Complex Non-Cyanotic Congenital Heart Disease

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Most of the complex non cyanotic CHD in adults are post repaired (eg post op TGA – Atrial and Arterial Switch, post op Fontan) and will be covered by Dr Broberg in the next talk.

Current talk with focus on non cyanotic CHD of moderate complexity.
Non-Cyanotic Congenital Heart Disease of moderate complexity

1. Atrioventricular Septal Defect (AVSD)
2. Ebstein Anomaly
3. Coarctation of the Aorta (CoA)
4. Shone Complex
1. **Atrioventricular Septal Defect (AVSD)**

   also known as

   - Atrioventricular Canal Defect (AVC)
   - Endocardial Cushion Defect
The essential morphological hallmark of an AVSD is

- The presence of a **COMMON atrioventricular junction** as compared to the **SEPARATE RIGHT and LEFT atrioventricular junction** in a normal heart.
What is an atrioventricular septal defect?

Diagram taken from ACHD Learning Centre Website; Hans Harmer : AVSD

1) Right anteriosuperior leaflet
2) Right Inferior Leaflet
3) Superior Bridging Leaflet
4) Inferior Bridging leaflet
5) Left Mural Leaflet

Important morphological features of AVSD

- Single common atrioventricular ring
  - Single orifice (Complete AVSD)
  - Dual Orifice
    - Partial / Incomplete
    - Transitional / Intermediate

- Site of shunting
  - Atrial and Ventricular (Complete AVSD)
  - Atrial only (partial AVSD)
  - Atrial mainly and small restrictive ventricular component (Intermediate / Transitional AVSD)

- Un-wedging of the aorta with elongation of LVOT
  - gooseneck deformity and possible LVOT obstruction
Shunting across an AVSD with common atrioventricular junction is dependent upon the attachment of the bridging leaflets to the septal components.

- If free floating with no attachment, then shunting at 2 levels through primum ASD and inlet VSD (complete AVSD)
- If attached to ventricular septum, then shunting through primum ASD only (partial AVSD)
- If attached to atrial septum, then shunting through inlet VSD only (rarer)
The Rastelli classification (1966) describes 3 types of complete AVSD based on the morphology of the superior bridging leaflet, its degree of bridging and its chordal attachments:

- **In Rastelli type A defect**, superior bridging leaflet (SL) is split into 2 with left portion (LSL) located entirely over the LV and the right portion (RSL) located entirely over the RV, chordal attachment to crest of ventricular septum.

- **In the rarer Rastelli type B defect**, LSL portion is larger and crosses over to RV side with anomalous papillary muscle attachment from the right of the ventricular septum to the LSL.

- **In Rastelli type C defect**, SL is not divided into left or right portion and floats without chordal attachment to the crest of the ventricular septum.

*Ann Thorac Surg 2000;69:S337*
AVSD (Atrioventricular Septal Defect)

• Incidence of AVSD varies from 0.24 – 0.31/1000 live births

• AVSDs accounts for 2.9-6.2% of all CHD

• Strong association with Down’s syndrome, 1/3 of patients with Down’s syndrome had a complete AVSD and 5% had an ostium primum ASD *(Marino B et al. Am J Dis Child 1990;144:1120–2)*

• Common Atrioventricular (AV) Junction with common valve (5 leaflets) which can have 1 or 2 separate orifices depending if bridging leaflets are free floating or attached to atrial or ventricular septum

• The left and right atrioventricular (AV) valves are not and should not be called mitral and tricuspid valves. Cleft in the left-sided AV valve

• Aorta unwedged and anteriorly positioned either inlet-outlet disproportion -> Goose neck appearance of LVOT
AVSD (Atrioventricular Septal Defect)

Defects associated with AVSD

- **LVOT obstruction** from un-wedging of the aorta or secondary to chordal attachments from LAVV across the LVOT

- **Tetralogy of Fallot (5%)**

- **Hypoplasia of one ventricle** especially with unbalanced AVSD
  - Eg. Unbalanced AVSD with Dominant RV and hypoplastic LV and aorta
  - Eg. Unbalanced AVSD with Double inlet LV, hypoplastic RV and pulmonary atresia

