Transient Pseudohypoaldosteronism (PHA) as a Complication of Infective Obstructive Uropathy in Infancy, a Case Series

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Transient PHA
- Renal resistance to aldosterone
- May be due to renal disease or medications
- Mainly in infants with obstructive uropathy or urinary tract infections

Genetic causes for persistent PHA
- AD mutations of mineralocorticoid receptor
  - Limited to kidney
- AR mutations of epithelial sodium channels
  - Systemic involvement: colon, sweat & salivary glands

Presentation
- Age: 1 2 3
- Sex:
- Seizures: 5m 4m 17d
- Looks "ill":
  - 1 + + +
- Vomiting + + +
- Pyrexia + + +
- Poor feeding + + +

Admission Bloods
- Sodium (mmol/L):
  - 1 113
  - 2 118
  - 3 123
- Potassium (mmol/L):
  - 1 6.0
  - 2 6.3
  - 3 8.0
- Urea (mmol/L):
  - 1 4.0
  - 2 6.0
  - 3 7.7
- Creatinine (mmol/L):
  - 1 21
  - 2 46
  - 3 41
- CRP (mg/L):
  - 1 100
  - 2 87.9
  - 3 58
- Neutrophilia:
  - 1 +++++
  - 2 ++
  - 3 ++

Acute USS
- 1 Infected urinary ascites secondary to renal tract rupture
- 2& Hydronephrosis
- 3 Obstructive infected renal tract

Micro/Virology
- 1 RSV +ve
- 2 Nil isolated
- 3 Urine Group B Streptococcus

Endocrine Studies
- Random Cortisol (µg/L):
  - 1 4186
  - 2 1749
  - 3 805 (T0)
- Short Synacthen Cortisol (µg/L):
  - 1 2348 (T30)
- Urinary Steroid Profile:
  - Normal
- Renin (pmol/L):
  - 1 240
  - 2 64000
  - 3 266.7
- Aldosterone (pmol/L):
  - 1 (0.5-4.5)
  - 2 (100-800)
  - 3 (<800)
- Aldosterone/PRA Ratio

Management
- Resolution of hypotension by day of treatment:
  - 1 138
  - 2 137
  - 3 141
- VUR (RFL):
  - 1 Normal
  - 2 Normal
  - 3 Normal

Follow Up
- Trimethoprim (Prophylactic):
  - 1 Yes
  - 2 Yes
  - 3 Yes

Differential Diagnoses
- SIADH
- Congenital Adrenal Hyperplasia
- Pyloric Stenosis
- PHA- genetic causes

Learning Points
- Consider Transient PHA in infants presenting with salt wasting, ill, febrile, FTT, dehydrated, once CAH is excluded
- Important tests include:
  - urine culture → renal USS
  - Urinary sodium
  - Serum aldosterone at presentation and follow up
- Chronic ↑Na corrects gradually with fluid and treatment of underlying cause
- Multidisciplinary Paediatric Team working- General, Surgery, Endocrine