He had polyuria and polydipsia for 2 weeks before presentation. Three days before presentation, he got increasingly exhausted and developed recurrent vomiting. At presentation his weight was 41.4 kg (P 25-50) and his height was 154 cm (P 25-50).

Type-1 diabetes was diagnosed and the boy treated with i.v. insulin, fluid and electrolyte substitution. However, despite a high potassium substitution of up to 6 mmol/kg body weight / 24h, therapy-resistant hypokalemia with hypokalemic metabolic alkalosis persisted after regression of ketoacidosis: pH 7.48, base excess 10.8 mmol/l, bicarbonate 33.7 mmol/l, Na 136 mmol/l, K 2.7 mmol/l.

Further lab diagnostics showed a plasma renin of >300 ng/l and a serum aldosterone of 141.1 ng/dl. The constellation of therapy-resistant hypokalemic alkalosis and secondary hyperaldosteronism made us think of the additional presence of Bartter’s syndrome. Bartter’s syndrome type 3 is caused by pathogenic changes in the CLCNKB gene and is inherited as an autosomal recessive trait. Analysis by next generation sequencing showed a complete deletion of the CLCNKB gene in the homozygous state, which was confirmed by multiplex ligation-dependent probe amplification analysis.

To the best of our knowledge, we describe the first case of persisting hypokalemia and alkalosis after regression of diabetic ketoacidosis due to a previously undiagnosed Bartter’s syndrome. During the course of long-term therapy with spironolactone and potassium substitution, the potassium levels normalized.

The constellation of therapy-resistant hypokalemic alkalosis and secondary hyperaldosteronism with normal blood pressures made us think of the additional presence of Bartter’s syndrome. Bartter’s syndrome is a clinically and genetically heterogeneous renal salt loss disorder with hypokalemic metabolic alkalosis and secondary hyperaldosteronism. Bartter’s syndrome type 3 is caused by pathogenic changes in the CLCNKB gene and is inherited as an autosomal recessive trait. Analysis by next generation sequencing showed a complete deletion of the CLCNKB gene in the homozygous state, which was confirmed by multiplex ligation-dependent probe amplification analysis.

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