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Title: A RARE CASREPORT OF MRKH (MayerRokitanskyKusterHauser SYNDROME).

INTRODUCTION :

Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome, also referred to as Müllerian aplasia is a congenital disorder characterized by aplasia or hypoplasia of the Müllerian duct derivatives with eugonadotropic primary amenorrhea in females with normal secondary sex characteristics,normal female hormonal profile and a normal female karyotype (46,XX).It can be isolated or associated with renal and skeletal abnormalities .It has an estimated prevalence of 1 in 5000 live female births..

AIMS AND OBJECTIVES:

Identify the type of primary amenorrhea and proper management of case.

MATERIALS AND METHODS

15 year old female with primary amenorrhea came to GMH Hanmakonda for evaluation . She had no history of cyclical abdominal pain.On examination,height,BMI,vitals within normal limits. Secondary sexual characteristics present as per age. On Examination, there is no mass palpable per abdomen .On inspection,Hymen is intact with depression seen just below hymen. On per rectal examination,uterus,cervix and upper 2/3rd vagina not palpable.All biochemical and microbiological tests with in normal limits. Hormonal profile of TSH,FSH,LH,prolactin,testosterone,estrogen within normal limits.whole skeletal X-ray shows no skeletal anomalies..IVP showed normal urinary tract.Karyotyping showed 46XX karyotype.USG findings confirmed with MRI showing absence of uterus and upper 2/3rd vagina

RESULTS:

Primary amenorrhea with normal female stature ,normal secondary sexual characteristics,normal hormonal profile,normal karyotype confirms eugonadotropic primary amenorrhea.USG,MRI confirmed absence of uterus and upper vagina which confirmed MRKH syndrome.

CONCLUSION:

The caring of patients with MRKH syndrome is complex and requires a patient-centered multidisciplinary approach in careful dialogue with the patient addressing all-together gynecological, sexual, psychological and infertility issues.The advent of Uterine transplantation as the first available fertility treatment for MRKH syndrome has provided new hope for these patients to become pregnant and achieve biological motherhood.

DISCUSSION:

Morcel K et al and Willemsen WN et al studies found that functional sexual outcomes were similar after surgical and non-surgical techniques.Callens Net al Study analysed 190 studies found that when anatomical success was defined as a length of ≥7 cm and functional success as coitus, all vaginoplasty techniques yielded significantly higher success rates (>90%) as compared to surgical techniques.

REFERENCE:

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