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Case Report

Mayer-Rokitansky-Küster-Hauser syndrome: a case report and surgical approach for neovagina construction

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ABSTRACT

Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome is a rare congenital anomaly characterized by agenesis or hypoplasia of the uterus and upper two-thirds of the vagina in phenotypically normal females with a 46, XX karyotype. Despite normal ovarian function and secondary sexual characteristics, affected individuals present with primary amenorrhea. We report the case of a 21-year-old female presenting with primary amenorrhea and normal secondary sexual development. Pelvic MRI demonstrated complete uterovaginal agenesis with small fibrofatty structures suggestive of uterine buds, and normal bilateral ovaries. Hormonal profile was within normal limits, except for mildly elevated thyroid-stimulating hormone. After multidisciplinary counselling, the patient underwent neovaginoplasty using a modified McIndoe technique with an amniotic membrane graft and foam mold support. Postoperative care included gradual mold dilations and psychological support. At follow-up, the neovagina maintained adequate depth (>8 cm) and caliber, with satisfactory functional outcome and no early complications. Surgical neovaginoplasty using amniotic grafts is a safe and effective option for MRKH patients when nonsurgical methods are unsuitable. Success depends on meticulous surgical technique, structured postoperative dilation, and comprehensive psychological counseling.

Keywords: Mayer-Rokitansky-Küster-Hauser syndrome, Müllerian agenesis, Primary amenorrhea, Vaginal agenesis, Müllerian duct anomalies, Neovagina

INTRODUCTION

Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome is a congenital Mullerian duct anomaly characterised by complete or partial absence of the uterus and upper two-thirds of the vagina in otherwise phenotypically and karyotypically normal females (46, XX). The condition has an estimated incidence of 1 in 4,500-5,000 female live births and represents the second most common cause of primary amenorrhea after gonadal dysgenesis.

MRKH syndrome is classified into two subtypes: Type I (isolated): uterovaginal agenesis without other malformations. Type II (syndromic or MURCS association): associated with renal, skeletal, and occasionally auditory orcardiac anomalies.[1]

Patients typically present during adolescence with primary amenorrhea despite normal breast and pubic hair development, normal external genitalia, and normal ovarian hormonal function. Imaging, particularly pelvic MRI, plays a pivotal role in confirming the diagnosis, delineating pelvic anatomy, and identifying associated anomalies.

Management of MRKH syndrome requires a multidisciplinary approach involving gynecologists, radiologists, psychologists, and reconstructive surgeons. First-line therapy for creation of a functional neovagina is nonsurgical dilation; however, surgical options such as McIndoe, Vecchietti, Davydov, or amnion graft-based techniques are indicated when conservative measures fail or when rapid results are desired. We present a case of type I MRKH syndrome in a young woman managed with

amnion graft neovaginoplasty, emphasizing surgical details, postoperative care, and a review of the literature.[2]

CASE REPORT

A 21-year-old unmarried female presented to the gynecology outpatient department of MMIMSR, Mullana, with a complaint of primary amenorrhea. There was no history of cyclical abdominal pain, urinary tract symptoms, bowel disturbances, or similar complaints in family members.

General examination revealed a height of 149 cm, weight 42 kg, and a BMI of 18.9 kg/m^2. Blood pressure was 120/70 mmHg. The patient was well nourished and in good general health.

Secondary sexual characteristics were fully developed, corresponding to Tanner stage V for both breast and pubic hair development. External genital examination revealed normally developed female genitalia. On closer inspection, a shallow vaginal dimple was present. A grooved urethra with slightly elevated margins was noted.

Speculum examination was deferred in view of the patient's age and virginity.

Investigations

Hormonal profile

FSH: 6 IU/L, LH: 11 IU/L, estradiol: 76 pg/mL, prolactin: 16 ng/mL-T3: 1.39 ng/mL, -T4: 7.8 ug/dL and TSH: 6.66 uIU/mL (mildly elevated).

Transabdominal ultrasonography: The uterus was not visualized; ovaries were poorly delineated.

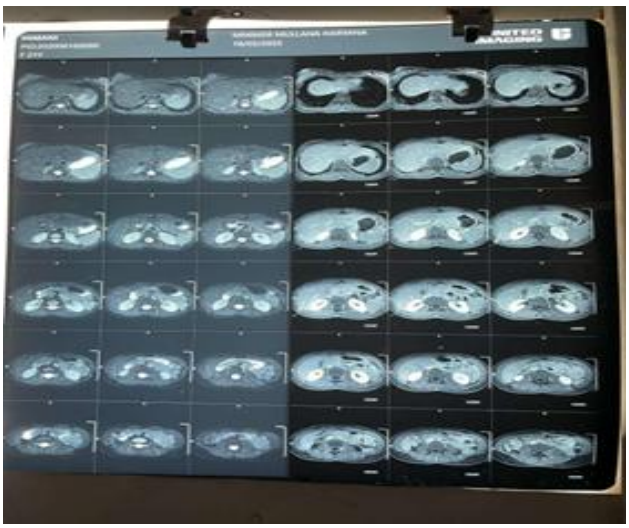

Figure 1: Pelvic MRI (axial sections) showing complete uterovaginal agenesis with normal bilateral ovaries, MRI sections confirming absence of uterus and upper vagina.

