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**[3] 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

	1	2	3
Age/Sex	5m	4m	17d
No Seizures			
Looks "ill"	+	+	+
Vomiting	+		
Pyrexia	+	+	+
Poor feeding	+		+

All born at term following normal antenatal scans, to non-consanguineous parents with no relevant family history

Admission Bloods

	1	2	3
Sodium (mmol/L)	113	118	123
Potassium (mmol/L)	6.0	6.3	8
Urea (mmol/L)	4.0	6.0	7.7
Creatinine (umol/L)	21	46	41
CRP (mg/L)	100	87.9	58
Neutrophilia	++++	++	++

Acute USS

1 Infected urinary ascites secondary to **renal tract rupture**

2& 3 **Hydronephrosis** **Obstructive infected** renal tract

Micro/Virology

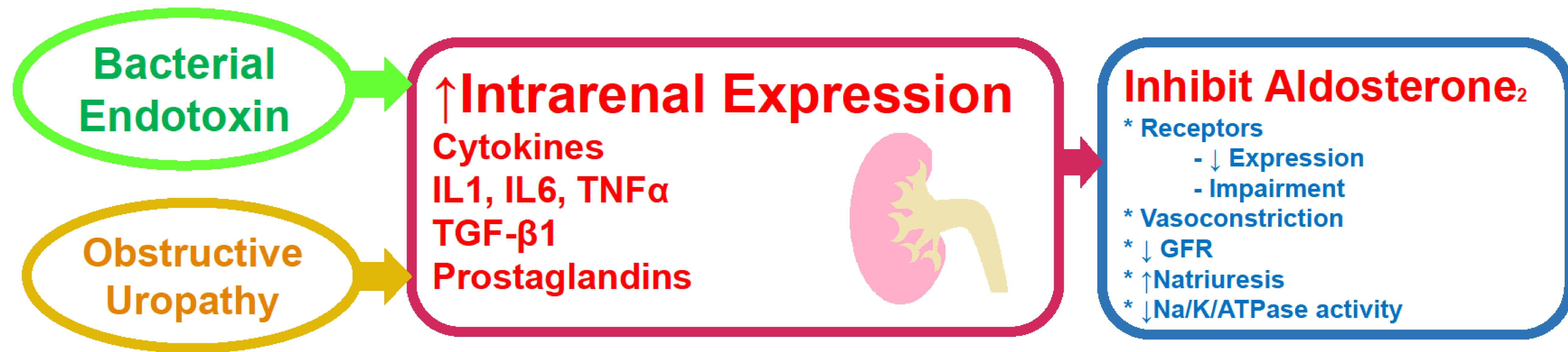
1 **RSV** +ve
Urine **Mixed coliforms**
Perinephric fluid **Citrobacter koseri**

2 Nil isolated

3 Urine **Group B Streptococcus**

Differential Diagnoses

SIADH
Congenital Adrenal Hyperplasia
Pyloric Stenosis
PHA- genetic causes[1]



Endocrine Studies

	1	2	3	
Random Cortisol (nmol/L)	4186	1749		
Short Synacthen (Cortisol, nmol/L)			805 2348	(T0) (T30)
Urinary Steroid Profile	Normal			
Urinary Sodium not consistently done				
Renin (pmol/ml/hr)			240	(0.5-4.5)
Aldosterone (pmol/L)			64000	(100-800)
Aldosterone/PRA Ratio			266.7	(<800)

Management

	1	2	3
Emergency Resuscitation	+	+	+
Empyrical antibiotics	+	+	+
Salbutamol for asymptomatic ↑K+			+
Covered with **steroids**			+
Management of Obstructive Uropathy			
-Conservative		+	+
-Surgical	+		

Follow Up

	1	2	3
Resolution of hyponatraemia	138	137	141
by day of treatment	7	2	2
MCUG	VUR (R+L) Grade 2-4	Normal	Normal
Trimethoprim (Prophylactic)	Yes	Yes	Yes

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**

54th Annual Meeting 1-3 October ESPE 2015 BARCELONA European Society for Paediatric Endocrinology

1. Bowden *et al*, Autosomal dominant PHA1 in an infant with salt wasting crisis assocaited with UTI and obstructive uropathy. Case Reports in Endocrinology. 2013
2. Bogdanovic et al, Transient Type 1 PHA: a report on an 8 patient series and literature review. Paed Nephrology. 2009
3. Kostakis *et al*, Syndromes of impaired ion handling in the distal nephron: PHA and familial hyperkalaemic hypertension. Hormones 2012

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DOI: 10.3252/pso.eu.54espe.2015
Poster presented at:

PosterSessionOnline.com