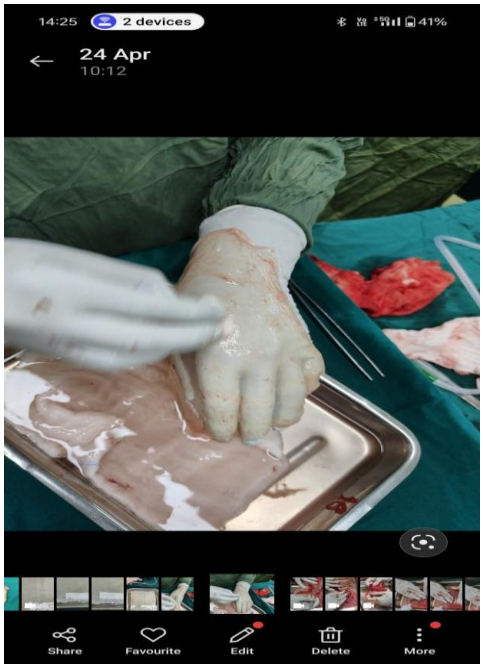
The patient was given antibiotics pre operatively and was planned for **Modified mcindoe vaginoplasty.** Informed consents taken regarding procedure, complications and chances of failures and documented.

CASE 2:
A 29 year old female , unmarried, nulligravida hailing from Pune, Maharashtra came to the OPD of Dr D Y Patil medical college and hospital with a history of failure to attain menarche. She had no significant family history, drug history, and no past surgical history. On physical examination her general condition was fair with stable vitals. Systemic examinations were normal.
On further examination, the patient indicated the development of secondary sexual characteristics including normal breast development of Tanner stage 5, normal stature, normal distribution of pubic hair, and normal external genitalia resembling her age.
She underwent preoperative investigations which included blood investigations ultrasound , MRI and Karyotyping.
On local examination, a 2 cm vaginal introitus was present. Ultrasonography showed an absent uterus, cervix, and upper two third of the vagina. Both ovaries were present. The patient was psychologically mature and was not sexually active. Her bowel and bladder habit
of pubic hair. Labia majora and minora well developed.

On Ultrasonography uterus was not visualized, and cervix and vaginal canal could not be delineated.
MRI revealed normal uterus distinctly not seen, a small ovoid shaped hypointense lesion measuring approximately 15x9 mm likely to be a uterine bud or remnant. Linear hypointense bands are noted extending laterally from it reaching jus caudal to ovary on either side with fibrous bands. Rt ovary is2.7x1.5x1.9cm and Lt ovary is 2.3x1.4x2.3cm. Upper third of vagina is not seen, lower third of vagina seen.(figure 6) Karyotyping of the patient was 46XX.
All her blood parameters and labs were within normal limits.

CASE 3:
Procedure:
Following initial blood investigations patient was taken up for OT, she was anesthetized spinally. Amniotic membranes was obtained under sterile condition from elective caesarean section 30 minutes before the procedure. Amnion donors were screened for Hepatitis B
& C, HIV. Inner amnion membrane was separated from outer membrane

