Sinus of Valsalva aneurysm and bicuspid aortic valve: detection and mechanism by cardiac magnetic resonance imaging

Jen Li Looi,1 Andrew J. Kerr1,2
1Department of Cardiology, Middlemore Hospital; 2Centre for Advanced MRI, The University of Auckland, Auckland, New Zealand

Abstract

Cardiac magnetic resonance imaging (CMR) demonstrated a sinus of Valsalva aneurysm (SVA) with severe dilatation of the right coronary sinus in association with a congenital bicuspid aortic valve (BAV) and sub-aortic membrane. The SVA had not been apparent on echocardiography as the dilatation was outside standard echo image planes. On both CMR and echo, blood flow was eccentrically directed into the right coronary sinus by the domed posterior leaflet of the BAV. The impact of the aortic jet on the wall of the right coronary sinus is probably important in the aetiology of the sinus dilatation. CMR proved valuable in demonstrating the SVA and understanding its aetiology.

Case Report

A 24-year-old man was diagnosed at birth with coarctation of the aorta and a patent ductus arteriosus and subsequently diagnosed with a bicuspid aortic valve (BAV) and a sub-aortic membrane. At the age of one month he had repair of the coarctation and closure of the ductus. In 1993 the BAV was noted on echocardiogram with a small sub-aortic membrane and a peak velocity of 1.6 m/s. Echocardiogram in 1994 showed a moderately dilated aortic root of 4.3 cm.

He re-presented to clinical attention in 2007. Repeat echocardiogram showed a BAV and sub-aortic membrane with no significant aortic regurgitation or aortic stenosis. There was moderate aortic sinus dilatation with a maximum diameter at 4.5 cm. This diameter is well under that at which surgical repair would be recommended. CMR was requested to better delineate the aortic dilatation. Surprisingly, the CMR demonstrated a sinus of Valsalva aneurysm (SVA), which had not been apparent on the echo images because it was outside standard imaging planes (Figure 1A, 1B; Supplementary files 1 and 2). There was aneurysmal dilatation of the right coronary sinus with a maximum dimension across the aortic sinus of 5.7 cm. On CMR flow was directed eccentrically by the domed posterior leaflet of the BAV into the right coronary sinus (Figure 1D; Supplementary file 3). The jet impacts on the supero-lateral wall of the right coronary sinus (Figure 1C). The jet eccentricity was also demonstrated on echo (Figure 2; Supplementary file 4). The sub-aortic membrane may also contribute to the eccentricity of the jet (Figure 2) by directing flow towards the belly of the posterior leaflet rather than more centrally.

He underwent successful repair of the SVA and resection of the sub-aortic membrane and remained well when followed up in clinic.

Correspondence: Jen Li Looi, Department of cardiology, Middlemore Hospital, Private Bag 933111, Otahuhu, Auckland, New Zealand. E-mail: ljl517@yahoo.co.nz

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Figure 1. A) CMR Short axis view using True-FISP demonstrating bicuspid aortic valve with a large postero-medial leaflet and smaller anterior leaflet (arrow). B) CMR short axis view (True-FISP) obtained just above the aortic valve plane demonstrating sinus of Valsalva aneurysm with aneurysmal dilatation of the right coronary sinus (arrow). C,D) An eccentric jet of flow directed eccentrically by the domed aortic valve leaflet (black arrow) into the right coronary sinus (white arrow).
Discussion

SVA are rare cardiac anomalies, which may be acquired or congenital. Congenital aneurysms are often caused by weakness at the junction of the aortic media and the annulus fibrosus. Acquired aneurysms are caused by conditions affecting the aortic wall, such as infections, degenerative diseases (atherosclerosis, connective tissue disorders or cystic medial necrosis) or trauma. The presence of BAV is associated with ascending aortic dilatation due to associated cystic medial necrosis. The importance of abnormal blood flow patterns and aortic dilatation in BAV remains unclear. den Reijer et al. recently provided new insights into the pathophysiology underlying aortic dilatation in BAV patients using CMR. The authors found a significant correlation between the systolic LV outflow jet angle with the aortic root channel and dilatation at the levels of the SVA, sinotubular (ST) junction, and the ascending aorta which could be important in the formation of aortic dilatation in BAV.

Our case describes the formation of SVA associated with an LV outflow jet which is eccentrically directed by a sub-aortic membrane through the BAV and into the right coronary sinus. It is probable that the asymmetric sinus dilatation is the result of many years of LV outflow impacting on the congenitally abnormal wall of the right coronary sinus.

Because it is out of the normal echocardiogram planes, the SVA may have been developing over many years but it has not been noted previously. CMR proves to be a valuable imaging modality in demonstrating the SVA and the likely mechanism of its development. CMR is useful in defining the three-dimensional anatomy of the aneurysm more precisely, including variations of flow direction during cardiac cycle. In addition, CMR may also provide better evaluation of aneurysm wall thickness or presence of thrombus, and better detection of small perforations.

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