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TLH in Rare Case of Didelphys Uterus with Hemi Vaginal Septum

Poonam Khera, Kanika Garg, Laxmi Mantri, Preeti Yadav

Uterus didelphys is a rare congenital Mullerian duct anomaly caused by failure of fusion of two Mullerian ducts (Class III anomaly). It is characterized by two hemi uterus, 2 endocervical canals and two cervices. Vagina can be single or double, depending upon the presence of septum. Each hemi uterus has one fallopian tube. The prevalence is reported as 8.3 % of all Mullerian duct anomalies. It can be associated with renal anomalies in 10% of cases. Herlyn-Werner-Wunderlich is the triad of didelphys uterus, obstructed hemi vagina septum and ipsilateral renal agenesis, involving both Mullerian duct and Wolffian structures.

A 50-year-old P4L4 presented with complaints of menorrhagia and metrorrhagia for 1 year, received medical treatment for 1 year but had no relief. D&C was done 2 months back and HPE report showed inflamed polyp. She had history of lap ligation done 22 years back. P/A - NAD. P/S - Cervix seen on left side of vagina, right side cervix not seen, only fistular opening seen on the vaginal septum right side from where blood was coming out during menstruation. USG and MRI report showed complete duplication of uterine horns and cervix with single vaginal, no renal abnormality. She had no relief with conservative management so was planned for total laparoscopy hysterectomy with BSO. Intraoperatively on inspection of the pelvic cavity, lots of omental adhesions were seen with anterior abdominal wall and right lateral pelvic wall, two uterine corpora almost same size with broad vesico-rectal fold were seen, anteriorly continuing with bladder peritoneum and posteriorly with peritoneum over the bowel. Right side tube and ovary were adherent with pelvic wall and posteriorly also. Because of adhesion all around the right corpora, decision was taken to do subtotal hysterectomy on right side to make surgery easy and to avoid ureteric and bladder injury as uterine elevator was not used on the right side due to vaginal septum. Vesicorectal fold opened, taking care of rectum posteriorly and bladder anteriorly. Bladder pushed down. Right corpora divided below the isthmus after coagulating the uterine arteries to do subtotal hysterectomy on right side. Cervical stump looked normal after subtotal hysterectomy. Then, left side TLH done, both uteri delivered vaginally. After delivering both uterus, right side cervix was seen from the colpotomy area which was held with vulsellum.

Para vaginally, paracervical clamps were applied after excision of vaginal septum, cervical stump was easily removed from vagina. Endo suturing was done, A drain put, cystoscopy was done after finishing surgery and patient was discharged after 48 hours.

Herculean Task of Removing More Than 100 Fibroids

Pooja Gupta, Dinesh Kansal

Diffuse uterine leiomyomatosis (DUL) is a rare condition that poses a unique management challenge. Hysterectomy is the standard treatment as myomectomy might be unable to achieve a complete clearance and repair of uniformly involved myometrium, but in women who want to preserve their fertility hormonal treatment, transcervical resection, uterine artery embolization and myomectomy are the other treatment modalities.

A 29 Years unmarried female came with complaints of abdominal distension, heaviness lower abdomen and increased bleeding during periods for last 2 years. We did a myomectomy for the same patient in 2015 and removed 48 fibroids. Her sister was also operated for multiple fibroids at some other hospital but expired due to DVT post operatively. Her uterus was enlarged up to 36 weeks size. Patient was counselled for hysterectomy but patient and their parent wanted to conserve the uterus as she was unmarried. All the risk of myomectomy was explained to the patient. Pre operatively hemoglobin was built up by Injectable iron and DVT prophylaxis given. Myomectomy was done by midline vertical incision. Dense bowel and omental adhesions were present, which were removed by sharp dissection. Injection vasopressin injected in the uterus. 106 Fibroids were removed of varying size from 0.5 cm to 9 cm. Bilateral uterine artery ligation was done. Blood loss was approximately 4 liters. Patient was shifted to ICU on ventilatory support. She was shifted to ward on 4^{th} day. She developed paralytic ileus on day 5, which was managed conservatively. She was discharged on day 8 in stable condition. She came for follow up on day 14, Stitches were removed and injection GnRH agonist 11.25 mg was given to prevent the recurrence.

