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# Diagnostic And Tactical Errors In The Management Of Patients With Vaginal And Uterine Aplasia At The Prehospital Level In The Republic Of Uzbekistan

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***Abstract: Diagnostics and treatment of genital abnormalities in girls remains relevant to the present day. The greatest scientific interest is uterine and vaginal aplasia. The vast majority of patients with Meier-Rokitans-Kuster-Hauser syndrome are diagnosed in adolescence and older age, as a rule, when complications appear, which worsens the prognosis. The treatment requires adequate psychosocial support of such patients throughout their life with the solution of the reproduction problem.***

***Keywords: vaginal and uterine aplasia, Mayer-Rocitan-Kuster-Hauser Syndrome, congenital defects of reproductive system organs development.***

## 1. INTRODUCTION

Uterine and vaginal abnormalities are congenital defects of the reproductive system, which are more common in recent years. Vaginal malformations in girls are one of the most urgent problems of reproductive health of children and adolescents. One of the most frequent abnormalities of uterus and vagina development is Mayer-Rocitans-Kuester-Hauser syndrome. This syndrome is characterized by uterine and vaginal aplasia.(5)The frequency of vaginal and uterine aplasia is 1 in 4000-5000 newborn girls. 90% of all vaginal aplasia observations are made with Mayer-Rocitans-Kuester syndrome, 7% of vaginal aplasia observations with a functioning uterus. (Kiera E.F. Politova A.K., Vyazmina K.Yu.) The causes of Mayer-Rokitans-Kuster-Hauser syndrome are not sufficiently studied, because the pathology of development occurs in the intrauterine period, due to the formation of fetal mesenchyma. Patients with this pathology are characterized by a female phenotype (normal development of mammary glands, proportional body, external genital organs are developed by a female type, ovaries have an unchanged structure and function and karyotype 46XX.(1,2). The hormonal background corresponds to a two-phase cycle. Despite the high occurrence of this pathology, it often remains unrecognized before menarche or is detected in adulthood.

There are many diagnostic methods to determine uterine and vaginal abnormalities, but despite this, the diagnosis of patients with Mayer-Rokitans-Kuester-Hauser syndrome remains relevant to this day and is a clinical problem with serious consequences in the management of these patients. Pathology of the reproductive system is identified and treated at an older age, as a rule, when complications appear, which worsens the prognosis. In 33% of cases, uterine and vaginal aplasia is combined with various variants of renal anomalies (3,4,6,7) and is classified as a complex malformation MURCS (Mullerian Renal Cervico-thoracic Somite anomalies,

MIM 601076) (7). Particular attention should be paid to complex combined vaginal, musculoskeletal and urinary system defects.

Diagnosable because of the rare occurrence of this pathology, the different nature of related diseases and malformations of other organs and systems. This leads to incomplete and untimely diagnosis, wrong management and treatment, unjustified surgical interventions in 23-24% of cases and as a result complications. Inability of sexual activity and infertility combined with psychosocial problems characterizes Mayer-Rokitans-Kuster-Hauser syndrome as a complex disorder of the female reproductive system. (5) The tactics of managing patients with Mayer-Rocitans-Kuster-Hauser syndrome is constantly in the focus of gynecologists, surgeons and pediatricians.

