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Case Report
Mayer-Rokitansky-Küster-Hauser

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Mayer-Rokitansky-Küster-Hauser Syndrome: Rare case diagnosis and management

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Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome is a rare congenital disorder characterized by vaginal agenesis or hypoplasia, often associated with uterine anomalies. We present a case of a 35-year-old woman with Type 1 MRKH syndrome, who underwent McIndoe vaginoplasty to create a functional vagina. The patient presented with primary amenorrhea and difficulty with sexual intercourse. Preoperative evaluation confirmed vaginal agenesis, and the patient underwent McIndoe vaginoplasty using a split-thickness skin graft. Postoperative follow-up showed successful creation of a functional vagina, with satisfactory anatomical and functional outcomes. This case highlights the effectiveness of McIndoe vaginoplasty in creating a functional vagina in patients with MRKH syndrome, improving their quality of life and sexual function.

Keywords: MRKH syndrome, vaginal agenesis, McIndoe vaginoplasty, vaginoplasty, congenital disorder

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Introduction

MRKH syndrome is a congenital anomaly characterized by aplasia of uterus and upper two-thirds of vagina in females with normal ovarian function and 46, XX karyotype. It occurs in approximately 1 in 4,500 to 5,000 females [1].

It is classified as Type 1- isolated Mullerian agenesis and Type 2- associated with other organ anomalies like renal, vertebral, and to a lesser extent, auditory and cardiac defects.

These patients present with primary amenorrhea, short or absent vaginal canal with normal external genitalia and secondary sexual characteristics. The condition can be diagnosed on ultrasound or MRI and can be treated by creating a neovagina by surgical or non-surgical means to enable normal sexual functions.

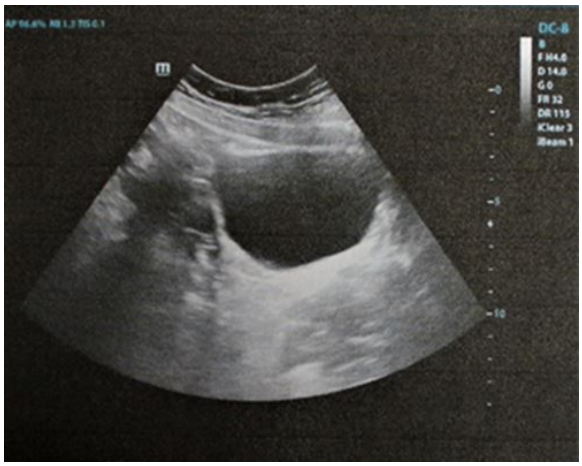


Figure 1: Ultrasound image of Patient with MRKH showing normal abdominal viscera but absent uterus and cervix

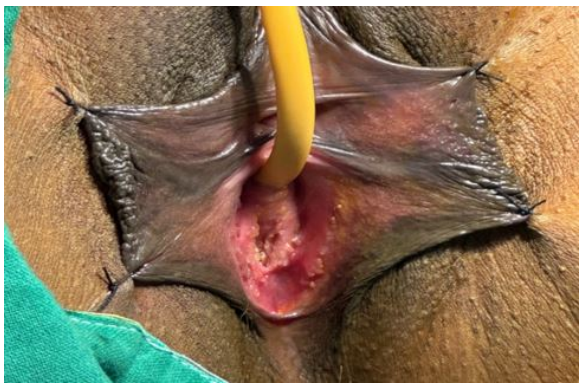
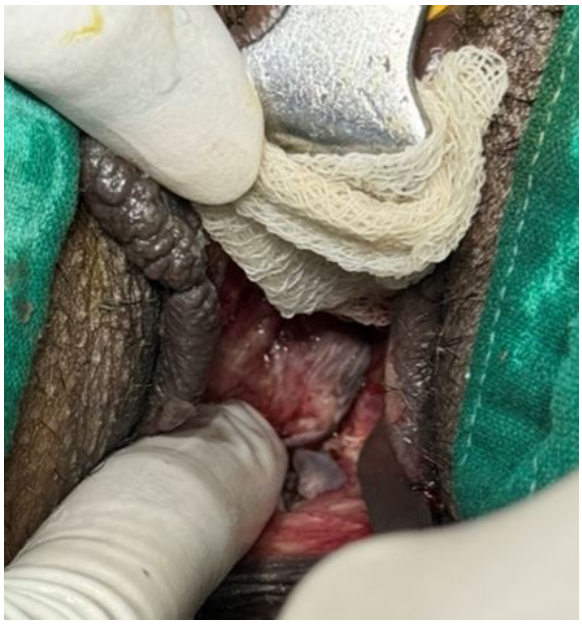


Figure 2: Preoperative image of vaginal dimple.

