V. The risk of the intercourse undertaken by a pacient with Mayer- Rokitansky- Küster-Hauser (MRKH) syndrome

*Vasile Nițescu
Medical Centre of Obstetrics-Gynecology and Sexology;

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Abstract:

A birth defect, the MRKH syndrome type 1 is characterized by uterine and vaginal agenesis.

It is the case of a patient aged 21 that had no uterus and the vagina had 5cm in the lower part, without any upper part, within the uterine and vaginal agenesis.

MRKH syndrome type 2, aside of the lack of uterus and vagina, indicates other defects, too, the most frequent affecting the kidneys, as in this case.

Abnormalities appear also in the morphophysiology of labia, in which the labia majora are hypotrophic and difficult to see, having an erotic sensitivity much under that of the labia minora, which are hypertrophic and have neurovascular and anatomic direct ties to the clitoris.

As well, also within the particulars, the vaginal introitus is small in size and determines difficult penetration and copulation, by the different defective structure of the vagina.

The short vagina (5cm in length) in its lower side, given its connections to the hyper-eroticism area located behind the pubis, and particularly to the clitoris, but to the other erotic areas, too, as well as the inner sides of the labia minora, determined a strong sexual arousal to the patient, which allowed the penetration of the vaginal introitus and copulation, within reasonable limits, but after the intercourse's completion (orgasm and ejaculation), given that the pleasurable sensation disappeared, the patient began completely feeling local pain and bleeding, which made her rush into the hospital.

Key words:

Muller agenesis, MRKH syndrome, partial vaginal aplasia, unilateral renal agenesis, urogenital folds.

*Correspondence to: *Nițescu Vasile MD., Ph.D, 9 Washington Street, 1st District, post code 011792, Bucharest, Romania

Description:

The MRKH syndrome is a malformation of the female genitalia, determined in the embryonic period, at the end of the gestation week 4 and 5, by ceasing the development of Müllerian ducts in the embryonic mesoderm. As consequence, disturbances occur in the organogenesis process, determining uterine and vaginal agenesis and the lack of uterus and virginal. As well, there can be an agenesis of the 2/3 upper parts of the vagina, various degrees of vaginal hypoplasia, stenosis, caliber modifications, all making up the type 1 of MRKH syndrome.

The lower parts of the Müllerian ducts are the first influenced in the malformation process. The defects, which are extremely varied, affect the vagina and are explained by the fact that the biggest part of the vagina does not develop from the Müllerian duct, but from the urogenital sinus, the fused part of the Müller *paramesonephric ducts*, together with the organs of the urinary apparatus, as the bladder and the urethra. Thus, it explains the situation of the vagina being present while the Müllerian (Kirsch) duct misses and the urethra and bladder that are present.

By the complexity of the embryonic structures concerned, during the defect-creating process, type 2 of the MRKH syndrome, other defects appear, too, which affect the urogenital apparatus, such as ectopic kidney, unilateral renal agenesis, horseshoe kidney, disturbances in emptying the bladder, urinary incontinence by sphincter assessment, deaf-

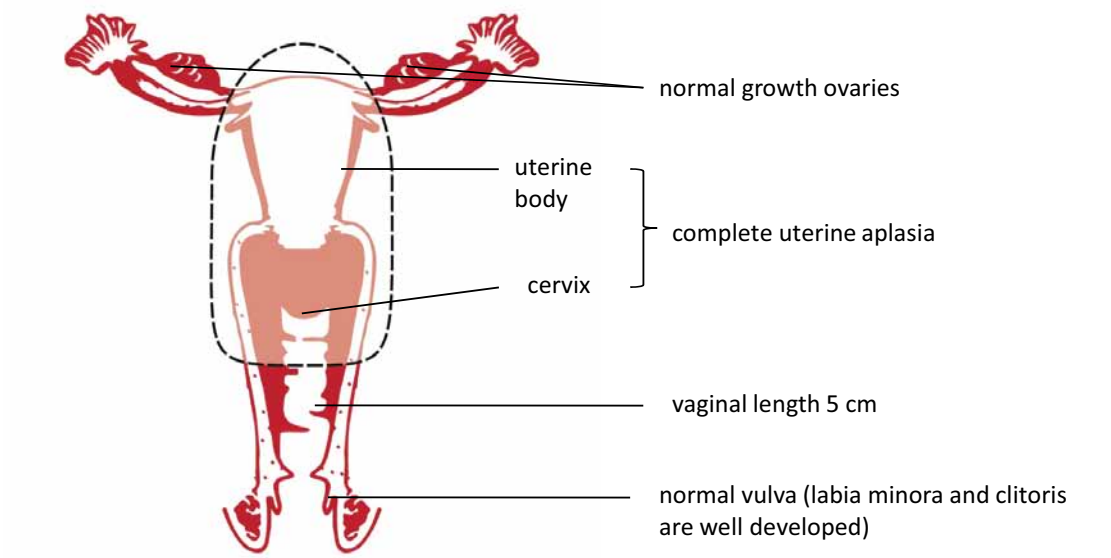

Fig. 1 - Complete uterine agenesis and partial vaginal agenesis (author's own sketch)

ness, heart defects, and skeletal abnormalities in the vertebrae, particularly C1-C7 cervical segment.