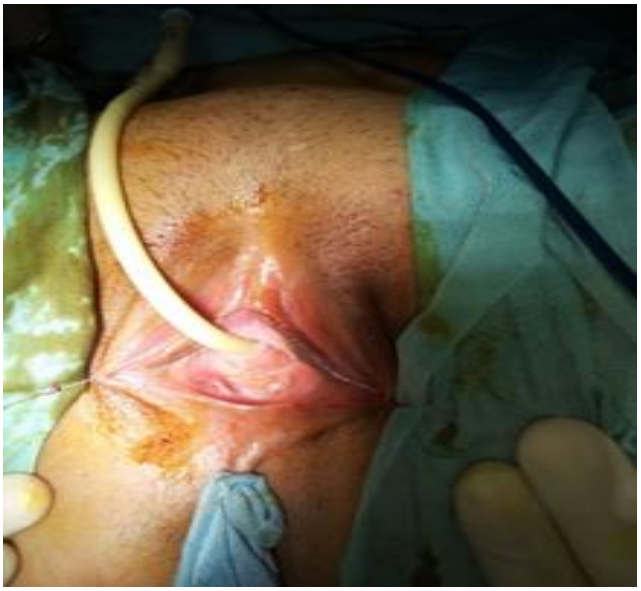
Difficulty in early diagnosis is due to insufficiently clarified clinical features of vaginal defects, diagnostic errors in the recognition of anomalies, and difficulties in early diagnosis (3,4). Improper diagnosis leads to ineffective therapy, psychoemotional trauma of patients and their relatives, irreversible metabolic disorders, which in turn affects the future fate of the patient.

This syndrome is the cause of primary amenorrhea, which is the first reason why patients go to specialists. Diagnosis of the disease is to collect anamnesis, examination of the external genitalia, assessment of physical and sexual development, laboratory and general clinical studies, bacteriological and bacterioscopic examination of excreta from the genital tract, vaginal probing, rectal abdominal examination, ultrasound of the urinary and sexual systems. Patients of older age usually worry about the inability to lead a sexual life. In some cases, Meier-Rokitans-Kuster-Hauser syndrome is detected as a result of emergency treatment of a woman due to severe perineal tears during an attempted sexual intercourse.

It is important to note that timely sexual desire and need for sexual life in women with Mayer-Rocitans-Kuster-Hauser syndrome serve as a traumatic factor that leads to the subsequent formation of neurosis-like disorders: depression, anxiety, as well as mistrustfulness with symptoms of obsessive states, feeling of own inferiority (1). Taking into account the emotional consequences of psychological stress (loss of biological motherhood, sense of abnormality, lack of equality with peers) and compassion of others after the diagnosis, professional psychological support should be an integral part of medical care at all stages of treatment, which is a guarantee of success. (4) Full understanding and interaction with the patient is important.

During the treatment, adequate psychosocial support for such patients is needed throughout their life cycle, with a solution to the problem of reproduction. After the therapy, the quality of life of the patients improves, and confidence in themselves is restored. Today, the combined medicine is able to significantly improve the life of a woman with this syndrome, but this requires long-term multi-level treatment. The therapy comes down to removing the defect through intimate plastics. As a result, the patient can get rid of psychological and physiological problems, enjoy life and even have children with the help of a surrogate mother(3,5)

**Purpose of our study:** Analysis of errors and improvement of diagnostics in patients with Mayer-Rocitans-Kuster-Hauser syndrome.



Patient M.N. 22 years old with Mayer-Rokitansky-Kuster-Hauser syndrome after 1/5 phemostone treatment.

## 2. RESEARCH MATERIALS AND METHODS

We carried out clinical and laboratory analysis of 38 patients who applied to the private clinic of "Samarkand Doctor Shifo-Baht" Limited Liability Company and to the gynecological department of Samarkand Maternity Complex № 3 in the period from 2013-2020. All investigated patients and their parents have carefully collected anamnesis. Complete vaginal and uterine aplasia was observed in 28 (73.7%), partial vaginal aplasia in a functioning uterus in 5 (13.1%), complete vaginal aplasia in a functioning uterus in 3 (7.9%), doubling of the uterus and vagina with partial vaginal aplasia in 2 patients (5.3%). In our study, the age range of patients under study was 15 to 30 years. By age range, the patients were distributed as follows: 15-20 years old 11 patients (28.9%), 20-25 years old 23 patients (60.5%), 25-30 years old 4 patients (10.6%). Of these, 7 patients (8.4%) were from the city and 31 (81.6%) were from the countryside. Studying the gynecological anamnesis in the study revealed the presence of burdened anamnesis in 23 patients (60.5%). According to the primary acces to specialists patients were distributed as follows: 6 patients (15.8%), 11 children's gynecologists (28.9%) and 21 gynecologists (55.3%).

