ACYANOTIC CONGENITAL HEART DISEASE
ATRIAL SEPTAL DEFECT (ASD)
ATRIAL SEPTAL DEFECT (ASD)

• Isolated anomaly In 10%

• M:F ratio : 1:2

• 30-50 % of children have ASD as part of cardiac defects.
ASD

- Increasingly referred as murmur and detected in infancy
- Auscultatory Findings helpful in detection
- ECG quite useful
ASD TYPES

• Three types –
  Secundum  (70 %)
  Primum  (20 )
  Sinus Venosus  (10)

• The Patent Foramen Ovale (PFO) does not ordinarily produce intracardiac shunts.
ASD - Types

Primum ASD

Secundum ASD

Sinus Venosus ASD

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ASD

Pre tricuspid  L  \( \rightarrow \)  R  shunt

Asymptomatic

Detected in Late infancy & Childhood

<table>
<thead>
<tr>
<th></th>
<th>Pulse</th>
<th>BP</th>
<th>JVP</th>
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</thead>
<tbody>
<tr>
<td></td>
<td>N</td>
<td>N</td>
<td>A=V</td>
</tr>
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</table>
ASD

Auscultation

Wide & Fixed Split of S₂ - No change with Respiration /

Standing. P₂ can be loud but no PAH

Ejection systolic murmur at Pulmonary Area

Mid Diastolic murmur at LLSB
ASD

• ECG : RAD / rSR : RSR : rR in V1 or V3 R (85%)

• CXR : Variable Cardiac size
  Right atrial enlargement

• Echocardiography : Location of ASD / Size
  Dilatation of RV / RA / PA
  Paradoxical motion of IVS
NATURAL HISTORY OF ASD

Spontaneous closure?

- ASD <4mm by 18months > 90% closure.
- ASD 4-8mm by 18m 75% closure.
- ASD > 8mm rarely close spontaneously.

- If untreated CHF and PAH develops in adults
- SBE prophylaxis is not required unless associated defects present.
ATRIAL SEPTAL DEFECT (ASD), OTHER THAN PRIMUM TYPE

• Mode of diagnosis:
  – Physical examination, ECG, X-ray Chest, transthoracic echocardiography

• Spontaneous closure: Rare if defect >8 mm at birth. Rare after age 2 years. Very rarely an ASD can enlarge on follow up.
PATENT FORAMEN OVALE

Patent foramen ovale:

• Echocardiographic detection of a small defect in fossa ovalis region with a flap with no evidence of right heart volume overload (dilatation of right atrium and right ventricle).
• Patent foramen ovale is a normal finding in newborns.
INDICATION FOR CLOSURE:
ASD ASSOCIATED WITH RIGHT VENTRICULAR VOLUME OVERLOAD

(i) In asymptomatic child: 2-4 yrs. (For sinus venosus defect - 4-5 yrs..)

(ii) Symptomatic ASD in infancy (CCF, severe PAH): seen in about 8%-10% of cases. Rule out associated lesions (e.g., total anomalous pulmonary venous drainage, left ventricular inflow obstruction, aorto-pulmonary window).

Early closure is recommended.
(iii) If presenting beyond ideal age: Elective closure irrespective of age as long as there is right heart volume overload and pulmonary vascular resistance is in operable range.

Method of closure:
Surgical: Established mode.
Device closure: More recent mode, may be used in children weighing >10 kg and having a central ASD.
VENTRICULAR SEPTAL DEFECT (VSD)
VSD – TYPES (location)

Location of the defect:

• Type I: Subarterial
  (outlet, subpulmonic, supracristal or infundibular)
• Type II: Perimembranous (subaortic)
• Type III: Inlet
• Type IV: Muscular
VSD – TYPES (SIZE)

• **Large (nonrestrictive):**
  
  *Diameter of the* defect is approximately equal to diameter of the aortic orifice
  
  Right ventricular systolic pressure is systemic
  
  Degree of left to right shunt depends on pulmonary vascular resistance

• **Moderate (restrictive):**
  
  *Diameter of the* defect is less than that of the aortic orifice
  
  Right ventricular pressure is half to two third systemic
  
  Left to right shunt is >2:1

• **Small (restrictive):**
  
  *Diameter of the defect* is less than one third the size of the aortic orifice
  
  Right ventricular pressure is normal
  
  left to right shunt is <2:1
VSD - HEMODYNAMICS

Ventricular Septal Defect (VSD)

