Bicuspid Aortic Valve Disease

A request for specific ICD-10-CM coding

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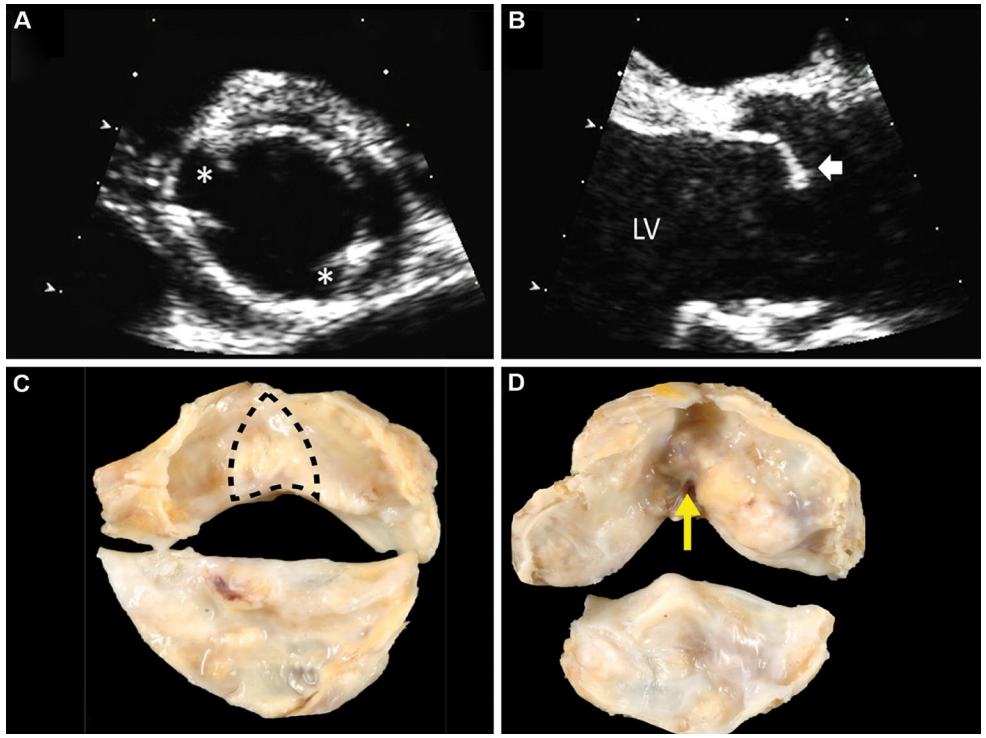
BAV Disease

- BAV - the aortic valve has two, not the typical three, cusps / leaflets
- Most common congenital heart valve defect, occurring in approximately 1 – 1.5% of the population



Michelena HI. Published Online: July 22, 2021
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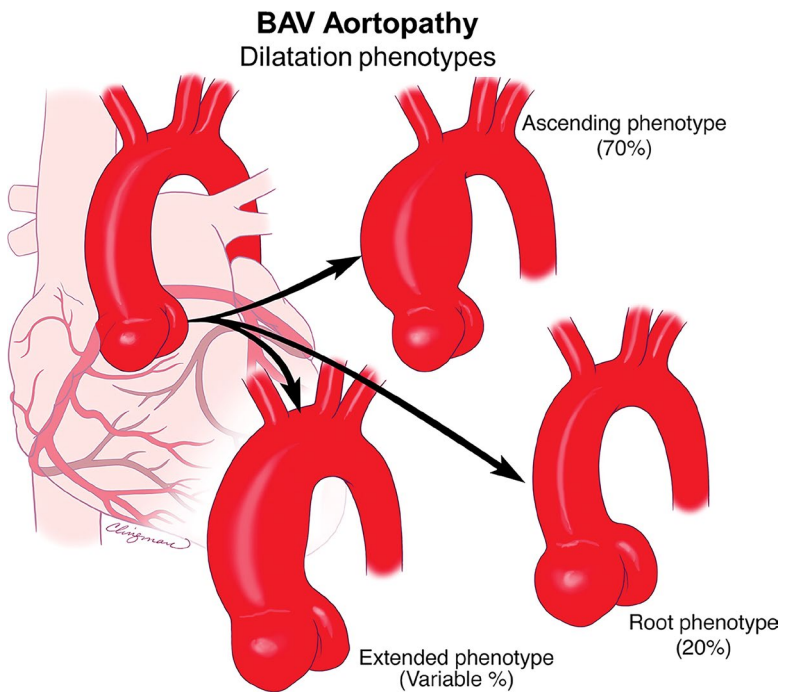
The Functional Consequences of BAV

- The functional consequences of BAV are:
  - The aortic valve becomes stenotic or incompetent in ~80% of BAV individuals during their lifetime
  - The bicuspid aortic valve has an increased risk of endocarditis
  - The ascending aorta of individuals with BAV is often aneurysmal with higher risk of aortic dissection and/or rupture



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BAV is associated with:

- ~ 80% lifetime risk of requiring aortic valve surgery ~19 years earlier than TAV
- ~ 20% lifetime risk of requiring aortic surgery for aortic aneurysm / dissection
- A mortality burden [mortality hazard ratio = 2.29 (1.93 - 2.72)]
- Personal and societal impact
- Increased risk of BAV and aortic disease in first degree relatives
- 36 associated gene loci, each with low population level impact

Societal Cost

- ~ 45% of aortic valve replacements are for a stenotic BAV, most commonly in late 50`s
- >100,000 AVRs per year in the US

- Without BAV-specific codes, how can we identify an individuals' outcomes specific to BAV and design interventions that are optimized for BAV patients?
- Improved coding will facilitate more efficient and responsible use of resources.

Guidelines and Implementation

- AHA and ACC Guidelines recommend longitudinal imaging of the aortic valve and ascending aorta (echocardiogram, CT scan, or MRI) in people with BAV
- Further, these Guidelines recommend echocardiographic imaging of first-degree relatives of probands with BAV
- These Guidelines are not rigorously followed because of difficulty implementing system-wide longitudinal monitoring, which a specific code would improve
- For example, of the 10,810 individuals with known BAV within the Mass General Brigham Healthcare System (Boston, MA), 55% do not have a recorded ICD-9-CM or ICD-10-CM code for BAV (e.g., from category Q23 or I35)

1. 2020 ACC/AHA Guideline for the Management of Patients with Valvular Heart Disease: A report of the American College of Cardiology/American Heart Association Joint Committee on Clinical Practice Guidelines. https://doi.org/10.1161/CIR.0000000000000923
2. 2022 ACC/AHA Guideline for the Diagnosis and Management of Aortic Disease: A Report of the American Heart Association/American College of Cardiology Joint Committee on Clinical Practice Guidelines. https://doi: 10.1016/j.jacc.2022.08.004.

Potential Patient Benefits from Improved ICD-10-CM Coding

- Higher rate of BAV individuals who are correctly coded

- Ability to generate individual guidance for timely long-term follow-up
  - Follow imaging Guidelines from AHA / ACC *et al*.
  - Generate automated Physician and Patient follow-up reminders
- Improve identification of first-degree relatives of probands with BAV
- Improved ability to perform rigorous system-wide research

Summary

- Bicuspid aortic valve is a frequent cause of morbidity and mortality
- BAV has direct personal impacts on those affected, and societal impacts related to loss of productivity as well as health care needs and family impacts on those affected (e.g., aortic stenosis can lead to heart failure if not treated)
- Guidelines recommend routine individual follow-up and family screening
- Improved ICD-10-CM coding of BAV will facilitate these improved Guideline adherence