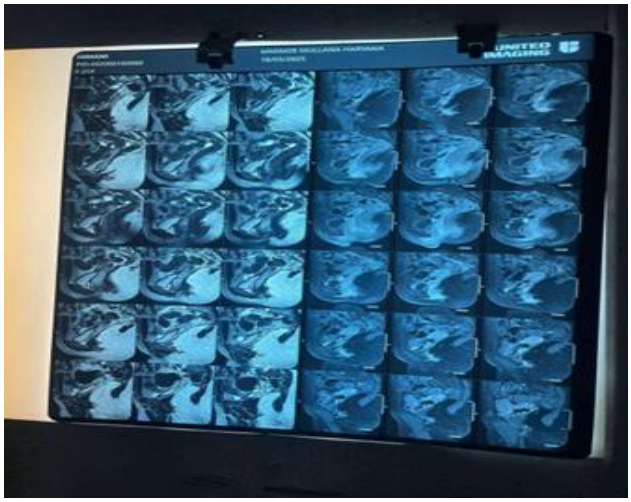

Figure 2: Pelvic MRI (sagittal) showing complete uterovaginal agenesis with normal bilateral ovaries, MRI sections confirming absence of uterus and upper vagina.

Pelvic MRI findings

Uterus: Not visualized-suggestive of complete uterine agenesis.

Cervix and vagina: Cervix and upper two-thirds of the vagina absent.

Soft tissue: Fibrofatty tissue posterior to the urinary bladder with mild enhancement, likely representing connective tissue in the expected uterine location.

Uterine buds: Small focal lesions adjacent to both ovaries-left: 40.5 30.5 mm, right: 38 19.5 mm suggestive of rudimentary Mullerian remnants.

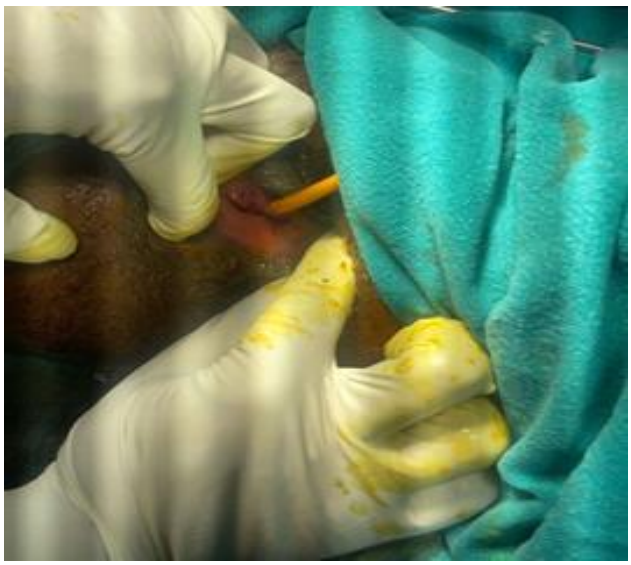

Figure 3: External genitalia with normally developed labia and urethral meatus; shallow vaginal dimple visible.

Figure 4: Prepared foam vaginal mold wrapped with amniotic membrane prior to insertion into neovaginal space during surgery.

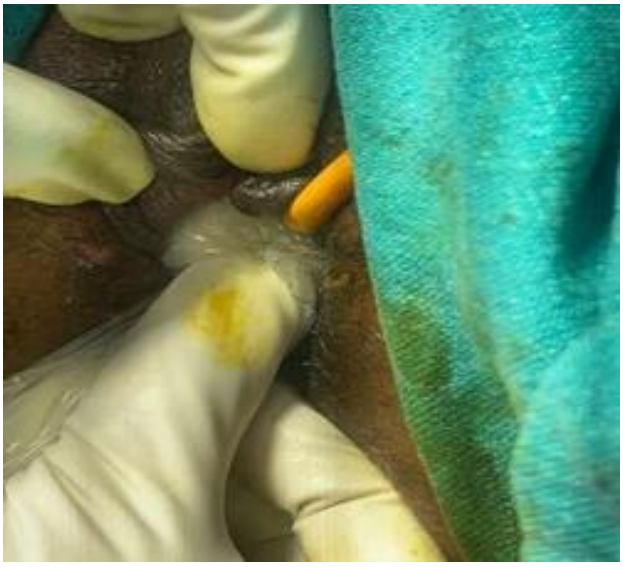

Figure 5 : Intraoperative view showing mold insertion into surgically created neovaginal canal.

