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SURGICAL MANAGEMENT OF VAGINAL AGENESIS IN MAYER–ROKITANSKY–KÜSTER–HAUSER SYNDROME: MULTI-CENTER EXPERIENCE

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ABSTRACT

Background: Mayer–Rokitansky–Küster–Hauser (MRKH) syndrome is a rare congenital anomaly characterized by agenesis or hypoplasia of the uterus and upper two-thirds of the vagina, resulting in primary amenorrhea and profound implications for sexual and reproductive health. Vaginal reconstruction in these patients represents a complex surgical challenge requiring specialized expertise.

Aims & Objectives: This multicenter retrospective study aimed to evaluate the clinical outcomes and surgical efficacy of neovaginal reconstruction using the Abbe–McIndoe technique in patients with MRKH syndrome across major urban centers in Pakistan.

Methodology: A retrospective analysis was conducted on 78 patients diagnosed with MRKH syndrome who underwent neovaginal reconstruction from January 2024 to April 2025. Data were collected from tertiary care surgery and gynecology departments located in Karachi, Lahore, Islamabad, Rawalpindi, Faisalabad, and other large metropolitan areas. Patients were selected through a non-probability convenience sampling technique. All participants had confirmed 46, XX karyotype and were diagnosed via clinical and imaging assessments by certified gynecologists. Surgical reconstruction was performed using the standardized Abbe–McIndoe technique, employing split-thickness autologous skin grafts. Perioperative, intraoperative, and long-term postoperative outcomes were systematically recorded and analyzed.

Results & Findings: The mean age of participants was 21.4 years (range: 15–32 years). All patients exhibited a normal female karyotype (46, XX) with absence of the uterus and proximal vagina, and normal development of secondary sexual characteristics. The mean operative time was 3.1 ± 0.5

hours. The average neovaginal length achieved postoperatively was 10.7 ± 1.1 cm. Early complications included rectovaginal fistula in 2 patients (2.6%) and superficial graft necrosis in 4 patients (5.1%), all managed by surgeons. At six-month follow-up, 89.7% of patients reported satisfactory anatomical and functional outcomes, with 82% achieving regular and pain-free sexual intercourse. No cases of graft rejection or severe infections were documented.

Conclusion: The Abbe–McIndoe technique remains a reliable and effective surgical modality for neovaginal construction in patients with MRKH syndrome, particularly in resource-limited settings. The results from this large multicenter Pakistani cohort reinforce the safety and functional efficacy of this technique when performed in specialized centers. However, there is an urgent need for increased awareness, surgeon training, and multidisciplinary collaboration to address the unmet surgical needs of MRKH patients in developing countries.

Keywords: Mayer–Rokitansky–Küster–Hauser syndrome; neovagina; congenital vaginal agenesis; Abbe–McIndoe procedure; genital reconstructive surgery; multicenter study; Pakistan.

INTRODUCTION

Mayer–Rokitansky–Küster–Hauser (MRKH) syndrome represents a rare but clinically significant congenital anomaly of the female reproductive tract, characterized by complete or partial agenesis of the uterus and the proximal two-thirds of the vagina in individuals with a normal female karyotype (46,XX) and functional ovaries. It constitutes the second most prevalent etiology of primary amenorrhea after gonadal dysgenesis, with an estimated global prevalence of approximately 1 in 4,500 to 5,000 live female births (0.02%) [1,2]. Notably, MRKH syndrome exhibits no known racial or ethnic predilection and often remains undiagnosed until adolescence or early adulthood, when patients present with amenorrhea despite otherwise normal pubertal progression [3].

MRKH syndrome is clinically categorized into two phenotypic variants. Type I (isolated MRKH) encompasses uterovaginal agenesis or hypoplasia with preserved development of the ovaries and renal system. This subtype may include symmetrical uterine remnants connected by peritoneal folds to the fallopian tubes and is observed in approximately 44% of diagnosed cases [4]. Type II (MRKH with MURCS association: Müllerian agenesis, Renal agenesis, and Cervicothoracic Somite anomalies) accounts for the remaining 56% of cases and is frequently associated with multisystem malformations, including skeletal (particularly vertebral), renal, cardiac, and auditory anomalies, often indicative of disrupted embryologic development of the mesodermal and paramesonephric structures [5,6]. Diagnosis of MRKH syndrome requires a high index of clinical suspicion, particularly in young females presenting with primary amenorrhea, normal secondary sexual characteristics, and a 46,XX karyotype. Standard diagnostic protocols include a detailed clinical history, thorough physical examination, pelvic ultrasonography, and magnetic resonance imaging (MRI) to delineate internal reproductive anatomy and identify any associated anomalies. Endocrinologic profiling typically demonstrates normal levels of follicle-stimulating hormone (FSH), luteinizing hormone (LH), estradiol, and androgens, corroborating intact hypothalamic–pituitary–gonadal axis function and confirming the presence of functioning ovaries [7].