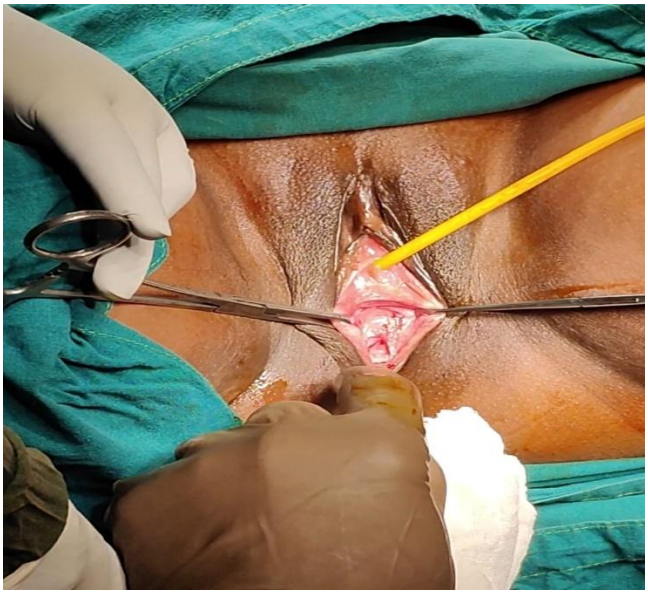


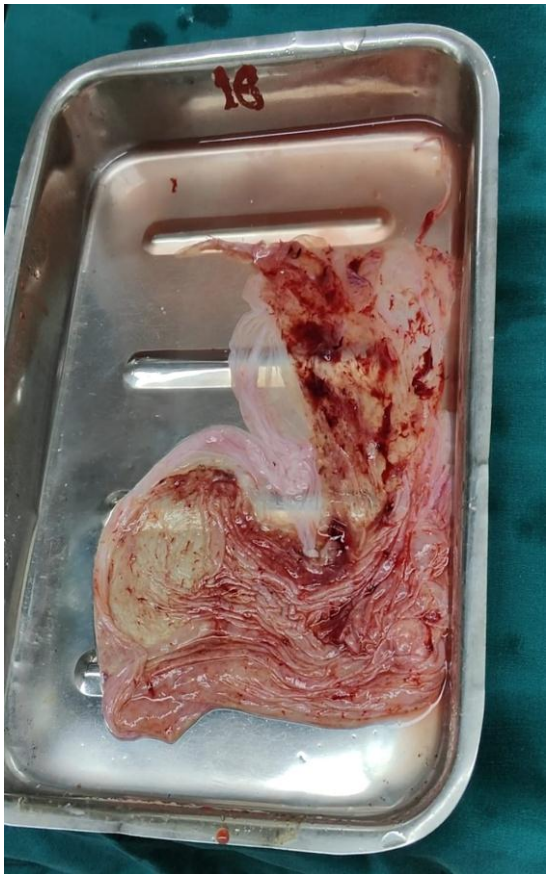


(figure 1) and rinsed in sterile normal saline solution repeatedly along with antibiotic wash with cefotaxime. The washed and prepared amnion was soaked in a solution with normal saline and cefotaxime and kept for 30mins before preparing the mould.

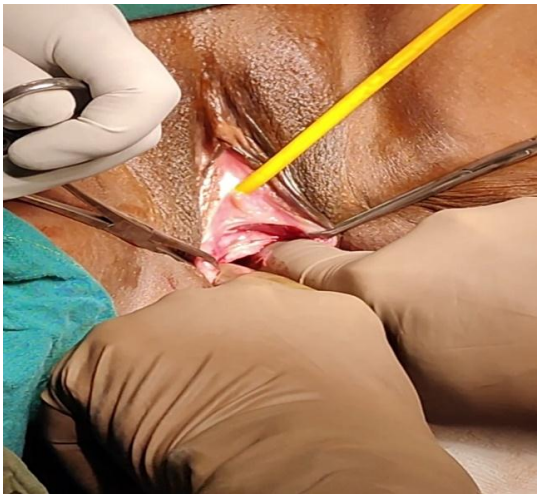
The patient was placed in the lithotomy position. To secure the urethra, a Foley catheter was placed and parts were painted

