Repair of Bicuspid Aortic Valves and Associated Aneurysms

from the Aortic Valve Repair Program
in collaboration with Professor Hans-Joachim Schäfers

The most common configuration of the aortic valve is tricuspid. In about 2% of the population the aortic valve is bicuspid and less frequently can be unicuspid or has a quadricuspid configuration (fig.1).

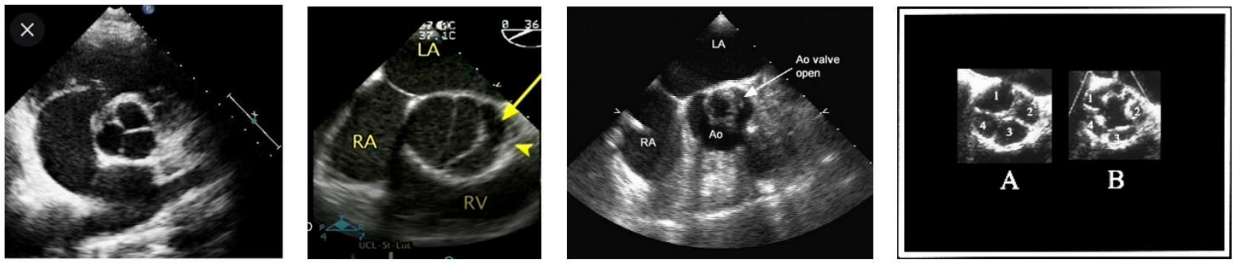


Fig 1: TEE of tricuspid, bicuspid, unicuspid and quadricuspid valves.

Bicuspid aortic valve (BAV) is the most common congenital cardiac anomaly and is three times more likely to affect men. Most patients with BAV can develop complications during their lifetime either from valve dysfunction or from expansion of the aorta (aortopathy). The valve dysfunction can be either aortic stenosis (AS) or aortic insufficiency (AI). The average age of BAV patients presenting with predominant AS is 65 yrs. Those with AI present at an earlier age (average 46 years) are usually associated with aortic annular dilatation (1).

About 30% of BAV patients present with aortopathy in various combinations of expansion of the aortic sinuses (aortic root), ascending aorta and aortic arch (Fig 2) (2).

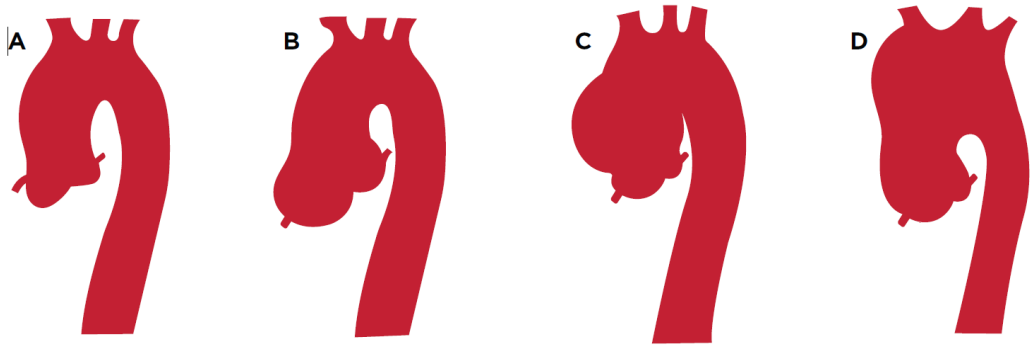


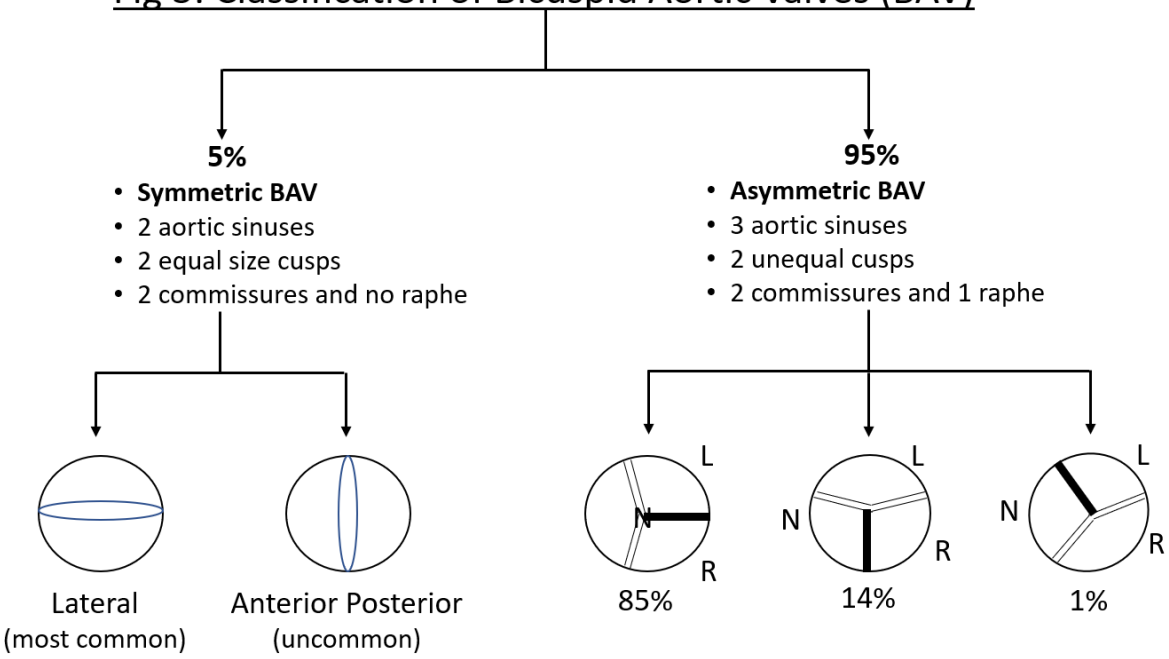
Fig 2: Patterns of aortopathy in patients with bicuspid aortic valve.
(A) Normal aorta **(B)** Aortic root aneurysm **(C)** Ascending aortic aneurysm **(D)** Aortic root, ascending & arch aneurysm