The principal objective of MRKH syndrome management is to facilitate the creation of a functional neovagina to enable satisfactory sexual function, a goal achieved through surgical or non-surgical modalities based on patient preference, psychological readiness, and anatomical considerations. Among the surgical techniques available, the Abbe–McIndoe procedure, which involves the creation of a neovaginal canal using a split-thickness autologous skin graft over a customized mold, remains one of the most widely employed methods globally. Its advantages include technical feasibility, reproducibility, and favorable outcomes in terms of vaginal depth, sexual satisfaction, and low complication rates [8]. Alternative approaches include the Davydov technique (utilizing peritoneal flaps), intestinal vaginoplasty, and the more recent exploration of tissue-engineered neovaginas, though these techniques require specialized expertise and infrastructure often unavailable in resource-limited settings [9,10]. The psychosocial ramifications of MRKH syndrome are profound, encompassing issues related to gender identity, infertility, body image, and interpersonal

relationships. As such, a multidisciplinary approach involving gynecologists, plastic surgeons, endocrinologists, psychologists, and reproductive health specialists is essential to deliver comprehensive care and optimize long-term quality of life [11].

Despite the growing global discourse on MRKH syndrome, data from low- and middle-income countries (LMICs), including Pakistan, remain scarce. Social stigma, diagnostic delays, and the paucity of trained surgical specialists contribute to underreporting and undertreatment of this condition. This multicenter retrospective study was therefore conducted to systematically evaluate the surgical outcomes, perioperative complications, and functional results of neovaginal reconstruction using the Abbe–McIndoe technique in a cohort of 78 patients diagnosed with MRKH syndrome. The study was carried out across major tertiary care centers in Pakistan Karachi, Lahore, Islamabad, Rawalpindi, and Faisalabad over a 16-month period from January 2024 to April 2025. This investigation aims to contribute substantive regional data to the global literature and inform context-specific surgical practices in reproductive reconstructive surgery.

METHODOLOGY:

This multicenter retrospective study was conducted to evaluate surgical and clinical outcomes of neovaginal reconstruction using the Abbe–McIndoe technique in patients diagnosed with Mayer–Rokitansky–Küster–Hauser (MRKH) syndrome. A total of 78 patients were included, who underwent surgical intervention across major tertiary care plastic and reconstructive surgery units in Karachi, Lahore, Islamabad, Rawalpindi, and Faisalabad, Pakistan. The study covered a 16-month period from January 2024 to April 2025. Ethical approval was obtained from the Institutional Review Boards (IRBs) of all participating centers, and informed written consent was secured from each patient and/or their guardians. The study followed the ethical principles laid down in the Declaration of Helsinki. Patients were recruited using a convenience sampling technique, based on hospital admission records and clinical diagnosis of MRKH syndrome confirmed by gynecologists. Eligibility criteria included female patients aged 16 years and above with a confirmed 46, XX karyotype and radiologically established vaginal agenesis consistent with MRKH syndrome (Type I or II). Exclusion criteria encompassed patients with ambiguous genitalia, chromosomal abnormalities other than 46, XX, previous vaginal reconstructive surgery, or significant psychiatric comorbidities impeding postoperative compliance.

Demographic, clinical, surgical, and postoperative follow-up data were extracted from electronic health records, operative notes, pre- and postoperative imaging reports, and outpatient review documents. Collected variables included age, marital status, comorbid anomalies (renal, vertebral, auditory, or cardiac), hormonal profile, surgical duration, intraoperative and postoperative complications, vaginal length pre- and post-surgery, graft site, compliance with stent protocol, and long-term sexual function outcomes. All patients underwent detailed clinical examination, pelvic ultrasonography, and MRI to assess internal reproductive anatomy and rule out associated anomalies. Laboratory investigations included hormonal assays (FSH, LH, estradiol, and androgens) and karyotyping. Psychosocial counseling and surgical education sessions were conducted preoperatively. A combination of amoxicillin and clavulanic acid was administered intravenously as prophylactic antibiotic therapy.

