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'.¹ 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

Case No	Sex	Gestation (weeks)	Birth weight (g)	Cardiac anomalies	Visceral anomalies	Operations	Outcome
1	Male	42	3770	Left atrial isomerism, double outlet right ventricle, bilateral superior vena cava, complete atrioventricular septal defect, patent ductus arteriosus	Symmetrical liver, gall bladder in midline, spleen on right and splenunculus, duodenojejunal flexure to left of midline, Ladd's transduodenal band, colon on right	Pulmonary banding, ligation of ductus, Ladd's procedure, duodenal fixation	Died at 20 months, cardiac failure
2	Female	34	1880	Double inlet left ventricle with double outlet, right atrial isomerism, pulmonary atresia	Spleen absent, stomach on left, pyloric atresia, duodenum and duodenojejunal flexure to right of midline, caecum in midabdomen, complete malrotation	Gastroduodenostomy, gastrostomy	Died at 14 days, septicaemia
3	Male	38	1740	Right atrial isomerism, complete atrioventricular septal defect with common atrioventricular valve, patent ductus arteriosus, pulmonary atresia	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	Ladd's procedure ileal resection, split ileostomy	Died at 26 days, necrotising enterocolitis
4	Male	38	3080	Dextrocardia, left atrial isomerism, complete atrioventricular septal defect	Situs inversus, symmetrical liver, spleen present, malrotation with Ladd's transduodenal band	Ladd's procedure, pulmonary banding	Died at 11 weeks, cardiac failure, bronchiolitis
5	Male	41	4220	Right atrial isomerism, double outlet right ventricle, complete atrioventricular septal defect, pulmonary atresia	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	Right Blalock shunt, Ladd's procedure, left Blalock shunt, Waterston shunt	Well at 8 years

represent 1% of congenital heart disease.¹ Deanfield *et al* found a 10% incidence of thoracic heterotaxy in children admitted to a cardiac unit,² and thoracic heterotaxy is thought to be more commonly associated with atrial isomerism than visceral heterotaxia. In studies of large numbers of cases severe cardiac disease still dominates the picture of atrial isomerism and, although recognised, extracardiac abnormalities are rarely reported as causing clinical problems.³ Rose *et al*, in a study of 60 patients with atrial isomerism, noted only two patients with appreciable gastrointestinal abnormalities other than asplenia.⁴ Moller *et al*, however, found that malrotation was always present in children with congenital heart disease who also had asplenia or polysplenia, though it occurred in less than 1% of cases in which the spleen was normal.⁵ Freedom concentrated on extracardiac problems and, in 23 cases, found 25% with important gastrointestinal and genitourinary anomalies.⁶ Three patients in his series presented with intestinal obstruction: two each had an annular pancreas and one had congenital fibrous bands. No child in his series with malrotation presented with obstruction.

Our collection of five children all had visceral heterotaxia and developed clinical intestinal obstruction. All had malrotation and four required division of Ladd's transduodenal band. Two had other appreciable gastrointestinal abnormalities that probably contributed to their deaths, but—as might be expected—intestinal operations seem to carry much greater risks in this group of children than in those with normal hearts. The poor survival in this group confirms other reports,⁴ but one child remains well at the age of 8 years on long term treatment

with antibiotics. Antipneumococcal vaccine is no use in congenital asplenia until the age of 2 years, but lifelong prophylaxis with half doses of phenoxymethylpenicillin is recommended. These children present early with serious cardiac problems, although in our unit antenatal diagnosis is becoming increasingly common. As treatment of these cardiac problems becomes more successful, the extracardiac associations become more important. Thus the occurrence of gastrointestinal symptoms (particularly bile stained vomiting) in infants with atrial isomerism warrants early investigation and consideration of operation.

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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.¹ 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 Boniface² found that children who used an 'attachment object',³ settled to sleep more easily than those without one ($p=0.07$), but they found no