- **Others defects – PDA, CoA**
Approach to echo imaging in AVSD

1. Assess the primum ASD
   • Size, direction of shunt

2. Assess the inlet VSD
   • Size, intact, direction of shunt, restrictive

3. Assess the AV valve/valves
   • Common, dual orifice
   • Regurgitation severity
   • Dual orifice of the Left AV valve
   • Rastelli classification

4. Balance or unbalance AVSD
   • Distribution of AV valve over the ventricles
   • Ventricular hypoplasia

5. Others
   • LVOT obstruction (goose neck)
   • RVOT obstruction (TOF)
   • PDA, CoA, etc
Complete AVSD
- Elongated LVOT
- Large VSD
- AVV with chordae attachments across LVOT
Uncorrected complete AVSD in an ACHD patient
- Pulmonary hypertension with mPAP 67 mmHg
Complete AVSD
- A4C best view for single AV valve in a common AV ring
- Large inlet VSD
- Large primum ASD
Partial AVSD

- Elongated LVOT
- No LVOT obstruction
- Dilated RV
Partial AVSD

- Left AV valve on the left
- Right AV valve below

NB: They should not be called mitral or tricuspid valve
Partial AVSD
- Common AV junction
- Primum ASD with left to right shunting
- Right and left AVV regurgitation
- RA and RV dilatation
- No LVOT obstruction
TEE Partial AVSD
- Primum ASD with L-R shunt
- Common AV junction
- Right AVV regurgitation
- 3D view of the left AVV showing superior and inferior bridging leaflets and left mural leaflet
Partial AVSD with slight unbalanced ventricles

- Primum ASD
- Inlet VSD closed by ventricular septal aneurysm
- 2 separate AV valves, larger left and smaller right
- Straddling of chordae
### Table 5  Indications for intervention in atroioventricular septal defect

<table>
<thead>
<tr>
<th>Indications</th>
<th>Class</th>
<th>Level</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Complete AVSD:</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>• Cardiac surgery must be avoided in patients with Eisenmenger physiology. In case of doubt, PVR testing is recommended. For indication of intervention see also VSD (Section 4.2)</td>
<td>III</td>
<td>C</td>
</tr>
<tr>
<td><strong>Partial AVSD:</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>• Surgical closure should be performed in case of significant volume overload of the RV. For further details see ASD (Section 4.1)</td>
<td>I</td>
<td>C</td>
</tr>
<tr>
<td><strong>AV valve regurgitation:</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>• Symptomatic patients with moderate to severe AV valve regurgitation should undergo valve surgery, preferably AV valve repair</td>
<td>I</td>
<td>C</td>
</tr>
<tr>
<td>• Asymptomatic patients with moderate or severe left-sided valve regurgitation and LVESD &gt;45 mm and/or impaired LV function (LVEF &lt;60%) should undergo valve surgery when other causes of LV dysfunction are excluded</td>
<td>I</td>
<td>B&lt;sup&gt;35&lt;/sup&gt;</td>
</tr>
<tr>
<td>• Surgical repair should be considered in asymptomatic patients with moderate or severe left-sided AV valve regurgitation who have signs of volume overload of the LV and a substrate of regurgitation that is very likely to be amenable for surgical repair</td>
<td>IIa</td>
<td>C</td>
</tr>
<tr>
<td><strong>SubAS:</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>• See Section 4.5.3</td>
<td></td>
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</tr>
</tbody>
</table>

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Summary of echo imaging in AVSD

• The hallmark of an AVSD is the presence of a common atrioventricular junction and ring.