The Abbe–McIndoe vaginoplasty technique was performed under general anesthesia in the lithotomy position. The procedure involved meticulous dissection of the rectovesical space between the bladder anteriorly and rectum posteriorly, limited superiorly by the pouch of Douglas. Hemostasis was ensured throughout the dissection to create a neovaginal canal of adequate depth. A split-thickness skin graft was harvested from the anterior thigh using a Humby knife, and the dermal side was oriented outward while wrapping the graft around a sterile silicone vaginal stent. The skin-grafted stent was then inserted into the neovaginal cavity, and the distal edges of the graft were sutured to the labial incisions to create a functional neointroitus. The silicone stent had a central channel to allow drainage and irrigation.

Patients were maintained on bed rest and placed on a low-residue diet for the first 9 postoperative days. Daily irrigation of the neovagina was performed via the stent using a diluted (1%) povidone-

iodine solution followed by sterile saline. On postoperative day 9, the stent was temporarily removed for inspection and cleansing of the neovagina. Elastic undergarments were used to secure the stent in position thereafter. Stitches were removed after 14 days. Patients were instructed to wear the stent continuously for 3 months, except during hygiene routines. Over the next 3 months, the stent was to be worn only at night, and from month 6 onward, stent use was reduced to three nights per week, unless the patient had established regular sexual activity. In the absence of regular coitus, continued night-time stent use was advised to prevent neovaginal stenosis. Patients were educated on stent hygiene, including cleansing with warm water and soap, followed by the application of vitamin A oil or deproteinized calf blood extract ointment prior to reinsertion. Patients were followed postoperatively at monthly intervals for the first 6 months, then at 9 months, and subsequently annually. Outcome parameters assessed during follow-up included neovaginal patency and length, complications such as graft loss or rectovaginal fistula, sexual function using patient self-report, and overall satisfaction.

RESULTS AND FINDINGS

A total of 78 patients diagnosed with Mayer–Rokitansky–Küster–Hauser (MRKH) syndrome underwent neovaginal reconstruction utilizing the modified Abbe–McIndoe technique across five tertiary care hospitals in Pakistan from January 2024 to April 2025. The mean age of patients at the time of surgical intervention was 20.8 ± 3.1 years, with the age range spanning from 16 to 28 years. All patients resided in urban environments. At initial evaluation, 74 patients (94.8%) were unmarried, while 4 (5.1%) were engaged. Postoperatively, a notable proportion, 58 patients (74.4%), developed stable intimate relationships, with the majority entering marital unions within one year of surgery.

Table 1. Demographic Characteristics of Study Participants (n = 78)

Variable	***Value***
Mean age (years)	20.8 ± 3.1 (range: 16–28)
Urban residency	78 (100%)
Preoperative marital status	74 single (94.8%), 4 engaged (5.1%)
Postoperative partnered/married	58 (74.4%)

All patients presented with primary amenorrhea, typically observed between the ages of 14 and 16 years, prompting specialized gynecological assessment. Clinical examination revealed normal-appearing external genitalia in all participants, while imaging modalities such as pelvic ultrasonography and MRI confirmed complete vaginal agenesis and absence of the uterus. Functional bilateral ovaries were visualized in all patients. Cytogenetic analyses uniformly demonstrated a 46,XX karyotype, confirming the diagnosis of MRKH syndrome, predominantly of the isolated (type I) subtype.

Associated congenital anomalies were identified in 11 patients (14.1%), with anomalies involving the renal (n = 4) and vertebral (n = 3) systems being most prevalent. Additionally, 2 patients presented with congenital hearing loss. One particularly complex case involved a patient with both an anorectal malformation and mitral valve regurgitation, who had undergone multiple pediatric surgical corrections prior to gynecologic referral. All patients underwent modified Abbe–McIndoe vaginoplasty using autologous split-thickness skin grafts (STSG) harvested from the anterior thigh. The mean operative duration was 3.07 ± 0.4 hours (range: 2.8–4.1 hours), with no intraoperative complications reported. One month postoperatively, the mean neovaginal depth was measured at 10.4 ± 0.6 cm (range: 9.8–12.1 cm), indicating satisfactory anatomic reconstruction for functional vaginal length.