- Pulmonary artery
- Left atrium
- Left ventricle
- Right ventricle
- Ventricular septal defect
- Right atrium
- Mitral Valve
- Aortic Valve
- Opening Between Ventricles
- Tricuspid Valve
- Pulmonary Valve

Legend:
- Red = Oxygen-rich Blood
- Blue = Oxygen-poor Blood
- Purple = Mixed Blood

AO = Aorta
PA = Pulmonary Artery
LA = Left Atrium
RA = Right Atrium
LV = Left Ventricle
RV = Right Ventricle

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CLINICAL PRESENTATION

- With small VSD - asymptomatic.

- With large VSD, delayed growth and development, repeated pulmonary infections and CHF.

- With long standing pulmonary hypertension, a history of cyanosis and a decreased activity.
MODE OF DIAGNOSIS

• Physical examination
• ECG
• X-ray chest
• Echocardiography
VSD AUSCULTATION

S 1 Normal. S 2 Loud ( P 2 )

Wide split with variable P2

Closely split when PAH develops

PSM at LLSB / MSB order

MDM at Apex. [ EDM at Aortic Area ]
## VSD AUSCULTATION

<table>
<thead>
<tr>
<th></th>
<th>Small</th>
<th>Moderate</th>
<th>Large</th>
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<tbody>
<tr>
<td><strong>S2</strong></td>
<td>Normal split</td>
<td>N split P2 + loud</td>
<td>Closely split P2 loud</td>
</tr>
<tr>
<td><strong>S3</strong></td>
<td>Physiologic</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td><strong>Murmur</strong></td>
<td>PSM or E SM</td>
<td>PSM</td>
<td>PSM or ESM</td>
</tr>
<tr>
<td><strong>MDM</strong></td>
<td>0</td>
<td>+</td>
<td>+</td>
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</table>
VSD - MURMUR

- **Moderate VSD**
- **Small, Closing VSD** (harsh)
- **Large, unrestrictive VSD** (less harsh)
NATURAL HISTORY

• About 10% of large nonrestrictive VSDs die in first year, primarily due to congestive heart failure.
• Spontaneous closure is uncommon in large VSDs.
• 30%-40% of moderate or small defects (restrictive) close spontaneously, majority by 3-5 years of age.
• Decrease in size of VSD is seen in 25%.
VSD - TIMING OF CLOSURE

- Large VSD with uncontrolled congestive heart failure: As soon as possible.
- Large VSD with severe pulmonary artery hypertension: 3-6 months.
- Moderate VSD with pulmonary artery systolic pressure 50%-66% of systemic pressure: Between 1-2 years of age, earlier if one episode of life threatening lower respiratory tract infection or FTT.
- Small sized VSD with normal pulmonary artery pressure, left to right shunt >1.5:1: Closure by 2-4 yrs..
- Small outlet VSD (<3mm) without aortic valve prolapse: 1-2 yearly follow up to look for development of aortic valve prolapse.
- Small outlet VSD with aortic valve prolapse without aortic regurgitation: Closure by 2-3 years of age irrespective of the size and magnitude of left to right shunt.
VSD -TIMING OF CLOSURE

• Small outlet VSD with any degree of aortic regurgitation: Surgery whenever aortic regurgitation is detected.
• Small perimembranous VSD with aortic valve prolapse with no or mild aortic regurgitation: 1-2 yearly follow up to look for any increase in aortic regurgitation
• Small perimembranous VSD with aortic cusp prolapse with more than mild aortic regurgitation: Surgery whenever aortic regurgitation is detected.
• Small VSD with more than one episode of infective endocarditis: Early VSD closure recommended.
• Small VSD with one previous episode of infective endocarditis: Early VSD closure recommended
VSD - MODE OF CLOSURE

• **Surgical** closure.

• **Device closure** for muscular VSD in those weighing >15 Kg & For perimembranous VSD.