• Classification into one of the 3 types of AVSD:
  • Complete AVSD
  • Partial AVSD and
  • Intermediate / Transitional AVSD

• A complete echo study of an adult AVSD patient should provide information on all aspects of the atrio-ventricular leaflets including chordal attachments, assessment of AV regurgitations, level and degree of shunting, LVOT obstruction, pulmonary hypertension etc.
2. Ebstein Anomaly
Wilhelm Ebstein

- German physician
- Born 27/11/1836, died 22/10//1912
- First published description in 1866 on a 19 year old man with history of palpitations, dyspnoe, cyanosis and cardiomegalgy
- At autopsy, Ebstein described ‘an enlarged and fenestrated anterior leaflet; hypoplastic and adherent septal and posterior leaflets with atrialized portion of RV and presence of PFO’

The essential morphological features of Ebstein Anomaly are

- Greater apical displacement of the TV annulus due to failure of delamination of the septal and posterior leaflets
- Anterior TV leaflet is redundant with tethering
- RV is atrialized with variable degree of RV dysplasia
• Incidence 1-5 per 200,000; < 1% of all CHD

• Most cases are sporadic, familial Ebstein is rare

• In rare cases, association with maternal lithium therapy

• Inferior displacement of the proximal attachments of the septal and the posterior leaflets of the TV valve from the atrio-ventricular ring
• 3 tricuspid valve leaflets – Septal (S), Anterior (A) and Posterior (P) leaflets
• 3 papillary muscles
  - s = Septal papillary muscle – joins septal and anterior leaflet, usually rudimentary
  - a = Anterior papillary muscle – joins anterior to posterior leaflet
  - p = Posterior (Inferior) papillary muscle – joins septal and posterior leaflet
• In Ebstein, there are no cords to suspend the septal and posterior leaflets, they arise from RV
• TV valve opening is more horizontal than vertical in position, resulting in atrialization of RV inlet area and reduction in functional RV size
PATHOLOGICAL FEATURES OF EBSTEIN ANOMALY

1. Adherence of TV leaflets to myocardium
2. Apical displacement of functional annulus (septal>posterior>anterior)
3. Dilatation of atrialized portion of RV with wall thinning
4. Anterior leaflet is often redundant, tethered and may be fenestrated with several accessory orifices
5. Chordae tendineae to the anterior leaflet are generally short and poorly formed
6. Dilatation of the true TV annulus (atrioventricular ring)
7. Inlet portion of RV is integrated with the RA (ARV - ‘atrialized portion of RV’) and functional RV constitute the trabecular and outlet portion of RV
**Ebstein Anomaly: Carpentier’s Classification (1988)**

**Type A** = Minimal displacement of the attachment of the septal leaflet. The atrialized RV is small.

**Type B** = Moderate displacement of the septal leaflet with a large atrialized RV. Mobile anterior leaflet.

**Type C** = Marked displacement of the septal and postero-inferior leaflet associated with a dyskinetic atrialized RV and restricted anterior leaflet motion.

**Type D** = Near-complete atrialization of the RV except for small infundibular component (Uhl's syndrome).

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*S. Chauvaud et al. EJCTS 13 (1998) 514–519*
Ebstein Anomaly

Associated cardiac anomalies

- PFO or ASD most common (80-90% in one reported series)
- Right ventricular outflow tract obstruction
- Association with chromosomal abnormalities are rare but have been reported in Trisomy 13 and Trisomy 21
- Other congenital lesions – VSD, PS, TOF, CoA, etc
- Wolf-Parkinson-White syndrome
- Can have left sided lesions in the setting of ccTGA with systemic RV on the left
Ebstein Anomaly: GOSE Score in Neonates

**GOSE (Great Ormond Street Ebstein) Score**
- Echo score used in neonates
- Published in 1992 by Celemajer and colleagues from GOS
- Ratio of area (RA + atrialized portion of RV) ÷ (functional RV + LV) from A4C view on echo at end-diastole

- Grade 1-4
  - Ratio < 0.5 = Grade 1
  - Ratio 0.5 to 0.99 = Grade 2
  - Ratio 1-1.5 = Grade 3
  - Ratio > 1.5 = Grade 4

- Grade 4 = 100% mortality
- Grade 1 or 2 = 92% survival
Approach to echo imaging in Ebstein Anomaly

