Hypertension in children

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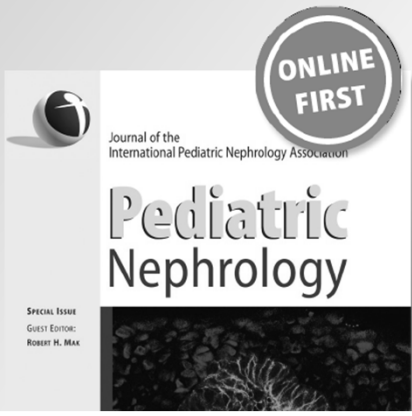


Case Report

- 17-month-old female toddler
- Presented with hypertensive encephalopathy
  - manifesting with convulsions
  - right hemiparesis and blindness.
- No family history of hypertension or kidney disease.

History

- Past: small for gestational age baby, 2 kg term, uncomplicated pregnancy
- Milestones: normal until the age of at 12 months- Not talking/playing

Clinical examination

- Her level of consciousness was depressed
- She appeared chronically ill ,Puffy eyes
- Stunted and wasted(length 72.5 cm, Z score < -3 SD, Weight for length Z score <-3 SD).
- All her pulses were normal and there was no radio-femoral delay.

Clinical examination

- BP was 148/92 mm Hg and was elevated in all four limbs
- Bulging precordium , displaced apex
- No murmur over the heart, the subclavian or carotid vessels, over the inter-scapular area or abdominal aorta.
- Loss of vision, but her other cranial nerves were intact.
- Fundoscopy revealed retinal haemorrhages, but no papilloedema

Laboratory investigations

- Hypokalaemic metabolic alkalosis (serum potassium 2.8 mmol/l, bicarbonate 30 mmol/l),
- Suppressed plasma immuno-reactive renin <1.8 mIU/l
- Low aldosterone level <31 pmol/l
- U- potassium - 21.7 mmol/l
- U-sodium <10 mmol/l
- U-chloride 20 mmol/l.

Laboratory investigations

- Steroid chromatogram- excluded endocrine causes of hypertension and hypokalaemic metabolic alkalosis.
- Genetic studies for ENaC: Heterozygous for βT594M mutation.

Imaging

- Kidney sonography: normal sized kidneys with decreased cortico-medullary differentiation.
- Renal artery stenosis was excluded on renal Doppler ultrasound.
- CT scan brain: mild cerebral atrophy.

DDX

- Differential diagnosis of severe hypertension and hypokalaemia
  - Hyperaldosteronism
  - Renal artery stenosis
  - Glucocorticoid – remediable aldosteronism (GRA)
  - Liddle syndrome
  - The syndrome of apparent mineralocorticoid excess

Treatment x

- Amlodipine
- Enalapril
- Spironolactone
- Prazosin
- Atenolol
- Hydrochlorothiazide
- Potassium Chloride

Challenges

- Polypharmacy
- Poor compliance
- Amiloride is not registered

Final diagnosis

- Liddle syndrome

Further treatment

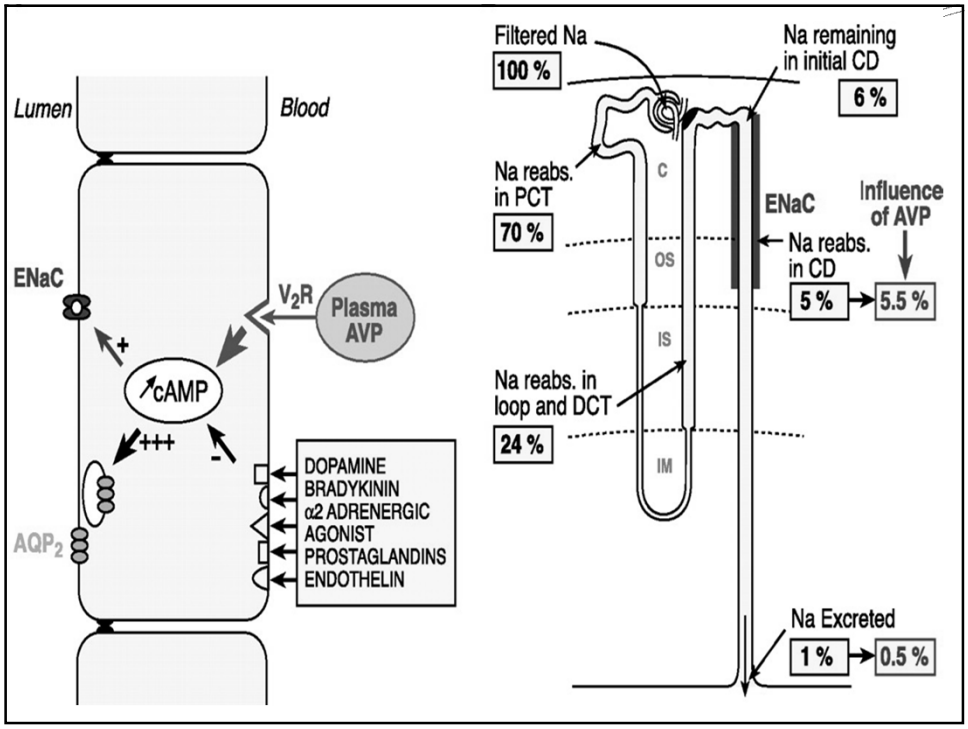
- Amiloretic (a combination of amiloride and hydrochlorothiazide) was obtained
- BP and serum K+ normalized within a week
- Other drugs withdrawn
- Current treatment: amlodipine + Amiloretic