• **Pulmonary artery banding** is indicated for multiple (Swiss cheese), or very large VSD, almost single ventricle, infants with low weight (<2 Kg), and those with associated co-morbidity like chest infection.
PDA
PDA

Patent Ductus Arteriosus (PDA)

Vessel connecting Aorta and Pulmonary Artery

AO = Aorta
PA = Pulmonary Artery
LA = Left Atrium
RA = Right Atrium
LV = Left Ventricle
RV = Right Ventricle

Left atrium
Right atrium
Left ventricle
Right ventricle
Ductus arteriosus
Aorta
Pulmonary artery

Oxygen-rich Blood
Oxygen-poor Blood
Mixed Blood
MODE OF DIAGNOSIS

- Physical examination
- ECG
- X-ray chest
- Echocardiography.
# PDA

<table>
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<th></th>
<th>Small</th>
<th>Moderate</th>
<th>Large</th>
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<tbody>
<tr>
<td><strong>Pulse</strong></td>
<td>++</td>
<td>+++</td>
<td>+++</td>
</tr>
<tr>
<td><strong>BP</strong></td>
<td>Wide PP</td>
<td>Wide PP</td>
<td>Wide PP</td>
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<tr>
<td><strong>CE</strong></td>
<td>0</td>
<td>+</td>
<td>+</td>
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<tr>
<td><strong>Apex</strong></td>
<td>N</td>
<td>LV</td>
<td>LV</td>
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<td><strong>Continuous m</strong></td>
<td>+</td>
<td>+</td>
<td>+</td>
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<tr>
<td><strong>Thrill</strong></td>
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<td>0</td>
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PDA

Moderate PDA

Large PDA. PAH (H)

Large PDA. Severe PAH
CONTINUOUS MURMUR, PINK CHILD

1. PDA
2. RSOV
3. AP Window
4. CAV Fistula
5. SAV Fistula
6. Aortic RA Fistula
7. ALCAPA
8. Lutembacher
9. PAB stenosis
10. Coarctation
VENOUS HUM

Soft blowing murmur
I - III Medium pitched
High R/L Sternal border or both
No Peaking around S2
↑ on sitting up with neck flexed
↓ on lying down, change in neck position

D.D: PDA, AVM, PAV fistula
Collaterals
SIZE OF PDA

Large PDA: Associated with significant left heart volume overload, CCF, severe PAH. PDA murmur is unlikely to be loud or continuous.

Moderate PDA: Some degree of left heart overload, mild to moderate PAH, no/mild CCF. Murmur is continuous.

Small PDA: Minimal or no left heart overload. No PH / CCF. Murmur may be continuous or only systolic

Silent PDA: No murmur, no PH. Diagnosed only on echo Doppler.

Spontaneous closure: Small PDAs in full term baby may close up to 3 mo of age, large PDAs are unlikely to close.
TIMING OF CLOSURE

• Large/ moderate PDA, with congestive heart failure, pulmonary artery hypertension: Early closure (by 3-6 months).

• Moderate PDA, no congestive heart failure: 6 months-1 year. *If failure to* thrive, closure can be accomplished earlier.

• Small PDA: At 12-18 months.

• Silent PDA: Closure not recommended.
MODE OF CLOSURE

Individualized.
Device closure, coils occlusion or surgical ligation in children >6 months of age.
Surgical ligation if <6 months of age.
Device/ coils in <6 months .
Indomethacin/ ibuprofen not to be used in term babies .
PDA IN A PRETERM BABY

• Intervene if baby in heart failure (small PDAs may close spontaneously).
• Indomethacin or Ibuprofen (if no contraindication).
• Surgical ligation if above drugs fail or are contraindicated.
• Prophylactic indomethacin or ibuprofen therapy: Not recommended.
CONGENITAL FORMS OF LVOT OBSTRUCTION

• **Subvalvular**
  Discrete membranous stenosis, Fibromuscular tunnel

• **Valvular**
  Unicuspid, Bicuspid, Quadricuspid and Dysplastic

• **Supravalvular**
  Discrete (membranous or hourglass)
  Aortic hypoplasia or atresia
  Interrupted aortic arch
  Coarctation of Aorta
DIFFERENT TYPES OF AORTIC VALVES
AV STENOSIS

Obstructive lesion. Usually asymptomatic

SCD / Syncope / Angina possible

Pulse Abnormal
JVP Normal
BP Near Normal

No Cardiomegaly. Heaving apex

Thrill ← Rt USB
Suprasternal
AS AUSCULTATION

S1 N  S2 N  Paradoxic Split?

