Uncommon cause of adult onset cyanosis: single left ventricle

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DESCRIPTION

A native American man aged 41 years presented with progressive shortness of breath and discolouration of his lips. Physical examination was significant for central cyanosis and clubbing. Given his cardiac history, echocardiogram and cardiac MRI were ordered to look at the cardiac anatomy and physiology. MRI revealed that the patient has laevorotation of the heart with the cardiac apex pointing posterior to the left midaxillary line with double-inlet morphologic left ventricle (DILV), which was enlarged and hypertrophied. There was a hypoplastic right ventricle which lies superior and anterior to the morphologic left ventricle with large ventricular septal defect. The aorta arises from the hypoplastic right ventricle and courses anterior and to the left of the pulmonary arteries (figures 1–3). There is no pulmonary stenosis or subpulmonary stenosis noted on transthoracic echocardiography, which makes this case even more unique given that is been shown to have a survival benefit in patients with DILV. MRI shows that he was born with DILV and D-transposition of the great vessels, which was never surgically corrected. This patient is truly remarkable, in that he has survived into his fourth decade of life without surgical intervention.

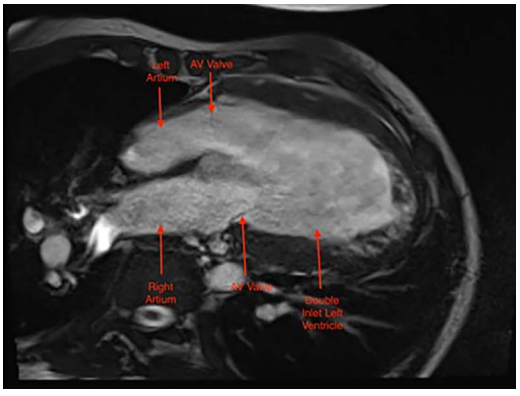

Figure 1 MRI of the heart double-inlet left ventricle (LV) with atrioventricular valves are inserted at the same level and draining into a common LV.

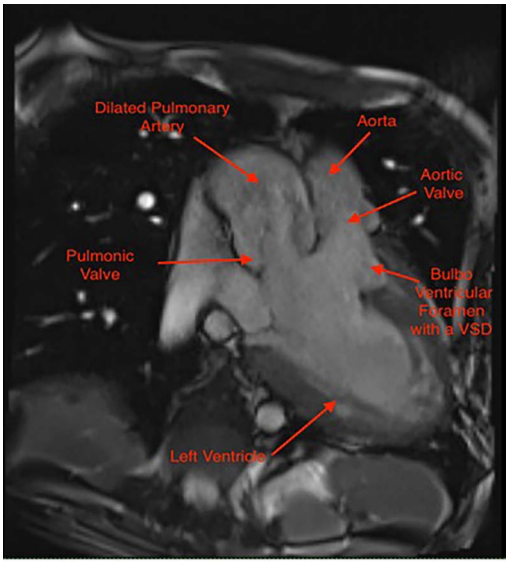

Figure 2 MRI showing the bulbo-ventricular foramen with ventricular septal defect providing systemic flow to the aorta and the pulmonary circulation with dilated pulmonary artery. There is complete mixing of systemic and venous circulation.

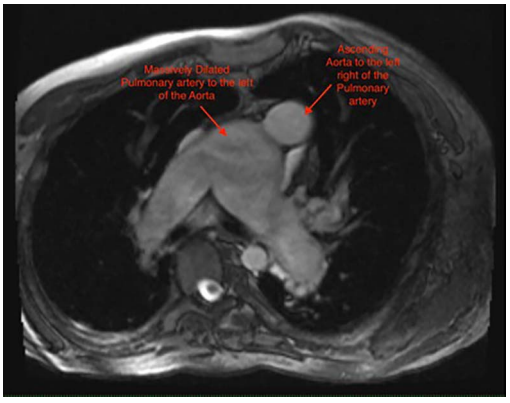

Figure 3 MRI showing the relationship of the great vessels consistent with D-transposition of great arteries. Notice massive dilation of the pulmonary artery consistent with pulmonary hypertension.

Learning points

- Adult onset cyanosis in this patient was due to Eisenmenger's syndrome.
- Cardiac transplantation is the only viable option for such patients.
- These patients should only be managed at specialised centres. In a retrospective review at Mayo Clinic, there was an annual death rate of 5%.[1]

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Competing interests None declared.

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1 Poterucha JT, Anavekar NS, Egbe AC, *et al*. Survival and outcomes of patients with unoperated single ventricle. *Heart* 2016;102:216–22.

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