Correspondence

Table Details of infants with bulging fontanelles

<table>
<thead>
<tr>
<th>Case No</th>
<th>Sex</th>
<th>Age (months)</th>
<th>Symptoms</th>
<th>Cerebrospinal fluid pressure (cm H2O)*</th>
<th>Cerebral imaging</th>
<th>No of recurrences</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>F</td>
<td>5</td>
<td>Upper respiratory tract infection</td>
<td>Not done</td>
<td>Not done</td>
<td>—</td>
</tr>
<tr>
<td>2</td>
<td>M</td>
<td>4</td>
<td>Upper respiratory tract infection</td>
<td>30–32</td>
<td>Computed tomogram, normal</td>
<td>—</td>
</tr>
<tr>
<td>3</td>
<td>F</td>
<td>9</td>
<td>Upper respiratory tract infection</td>
<td>30</td>
<td>Ultrasound normal</td>
<td>1</td>
</tr>
<tr>
<td>4</td>
<td>F</td>
<td>4</td>
<td>Gastroenteritis (Echo 18 virus)</td>
<td>16</td>
<td>Ultrasound normal</td>
<td>2</td>
</tr>
<tr>
<td>5</td>
<td>M</td>
<td>5</td>
<td>Febrile, rash over shoulders</td>
<td>Not done</td>
<td>Not done</td>
<td>—</td>
</tr>
<tr>
<td>6</td>
<td>M</td>
<td>4</td>
<td>Gastroenteritis</td>
<td>Not done</td>
<td>Not done</td>
<td>—</td>
</tr>
<tr>
<td>7</td>
<td>F</td>
<td>2</td>
<td>Upper respiratory tract infection</td>
<td>Not done</td>
<td>Not done</td>
<td>—</td>
</tr>
</tbody>
</table>

*Normal range for infants 4–15 cm H2O.

The Family Fund

Sir,

Paediatricians have supplied reports to the Family Fund for many years in support of families of severely disabled children. About 105 000 families have benefited since 1973. Washing machines, dryers, and holiday relief are the most needed and the present budget is £8 million of government money.

Recently the Fund has been unable to help the parents of two of my patients (the only children of the family). They are severely mentally retarded, undiagnosed with marked involuntary movements, have very little independent mobility, and severely limited communication at the ages of 5 and 7 years. The Fund accepts that the children’s disabilities come within its medical criteria but is unable to offer the family help on the grounds that their economic circumstances do not justify it.

The Fund reports that only 0–9% of families have been refused help on the grounds that ‘they are too wealthy’ for the purposes of the Fund. It also accepts that parents who are denied on these grounds feel angry, and observes that all families tend to live up to the edge of their income: ‘Relatively well off families have understandably high expectation in material and personal terms. The stress they experience can be no less acute than they experienced in less favourable circumstances.’ The Fund stresses that it does not operate a means test but justifies each grant individually. It comments on its sensitivity to the exceptional expense of having more than one handicapped child, or unusual expenses because of particular disabilities.

The Family Fund grew out of the appreciation of the cost of caring for disabled children. If it is accepted that low income families should have greatest priority.

If my patients’ families were ‘wealthy’ in the ordinary sense of the term, this letter would not have been written. ‘Too wealthy’ is a threshold under regular review but seems to be around a gross income of £12 500 per annum.

References


W Barry, W Lenney, and G Hatcher
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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

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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 glutam-oxaloacetate 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

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 electrocardiogram was also normal. Bacterial cultures and viral serology were negative, serum immunoglobulins, complement and anti-streptolysin O titres 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.