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Complex transposition of the great arteries with pulmonary arterial hypertension and giant pulmonary artery aneurysm

Kai'En Leong*, Subodh Joshi, and Leeanne Grigg

Department of Cardiology, The Royal Melbourne Hospital, 300 Grattan Street, Parkville 3050, Melbourne, VIC, Australia

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A 41 year-old male patient with transposition of the great arteries (TGA) and a large muscular ventricular septal defect (VSD), status-post Mustard repair, has attended our congenital cardiac service since immigration from Europe a decade earlier. His cardiac diagnoses were made aged 1 month with high pulmonary vascular resistance (PVR) noted then (6.8 WU).

Palliative atrial septostomy was performed soon after birth. Definitive Mustard repair with Dacron patch VSD closure were undertaken at age 9 months. Pulmonary arterial hypertension (PAH) and pulmonary arterial (PA) dilatation were recognized in his early adult years, and surveillance continued.

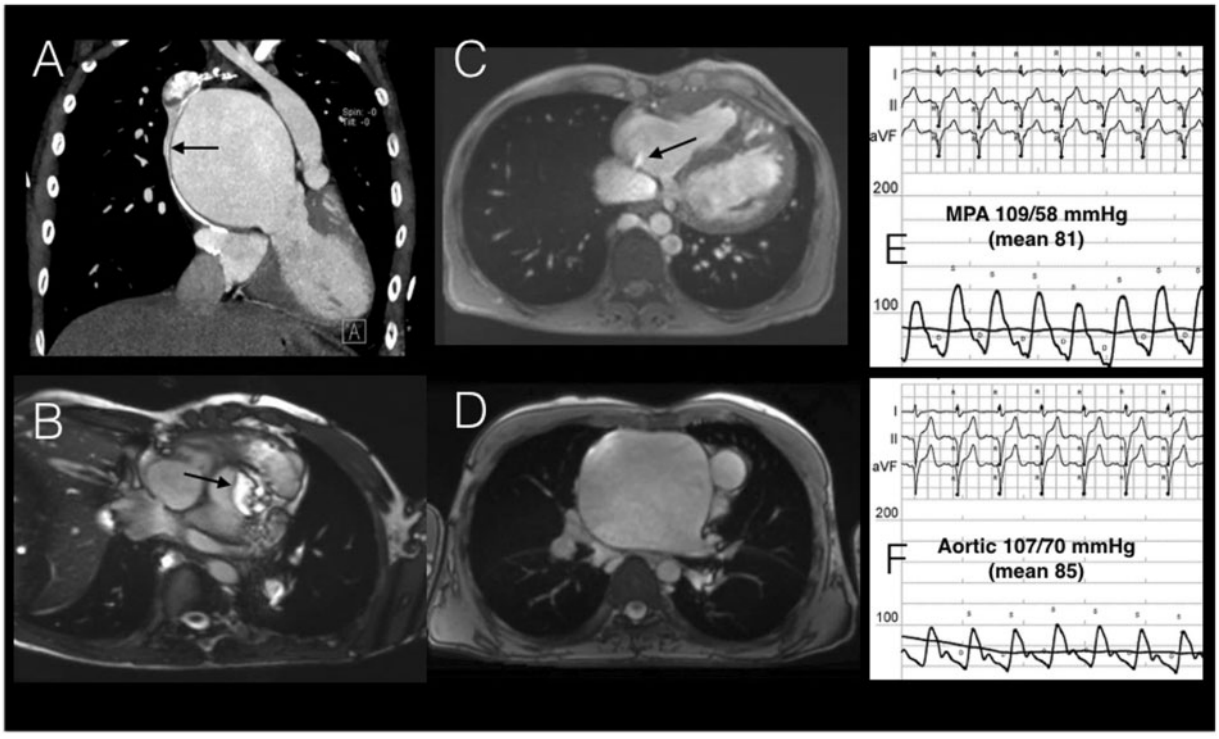

Figure 1 Complex TGA with giant PA aneurysm; severe PAH with Eisenmenger's physiology.

* Corresponding author. Tel: +61 9342 700, Fax: +61 9347 2808, Email: kai'en.leong@mh.org.au. This article was peer reviewed by George Giannakoulas and Cemil Izgi when submitted to "*Europace*". Accepted to EHJ Case Reports.

In the last 24 months, progressive hypoxemia and erythrocytosis developed with severe PAH noted on catheterization (mean PA pressure 81 mmHg, PVR 23 WU) (*E* and *F*, Supplementary material online, *Video S1*). Eisenmenger's physiology was demonstrated with physiologic right (pulmonic LV) to left (systemic RV) shunting via an inferior baffle leak (*C*, Supplementary material online, *Video S2*). The patient's pulmonary valve was suspicious for bicuspid morphology, albeit unrestricted (B). Macitentan therapy improved his walk distance with unchanged oxygen saturations. The patient's PA aneurysm now measures 10 cm in diameter, increasing by 2 cm every 7 years on serial MRI (*D*, Supplementary material online, *Video S3*). Consequently, SVC baffle distortion has occurred (*A*, Supplementary material online, *Video S4*), with decompression down a dilated azygos system.

His severe PAH presented prohibitive risk for surgical PA replacement. A percutaneous strategy was infeasible secondary to lack of appropriately sized and configured stents/grafts. The patient has been referred for heart–lung transplantation.

TGA patients typically avoid development of PAH with early repair. However, some develop PAH regardless; the aetiology of which is unclear. Bicuspid pulmonary valve (BPV) with TGA is an uncommon but recognized association, with incidence estimated at 4% in a large series.[1] PA aneurysm is a further rare association with BPV.[2] The triad of TGA associated PAH, BPV, and pulmonary arteriopathy presents a therapeutic challenge with limited operative options for progressive PA dilatation.

Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

Consent: The author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: none declared.

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