Quantitative urinary GC-MS based steroid analysis for treatment monitoring of adolescents and young adults with autoimmune primary adrenal insufficiency

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Background

Primary adrenal insufficiency (PAI) is a rare and life-threatening disease. About 50% of adolescents and young adults (AYAs) with chronic diseases are non-adherent to their prescribed treatment regimens [1,2]. Therefore, suitable hormonal monitoring of glucocorticoid replacement would be desirable in AYAs with PAI.

Objective and hypotheses

Quantitative targeted GC-MS urinary steroid metabolome analysis for treatment monitoring for AYAs with autoimmune PAI [3].

Methods

We retrospectively analyzed 21 24-hr urinary steroid metabolome analysis of four AYAs aged 15.6 ± 2.0 years with autoimmune PAI on hydrocortisone and fludrocortisone treatment. 24-hr urinary cortisol metabolite excretion rates (CMER) were calculated and transformed into z-scores (Fig. 1).

Results

Three patients (#1-3) showed good treatment adherence (17 of 21 samples). Mean CMER of these samples was 7.4 ± 1.8 mg/dL, corresponding to a z-score of 1.8 ± 1.1 (Fig. 2B). CMER reflected 59.7 ± 14.5% of prescribed hydrocortisone dosages (Fig. 2A).

The forth patient (#4, female) initially showed an adequate treatment-adherence with a CMER of 5.2 mg/dL, consistent with z-scores of 0.8 and 0.8 for absolute and BSA-corrected values, respectively (Fig. 2B, green circle). CMER reflected 49.4% of the prescribed hydrocortisone dosage (Fig. 2A: green circle).

The patient later displayed clinical symptoms of PAI during treatment (marked by an arrow). Her CMER was only 0.3 mg/dL (-3.4 z), reflecting only 3.1% of prescribed hydrocortisone dosage. She admitted that she had interrupted treatment. Later controls showed somewhat better but still inadequate adherence. CMERs were 3.6 mg/dL (-0.3 z; 27.4 %) and 1.9 mg/dL (-1.6 z; 17.2%), respectively.

Conclusions

We could demonstrate that quantitative targeted GCMS steroid metabolome analysis is excellent suited to monitor glucocorticoid replacement treatment in patients with PAI. It allows assessment of treatment adherence and it helps to avoid over- and under-treatment.

References:


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