Effect of treatment of double precocious puberty in a 9-year-old girl- case report

Beata Sawicka¹, Hanna Borysewicz- Sańczyk¹, Justyna Michalak¹, Artur Bossowski¹

¹Department of Pediatrics, Endocrinology, Diabetology with Cardiology Division, Medical University in Białystok, Poland

Background

Precocious puberty means an abnormally early onset of puberty (before age 6 in girls and before age 9 in boys). There are two main types of this condition:
- central precocious puberty
- peripheral precocious puberty

Congenital adrenal hyperplasia due to 21-hydroxylase deficiency is the most common disorder, in which lack of the enzyme causes deficiency aldosterone and cortisol. The result of this deficiency is an increased level of ACTH. The classic form appears in early childhood and may be associated with the development of abnormal genitalia. Clinically older patients present the GnRH-independent precocious puberty with rapid growth and advanced bone age.

Non-classic form of congenital adrenal hyperplasia due to 21-hydroxylase deficiency is the most common disorder, which is associated with the use of hydrocortisone. Clinically patients present the GnRH-independent precocious puberty with rapid growth and advanced bone age. Inappropriate inhibition of androgen secretion can induce precocious central puberty.

Case report

We present the case of 9-year-old girl, who was treated of the congenital adrenal hyperplasia due to 21- hydroxylase deficiency. When the girl was 5 years old, she was diagnosed because of precocious puberty. The height and weight were over 97 percentile, bone age – 11 years, advanced puberty by Tanner stages: Th 1, Pub 3, Ax 1, enlarged clitoritis. In laboratory tests the patient had normal cortisol level in serum and elevated adrenal androgens. Congenital adrenal hyperplasia was confirmed in steroid profile in urine. The hydrocortisone had been administered orally in dosages of 8-12mg/m²/day in three divided doses. The side effect of that therapy was hypertension. The result of using hydrocortisone was slow down progression of bone age. When the girl was 7.5 years old, because of progress precocious puberty, the test with GnRH was performed. Central precocious puberty was treated with triptorelin to achieve gonadotropin inhibition. Due to insufficient improvement after treatment with hydrocortisone the using therapy with dexamethasone (0.5 mg daily orally) caused decreased level of androgens, slowed growth during first year of treatment and normalization steroid profile in urine.

Conclusions

1. The preferred glucocorticoid for chronic treatment of the congenital adrenal hyperplasia due to 21-hydroxylase deficiency is hydrocortisone.
2. Sometimes, precocious puberty should be treated centrally in children.
3. Proper treatment of precocious puberty prevents final short stature.