Vesico-vaginal Fistula in a Patient with Meyer-Rokitansky-Küster-Hauser syndrome

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Summary
This was a case of Gishiri cut in a patient with Meyer-Rokitansky-Kuster-Hauser syndrome resulting in a vesico-vaginal fistula and urethral loss. This followed an attempt to enlarge and lengthen the vagina to enhance penile penetration. Few cases of MRKH syndrome presenting with complications after an attempt at treatment by traditional birth attendants have been reported. This report is particularly of essence as most urinary fistulae in Nigeria are obstetric fistulae following prolonged obstructed labour.

Key words: Primary amenorrhoea, Meyer-Rokitansky-Kuster-Hauser syndrome, urinary fistula

Meyer-Rokitansky-Kuster-Hauser (MRKH) syndrome or vaginal agenesis occurs in 1:4000 female births and usually presents as primary amenorrhoea[1]. It may, occasionally, present with difficulty during coitus due to the short length of the vagina. This communication is made to draw attention to vesico-vaginal fistula in a patient with MRKH syndrome whose short vagina was treated with Gishiri cut by a traditional birth attendant.

Mrs. T.A was a married 25 year old nullipara who presented with failure to achieve menarche, inadequate penile penetration at coitus and involuntary leakage of urine of 3 years duration. She was divorced by her first husband because of difficult penile penetration during coitus and primary infertility but the problem remained with the new husband. This made her present to a traditional birth attendant who gave her a Gishiri cut as a form of treatment. This led to involuntary voiding of urine.

Her weight and height was 46kg and 151cm respectively. Breast, pubic hair and axillary hair developments were respectively Tanner stage 4, 4 and 3. Examination of the vulva showed a moist well developed vulva with no urethral orifice. There was ammoniacal dermatitis of the proximal thigh bilaterally. The vagina ended blindly about 2cm from the vestibule and had a defect of about 2cm in diameter at its superior anterior part. Rectal examination was essentially normal.

Abdomino-pelvic ultrasound scan reported absence of the uterus with an absent right kidney but the left kidney was located in the pelvis, proximal to the urinary bladder as in figure 1. There was a rudimentary right ovary. An Intravenous urogram after the repair of the fistula as in figure 2 showed a non-functioning right kidney and a left kidney located in the pelvis with moderate dilatation of the calyces. There was also a left short, kinked ureter ectopically inserted on the bladder neck. Her hormonal profile was within the reference range.

Surgical Repair of fistula

In exaggerated lithotomy position, under spinal anaesthesia, patient was cleaned and draped. The vaginal was too small to admit a speculum. Using scalpel, a circumferential incision was made around the edges of the

fistula. The vaginal wall was dissected and separated from the bladder wall with a pair of curved scissors. The bladder wall was repaired, with the edges inverted, in the longitudinal axis over a size 12 Foley catheter starting from the proximal point with interrupted Vicryl suture, size 2/0 leaving a small opening approximately 0.5cm just below the clitoris. The vaginal wall was also repaired with the same suture using interrupted stitches in the same axis leaving an opening of 0.5cm at the same point as on the bladder wall. The edges of the two openings around the catheter were sutured together using interrupted Vicryl suture size 2/0, thereby creating a neourethra and orifice.

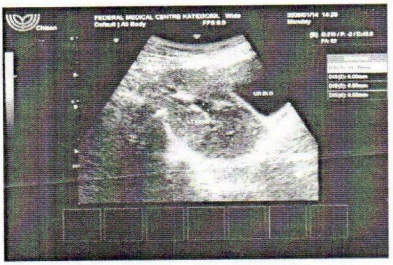

Fig. 1: Pelvic ultrasound scan showing the left pelvic kidney proximal to the urinary bladder

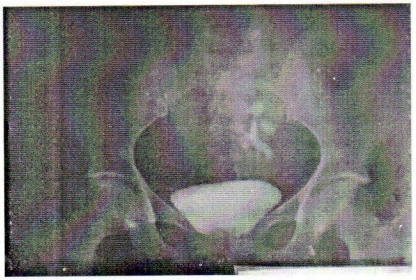

Fig 2: Urogram showing pelvic kidney with dilated calyces

To the end of the catheter was connected an infusion set which drained into a bed pan. Intravenous infusion was administered to ensure adequate urinary output in the first 48 hours and she had liberal oral fluid intake thereafter. The catheter was removed after bladder training on the 21st post operative day after successful bladder training which began on the 14th post operative day. She was continent of urine post surgery and was subsequently discharged home for follow up at the Gynaecological clinic. Six months after the surgery, improvised dilators (plastic syringes) were used to enlarge the vagina. This was gently introduced into the vagina. She had the dilator applied for 20 minutes daily initially, then twice daily later.