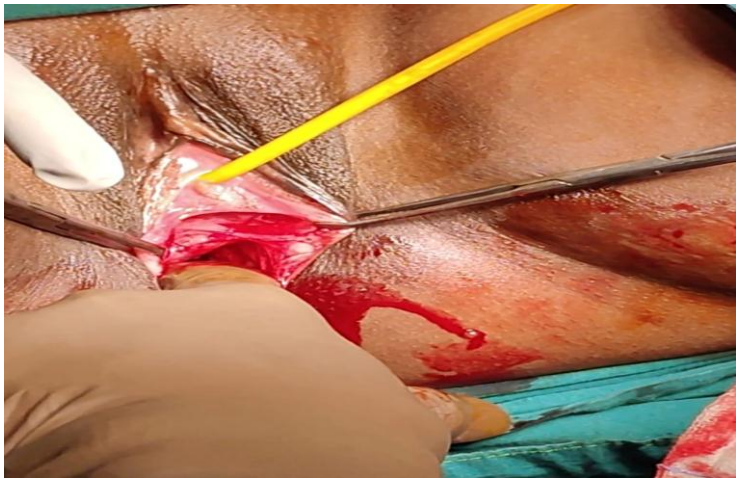
and draped. The labia majora were sutured on to the adjacent skin for better visualization of operative field.



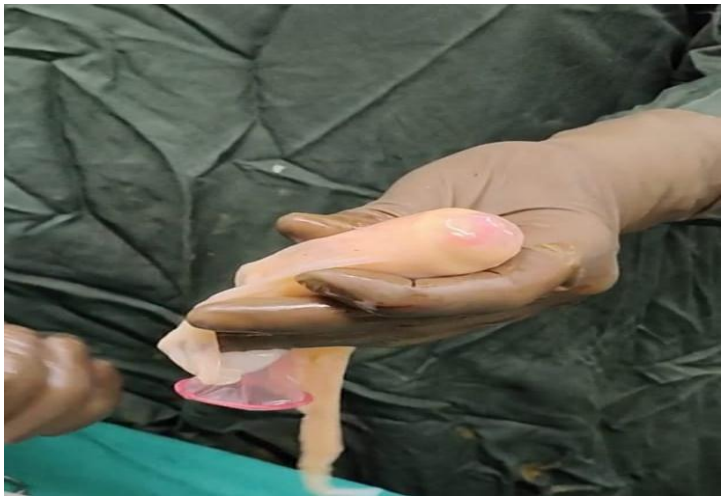


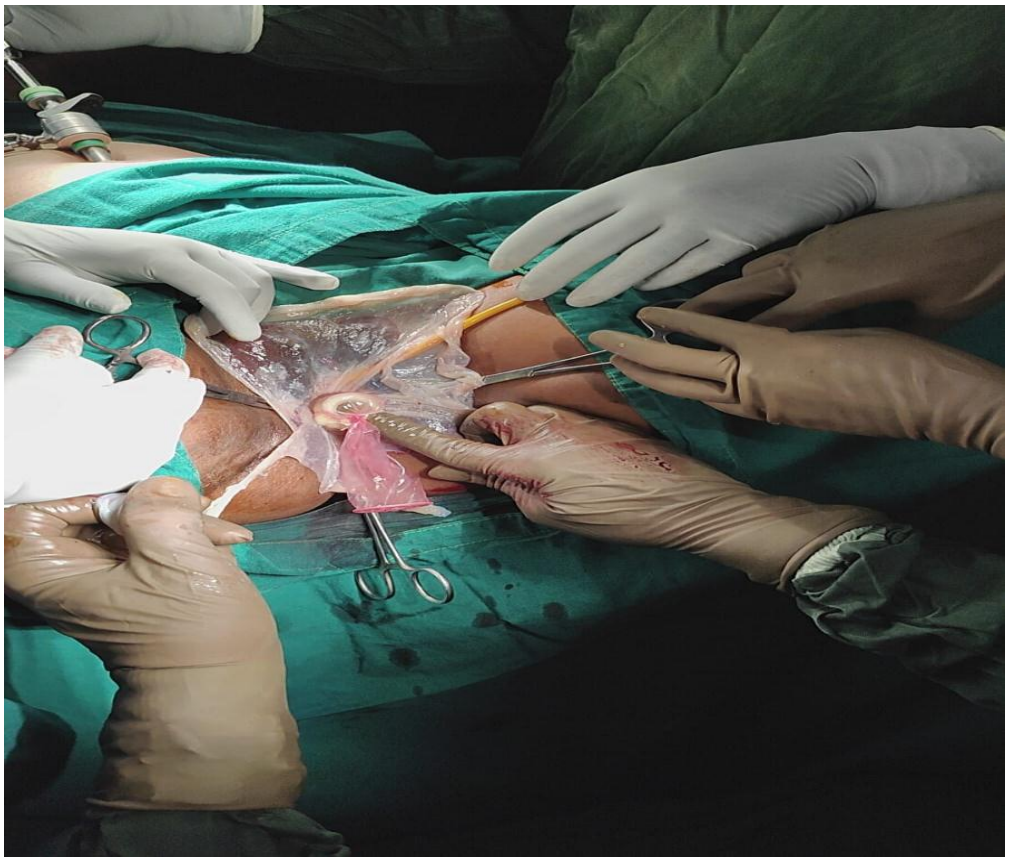
(figure 2) The labia minora were separated and a 3 cm midline incision was made, meticulous dissection was done between vesicorectal space after infiltration of 60 to 80 ml of prepared solution(100ml NS+ 0.5cc of adrenaline) to create a space in the loose areolar connective tissue between bladder and rectum. The cavity is made with blunt dissection accomadating the size of two fingers, without directing the fingers towards bladder or rectum (similar to swimming breaststroke).

