



The case of R

A slush puppy mystery

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Alder Hey Children's Hospital

History

- Male, born 5/11/05
- Normal delivery at term
- Mum used recreational drugs while pregnant
- R hospitalised 8 weeks, withdrawal
- Inguinal hernia repair as neonate
- Height and weight between 50th and 75th centile
- Head circ between 75th and 91st centile
- Lives with grandparents, grandad diabetic on metformin

General health

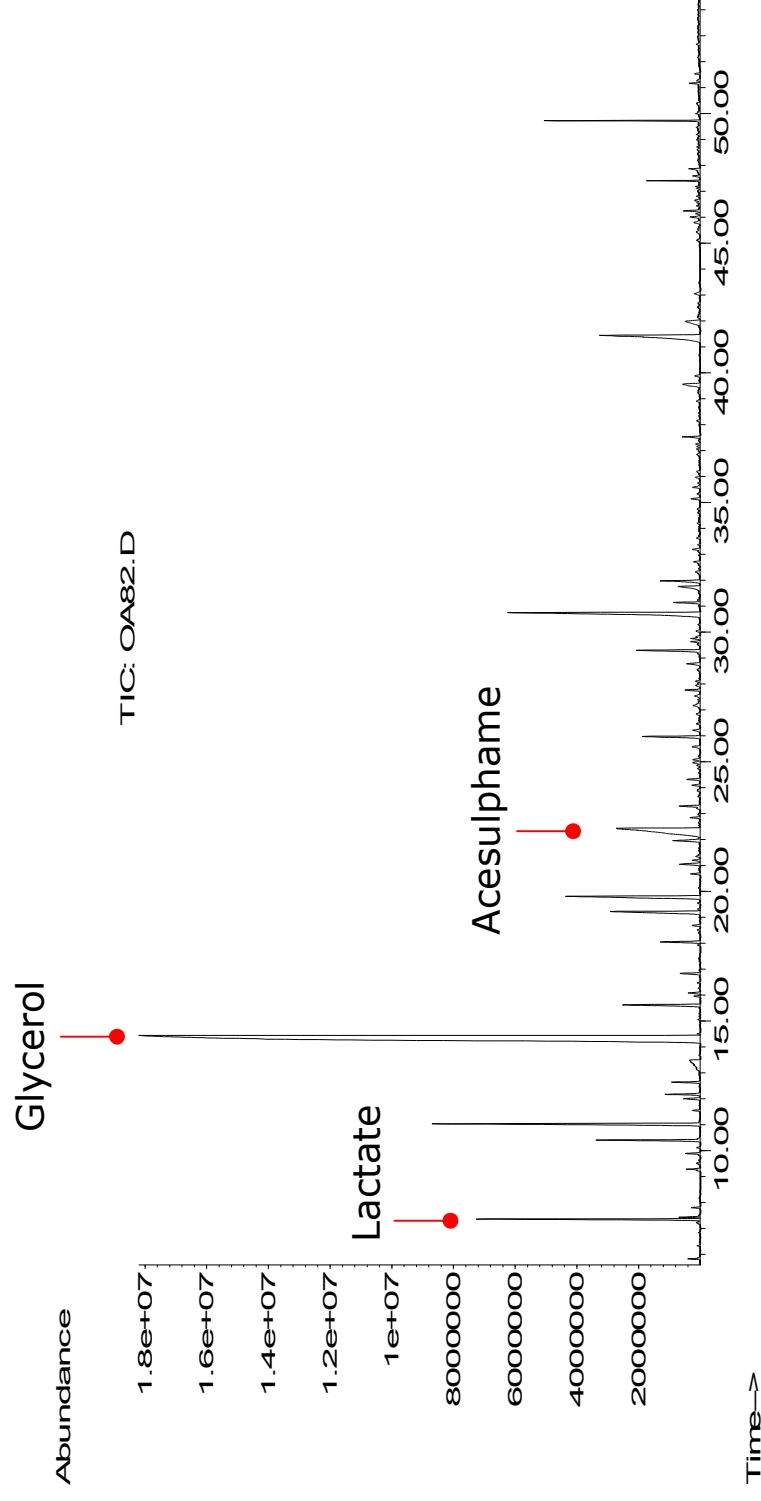
- Mild asthma managed with salbutamol
- Sleeps through – no symptoms suggesting hypoglycaemia before breakfast
- No previous problems with sugary drinks
- Mid Jan 2009 had diarrhoea and vomiting for 7 days but had recovered for 7 days prior to admission

Slush puppy incident

- 25/1/09 visited Wacky Warehouse
- About 3 hrs after lunch had slush puppy
- 15 – 20 mins later pale, headache, passed out
- No obvious seizure activity
- Glucose 2.3mmol/l but normalised after IV gluc
- Glasgow coma score of 11 for 2 hrs
- By 4 hours full recovery

