



## Rare cause of severe hypertension in an adolescent boy presenting with short stature: Questions

Zehra Yavas Abali<sup>1</sup> · Gozde Yesil<sup>2</sup> · Tarik Kirkgoz<sup>1</sup> · Neslihan Cicek<sup>3</sup> · Harika Alpay<sup>3</sup> · Serap Turan<sup>1</sup> · Abdullah Bereket<sup>1</sup> · Tulay Guran<sup>1</sup>

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### Case summary

A 14-year-old boy was referred to the pediatric endocrinology clinic for short stature and excessive weight gain. History revealed that weight gain started from the age of 4 years and slow growth was noticed especially over the last year. He was born at 39 gestational weeks with a birth weight of 2750 g (−1.7 SDS). His parents were not related. His father was diagnosed with hypertension at the age of 32 years and died due to intracranial hemorrhage at 37 years of age. His paternal uncle also had hypertension.

At presentation, his weight, height, and body mass index were 55.1 kg (−0.1 SDS), 143.3 cm (−2.8 SDS), and 26.8 kg/m<sup>2</sup> (+1.6 SDS), respectively. Target height was 167.6 cm (−1.2 SDS). He had acanthosis nigricans and did not have any striae or buffalo hump. Cardiac and abdominal examinations were unremarkable. His pubic hair was Tanner stage 3 and testicular volumes were 2 ml/2 ml. He had mild developmental delay and speech

disturbance. Blood pressure (BP) was 140/80 mmHg (99th percentile for age, gender, and height is 128/87 mmHg). Fasting plasma glucose (83 mg/dl), serum potassium (6.4 mmol/L), serum sodium (138 mmol/L), serum creatinine (0.48 mg/dl), and fasting lipid profile were normal at initial evaluation. Cushing's syndrome was excluded by normal 24-h free urinary cortisol and overnight dexamethasone (1 mg) test. Amlodipine treatment (10 mg/day) was initially commenced. He had no symptoms such as headache or dizziness, but the patient's BP remained high for age during this treatment. Hyperkalemia and hyperchloremic metabolic acidosis (potassium, 8.2 mmol/L; chloride, 113 mmol/L; pH, 7.28 HCO<sub>3</sub>, 17.7 mmol/L) with normal renal function (serum creatinine, 0.53 mg/dl) was detected 1 month after the amlodipine treatment. High amplitude "T" wave was observed on electrocardiography. Potassium-binding resin, calcium gluconate and inhaled salbutamol treatments were commenced to lower potassium level. Fundoscopic examination, renal doppler ultrasonography, and echocardiography were all normal. Aldosterone and renin concentrations before amlodipine treatment gave the following results: 180 pg/ml; *N*:13.3–231.4) and (< 0.01 pg/ml; *N*:0.8–16.5), respectively. Ambulatory blood pressure monitoring revealed severe hypertension.

The answer to this question can be found at <https://doi.org/10.1007/s00467-019-04352-1>.

✉ Tulay Guran  
tulayguran@yahoo.com

<sup>1</sup> School of Medicine, Department of Pediatric Endocrinology and Diabetes, Marmara University, Fevzi Çakmak Mahallesi, Muhsin Yazıcıoğlu Caddesi, No:10 34899 Pendik, Istanbul, Turkey

<sup>2</sup> Department of Medical Genetics, Bezmialem Vakıf University School of Medicine, Istanbul, Turkey

<sup>3</sup> School of Medicine, Department of Pediatric Nephrology, Marmara University, Istanbul, Turkey

### Questions

1. What is the most likely diagnosis of our patient with low renin hypertension?
2. How should this patient be managed?

## Compliance with ethical standards

**Conflict of interest** The authors declare that there are no conflicts of interest.

**Patient confidentiality** The patient's parents provided informed consent for publication of the submitted article and the results of the accompanying analyses.

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