Dextrocardia and situs inversus

VIVIENNE MILLER MB BS, FRACGP, DRACOG, DCH, MACPM, MWAME

Articles in this section are inspired by, but not based on, real cases to illustrate the importance of knowledge about ECGs in relation to clinical situations in general practice. Management is not discussed in detail.

Zac is an 18-year-old man who was diagnosed at birth with dextrocardia and situs inversus. He had a small ventriculoseptal defect that is now almost completely closed over, but this still causes him a great degree of anxiety about his heart generally. Zac has come to see you, his GP, today with symptoms typical of gastro-oesophageal reflux but is requesting 'a test of his heart in case it was a heart attack'. Although a recent previous echocardiogram found in his medical records shows he has now essentially a normal heart apart from it being located in the right side of the chest, you perform an ECG to reassure him. The ECG is shown in the Figure.

Q1. What are the features of this ECG that confirm dextrocardia?

The features shown on this ECG that confirm dextrocardia are:

- right axis deviation
- positive QRS complexes (with upright P and T waves) in lead aVR
- lead I – inversion of all complexes (known as 'global negativity'); the P wave and T waves are inverted and the QRS is negative
- absent R-wave progression in the chest leads (dominant S waves throughout).

These features can be reversed by placing the precordial leads in a mirror-image position on the right side of the chest and reversing the left and right arm leads.

It should be noted that dextrocardia can be mimicked by incorrect lead placement. Reversal of the left and right arm electrodes may produce a similar ECG appearance in the limb leads (but a normal appearance of the precordial leads).

Q2. What are the types of dextrocardia?

There are several types of dextrocardia, including those listed below.

- Mirror-image dextrocardia (situs inversus) – the most common type of dextrocardia, presenting in one in 8000 to 25,000 live births.[1] The right to left orientation of the heart sections are reversed but the anterior–posterior relation is normal. By definition, there is always a degree of abdominal situs

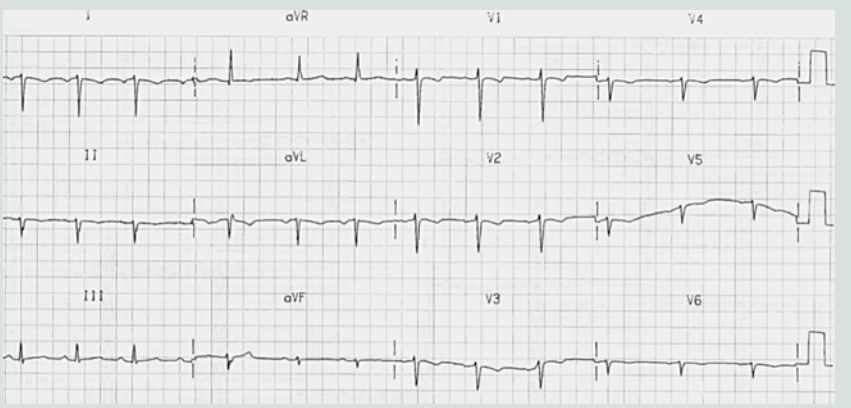

Figure. ECG showing dextrocardia.
Image courtesy of Dr Ed Burns and Life in the Fast Lane. http://lifeinthefastlane.com

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Dr Miller is a GP in Sydney, a medical journalist and author, and the Medical Editor of *Cardiology Today*.

SERIES EDITOR: Dr Richard Hillock MB ChB, FRACP, FCSANZ is a Cardiologist and an Electrophysiologist at SA Heart Centres, South Australia.

inversus (i.e. reversal of the normal relationship between the atria and the abdominal organs); however, other cardiac abnormalities are rare.
- Isolated mirror-image dextrocardia without situs inversus – extremely rare and patients are at risk of other cardiac malformations as well as abdominal obstruction.
- Dextroposition – in which dextrocardia occurs because of a physical shift of the heart into the right chest due to mechanical factors such as eventration of the left diaphragm, extensive scarring or malignancy of the right lung, or pneumothorax.
- Dextroversion – in which the heart is located in the right side of the chest with the ventricles rotated to the right but the atria in a normal position, and so the direction of spread of atrial depolarisation is the same as in a normal individual. As a result, the P wave in lead I is upright, differentiating it from mirror-image dextrocardia. Dextroversion is associated with typically serious cardiac abnormalities.