It was also described the existence of a facial asymmetry, which together with the uterine and vaginal agenesis, obviously enhances the depression of the patient during her puberty and adolescence in connection to whether her image as a female exists in the social environment, family, and in relation to her future.

Within the MRKH syndrome, the ovaries and Fallopian tubes have a normal morphophysiology, and the secondary sexual characters are present.

In fact, the external genitalia, the development of breast, pubic and head hair, respectively the value of female gonadotropin are normal.

The uterus aplasia determines primary amenorrhea (reason for the patient going to a doctor) and infertility, and the vaginal aplasia renders impossible to have intercourse.

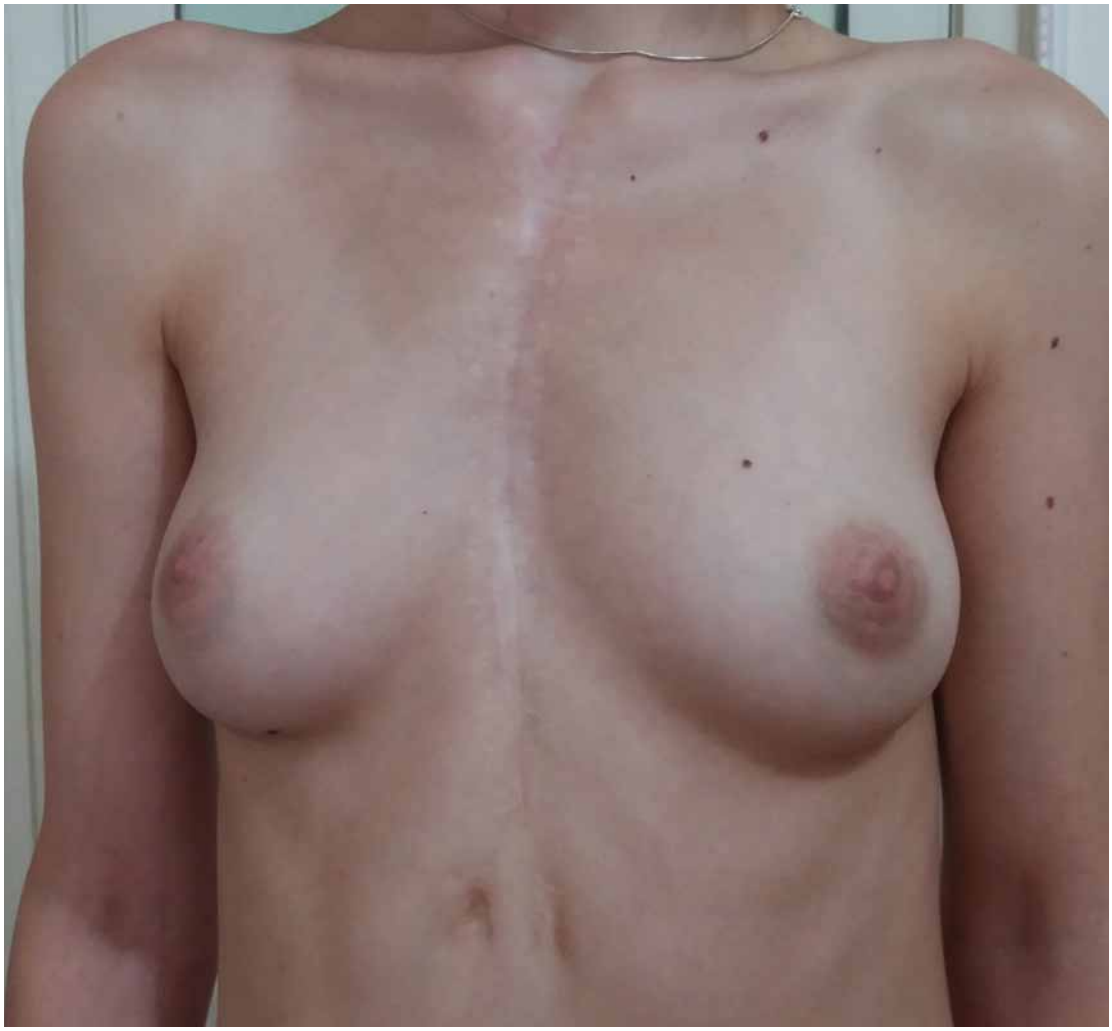

Fig. 2 - Pre-sternal scar, post cardiotomy for cardiac malformation

The existence of a "uterus leftover" determines cyclical abdominal pain, the intensity being reported to it.

