DIABETIC KETOACIDOSIS RESULTING IN TREATMENT-RESISTANT HYPOKALEMIC ALKALOSIS

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INTRODUCTION

From time to time pediatric endocrinologists may be confronted with paradoxical constellations. Here, we report on an 13-year-old-boy whose treatment of ketoacidosis at manifestation type-1 diabetes resulted in hypokalemic alkalosis.

CASE

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).

Initial labs were HbA1c 10.2%, glucose 530 mg / dl, pH 7.15, base excess - 21.1 mmol / l, bicarbonate 9.8 mmol / l, Na 131 mmol / l, K 2.4 mmol /l.

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 >300ng/l and a serum aldosterone of 141.1 ng/dl.

	pH	Bicarbonate (mMol/l)	BE (mMol/l)	K (mMol/l)
Hour 0	7,15	9,8	- 21,1	2,4
Hour 1	7,11	9,8	- 20,8	2,6
Hour 3	7,14	9,4	- 21,8	2,6
Hour 5	7,15	9,6	- 21,7	2,6
Hour 12	7,29	15,9	- 11,0	2,6
Hour 24	7,43	22,7	- 1,2	2,6
Hour 48	7,49	34,3	11	2,9
Hour 72	7,53	33,2	9,5	3,2
Hour 96	7,48	29,9	7,9	3,5

RESULTS

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.

CONCLUSIONS

To the best of our knowledge, we describe the first case of persisting hypokalaemia 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.

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