Q3. What are the associated cardiac defects commonly found with dextroversion?

Cardiac defects commonly found with dextroversion are:
- functional single ventricle
- ventriculoarterial discordance – for example, transposition of the great vessels
- atrioventricular discordance – for example, right ventricle is on the left (attached to the left atrium) and vice versa
- significant ventriculoseptal or atrioseptal defect – when these co-exist it is called an endocardial cushion defect
- double outlet right ventricle – for example, the supply to the pulmonary artery and the aorta is via a significant ventriculoseptal defect
- pulmonary stenosis or valvular atresia
- supraventricular tachycardia.

Q4. What is heterotaxy?

Heterotaxy is the malformation of the chest and abdominal organs found in conjunction with dextrocardia and situs inversus, and is also termed situs ambiguus. The most common abnormalities are in the gallbladder, the lungs, the structure and posture of the intestines (leading to malrotation and obstruction) and the spleen (both asplenia and small, ineffective polysplenia, both requiring immunisation to prevent pneumococcal disease).

Q5. Cyanotic congenital heart disease may occur with cardiac malformations associated with dextrocardia. What are the noncardiac clinical signs of cyanotic congenital heart disease?

Tachycardia, tachypnoea, cyanosis (central and peripheral), fatigue and dyspnoea on minimal exertion, peripheral oedema from congestive cardiac failure, hypotension, poor peripheral pulses and clubbing of the digits are the noncardiac clinical signs of cyanotic congenital heart disease in adults. Babies show these signs in addition to having failure to thrive, aspiration and poor feeding, irritability and developmental delay.

Q6. What is Kartagener's syndrome?

Kartagener's syndrome is a clustering of situs inversus, chronic sinusitis, immotile sperm and bronchiectasis. Approximately half of patients with immotile cilia (primary ciliary dyskinesia) have dextrocardia and situs inversus, and are classified as having Kartagener's syndrome; it is thought that the abnormal ciliary beating impedes organ rotation during embryonic development. The main clinical features derive from poor mucociliary clearance and are a thick, chronic mucoid nasal discharge that frequently develops bacterial infection (noted from childhood), nasal polyps, otitis media, anosmia, chronic bronchitis, recurrent pneumonia and, in males, bronchiectasis and infertility, and in females, reduced fertility. Congenital cardiac abnormalities may also occur.

Outcome: Zac is reassured by your explaining the ECG results to him and showing him that the changes can be reversed by placing the precordial leads in a mirror-image position on the right side of the chest and reversing the left and right arm leads. You also tell him that his recent echocardiogram was essentially normal and that his very small ventriculoseptal defect was almost completely closed. He was given 30 mL of liquid antacid with immediate effect, and was advised to avoid precipitants of reflux oesophagitis. CT

Key points

- **Mirror-image dextrocardia is the most common form of dextrocardia and occurs with situs inversus.**
- **Dextroposition is where dextrocardia occurs because of a physical shift of the heart to the right due to mechanical factors.**
- **Dextroversion is the rarest form of dextrocardia, and occurs along with typically serious cardiac abnormalities.**
- **Isolated mirror-image dextrocardia without situs inversus is extremely rare.**
- **Heterotaxy (situs ambiguus) is the malformation of the chest and abdominal organs found in conjunction with dextrocardia and situs inversus.**
- **Noncardiac clinical signs of cyanotic congenital heart disease in adults include tachycardia, tachypnoea, cyanosis, fatigue and dyspnoea on minimal exertion, peripheral oedema from congestive cardiac failure, hypotension, poor peripheral pulses and clubbing of the digits.**
- **Noncardiac clinical signs of cyanotic congenital heart disease in babies also include failure to thrive, aspiration and poor feeding, irritability and developmental delay.**
- **Kartagener's syndrome is a clustering of situs inversus, chronic sinusitis, immotile sperm and bronchiectasis.**

Reference

1. Casey B. Two rights make a wrong: human left–right malformations. Hum Mol Genet 1998; 7: 1565-1571.

COMPETING INTERESTS: None.