Management and surgical procedure

Given the patient's upcoming marriage and expressed desire for satisfactory sexual function, surgical creation of a neovagina was considered. Although primary vaginal elongation by patient- controlled dilatation is the first-line approach in most women with Mullerian agenesis, the patient opted for surgical intervention after detailed counselling. Preoperative counselling included risks of haemorrhage, infection, injury to bladder/urethra/rectum, graft necrosis, vaginal stenosis, fistula, dyspareunia, anesthetic complications, and revision surgery. Psychological counselling was also provided.

Surgical technique-modified McIndoe procedure with amniotic membrane graft: Under general anesthesia, patient in dorsal lithotomy position, Foley catheter *in situ*. Transverse incision at vaginal dimple; sharp and blunt dissection in vesicorectal space to ~8-10 cm depth. Fresh amniotic membrane obtained from consenting donor placenta, rinsed in antibiotic solution, trimmed, and epithelial side oriented outward around a foam vaginal mold. Mold with graft inserted into neovaginal space, graft margins sutured to introitus with 4-0 Vicryl, labia approximated with nonabsorbable sutures. Foley removed on day 3, mold removed on day 10, daily povidone-iodine irrigation and self dilatation initiated.

Outcome

Neovagina depth >8 cm with adequate width, no early postoperative complications, and good compliance with mold use.

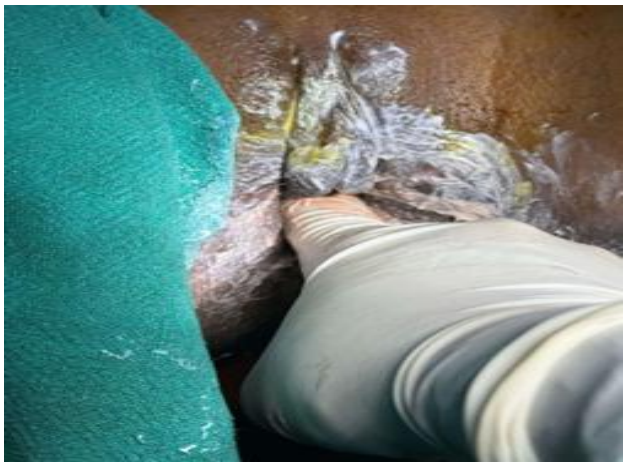

Figure 6: Demonstrating adequate depth and caliber of the neovagina after healing.

DISCUSSION

MRKH syndrome is a rare congenital anomaly, with an estimated incidence of 1 in 4,500 to 5,000 female live births (Morcel et al).[3,4,6] It occurs due to failure of Müllerian duct development, resulting in agenesis or hypoplasia of the uterus and the upper two-thirds of the vagina in otherwise phenotypically and karyotypically normal females (46, XX). Condition is usually diagnosed during adolescence, most often when a patient presents with primary amenorrhea, as happened in our case.

Most Müllerian anomalies go undiagnosed because of low clinical suspicion and the absence of specific clinical or radiological signs. All cases with Müllerian agenesis have primary amenorrhea as the main symptom. MRI and 3D ultrasonography are considered the gold standard for diagnosis.

In our study, the diagnosis of MRKH syndrome was confirmed through clinical evaluation and imaging, especially pelvic MRI. This agrees with the findings of Oppelt et al who highlighted the value of MRI in

distinguishing MRKH from other causes of Müllerian anomalies such as androgen insensitivity syndrome or transverse vaginal septum.[5] Similar to the case reported by Pan et al our patient had fully developed secondary sexual characteristics and normal ovarian morphology, which is consistent with type I MRKH syndrome (isolated uterovaginal agenesis without associated renal or skeletal problems).[9] By contrast, type II MRKH syndrome, also called MURCS association, presents with additional renal, vertebral, or auditory abnormalities.

