

Abdominal right isomerism with normal intracardiac anatomy in the fetus

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Objectives

In the postnatal sequential segmental diagnosis of cardiac abnormalities, it has historically been recognised that atrial situs usually follows that of the abdominal situs. Recognition of abdominal right isomerism (RI) or left isomerism (LI) is often associated with complex congenital heart disease (CHD). However, whilst 'less complex or minor CHD' and 'normal hearts' have been reported with LI, the same has not been previously reported in the setting of abdominal RI. We had identified a number of such cases in our fetal practice and therefore sought to clarify their features.

Methods

Patients were identified from the electronic databases in our tertiary fetal cardiac service comprising three fetal cardiology units. Prenatal echo reports and postnatal reports were reviewed.

Results

Six cases of abdominal RI without complex CHD were identified. Gestational age at diagnosis ranged from 18+0 to 23+1 weeks gestation. Four cases were livebirths; one pregnancy was terminated and one lost to follow up. The stomach was right sided with levocardia and apex to the left in 5 cases and the stomach left sided with dextrocardia and apex to the right in 1 case.

The position of the aorta and inferior vena cava (IVC) is summarised in table 1:

Case	Position of stomach	Position of aorta	Position of IVC	Position of heart	Position of Apex
1	R	Midline	R	L	L
2	R	Midline/R	R	L	L
3	R	Midline/R	R	L	L
4	R	L	L	L	L
5	L	R	R	R	R
6	R	L	L	L	L

Table 1: Ultrasound findings of six fetuses with abdominal features of RI

Key: R=right, L=left; IVC = inferior vena cava

All cases had biventricular atrio-ventricular connections. There was right hand topology in the cases with laevocardia and apex to the left and left hand topology in the case with dextrocardia and apex to the right. Ventriculo-arterial connections were concordant in all cases. In four cases, no cardiac abnormalities were seen. In two cases, abnormalities of systemic venous return were identified: One fetus had a left superior vena cava (SVC) and IVC draining to a dilated coronary sinus; a second fetus had anomalous drainage of the IVC to the left sided atrium, a left SVC draining to the coronary sinus and abnormal drainage of hepatic veins. The presence of a coronary sinus in these 2 cases indicated discordance between abdominal and thoracic situs. One case developed bowel obstruction postnatally secondary to malrotation.

Conclusions

Abdominal RI can occur with normal intracardiac anatomy. This may be explained in some cases by discordance between abdominal and thoracic situs. Detailed sequential segmental analysis of the fetus is important to identify such cases as these fetuses are at risk of the postnatal complications of isomerism such as gut malrotation, splenic abnormalities and primary ciliary dyskinesia.