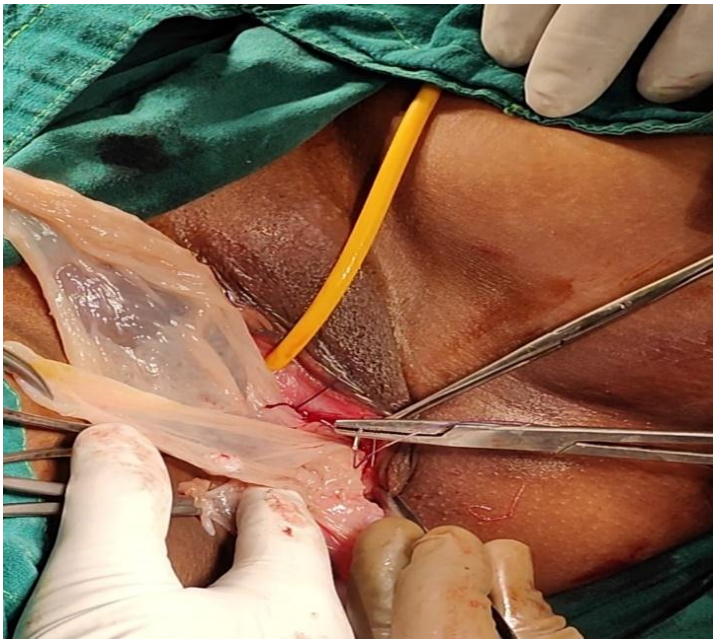




The vagina was enlarged 7-11 cm in depth and 3-4 cm in width so mould can be easily inserted. A vaginal mould was made using a sterile 10cc syringe wrapped with sponge covered with sterile latex condom