1. Confirm Ebstein Anomaly
   • Confirm that the septal displacement index of the TV leaflet > 8mm/m2 index to BSA from insertion of anterior mitral leaflet

2. Assess and describe all the TV leaflets
   • Size, site of tethering, attachment of chordae, etc

3. Severity of TR
   • Image at different planes, severity can be under appreciated

4. RV size and function
   • Atrialized portion of RV (ARV)
   • Exclude RVOT obstruction from large anterior TV leaflet

5. Others
   • ASD, PFO, other lesions

Echo in Ebstein: 1) SEPTAL DIPLACEMENT INDEX

- The septal and posterior leaflets of the tricuspid valve are displaced inferiorly towards the RV apex (Displacement of septal TV leaflet >8mm/m2 index to BSA from insertion of anterior mitral leaflet)

\[
\text{Displacement index} = \frac{35}{1.6} = 22 \text{ mm/m}^2
\]
Echo in Ebstein: 2) FUNCTIONAL and TRUE ANNULUS

- The area of right ventricle between the true tricuspid annulus and the displaced attachment of the leaflets is thinned and dilated (atrialized)

- The remainder of the right ventricular cavity is usually small

True annulus (TA) 5.3cm, Functional annulus (FA) 7.4cm

Dimensions of the ‘true’ RA
Echo in Ebstein:
3) TRICUSPID REGURGITATION

- TR often results from incomplete coaptation between the large sail-like anterior leaflet and the tethered septal and/or posterior leaflet.
- Very rarely can also come from multiple fenestrations in the large anterior leaflet.
- Functional tricuspid annulus can be in > 1 plane, difficulty in imaging to see the full TR jet.
- RA large, cannot rely on TR jet area alone.
Functional tricuspid annulus can be in > 1 plane, difficulty in imaging to see the full TR jet.

Need to use all available echo windows and scan in different planes to see full TR jet.
In severe TR, CW Doppler echo is usually dense and triangular in shape, looking like aortic CW Doppler echo pattern.
• As RA is large, we cannot rely on TR jet area alone to assess severity of TR

• Hepatic vein systolic flow reversal seen on either colour or PW Doppler would also suggest severe TR

• Dilated and plethoric IVC is an indirect reflection of raised RA pressure
• Affects surgical management and prognostic outcome

• Accessory attachment (chords) and degree of tethering of each leaflet component will affect leaflet motion and alignment

• Result in significant residual TR even after repair
- The anterior leaflet is large and sail-like with abnormally numbered and placed chordal attachments
- The valve leaflets may be adherent to the right ventricular wall
In Ebstein’s Anomaly, it is often possible to see all 3 leaflets in one view.

Important to see the relationship of all 3 leaflets if TV repair is to be considered.

2 patients with Ebstein’s Anomaly presenting with different degree of leaflet involvement.
Echo in Ebstein: 4) RV DYSPLASIA

- RV dilatation in 2/3 of EA patients (atrialized portion of RV and also functional RV)
- RV wall thinning
- RV dyskinesia
  - paradoxical septal motion
  - paradoxical systolic expansion of atrialized RV
  - Decreased RV wall motion
- Paradoxical septal motion
- Paradoxical systolic expansion of atrialized RV
- RV wall thinning
- Decreased RV wall motion
Echo in Ebstein: 5) LV INVOLVEMENT

• LV systolic and well as diastolic function affected
  o Daliento L, Ho SY, etc. Angiographic and morphologic features of left ventricle in Ebstein’s malformation. Am J Cardiol 1997;80:1051-9

• Change in RV geometry can lead to paradoxical septal motion, regional contraction abnormalities and subsequent changes in LV shape, size and function

• LV filling pattern: Decrease E/A, prolong DT and IVRT

• Chronic cyanosis and RV overload can lead to LV interstitial fibrosis and subsequent diastolic dysfunction

• Association with LV non-compaction
Change in RV geometry can lead to paradoxical septal motion, regional contraction abnormalities and subsequent changes in LV shape, size and function.
Patient with Ebstein anomaly, atrial septal aneurysm and secundum ASD

In this patient, the shunt is mainly left to right.