Variations in the configuration of BAV

The bicuspid valve is composed of two leaflets. On echo imaging, the two commissures reach all the way to the annulus. It should be differentiated from a unicuspid valve which can appear like bicuspid on echocardiography, but only one commissure reaches the annulus (Fig.1).

Only in 5% of the BAV patients, the two leaflets are equal without any raphe-symmetric BAV (1). In 95% of the patients the two leaflets are unequal in proportion, and the fused leaflet has a raphe (fused rudimentary commissure) - asymmetric BAV. The most common pattern of fusion is left and right-85%, followed by right/non-14% and most infrequently non/left-1%. (Fig. 3).

Fig 3: Classification of Bicuspid Aortic Valves (BAV)



The commonly quoted classification of bicuspid aortic valves by Sievers, type 0, type I and II is inaccurate. As mentioned above, type 0 (equal leaflets with no raphe in either leaflet) is present in only 5% of patients. Type I (one normal size and one fused larger leaflet with a raphe) is present in the remaining 95% of patients. Type II is actually a unicuspid valve, which is frequently misdiagnosed as a bicuspid valve.

Patients with any regurgitant bicuspid valve with pliable leaflets and with or without associated aortic aneurysms can be repaired. Repair if done well is ideal to replacement as there is no ideal valve substitute for these young patients, and the results are excellent (4). Our surgical approach is summarized in Fig. 4.

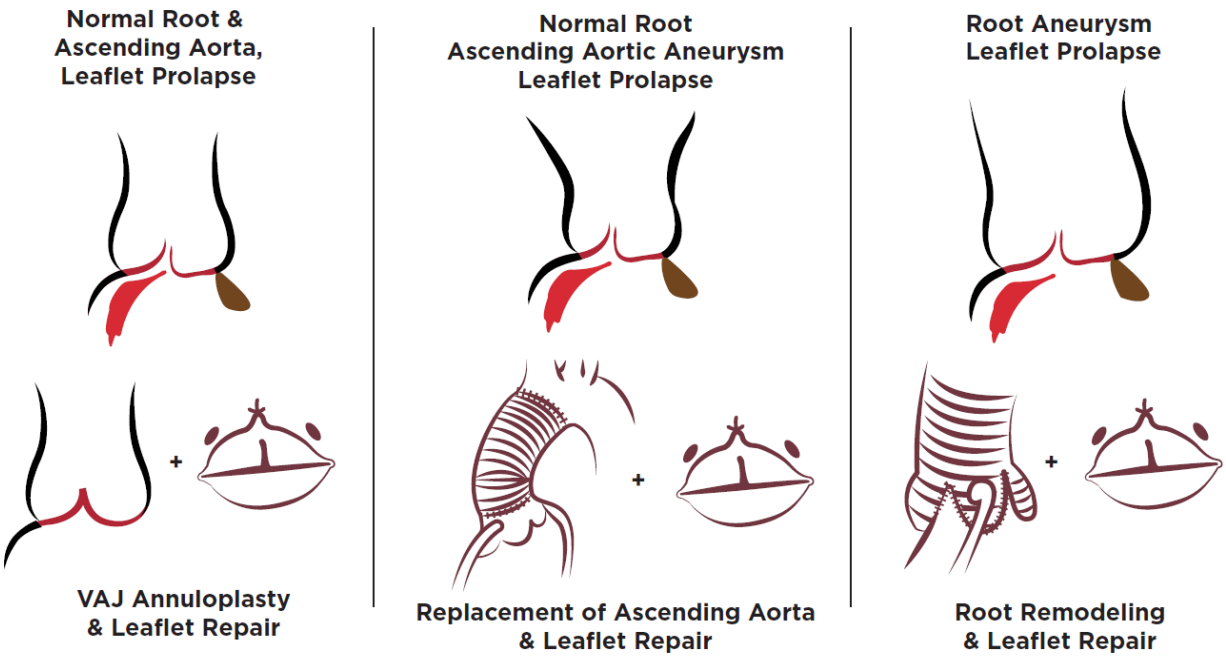


Fig. 4: Associated aortopathy and surgical management.

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To refer a patient, please call 925-676-2600 or Ramesh Veeragandham, M.D. at 925-336-7774.

References:

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