A rare case of an infant with dehydration

4 month old male infant was brought by mother,reliable history

With complaints of Poor activity for 2 weeks,Fever for 10days &Seizure 1episode

Child was apparently normal 2weeks back The mother noticed decreased activity of the child with poor feeding .After two days child developed fever, low grade ,on and off, not associated with chills or rigorsChild had one episode of seizure – upward gaze with tonic posturing of upper limbs, lasted for few seconds , regained consciousness after 15min

No H/o vomitings,diarrhoea, head injury,ear discharge, oliguria,facial puffiness polyuria,SOB or jaundice. Baby was Born to 3rd degree consanguineous parents 1st in birth order Born through LSCS at term Birth weight 2.8 kgs.Immunization History was complete. Developmental History:

Attained partial neck holding

Family History: No family h/o seizures

General Examination: Child is dull , afebrile , dehydrated, pallor, icterus,clubbing, cyanosis, lymphadenopathy, pedal edema,No dysmorphic facies/ hyperpigmentation

AF is flat, Genitalia normal.

Vitals:pulse rate - 110/ min , normal volume, RR- 36/ min , BP- 90/60 mmhg pulses felt in all 4 limbs

Anthrapometry: weight – 4.5 kgs(< 3rd percentile). length – 57 cm( <3rd percentile) Head circumference – 39cms( < 3rd percentile), Chest circumference – 38cms

CVS: S1 , S2 heard, no murmurs , RS: Bilateral air entry equal, clear,P/A : Sof liver is palpable 3cm below right costal margin,spleen not palpable ,CNS: Normal No signs of meningeal irritation.No cranial nerve palsies.GRBS : 90 Hb :10.6gm% RBC counts :4.16 mill/cumm WBC :17,300cells/cumm ,N 74%, L20%,E 04%,M 02%, B 00%,Platelet count :2.0 lakhs/cumm,CRP : 12.5 .Renal function tests.Sr creatinine 0.5,Bld urea 15,PT 14 seconds,APTT 31 seconds ,INR,Liver Function Tests,Bilirubin total 0.8,conjugated 0.2.,unconjugated 0.6.Alikaline phosphotase 805 mg/gl.S.G.P.T 408U/L.,S.G.O.T 847U/L,Total protein 6.1gms,Albumin 3.8gms,Globulin 2.3 gms,A/G Ratio 1.6%

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| **Serum electrolytes**  **mmol/L** | Day 1 | Day 2 | Day 3 |
| Sodium  (135-145) | 123 | 125 | 124 |
| Potassium  (3.5 – 5) | 1.8 | 1.7 | 1.4 |
| Chloride  (95-105) | 60 | 80 | 86 |

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| **ABG** | **20-01-2014** | **23-01-2014** | **25-01-2014** |
| PH  (7.35 – 7.45) | 7.636 | 7.64 | 7.70 |
| Pco2  (32 – 48) | 42.7 | 35.6 | 30.6 |
| Hco3 | 46.1 | 39.2 | 38.3 |
| Anion gap | 18.7 | 14.7 | 10.6 |

Sr calcium – 8.8,Sr magnesium – 0.9 ( 0.6 -1.2),24 Hrs urinary electrolytes,Urine output – 350 ml,Urinary sodium – 31 mmol/day (40 – 220),Urinary potassium,3.8mmol/day (25 – 125),Urinary chloride – 35 mmol/day (110 – 250),Urinary calcium – 24 mg/ day (less than 300mg/day),USG abdomen – Mild hepatomegaly ,NSG – Striate vasculopathy ,CT Brain – Diffuse cerebral atrophy

In consultation with the nephrologist, we have come to a conclusion of Bartter syndrome, as there was hypokalemic, hypochloremic metabolic alkalosis with normal blood pressure. Serum Renin and Aldosterone levels could not be sone due to financial constraint.Baby was treated with Potassium supplements,Spironolactone for 15 days, indomethacin was planned at a later date .

Bartter syndrome,Bartter syndrome, originally described by Bartter and colleagues in 1962, represents a set of closely related, autosomal recessive renal tubular disorders characterized by hypokalemia, hypochloremia, metabolic alkalosis, and hyperreninemia with normal blood pressure. The underlying renal abnormality results in excessive urinary losses of sodium, chloride, and potassium.

SYMPTOMS

Polyuria,Polydipsia,Vomiting,Constipation,Salt craving,Tendency for volume depletion,Failure to thrive,Linear growth retardation Other symptoms, which appear during late childhood, include fatigue, muscle weakness, cramps, and recurrent carpopedal spasms.CLINICAL FEATURES:Patients are thin and have reduced muscle mass and a triangularly shaped face, which is characterized by a prominent forehead, large eyes, protruding ears, and drooping mouth. Strabismus is frequently present. Blood pressure is within the reference range.

A subset of patients with Bartter syndrome (types IV) develop sensorineural deafness, which is detectable with audiometry

MANAGEMENT: DIET: Adequate salt and water intake is necessary to prevent hypovolemia, and adequate potassium intake is essential to replace urinary potassium losses. Patients should consume foods and drinks that contain high levels of potassium (eg, tomatoes, bananas, orange juice). MEDICAL MNG: Sodium and potassium supplements - Used for the electrolyte imbalances.Aldosterone antagonists and diuretic spironolactone - Are mainstays of therapy.Angiotensin-converting enzyme (ACE) inhibitors - Used to counteract the effects of angiotensin II (ANG II) and aldosterone.Indomethacin - Used to decrease prostaglandin excretion,Growth hormone (GH) - Used to treat short stature,Calcium or magnesium supplements.