The rare cause of hypokalaemia

Sir,

We have read with interest the article by Reimann and Gross [1]. Hypokalaemia is a frequent problem among hospitalized paediatric patients also. However, in children with chronic hypokalaemia establishing the diagnosis may be difficult. The most common causes of chronic hypokalaemia in children are related to chronic diarrhoea from inflammatory bowel disease, gastrointestinal infection or laxative abuse. A specific paediatric problem is cystic fibrosis. In this autosomal recessive disease hypokalaemia is frequently seen. It is usually associated with hyponatraemia, hypochloraemia and metabolic alkalosis. It is sometimes called Pseudo-Barter’s syndrome: it must be distinguished from true Bartter’s syndrome [2]. Renal potassium wasting causing chronic hypokalaemia can also arise from primary or secondary hyperaldosteronism, diuretic use/abuse and primary tubular disorders [3]. The article by Reimann and Gross [1] provided diagnostic guidelines for chronic hypokalaemia. We would like to present a clinical case to complete these guidelines.

An 8-year-old white boy was examined by his paediatrician for fatigue, headache, occasional fever up to 38°C, poluria (2600–2900 ml/24 h) and nycturia. Clinical examination revealed no anomalies except elevated blood pressure (BP). Serum biochemistry showed hypokalaemia (2.3 mmol/l). Persistent hypertension with hypokalaemia led to a therapeutic trial with spironolactone. However, there was no improvement of BP or hypokalaemia in the course of 2 months. Therefore, the boy was referred to our department. On admission body weight was 27 kg (10–24 percentile), height 135 cm (25–50 percentile), and BP 156/98 mmHg (95 percentile). Ultrasound examination of the kidneys and renal blood vessels was normal. Table 1 shows the laboratory results on a normal diet, in the absence of spironolactone treatment. The concentrations of urinary catecholamines, 17-ketosteroids, 17-OH-corticosteroids, and plasma cortisol were normal. Glomerular filtration rate (Schwartz formula) was 81.4 ml/min/1.73 m². Urinalysis was normal, urine pH was 6.5. Urine osmolality was between 204 and 520 mmol/kg. We made the diagnosis of Liddle’s syndrome (LS) and began therapy with amiloride. A follow-up examination 2 weeks later showed complete normalization of BP, blood and urinary parameters.

LS is caused by mutations in beta or gamma subunit of epithelial sodium channel in the distal tubule (amiloride-sensitive) and has an autosomal dominant mode of inheritance. This tubular defect leads to increased reabsorption of sodium, and the subsequent stimulation of potassium secretion and hydrogen ions. Hypokalaemia, including kaliuric nephropathy, elevated BP and metabolic alkalosis are the main consequences. The first symptoms of LS can appear as early as the infant period, but they occur more frequently in older children. The presence of hypertension, which can be asymptomatic, is typical. However, it is often accompanied by fatigue, headache, paresthesia, weakness and abdominal pain. Published reports indicate a marked variability in the severity of hypertension and hypokalaemia in patients with LS [4]. This leads to the assumption that LS is underdiagnosed among patients with essential hypertension. In contrast to primary hyperaldosteronism (Conn’s syndrome), resistance to spironolactone treatment is characteristic for patients with LS. Limited sodium intake, increased potassium intake and use of amiloride is the recommended therapeutic option.

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Table 1. Abnormal laboratory findings in the patient

<table>
<thead>
<tr>
<th>Serum</th>
<th>Normal range</th>
</tr>
</thead>
<tbody>
<tr>
<td>Na⁺ (mmol/l)</td>
<td>149.3</td>
</tr>
<tr>
<td>K⁺ (mmol/l)</td>
<td>2.4</td>
</tr>
<tr>
<td>Ca²⁺ (mmol/l)</td>
<td>2.01</td>
</tr>
<tr>
<td>HCO₃⁻ (mmol/l)</td>
<td>30.3</td>
</tr>
<tr>
<td>Cl⁻ (mmol/l)</td>
<td>76.6</td>
</tr>
<tr>
<td>Blood pH</td>
<td>7.5</td>
</tr>
<tr>
<td>PA (nmol/l)</td>
<td>0.06</td>
</tr>
<tr>
<td>PRA (nmol/l/h)</td>
<td>0.135</td>
</tr>
</tbody>
</table>

PA, plasma aldosterone; PRA, plasma renin activity.

Reply

Sir,

Dolezel et al. address the causes of hypokalaemia in children. We welcome their emphasis on syndromes and presentations in a paediatric population which are different from those of hypokalaemia in adults. In particular the hypokalaemia of cystic fibrosis is intriguing. In the absence of a more detailed description we hypothesize that intestinal chloride losses during periods of metabolic stress together with secondary hyperaldosteronism and enhanced renal potassium excretion is responsible. Our diagnostic guide [1] should be applicable to this situation also, since it would probably demonstrate a low urinary chloride excretion rate.

We agree with Dolezel et al. that Liddle’s syndrome may present with hypokalaemia of some duration [2]. However, the associated hypertension, which will be even more unusual in a child than in an adult, will place Liddle’s syndrome-patients in the category of presumed mineralocorticoid excess.

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In our experience such patients are diagnosed more rapidly than the patients in our article.
In the latter, the emphasis was on disorders that are associated with normal or low normal blood pressure. It is a common observation in these patients that often a specific diagnosis of the hypokalaemia is not established for several years.

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