Shunt can reversed from right to left when RA pressure is raised, resulting in cyanosis.
### Table 14: Indications for intervention in Ebstein’s anomaly

<table>
<thead>
<tr>
<th>Indications</th>
<th>Class</th>
<th>Level</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Indications for surgery</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Surgical repair should be performed in patients with more than moderate TR and symptoms (NYHA class &gt;II or arrhythmias) or deteriorating exercise capacity measured by CPET</td>
<td>I</td>
<td>C</td>
</tr>
<tr>
<td>If there is also an indication for tricuspid valve surgery, then ASD/PFO closure should be performed surgically at the time of valve repair</td>
<td>I</td>
<td>C</td>
</tr>
<tr>
<td>Surgical repair should be considered regardless of symptoms in patients with progressive right heart dilation or reduction of RV systolic function and/or progressive cardiomegaly on chest X-ray</td>
<td>IIa</td>
<td>C</td>
</tr>
<tr>
<td><strong>Indications for catheter intervention</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Patients with relevant arrhythmias should undergo electrophysiologic testing, followed by ablation therapy, if feasible, or surgical treatment of the arrhythmias in the case of planned heart surgery</td>
<td>I</td>
<td>C</td>
</tr>
<tr>
<td>In the case of documented systemic embolism probably caused by paradoxical embolism, isolated device closure of ASD/PFO should be considered</td>
<td>IIa</td>
<td>C</td>
</tr>
<tr>
<td>If cyanosis (oxygen saturation at rest &lt;90%) is the leading problem, isolated device closure of ASD/PFO may be considered but requires careful evaluation before intervention (see text)</td>
<td>IIb</td>
<td>C</td>
</tr>
</tbody>
</table>

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Summary of echo imaging in Ebstein

• Ebstein Anomaly is a complex form of congenital heart disease with a wide spectrum of morphological features and presentation

• Accurate assessment of the anatomical and hemodynamic features can be accurately obtained with echocardiography

• A complete imaging of Ebstein Anomaly should provide information on all the TV leaflets, the degree of TR, RV and LV functional assessment, the presence of other associated cardiac lesion and the reparability of the TV valve
3. Coarctation of the Aorta (CoA)
COARCTATION of the AORTA (CoA)

- 5-8% of all congenital heart defects
- Male predominance 1.5:1
- Coarctation means ‘drawing together’
- 2 types
  - Discrete, localized shelf of endothelial tissue
  - Diffuse, tubular hypoplasia
- Most commonly located at the aortic isthmus, between LSA and PDA
COARCTATION of the AORTA (CoA)

• **Simple CoA** – Isolated CoA

• **Complex CoA** – Associated with
  # Bicuspid aortic valve
  # VSD
  # PDA
  # Mitral valve anomalies
  # Turner Syndrome

• **Discrete/Focal vs long segment** narrowing (tubular hypoplasia)

• **Describe with its relationship to the PDA:** Pre-ductal, para-ductal or post ductal in relation to the PDA

• The CoA can be very tight with no continuity in flow between the aortic arch and descending thoracic aorta – **Interrupted Aortic Arch**
Echo Evaluation of Aortic Coarctation

Aim of echocardiography in CoA

1. Define the location, extent and severity of the CoA
2. Look for restenosis or residual obstruction post surgical repair or post balloon dilatation or post stenting
3. Gradient across the stenosis / restenosis
4. Measure and document all aortic dimensions
5. Left ventricular size, function, LVH and LV mass
6. Associated cardiac defects ie. bicuspid aortic valve, VSD
7. Aneurysmal dilatation, pre or post CoA
1) Site, extent and severity of the CoA

2D Echo
- Suprasternal view to assess CoA
- High left parasternal views
- Often suboptimal views in adult patients

Doppler Echo
- CW Doppler – Peak gradient across stenosis
  - Diastolic pattern and velocity flow profile
- PW Doppler - Abdominal aortic flow pattern
• This patient’s CoA has a discrete shelf like narrowing at the Ao isthmus

• Post CoA repair with jump graft from ascending to desc thoracic aorta

• Residual gradient of 26 mmHg but no diastolic tail
Is peak systolic gradient alone adequate?