The relaxation of the narrow vaginal caliber or of the vaginal stenosis is painful and long lasting, and some of them determines, at times, impossibility of favorable relaxation and thus, the penile-vaginal intercourse cannot unfold.

Case study

Patient aged 21 required gynecologic consult for vaginal bleeding that appeared after intercourse. Given that until 16 years she had no menstruation, she went for a checkup. After a specialty consult, the MRKH syndrome was diagnosed to the patient, which involved uterine agenesis, it being responsible for primary amenorrhea and agenesis of the upper part of the vagina (fig. 1).

When examining the patient, the next matters were found:

Minor facial asymmetry;

Flattened sternum, with a long scar resulting from surgery (fig. 2), following a surgery performed for a heart defect;

Frontal deformation of the dorsolumbar spine, with a moderate primary curve to the right and two adjacent compensation curves;

Bilateral mammary glands with normal appearance;

Gynecology examination: mons pubis covered with hair; under the tegument, adipose tissue can be found; labia majora are hypotrophic, they can be separated and seen with difficulty; labia minora are hypertrophic (fig. 3a, b) and stretch from the base of the clitoris obliquely down.

On the right labia minor, on the median side and towards the clitoris region, slightly bleeding tears were seen, which have been determined by the masturbation means used, as the patient reported.

The vaginal orifice (fig. 4) is oval and small in size, with fibrous edges and highly resistant. On the edges, in various spots, there are bleeding scars (fig. 4), which give the impression of profoundness right next to the hymeneal caruncle. The rigidity of the vaginal introitus renders difficult the penetration of the vaginal cavity by the use of valve speculum. The vaginal fornix can be seen through the vaginal orifice;

The vagina, which is present in the lower part, above the vaginal orifice, is 5cm in length. The vaginal walls have strictures and stenoses, which alter the vagina caliber. Bleeding lesions can be seen on the vaginal walls next to the scarred stenosis. The observed stenoses, scars and muscle structure modifications decrease the elasticity of the tissues;

The upper side of the vagina (its end) is an arch given the lack of cervix and obviously, of the Douglas pouch. Instead of the uterus, which misses given its agenesis, there are 2 uterine leftovers of 1/1.3 mm, without any cavity, located asymmetrically on one side and the other of the median line between the rectum and the bladder.

The Fallopian tubes and the ovaries are normal and located in the pelvic region;

Unilateral renal agenesis (left kidney);

Heart defect – surgery performed.

Discussions

The labia minora come from the urogenital folds and are separated; the labia majora come from the labioscrotal folds. The labia develop while the specific hormones lack. Given that the morphophysiological differences between the 2 types of labia are obvious (labia major are hypotrophic, difficult to notice, and the labia minora are hypertrophic and greatly surpass the labia majora), one can deem that this abnormality also represents a birth defect ancillary to the MRKH syndrome, with a different morphofunctional significance. The labia minora, which are over

Fig. 3a

Fig. 3b

Fig. 3 a, b- Hypertrophic labia minora in the MRKH syndrome

developed (fig. 3a, b), spread from the base of the clitoris down, and have direct neurovascular and anatomical ties to the clitoris, by the clitoral hood, thus making up an important eroticism area. This is the explanation of the fact that any manual maneuver on that area, as in the case presented, induces strong sexual arousal, easily leading to 1-2 orgasms, fellatio and penile-anal intercourse being preferred and rarer the penile-vaginal intercourse, which is uncomfortable and painful to the patient.

On the surface of labia minor, special tactile corpuscles are found, which determine a high erotic sensation, the pain perceived being much more reduced, given that endorphins are discharged.

The sebaceous, sudoriferous and small vestibular glands, which are very numerous on the mucosa of labia minora, together with

