A case of hyperinsulinemic hypoglycemia, associated with insulin autoimmune syndrome (IAS) in 3.5 year old girl.

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Background: IAS is a rare cause of hyperinsulinemic hypoglycaemia with only few descriptions in children in the literature. Drugs containing the sulphydryl group, such as methimazol, are known to be a causative factor of this syndrome. Diazoxide and octreotide are usually ineffective in such patients.

Objective: We aim to describe a rare case of AIS in a child, with a good response to a short course of glucocorticoid therapy.

Case study: A previously healthy 3.5 year old Caucasian girl presented with hypoglycemic seizures. It is known that she had two courses of Piritinol treatment before the onset of the disease- for 1 month (6 months before) and for 2 weeks (10 days before). On admission blood glucose monitoring showed recurrent episodes of fasting hypoglycemia (1.7-2.8 mmol/l) and postprandial hyperglycemia (11-16 mmol/l), fasting tolerance was no longer than 1.5-2 hours. Fasting test revealed non-ketotic hyperinsulinemic hypoglycemia (tab.1). OGTT showed hyperglycemia (14.7 mmol/l) at 90 minute, but normal glucose levels at 120 min. (6.9 mmol/l) (Fig.1).

To confirm the diagnosis we performed an examination insulin antibodies (AIAb) and HLA-typing

<table>
<thead>
<tr>
<th>AIAb</th>
<th>&gt;100 U/ml</th>
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<tbody>
<tr>
<td>HLA alleles</td>
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<tr>
<td>DQB 1*02</td>
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Final diagnosis: Insulin autoimmune syndrome

Treatment: Prednisone 1.4 mg/kg/day.

Follow up

2 months later (after the end of treatment)
No hypoglycaemias
14-hours fasting test was negative: BG 4.6 mmol/l, Insulin 29 µU/ml
Mildly elevated AIAb (24,4 U/ml).

8 months later (after the end of treatment)
No hypoglycaemias
13-hours fasting test was negative: BG 3.7 mmol/l, Insulin 4.3 µU/ml
Normal AIAb (4,5 U/ml).

Piritinol contains disulfide bond (fig.2). We suspect that the likely trigger factor of the disease in this case was treatment of Piritinol, although we haven't found information about the same cases in the literature.

Conclusion:
To our knowledge this is a first description of IAS in children in Russian Federation.
A short course of glucocorticoid treatment was effective in our case and might be recommended as an immunosuppressive therapy to achieve the remission rapidly.