IMAGE CHALLENGE

Heart block in a young man

CLINICAL INTRODUCTION

A young adult presented with a short history of subacute dyspnoea and dizziness. He had a sore throat and a rash 4 days before the onset of his current complaints, but this illness receded after a course of penicillin. Arthritis was absent, and there was no history of recent foreign travel or zoonosis. Bradycardia (46 beats per minute) was the main physical finding. His serum electrolytes, troponin, inflammatory markers, thyroid function, throat swab and chest X-ray were all normal or negative. His ECG, echocardiogram and cardiac CT scan are shown in figure 1A–C. (see also online supplementary video 1).

QUESTION

What is the most probable cause of this man's bradycardia?

A. Congenital complete heart block.
B. Acute rheumatic fever.
C. Sarcoidosis.
D. Congenitally corrected transposition of the great arteries (ccTGA).
E. Ebstein's anomaly.

For answer see page 1866

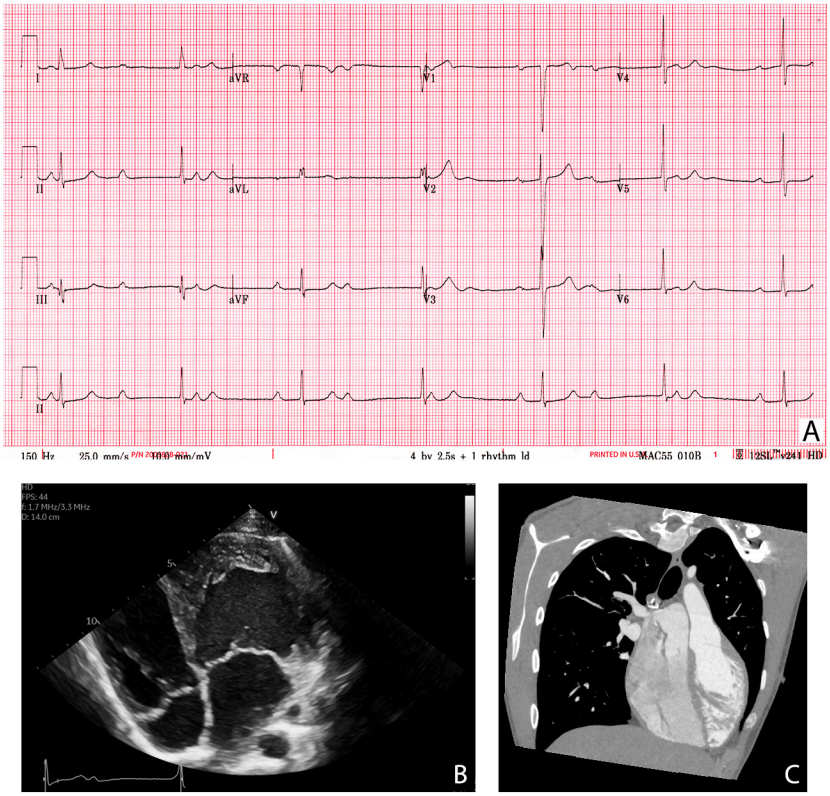

Figure 1 (A) ECG of bradycardia. (B) Four-chamber apical echocardiogram. (C) Reformatted arterial-phase cardiac CT section. aVR, augmented vector right; aVL, augmented vector left; aVF, augmented vector foot.

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ANSWER

D. Congenitally corrected transposition of the great arteries (ccTGA).

Figure 1A demonstrates complete heart block, for which A–E are all plausible causes. What is unique here is the concurrent absence of 'septal' q wave in leads V5/V6, and the large Q and S waves in V1 and V2, respectively. In the context of heart block, these oddities raise the possibility of ccTGA, a rare congenital malformation in which the cardiac ventricles 'switch places'. In situs solitus, the morphological left ventricle (LV) takes up the subpulmonic position, beneath the sternum, while the morphological right ventricle (RV) lies leftwards and propels the systemic circulation. The substernal location of the LV and the reversal of normal septal depolarisation produce the characteristic ECG profile. The atrioventricular node (AV) node and His bundle are malformed and located ectopically in ccTGA; they are prone to conduction block, which may occur years after birth.[1] In figure 1B, the morphological RV—the systemic ventricle—may be identified by the more apical location of its atrioventricular valve and by its coarse apical trabeculations; it is dilated. Figure 1C demonstrates ventriculoarterial discordance (RV—containing contrast and moderator band—connected to the aorta, LV to the pulmonary artery). While congenital complete heart block can theoretically present in adulthood, it does not explain the clinical images, and likewise for acute rheumatic fever and sarcoidosis (the absence of carditis and arthritis also making acute rheumatic fever remote). In Ebstein's anomaly, the ECG typically displays large P waves and right bundle branch block[2]; the tricuspid valve is displaced apically, but there is ventriculoarterial concordance. The ECG may hold the first clues of ccTGA.

Vickram Singh [1], **Heather J Edwards**,[1] **Fong T Leong** [2]

[1]Cardiology, University Hospital of Wales Healthcare NHS Trust, Cardiff, UK
[2]Heart Rhythm Service, University Hospital of Wales Healthcare NHS Trust, Cardiff, UK

Correspondence to Dr Vickram Singh, Cardiology, University Hospital of Wales Healthcare NHS Trust, Cardiff CF14 4XW, UK; vickram.singh@wales.nhs.uk

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ORCID iDs
Vickram Singh http://orcid.org/0000-0001-5944-0519
Fong T Leong http://orcid.org/0000-0002-8161-6928

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