

**G/THURS/GEM1 Table 1** Tests to exclude non-metabolic disorders

Test	% Patients	Test	% Patients
Virology	23.7	Toxicology screen	13.5
Bacterial culture	44.1	Carbon monoxide	0

**G/THURS/GEM1 Table 2** First-line metabolic investigations

Test	% Patients	Test	% Patients
Liver function tests	71.2	Urine organic acids	20.3
Glucose	33.9	Free fatty acids	6.8
Serum lactate	40.7	3-hydroxybutyrate	6.8
Acylcarnitines	23.7	CPTII mutation	6.8
Blood amino acids	20.3	McArdle's mutation	6.8
Urine amino acids	18.6	XP21 mutation	0

26.7 s). Wilson's disease, alpha-1-antitrypsin deficiency, haemochromatosis, viral and autoimmune hepatitis were all excluded. The ammonia was subsequently noted to be elevated (303  $\mu\text{mol/l}$ ) and testing for urea cycle defect confirmed a diagnosis of ornithine transcarbamylase deficiency and treatment was instituted, with resolution of the abnormal liver function tests. Subsequent mutational analysis confirmed a mutation of the ornithine transcarbamylase (OTC) gene (P225L).

The second case presented to hospital at 12 months of age, with a history of intermittent lethargy and vomiting for 1 month. She was noted to have elevated liver enzymes (ALT: 5143 IU, AST: 284 IU) and abnormal coagulation profile. Ammonia was slightly elevated (105  $\mu\text{mol/l}$ ). Viral and autoimmune hepatitis was excluded. Following a protein challenge, the ammonia rose to 224  $\mu\text{mol/l}$  and the urine organic acid profile detected increased orotic acid suggestive of OTC deficiency. This was confirmed by enzymatic assay of a liver biopsy. The abnormal liver function tests subsequently resolved with institution of dietary intervention.

X-linked OTC deficiency is the commonest urea cycle defect, with a predicted incidence of 1 in 14 000 births. Males may present with severe life threatening neonatal hyperammonaemia or with a milder late onset form. The presentation in females can be subtle. OTC deficiency should be considered in the differential diagnosis of children presenting with abnormal liver function tests, in particular with a subtle presentation in females.

Haycock G, Greenough A. Sudden infant death, bed-sharing and dummies: authors' reply. *Arch Dis Child* 2007;**92**:560. This was a comment on an earlier article in *ADC*: Fleming P, *et al*. New knowledge, new insights and new recommendations. *Arch Dis Child* 2006;**91**:799–801. It was *not* an authors' reply since the original article did not refer to any previous publications. We apologise for this error.

Owing to incorrect authorship listed in the online March issue the following letters are republished here.

### Intravenous rehydration of children with gastroenteritis: which solution is better?

Recent publications have suggested that hyponatraemia may develop in children with gastroenteritis treated with intravenous hypotonic saline.<sup>1–3</sup>

Even though we believe these papers have been well designed and developed, we cannot agree with their results for we are carrying out a similar study in our centre (81 cases up to now) that is leading to the opposite conclusion: our children with gastroenteritis did not develop hyponatraemia even though they were all treated with hypotonic intravenous solutions (0.3% saline with 5% glucose), while isotonic fluids were only used in "preshock" situations.

The incidence of hyponatraemia at the time of diagnosis is lower in our study (9%) than in those published previously (range 30–50%); this could be due to differences in climate or diet.

In the analysis, we separated children according to whether they were hyponatraemic, normonatraemic, or hypernatraemic at presentation. In the first group, hypotonic intravenous saline increased mean plasma sodium (from 132.4 (SD 2.07) to 135.3 (SD 2.21) mEq/l); it was decreased slightly in the second group, without leading to hyponatraemia (139.2 (SD 2.9) to 137.3 (SD 2.9) mEq/l), and also in the third group (150.4 (SD 4.12) to 140.6 (SD 3.6) mEq/l). No cases of hyponatraemia post-infusion were seen. Hoorn and colleagues,<sup>4</sup> in a sample of 1586 children, showed that the cases of hyponatraemia in their study were due to incorrect treatment, with higher volumes of fluid than needed.

In our study, 10 children (16.3%) presented with glucose levels lower than 70 mg/dl (40 mg/dl in one case). If these children were treated with isotonic fluids without adequate glucose, levels would never increase, with serious consequences.

These data are not definitive, but should be taken into consideration before selecting an appropriate solution in these patients. Further studies with different designs are required.

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*Arch Dis Child* 2006;**91**:716.

### References

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- 2 Duke T, Molyneux EM. Intravenous fluids for seriously ill children: time to consider. *Lancet* 2003;**362**:1320–3.
- 3 Neville KA, Verge CF, O'Meara MW, *et al*. High antidiuretic hormone levels and hyponatraemia in children with gastroenteritis. *Pediatrics* 2005;**116**:1401–7.
- 4 Hoorn EJ, Geary D, Robb M, *et al*. Acute hyponatremia related to intravenous fluid administration in hospitalized children: an observational study. *Pediatrics* 2004;**113**:1279–84.

### Intravenous rehydration of children with gastroenteritis: which solution is better? Authors' response

The letter by Sanchez-Bayle *et al*<sup>1</sup> states that the administration of hypotonic saline to children with gastroenteritis is not, in their view, associated with an increased risk of hyponatraemia. This is in sharp contrast with our findings,<sup>[2 3]</sup> and those of others, showing that the risk is real. Unfortunately, the data provided by Sanchez-Bayle are insufficient for analysis and we look forward to their findings being published in full. On the other hand, we also concluded from our studies that any isotonic solution used should contain added glucose. In two studies of children with gastroenteritis (n = 154), we have documented a 4% rate of hypoglycaemia (blood glucose concentration, 2.6 mmol/l) at presentation.<sup>[2 3]</sup> In both studies, the hypoglycaemia responded to the 2.5% dextrose content of the intravenous fluid prescribed at either a slow or rapid rehydration rate. Much of the recent literature on isotonic versus hypotonic saline solutions for children ignores the need for glucose, and we welcome this focus.

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- 2 Neville KA, Verge CF, O'Meara MW, *et al*. High antidiuretic hormone levels and hyponatremia in children with gastroenteritis. *Pediatrics* 2005;**116**:1401–7.
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## Intravenous rehydration of children with gastroenteritis: which solution is better? Authors' response

Kristen A Neville, Charles F Verge, Andrew R Rosenberg, et al.

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