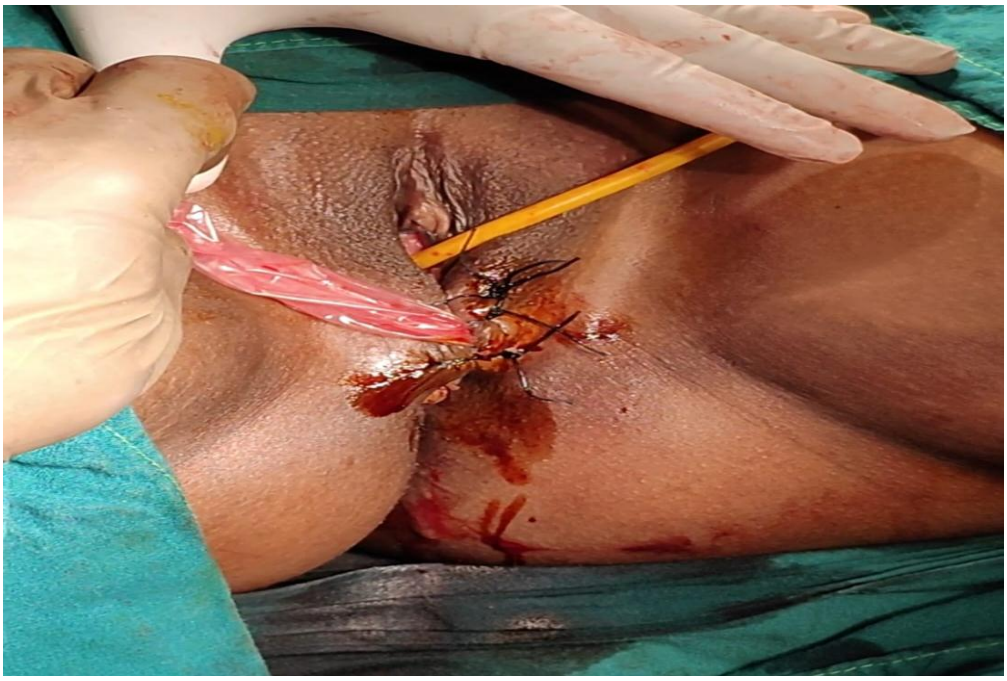






(figure 3). Then amnion membrane was mounted on the mold with a mesenchymal surface outward to adhere surface of the raw neovagina and shiny surface towards condom. The prepared mould using sponge helps in minimising pressure on the tissue but optimises compression of amnion to resected raw area of vagina. After obtaining hemostasis , the mold along with the graft introduced into the vagina and the amnion graft was fixed to the cavity created by vicryl 2-0 simple sutures and edges were sutured onto the mould for stability of the graft.

(figure)Closure of the labia minora secured the mold in place for 7 days.



(figure 4) Patient was given antibiotic for 5 days. On the 6th postoperative day, the labial sutures were cut mold was removed along with the foleys catheter.
In the postoperative examination, there was no pressure necrosis or ulcer. The amnion graft was seen well taken with resultant 6 to 7 cm of vaginal canal when mould was removed on 6th day.
had been taken up and the cav
was discharged with the advice to follow up every fortnight
A polymer vaginal mould available in different sizes

(figure 5) was given to the patient. Markings were made on the mould with permanent marker at 5cm, 7.5cm and 10cm and the moulds were kept in savlon solution and rinsed with NS and sterile condom was applied on the mould and with help , mould was gradually inserted into the neovagina for 15mins daily, 4times a day to ensure adequate patency and taught to correctly sterilize the dilator remove and insert the mould to prevent vaginal narrowing and maintain the length , removal and washing of mould . Patient was discharged with advice to wear the mould for 3 month and to visit forthnightly. Overall post operative period was uneventful.

rate hymen and atresia of the vagina. All other routine investigations were within normal limits. With the diagnosis of MRKHsyndrome she was planned for vaginoplasty with amnion graft.

2. Discussion

The commonest cause of vaginal agenesis is MRKH syndrome
where there is aplasia or severe hypoplasia of vagina resulting in a vaginal dimple of various depths and variable development of uterus. It maybe isolated or associated with skeletal and renal anomalies.
in a vaginal dimple with variable depth and with variable development of uterus. It may be isolated or sometimes associated with renal or skeletal abnormalities.
Vaginoplasty by modified Abbe McIndoe procedure using amnion graft

Patients with MRKH syndrome have a normal female karyotype with normal ovaries and ovarian function, thus they develop normal secondary sexual characteristics. Most of the patients have rudimentary uterus which are nonfunctional while 7-10% may have functional endomentrium.