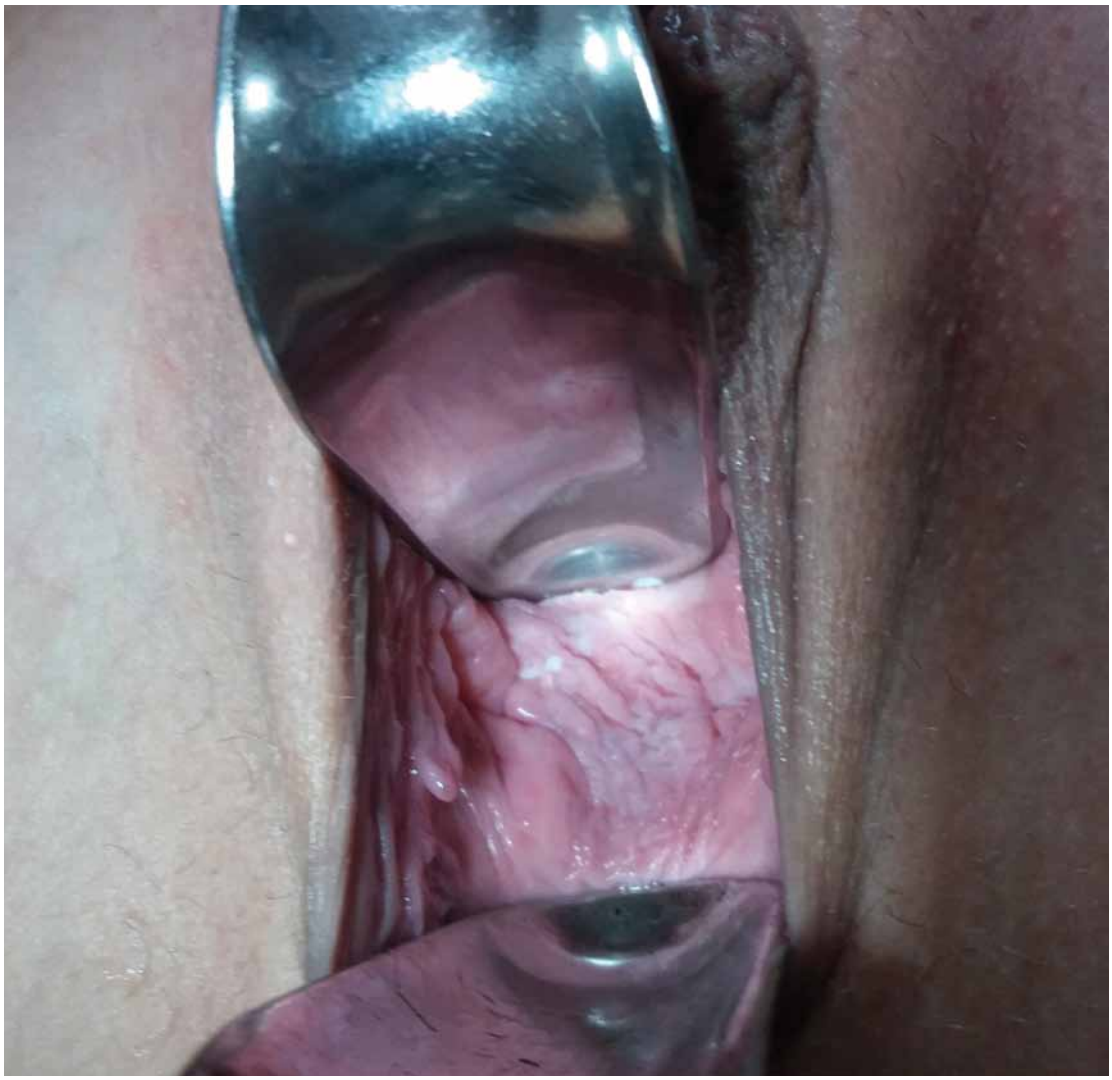

Fig. 4 - The appearance of the vagina in the MRKH syndrome

the secretion of the ancillary glands, which spill into the vaginal vestibule, lubricate the surface of the labia and vaginal introitus, thus favoring not only penetration, but also completion of intercourse in the case presented. Given the content of female pheromones of the patient, inducing the state of sexual attraction is determined, as well as increasing the excitation state and erection of the partner. Thus, the strong tone of the penis determined the vulvovaginal lesions described above.

The labia minora, as well as the lower third of the vagina, which has a higher bio excitability by comparison to the upper two thirds of the vagina, benefits of the direct connection to all its surrounding erogenous areas (please refer to Hyper-eroticism area –Treaty of Clinical Sexology), thus offering enhanced erectile traits, present in the patient as well, as reported by her.

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Conclusions

The high number of sebaceous, sudoriferous and small vestibular glands, by the mucus they discharge, as well as the large vestibular ancillary glands, which open into the vulva, by lubricating the surface of the labia and vaginal introitus have allowed the vaginal orifice to be penetrated with difficulty and caused the bleeding lesions. On the vagina level, its penetration and the copulatory movements have determined bleeding tears on the relaxed muscle fibers, by stretching the walls of the vagina affected by hypoplasia, by affecting the stenoses and scars and by recalibrating the defective vagina.

The pain and the bleeding tears, weakly perceived during sexual arousal, due to the endorphins discharged, were perceived as such following orgasm and ejaculation and as consequence, the patient rushed into the emergency room, where she was admitted for hemostasis and analgesic treatment.