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The surgical management of mayer- rokitansky kuser hauser syndrome at Lagos University teaching hospital: A ten year review

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Abstract

The introduction of a purposeful vagina could be very crucial in sufferers with mayer-rokitansky-hauser-syndrome (mrkh). Many surgical techniques had been defined for vaginal reconstruction in mrkh. They may be classified as nearby, nearby and distant flaps. We evaluated the surgical techniques utilized in our center and in comparison the outcome and complications. The mayer-rokitansky-kuster-hauser (mrkh) syndrome is a unprecedented anomaly characterised with the aid of congenital aplasia of the uterus and vagina in girls showing everyday development of secondary sexual characters and ordinary 44 xx karyotype. We report our revel in inside the management of two sufferers with congenital absence of the vagina due to the mrkh syndrome. The primary case become a 24-year-antique scholar, who supplied with number one amenorrhea, uterovaginal agenesis, proper pelvi-ureteric junction obstruction, and left renal agenesis. The second one affected person become a 24-12 months-old housewife, who offered with primary amenorrhea and inability to obtain penetrative sexual sex. She had vaginal atresia and a grossly hypoplastic uterus. Both had a hit sigmoid colovaginoplasty and are sexually active. Vaginal reconstruction the usage of the sigmoid colon noticed a direct and first-class final results in both sufferers the congenital aplasia or intense hypoplasia of mullerian systems is infrequent. However, the functions of everyday woman endocrine characteristic paired with the absence of a useful uterus and vagina makes it a fascinating entity. The prognosis and paintings-up in these sufferers has emerged as very efficient, way to the use of imaging, and there are a couple of hit methods for the introduction of a neovagina. In recent years, infertility remedy options thru in vitro fertilization have also emerge as available as a part of the long-term care of those patients.This shape of mrkh syndrome is also known as isolated mullerian aplasia, or rokitansky sequence. The disease is characterised via the failure of the uterus and the vagina to increase well. The severity of mrkh syndrome kind i may also range greatly from one man or woman to some other. In most cases, the uterus and/or the vagina have not developed (aplasia); in different uncommon cases, there can be narrowing (atresia) of the top part of the vagina and an underdeveloped or rudimentary uterus. In a few instances, the fallopian tubes can be affected as well. The ovaries of ladies with mrkh syndrome are unaffected and characteristic generally. In maximum cases, the preliminary symptom of mrkh syndrome kind i is the failure to start menstrual cycles (number one amenorrhea). No matter amenorrhea, affected women do revel in regular secondary sexual improvement consisting of breast improvement, the growth of hair underneath the palms and within the public place, and an increase in frame fats across the hips and different regions. Sex steroid levels, lady sexual identification, and level of sexual choice (libido) are all additionally everyday. However, because of the absence of the uterus and properly advanced fallopian tubes, all affected ladies are unable to endure kids (infertile). Many affected ladies additionally revel in trouble at the same time as attempting sexual

Intercourse due to the shortness of the vagina. Some women may additionally experience pain at some point of sex. Mrkh syndrome kind i is every now and then known as mullerian aplasia due to the fact the mullerian ducts are a twin structure within a developing embryo that in the end develops into the uterus, fallopian tubes, cervix and the top portion of the vagina. It's far believed that unsuitable development of tissues derived from the mullerian ducts happening at some stage in embryogenesis, in the long run causes the signs of mrkh syndrome. When the abnormalities that signify mrkh syndrome kind i occur in association with extra bodily findings, the disorder is assessed as mrkh syndrome type ii or (mu)llerian duct aplasia, (r)enal dysplasia and (c)ervical (s)omite anomalies or murcs association. The maximum common abnormalities related to mrkh syndrome kind ii are failure of the kidneys to improvement nicely (renal adysplasia) and numerous skeletal malformations, mainly vertebral. A whole lot much less common defects include coronary heart malformations and listening to impairment.

Women with MRKH syndrome type II may exhibit absence of a kidney (unilateral renal agenesis), malformation of one or the two kidneys (renal dysplasia), underdeveloped (hypoplastic) kidneys and/or improper positioning within the body of one or both kidneys (renal ectopia). Renal abnormalities can cause growth deficiency, kidney stones, an increased susceptibility to urinary tract infections and abnormal accumulation of urine in the kidney due to obstruction (hydronephrosis). Many females with MRKH syndrome type II also exhibit skeletal malformations. For example, bones (vertebrae) in the spinal column within the neck (cervical vertebrae) and the upper back (thoracic vertebrae) may develop improperly (dysplasia). As a result, some of the vertebrae within the neck may be missing and/or fused, causing shortness of the neck, limited neck motion, and an abnormally low hairline (Klippel-Feil syndrome). In addition, affected females may exhibit asymmetric, fused or wedge vertebrae; malformed or missing ribs; abnormal sideways curvature of the spine (scoliosis); elevation of the shoulder blade (scapula), due to the scapula's failure to move into the appropriate position during fetal development (Sprengel deformity). (For more information on Klippel-Feil syndrome and Sprengel deformity, please see the Related Disorders section of this report.). Abnormalities of the head and face may also occur including an abnormally small jaw (micrognathia), cleft lip, cleft palate and underdevelopment of one side of the face causing facial asymmetry.

Results: Twelve patients were seen and managed. One patient was treated with the Gracilis Myocutaneous Flap (GMCF), and one patient had colonic vaginoplasty (CV). Eight patients were treated with bilateral pudendal thigh flap (BPTF). Two patients accepted vaginal dilatation, however, one defaulted from treatment. We found it easier to raise the PTF with minimal blood loss and also easier to reduce the flap under the pubic arch. The colonic vaginoplasty has the advantage of lack of dryness during sexual intercourse

Conclusion: Vaginal dilatation is not culturally acceptable in our patients. The PTF is easier to raise and inset as neovagina compared with GMCF. The GMCF tend to be bulkier. However, the three flaps described are good choices of vagina replacement with little or no risk of vaginal stenosis.