

Visceral heterotaxy and malrotation in a neonate

A term infant with antenatal diagnosis of visceral heterotaxy presented on the third day of life with bilious vomiting and rectal bleeding. Abdominal radiograph (*Figure 1*) and upper gastrointestinal contrast study (*Figure 2*) demonstrated intestinal malrotation with suspicion of midgut volvulus. Findings were confirmed at laparotomy where a right-sided stomach was seen, with the duodeno-jejunal flexure lying to the left and volvulus of the midgut. The liver was on the left side of the abdomen and the spleen in the midline with the hilum facing left. Two additional splenunculi (polysplenia) were found. A Ladd's procedure with inversion appendicectomy was performed.

'Situs ambiguus' or heterotaxy refers to a disordered arrangement of the truncal organs in which the atrial situs and visceral situs correspond, associated with symmetrical morphology of normally asymmetrical structures ('isomerism'). Heterotaxy syndrome is generally classified as either

right isomerism (associated with asplenia syndrome – also termed Ivemark's syndrome) or left isomerism (associated with polysplenia) (Applegate et al, 1999).

Intestinal malrotation is a common association of heterotaxy syndrome (Ditchfield and Hutson, 1998). The majority of malrotations are asymptomatic but there is a risk of midgut volvulus leading to significant morbidity and mortality (Ruben et al, 1983). Most authors agree that infants with heterotaxy should have an upper gastrointestinal contrast study before discharge from the postnatal ward and if malrotation is identified should undergo correction to

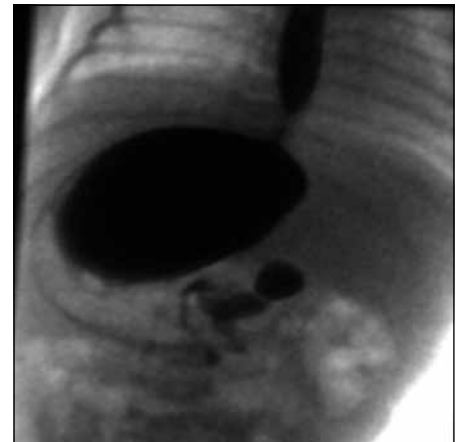
Figure 1. Abdominal X-ray demonstrates the heart is normally positioned. A large gas-filled visceral structure is seen in the right upper quadrant.



reduce the risk of life-threatening complications in later life (Yu et al, 2009). [BJHM](#)

Applegate KE, Goske MJ, Pierce G, Murphy D (1999) Situs Revisited: Imaging of the Heterotaxy Syndrome. *RadioGraphics* **19**: 837–52
 Ditchfield MR, Hutson JM (1998) Intestinal rotational abnormalities in polysplenia and asplenia syndromes. *Pediatr Radiol* **28**: 303–6
 Ruben GD, Templeton JM, Jr, Ziegler MM (1983) Situs inversus: the complex inducing neonatal intestinal obstruction. *J Pediatr Surg* **18**: 751–6
 Yu DC, Thiagarajan RR, Laussen PC, Laussen JP, Jaksic T, Weldon CB (2009) Outcomes after the Ladd procedure in patients with heterotaxy syndrome, congenital heart disease, and intestinal

Figure 2. Upper gastrointestinal contrast study showing pooling of contrast in a right-sided stomach with the duodenal-jejunal flexure lying to the left of the midline consistent with malrotation of the intestine. Only a small trickle of contrast was demonstrated in non-dilated bowel loops suggesting a bowel obstruction.



metrical polyarthritis, eye inflammation and

Mr A MacDonald is Senior House Officer, **Mr NS Johal** is Specialist Registrar, **Mr M Haddad** is Consultant Paediatric Surgeon and **Mr MS Choudhry** is Consultant Paediatric Surgeon in the Department of Paediatric Surgery, Chelsea and Westminster Hospital, London SW10 9NH

Correspondence to: Mr A MacDonald
(alexander.macdonald@mac.com)