

Island Singapore Flap Vaginoplasty of Two Adult Cases of Mayer–Rokitansky–Küster–Hauser Syndrome Type I

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Summary

Mayer–Rokitansky–Küster–Hauser Syndrome Type I is an emotionally distressful rare condition that limits normal sexual functioning and relationships. First described in 1989 by Wee and Joseph, the neurovascular island pudendal thigh flap (Singapore flap) has been variously utilized and modified for use in vaginal reconstruction. We report two adult cases of vaginal agenesis who underwent vaginal reconstruction using the Singapore flap. Both patients reported improved sexual lifestyle.

Key words: Island Singapore flap, Vaginoplasty, Mayer–Rokitansky–Küster–Hauser syndrome Type I

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Introduction

Vaginal agenesis can present as isolated Müllerian aplasia or Mayer–Rokitansky–Küster–Hauser syndrome type I with an incidence of 1 per 4,500–5,000 females (1). This embryologic underdevelopment of the Müllerian duct results in isolated absence of the uterus and the proximal two-thirds of the vagina (2). The normal hormonal profile and normal secondary sexual development found in such patients is due to different embryologic origin of the ovaries. Despite the obvious primary infertility it confers, vaginal agenesis has significant psychological impact on the patient due to inability to experience a satisfactory sexual lifestyle. The Singapore flap vaginoplasty provides a sensate and pliable (thin and elastic) neo-vagina that enables sexual functioning (3). We present two adult cases of Mayer–Rokitansky–Küster–Hauser syndrome type I that underwent vaginoplasty using the island Singapore flap: the first such report in Kenya.

Case Presentation

We present two cases; a 27-year-old female who presented to us accompanied by her sister in November 2016, and a 30-year-old female accompanied by her spouse presenting in September 2017. The patients had similar clinical presentation. Chief complaints were primary amenorrhea and inability to experience sexual intercourse due to shortened vagina. Physical examination revealed normal secondary sexual characteristics, blind ending distal vaginal canal of 4 cm and an absent cervix, in both patients. An

absent uterus with normal bilateral ovaries was reported on ultrasonography. Hormonal profile was normal (luteinizing hormone, follicle-stimulating hormone, prolactin, estradiol and progesterone levels were all within normal ranges). Chromosomal analysis was not performed though no associated congenital anomalies were present.

A standard perioperative protocol was adhered to: preoperative psychological counselling, surgical technique and postoperative management. Surgical marking was done with assistance of a vascular Doppler ultrasound.

The recto-vesical pouch was bluntly developed and a modified island Singapore flap vaginoplasty, based on perineal vessels (branches of internal pudendal vessels) and perineal nerve (branch of pudendal nerve), was performed with incremental neo-vaginal canal length of 10 cm.

The flap was inset using Polyglactin 910 and a vaginal pack placed. The pack was removed after 72 hours; thereafter 6 hourly warm saline sitz baths were prescribed. Drains were removed by the third postoperative day. After discharge, follow-up reviews were done at 3-month intervals for 1 year. The first patient had an uneventful recovery and she was discharged 12 days post-surgery. Self-dilatation was initiated at 3 weeks post-surgery and continued for 3 months. Thereafter, sexual intercourse was encouraged.

The patient reported ability to engage in vaginal sexual intercourse at the 6-month review, and during her annual review further asserted that she was happier with sexual lifestyle.



markings of internal pudendal artery (bilaterally) exiting the pudendal canal after vascular doppler



design of bilateral singapore flaps

Figure 1: Preoperative markings



subfascial elevation of flaps



neurovascular bundle of flap: internal pudendal vessels & pudendal nerve



island flaps tunneled and posterior wall sutured

Figure 2: Intraoperative image 1



cylindrical-fashioned neo-vagina: length of 10 cm



donor site wounds



flaps inset with vaginal pack in-sit & primary closure of donor sites

Figure 3: Intraoperative image 1



1 month post-surgery



3 months post-surgery: donor site well healed

Figure 4: Postoperative images

The second patient developed recipient site surgical site infection that resulted in neo-vaginal stenosis. Intralesional steroid (Triamcinolone 40 mg monthly for 2 months) injection was given to soften the introitus of the neo-vaginal canal. Further self-dilatation coupled with monthly dilatation under anesthesia for 5 months was performed until the patient was comfortable to attempt vaginal sexual intercourse. The donor site healed uneventfully (Figure 4).

She reported ability to engage in sexual intercourse during her 6-month review.

Discussion

The presentation of both of patients was consistent with Mayer-Rokitansky-Küster-Hauser syndrome type I. The cause is genetic with autosomal dominant inheritance, incomplete penetrance and variable expressivity (4). A multifactorial pathogenesis has also been postulated (5). Because of the significant cost implications, the patients were unwilling to undergo karyotyping. First described by Wee and Joseph for vaginal reconstruction after pelvic exenteration, the Singapore flap has been variously used for

vaginoplasty in vaginal agenesis (6-8). This is the first report in our set up, Kenya. Use of a vascular Doppler to map the vessels is a critical step that guides the design of a viable flap. Advantages of the Singapore flap include minimal donor site morbidity, pliable sensate flap, having a reliable blood supply and single-stage procedure (9, 10). Furthermore, sexual intercourse is initiated earlier compared with other techniques (11). Initiation of sexual intercourse facilitates molding and

remodeling of the neo-vaginal canal, limiting stenosis. In contrast to other reports of not using dilators, we used the vaginal pack in the early postoperative period and later on dilators to prevent neo-vaginal canal stenosis (12). This 'cylindrical designed' wound definitely contracts during healing. By 3 months, remodeling is well underway and introduction of sexual intercourse likely assists in this process. If stenosis develops, we recommend combining dilation under anesthesia with intralesional steroid injection to fibrotic regions. Sexual satisfaction was assessed as: ability to engage in sexual intercourse and the patient's emotional expression regarding their sexual lifestyle, to which both patients reported to be positive.

Our study is limited by the low number of cases; not using a validated scale for assessing sexual satisfaction (female sexual function index–FSFI, female sexual distress scale–revised–FSDS-R, female genital self-image scale–FGSIS, sexual satisfaction scale for women–SSS-W)); not performing karyotyping; and not providing radiological images (13-16).

Conclusion

The island Singapore flap is a good option for vaginoplasty in vaginal agenesis.

Conflict of interest

Dr Wamalwa joined the editorial team while this paper was under review. He was excused from the final manuscript meeting where the final decision was made on publication.

Ethical approval

The patient gave informed consent to publish the anonymized images and further approval was granted by the Kenyatta National Hospital/University of Nairobi Ethics Research Committee (KNH/UoN-ERC) – Ref. no. KNH-ERC/PUB/2.

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