NO

Because besides severity of CoA

Doppler velocities are also affected by

1. Cardiac output
2. Lesion length
3. Collateral networks
4. Aortic compliance
Diastolic tail (DT)

- In significant CoA, systolic pressure gradient results in high velocity flow during ventricular systole.

- This pressure gradient persists in diastole.

- Diastolic pressure gradient across the coarctation site results in forward flow during diastole \(\Rightarrow\) Diastolic Tail.

- The higher the diastolic gradient, the longer the Diastolic tail.
Doppler Echo

Peak systolic gradient = $4V^2$

If $V1 > 1.5\text{m/s}$, use the full Bernoulli’s Equation:

$4 \left(V^2 - V1^2\right)$
Diastolic flow profile as a marker for CoA severity

Doppler flow profile in the same patient before and after stenting for aortic coarctation

- Diastolic tail regresses
- Peak systolic velocity ↓
Diastolic flow profile as a marker of CoA severity

Tan JL et al
JACC 2005;46:1045-53

- Systolic peak gradient
- Diastolic velocity
- Diastolic half-time indices

All the above decrease after stenting
Table 2. Coarctation Index and Echocardiographic Measurements of Group 1 and Group 2 Patients

<table>
<thead>
<tr>
<th></th>
<th>Group 1 Pre-Stenting Mean ± SD (n = 13)</th>
<th>Group 1 Post-Stenting Mean ± SD (n = 13)</th>
<th>Group 2 (Post-Surgical Repair) Mean ± SD (n = 11)</th>
<th>p Value (Group 1 Pre- Versus Post-Stenting)</th>
<th>p Value (Group 1 Post-Stenting Versus Group 2)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Peak systolic gradient, mm Hg</td>
<td>59 ± 13</td>
<td>27 ± 10</td>
<td>13 ± 4</td>
<td>0.001</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>DV, cm/s</td>
<td>256 ± 38</td>
<td>120 ± 38</td>
<td>63 ± 15</td>
<td>0.001</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>DV/SV index</td>
<td>0.71 ± 0.06</td>
<td>0.43 ± 0.09</td>
<td>0.34 ± 0.06</td>
<td>0.001</td>
<td>0.01</td>
</tr>
<tr>
<td>EDTV, cm/s</td>
<td>73 ± 30</td>
<td>28 ± 10</td>
<td>23 ± 12</td>
<td>0.005</td>
<td>NS</td>
</tr>
<tr>
<td>SVHTi, ms</td>
<td>189 ± 59</td>
<td>151 ± 18</td>
<td>147 ± 24</td>
<td>0.75</td>
<td>NS</td>
</tr>
<tr>
<td>SPHTi, ms</td>
<td>127 ± 34</td>
<td>99 ± 38</td>
<td>103 ± 21</td>
<td>0.10</td>
<td>NS</td>
</tr>
<tr>
<td>DVHTi, ms</td>
<td>123 ± 65</td>
<td>45 ± 17</td>
<td>56 ± 38</td>
<td>0.001</td>
<td>NS</td>
</tr>
<tr>
<td>DPHTi, ms</td>
<td>69 ± 31</td>
<td>27 ± 13</td>
<td>32 ± 23</td>
<td>0.001</td>
<td>NS</td>
</tr>
<tr>
<td>DT pattern, n</td>
<td>10 prominent DT, 3 small DT</td>
<td>1 prominent DT, 12 small DT</td>
<td>None had prominent DT, 9 small DT, 2 no DT</td>
<td>0.003</td>
<td>NS</td>
</tr>
<tr>
<td>Abdominal aortic flow pattern, n</td>
<td>10 continuous flow; 3 pulsatile flow</td>
<td>3 continuous flow; 10 pulsatile flow</td>
<td>All pulsatile flow</td>
<td>0.005</td>
<td>NS</td>
</tr>
</tbody>
</table>

