A child with polyuria and polydipsia

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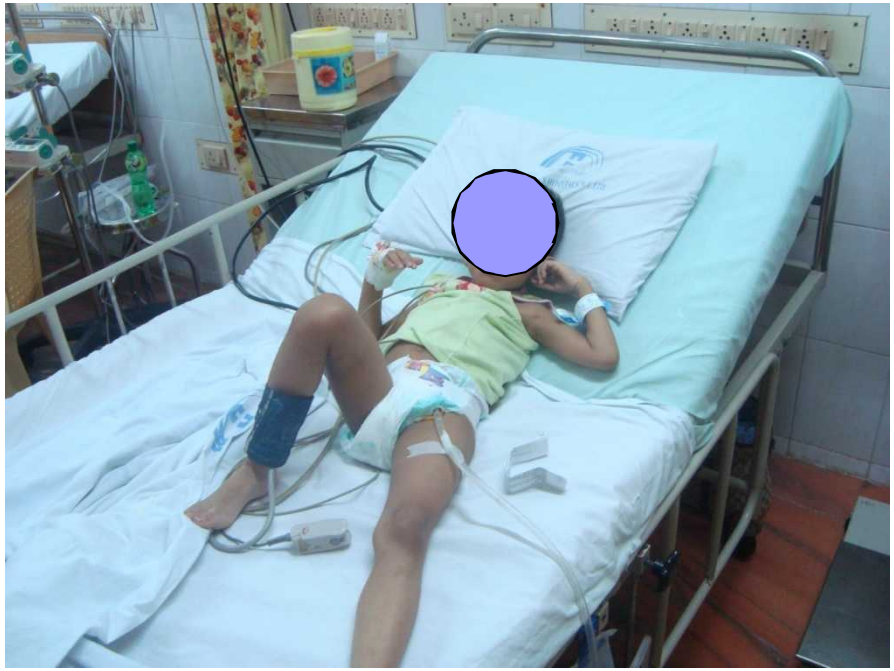
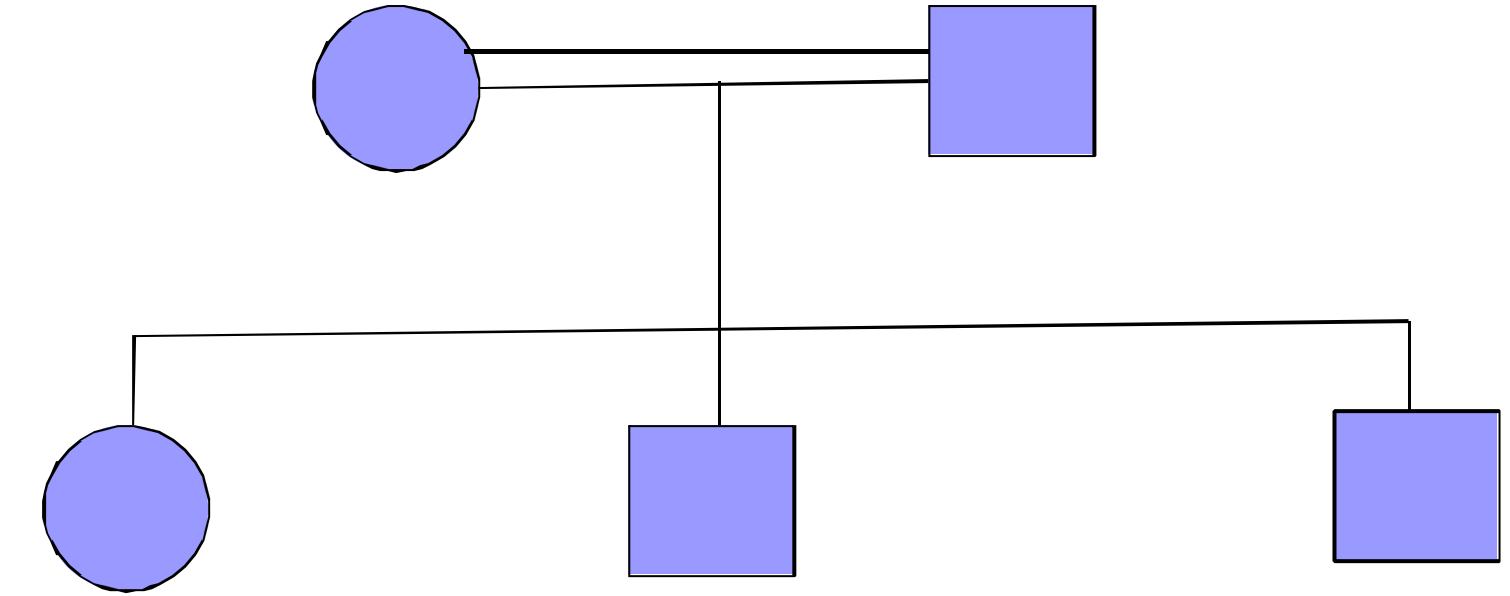
- 4 ½ yr old girl brought for polydipsia, polyuria, not gaining weight-past 2 ½ yr
- Never had features of acidosis/neurological manifestations