Women with MRKH syndrome have normal development of secondary sexual characteristics because ovarian function is unaffected. They also have normal growth. However, they have a short vagina and an absent uterus. What was unique in this case was the short vagina and attendant difficulty with sexual intercourse. This led to her divorce and also made her seek traditional Gishiri cut in the new marriage which caused her urinary incontinence. Short vagina in patients with MRKH syndrome has been reported to cause bruises during coitus. This caused Human Immunodeficiency Virus (HIV) infection in a woman with the syndrome[2]. This has long term implications unlike the urinary fistula in our patient which was successfully repaired. Gishiri cut is usually made by traditional birth attendants. It is a documented practice during obstructed labour, occurring primarily, in Northern Nigeria. A knife, razor blade or a piece of broken glass is used to make a series of random cuts on the vagina in an attempt to relieve the obstruction and make way for the baby [3]. This practice can directly injure the bladder or urethra and may account for as many as 15% of fistula cases in Northern Nigeria [3].

We observed that the urethra was likely to have been incised and the incision extended into the bladder. This resulted in posterior urethral loss and an absent orifice. Following the repair as described above, the re-establishment of continence after repair in this case was indicative of a successful anatomic and physiologic restoration of the urethra. This is of note as the normal female urethra is about 3cm long and there may be a shorter urethra in MRKH syndrome due to associated genito-urinary anomaly. This is because forty per cent of these patients have renal anomalies, 15% of which are major anomalies including absence of the kidney[4]. Intravenous urogram (IVU) gives better demonstration of urinary abnormalities [5] and is more sensitive than ultrasonography in the diagnosis of urinary anomalies [4]. IVU was done in this case report as it aids the diagnosis of the syndrome. While ultrasound scan suggested an absent right kidney, the urogram showed that it was present but non-functioning.

When patients with MRKH syndrome present, usually, with primary amenorrhoea, making a diagnosis may be a challenge. This challenge extends to the management. Therefore, these women require a lot of reassurance and support. Management challenges include sexual satisfaction and future fertility. For those already married, sustenance of their marriage is of concern. They require an adequate length of vagina for satisfactory coitus to reduce marital disharmony. The vagina can be enlarged mechanically with dilators or surgically by creation of neovagina. Mrs. AT was offered mechanical gradual dilatation by dilators using Frank's method. While this was being done, we were mindful of the successful repair of her fistula, hence the application of pressure away from the point of repair and the initial daily application for 20 minutes, 6 months post surgery. The pressure of the dilator

was applied on the inferior posterior part of the vagina.

She was also counseled on the duration of regular gradual dilatation of the vagina required to achieve a vaginal length for good sexual activity. This was to enhance her cooperation and compliance with treatment. We chose vaginal dilatation as good follow up on sexual satisfaction was not reported after surgical creation of neovagina[6]. The fertility choices for these patients need to be brought to the fore. These include surrogacy and adoption. Surrogacy involves oocytes collection from the patient and *in-vitro* fertilisation using semen from her husband. This is followed by a transfer of the fertilised zygote into a surrogate carrier. This offers them genetic offspring. Surrogate pregnancy has been reported not to cause inheritance in a dominant fashion in analysis of 34 surrogacy live births[7].

However, she cannot afford the high cost of *in-vitro* fertilisation and surrogacy is alien to our culture. Therefore, the way out of the fertility challenge may be traditional adoption, in which case, a relative gives her a child to cater for. A differential diagnosis which needs consideration is the androgen insensitivity syndrome. Patients with this condition have genetic sex XY and are usually very tall and attractive but with absent or at best scanty pubic and axillary hair[1]. Although we did not do karyotyping for the patient, patients with androgen insensitivity syndrome do not have renal malformations as was in this case. She also had normal hormone profile which is not found in patients with androgen insensitivity. This case shows the wide gap in the knowledge of the populace on ambiguity of the genital tract. The need for community enlightenment on the subject in order to seek help at the appropriate place cannot be overemphasised.

References

1. Speroff L., Glass R. H., Kase N. G. Amenorrhea. Clinical Gynaecologic Endocrinology and Infertility 6th ed. Charles Mitchell. Linpincott Williams and Wilkins Ltd 1999; 421 – 485.
2. Kell P. D., Barton S. E., Edmonds D. K., Boag F. C. HIV infection in a patient with Meyer-Rokitansky- Kuster-Hauser syndrome. *J R Soc Med* 1992; **85:** 706–707.
3. The info project. Johns Hopkins Bloomberg School of Public Health Center for communication programs. Obstetric fistula. Ending the silence, Easing the suffering, Baltimore, Issue No 2, 2004.
4. Edmonds D. K. Primary Amnorrhoea. Dewhurst's Textbook of Obstetrics and Gynaecology for Postgraduates 6th ed. 1999. Ed. D. K. Edmonds Blackwell Sciences Ltd 1999; 34–41.
5. Rzymski P., Szpakowska-Rzymska I., el Yubi R., Wilczak M., Sajdak S. *et al.* Coexistence of female sexual organ malformation and urinary tract anomalies. *Ginekol Pol* 2001; **72:** 67–72.
6. Emilio I., Alessandro A., Giovanni A., Anton L. M., Arsenio S. Long-term results of sigmoid vaginoplasty in a consecutive series of 62 patients. *International urogynecology Journal* 2007; **18:** 1465–1469.
7. Batzer F. R., Corson S. L., Gocial B., Daly D. C., Go K. et al. Genetic offspring in patients with vaginal agenesis: specific medical and legal issues. *Am J Obstet Gynecol* 1992; **167:** 1288.