DT = diastolic tail; EDTV = end-diastolic tail velocity; NS = not significant; other abbreviations as in Table 1.
Diastolic flow profile as a marker of CoA severity

Tan JL et al
JACC 2005;46:1045-53

- MRI CoAi as gold standard for CoA severity
- **Diastolic velocity of >193 cm/s** (100% sensitivity and 100% specificity in predicting patients with severe CoA (CoAi <0.25)
- Diastolic velocity may be affected by preceding SV. To correct this, DV was indexed to SV, this DV/SV ratio correlated with CoAi
- **DV/SV ratio of 0.53** had a 100% sensitivity and 96% specificity for predicting severe CoA

**CoAi** = Area at the narrowest coarctation site

Area of the descending aorta at the diaphragmatic level

= (Diameter X / Diameter Y)²

Significant aortic coarctation is taken as CoAi of <0.25
Abdominal aortic flow profile in severe CoA

Doppler flow profile in the same patient before and after stenting for aortic coarctation

- Continuous abdominal aortic flow may revert back to pulsatile flow after relieve of CoA
Measure and document all aortic dimensions

Measure the Sinus of Valsalva, ST Junction and proximal Ascending Ao

↑ - Distal Ascending Aorta
↑ - Aortic Arch
Left ventricular size, function, LVH and LV mass

- LVH from CoA
- LVH from AS from BAV
- LVH from hypertension
- Premature CAD
- LV failure from all above
Associated cardiac defects

- Bicuspid AV (50% - 85%)
- VSD
- PDA
- Mitral anomalies
  - Parachute MV
  - Supramitral Ring
- Turner Syndrome
Associated cardiac defects

Repaired CoA patient with BAV and severe AR
Aneurysmal dilatation

- Aneurysm formation usually occurs in 2 regions
  - Ascending aorta (esp. Bicuspid aortic valve, from hypertension)
  - Aortic isthmus at site of surgery or balloon dilatation or stenting

- Type of surgery:
  - Dacron patch aortoplasty (30-50%) repair has higher occurrence than end-end anastomosis type of repair

- Not easy to see on echo, better seen on other imaging modality such as CT or CMR

Patient with aneurysmal dilatation at site of previous patch aortoplasty for CoA
### Indications for intervention in coarctation of the aorta

<table>
<thead>
<tr>
<th>Indications</th>
<th>Class</th>
<th>Level</th>
</tr>
</thead>
<tbody>
<tr>
<td>All patients with a non-invasive pressure difference ≥20 mmHg between upper and lower limbs, regardless of symptoms but with upper limb hypertension (&gt;140/90 mmHg in adults), pathological blood pressure response during exercise, or significant LVH should have intervention</td>
<td>I</td>
<td>C</td>
</tr>
<tr>
<td>Independent of the pressure gradient, hypertensive patients with ≥50% aortic narrowing relative to the aortic diameter at the diaphragm level (on CMR, CT, or invasive angiography) should be considered for intervention</td>
<td>IIa</td>
<td>C</td>
</tr>
<tr>
<td>Independent of the pressure gradient and presence of hypertension, patients with ≥50% aortic narrowing relative to the aortic diameter at the diaphragm level (on CMR, CT, or invasive angiography) may be considered for intervention</td>
<td>IIb</td>
<td>C</td>
</tr>
</tbody>
</table>
Summary of echo imaging in CoA

• Define the anatomy and associated defects in patients with CoA

• Echo is an important initial screening modality but there are important limitations in the following
  ✓ 2D Window (Analysis of CW Doppler more useful)
  ✓ Detection of aortic aneurysm in the isthmus region

• CT and MRI can be used to complement echo in providing information on
  ✓ Aortic size and tortuosity, collateral circulation
  ✓ Aortic aneurysm, post stenting and post surgical assessment
4. Shone Complex
FOUR LESIONS

1) Supramitral Ring
2) Parachute Mitral Valve
3) Subaortic Stenosis
4) Coarctation of the Aorta
Shone Complex/ Shone Syndrome/ Shone Anomaly

- Shone complex is a very rare condition and consists of multi-level of left-sided obstructive lesions. First described by Shone and colleagues in 1963.

