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IMAGE

Tetralogy of Fallot, Bicuspid Valves and a Single Origin Coronary: Uncorrected and Asymptomatic

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A 79-year-old asymptomatic man was referred to our clinic for routine immigration screening. His past medical history was significant only for remote treated pulmonary tuberculosis and heavy tobacco use. Examination revealed oxygen saturations of 90%, blood pressure of 150/90 mmHg, and a prominent right ventricular heave. There was no digital clubbing. Chest X-ray demonstrated an enlarged cardiothoracic ratio with a prominent right heart border and left hilar enlargement (Figure 1A). His electrocardiograph (ECG) showed atrial fibrillation, right axis deviation and anterior T-wave inversion consistent with right ventricular strain (Figure 1B).

Transthoracic echocardiography (TTE) demonstrated marked right ventricular hypertrophy (Figure 1C and D). There was right ventricular outflow tract (RVOT) narrowing, severe calcific pulmonic stenosis (peak-gradient 91 mmHg, Figure 1E), a large ventricular septal defect (VSD) with bidirectional shunting, and an overriding aorta (Figure 1C and Supplementary Video S1 in the online version at DOI: 10.1016/j.hlc.2018.07.016). Pulmonary hypertension was only mild with a corrected pulmonary artery systolic pressure (PASP) of 49 mmHg. These findings were consistent with uncorrected Tetralogy of Fallot (TOF). A minimally calcified bicuspid aortic valve (Figure 1F) without stenosis and associated aortopathy was also noted (aortic root measuring 49 mm, Figure 1G).

Claustrophobia precluded cardiac magnetic resonance imaging, so cardiac computed tomography (CT) was performed to assess for aorto-pulmonary shunting and other lesions. CT confirmed the findings of TOF (Figure 1H, I and J) with no aorto-pulmonary collaterals visualised. Cardiac CT was able to identify additional lesions with cine sequences showing bicuspid pulmonic valve morphology (Figure 1K, Supplementary Video S2 in the online version at DOI: 10.1016/j.hlc.2018.07.016). There was also a single origin coronary artery arising from the anterior sinus (Figure 1K), giving rise to a left anterior descending and right coronary artery. The left circumflex originated from the right. No significant coronary stenosis was demonstrated.

Tetralogy of Fallot represents the most common form of cyanotic congenital heart disease but uncorrected cases diagnosed in adulthood are rare. One-year survival without surgical correction is estimated to be 66%, with 40% surviving to 3 years and only 3% to 40 years [1]. Bicuspid pulmonic valves, though rare, can be associated with TOF, with up to 13% having valve abnormalities [2]. Coronary anomalies are relatively common (36%), with a single origin coronary system occurring in up to 6% [2]. Aortic dilatation has previously been reported as a late occurrence following Tetralogy repair, with a proposed mechanism of sustained volume overload [3]. Our patient had a non-obstructive bicuspid aortic valve, and this was also likely contributory given the known association.

Survival into adulthood suggests a balanced circulation dependent on the degree of pulmonic obstruction and collateral formation [4]. Factors contributing to prolonged survival include initially mild pulmonary stenosis with slow progression and adaptations that reduce right-to-left shunting such

