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Bicuspid pulmonary valve - an unusual cause of dilated pulmonary artery

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Topic(s):
Imaging: Valve Disease

Citation:

Introduction
Dilated pulmonary artery is a common finding with a variety of possible aetiologies. We present a case of unexplained dilated pulmonary artery that was associated with bicuspid pulmonary valve identified by CMR.

Case Presentation
A 67-year-old female underwent mitral valve repair for severe mitral regurgitation. She had presented with breathlessness, and had severe mitral regurgitation due to a myxomatous valve with bileaflet prolapse, confirmed with TOE. Echo showed: normal LV dimensions, LVEF 66%. RV size and systolic function normal. Coronary angiography was normal and pulmonary hypertension was not present on right heart catheterization.

At postoperative follow up she was well and asymptomatic, with echo showing good LV systolic function, intact mitral valve repair, and normal systolic pulmonary artery pressures (36mmHg). New T wave inversion was identified across the precordial leads so a CMR was performed.

CMR showed normal LV volumes and function with no wall motion abnormality or LVH. There was no fibrosis or infarction in the late phase after Gadolinium contrast. The RV was dilated (73mm diameter at base) with normal systolic function. The main pulmonary artery was dilated at 42mm. Flow mapping showed no evidence of shunt (Qp:Qs = 1:1) and the atrial septum was intact. Pulmonary valve function was normal but the valve was bicuspid in morphology.

Discussion
Bicuspid pulmonary valve is a rare finding. In our case this was identified by CMR and was associated with a dilated pulmonary artery.

Bicuspid pulmonary valve is most commonly identified in the context of pulmonary valve stenosis. Isolated pulmonary valve stenosis is a rare finding; it is most often associated with other congenital defects like Fallot. Pulmonary stenosis has been associated with pulmonary artery aneurysm, attributed to hemodynamic changes caused by the stenotic valve. However as with bicuspid aortic valve it is recognized that the "post stenotic dilation" is part of the pathology of bicuspid valve and does not need the changed haemodynamics of a stenotic valve.

During embryogenesis neural crest cells are necessary for the normal development of the semilunar valves, the septation of the outflow tract into the aorta and pulmonary artery, and the remodeling of the aortic arch. Any defect in this migration can cause abnormalities or aneurysm of the aorta or pulmonary artery together with semilunar valve defects. A study of 3,861 donor hearts dissected at the European Homograft Bank revealed only four cases (0.1%) of bicuspid pulmonary valves.

Bicuspid pulmonary valve with normal function can be difficult to identify at echocardiography. It can be well visualized by CMR and this can be combined with an assessment of the pulmonary arteries. It should be remembered as a rarer cause of dilated pulmonary artery when other causes have been excluded.
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Figure: A. Dilated MPA. B. Dilated RV. C. and D. Bicuspid pulmonary valve.