

IMAGES IN CARDIOLOGY

Demonstration of supravalvar aortic stenosis by different cardiac imaging modalities in Williams syndrome

A 31 year old man was admitted for evaluation of dyspnoea and heart murmur. He had had exertional dyspnoea for several years. He had a typical posture with short stature and elfin face typical of Williams syndrome, characterised by protruded forehead, underdeveloped mandible and bridge of nose, and epicanthal folds. On auscultation, a mid systolic ejection murmur with 4/6 grade was best heard on the first right interspace and transmitted into the jugular notch and along the carotid vessels. ECGs revealed left ventricular hypertrophy, and chest radiography showed mild cardiomegaly and engorged pulmonary vasculature. A transoesophageal echocardiogram showed tubular narrowing and thickening of the aortic wall above the sinus (arrowhead in panel A; LA, left atrium; LV, left ventricle). Continuous wave Doppler across the supravalvar narrowing

revealed a maximal pressure gradient of 126 mm Hg. T1 weighted image on cardiac magnetic resonance imaging showed typical hour glass shaped narrowing of the aorta just above the sinus (arrowhead in panel B). This resulted in the typical appearance of a tubular deformity (arrowhead in panel C) on aortic root angiography. Left ventricular catheter pullback to the aorta revealed a peak-to-peak gradient between the left ventricle and ascending aorta of over 70 mm Hg. The patient was transferred for surgery.

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Exercise induced ST elevation and T wave alternans associated with critical disease of left anterior descending coronary artery

A 52 year old woman with hypercholesterolaemia and hypertension had a history of typical chest pain on exertion. An exercise treadmill test was performed, and her baseline ECG was unremarkable. She developed chest pain typical of angina at one minute of standard Bruce protocol associated with ST segment elevation in leads II, III, aVF, V1–V6, and T wave alternans (below left). The chest pain and ST segment elevation resolved with sublingual glyceryl trinitrate spray. The patient subsequently underwent coronary arteriography which showed a 99% diameter stenotic narrowing of the mid left anterior descending coronary artery (below right). Balloon

angioplasty with stenting was successfully performed without residual stenosis. The patient had no symptoms after the procedure. A repeat exercise test was performed, the results of which were negative without symptoms or ST changes.

T wave alternans is a rare phenomenon. Patients who develop T wave alternans have an increased chance of malignant ventricular arrhythmia and sudden cardiac death.

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