

BICUSPID AORTIC VALVE

EPIDEMIOLOGY AND PATHOPHYSIOLOGY

1-2% population

Men >>> women (70-80% cases)

Familial clusters – [autosomal dominant inheritance with incomplete penetration \(20-30%\)](#)

2 cusps:

- Fusion of R and L cusps in 70-80%
- Fusion of R and NC cusps in 20-30% (more dilatation of ascending aorta)
- Fusion of L and NC = rare

Diagnosis based on systolic opening of the valve with only 2 commissures in TTE

Unicuspid valves: 1 leaflet with 1 aortic commissure

Associated with **aortopathy, dilatation of ascending aorta and sinus of Valsalva**

- Related to valve morphology but not the severity of valve dysfunction
- Risk of aortic dissection 5-9 times higher than general population

CLINICAL MANIFESTATIONS

Normal valve function until later in life – related to valvular dysfunction

Risk factors of cardiac events: [> 30 y-o, moderate to severe AR, and moderate or severe AS](#)

- **20%** of bicuspid will develop severe AR requiring AVR between 10-40 y-o
- Increase risk of endocarditis
- Severe stenosis later in life > 50 y-o
 - o Pathology identical to trileaflet valves **BUT**
 - o **Earlier** in life because of turbulent flow through abnormal architecture that cause accelerated valvular changes leading to AS

DIAGNOSIS & FOLLOW-UP

No specific gene testing.

Recommendation: [ETT - 1st degree family members if associated aortopathy, family history of VHD or aortopathy.](#)

Angio CT and MRI – aortic diameters 1-2 mm larger because it includes the aortic wall

★ **Suggested reference:** Nishimura R. A. et al. 2014 ACC/AHA guidelines for the management of patients with valvular heart disease. J Am Coll Cardiol 2014;63:e57-185.

TREATMENT

- No medical treatment proven to reduce the aortic dilatation progression
- Treat if hypertension

Surgical intervention

★ **Suggested reference:** Nishimura R. A. et al. 2014 ACC/AHA guidelines for the management of patients with valvular heart disease. J Am Coll Cardiol 2014;63:e88-90.

Content of this summary from these references:

- Otto C & Bonow R. Valvular Heart Disease. (2012) In Bonow R. *et al.* Braunwald's Heart Disease, 9th edition, pp. 1468-1539. Philadelphia, PA: Elsevier.
- Nishimura R. A. et al. 2014 ACC/AHA guidelines for the management of patients with valvular heart disease. J Am Coll Cardiol 2014;63:e57-185.