

P2-268 MODY-GCK and MODY-HNF1A in children and adolescents in Russian population



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BACKGROUND

The most common forms of maturity-onset diabetes of the young (MODY) are MODY-GCK and MODY-HNF1A. Prevalence of MODY in Russian population is unknown.

AIMS

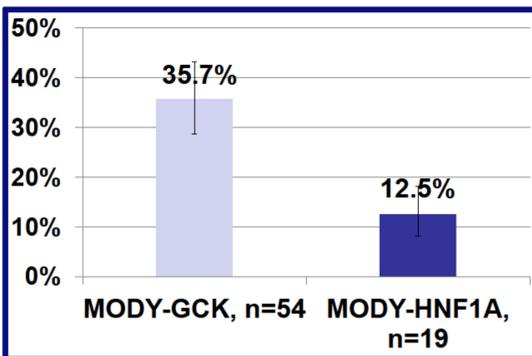
- to compare clinical laboratory characteristics of MODY-GCK and MODY-HNF1A in children and adolescents
- to estimate prevalence of MODY.

METHODS

- 151 children and adolescents were screened for mutations in *GCK* and *HNF1A*.
- HbA1c, fasting and stimulated glucose, C-peptide, insulin levels (OGTT) were measured.
- The data is presented as medians (25;75 percentile).
- Mann-Whitney U-test was used to compare medians.

RESULTS

GENETIC ANALYSIS



The prevalence MODY-GCK was **2.8 times** higher than MODY-HNF1A in Russian population.

Table 1. Results of genetic analysis

CLINICAL DATA OF PATIENTS WITH MODY-GCK AND MODY-HNF1A

	MODY-GCK	MODY-HNF1A	p
Age at diagnosis with DM, years	8.0 (4.0; 11.3)	11.8 (9.7; 13.5)	p<0.01
Positive family history, %	85.2	94.4	p>0.05
Male gender, %	61.2	31.6	p<0.05
Fasting glucose level at diagnosis with DM, mmol/l	6.8 (6.5; 7.4)	7.7 (6.9; 9.3)	p<0.01
HbA1c at diagnosis with DM, %	6.5 (6.1; 6.7)	6.8 (6.5; 7.8)	p<0.05
Prevalence of obesity (SDS BMI≥2), %	7.4	31.6	p<0.05

Table 2. Clinical and biochemical characteristics of the patients with MODY-GCK and MODY-HNF1A

MODY was confirmed by genetic analysis at 12.4 years (9.1; 15.4), disease duration was 2.4 years (0.8; 4.3).

Fasting glucose level was higher in MODY-GCK. Stimulated glucose level was lower in MODY-GCK. All MODY-HNF1A patients had diabetes (fig. 1).

Fasting serum C-peptide and insulin didn't differ between MODY-GCK and MODY-HNF1A. Stimulated (OGTT) serum C-peptide and insulin were significantly higher in MODY-GCK than MODY-HNF1A (fig.1).

