Intestinal obstruction caused by malrotation of the gut in atrial isomerism

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SUMMARY Five children with atrial isomerism developed intestinal obstruction caused by malrotation of the gut. Other than asplenia, the extracardiac anomalies in these syndromes are rarely regarded as important as the outcome after intestinal surgery is poor. As cardiac treatment improves, early investigation and intervention for intestinal symptoms becomes more important.

Children with atrial isomerism usually have other severe cardiac abnormalities that dominate their clinical presentation. Associated thoracic and visceral heterotaxia were originally recognised at necropsy and have been described as 'the cardio-splenic syndrome'. Apart from asplenia, however, the visceral abnormalities have been considered predominantly of pathological interest. We report five patients with atrial isomerism and severe cardiac disease in whom the clinical course was complicated by intestinal obstruction caused by malrotation of the gut.

Patients

Between 1980 and 1988 five patients with atrial isomerism developed intestinal obstruction as a result of malrotation of the gut; details of the individual cases are given in the table. All five started vomiting within six weeks of birth; the vomit was bile stained in four, the fifth having pyloric atresia as well as malrotation, and all required operation to relieve the obstruction. Two children, both without spleens, died of septic complications within three weeks of operation. One had a further episode of intestinal obstruction caused by adhesions at the age of 14 months and died six months later from his cardiac disease. Another died of bronchiolitis and cardiac failure at 11 weeks of age. One child remains well at the age of 8 years on long-term treatment with antibiotics after one abdominal and three cardiac operations.

Discussion

Right atrial isomerism with asplenia is reported to

Table Five cases of atrial isomerism with intestinal malrotation

<table>
<thead>
<tr>
<th>Case No</th>
<th>Sex</th>
<th>Gestation (weeks)</th>
<th>Birth weight (g)</th>
<th>Cardiac anomalies</th>
<th>Visceral anomalies</th>
<th>Operations</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Male</td>
<td>42</td>
<td>3770</td>
<td>Left atrial isomerism, double outlet right ventricle, bilateral superior vena cava, complete atrioventricular septal defect, patent ductus arteriosus</td>
<td>Symmetrical liver, gall bladder in midline, spleen on right and splenunculus, duodenojejunal flexure to left of midline, Ladd's transduodenal band, colon on right</td>
<td>Pulmonary banding, ligation of ductus, Ladd's procedure, duodenal fixation</td>
<td>Died at 20 months, cardiac failure</td>
</tr>
<tr>
<td>2</td>
<td>Female</td>
<td>34</td>
<td>1800</td>
<td>Double inlet left ventricle with double outlet, right atrial isomerism, pulmonary atresia</td>
<td>Spleen absent, stomach on left, pyloric atresia, duodenum and duodenojejunal flexure to right of midline, caecum in midabdomen, complete malrotation</td>
<td>Gastroduodenostomy, gastrostomy</td>
<td>Died at 14 days, septicaemia</td>
</tr>
<tr>
<td>3</td>
<td>Male</td>
<td>38</td>
<td>1740</td>
<td>Right atrial isomerism, complete atrioventricular septal defect with common atrioventricular valve, patent ductus arteriosus, pulmonary atresia</td>
<td>Symmetrical liver, gall bladder in midline, absent spleen, portal vein anterior to first part of duodenum, malrotation with common mesentery, meconium ileus, microcolon, perforated terminal ileum</td>
<td>Ladd's procedure ileal resection, split ileostomy</td>
<td>Died at 26 days, necrotising enterocolitis</td>
</tr>
<tr>
<td>4</td>
<td>Male</td>
<td>38</td>
<td>3080</td>
<td>Dextrocardia, left atrial isomerism, complete atrioventricular septal defect</td>
<td>Situs inversus, symmetrical liver, spleen present, malrotation with Ladd's transduodenal band</td>
<td>Ladd's procedure, pulmonary banding</td>
<td>Died at 11 weeks, cardiac failure, bronchiolitis</td>
</tr>
<tr>
<td>5</td>
<td>Male</td>
<td>41</td>
<td>4220</td>
<td>Right atrial isomerism, double outlet right ventricle, complete atrioventricular septal defect, pulmonary atresia</td>
<td>Spleen absent, symmetrical liver, gall bladder on right, portal vein and common bile duct anterior to first part of duodenum, Ladd's transduodenal band, malrotation with common mesentery</td>
<td>Right Blalock shunt, Ladd's procedure, left Blalock shunt, Waterston shunt</td>
<td>Well at 8 years</td>
</tr>
</tbody>
</table>
Comforters and night waking

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SUMMARY Among 320 low birthweight infants seen at nine months post-term those using a soft object, thumb, or fingers as comforter were significantly less likely to wake at night (9/96, 9%) than those with no comforter or using a dummy (66/224, 29%). Dummy users were as likely to wake (27/93, 29%) as those without a comforter (39/131, 30%).

One of the commonest reasons for parents to seek professional advice is that their child wakes them at night. Sleep disturbance can cause serious family problems and many strategies have been advocated to alleviate the problems. One of us (CJM) observed that children who were able to soothe themselves with a thumb or cloth were least likely to wake their parents at night.

In a survey of 3 year old children, Graham and Boniface found that children who used an 'attachment object', settled to sleep more easily than those without one (p=0.07), but they found no

References


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Arch Dis Child 1989 64: 1623-1624
doi: 10.1136/adc.64.11.1623

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