S4 .  S3  rare ( ominous )

Ejection  Click ( constant )

Ejection Systolic murmur RUSB

EDM ±
AS SEVERITY ASSESS

Pulse  Low volume
Heaving  Apex
S1  E click  Distance
S4
Murmur  Length
         Harshness
         Late Peaking
Suprasternal Thrill
Thrust
NATURAL HISTORY OF AS

• Mild AS and Moderate AS - asymptomatic.

• Severe AS - heart failure in newborns, chest pain, syncope & sudden death.

• Pressure gradient increases with growth.
• Worsening of AR may occur in subaortic stenosis.
• SBE is 4% in valvar AS.
TIMING OF INTERVENTION: VALVULAR AS

For infants and older children:
– Left ventricular dysfunction:
  Immediate intervention by balloon dilatation, irrespective of gradients.
– Normal left ventricular function:
Balloon dilatation if any of these present:
(i) gradient $>80$ mmHg peak and $50$ mmHg mean by echo-Doppler;
(ii) ST-T changes in ECG with peak gradient of $>50$ mmHg;
(iii) symptoms due to AS with peak gradient of $>50$ mmHg. In case of doubt about severity/symptoms, an exercise test may be done for older children.
• For neonates: Balloon dilatation if symptomatic or there is evidence of left ventricular dysfunction / mild left ventricular hypoplasia, or if Doppler gradient (peak) $>75$ mmHg.
COARCTATION OF THE AORTA
COARCTATION OF THE AORTA
COARCTATION OF AORTA (COA)

- 8 % of all CHD.
- M:F = 2:1. 30% of Turner Syndrome.
- 85% of COA have bicuspid valve.
- Poor feeding, dyspnea & poor weight gain, & acute circulatory shock in first 6 weeks.
- 20-30% of COA develop CHF by 3 months
COARCTATION OF AORTA

Stenotic lesion. Asymptomatic in many Infancy to Adulthood

Pulse discrepancy
BP discrepancy

Normal JVP
• Radio femoral delay

  Strong radials; Weak Femorals

  ‘Touch the Feet of Each Infant’

• Upper limb hypertension; Normotensive Lower limb

  SBP of Lower limb 10 mm or more

  Less than SBP of Upper limb
COA AUSCULTATION

S1  S2  N

S3  S4  not  usual

Ej  Click +

Ejection  murmur  /  continuous  murmur

No  murmur

Clinical:  Radio  femoral  delay  ;  Pulse  discrepancy

Never  mind  the  murmur  !
Infant
Cardiomegaly. PVH
Aorta +

Child
No Cardiomegaly. PVH +
Aorta ++ & 3 signs
Rib notching
Diagnostic finding

Aortic lumen is narrowed, typically distal to the left subclavian artery.
Hypoplastic aortic arch
Post stenotic dilatation of the aorta.
Bicuspid aortic valve.
Doppler will show the severity of obstruction.
NATURAL HISTORY OF COA

- Bicuspid valve may cause stenosis or regurgitation with age.
- SBE may occur on either aortic valve or on coarctation.
- LV failure, rupture of aorta, ICH, hypertensive encephalopathy may develop during childhood.
TIMING & MODE OF INTERVENTION

**Timing**
- With left ventricular dysfunction / congestive heart failure or severe upper limb hypertension (for age): Immediate intervention.
- Normal left ventricular function, no congestive heart failure and mild upper limb hypertension: Intervention beyond 3-6 months of age.
- No hypertension, no heart failure, normal ventricular function: Intervention at 1-2 years

**Mode of intervention**
- Balloon dilatation or surgery for children >6 mo of age.
- Surgical repair for infants <6 mo of age.
- Balloon dilatation with stent deployment can be considered in children >10 years of age if required.
- Elective endovascular stenting of aorta is contraindicated for children <10 years of age
THANK YOU