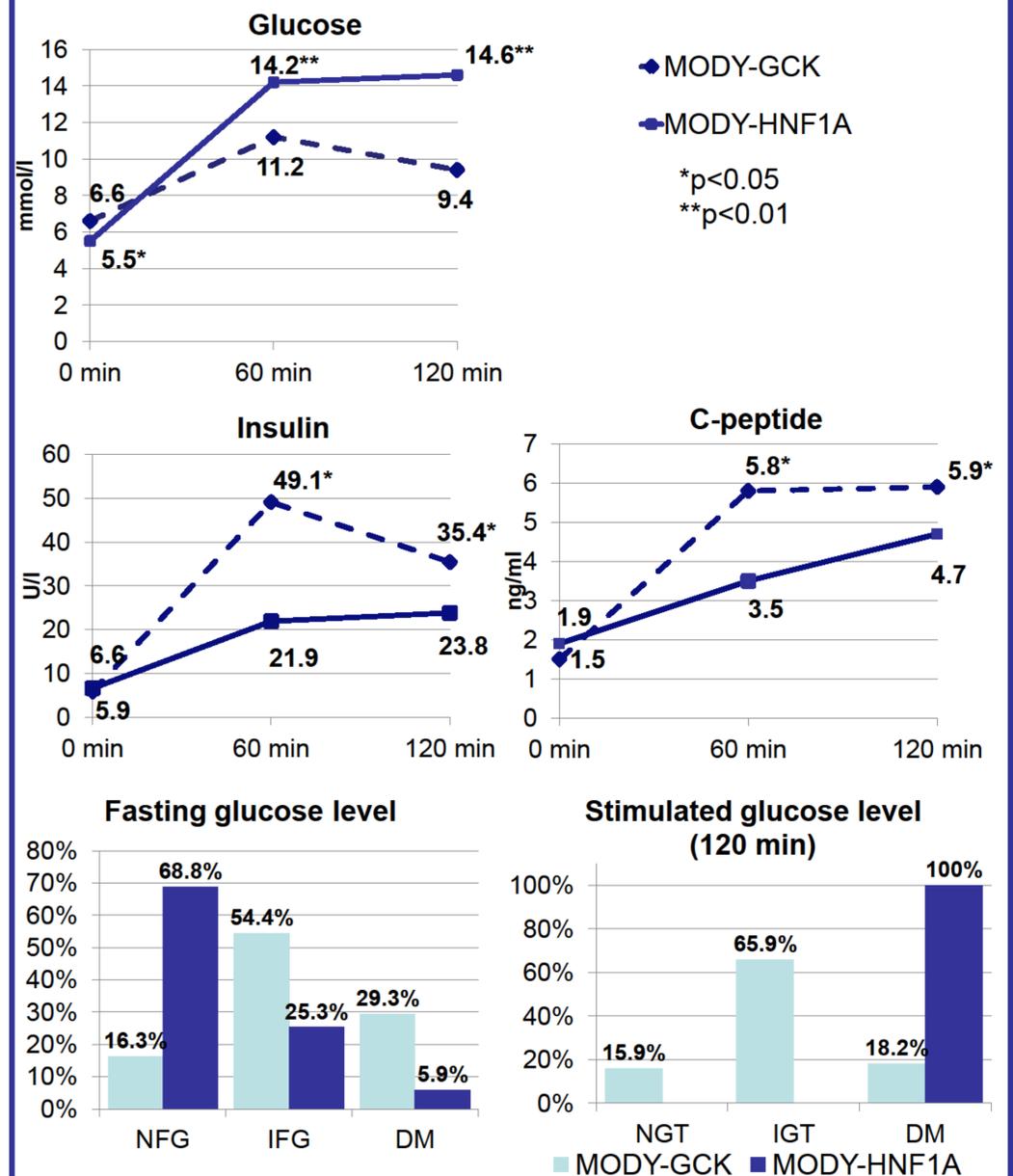


Figure 1. Fasting and simulated (OGTT) glucose, insulin, C-peptide

NFG – normal fasting glucose
IFG – impaired fasting glucose
DM – diabetes mellitus

NGT – normal fasting glucose
IGT – Impaired glucose tolerance [ISPAD, 2014]

TREATMENT

	MODY-GCK	MODY-HNF1A	p
Insulin, %	7.4	5.3	p>0.05
Sulfonylurea, %	3.7	63.2	p<0.01
Metformin, %	3.7	10.5	p>0.05

Table 3. Treatment in children and adolescents with MODY-GCK and MODY-HNF1A

CONCLUSIONS

The prevalence MODY-GCK was 2.8 times higher than MODY-HNF1A in Russian population. Patients with MODY-GCK were diagnosed earlier than MODY-HNF1A, and had milder presentation and higher stimulated C-peptide and insulin to compare to MODY-HNF1A.