Liddle syndrome

- Rare autosomal dominant disorder
- First described in 1963 by Grant Liddle
- Early onset of hypertension associated
  - hypokalaemic metabolic alkalosis
  - volume expansion
  - reduced plasma rennin activity
  - Low aldosterone level

Liddle syndrome

- Caused by hyperactivity of the amiloride-sensitive epithelial sodium channel (ENaC)
- ENaC is expressed in the distal connecting tubules and cortical collecting tubules of the nephron where it stimulates reabsorption of sodium.
- ENaC expression is regulated by mineralocorticoids and glucocorticoids



ENaC

- ENaC is highly selective for Na+ and the channel pores are blocked by K+ sparing diuretic- Amiloride
- Several mutations that increase the activity of ENaC has been described in the literature e,g. C618F an A663T in the C-terminal tail of the α subunit, **T594M** mutation in the in the C-terminal tail of the β subunit

Treatment

- Salt restricted diet
- Amiloride

Hypertension in children

- Hypertension is defined as systolic or diastolic BP above the 95th centile for age, gender and height.
- Any child with a BP exceeding the 90th percentile requires scrutiny
- Physiology matters
- BP is determined by the balance between cardiac output and vascular resistance

Goals

- History taking and clinical examination
- Assess the presence and severity of target end organ damage
- To define the aetiology

Age and hypertension

Infants

- Thrombosis of renal artery or vein
- Congenital renal anomalies
- Coarctation of aorta
- Bronchopulmonary dysplasia
- Drugs- corticosteroids

Age and hypertension

1-5 years

- Renal artery stenosis
- Reflux nephropathy
- Glomerulonephritis
- Wilms tumor
- Monogenic hypertension
- Neuroblastoma
- Coarctation of aorta

Age and hypertension

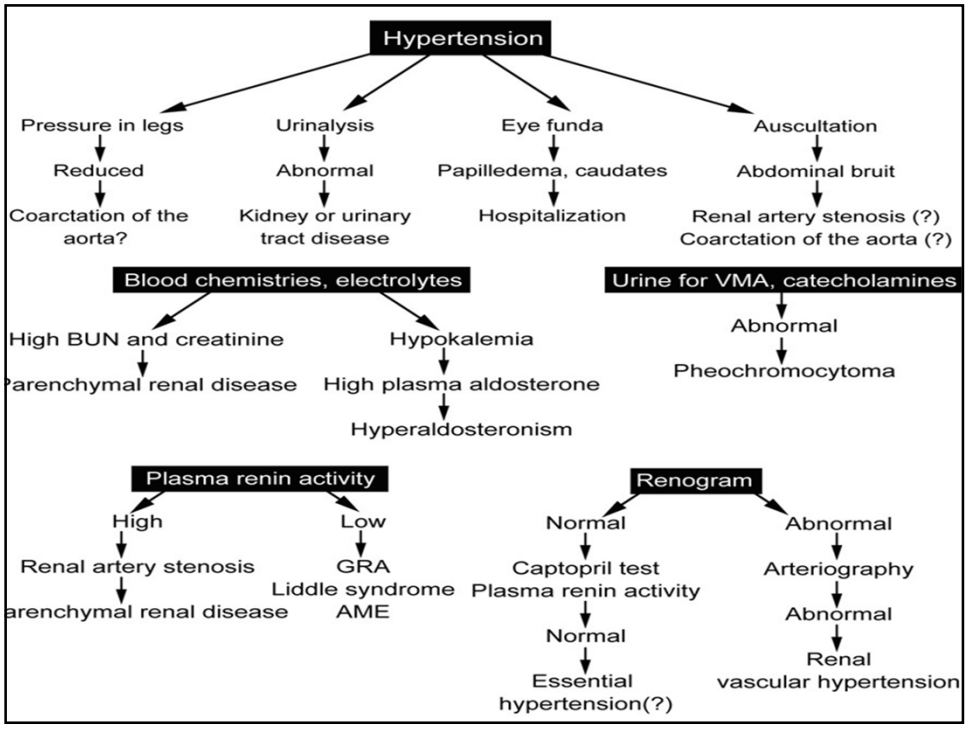
5-10 Years

- Renal parenchymal disease
- Renovascular abnormalities
- Endocrine causes- neuroblastoma, pheochromocytoma
- Essential hypertension

Age and hypertension

>10 years

- Obesity
- Essential hypertension
- Reflux nephropathy
- Glomerulonephritis
- Renal artery stenosis
- Drugs
- Endocrine tumours



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