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 (DIL V), 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 DIL V MRI shows that he was born with DIL V 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.

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%.

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REFERENCE