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# TREATMENT AND DIAGNOSIS AND ADIPOSOGENITAL DYSTROPHY

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**Annotation:** Adiposogenital dystrophy is a neuroendocrine pathology manifested by overweight and underdevelopment of the sex glands. It is accompanied by gynoid-type obesity and delayed sexual development. Diagnosis is based on a physical examination by a pediatrician or endocrinologist, a study of the hormonal background. Additionally, radiography and tomography of the hypothalamic-pituitary region are prescribed. Therapy of the underlying disease that caused dystrophy is indicated. Symptomatic treatment includes administration of chorionic gonadotropin, hormone replacement therapy, and dietary nutrition.

**Key words:** Adiposogenital dystrophy , Alfred Froelich , toxoplasmosis, chlamy dia, scarlet fever.

Adiposogenital dystrophy (Pehkrantz-Babinsky-Frohlich syndrome) is a pathological condition that occurs as a result of disorders of the hypothalamic-pituitary system. The disease was first described in the late nineteenth century by the Russian doctor Pehkrantz. A few years later, the symptoms of the disease were reflected in the works of the French neurologist Joseph Babinsky and the Austrian pharmacologist Alfred Froelich. The pathology occurs in childhood or puberty and more often affects boys aged 6 to 13 years. The syndrome can be an independent disease or a manifestation of another pathological process. In the latter case, symptoms can appear at any age.

Despite the fact that the symptoms of the disease usually manifest themselves in adolescence, the formation of the pathological process can begin even at the stage of intrauterine development. The main causes of pathology include:

- Brain tumors. The syndrome can occur with craniopharyngioma, pituitary chromophobe adenoma, and other malignant and benign neoplasms located in the hypothalamus and pituitary areas.
- Intrauterine infections. Pathology is provoked by viral and bacterial infections transferred by a woman during pregnancy: flu, toxoplasmosis, chlamydia, scarlet fever, measles, typhoid.
- Some infectious diseases. Among the infections that can cause the development of dystrophy are meningitis, encephalitis, tuberculosis, syphilis, transferred in childhood.
- Traumatic injuries. The development of the syndrome is caused by birth injuries of newborns, closed and open TBI with damage to the hypothalamus and pituitary gland.

Pathogenesis

The main cause of pathology is disruption of the pituitary gland and hypothalamus. As a result of a disorder of endocrine function, the secretion of gonadotropic hormones by the adenohypophysis decreases, hypogonadism is formed. In some cases, the delay in sexual development is combined with a violation of the production of thyroid-stimulating, somatotropic and antidiuretic hormones. Damage to the hypothalamus leads to pathological irritation of its nuclei and increased appetite. Bulimia episodes occur, patients consume excessive amounts of food, and obesity develops.

Symptoms of adiposogenital dystrophy

The disease begins in childhood and adolescence, manifests itself as weight gain, increased fatigue, drowsiness, and reduced learning ability. Patients have a bulimia-type appetite disorder. During puberty, boys develop gynecomastia, cryptorchidism, and penile underdevelopment. In girls, there is a delay in the onset of menstruation up to amenorrhea, underdevelopment of the mammary glands, uterus and appendages. There are no secondary sexual characteristics (the presence of hair in the armpits and pubis, on the face of boys).

The formation of the osteoarticular system is disrupted, flat feet are detected, hallux valgus curvature of the lower legs. The formation of the skeleton is characterized by the eunuchoid type. Patients are tall, have disproportionately long limbs, and have large feet. Libido is reduced or absent. Due to obesity and insufficient nutrition of the heart muscle, myocardial dystrophy develops, which leads to a decrease in cardiac output and rhythm disturbances. The syndrome does not affect mental abilities, intellectual development corresponds to age.

The skin is pale, smooth, and dry. Often there is a violation of skin pigmentation, corneal inflammation and dry eyes. Fat deposits are localized in the abdomen, thighs, chest, and face. In boys, weight gain occurs according to the gynoid (female) type. Adiposogenital dystrophy is often combined with other endocrine pathologies: autoimmune thyroiditis, hypothyroidism, acromegaly, hypersomnia. In most cases, patients lead a sedentary lifestyle, which worsens obesity.

