Clinical update no. 543

21 August 2019

Children are mostly little adults.

It's hard having to always explain that to a paediatrician.

Not always – children get metabolic problems

D&V FOR 4/7 AND MORE LETHARGIC THAN USUAL. LAST EPISODE OF DIARRHOEA LAST NIGHT, BUT HAS ONGOING VOMITING. BGL 2.6 AND KETONES 4.7 AT

pH	7.29L
pCO2	34L
HCO3	17.3L
Base Excess	-10.3L
Potassium	3.6
Sodium	131L
Chloride	104
Glucose	4.0
Lactate	1.0
Anion Gap	14

4 year old child presented to ED with a BGL of 2.3 and ketones of 4.9 on a back ground of vomiting and diarrhoea for 3 days, with poor oral intake. An older sibling was diagnosed with idiopathic ketotic hypoglycaemia.

Improved with 2 ml/kg IV 10% dextrose and rehydration with 0.9%saline/5% dextrose.

Always check the glucose; ketones as well, as a workup may be needed.

Hypoglycaemia

www.rch.org.au/clinicalguide/guideline_index/Hypoglycaemia/

Clinically hypoglycaemia is BSL low enough to cause symptoms and/or signs of impaired brain function, generally <2.6 mmol/L. Long term neurological complications is a risk.

Hyperinsulinism is the most common cause < 2 years, though is unlikely with ketonaemia.

Accelerated starvation (previously known as "ketotic hypoglycaemia") is the most common cause beyond infancy, presenting at 18 mth – 5yr and resolves before age 10yr. It is generally after fasting and precipitated by mild illness. There is low BSL with ketones. Diagnosis requires exclusion of other metabolic and endocrine causes which require testing depending on the clinical picture.

Management involves maintaining dietary intake and avoiding prolonged fasting. Ketonuria precedes hypoglycaemia and can help monitor and prompt increased feeding.

Age at onset guides investigation and cause; neonates are separate.

Beyond neonatal period to 2 years: congenital hyperinsulinism, inborn errors of metabolism, congenital hormone deficiencies

Child: accelerated starvation, hypopituitarism

Adolescent: insulinoma, adrenal insufficiency.

An initial screen for inborn errors includes BSL, ketones, lactate, and ammonia. If all normal then relax; if not get help. Expect a long list of tests, which need to be sent on ice.

www.rch.org.au/clinicalguide/guideline_index/Hypoglycaemia/

Discharge requirements

- A cause for hypoglycemia **must** be known prior to discharge
- A reasonable time between feeds/meals (at least 4 hours) must be safely tolerated

PT VOMITING SINCE YESTERDAY NIL DIARRHOEA

	ARTERIAL
pH	7.11L
pCO2	9L
HCO3	6.1L
Base Excess	-26.6L
Potassium	3.3L
Sodium	126L
Chloride	101
Glucose	21.0H
Lactate	1.1
Anion Gap	25H
Ketones	6.8

12yr girl presents with vomiting without diarrhoea– it is not "gastro". There had been polyuria for a few weeks leading up to becoming critically unwell with DKA presenting with non-specific symptoms.

A high AG metabolic acidosis, and likely added respiratory alkalosis with a pCO2 9 at the extreme low range (was an ABG, though

didn't need to be). Winters formula would calculate respiratory compensation for a HCO3 6 as pCO2 17 ±2.

The delta gap suggested an added non-gap gap metabolic acidosis. The extreme low range HCO3 of 6.1 is 18 below normal (24), and falling by more than the elevation in AG of about 13 from the upper range (8-12).

Checking BSL and ketones can be diagnostic – BSL can be high or low with +ve ketones.

Case: 4mth old boy presents with vomiting and reduced feeding. He had been started antibiotics from the GP for an *E coli* UTI. There had been some diarrhoea which had resolved.

Born at term, with no problems.

He was alert and active, RR 40 O2 sats 100% on air, HR 122, afebrile.

Chest clear, no increased work of breathing. HS dual, abdo soft/not distended.

WCC 26.7 Hb 122 CRP 43

Na 102

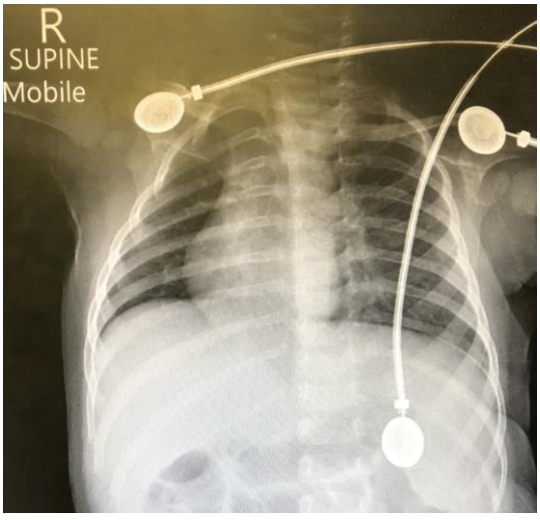
K 7.8

Cl 71 HCO3 11 urea 8.1 creatinine 23

VBG: pH 7.34 pCO2 28 HCO3 15 BE n/a lactate 3.4

AG 20 Glucose 5.2

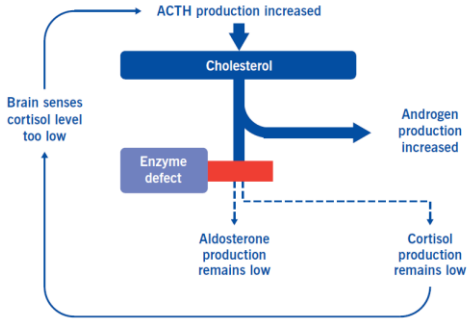
CXR: dextrocardia.



Treated for an Addisonian crisis, based on hypo-Na and hyper-K, and given broad spectrum antibiotics for likely sepsis.

Given hydrocortisone 4mg/kg IV. It was advised that the high K would likely correct itself with replacement and didn't need anything else acutely to reduce. Clearly nothing active to raise the critically low Na.

There is a range of underlying causes. Congenital Adrenal Hyperplasia is one.



An enzyme defect leads to reduced aldosterone and cortisol, with a feedback loop increasing ACTH which increases androgens. Females may have ambiguous genitalia. It typically presents in the first few weeks of life with shock and electrolyte derangement.

PediatricsinReview

Congenital Adrenal Hyperplasia: Diagnosis, Evaluation, and Management
Zoltan Antal and Ping Zhou
Pediatr. Rev. 2009;30;e49-e57

The common defect in what is a range of disorders comprising CAH is impaired cortisol secretion. About 75% with the more severe classic form also have salt wasting due to inadequate aldosterone production. The 2 main categories are classic simple virilising and classic salt-wasting forms.

Treatment
Acute Adrenal Crisis

Volume replacement with 0.9% saline; and dextrose to treat/prevent hypoglycaemia. Hydrocortisone replacement: stress dose is 1-2 mg/kg q 6hr; use double or more if hyponatraemia/hyperkalaemia/hypovolaemia. Dexamethasmone does not have mineralocorticoid effect, whereas hydrocortisone does. There is no IV mineralocorticoid (only fludrocortisone orally).