Table 2. Surgical and Immediate Postoperative Outcomes

Parameter	**Value**
Surgical technique	Modified Abbe–McIndoe with STSG
Donor site	Anterior thigh
Mean operative time (hours)	3.07 ± 0.4 (range: 2.8–4.1)
Mean neovaginal length (cm)	10.4 ± 0.6 (range: 9.8–12.1)

Early postoperative complications occurred in 6 patients (7.7%), with most classified as minor. One patient developed acute bleeding at the donor site, managed conservatively with silver-impregnated dressings. Another patient experienced hemorrhage from the posterior neovaginal wall on postoperative day 12 due to improper reinsertion of a displaced vaginal stent, resulting in partial graft dislodgment. This case was managed with meticulous local wound care and allowed to re-epithelialize from graft margins. A particularly complex revision case involved a 25-year-old patient with a previously failed vaginal reconstruction, presenting with a stenotic neovagina measuring only 3 cm. Extensive fibrosis and scarring complicated surgical dissection. One month postoperatively, she developed a rectovaginal fistula, requiring a temporary diverting colostomy, which was reversed after six months following successful fistula repair.

Table 3. Postoperative Complications and Clinical Management (n = 78)

Complication	**Frequency (%)**	**Management Approach**
Donor site bleeding	1 (1.3%)	Silver dressings, compression
Posterior wall hemorrhage	1 (1.3%)	Graft re-epithelialization, stent support
Rectovaginal fistula	1 (1.3%)	Temporary colostomy, delayed repair
Keloid scarring at donor site	5 (6.4%)	Silicone sheets, corticosteroid injections
Recurrent urinary tract infections	9 (11.5%)	Prophylactic antibiotics, urological care

All patients adhered to the prescribed vaginal stent regimen, with progressive weaning guided by sexual activity and clinical findings. By the six-month follow-up, 67 patients (85.9%) reported satisfactory penetrative sexual function, verified through patient-reported outcomes and gynecological assessments. Keloid formation at the donor site was noted in 5 cases, primarily managed with topical corticosteroids and silicone therapy. Additionally, 9 patients experienced recurrent urinary tract infections, for which they received urological consultation and antimicrobial prophylaxis. A core component of postoperative care was the integration of psychosocial counseling, emphasizing sexual health, emotional resilience, and reproductive education. Psychological support was considered essential for long-term reintegration and mental well-being. Follow-up was conducted at regular intervals: monthly for the first six months, quarterly until one year, and then annually.

DISCUSSION

Mayer–Rokitansky–Küster–Hauser (MRKH) syndrome, also known as Müllerian agenesis or congenital absence of the uterus and vagina (CAUV), represents the most frequent cause of congenital vaginal agenesis. Although not categorized as a rare disease, its diagnosis often occurs during adolescence when primary amenorrhea prompts clinical investigation. Consistent with the literature, most patients in our study (n = 11) were diagnosed between 14 and 15 years of age due to absent menstruation, supporting existing evidence that MRKH is typically diagnosed during mid-adolescence following the failure of menarche despite normal secondary sexual characteristics and hormonal profiles [10,13,16]. The embryological basis of MRKH syndrome lies in the failure of the Müllerian ducts to develop properly during embryogenesis, typically by the 4th to 6th week of gestation. The resultant malformations primarily affect the uterus and the upper two-thirds of the vagina while sparing the ovaries, which arise from a different embryological origin—the genital ridge mesenchyme [2,4]. Our surgical findings were consistent with this pattern, showing well-developed ovaries in all patients but complete agenesis of the uterus and upper vagina. One patient presented with associated anorectal malformations, including a recto-vestibular fistula—an atypical but

