



Case report of pudendal thigh flap for vaginal atresia in Mayerrokitansky- Kuster hauser syndrome

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Abstract : Mullerian and wolffian ducts are the primordial for the internal reproductive system of female and male and coexist in embryo until genetic sex trigger differentiate of either testis and ovaries. The Mayer- rokitansky-kuster hauser syndrome affect atleast 1 out of 4500 woman . A 20 years old unmarried woman presented with primary amenorrhoea and normal development of secondary sexual characteristics. Diagnostic laparoscopy revealed uterus with two horns non communicating with both ovaries and fallopian tubes normal and vaginal atresia. Six months following this procedure she underwent neo vaginal creation with pudendal thigh fasciocutaneous flap for vaginal atresia.

Keyword :Primary Amenorrhoea, Mayer Rokitansky kuster Hauser Syndrome, Vaginal Atresia.

INTRODUCTION

Vaginal agenesis is a rare congenital condition often associated with Mayer- rokitanskykuster- hauser syndrome. Incidence 1 in 4500 female birth¹. It consists of primary amenorrhoea with normal female phenotype, genotype, normal ovarian function and endocrine status. The uterus is either completely absent or it is represented by two rudimentary horns. There are 2 subtypes of Mayer- rokitansky - kuster- hauser syndrome .The typical or type 1 form characterised by laparoscopic findings symmetrical muscular bands and normal fallopian tubes so called rokitansky sequence. Atypical form are type 2 shows in addition asymmetric hypoplasia of 1 or 2 buds with or without dysplasia of fallopian tubes and is often associated with other anomalies including mainly renal defects cervico thoracic hearing defects. The treatment of vaginal agenesis includes graduated dilators², split skin graft³, full thickness graft⁴, buccal mucosa graft⁵, colon interposition vaginoplasty, abdominal pull through technique, myocutaneous flap, vulvovaginoplasty using labia minora, infragluteal thigh and pudendal fasciocutaneous flap.

Case report:

A 20 years old unmarried woman admitted in our department with complaint of primary amenorrhoea. On general examination moderately built and nourished, height 158 cm, weight 56kg , breast tanner stage 3, axillary hair present ,

pubic hair tanner stage 4. CVS- s1s2 heard, RS-Bilateral air entry present, PER ABDOMEN -Soft. Local examination of external genitalia appears normal, vaginal orifice like a dimple ,per rectum normal

Past history

She was evaluated for this condition six months back, then she underwent diagnostic laparoscopy.

Intra operative findings –

Right side ---Rudimentary horn of hypoplastic uterus with fallopian tube and ovary.

Left side ---Rudimentary horn of hypoplastic uterus with fallopian tube and ovary normal. Both ovaries present at normal site.

INVESTIGATIONS:

- HEMOGLOBIN- 11.5 gms/dl

- PCV- 34%

- HORMONAL ASSAY: T3 - 2.96

- T4- 1.59

- TSH -1.08

- FSH- 5.9,

- LH- 5.52,

- SERUM PROLACTIN -7.72,

- KARYOTYPING : 46XX

USG ABDOMEN AND PELVIS:

Uterus not visualised , right ovary 2.2x1.2cm Left ovary 1x 0.9cm ,other organs normal.

MRI:

Shows absent uterus and no distinct vagina.

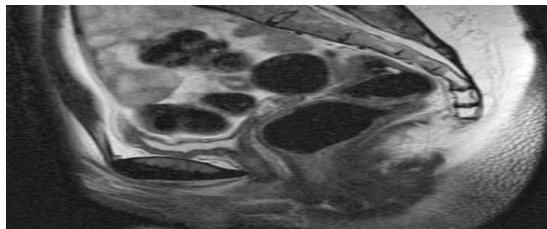


fig-1 MRI Shows Absent Uterus And No Distinct Vagina.

TREATMENT:

She underwent vaginal reconstruction surgery with pudendal thigh fasciocutaneous flap .

PROCEDURE:

Transverse incision made posterior to the urethral opening just behind the dimple, through hydrodissection a blind pouch created between the bladder and rectum of 8 cm. Bilateral pudendal flap marking(fig-3) done adjacent to labia majora with axis centered the inguinal creases and base near the perineal body . Bilateral flap tunnelled under labia majora. Posterior suture line was completed first (fig-5) and after the tip was reached then anterior suture line was commenced(fig-6). Sutured flaps were invaginated as a skin lined tube into the previously created rectovesical space. Donar site was closed primarily without tension. **Fig2-8.**

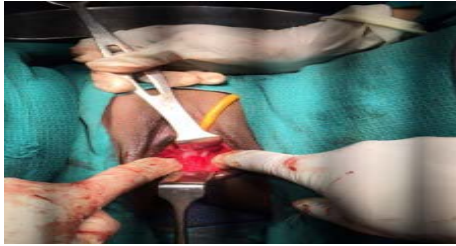


fig-2 Image Shows Vaginal Atresia

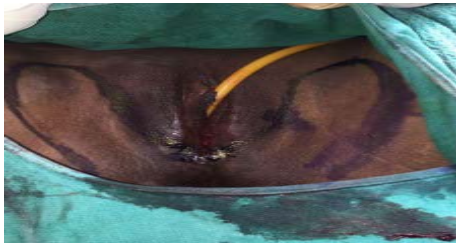


fig-3 Bilateral Pudendal Thigh Flap Marking



fig-4 Elevation Of The Flap



Fig-5 Suturing Of The Posterior Aspect Of Flap

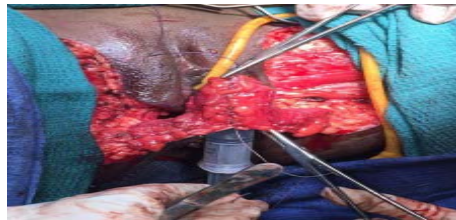


fig-6 Suturing Of Anterior Aspect Of The Flap Around A Syringe To create A Tubular Canal



fig-7 sutured flap were invaginated as a skin lined tube into the previously created rectovesical space.



fig-8 vaginal mould to maintain the neovagina from collapsing

DISCUSSION :

Vaginal reconstruction surgery is necessary in congenital absence of vagina, gender assignment surgery, vaginal contracture and stenosis, and reconstruction following neoplastic resective surgery or radiotherapy. One of the most widespread surgical technique performed for creation of neovagina was McIndoe operation⁶ in which the space is surgically created between rectum and urethra, then mould covered with a full or split thickness skin graft is placed in it. Here, in our case vaginal atresia is corrected by pudendal thigh flap .It is an axial pattern fasciocutaneous flap based on the terminal branch of superficial perineal artery with reliable vascularity and sensation at its lower part of the flap. This technique is simple and safe with no need to use stent or dilators after surgery. Moreover longterm use of stent or dilators can traumatise the posterior vaginal wall and has a risk of neovaginal rectal ulcer. The reconstructed vagina has a natural physiological angle and a correct anatomic axis to facilitate intercourse. It is observed that the risk of vaginal contracture and stenosis are reduced as compared to the other methods of vaginal reconstruction surgery⁷. The functional outcome was excellent in all patient as noticed by the relief of menstrual flow blockage, and abdomen pain, sexual and psychological satisfaction⁸.

CONCLUSION:

Hence, the condition carries a serious sexual, psychological, and even social burden. So the aim of reconstruction are to provide the cosmetically satisfactory introitus, a conduit for normal menstruation and also to facilitate pain free sexual intercourse.

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