



A case of  
DYSELECTROLYTEMI A

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# CASE SUMMARY

- 4 month old, female infant
- 1<sup>st</sup> born to NC parents, term, b.wt: 3.25kg
- No neonatal hospitalization
- Attained head control
- Weight 6.1 kg

h/o loose stools

decreased urine output

decreased activity

# AT ADMISSION [18-4-12]

- Ill looking, signs of dehydration
- Normal blood pressure [ 101/50 (71) ]
- Normal systemic examination, Dx- 121
- Normal external genitalia

# INVESTIGATIONS

- TC 13300 [ N 59, L38, M 03 ]
- Hb 11.4
- RBS 99
- Urea 71
- S. creatinine 1.1
- Na 109
- K 6.5
- Cl 81
- HCO<sub>3</sub> 14
- Ca 1.14
- Anion gap 15

## Appropriate IVF to correct dehydration

- Urea 71 → → 54 → 37
- Creat 1.1 → → 0.9 → 0.4
- Na 109 → 128 → 135 → 135
- K 6.5 → 4.4 → 3.7 → 3.3
- HCO<sub>3</sub> 14 → 15 → 9 → 15

# FURTHER EVALUATION

- S. cortisol 0.14 [ 0.5- 1.2 ]
- S. 17 OH progesterone 9.0 [ 10- 240 ]

- USG abdomen was normal

normal kidneys

normal uterus and ovaries

DISCHARGED AS

HYPONATREMIC DEHYDRATION

## One month later 22-5-12

- loose stools
- Vomiting
- Signs of some dehydration
- Weight - 6.2 kg

## INVESTIGATIONS

- RBS 121
- B. urea 78
- Creatinine 2.0
- Na 110
- K 7.9 [ tall T waves in ECG ]  
Ca gluconate, k bind,  
salbutamol nebulization
- cl 76
- Hco<sub>3</sub> 14



4 month old infant with history of recurrent episodes

- Dehydration
- Prerenal azotemia
- Acidosis
- Hyponatremia
- Hyperkalemia
- ? CAH / ? ADDISON'S / MINERALOCORTICOID DEFICIENCY
-

Endocrinologist and Nephrologist opinion was sought

Urine electrolytes

- sodium 65
- Potassium 35.3
- Chloride 95
- Bicarbonate 2.2
- pH 6.0

● SERUM OSMOLALITY 280

● URINE OSMOLALITY 360

# ACTH STIMULATION TEST

8.00 am : Cortisol 206 [10 - 240 ]

8.15 am : Inj. Synactin IV

9.00 am : Cortisol 367 [10 - 240 ]  
17 OH progesterone 1.31 [0.5 - 1.2 ]

FREE T4, TSH normal

- Plasma renin activity
- Serum Aldosterone levels sent
- Mantoux
- Chest X ray



- STARTED ON ORAL FLUDROCORTISONE
- DISCHARGE DIAGNOSIS
- SUSPECTED MINERALOCORTICOID DEFICIENCY

<1 week 28-5-12

- Vomiting
- Decreased activity
- Signs of some dehydration

# INVESTIGATIONS

- RBS 171
- B. urea 73
- Creatinine 1.9
- Na 117
- K 8.9 [ tall T waves in ECG ]  
Ca gluconate, k bind, salbutamol nebulisations
- cl 79
- Hco3 14

## MOST AWAITED RESULTS

- Plasma rennin activity - > 37
- Serum aldosterone level - > 250  
[5-90]



- FINAL DIAGNOSIS

- PSEUDOHYPOALDOSTERONISM



# TREATMENT

- STOP Fludrocortisone
- Common salt supplementation
- K - bind
- Regular electrolyte monitoring

# AT REVIEW

DATE	31-5-12	11-6-12	25-6-12	25-7-12
SODIUM	135	138	137	135
POTASSIUM	4.7	4.8	4.5	4.4

# DISCUSSION

## Electrolyte of concern – K<sup>+</sup>

- SPURIOUS LABORATORY VALUE
- INCREASED INTAKE
- TRANSCELLULAR SHIFTS
- DECREASED EXCRETION
  - Renal failure
  - Primary adrenal disease
  - Hyporeninemic Hypoaldosteronism
  - Renal tubular disease
  - Medications

# Associated with Hyponatremia

- Primary Adrenal disease [ congenital/ acquired ]  
CAH

Most common is 21-OH deficiency in male,  
girl- ambiguous genitalia

- Adrenal insufficiency : Adrenal hypoplasia  
Adrenoleukodystrophy

- Renal tubular diseases  
Hypoaldosteronism [ RTA IV ]  
Aldosterone resistance

## TTKG [TRANS TUBULAR POTASSIUM GRADIENT]

- Evaluate renal response to hyperkalemia
- To differentiate decreased K<sup>+</sup> excretion from other etiologies

$$\frac{[K] \text{ urine}}{[K] \text{ plasma}} \times \frac{\text{plasma osmolality}}{\text{urine osmolality}}$$

- TTKG > 10 - Renal excretion is normal
- TTKG < 8 - Defect in excretion of potassium  
[deficiency or resistance of aldosterone]

# PSEUDOHYPOALDOSTERONISMO

- Incidence of PHA 1 in 80000
- Incidence of CAH 1 in 15000

# PSEUDOHYPOALDOSTERONISM

- Type I PHA

AD - renal Type 1 PHA

spontaneous remissions

AR - Systemic Type 1 PHA

defects in salt reabsorption in other organs

lungs (mimic cystic fibrosis)

cholelithiasis, Meibomian glands,

skin and placenta

- Type II PHA

Gordon syndrome

familial hyperkalemia and hypertension

only salt retending variety

low doses of thiazide diuretics

- Type III PHA

transient resistance

secondary to nephropathies

obstructive uropathy or UTI

main characteristic of this type of PHA is a  
decreased GFR



# MANAGEMENT

- Initial Supportive Measures

Dehydration:

Fluid boluses

(Ensure that, IVF contain no potassium)

Hyperkalemia

- Appropriate nephrologist and endocrinologist consult.

○ Dietary sodium supplementation

○ Potassium restriction

○ Potassium-binding resins

○ Alkalinizing agents ( sodabcarb / citrate )

○ Prostaglandin inhibitors

Indomethacin ( dec Na excretion)

some patients may not benefit

○ Hydrochlorothiazide (in PHA type II )

## POINTS TO REMEMBER

- DEHYDRATION WITH LETHARGY  
Try to preserve a blood sample at admission  
Will be useful to evaluate dysectrolytemia
- Urine electrolytes will help in finding out etiology.