

HORMONE REPLACEMENT THERAPY IN PATIENTS WITH SHERESHEVSKY-TURNER SYNDROME

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Abstract. This article discusses hormone replacement therapy in patients with Shereshevsky-Turner syndrome.

Key words: Shereshevsky-Turner syndrome, estrogen, dysgenetic ovaries, cardiovascular diseases, cytogenetic, pelvic organs, osteodensitometry, biochemical blood test, endometrial thickness, patients, estradiol valerate, cholesterol, osteoporosis, lipid metabolism disorders.

ЗАМЕСТИТЕЛЬНАЯ ГОРМОНАЛЬНАЯ ТЕРАПИЯ У БОЛЬНЫХ С СИНДРОМОМ ШЕРЕШЕВСКОГО-ТЕРНЕРА

Аннотация. В статье обсуждается заместительная гормональная терапия у больных с синдромом Шерешевского-Тернера.

Ключевые слова: синдром Шерешевского-Тернера, эстроген, дисгенетические яичники, сердечно-сосудистые заболевания, цитогенетический, органы малого таза, остеоденситометрия, биохимический анализ крови, толщина эндометрия, пациенты, эстрадиола валерат, холестерин, остеопороз, нарушения липидного обмена.

Introduction: Shereshevsky-Turner syndrome (STS) is the most common cause of impaired sexual differentiation. Due to the lack of estrogen production by dysgenetic ovaries, the formation of normal puberty does not occur, and a decrease in bone mineral density (BMD) and early progression of cardiovascular diseases are observed. Among patients with TTS, 90% require hormone replacement therapy (HRT) to initiate and/or achieve progress in sexual development and prevent late complications of estrogen deficiency.

Materials and methods: 14 teenage girls aged 12–19 years (average age 16.4 ± 3.2 years) were examined. The diagnosis of STS was made based on the results of a cytogenetic study. All patients had anthropometric data measured and bone age assessed. The degree of sexual development was assessed using the J. Tanner scale. A study of blood hormones (FSH, LH, prolactin, TSH, estradiol, AMH), ultrasound of the pelvic organs, and osteodensitometry were performed.

The degree of sexual development was assessed using the J. Tanner scale. A study of blood hormones (FSH, LH, prolactin, TSH, estradiol, AMH), ultrasound of the pelvic organs, and osteodensitometry were performed. In a biochemical blood test, glucose levels and lipid profile were assessed. HRT was started with estrogen monotherapy: an oral preparation of estradiol valerate 0.5 mg/day or a transdermal preparation of 0.1% estradiol hemihydrate gel 0.25 mg/day was chosen.

Subsequently, a gradual (every 6 months) dose increase was carried out to 1 mg/day, and then 1.5 and 2 mg/day.

When a spontaneous menstrual-like reaction appeared or after 12–18 months, when sufficient endometrial thickness was achieved according to pelvic ultrasound (8–10 mm), the patients were transferred to combined HRT in a cyclic mode by adding progesterone preparations from 16 to 25 days. 1st day of the menstrual cycle or prescribing a combined drug of cyclic HRT (usually 17β -estradiol 2 mg and dydrogesterone 10 mg). During HRT, a control examination of patients was carried out, including an assessment of the dynamics of sexual development, a lipid profile, ultrasound of the pelvic organs (size of the uterus and endometrial thickness over time), and densitometry.

Results: The karyotype of patients with STS was represented by various options: 45X; 45X/46XX; 45X/46X+der (X); 46X+mar/45X; 45X/46Xi, (Xq); 46X, del (X). The average height of the patients was 131.5 ± 4.6 cm. At the time of treatment, the mammary glands did not exceed the development stage according to Tanner Ma3, and after 2 years while taking HRT they corresponded to Ma4-Ma5. The dimensions of the uterus before the start of HRT $(28.4 \pm 0.8) \times (20.4 \pm 0.4) \times (29.6 \pm 0.5)$ mm, after 2 years of HRT they increased to sizes corresponding to the norm for women of reproductive age, — $(42.3 \pm 1.3) \times (25.2 \pm 0.6) \times (44.1 \pm 1.1)$ mm. Hypercholesterolemia and dyslipidemia were detected in 57.1% of cases.

Treatment with sex steroids, regardless of the route of administration, led to a decrease in elevated levels of cholesterol and atherogenic fractions of lipoproteins. All patients with STS had reduced BMD both in the lumbar spine and in the proximal femur (Z-score ranged from -1.9 to -2.6). Estrogen replacement therapy for two years resulted in improved bone quality (Z-score -1.2 to -1.9).

Conclusion: Timely and adequate hormone therapy makes it possible to achieve the formation of secondary sexual characteristics, ensure full function of the uterus before a planned pregnancy, and prevent the progression of osteoporosis and lipid metabolism disorders.

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