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IMPROVING THE PRACTICE OF FEMINIZING SURGERY IN THE DEVELOPMENT OF GENITAL ORGANS

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ANNOTATION

Despite the progress made in the treatment of disorders of sex development, many aspects of birth defects remain controversial and controversial. In particular, the methods of reconstructive plastic surgery that feminize them have not beyen fully resolved. The following article presents the clinical manifestations of several types of defects in the formation of the genital organs in girls.

Key words: adrenal glands, malformations of the genital organs, congenital adrenal hyperplasia.

Relevance of the topic. We all know that in recent years, modern examination and treatment methods have been applied in all fields of our medicine, but the treatment of diseases of genital development defects among girls remains one of the urgent problems of children's gynecology. Despite the progress made in the treatment of gender dysphoria, many aspects of the correction of developmental defects remain controversial and controversial. Including methods of reconstructive plastic surgery that feminizes them are not fully resolved

Disturbances of the development (formation) of sexual organs (JAShB) create a unique problem for the individual in the society in terms of domestic and spiritual aspects. JAShB occurs in 1 in 5000-10000 births. Genetically, such diseases repeat in one family up to 20-25%. According to WHO, intersexual development occurs in 1 in 1,000,000 births. However, this pathology also increases in the postnatal period.

The cause of any form of congenital hyperplasia of the adrenal gland is mutations in the genes responsible for the synthesis of enzymes or transport proteins involved in the synthesis of cortisol. The most common cause is a lack of the enzyme 21-hydroxylase, which accounts for 95% of all cases, which is caused by mutations in the CYP21 gene; Then comes 11-β-hydroxysteroid dehydrogenase deficiency and 3β-hydroxysteroid dehydrogenase deficiency. They make up 4%, the rest are special cases.

Adrenal hormones include mineralocorticoids (aldosterone), glucocorticoids (cortisol), and sex steroids (testosterone and estrogen). The syndrome occurs when enzyme deficiency causes a decrease in the synthesis of glucocorticoids in the adrenal cortex. The effect of adrenal cortex hormones on the pituitary gland decreases, so the

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synthesis and secretion of adrenocorticotropic hormone (ACTG) increases through a negative feedback mechanism.

Adrenogenital syndrome (AGS), (congenital hyperplasia of the adrenal gland), testicular feminization (TF), (Morris syndrome) are the main causes of sexual dysfunction. An example of this is the late diagnosis of AGS and TF diseases or the presentation of patients in adolescence. it is common in our region. Unfortunately, scientific fundamental works aimed at this problem are not found in practical terms. This makes the problems of early diagnosis and treatment of developmental defects of sexual organs even more difficult in medicine.

There are 3 types of congenital adrenal hyperplasia:

21-hydroxylase deficiency with salt loss syndrome (salt-resolving type); the most common form of the disease. This is represented by a defect in the development of the external genitalia. It manifests itself as somatic diseases in the form of diarrhea, nausea, seizures and disturbances in the cardiovascular system.

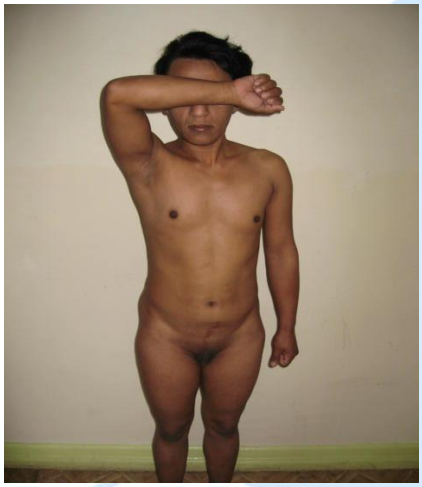
Simple viril form ((Простой virilnyy.)incomplete 21-hydroxylase deficiency); along with male development in girls. Despite the "female" genetic makeup, the external genitalia of girls born more or less resemble those of men. This is called false female hermaphroditism.

Non-classic form (pubertal and post-pubertal).

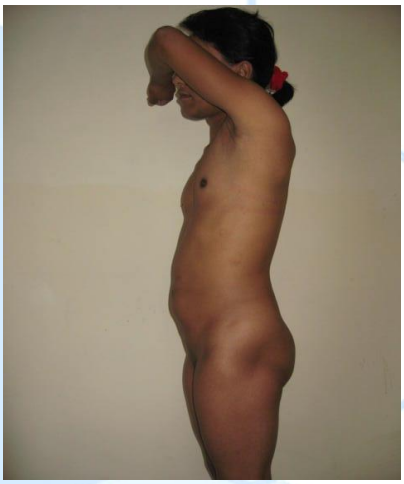
The pubertal form manifests itself in adolescents. Underdevelopment of mammary glands, enlargement of the clitoris, late onset of the first menstruation, lengthening of the menstrual period, hirsutism, acne are observed in girls.

The postpubertal form develops in women, often in the form of increased adrenogenital syndrome after an aborted pregnancy. Menstrual cycle lengthens, menstruation shortens. Hirsutism.

Depending on the type of disruption of the 21-hydroxylase gene (mutation status, number of mutations, zygosity), different levels of enzyme activity loss are observed.



simple virile form



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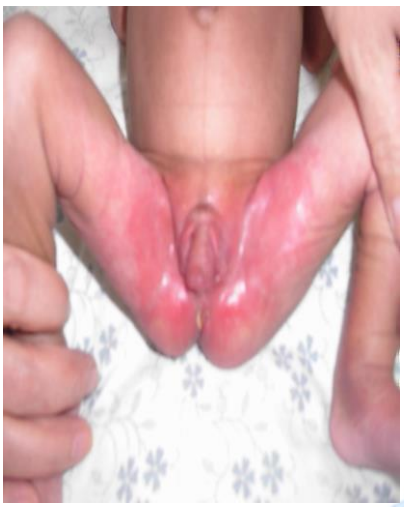

salt loss syndrome

Testicular feminization (TF), the feminization syndrome of (Morris syndrome), the full form (TTFp) is an androgen insensitivity syndrome and is the most common type of false male hermaphroditism. It occurs at a frequency of 1:20,000-1:60,000 in the general population [4]. Patients have a female phenotype with male gonads and a 46,XY karyotype. The external genitalia are female, the vagina is shortened, the beak ends, there are no pubic hairs; At puberty, the formation of mammary glands lags behind and primary amenorrhea develops.

Purpose of research. Selection and improvement of surgical methods of treatment of feminizing disorders in gender formation in children and adolescents.

Inspection materials and methods. We conducted examinations on 45 patients aged 5 to 31 years who were treated in 2015-2023 in the gynecology department of the 3rd maternity complex and in the endo-gynecology department of BKTTM. Investigations were conducted retrospectively and prospectively.

Inspection results. According to our investigations, 39 of the 45 patients we are researching were diagnosed with adrenogenital syndrome, and 6 were diagnosed with testicular feminization.

As a result of our research, it became clear that the anatomy of the genital organs was not preserved despite the fact that the patients had undergone age-appropriate hormonal therapy. In the case of dome genital virilization, plastic surgery is performed: removal of the penis and clitoris, cutting of the front wall of the urogenital sinus and the formation of the entrance to the vagina. Regardless of the patient's age, the penile clitoris is removed after the diagnosis. Plastic operations aimed at the formation of the entrance to the vagina should be performed before the age of 10-11. Glucocorticoid treatment allows patients to grow and develop the soft tissues of the urogenital sinus, to a certain extent, the formation of the entrance to the vagina and the risk of injury to the urethra. reduces

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