Systemic vasculitis complicating infantile autoimmune enteropathy

EDITOR.—We read with interest the case reported by Jenkins et al in which they highlight the association of autoimmune enteropathy with sideroblastic anaemia.1 We too do not know of any reported cases of these two associations and believe that they are most likely to be separate clinical entities. The sideroblastic anaemias are a heterogeneous group of disorders that result from different pathophysiological mechanisms impairing haem synthesis. In young children the inherited forms of sideroblastic anaemia of which X linked transmission is the commonest, account for the vast majority of cases when lead toxicity had been excluded. Defining the type of sideroblastic process from blood indices and bone marrow morphology can be difficult as the vast majority of cases have variable hypochromia with macrocytosis and less frequently a dimorphic blood picture. In cases where the blood is macrocytic and the anaemia is refractory to pyridoxine then Pearson’s syndrome should be considered in the differential diagnosis.2 This is a multisystem disorder characterised by refractory sideroblastic anaemia, with or without vacuolisation of bone marrow precursors, and varying insufficiency in exocrine pancreatic (malabsorption), hepatic (fibrosis, steatosis), renal (proximal tubulopathy), and gastrointestinal (watery diarrhoea, partial villous atrophy) function.3 4 Like other mitochondrial cytopathies, Pearson’s syndrome is characterised by mitochondrial dysfunction occurring secondary to deletions/arrangements of mitochondrial DNA (mtDNA). It is usually fatal within the first three years of life, the majority succumbing to end organ failure. Clearly the case reported has not inherited her sideroblastic anaemia in an X linked manner as she is female. It would be interesting to know whether lead concentrations were established, what the red cell indices were at presentation and if postmortem tissue was stored as mtDNA analysis could be carried out. We suggest that deletions/arrangements of mtDNA should be sought in all young children presenting with anaemia secondary to a sideroblastic process.

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Dr Jenkins and coauthors comments:
We thank the authors for their letter regarding our case report highlighting the association of sideroblastic anaemia and autoimmune enteropathy. Like them we too had considered a wide variety of differential diagnoses and can confirm that lead deficiency had been excluded (normal lead concentrations were found on several occasions) as had Pearson’s syndrome. As we stated in our report, there was normal pancreatic function on formal testing and blood had already been sent for mtDNA analysis to two different centres (one in the UK and one in the USA). Neither centre found any abnormality in mtDNA (in particular no deletions) and further specimens were sent to a laboratory in New York which carried out extensive sequence analysis of the child’s genes and found no abnormalities (in particular no abnormality of erythroid ALA synthetase). We can therefore reassure the correspondents that deletions or rearrangements of mtDNA were excluded in our child and agree with them that this should be undertaken in any case of sideroblastic anaemia.

Potentially dangerous sleeping environments and accidental asphyxia in infancy and early childhood

EDITOR.—Byard and colleagues are right to draw attention to the sleeping environment as a cause of accidental death in infancy.1 I have analysed the Department of Trade and Industry’s Home Accident Deaths Database (HADD) for England and Wales for the most recent available year, 1992. There were 10 comparable cases for this single year. The children were aged between 6 months and 2 years and were all found dead in sleep settings (table). These deaths took place despite the existence of safety standards (BS 1753, Cots and BS EN 747 1993, Bunk Beds) or safety regulations (Bunk Bed Entrapment Hazards 1987) designed to minimise such incidents.

Circumstances of deaths in 10 cases of childhood asphyxia (HADD 1992)

<table>
<thead>
<tr>
<th>Age (months)</th>
<th>Sex</th>
<th>Circumstances</th>
</tr>
</thead>
<tbody>
<tr>
<td>11</td>
<td>F</td>
<td>Cord in cot tangled round neck</td>
</tr>
<tr>
<td>10</td>
<td>M</td>
<td>Cord of blind wrapped round neck in cot</td>
</tr>
<tr>
<td>17</td>
<td>M</td>
<td>Hooked shirt on cot</td>
</tr>
<tr>
<td>9</td>
<td>M</td>
<td>Cardigan caught in cot side</td>
</tr>
<tr>
<td>9</td>
<td>M</td>
<td>Found hanging over side of bed</td>
</tr>
<tr>
<td>12</td>
<td>M</td>
<td>Cord with mittens at each end caught round neck and part of head</td>
</tr>
<tr>
<td>6</td>
<td>M</td>
<td>Head and chest in 5 inch gap between mattress and cot</td>
</tr>
<tr>
<td>11</td>
<td>M</td>
<td>Slipped between bed and wall</td>
</tr>
<tr>
<td>14</td>
<td>M</td>
<td>Neck trapped in cot</td>
</tr>
<tr>
<td>24</td>
<td>M</td>
<td>Slipped from top bunk, wedged chin between bunk bed and other furniture</td>
</tr>
</tbody>
</table>

I would endorse the view of Byard et al that parents should receive advice about the appropriate sleep environment for their infants, including avoiding the use of clothing that could be snapped. However, individualised advice delivered on a one to one basis, perhaps by a health visitor or midwife, is more likely to be effective than the display of pamphlets.

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