FAMILY HISTORY



Present Child

Male,died in newborn period due to acidosis, IEM work up-inconclusive

Male with vit-D deficiency rickets

- No family H/O diabetes mellitus/endocrine disorder

Examination

- No dysmorphic features/thyroid enlargement/ acanthosis nigricans/features of poly endocrine disorder
- No midline defects. HR, RR, BP, Normal
- Weight:< 3 rd centile, Height:3 rd centile
- Systemic examination-not significant except distended bladder
- U.O : 3-4 L/day
- Fundus: Normal

Evaluation

- Features of type 1 DM- glucose-165 mg/dl
  Increased HbA1c (8.8%)
  Low c-peptide level (0.53)
  Urine sugar ++++
  GAD Negative
- Odd points-long history, no DKA
  marginal lab abnormalities
  mild hyperglycemia
  low insulin requirement

- Started on insulin
- Requirements were very low (0.6 units/kg/day)

- Symptoms did not resolve (polyuria)

Other Investigations

- Serum Na+-144 meq/l
- Serum osmolality-303
- Urine osmolality-61
- Calcium, Urea, Creatinine: Normal
- USG: Bilateral hydroureteronephrosis

- Other causes of polyuria-Psychogenic polydipsia
  diabetes insipidus
  renal tubular defects
  hypercalcemia
  hypokalemia
  drugs

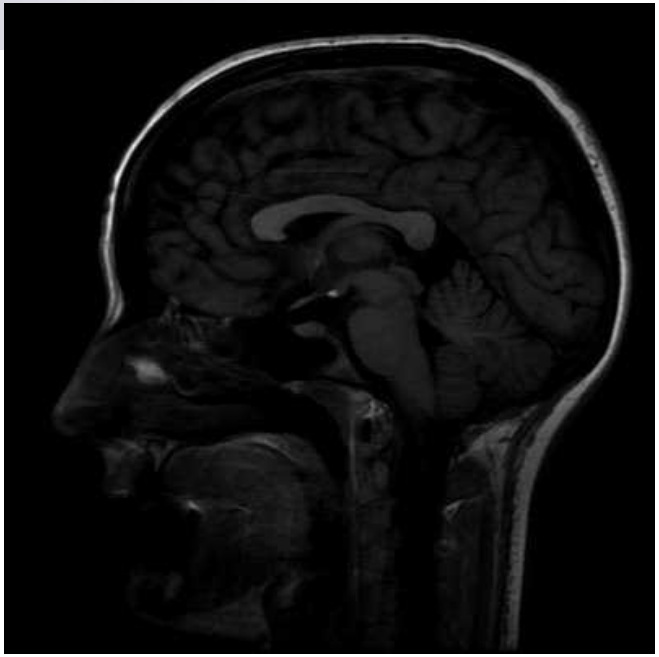
WATER DEPRIVATION TEST

Tanmika

4 ½ yrs. F

s.no	time	weight kg	HR	RR	BP	U.O	s.osm	s.sodium	u.osm	CBG	
1.	10·30	12·3	126	24	114/78	300 ml (voided)					
2.	11 am	12·1	125	22	111/75	Catheterized ~~350 ml~~	303	144	61		
3.	12 noon	11·6	132	24	136/99	350 ml	298.	142	56	187	Se. ADH levels
4.	1 pm	11·2	124	24	128/78	350 ml	307	149	57.	90	
5.	2 pm	11·2	121	26	111/76	260 ml	Inj. Vasopressin		6 U sc	178	Food-80 water-100 ml
6.	3pm	11·2	117	24	100/73	20ml.	313	149	126	228.	
7	4pm	11·1	124	26	122/89	35ml	321	150	217	195	
8	5pm	11·1	117	24	101/69.	60ml	318	153	243.	154	H20 50
					109/80	30ml	322	152.	224	122	50ml H

