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Clinical Image

Sudden Cardiac Death in Williams Syndrome

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Clinical Image

A 22-year-old man with a history of Williams syndrome and surgical repair of supra and subvalvaraortic stenosis, presented to the emergency department with a 3-month history of progressive dyspnoea. Chest x-ray showed signs of previous median sternotomy and cardiomegaly (Panel A). Transthoracic echocardiography showed relapse of the subaortic stenosis with significant aortic regurgitation (Panel B), severe mitral regurgitation (Panel C) and moderately impaired left ventricular systolic function with an ejection fraction of 40%. The patient was discharged on beta-blockers and vasodilators and was scheduled for an elective cardiac operation (mitral valve repair, aortic valve replacement and repair of the subaortic stenosis). However, while being at home, the patient experienced sudden cardiac death. Retrospectively, after reviewing his 12-lead surface ECG, we realised that he had prolonged QTc interval (QTc in lead V4=500ms, Panel D). Abnormalities of cardiac repolarization are frequent in patients with William's syndrome and may be associated with the elevated risk of sudden cardiac death in these patients.

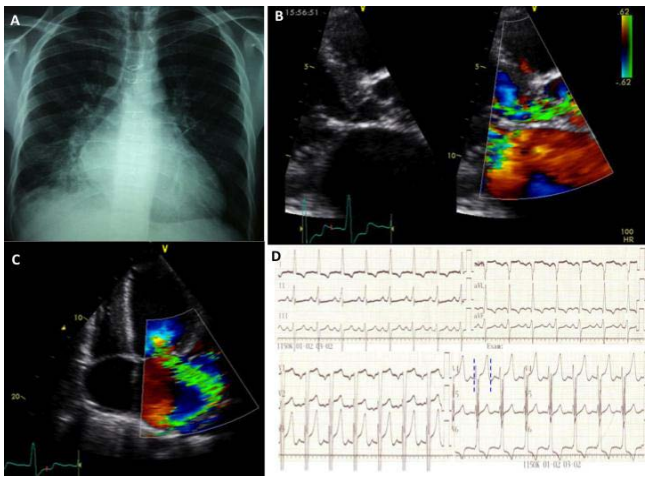

Figure:

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