When abnormal is normal …

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Case history

A 36-year-old woman was seen at the Cardiac Congenital Clinic for her annual visit. She was born with a d-transposition of the great arteries (d-TGA) and underwent a Rashkind procedure shortly after birth, followed by a Senning procedure (atrial switch) in 1978. She recently went through two uncomplicated pregnancies and is asymptomatic except for occasional palpitations documented as ventricular extrasystoles and bigeminy. She does not take medication.

Her clinical status is unremarkable and cardiac auscultation reveals typical single B2.

The 12-lead ECG (figs 1 and 2) shows bradycardia at 40–45 bpm with sinus rhythm alternating with junctional rhythm. There is right axis deviation with QRS axis at 170°. The PR interval is normal in sinus rhythm, and QRS is narrow. Precordial leads demonstrate right ventricular (RV) hypertrophy and there is no Q wave with a small R wave and deep S wave in lead V6.

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Questions

What is normal/abnormal on the 12-lead ECG when taking into account the congenital heart disease?
Does junctional rhythm warrant further investigations/interventions?

Commentary

In complete transposition of the great arteries, or d-TGA, there is atrioventricular discordance, which means that the aorta arises from the morphological right ventricle and the pulmonary artery arises from the morphological left ventricle[1]. Nowadays, the arterial switch is the procedure of choice, as it restores the normal anatomic arrangement, but the majority of adult patients seen in adult congenital heart disease clinics underwent an atrial switch procedure. First described by Senning in 1958, it involves creation of an atrial baffle to direct the venous return to the contralateral atrioventricular valve and ventricle. Deoxygenated blood is directed to the mitral valve and left ventricle, and oxygenated blood goes to the right ventricle through the tricuspid valve. This means that the systemic ventricle is the morphological right ventricle and explains why we can see RV hypertrophy and right axis deviation. The left ventricle (the subpulmonary ventricle) is diminutive, which is reflected by the absence of Q waves, small R waves and deep S waves over the left precordial leads [2].

As a result of extensive atrial surgery, asymptomatic loss of sinus rhythm and supraventricular arrhythmias are extremely frequent after atrial switch for d-TGA [3]. In the absence of clinical repercussions and echocardiographic abnormalities, we recommend annual follow-up in a tertiary care centre with clinical examination, ECG, echocardiography, Holter monitoring and an exercise stress test to rule out chronotropic incompetence and/or symptomatic sinus node dysfunction.

References