The principal aim of the treatment is to restore the sexual
The principle aim of the treatment is to restore the sexual function by creating a vaginal canal at the normal anatomical
position and axis. This neovaginal canal is created within an avascular space between urethra and bladder above and rectum below.
There are non-surgical options such as progressive dilation (Frank technique)
(figure)
surgical options, and a combination of surgical and conservative methods (developed by Veccihetti). (D)

Choosing a technique depends on several factors including the patient's condition, the surgeon's experience, and preference. Despite multiple methods being suggested for the creation of a neovagina, the modified Abbe-McIndoe procedure is the preferred technique in developing countries.

Surgical procedures are often performed in late adolescence or young adulthood when the patient is mature enough to agree to the procedure and to be able to adhere to postoperative dilation.

Original McIndoe procedure describes the use of split thickness skin graft in the newly created vagina but several investigators have sought for the modification on the use of graft like peritoneum, buccal mucosa, intestines, amnion etc.[E]

amnions e

avascular space between the urethra and bladder above and

the rectum below. Various non-surgical and surgical methods

have been described. Non-surgical treatment includes self-selfdilata□on with graduated dilators as described by Frank which

may take months to develop a canal. Success depends

Amniotic membrane has been used in clinical medicine for

a long time starting from the use as a skin substitute to uses in wound care and orthopedics and various gynecological procedures.

Amnion as a graft: Amnion is non immunogenic and has been shown to reduce inflammation and pain. It serves as a matrix for cell deposition. The amniotic membrane contains the following : growth factors, cytokines, vascular endothelial growth factors, platelet derived growth factors AA and BB, transforming growth factors alpha & beta, basic fibroblast growth factor, epidermal growth factor, granulocyte – colony stimulating factor and interleukins 4,6,8 and 10.

.[B]

in ophthalmic surgery, and now with uses in wound care,

orthopedic, and various other disciplines like gynecology. It

It is very cheap and easily available and its anti-inflammatory

and antiscarring properties helps in regenerative and healing

7

processes. Acute immune rejection does not occur after the

Faulke et al have demonstrated microscopic evidence of new vessel formation and suggested that an angiogenic factor is produced by amnion.

Morton and Dewhurst reported more than 90% success rate when they operated with amnion graft in 27 patients suffering from MRKH syndrome. Similarly Sharma et al after performing amnion graft vaginoplasty in 17 patients showed 88% success rate. Both the investigators reported complete epithelialization and metaplasia of the amnion into squamous cells. This provides a natural layer for wound healing and contains various important growth factors and biological macromolecules important in wound healing so that postoperative scarring, pain and infections are also less than that seen in skin graft due to its anti-inflammatory properties.

After 2 weeks of the surgery, our patient had 4 to 5 cm of vaginal length but the success depends upon compliance of the patient on regular self-dilatation of the newly created vagina and regular sexual activity to maintain the patency of the neovagina.

success

The psycological impact of the condition must not be underestimated, hence the requirement of proper counselling is essential.

3. Conclusion

Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome is a rare congenital malformation, associated with absence of Mullerian structures, uterus, and upper two-thirds of the vagina, in

presence of normal secondary sexual characteristics, primarily, presenting with amenorrhea. Various surgical and non-surgical vaginoplasty methods have been developed.
Modified Abbe-Mclndoe procedure with amnion graft is an effective, rapid and simple technique with the amnion membrane readily available, easily stored and inexpensive and can be used without sterilization as a graft for vaginal reconstruction with little chance of vaginal restenosis in the future, given the good
patient compliance.
Amnion graft vaginoplasty has with good success rate with minimal postoperative pain, infection and scarring and thus is a rewarding procedure especially in developing countries like our own..
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9. is simple and rewarding procedure especially in developing country
10. by embryologic underdevelopment of the müllerian duct, with
11. resultant agenesis or atresia of t