Quadricuspid aortic valve associated with unruptured sinus of Valsalva aneurysm

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Quadricuspid aortic valve is a rare congenital anomaly. It may present as an isolated anomaly but is occasionally associated with aortic regurgitation. Sinus of Valsalva aneurysm (SVA) is also an infrequent congenital anomaly, typically associated with tricuspid aortic valves. There are only a few reported cases of SVA (ruptured) associated with quadricuspid aortic valves in the literature. We report the first case of the association of quadricuspid aortic valve with an unruptured SVA in an adult patient.

KEYWORDS
Quadricuspid Aortic Valve; Sinus of Valsalva Aneurysm; Echocardiography

Case report
A 40-year-old African-American female presented with shortness of breath and haemoptysis. Past medical history was unremarkable. Pertinent exam findings included a blood pressure of 118/40 mmHg, a water hammer pulse, jugular venous distension to the angle of jaw, a prominent early diastolic murmur in the aortic area, and a hyperdynamic apical impulse. A transthoracic echocardiogram showed a possible quadricuspid aortic valve, sinus of valsalva aneurysm (SVA) and severe aortic regurgitation. These findings were confirmed on transesophageal echocardiography (Figure 1A and B) and later intraoperatively, following which, our patient uneventfully underwent surgical resection (Figure 2) of the SVA and aortic valve replacement with a 21 mm Carbomedics mechanical valve.

Figure 1  (A and B) Transesophageal echocardiogram (TEE) short axis views in the vicinity of the aortic valve plane, depicting the four symmetric cusps and SVA (A, left panel) and following agitated saline injection (B, right panel), delineating the shape and extent of the windsock SVA (~2.5 cm in length), and origin from the accessory sinus of Valsalva (4th sinus).

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Discussion

Quadricuspid aortic valve is a rare congenital anomaly that predominantly affects males and has a reported incidence of 0.008–0.033%.\textsuperscript{1} Hurwitz and Roberts\textsuperscript{2} classified quadricuspid valves based on cusp size and degree of cusp equality into seven subtypes. The competency of a quadricuspid valve has been shown to be related to aortic cusp morphology and symmetry. The risk of aortic insufficiency and infective endocarditis is reportedly lowest in valves with four symmetric cusps.\textsuperscript{1} It is estimated that ~ 44% of quadricuspid valves have associated aortic insufficiency.\textsuperscript{3} Congenital SVA is a very rare anomaly with a reported incidence of 0.09% in the US population.\textsuperscript{4} SVA is associated with multiple conotruncal congenital malformations because of a single embryologic event affecting conbulbar septation and aortic arch development occurring at the level of neural crest development.\textsuperscript{4} Only a few published cases\textsuperscript{5,6} have described the unusual association of a quadricuspid aortic valve with SVA, all of which reported ruptured SVAs. To the best of our knowledge, our case is the first to report the association of quadricuspid valve with an unruptured SVA (see supplementary data online). Our case illustrates the utility of conventional and contrast echocardiography in precisely defining the morphologic abnormalities preoperatively and emphasizes the importance of early recognition in the prevention of potential complications such as SVA rupture and infective endocarditis.

Supplementary data

Supplementary data are available at European Journal of Echocardiography online.

Conflict of interest: none declared.

References


Figure 2  Operative specimen confirming preoperative diagnosis of SVA and quadricuspid morphology of the aortic valve.