

## References

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M D LOUBSER, M C HOH, and N SREERAM  
 Children's Department,  
 Newcastle General Hospital,  
 Westgate Road, Newcastle upon Tyne  
 NE4 6BE

## Late onset ornithine carbamoyl transferase deficiency in males

Sir,

We were interested that Drogari and Leonard did not mention sodium valproate as a possible precipitant of symptoms in late onset carbamoyl transferase deficiency in males.<sup>1</sup> We wish to report such a case; to our knowledge this has not been reported before.

A 10 year old boy, who had no previous medical or family history, was started on sodium valproate 200 mg twice a day for worsening photosensitive absence seizures. After three weeks a subtherapeutic concentration of the drug prompted an increase in dose to 200 mg three times a day. Five days later the boy vomited twice. Solids and sodium valproate were withheld. The next day he vomited three more times and became difficult to rouse in the general practitioner's surgery. He was immediately admitted to hospital but remained easily rousable overnight. Twelve hours after admission his conscious level deteriorated. Blood glucose concentration was 5.5 mmol/l and ammonia 145  $\mu$ mol/l, his prothrombin ratio was 1.5 and aspartate aminotransferase activity 47 IU/l. All other liver function tests and a computed tomogram were normal. Blood sodium valproate concentration was 0.16 mg/l (subtherapeutic).

Eighteen hours after admission he was transferred to another hospital for intensive care. Assisted ventilation was begun. Intracranial pressure was low initially. Thirty six hours after his first hospital admission the blood ammonia concentration was 245  $\mu$ mol/l and intracranial pressure 2.67-4.00 kPa. Chromatography of the urine showed a peak of orotic acid. This was initially a very small peak but after reversal of the effect of sodium valproate with sodium benzoate, became a very large peak typical of ornithine carbamoyl transferase deficiency. Despite aggressive management of his intracranial pressure and appropriate doses of dextrose, sodium benzoate, arginine, and growth hormone, intracranial pressure rose to 7.33 kPa and ammonia to 918  $\mu$ mol/l over the next 12 hours before coming under control. His cerebral perfusion pressure could not subsequently be maintained and he died on the sixth hospital day. Very low residual ornithine carbamoyl transferase activity of 1.6  $\mu$ mol/mg/hour (and normal carbamoyl phosphate synthetase activity) was confirmed in liver obtained immediately postmortem.

Hyperammonaemia and brain swelling in valproate toxicity complicating urea cycle disorders are more often rapidly progressive and fatal than in classical Reye's syndrome even if, as in the present case, early and aggressive treatment is given. Such deaths are one of several metabolic hazards of sodium valproate that must be weighed against the potential benefit of treatment.<sup>2</sup>

## References

- Drogari E, Leonard JV. Late onset ornithine carbamoyl transferase deficiency in males. *Arch Dis Child* 1988;**63**:1363-7.
- Brown JK. Valproate toxicity. *Dev Med Child Neurol* 1988;**30**:121-5.

C R KENNEDY and J J COGSWELL  
 Department of Child Health,  
 Southampton General Hospital,  
 Shirley, Southampton SO9 4XY

## Rubella immunisation for girls over 14 years

Sir,

I was interested to read Dr Hodes' article on rubella immunisation for girls over 14 years,<sup>1</sup> but believe that her findings may not justify her recommendation that rubella vaccination should be offered to girls until they leave school at 16 years of age.

Dr Hodes offered her programme to 56 of 83 girls whose rubella immunity status was unknown, but she does not tell us why the other 27 were not invited. Only 25 of the 56 accepted, and 21 of them (84%) were found to be seropositive. The other four were vaccinated. The increase in the proportion of girls vaccinated or immune from 86% to 90% was due largely to the finding of seropositive girls, and much less to vaccination.

If it is assumed that the same proportion (84%) of all 83 girls was immune, 566 girls (96.6%) were vaccinated or immune before Dr Hodes' programme was introduced, and 570 (97.2%) afterwards. While applauding every effort to increase rubella immunity in the community, I wonder whether this marginal increase justified the expense of the enterprise.

## Reference

- Hodes D. Rubella immunisation for girls over 14 years. *Arch Dis Child* 1988;**63**:983-5.

R W SMITHELLS  
 University Department of Paediatrics and Child Health,  
 Leeds General Infirmary,  
 Belmont Grove, Leeds LS2 9NS

Dr Hodes comments:

Professor Smithells rightly criticises the fact that only 56 of the 83 eligible girls were invited. This was due to many practical problems that were described in the second and third paragraphs of the discussion. This is a shortcoming of the study and in future one would have to try harder to invite all eligible girls.

Assuming that the same proportion (84%) of all 83 girls was immune, 70 girls would be seropositive and 13

seronegative. The total number of girls vaccinated or immune would have been (503+70) 573 (97.8%) before the programme and (573+4) 577 (98.5%) afterwards. Professor Smithell's figures of 566 and 570 respectively are probably due to an inadvertent miscalculation. In other words, there were 13 (2.2%) seronegative girls before and nine (1.5%) after the programme. The vaccination of four girls is worth the estimated cost of at most a few hundred pounds. There are still 20 cases a year of congenital rubella syndrome.<sup>1</sup> Until rubella disease is eliminated it is extremely important to achieve an uptake of 100% in women of child bearing age. If the study had been completely successful all 13 girls would have been vaccinated.

#### Reference

- <sup>1</sup> Joint Committee on Vaccination and Immunisation. *Immunisation against infectious disease*. London: HMSO, 1988.

## Reflex anal dilatation associated with severe chronic constipation in children

Sir,

Reflex anal dilatation is an important sign associated with

anal abuse in children.<sup>1</sup> The buttocks are parted gently and the examiner waits for up to 30 seconds. Where the sign is present, the anal sphincter dilates to up to 2–3 cm and a view of the interior of the rectum is observed. The anus then frequently closes and then reopens in a dynamic fashion.

Dr Clayden describes a 'visibly relaxed anus such as one sees when a large faecal mass has chronically distended the rectum leading to a sphincter which is dilated'—there is no suggestion of the dynamic findings of reflex anal dilatation described in his paper.<sup>2</sup>

Perhaps if doctors could agree a common vocabulary,<sup>1</sup> then these kind of irrelevant disagreements would not exist.

#### References

- <sup>1</sup> Butler-Sloss E. *Report of the inquiry into child abuse in Cleveland 1987*. London: HMSO, 1988.  
<sup>2</sup> Clayden GS. Reflex anal dilatation associated with severe chronic constipation in children. *Arch Dis Child* 1988;**63**:832–6.

C J HOBBS and J M WYNNE  
*St James's University Hospital,*  
*Beckett Street,*  
*Leeds LS9 7TF*