Simple Congenital Defects
BAV and ASD

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No Disclosures
Common “Simple” Defects

Bicuspid Aortic Valve

Atrial Septal Defect
Bicuspid Aortic Valve

- Echocardiography
- Magnetic Resonance
- Computed Tomography
Bicuspid Aortic Valve: A Simple Lesion?

“BAV disease is responsible for more deaths and morbidity than all other congenital heart defects combined”

Verma S et al, Toronto
A-P orientation
Fusion of the R and L coronary cusps

R-L orientation
Fusion of the R and N coronary cusps
A-P orientation
Fusion of the R and L coronary cusps
Type 1

R-L orientation
Fusion of the R and N coronary cusps
Type 2

80%

19%
Fused Raphe’

Present in ~30% of cases
Long-Axis View Doming

Mostly appreciated in Type 1 (R-L fusion)
How Common is Bicuspid Valve?

What is the Incidence of BAV?

a) 0.5%
b) 0.8%
c) 1%
d) 2%
e) 4%
How Common is Bicuspid Valve?

Screening of all newborns in Ankara, Turkey

\[
\frac{5 \text{ BAV found}}{1,075 \text{ neonates}} = 0.46\%
\]

0.71% in males
0.19% in females

Tutar E, Ekici F, Am Heart J, 2005, 150(3); 513-515
Valve Complications

Stenosis  present in 50%  24% will need surgery
Regurgitation  ~ will need surgery
Endocarditis  2%
Aortic Enlargement  present in 50%  5% will need surgery  0.03% dissect

Michelena H, Circulation. 2008 May 27;117(21):2776-84
Valve Function

Tricuspid
Valve Function

Bicuspid  Tricuspid  Quadricuspid
Valve Function

Bicuspid  Tricuspid  Quadricuspid
Valve Function

Bicuspid  Tricuspid  Quadricuspid
Valve Stenosis

Leaflet motion may be better than the measured gradients
Valve Regurgitation

Jets may be eccentric and difficult to gage severity
Endocarditis

Prophylactic antibiotics no longer recommended before dental procedures
Bicuspid Valve Aortopathy

Aortic diameter will be *larger* at nearly every level.

Aortic Enlargement Patterns

- None
- Root
- Mid Ascending
How Common is Aortic Enlargement?

Screening of Recruits for Italian Police Force

66 with BAV and normal function
34 (52%) had abnormal aortic root

Prevalence
- Ascending: 43.9%
- ST junction: 15%
- Sinuses: 19.6%

5 people in 100 of general population

Factors Related to Enlargement

Diameter increases with age


Expected growth of 0.5 – 0.9 mm/year


Rate of growth increases exponentially

Of all dissections, how many are in BAV patients?

6 – 9%

Of all BAV patients, how many will have a dissection?

~0.1 – 0.2%
Measuring the Aorta
The Problem: Excessive Variation

Reported diameter vs. measured diameter

Measured maximum diameter (MRI core)

Reported maximum diameter (chart)
Echo, MRI, and CT all done in same day:

Inter-modality Differences

Difference (mm)

MRI maximum (mm)

echo difference  CT difference
Inner vs. Leading Edge

2015 ASE (Adult)
Inner Edge Technique

2010 ASE (Pediatric)
Leading Edge Technique

Son K and Oh J, Cardiovascular Ultrasound 2013, 11:28
Diastole vs. Systole

2015 ASE (Adult)
Inner Edge Technique
Diastole

2010 ASE (Pediatric)
Leading Edge Technique
Systole

Son K and Oh J, Cardiovascular Ultrasound 2013, 11:28
2015 ASE (Adult)
“Root measurements should be obtained from the parasternal long-axis view”
“This plane is slightly different from the parasternal long axis of the left ventricle.”

Cusp-cusp vs. Commissure-cusp

For annular sizing requires both (TAVR experience)

For root diameter, recommendations do not specify

Most report the widest diameter measured (cusp to cusp)

Thoracic Aortic Guidelines (2010)

Measurements of aortic diameter should be:
“taken at reproducible anatomic landmarks”
“perpendicular to the axis of blood flow”
“direct image to image comparison”

No mention of
Diastole vs. systole
Edge selection
Imaging planes

Hiratzka, JACC 2010 55 (14):e27-129
When to Intervene

<table>
<thead>
<tr>
<th>Indications for elective aortic surgery</th>
</tr>
</thead>
<tbody>
<tr>
<td>&gt;55 mm</td>
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<tr>
<td>50 - 54 mm</td>
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<td></td>
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<td>&gt;45 mm</td>
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Also consider expertise of surgical team

Common “Simple” Defects

- Bicuspid Aortic Valve
- Atrial Septal Defect
Atrial Septal Defect
## Intra-atrial Defects

Communication between the right and left atria

<table>
<thead>
<tr>
<th>ASD</th>
<th>PFO</th>
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<tbody>
<tr>
<td>Located Anywhere</td>
<td>At the foramen ovale</td>
</tr>
<tr>
<td>Continuously open hole</td>
<td>Flap-like communication</td>
</tr>
<tr>
<td>Allows continuous left-to-right shunt</td>
<td>Allows intermittent right-to-left shunt only</td>
</tr>
<tr>
<td>Causes chamber enlargement</td>
<td>Chamber sizes are normal</td>
</tr>
</tbody>
</table>

“Stretched out” PFO such as with atrial septal aneurysm
Intra-atrial Defects

Cleveland Clinic (http://my.clevelandclinic.org/heart/disorders/congenital/pfo.aspx)
Intra-atrial Defects

Cleveland Clinic (http://my.clevelandclinic.org/heart/disorders/congenital/pfo.aspx)
Atrial Septal Defect

Different Types of Defects

- sinus venosus
- secundum
- caval
Viewing the Defect
Negative contrast from Coronary Sinus or IVC are expected
2.5.2. Recommendations for Interventional and Surgical Therapy

CLASS I

1. Closure of an ASD either percutaneously or surgically is indicated for right atrial and RV enlargement with or without symptoms. *(Level of Evidence: B)*
RV Enlargement

Parasternal Long Axis

Parasternal Short Axis
RV Enlargement
Estimating RV size

Rudski LG, J Am Soc Echocardiogr 2010;23:685-713
Transesophageal Echo
Transesophageal Echo

Need to evaluate adequacy of rims
Transesophageal Echo

Need to evaluate adequacy of rims
3D visualization

Rim is often too thin, images suggest larger defect than actual, or multiple holes
Multiple ASDs
Transesophageal Echo

Bi-Caval view on TEE
Look for this in setting of unexplained RV enlargement.

Usually associated with anomalous right upper pulmonary venous drainage
Conclusions

BAV

Note leaflet orientation
Thank You
Visualization