The psychosocial impact of MRKH syndrome is significant, especially in cultures where early marriage and childbearing are important. Psychological counseling, which we provided to our patient, plays an important role in helping with issues related to identity, femininity, and fertility (Fliegner et al). Our multidisciplinary approach was aimed at supporting the patient emotionally and socially in preparation for marriage. Both surgical and non-surgical options exist for creating a neovagina. Non-surgical dilation methods, such as the Frank method, are often preferred first because they have a low risk of complications (ACOG). However, in our case, the patient chose surgical intervention due to personal and cultural reasons. We performed neovaginoplasty using an amnion graft, which is known for its biocompatibility, anti-inflammatory properties, and ability to support epithelialization (Pan et al). For achieving pregnancy, assisted reproductive technologies such as IVF with gestational surrogacy can give good results for women with Müllerian anomalies.[7] Adoption is another option. In the future, uterine transplantation also seems promising for giving women with uterine agenesis the chance for biological motherhood. The outcome in our case was similar to the results reported by Bhatt et al who had success with the same technique, showing good long-term function and minimal complications.[10] Callens et al compared long-term outcomes of various surgical techniques and found no major difference in function between the Vecchietti procedure and amnion graft vaginoplasty, but the latter often caused less postoperative pain and required simpler aftercare.[11] Our experience matched this, as our patient healed well and complied with regular mold dilations after surgery. Postoperative care is very important-especially the continued use of vaginal molds and following the dilation routine. Poor compliance can lead to narrowing or collapse of the neovagina, as seen in other reports (Banerjee et al).[12] Thankfully, our patient was motivated and understood the importance of care, and she did not have any early complications.

Table 1: Comparison with literature.

Study	Sample size	Technique used	Success rate	Complications
Pan et al	47 patients	Amnion graft vaginoplasty	93.6%	Minor infections, mold
Bhatt et al	28 patients	Amnion with mold therapy	92.8%	None significant
Callens et al	110 patients	Vecchietti vs amnion	Comparable	Vecchietti: more pain
Our case	1 patient	Amnion graft + mold	Early good outcome	Not reported

CONCLUSION

Our case supports current literature in demonstrating that MRKH syndrome, though rare, can be effectively diagnosed through imaging and clinical examination. Surgical vaginoplasty using an amnion graft, combined with regular mold dilation, offers excellent anatomical and functional out- comes. Early psychological support, patient centered care, and proper counselling remain the cornerstone of management in such complex and life-impacting conditions.

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REFERENCES

1. Bhatia R, Aishwarya KK, Kalyanam R. Primary amenorrhea with aplastic uterus, bilateral non-communicating rudimentary horns with hematometra in left horn and cervicovaginal aplasia (USA C4 V4)-a rarest case report. Int J Pharm Sci Res. 2023;14(12):5722-4.
2. Bhatia R, Gupta R, Arora T, Jain S. Davydov's procedure: innovative method for creation of neovagina. Int J Med Sci Innov Res. 2020;5(4):258-62.
3. Morcel K, Guerrier D, Watrin T, Pellerin I, Levêque J. The Mayer-Rokitan-Sky-Küster-Hauser (MRKH) syndrome: clinical description and genetics. J Gynecol ObstetBiol Reprod (Paris). 2008;37(6):539-46.
4. Griffin JE, Edwards C, Madden JD, Harrod MJ, Wilson JD. Congenital absence of the vagina. The Mayer-Rokitansky-Kuster-Hauser syndrome. Ann Intern Med. 1976;85(2):224-36.
5. Oppelt P, Renner SP, Kellermann A, Sara B, Georges AH, Kurt SL, et al. Clinical aspects of Mayer-Rokitansky-Küster-Hauser syndrome: recommendations for clinical diagnosis and staging. Hum Reprod. 2006;21(3):792-7.
6. Morcel K, Camborieux L, Programme de Recherche sur les Aplasies Mülléri-pennes, Guerrier D. Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome. Orphanet J Rare Dis. 2007;2:13.
7. Carr NR, Hann LE, Brotman S. MR imaging of müllerian duct anomalies: comparison with US,

surgical, and clinical findings. Radiology. 1995;194(3):797-802.
8. Edmonds DK. Congenital malformations of the genital tract and their man- agement. Best Pract Res Clin Obstet Gynaecol. 2003;17(1):19-40.
9. Pan HX, Zhu L, Chen J, Lang JH. Comparison of amnion and split-thickness skin grafts in vaginoplasty. Int J Gynaecol Obstet. 2013;120(2):183-6.
10. Bhatt NR, Gupta S, Singh P. Amnion graft vaginoplasty in MRKH syndrome: a case series. J Obstet Gynaecol India. 2021;71(3):283-8.
11. Callens N, De Cuypere G, De Sutter P, Stan M, Steven W, Piet H, et al. An update on surgical and non-surgical treatment options for vaginal agenesis in MRKH syndrome. Hum Reprod Update. 2014;20(5):775-801.
12. Banerjee S, Singhal S, Agarwal N. Evaluation of vaginal mold use post vaginoplasty in MRKH syndrome. J Clin Diagn Res. 2016;10(9):QR01-3.

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