

INTRODUCTION

- MRKH Syndrome is caused by embryologic underdevelopment of the Mullerian duct
- It results in agenesis or atresia of the vagina, uterus, or both

OBJECTIVES

- Correct diagnosis of the underlying condition
- Evaluation for associated congenital anomalies
- Psychosocial counseling in addition to treatment or intervention to address the functional effects of genital anomalies

NO CONFLICT OF INTEREST

CASE

HISTORY

- 20 year old came with c/o primary amenorrhea associated with cyclical lower pain abdomen, every month for 3-4 days since 5 years.

EXAMINATION

- Vitals were stable, secondary sexual characteristics were of tanner stage 4 ,normal abdominal examination and normal external genitalia findings

TREATMENT

- Progesterone only pills given.
- Counselling regarding the future fertility options of adoption and gestational surrogacy given
- Treatment options include vaginal elongation and the surgical creation of a neovagina before resumption of sexual activity were given

INVESTIGATION

- USG ABDOMEN/PELVIS revealed ectopic left kidney, cystitis, hypoplastic uterus, polycystic pattern of right kidney, left ovarian complex cyst.
- MRI ABDOMEN/PELVIS confirmed-hypoplastic uterus with bicornuate configuration, right polycystic ovarian patter ,functional left ovarian cyst, caudate lobe hypertrophy of liver, ectopic malrotated left kidney S/O MRKH syndrome type 2
- Echo showed normal findings
- Peripheral blood karyotyping 46xx and normal hormonal levels(FSH,LH,PRL,PROGESTERONE, TESTOSTERONE)

CONCLUSION

- Timely identification and multidisciplinary approach care is needed
- Continuous research is needed to advance understanding and refine treatment approaches ,enhancing outcomes, and quality of life for patients of mrkh syndrome

DISCUSSION

- Incidence of MRKH Syndrome is 1 per 4,500–5,000 females
- Adolescent patient with primary amenorrhea must be evaluated thoroughly
- Androgen insensitivity syndrome and obstructive vaginal/uterine anomalies must be excluded

REFERENCES

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