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

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

Table 1. Results of genetic analysis

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

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).

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.