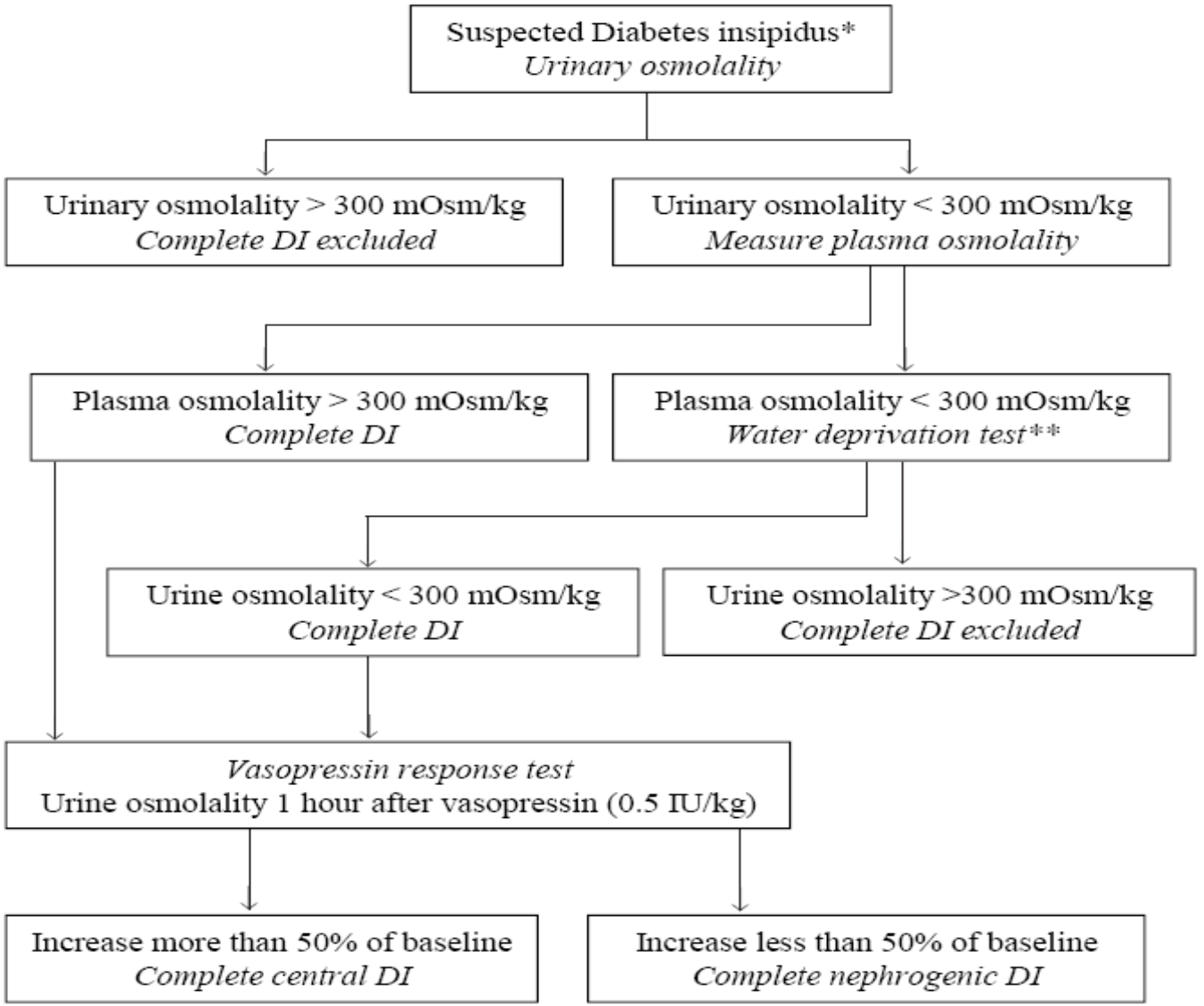
Further evaluation..



