Abstract: Herlyn Werner Wunderlich Syndrome is a very rare Type III Mullerian anomaly associated with obstructed unilateral vagina and ipsilateral renal and ureter agenesis. We are discussing a case of Uterine didelphys obstructive type presenting as Hematocolpos Hematometra. MRI revealed Renal agenesis and diagnostic laparoscopy was done. Drainage of hematometra and marsupialization was done.

Keywords: Uterine didelphys, Haematocolpos, Excision, Marsupialization

INTRODUCTION
Uterine diadelphys constitute about 11% of uterine malformation. Didelphys uterus arises when midline fusion of the Mullerian duct is arrested either completely or incompletely. The incidence of renal anomalies is about 20%. Obstructed unilateral vagina in patients with didelphys is frequently a HERLYN WERNER WUNDERLICH SYNDROME. It is approximately 1/2000 to 1/28,000 cases.

CASE REPORT
14 Year old girl who attained menarche 1 year back admitted with complaints of severe lower abdominal pain for the past 3 days associated with difficulty in micturition. She got regular cycles since menarche 3/30 scanty flow Her last menstrual period was 09.05.13 On Examination of the abdomen A swelling of size about 15x12 cm occupying the hypogastrium right iliac and left iliac fossa was present which was not tense and tender mobility restricted Considering her age, Examination under anesthesia was done.

Bimanual Pelvic Examination reveals a tense swelling was present in the right lateral wall which was extended superiorly with the uterus was felt. Ultra sound was showing Haematomata and right renal agenesis. MRI was showing Right renal agenesis, uterine didelphys with two uterine horns widely separated two endocervical canals and two separate vaginal cavities. Right sided vagina grossly distended with blood measuring 12.5x7x6.5 displacing the uterus superiorly suggestive of Haematocolpos, the ovaries are normal, no ovarian adnexal mass lesion.

Because of Haematometra with severe pain, diagnostic Laparoscopy was done the findings are Uterine didelphys. left horn normal, left tube and ovary normal. Right horn was distended up to size 16x15cm. Right tube and ovary congested and hemorrhagic spots seen. Pouch of douglas was occupied by distended vagina. On local examination blush distended vagina was seen. Incision was made and of about 800 ml of altered blood drained followed by excision of the septum and marsupialization was done post operative period was uneventful.

HAEMATOCOLPOS
DISCUSSION
Herlyn–Werner Wunderlich (HWW) syndrome is a very rare congenital anomaly of the urogenital tract involving both Mullerian ducts and Wolffian structures. It is characterized by the triad of uterine didelphys, obstructed hemivagina, and ipsilateral renal agenesis. Most of the patients are asymptomatic. Obstructive variety are presenting with complaints of sudden lower abdominal pain, urinary retention because of haematocolpos.
Mullerian duct anomalies have an overall prevalence rate of 2-3% of all women of which constitutes about 11%. It is associated with complete/partial vaginal septum in 75% of uterine didelphys. The potential complications of this syndrome are pyo-haematocolpos, pyosalpinx, pelvic peritonitis and long term complications such as endometriosis, pelvic adhesions and increased risk for abortions and infertility. When the patient is having Mullerian duct anomalies in order to obtain accurate MDA classification MRI must be performed. MRI is most accurate in diagnosing, thereby allowing most appropriate treatment. It is very sensitive to detect the uterine contour. Character of the septum, and associated endometriosis, pelvic inflammation & adhesions.

UTERUS DIDELPHYS

Laparoscopy should now be considered the gold standard for evaluation of Reproductive Tract Anomaly, it could be reserved for cases presenting with Haematocolpos haematometra & pyometra. It can be therapeutic in drainage of haematometrocolpos or marsupialization of blind vagina, failure of treatment results in urinary retention, haematosalphinx, endometriosis(4). If the patient is having didelphys uterus, HWWS Syndrome should be excluded to check for ipsilateral kidney, another common condition is associated endometriosis. Another kind of Obstructive Mullerian anomaly is OHVIRA Syndrome obstructed hemi vagina other renal anomalies such as duplicated kidneys, dysplastic kidneys also be present.

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