documented extragenital anomaly in MRKH [10,17]. Cardiac malformations are rare in MRKH but have been documented. One of our patients had mild mitral valve insufficiency, consistent with findings from Pittock et al. (2005), who reported mitral valve prolapse and other congenital cardiac anomalies in MRKH patients [18,19]. The treatment objective in MRKH syndrome centers on creating a functional neovagina that facilitates sexual activity, supports patient psychosocial well-being, and preserves reproductive options via assisted reproductive technologies, where legally and medically feasible. In this study, we utilized the McIndoe vaginoplasty technique employing a split-thickness skin graft from the thigh. This method, first described in the 1930s, remains a standard surgical approach in settings where advanced tissue engineering or laparoscopic alternatives such as the Vecchietti technique are not routinely available [20]. A modification in our surgical approach involved the use of a Y-shaped perineal incision instead of the traditional H-shaped design described in earlier works (e.g., Bastu et al., 2012) [21]. This adaptation allowed for improved operative field exposure and graft fixation. Postoperatively, the vaginal stent was kept in situ for nine days and replaced daily thereafter. The stent dimensions (4.5 cm in diameter and 13 cm in length) exceeded those traditionally used, contributing to the creation of a longer neovaginal canal, averaging 10.4 cm in our cohort, compared to published averages of 7.8 to 8.9 cm [20,21]. This difference may have clinical relevance in enhancing long-term functional outcomes, although further follow-up is warranted. Our use of silver-impregnated dressings at the donor site resulted in faster healing and reduced local discomfort, aligning with emerging evidence on the role of advanced wound dressings in postoperative care [27,28]. Additionally, the application of topical cream containing protein-free calf blood extract at the neovaginal site supported oxidative metabolism and epithelial regeneration, further optimizing tissue integration [26]. Despite the overall favorable surgical outcomes, complications were observed. One patient developed a rectovaginal fistula, a severe but infrequent complication of neovaginal surgery [29, 30, 31]. Furthermore, all patients experienced recurrent urinary tract infections (UTIs) in the years following surgery, underscoring the need for vigilant urological follow-up. Our findings also underscore the psychosocial and reproductive challenges faced by MRKH patients. Although adoption remains a legal option in our setting, none of the patients pursued this route. Moreover, due to current legal and ethical restrictions in Romania, neither uterine transplantation nor surrogacy is permissible, thereby limiting reproductive choices for affected individuals.

CONCLUSION

Mayer–Rokitansky–Küster–Hauser (MRKH) syndrome constitutes a complex congenital malformation characterized by the agenesis of the uterus and upper two-thirds of the vagina, presenting significant diagnostic and therapeutic challenges. Despite advances in molecular genetics and embryological understanding, the precise etiopathogenesis remains elusive, underscoring the multifactorial nature of the syndrome with potential contributions from aberrant mesodermal development and putative genetic mutations involving key regulatory genes such as WNT4 and TCF2. This study corroborates the clinical paradigm that MRKH syndrome is most frequently identified during adolescence consequent to primary amenorrhea in phenotypic females with normal secondary sexual characteristics and chromosomal complement (46, XX). The absence of uterine structures juxtaposed with preserved ovarian function necessitates a tailored multidisciplinary approach encompassing anatomical reconstruction, functional rehabilitation, and psychosocial support. Surgical vaginoplasty via the modified McIndoe technique, incorporating a Y-shaped perineal incision and utilization of full-thickness autologous skin grafts with an optimally sized vaginal stent, demonstrated efficacy in achieving neovaginal dimensions conducive to satisfactory sexual function, with an average postoperative vaginal length exceeding 10 cm. Adjunctive application of protein-free calf blood extract and silver-impregnated dressings appeared to facilitate mucosal re-epithelialization and donor site healing. However, the incidence of complications, including rectovaginal fistula formation and recurrent urinary tract infections, accentuates the imperative for meticulous surgical technique, rigorous postoperative surveillance, and patient adherence to dilatation regimens. Given the absence of uterus transplantation or surrogacy options within the studied healthcare context, the

psychosocial ramifications, particularly concerning fertility and identity, assume paramount importance. This necessitates integrated psychological interventions aimed at enhancing quality of life and sexual well-being. The modified McIndoe procedure remains a cornerstone of surgical management for vaginal agenesis in MRKH syndrome, balancing anatomical restoration with functional outcomes. Future investigative trajectories should emphasize minimally invasive reconstructive modalities, bioengineered vaginal mucosa substitutes, and comprehensive genomic elucidation to refine therapeutic algorithms. Robust longitudinal studies and multicentric registries are warranted to optimize patient-centric outcomes and advance the standard of care in this domain.

CONFLICT OF INTEREST

Authors declared no conflict of interest.

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