Mimicer of aortic stenosis: subvalvular stenosis

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Case presentation:

30M with history of bicuspid aortic valve and family history of congenital heart disease. He is asymptomatic.
- Mid to late peaking systolic cres-decrescendo murmur at RUSB
- Absence of systolic ejection click
- Soft holodiastolic murmur
- No radial femoral delay
TEE in deliniation of subvalvular obstruction


Discrete subaortic membrane

- Discrete fibrous ring or fibromuscular narrowing
- M:F ~ 2:1; 6.5% of all ACHDs
- May extend to the anterior leaflet
- Usually a solitary congenital defect but may be associated with other defects (VSD, BAV, coarctation)
- Triad: Subaortic membrane, VSD and double chamber RV
- Never seen in infancy
- May be acquired (after VSD patching)


- Septal hypertrophy may develop below the level of the membrane
- Obstruction is usually fixed, but may be dynamic if septal hypertrophy is present
- Aortic valve degeneration occurs and aortic regurgitation develops in 50%
When to intervene?

Warnes et al.  
ACC/AHA 2008 Guidelines for ACHD.

- Class I: Surgical intervention is recommended
  - Peak instantaneous gradient > 50mmHg or
  - Mean gradient of 30mmHg
  - Or lesser gradients with progressive AR or LVEFd >50mm

- Class IIb: Surgery may be considered with lesser gradients if:
  - LVH
  - Pregnancy is planned
  - Competitive sports

- Catheter based intervention is not recommended

Surgical complications:

- Injury to aortic or mitral valve (membranes that extend to the anterior leaflet) and need for mitral valve reconstruction or replacement

- Complete heart block

- Creation of a VSD

- Endocarditis
Rule of 15%....

• 15% familial
• 15% of all congenital LVOT obstruction
• 15% risk of endocarditis
• 15% may recur post operatively