Admittedly a quarter of all children in Britain have no annual holiday (half in Manchester). One in 20 has no washing machine (15% in Manchester). And there are families with quite limited incomes who fund holidays and laundry equipment. But if children meet the medical criteria for severe disability should there be exclusions at such moderate levels of income? If so, applications should be discouraged.

References

I AMcKINLEY
Royal Manchester Children’s Hospital, Pendlebury, Manchester M27 1HA

Transient neonatal galactosaemia

Sir,

The case reported by Taylor et al reminds me of a child I saw in 1980 with a similar syndrome in association with cystic fibrosis.1

Case report

A first born girl was admitted to hospital at the age of 3 months because of failure to thrive (birth weight 3100 g, admission weight 3900 g: less than the third percentile). There were no specific clinical abnormalities. She was anaemic with a haemoglobin of 80 g/l and a reticulocyte count of 13%. She had abnormal liver function tests with a low plasma albumin concentration of 25 g/l and a raised serum glutamo-oxaloacetic transaminase activity of 169 IU/l. Tests for bleeding, jaundice, congenital infection, hepatitis B antigen, and α1 antitrypsin deficiency gave negative results. Her urine contained galactose and a plasma galactose concentration was 5.8 mmol/l (upper limit of normal 0.24 mmol/l). Subsequently red cell enzymes were studied and she was shown not to have deficiencies of galactose-1-phosphate uridyl transferase, galactokinase, or epimerase.

She was treated with a galactose free diet, and galactose concentrations were normal within three days. She thrived so that her weight achieved the 10th percentile at 6 months of age and her liver function returned to normal. At 9 months a milk diet was reintroduced and her urine remained galactose free. She was admitted to hospital at the age of 3 years because of weight loss and pneumonia. Sweat sodium concentrations were 84, 88, 117, and 115 mmol/l. She has responded well to treatment for her cystic fibrosis.

Comment

This case is very similar to that reported by Taylor et al. The response to a galactose free diet suggests that her liver abnormality was secondary to the raised blood galactose concentration. There were no clinical stigmata of cystic fibrosis before admission at the age of 3 years with chest symptoms and weight loss. Though cystic fibrosis may be associated with neonatal jaundice, this had not occurred.

Reference

C H CHEETHAM
Amersham General Hospital, Amersham, Bucks HP7 0JD

Sudden death in incomplete Kawasaki disease

Sir,

We read with interest the case report by McCowen and Henderson regarding sudden death in incomplete Kawasaki disease.1 Recently we managed a similar patient presenting with this syndrome who died suddenly despite a normal echocardiogram.

A 2 year old Asian girl was admitted with a six day history of fever and four days of rash. The rash was maculopapular, starting on the chest and spreading to the limbs. She had mild palmar erythema, cervical lymphadenopathy, stomatitis, and mild hepatomegaly. On admission the erythrocyte sedimentation rate was 125 mm in the first hour. The C reactive protein was 83 mg/l and the platelet count was 448×10^9/l rising to 762×10^9/l on the eighth day after admission. Cross sectional echocardiography confirmed normal ventricular dimensions and function. The proximal coronary arteries were well visualised and were of normal calibre. The echocardiogram was also normal. Bacterial cultures and viral serology were negative, serum immunoglobulins, complement and anti-streptolysin O titles were normal. Neutrophil cytoplasmic antibodies were not present. On the ninth day in hospital her temperature settled and she was clinically improved. There was desquamation of the truncal rash, but not of the fingers. Forty eight hours later the patient collapsed suddenly and could not be resuscitated.

At necropsy the right coronary artery was occluded by recent thrombus close to its origin. The wall of the left anterior descending branch was thickened and the lumen narrowed. Histology showed coronary arteritis without aneurysmal dilatation.

Children with Kawasaki disease who have a high risk of developing coronary complications are Asians, aged less than 5 years, with fever longer than 14 days, and high erythrocyte sedimentation rates and platelet counts.2,3 This case illustrates the limited predictive value of echocardiography in the acute stage with regard to cardiac complications. It also raises the question of when to institute antiplatelet treatment in children presenting with fever, a rash, and thrombocytosis.
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. We wish to report 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 μmol/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 μmol/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 μmol/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 μmol/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.

References


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, 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


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...