- Besides the 4 lesions other associated obstructions include:
  - Other mitral valve abnormalities – congenital MS. etc
  - Bicuspid aortic valve with aortic stenosis
  - Supravalvular aortic obstruction
  - Hypoplastic aortic arch

- Other associated congenital defects include VSD, ASD, PDA, vascular rings and infundibular RVOT obstruction

- Subaortic stenosis in Shone Complex can occur in the form of fibromuscular tunnel or discrete subaortic membrane

Supramitral Ring
- Fibrinous ridge of tissue on the LA side of the MV, inferior to LAA
- 2 variants
  - Supramitral – above valve, not adherent to MV leaflets
  - Intramitral – membrane located within MV tunnel and adherent to leaflet affecting mobility
Parachute Mitral Valve (PMV)

- In PMV, chordae tendinae from both mitral valves leaflets converge on a single, centrally placed papillary muscle instead of 2 papillary muscles.
- The chords in PMV are short, thick and adherent to MV causing stenosis.
- Interchordal space also narrowed resulting in reduced secondary mitral orifice and further obstruction.
Subaortic Stenosis (SAS)

- Majority of cases, due to a discrete fibrous or fibromuscular ridge encircling the LVOT below the AV
- Can be ring-like or crescentic
- Rarely (10%), SAS is diffuse forming a tunnel-like LVOT
- Besides LVOT obstruction, progressive AR may also develop
### Table 10  Indications for intervention in subaortic stenosis

<table>
<thead>
<tr>
<th>Indications</th>
<th>Class</th>
<th>Level</th>
</tr>
</thead>
<tbody>
<tr>
<td>Symptomatic patients (spontaneous or on exercise test) with a mean Doppler gradient ≥50 mmHg or severe AR should undergo surgery</td>
<td>I</td>
<td>C</td>
</tr>
<tr>
<td>Asymptomatic patients should be considered for surgery when:</td>
<td></td>
<td></td>
</tr>
<tr>
<td>• LVEF is &lt;50% (gradient may be &lt;50mmHg due to low flow)</td>
<td>IIa</td>
<td>C</td>
</tr>
<tr>
<td>• AR is severe and LVESD &gt;50mm (or 25 mm/m² BSA) and/or EF &lt;50%^d</td>
<td>IIa</td>
<td>C</td>
</tr>
<tr>
<td>• mean Doppler gradient is ≥50 mmHg^c and LVH marked</td>
<td>IIa</td>
<td>C</td>
</tr>
<tr>
<td>• mean Doppler gradient is ≥50 mmHg^c and blood pressure response is abnormal on exercise testing</td>
<td>IIa</td>
<td>C</td>
</tr>
<tr>
<td>Asymptomatic patients may be considered for surgery when:</td>
<td></td>
<td></td>
</tr>
<tr>
<td>• mean Doppler gradient is ≥50 mmHg^c, LV normal, exercise testing normal, and surgical risk low</td>
<td>IIb</td>
<td>C</td>
</tr>
<tr>
<td>• progression of AR is documented and AR becomes more than mild (to prevent further progression)</td>
<td>IIb</td>
<td>C</td>
</tr>
</tbody>
</table>

ESC Guidelines on Management of GUCH patients 2010
Summary of echo imaging in Shone Complex

- Shone Complex is rare but need to considered as a differential diagnosis in congenital patients with more than 1 level of left sided obstructions.
- Echo is used to assess and define the anatomy and severity of each congenital lesion: supramitral membrane, parachute mitral valve, subaortic obstructions and coarctation of the aorta.
- Echo is also useful to evaluate other associated obstructive and non-obstructive cardiac defects.
THANK YOU