R's organic acids – 25/01/09

- Moderately increased glycerol & lactate
- Acesulphame present



- OAs 3 days later – normal, trace glycerol

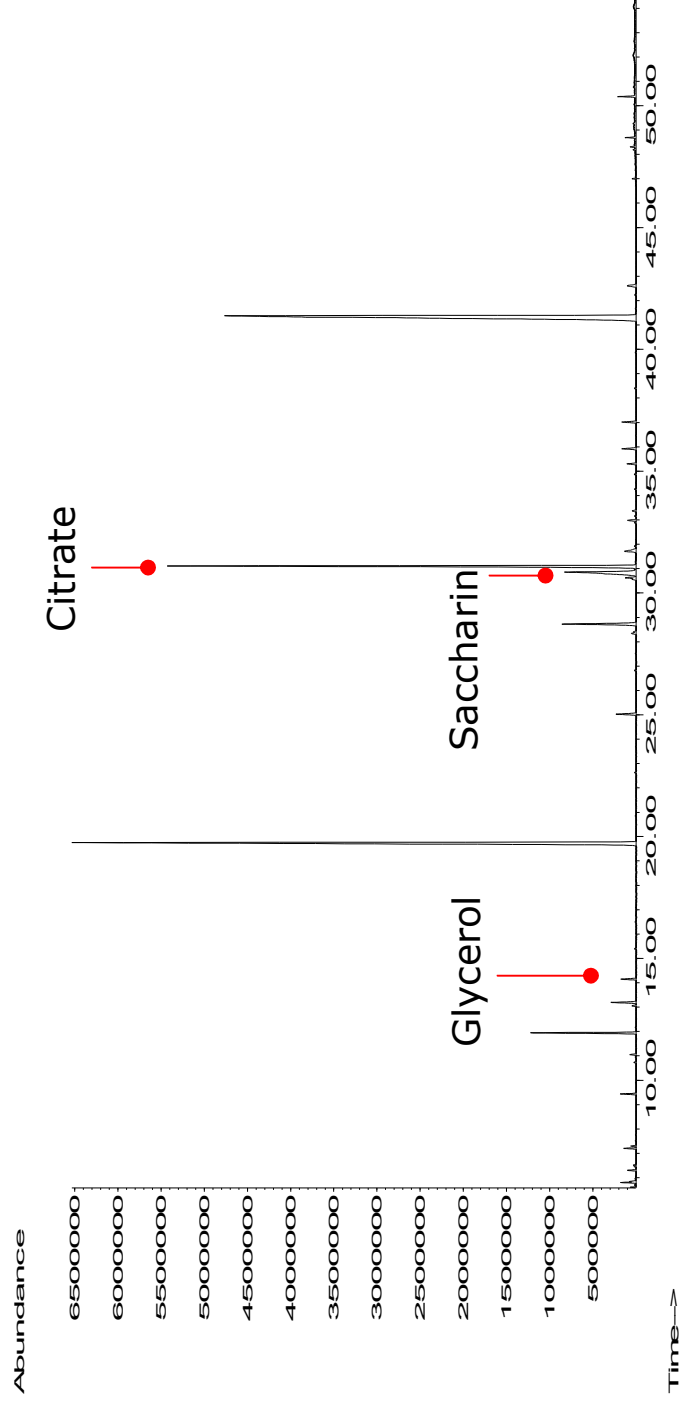
Lab results

	Admission	6hrs	29/1/09
K+	2.4 mmol/l	4.1 mmol/l (no K+ given)	
PO₄	1.34 mmol/l	2.76 mmol/l (no PO ₄ given)	
Trigs	25.6 mmol/l (not lipaemic)		2.4 mmol/l
Blood gases	Mild metabolic acidosis		

- Urine sugars — trace lactose, trace fructose
- Blood acylcarnitine and synacthen tests normal
- Urine tox normal
- Negative for fructose 1,6 bisphosphatase deficiency

Slush puppy organic acids

- Small amount glycerol
- Saccharin and citrate present
- No acesulphame



- Sugar TLC – heavy glucose band, fructose present

Possibilities

- F16BPD – intermittent glyceroluria, episodes of ↓gluc and ↑lact
- GK deficiency – but glyceroluria and glycerolaemia would be constant
- Hereditary fructose intolerance – but tolerated large fructose loads in past
- Metformin ingestion – awaiting result

Recent health

- Generally well
- For 12 days up to clinic visit diarrhoea, abdo pain, 4 or 5 watery stools per day
- Not off food
- No vomiting
- Slightly wheezy when seen
- Developing normally