Case Report

A 32-year-old female married for 10 years with a known case of type 2 diabetes mellitus presented with primary amenorrhea. On clinical examination secondary sexual characteristics and external genitalia appeared normal with a blind dimple at vaginal orifice. Ultrasound was done which was suggestive of MRKHS type 2, with absent uterus and upper vagina with cross fused ectopic left kidney.



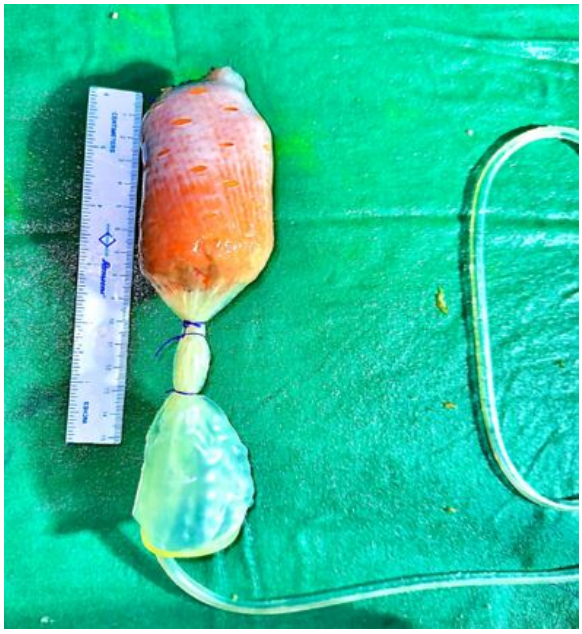


Figure 3: about 9 cm of neovagina created. Mould was removed on 8th post-operative day.

As the patient was keenly interested in sexual activity, with the patient's counselling and consent, McIndoe Vaginoplasty was performed. The blind pouch was dissected, creating a canal of approximately 9cm in length. A mould of about 9cm was prepared with autoclaved sponges and covered with a condom. A split thickness skin graft harvested from the anteromedial aspect of the thigh was lined into the neovaginal space using a mould. Postoperatively patient was kept on strict bed rest on a water bed, immobilised. At most care was taken to avoid any activity which increases intraabdominal pressure, viz, coughing or straining. She was catheterised for 10 days and was on a liquid diet for 7 days. On the 6th post-operative day, about 70-80% graft acceptance was achieved, and a new mould was placed into the neovagina. On 8th post operative day the mould was removed and a vaginal canal of ~9cm was achieved. On discharge patient was advised self dilatation with vaginal dilators and abstainence from intercourse till 6 months. On follow-up visits patient showed satisfaction. The patient started sexual activity after 6 months with satisfaction.

Discussion

Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome occurs due to defective Mullerian duct fusion during embryonic development. [2] Type I MRKH syndrome is characterized by variable underdevelopment of vagina and uterus. Type II MRKH also incorporates extragenital / extramüllerian malformations, which include vertebral, cardiac, urologic (upper tract), and otologic anomalies.[3] MRKH syndrome usually remains undetected until patient presents with primary amenorrhea despite normal female sexual development. Differentials include XY disorders of sexual development, Androgen insensitivity syndrome, imperforate hymen, congenital adrenal hyperplasia, etc.

The diagnosis is by involves a combination of physical examination, pelvic imaging (ultrasound or MRI), and sometimes chromosomal analysis.

Patients can benefit by creating a neovagina by surgical or non-surgical methods. Non surgical methods mainly include Frank technique using vaginal dilators (78% success rate) [4] Surgical methods include McIndoe vaginoplasty, Williams vaginoplasty, Intestinal neovagina, Vecchietti technique etc.

Infertility patients can undergo assisted reproduction techniques, and uterine transplantation has also been reported. MRKH patients are at a great risk of depression and anxiety problems after artificial vaginoplasty. Early psychological intervention can alleviate these symptoms.[5]

Conclusion

Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome occurs due to defective Mullerian duct fusion during embryonic development with variable involvement with only uterus and vagina and sometimes extending to other systems like skeletal, auditory or cardiac. Primary amenorrhea is the commonest presentation, with definitive diagnosis by physical examination and ultrasound with or without MRI. Treatment aims at the creation of a neovagina for sexual well-being, whereas psychological counselling plays a crucial role as far as postoperative recovery and starting sexual activity are concerned. A couple needs counselling for fertility options, as legal adoption, gestational surrogacy (GS), and uterine transplantation (UTx) have become available to these patients.[6]

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