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Question

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Case summary

A 12-year-old girl was sent to the nephrology unit for evaluation of high blood pressure. This was detected at school; she was asymptomatic. Repeat blood pressure measurements ranged between 140 and 160/110 mmHg. She was on no medications and ate a normal diet. She had had recurrent episodes of pyelonephritis in infancy and was diagnosed with complete bilateral ureteral duplication and right vesicoureteral reflux. This was surgically corrected at 10 years of age. Hypertension was confirmed in the clinic (143/93 to 174/117 mmHg).

Physical examination was normal, with no abdominal bruits or heart murmurs. There were no hypo- or hyperpigmented skin lesions, striae, or buffalo neck. Her weight was 44.7 kg (25th–50th percentile), and her height was 152 cm (10th–25th percentile).

Laboratory studies revealed sodium 140 mmol/l, potassium 2.8 mmol/l, chloride 105 mmol/l, bicarbonates 28.4 mmol/l, base excess +3.3 mmol/l, creatinine 71 μ mol/l, blood urea nitrogen 4.3 mmol/l, and glucose 5.3 mmol/l. Analysis of a 24-h urine sample revealed volume 460 ml, sodium 113 mmol, potassium 75 mmol, and creatinine 10.46 mmol. 123 I-Hippuran scintigraphy showed indirect signs of reflux nephropathy in the right kidney, with relative function of 65% left kidney and 35% right kidney, and normal global renal function but with numerous defects in the right inferior pole.

Family history was remarkable for severe hypertension in numerous members of the family (Fig. 1). Four family members had died from subarachnoid hemorrhages. The girl's father was severely hypertensive (155/100 to 165/110 mmHg), but without symptoms. His serum plasma potassium was 3.4 mmol/l, bicarbonate 27 mmol/l, chloride 108 mmol/l, and base excess +1.6 mmol/l. His daily urine potassium excretion was 36 mmol.

Questions

1. Can the reflux nephropathy explain all the clinical and laboratory abnormalities in this patient?
2. In view of the family history, which other diagnosis should be considered?
3. Which laboratory tests will help to make the correct diagnosis?
4. Which is the best treatment of her condition?

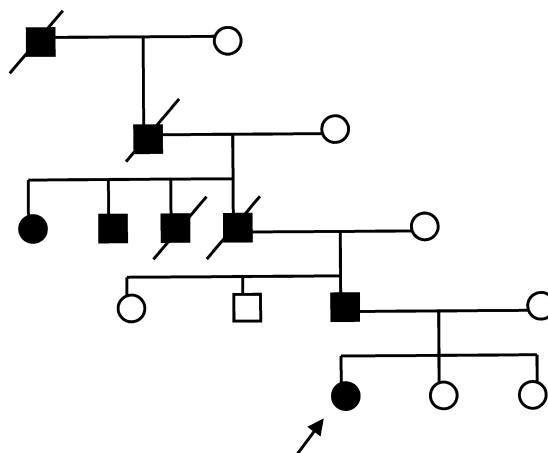


Fig. 1 Black circles or squares denote hypertension. Diagonally crossed circles or squares denote death secondary to subarachnoid hemorrhage

The answer to this question can be found at <http://dx.doi.org/10.1007/s00467-004-1437-9>

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