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

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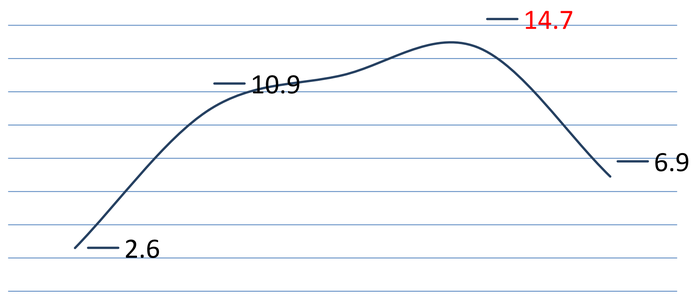
Background: IAS is a rare cause of hyperinsulinemic hypoglycaemia with only few descriptions in children in the literature. Drugs containing the sulfhydryl group, such as methimazol, are known to be a causative factor of this syndrome. Diazoxide and octreotide are usually ineffective in such patients.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

Tab .1 Fasting test

Blood glucose (BG)	2.9 mmol/l
Insulin	>1000 µUn/ml
C-peptide	16.8 ng/ml
Ketones	0.1 mmol/l

Fig. 1 Oral glucose tolerant test



atypical manifestation of the disease | **extremely high levels of serum insulin** | **discordant C-peptide level** | **absence of sulfonylurea intake**

IAS was suspected

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

AIAb	**>100 U/ml**
HLA alleles	**DRB 1 04- DQA 1* 0301- DQB 1 *0401/0402** **DRB 07 - DQA 1* 0201- DQB 1 *02**

Final diagnosis: Insulin autoimmune syndrome

Treatment: **Prednisone** 1.4 mg/kg/day.

Normoglycemia achieved in 3 days.
1 week later **insulin (>1000 µUn/ml)** and **insulin antibody levels (>100 U/ml) remained elevated.**
Dose of prednisone was gradually reduced, **treatment was stopped in 6 weeks**.

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 µUn/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 µUn/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.

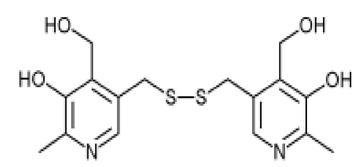


Fig. 2 «The structure of Piritinol»

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.

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