- MRI brain- normal
- Fundus - Normal
- Low serum ADH level: 2.4 pmol/L (Normal – 13 pmol/L)

- Confirmed central DI and started on intra nasal desmopressin (10 mcg BD)
- Objective: Good sleep in the night, One hour of diuresis
- Follow up-optic atrophy,hearing loss
- Plan-genetic studies

  hearing evaluation


Suspected Diabetes insipidus*
Urinary osmolality
Urinary osmolality > 300 mOsm/kg
Complete DI excluded
Urinary osmolality < 300 mOsm/kg
Measure plasma osmolality
Plasma osmolality > 300 mOsm/kg
Complete DI
Plasma osmolality < 300 mOsm/kg
Water deprivation test**
Urine osmolality < 300 mOsm/kg
Complete DI
Urine osmolality >300 mOsm/kg
Complete DI excluded
Vasopressin response test
Urine osmolality 1 hour after vasopressin (0.5 IU/kg)
Increase more than 50% of baseline
Complete central DI
Increase less than 50% of baseline
Complete nephrogenic DI

Essentials features of central DI

- U.O (> 2 L/m2/d), dehydration, and hypernatremia.
- Inability to concentrate urine after fluid restriction (urine specific gravity < 1.010; urine osmolality < 300mOsm/kg).
- Plasma osmolality > 290 mOsm/kg with urine osmolality< 300 mOsm/kg.
- Low plasma vasopressin with antidiuretic response to exogenous vasopressin

Causes of central DI:

- Genetic (autosomal dominant)
- Acquired

  trauma (surgical or accidental)

  congenital malformations

  neoplasms

  infiltrative,autoimmune,and infectious causes

  drugs-phenytoin,halothane

Desmopressin (dDAVP)

- Analog of vasopressin
- Intra nasal(onset 5-10 min)

  Delivers 10 mcg (0.1 ml) per spray

  To prevent water intoxication,patients should have at least 1 hr of urinary breakthrough between doses each day
- Tablet (onset 15-30 min)

  25-300 mcg,every 8-12 hr

Management of infants with DI

- Can present with irritability,failure to thrive, constipation and intermittent fever
- Should not be treated with DDAVP
- Can cause water intoxication.
- treated with extra free water, to maintain normal hydration.
- A formula with a low renal solute load and chlorothiazides may be helpful

Wolfram syndrome

- First described in four siblings in 1938 by Dr. Don J. Wolfram
- Also called **DIDMOAD** (Diabetes Insipidus, Diabetes Mellitus, Optic Atrophy, and Deafness)
- Rare disease with wide variety of CNS and renal abnormalities
- Autosomal recessive trait affects males and females
- Chromasome 4p, ? Mitochondrial

Symptoms

- Type 1 Diabetes, usually starting between ages 5 and 15.
- Visual impairment, starting with wearing glasses,colour blindness but rapidly increasing.(slow reacting pupils-first observable symptom in some cases)
- High frequency hearing loss or tonal deafness becomes evident
- Easy to become emotionally agitated or upset.

Cases Journal 2009, **2:**9355doi:10.1186/1757-1626-2-9355

- Patients demonstrate diabetes mellitus followed by optic atrophy in the first decade, diabetes insipidus and sensorineural deafness in the second decade, dilated renal outflow tracts early in the third decade, and multiple neurological abnormalities early in the fourth decade.

Indian Journal of Clinical Biochemistry, 2009 / 24 (4) 436-438

- We describe two cases of wolfram syndrome belonging to same family; 25 year old female and her only 15 year old brother. In female, **diabetes mellitus and optic atrophy were manifested in 1st decade, diabetes insipidus in 2nd decade and hypoacusis at the age of 25 years.** Her ophthalmic evaluation revealed bilateral optic atrophy, decreased vision and peripheral constriction of visual field. However she didn't have any renal dysfunction which is also considered to be one of the features of the syndrome.

To summarise

- Is it polyuria? (wt gain, measure U.O)
- Exclude UTI, DM. CBG, Urine sugar, ketones
- Check calcium, urea, creatinine, serum Hco_3 ABG
- Serum sodium, serum osmolality, Urine osmolality (evaluate to exclude DI)
- Water deprivation test

Take home message

- DI though rare is an important cause of polyuria in children
- DI can be an association with DM
- Suspect DI with DM when symptomatogy out of proportion to the severity of DM


THANK
YOU!