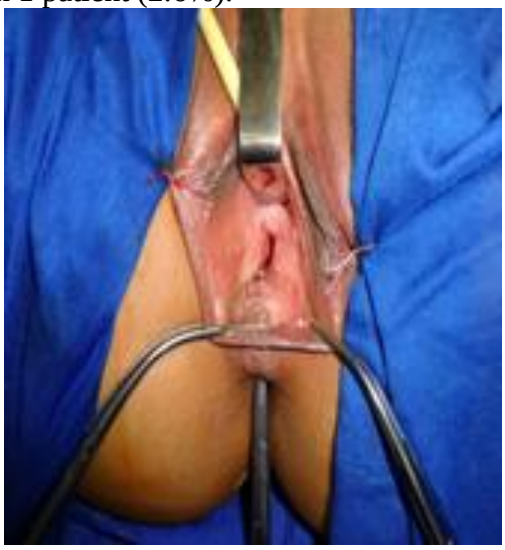
Patients more often complained about the absence of menarche, which was observed in all 38 patients, as well as cyclical lower abdominal and lower back pain in 27 patients (71%), the inability to have sex coitus in 19 patients (50%), difficulty in sexual intercourse in 8 patients (21%), incontinence of feces and stools in 8 patients (21%). All patients (with Meier-Rokitans-Kuster-Hauser syndrome) had well developed secondary signs corresponding to the age norm. During the gynecological examination, the structure of the external genitalia was correct, female type. In 31 girls there was no entrance to the vagina, 7 girls had a normal structure of the gimen, followed by a deepening of 0.5-3 cm. In a two-handed rectoabdominal study, the uterus was not determined in 28 patients. In the study of ovarian function, it was found that the hormonal profile of girls with Mayer-Rocitans-Kuester-Hauser syndrome did not differ significantly from that of healthy peers.

## 3. RESULT AND DISCUSSION

In the ultrasound study in 9 (23,6%) of patients, the uterus was determined in the center of the small pelvis as a formation of cylindrical shape, the size of which on average did not exceed the values typical for 2-7 years of girls with normal sexual and physical development. In 5 patients the uterus was determined in the form of two muscular rollers located in the parietal cavity of small pelvis with sizes not exceeding 3.1x1.4x2.8 cm. The ovaries were located high at the walls of the small pelvis and their size corresponded to the age norm. Follicles with diameter from 1.5 to 2.7 cm on ovulation days were determined in 5 patients. In the course of kidney ultrasound examination it was found: aplasia of one kidney - 7 patients, expansion of calyx-fibrous complex in 1 patient, kidney size reduction in 1 patient, dystopia of one kidney in 1 patient, doubling of calyx-fibrous system in 2 patients.

## 4. CONCLUSION

Having not diagnosed Mayer-Rokitans-Kuester syndrome, 23 patients (60,5%) received "outpatient treatment" on different links. Patients mainly received hormonal therapy for functioning ovaries, which resulted in metabolic disorders and obesity of various degrees in 21 patients (55.2%). 17 patients received hormonal therapy with 1/5 phemostone. Most of them, on the advice of doctors, "get married and everything will go away", did not consult specialists before sexual activity. As a result, after the beginning of sexual life, patients were treated after complications: Direct bowel injuries, which were observed in 12 patients (31.5%), recto-vaginal fistulas in 3 patients (7.7%), perineal tears of various degrees in 5 patients (13.1%), urethral and bladder tears in 1 patient (2.6%).



**Patient Z.O. 1 year was treated with phemostone 1/10, a year after the beginning of sexual life turned to Samarkand Doctor Shifo-Baht Ltd., after the formation of recto-vaginal fistula.**

Thus**,** late diagnosis, irrational patient management and prescription of hormonal therapy leads to psycho-emotional and physical trauma of patients, metabolic disorders. Late surgeries, especially after marriage, degrade the quality of life of patients.

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