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Citation

Abstract
Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome is characterized by Mullerian duct system anomalies, vaginal atresia is the most frequent variant. It can be associated with renal, skeletal, spine and other malformations.

Objective: The first documented case of MRKH syndrome on the West Indian island of Nevis is reported. A 54 year-old female presented describing primary amenorrhea, infertility and protrusion of the anterior vaginal wall. Clinical and sonographic examination confirmed vaginal atresia, agenesis of the uterus, as well as a moderate cystocele. Surgical correction of the cystocele was performed, further corrective dilatory surgery of the vagina was recommended and psychological support was provided.

INTRODUCTION
Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome was first described by Mayer in 1829 and Rokitansky in 1838.1,2 Its incidence is 1 in 4,000 to 5,000 females and affects all races equally.3 The syndrome was described as the agenesis of the uterus and vagina due to abnormal development of the müllerian ducts; uterine and vaginal agenesis were reported by Rokitansky, while Mayer described some vaginal duplications.1,12,13,14 In 1910, Küster recognized urologic associations, such as renal ectopy or agenesis.5 In 1961, Hauser differentiated MRKH from testicular feminization6. Although rare, MRKH syndrome represents 15% of cases of primary amenorrhea and as such is usually not suspected or diagnosed until adolescence.7 Given the reproductive, psychological and sexual repercussions that this entity has on the affected patients, continuity of medical and psychological care is a must. A variety of surgical procedures are now available that attempt to correct the deficiencies of the vaginal canal, allowing these patients to benefit from a normal sexual functioning.8 She denied any family history of congenital disorders or exposure to teratogenic agents. Hormonal tests (Estradiol, FSH, LH, TSH, Prolactin) were all within normal limits for a adult female in the follicular phase. Preliminary clinical examination revealed a normal female phenotype, normal labia majora and minora in addition to a shallow and blind vaginal pouch, measuring 4.5 centimeters in length and 3 centimeters wide. The cervix was not visualized on speculum exam. Descent of the anterior vaginal wall was diagnosed as a moderate cystocele. Transabdominal and transvaginal sonographic scan of the abdomen and pelvis confirmed congenital agenesis of the uterus and the presence of ovaries as well as the absence of urinary tract anomalies. The cystocele was corrected surgically under general anesthesia by anterior colporrhaphy. Further corrective dilatory surgery of the vagina was recommended and psychological support was provided. It is of interest to note that the Island of Nevis has a population of 11,000 and this is the first documented and reported case of Mayer-Rokitansky-Küster-Hauser syndrome.

CASE REPORT
A 54 year-old black female presented complaining primarily of a vaginal protrusion restricting further an already limited sexual function. Upon further interview, she confirmed that she has been presenting primary amenorrhea and infertility.
DISCUSSION

Under-development of the Müllerian duct system may result in different urogenital anomalies. These malformations are the source of intense concern for both the patient and her family due to their repercussions on reproductive health as well as psychological and sexual impact. Initial diagnosis of these anomalies is made usually during adolescence. Uterine anomalies have been reported to occur in 1 in 594 fertile women (0.17%) and in 1 in 29 infertile women (3.5%). The MRKH syndrome can be regarded as resulting from the cessation of development of the Mullerian duct between the sixth and seventh week of gestation, which in this condition extends only as far as its attachment to the caudal mesonephric ligament, the round ligament. The adjacent structures beyond this point, which include the connecting strand and smooth muscle bundles dorsal to the bladder and vaginal rudiment, are derivatives of the Wolffian duct or Gartner's duct, respectively and renal ducts.

Figure 1: Transabdominal pelvic sonogram showing absence of uterus.

References

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