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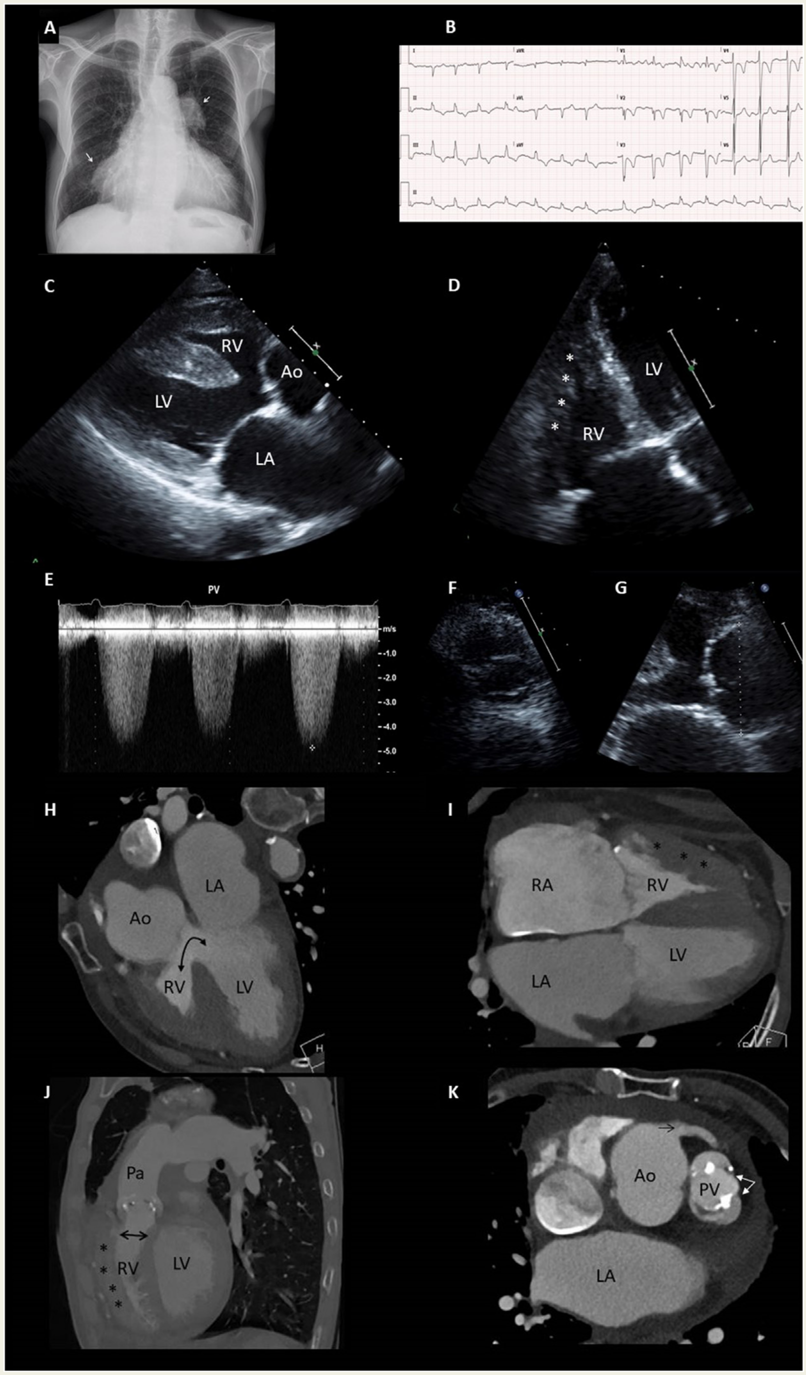

Figure 1 Panels.
(***A***): Chest X-ray shows an enlarged cardiothoracic ratio with a prominent right heart border (*arrow)* and left hilar enlargement (*arrow*). (***B***) Electrocardiograph (ECG**)** demonstrating atrial fibrillation, right axis deviation and anterior T wave inversions consistent with right ventricular strain.
(***C* – *G***): Transthoracic echocardiography of case. (***C***) Parasternal long axis view showing moderate left ventricular wall thickening, marked right ventricular hypertrophy with a clear ventricular septal defect. (***D***) Modified right ventricular view showing marked right ventricular hypertrophy (*). (***E***) Spectral Doppler signal through the pulmonic valve showing severe pulmonic stenosis with a peak gradient of 91 mmHg. (***F***) Short axis view at aortic valve level showing a minimally calcified bicuspid valve and ***(G)*** parasternal view of dilated aortic root measuring up to 49 mm.
(***H* - *K***): Cardiac computed tomography (CT) of case. (***H***) Sagittal oblique view demonstrates the aortic root overriding the right ventricular and left ventricular outflow tracts and ventricular septal defect (*double-headed arrow).* (***I***) Horizontal long axis view demonstrates marked right ventricular hypertrophy (*). (***J***) Sagittal view showing infundibular narrowing/obstruction

as the formation of aorto-pulmonary collaterals, persistent ductus arteriosus or systemic hypertension. Whilst severe RVOT obstruction can protect against pulmonary vascular disease, it also increases right-to-left shunting through the VSD. Hence, the circulatory adaptations that occur in uncorrected TOF certainly represent a 'fine line' of physiological balance. We speculate slowly progressive pulmonic obstruction and systemic hypertension contributed to our patient's extreme longevity.

Cardiac CT helped to identify further cardiac anomalies in this patient, highlighting the need for advanced cardiac imaging in all cases of congenital heart disease due to the multiplicity of lesions. Particularly in cases where intervention is being planned, identifying all anomalies is paramount for effective long-term outcomes. Indeed, given our patient's asymptomatic longevity, there is no indication to pursue any intervention at this stage. This case demonstrates a constellation of uncorrected congenital anomalies in an elderly asymptomatic patient, which represents a rarity in the literature and is a valuable lesson in adaptive circulatory balance.

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(*double-headed arrow)* and pulmonic valve calcification. (***K***) Axial view showing a single origin coronary artery arising from the anterior sinus (*single-headed arrow*) and calcified bicuspid pulmonic valve (*double arrow)*.
Abbreviations: LV, left ventricle; RV, right ventricle; LA, left atrium; Ao, aorta; Pa, pulmonary artery; PV, pulmonic valve.