Urosepsis or Pseudohypoaldosteronism in a Neonate?

TEL AVIV UNIVERSITY

Sheba - Academic Medical Center Hospital

Noah Gruber[1,2], Einat Lahav[2,3], Reut Kassif-Lerner[1,2], Orit Pinhas-Hamiel[1,2]

[1]Pediatric Endocrine and Diabetes Unit, Edmond and Lily Safra Children's Hospital, Chaim Sheba Medical Center, Tel-Hashomer, Israel. [2]Sackler School of Medicine, Tel-Aviv University, Israel. [3]Pediatric Nephrology Unit, Edmond and Lily Safra Children's Hospital, Chaim Sheba Medical Center, Tel-Hashomer, Israel

Background and Objective

Pseudohypoaldosteronism is a disorder of impaired renal electrolytes re-absorption and excretion. It can be divided to:

1. **Primary, which has two clinically and genetically distinct forms:**
   - **A. Renal autosomal dominant form, which involves a mutation in the mineralocorticoid receptor.**
   - **B. Systemic autosomal recessive form, which involves a mutation in the epithelial sodium channel.**
2. **Secondary, which can be transient secondary to urinary tract infections/malformations.**

- **We present a neonate with a salt-wasting crisis and emphasize the nature of secondary pseudohypoaldosteronism and the need for long-term follow-up.**

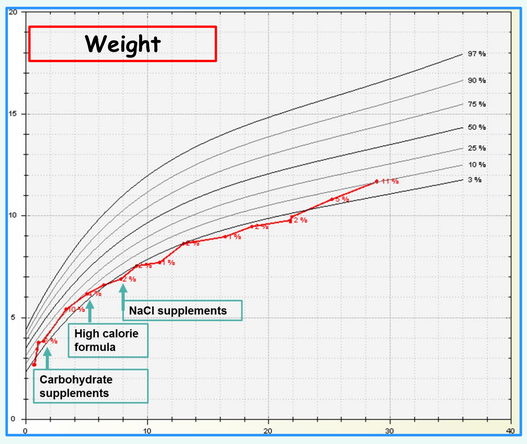
Case Presentation:

- **A 17 day-old male presented with a two-day history of poor feeding, irritability and dyspnea.**
- **Physical examination - a baby in septic shock: pale-grey looking, grunting, dehydrated, apathic and with poor perfusion.**
- **Labs - Severe hyponatremia, hyperkalemia, metabolic acidosis, acute renal failure and elevated urine Na/K ratio. Cortisol and androgens levels were normal. Extremely high aldosterone (27-fold higher than upper normal limit) and high plasma renin activity (Table 1). Microbiology cultures were positive for E.coli bacteremia and urinary tract infection (UTI). For further evaluation, sweat test was performed, to exclude the systemic type of pseudohypoaldosteronism, and was normal. Parental aldosterone and renin levels were normal. Genetic whole exome – pending.**

Table 1: *Initial laboratory results*

Na - 121 mEq/l	Urinary Na - 76 mEq/l	Cortisol - 1630 nmol/l
K - 9.2 mEq/l	Urinary K - 21 mEq/l	Aldosterone - 109,000 pmol/l
Glucose - 91 mg/dL	pH - 6.6	Renin activity >50 ng/ml/hr
Cr - 4.2 mg/dL	pCO2 - 23.6 mmHg	
Urea - 267 mg/dL	HCO3- 2.2 mmol/l	

- **Imaging - Renal ultra-sound was normal; cystogram showed vesico-urethral reflux.**
- **Differential diagnosis: renal and system types of pseudohypoaldosteronism, congenital adrenal disorders and Barter's syndrome.**
- **Management –**
  - **Acute - The baby was treated intensively in the PICU with intravenous fluids, electrolyte and acid-base corrections, antibiotics and hydrocortisone.**
  - **Long-term - Due to failure to thrive (Fig. 1) and persistently high aldosterone and renin levels, despite normal electrolytes, we added salt supplements. Weight gain improved, and finally, only at the age of 2 years aldosterone and renin levels returned to normal (Fig. 2).**



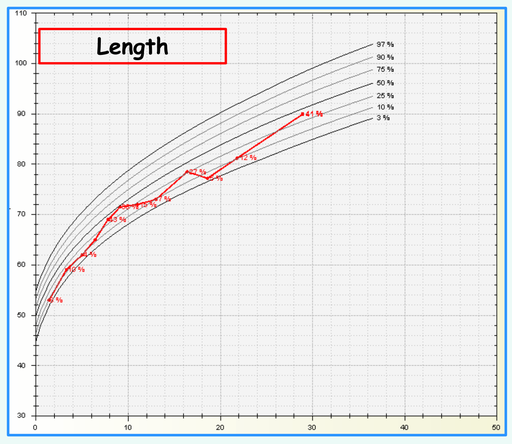


Figure 1: *Weight and length growth charts*

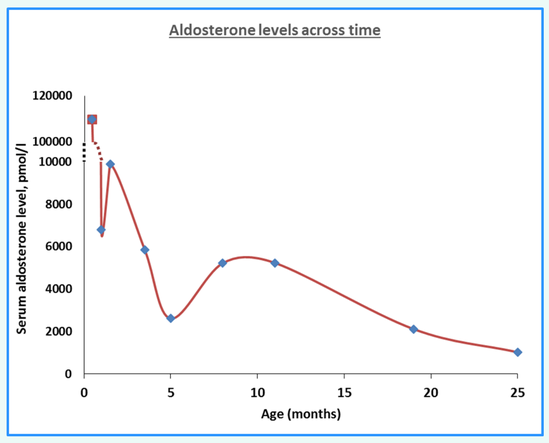


Figure 2: *Aldosterone levels across time. Normal values at birth are up to 4000 pmol/l and at age 2 years up to 1200 pmol/l.*

Conclusion: **We present a case of secondary pseudohypoaldosteronism due to E.coli urosepsis in a baby with vesico-urethral reflux. It emphasizes how secondary pseudohypoaldosteronism is part of the differential diagnosis of a salt-wasting crisis. Aldosterone and renin levels continue to be elevated for a long period, therefore a long-term follow-up is needed.**

55ESPE 87--P2

Adrenal

Noah Gruber

DOI: 10.3252/pso.eu.55ESPE.2016

Poster presented at: ESPE 2016 PARIS 10-12 SEPTEMBER

PosterSessionOnline.com