



The Impact of 21 hydroxylase deficiency on Cardiac Repolarization Changes in Children with 21-hydroxylase-deficient congenital adrenal hyperplasia



Hüseyin Anıl Korkmaz¹, Rahmi Özdemir², Mehmet Küçük², Cem Karadeniz², Timur Meşe², Behzat Özkan¹

¹ Pediatric Endocrinology Clinic, Dr.Behcet Uz Children Disease and Surgery Training and Research Hospital, Izmir, Turkey.

² Pediatric Cardiology Clinic, Dr.Behcet Uz Children Disease and Surgery Training and Research Hospital, Izmir, Turkey.

Introduction:

The purpose of this study is to compare the 12-lead electrocardiographic measures such as PWd, QT interval, QTd, Tp-e interval, Tp-e/QT and Tp-e/QTc ratio in patients of 21-hydroxylase-deficient congenital adrenal hyperplasia with that in healthy control subjects matched for age, sex, height, weight and body mass index (BMI).

Methods:

In the Endocrinology Outpatient Clinic of Dr. Behcet Uz Children's Hospital, Twenty-five patients of 21-hydroxylase-deficient congenital adrenal hyperplasia and twenty-five control subjects were enrolled into this observational, cross-sectional, controlled study. The evaluation consisted of anthropometric measurements, biochemical parameters, and electrocardiographic (ECG) measures. The standard 12-lead electrocardiography was performed in all patients and P-wave dispersion (PWd), QT interval, QTd, QTcd, Tp-e interval, Tp-e/QT and Tp-e/QTc ratios were calculated.

Results:

There were no significant differences in the groups for age, sex, height, weight and BMI (median age 112.8 (90.4) vs. 80.7 (109.5) months, mean weight 37.6±21.5 vs. 27.9±18.3 kg, mean height 125.4±28.9 vs. 114.7±31 cm, mean BMI 21.4±5.7 vs. 18.9±3.4 kg/m²).

Table 1. Demographic, clinical and laboratory characteristics of patients and controls

Variables	Patients (n:25)	Controls (n:25)	p
Age, months*	112.8 (90.4)	80.7 (109.5)	0.28
Male/Female	9/16	8/17	
Height, cm	125.4±28.9	114.7±31	0.21
Height, SDS*	-0.77±1.3	-0.04±0.8	0.67
Weight, kg	37.6±21.5	27.9±18.3	0.09
Weight, SDS*	0.84±1.4	0.34±1.1	0.17
Body mass index, kg/m ²	21.4±5.7	18.9±3.4	0.74
Body mass index, SDS*	1.2 (1.4)	0.8 (1.8)	0.12
Systolic blood pressure, mmHg	110.8±15	90.2±11.7	0.008
Diastolic blood pressure, mmHg*	65 (17)	60 (17)	0.41
Triglyceride, mg/dL	108±27	92.7±31.8	0.73
Cholesterol, mg/dL	160±19.7	155±25.7	0.47
LDL, mg/dL	88.5±15.3	85.6±13.5	0.47
HDL, mg/dL	48.9±9.9	52.3±13.4	0.32

HDL: high-density lipoprotein; LDL: low-density lipoprotein

* Variables without normal distribution are shown as median (interquartile range)

Table – 2: Comparison of electrocardiographic measurements between patients and controls

Variables	Patients (n:25)	Controls (n:25)	p
Mean heart rate (beats/min)	93.6 ± 18.1	101.6 ± 19.2	0.13
P dispersion (ms)	50 (25)*	40 (40)*	0.01
QT interval (ms)	371.6 ± 41.9	369.8 ± 36.8	0.94
QT dispersion (ms)	67.6 ± 33.6	62 ± 31.2	0.54
QTc (ms)	432.3 ± 20.8	422.8 ± 21.9	0.12
QTc dispersion (ms)	58.1 ± 27.2	48.6 ± 19.9	0.16
Tp-e interval (ms)	94.4 ± 15.4	88 ± 20.8	0.22
Tp-e dispersion (ms)	48 ± 15.5	35.2 ± 17.5	0.009
Tp-e /QT	0.25 ± 0.04	0.23 ± 0.05	0.19
Tp-e/QTc	0.21 ± 0.03	0.20 ± 0.05	0.43

QTc : Corrected QT, ms: millisecond

* Variables without normal distribution are shown as median (interquartile range)

Results:

The patient group did not differ from the control group in terms of age, body mass index, diastolic blood pressure, or plasma levels of triglyceride, cholesterol, LDL, and HDL. However, systolic blood pressure was significantly higher in the patient group when compared to the control group (Table 1). Mean heart rate (beats/min), QT interval (ms), QT dispersion (ms), QTc (ms), QTc dispersion (ms), Tp-e interval (ms), Tp-e /QT and Tp-e/QTc were similar between patient and control groups. P dispersion and Tp-e dispersion were significantly higher in patients of 21-hydroxylase-deficient congenital adrenal hyperplasia compared to the controls (median P dispersion 50 (25) vs. 40 (40) ms, mean Tp-e dispersion 48±15.5 vs. 35.2±17.5 ms).

Conclusions:

In this study, we have suggested that 21-hydroxylase deficient CAH is associated with detrimental effects on electrocardiographic measures of children and adolescents. To our knowledge, this study is the first to assess atrial and ventricular arrhythmias in children with congenital adrenal hyperplasia 21-hydroxylase-deficient using standard 12-lead.

Our study revealed that P and Tp-e dispersion were increased in 21-hydroxylase deficient children. P dispersion and Tp-e dispersion Tp-e ratio might be a useful marker of atrial and ventricular arrhythmias in patients with 21-hydroxylase-deficient.