Complications

In case of late diagnosis or insensitivity to therapy, complications from the cardiovascular system occur: heart failure, sclerotic vascular changes, and rhythm disorders. With an increase in the size of the tumor, compression of the surrounding tissues is noted, accompanied by visual disturbances (high degree of myopia and hyperopia, corneal ulcers) and increased intracranial pressure.

Lagging in the formation of the sex glands leads to erectile dysfunction and impotence in men, amenorrhea and infertility in women. Typical complications associated with the development of obesity: type 2 diabetes mellitus, coronary heart disease. Dyskinesia of the biliary tract contributes to bile stagnation and the occurrence of cholelithiasis. With the growth of the tumor, behavior changes, increased nervous excitability, impaired consciousness and the development of coma are possible.

Diagnostics

Diagnosis of adiposogenital dystrophy is based on the examination data of a pediatrician and an endocrinologist, the study of the history of life and the course of pregnancy of the mother, laboratory and instrumental studies. If a delay in sexual development is suspected, patients are referred for consultation with a gynecologist or andrologist. The diagnosis is confirmed in the presence of pre-obesity or obesity with a predominant deposition of fat in the upper half of the body, hypoplasia of the gonads and the absence of secondary sexual characteristics. During the survey, conduct:

- Determination of the hormonal status. Perform a blood test for pituitary hormones, sex hormones (FSH, LH, testosterone and estrogen).
- X-ray and tomographic studies. X-rays of the skull, CT scan of the Turkish saddle, MRI of the brain and pituitary gland can visualize tumors, hemorrhages, dropsy, enlargement and deformity of the Turkish saddle.

Differential diagnosis of dystrophy is carried out with Itsenko-Cushing's syndrome and disease, which are characterized by normal sexual development. Pathology should be distinguished from hereditary alimentary-constitutional obesity, which has a family character and normal stages of puberty. The disease is also differentiated with the Lawrence-Moon-Beadle, Morgagni-Stewart-Morel, Shereshevsky-Turner, and Klinefelter syndromes. To diagnose these pathologies, genetic studies are used to identify chromosomal abnormalities.

Treatment of adiposogenital dystrophy

Etiotropic treatment is aimed at eliminating the disease that led to the development of a pathological syndrome. During the tumor process, surgical intervention, X-ray therapy, and chemotherapy are performed. Drug therapy for infectious and inflammatory diseases includes the appointment of antibacterial or antiviral drugs, vitamin and mineral complexes. Symptomatic

treatment involves a change in the diet and lifestyle, the introduction of hormonal drugs. In the absence of contraindications from the musculoskeletal system, physical therapy, swimming, and Nordic walking are indicated.

All patients are prescribed a low-calorie diet with a reduced content of easily digestible carbohydrates and vegetable fats. The diet includes fresh vegetables and complex carbohydrates, which increase the feeling of satiety. It is recommended to take food 6-7 times a day, avoiding prolonged fasting. With the development of bulimia, anorexic drugs that suppress appetite are indicated. Therapy for hypogonadism involves the introduction of hCG. When reaching puberty, boys are prescribed gonadotropins in combination with testosterone, girls-estrogens with progesterone.

Prognosis and prevention

The course of the syndrome depends on the etiology of the underlying disease. With timely diagnosis and treatment, the prognosis is favorable. Late detection of the syndrome, the development of complications, and progressive obesity cause disability and hinder normal functioning.

Prevention of the disease consists in the rehabilitation of chronic foci of inflammation in children and planning the pregnancy of the expectant mother. Proper nutrition, breaking bad habits, and hormone replacement therapy can slow the development of the disease. With proper treatment in the pubertal period, patients develop by age. If you follow a diet, you can control your body weight and avoid complications associated with obesity.

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