Lap Creation of Neovagina in MRKH by Peritoneal Pull Through

Dinesh Kansal, Pooja Gupta

Type I or Typical MRKH refers to isolated symmetrical uterovaginal aplasia or hypoplasia with normal ovarian function. For many years, McIndoe's method has been the gold standard. However, laparoscopic methods are being utilized more often to avoid an unsightly scar at the

site of skin graft, incomplete uptake of graft and high incidence of rectal-bladder complications associated with McIndoe's. Among the two laparoscopic methods, Davydov's is being preferred due to less pain, short hospital stay, longer vagina and higher sexual satisfaction.

Our patient was 26 years old, married and had 1-inch vaginal dimple. A neovagina was created for sexual function by using peritoneal pull through method- Davydov's technique. Counselling is of paramount importance in these patients regarding need of regular dilatation, expectations from surgery and future fertility options. In this patient, complete dissection was done laparoscopically.

Initially, releasing incision was given posterior to urinary bladder which also helps in keeping it safe from harm. Peritoneal incision was given at the potential POD just anterior to rectal fat and rectum was pushed down. A sponge on holder was used to push through vaginal dimple. Laparoscopically, an incision was made transversely over the bulge with harmonic from one uterosacral ligament to the other. Vaginal space was thus created to reach the skin at introitus. The space was lined anteriorly and posteriorly by pulling down peritoneum. Peritoneal edges were fixed to introitus with absorbable sutures. Vaginal vault was formed by taking a purse string suture that included uterosacral ligaments, sero-muscular layer of rectum and uterine horns. A soft mould was kept in vagina.

A Rare Case of Harlequin Ichthyosis

Laxmi Mantri, Manali Paul, Poonam Khera

Harlequin Ichthyosis (HI) is a rare genetic condition affecting the skin. The skin of the newborn is covered with thick diamond-shaped plates that resemble fish scales. These hard plates can make it difficult for the baby to breathe and eat. It is a life-threatening condition requiring immediate intensive care. Its incidence is 1 in 300,000 births. Approximately 200 cases of HI have been reported worldwide. Protective or the barrier function of the skin is disrupted, making it more difficult for affected infants to control water loss, regulate their body temperature, and fight infections. Infants often experience excessive dehydration and develop life-threatening infections. HI has an autosomal recessive inheritance. It is due to mutation in adenosine triphosphate binding cassette A 12 gene (ABCA 12 gene). This ABCA 12 gene is responsible for the exocytosis of the lipid-containing lamellar granules that control the desquamation process. The locus for ABCA 12 gene is located on chromosome 2q35. The recurrence of this condition

in the following pregnancy is 25%.

Our patient was 28 years old, second gravida, presented at 34 weeks pregnancy with labor pains. Her antenatal ultrasound at 32 weeks showed that baby had the possibility of Harlequin syndrome. The patient had a spontaneous preterm vaginal delivery, and a female baby weighing 1.9 kg with all features of Harlequin Ichthyosis was delivered. The baby was shifted to NICU immediately for conservative management. The baby had white porcelain-like skin with deep creases all over the body with bleeding from the creases. Eyelids and lips were everted, showing ectropion and eclabium. The mouth was open with thick lips. Genetic counseling was offered to the patients. Next genome sequencing was done. Both parents turned out to be heterozygous (carriers) of the ABCA 12 gene, and the affected baby was homozygous for the mutation.

Although HI is a rare and life-threatening condition, yet no definitive cure has been established. As it is a rare genetic disorder, prenatal genetic testing in the form of CVS or amniocentesis to check for the mutation in ABCA 12 gene is the mainstay of diagnosis.

Quiz

A quiz based on the May month Bulletin was held after the above presentations. The answers are highlighted in bold italic font.

Q1. Second stage arrest is diagnosed after at least 2 hours of pushing in nulliparous women

a. True
b. False

Q2. Suspicious and pathological tracings have a limited capacity to predict low APGAR score

a. True
b. False

Q3. Patient Controlled Intravenous Analgesia is programmed dose of IV medication

a. Self administrated with lockout intervals between doses
b. Self administrated without lockout intervals between doses
c. Administrated by nurse with lockout intervals between doses
d. Administrated by nurse without lockout intervals between doses

Q4. When oxytocin is used for induction of labour, the target is to achieve

a. Strong uterine contractions every 2-3 minutes
b. Uterine activity of 200-250 Montevideo units

c. a & b

d. None of the above

Q5. Shock index is calculated by dividing

a. Heart rate divided by diastolic blood pressure

b. Diastolic blood pressure divided by heart rate

c. Heart rate divided by systolic blood pressure

d. Systolic blood pressure divided by heart rate

The winners of the quiz were- 1st position- Dr Neha Katre, 2nd position- Priyanka Chauhan, 3rd position- Shakti Sharma.