Paediatric Cardiology

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Introduction

• Daunting topic which is very easy to get confused by
• Try and understand the basics of congenital heart defects to logically approach the problem!
• General introduction to murmurs
• Go back over the different types of heart failure
  – Clinical signs
  – Findings
  – Treatments
Learning Objectives

• **Age related changes in heart rate and blood pressure**
  - Know approximate values for infants and toddlers

• **Innocent murmurs**
  - Definition and how to distinguish from pathological murmurs

• **Heart failure**
  - Symptoms and signs in infants and children
  - Possible causes

• **Common forms of congenital heart disease**
  - Natural history and management of:
    - Acyanotic
      - ASD, VSD, PDA, coarctation
    - Cyanotic
      - Fallots, transposition

• **Infective endocarditis**
  - Which children are at risk?
  - Preventative measures
Age Related Changes in HR & BP

• Important to remember that:
  – Paediatric HR is much faster than adult normal
  – BP is lower

<table>
<thead>
<tr>
<th>Age</th>
<th>RR</th>
<th>HR</th>
<th>SBP (50th centile)</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt;1</td>
<td>30-40</td>
<td>110-160</td>
<td>80-90</td>
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<tr>
<td>1-2</td>
<td>25-35</td>
<td>100-150</td>
<td>85-90</td>
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<td>2-5</td>
<td>25-30</td>
<td>95-140</td>
<td>90-95</td>
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<tr>
<td>5-12</td>
<td>20-25</td>
<td>80-120</td>
<td>100-105</td>
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<tr>
<td>&gt;12</td>
<td>15-20</td>
<td>60-100</td>
<td>110-120</td>
</tr>
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</table>

*Source: APLS guidelines*
Murmurs Intro – Location

- Remember that “All Physicians Take Money” to have some logic to approaching where the murmurs will be heard loudest.

- Discuss Erb’s point.
Murmurs Intro – Heart Sounds

• Heart sounds are known as S1 and S2. Additional sounds are S3 and S4.
• S1 >> mitral (M1) and tricuspid (T1) components
• S2 >> aortic (A2) and pulmonary (P2) components

• S2 splitting is physiological during inspiration
• Inspiration causes thorax pressure to fall, increasing venous return to RA
• Opens the pulmonary valve for longer, “splitting” S2 – heard in pulmonic region
• Pathological splitting of S2 is due to increased loading on the R side
Murmurs Intro – Innocent

- Occur in 80% of normal children at some point in childhood
- May be exacerbated by febrile illness
- No associated symptoms
  - No breathlessness, cyanosis, no Hx of fainting
- Normal pulses

- Simple version: *murmurs are innocent*
  - aSymptomatic
  - Soft blowing murmur
  - Systolic murmur only
  - left Sternal edge
Heart Failure

• Heart is no longer adequately perfusing the tissues

• Diverse presentation:
  – Spectrum from severely ill new-born requiring ICU
  – Asymptomatic child with murmur

• Infants:
  – Central cyanosis
  – HF symptoms: breathlessness, sweating, fatigue on feeding, poor weight gain, circulatory collapse with hypotension and acidosis

• Children:
  – Palpitations, dizziness/syncope on exertion, exercise limitation, chest pain (very rarely)
Acyanotic HF: ASD, AVSD

- Atrial septal defect
  - Ostium *secundum* defect
  - More common

- Atrio-ventricular septal defect
  - Ostium *primum* defect
  - Associated with Down's syndrome

- Usually asymptomatic in childhood
- May develop HF symptoms
Acyanotic HF: ASD, AVSD

• Clinical features
  – Palpitations, RV heave

• MURMURS:
  – (1) Diastolic flow murmur across tricuspid valve (increased volume in RA)
  – (2) Ejection systolic murmur at upper left sternal edge (pulmonic region – RV)
  – (3) Fixed and widely split S2 (increased volume in RV)
  – AVSD, (4) apical pansystolic murmur

• ECG:
  – ASD
    • RV volume overload: RSR in V1, right axis deviation
    • RA enlargement: Tall peaked P waves
    • Right BBB
  – AVSD
    • RV hypertrophy / right BBB
    • LV hypertrophy
    • 1st degree heart block
Acyanotic HF: ASD, AVSD

• CXR (both):
  – RA and RV enlargement
  – Prominent pulmonary trunk
  – Increased pulmonary vascular markings

• Mx:
  – ASD:
    • 80% of small defects Dx in infancy close spontaneously by 18 months
    • RV volume-loading necessitates closure with a transcatheter device
  – AVSD:
    • Requires elective surgical repair between 2 and 5 years
Acyanotic HF: VSD

- VSD
  - Flow of blood through ventricular septum at lower L sternal edge
  - When large, cardiac output must increase
Acyanotic HF: VSD

• Clinical features:
  – Small – Asymptomatic
  – Moderate/Large – full HF symptoms

• MURMURS
• Small:
  – Grade 1-4/6 pansystolic murmur at LLSE
  – Palpable thrill

• Moderate/Large:
  – Blowing, pansystolic murmur at lower left sternal edge
  – Apical, mid-diastolic, low-pitched rumble due to increased flow across mitral valve
  – N.B. S2 is NOT split
Acyanotic HF: VSD

- **ECG:**
  - Biventricular hypertrophy
  - Notched/peaked P waves (think atrial hypertrophy)

- **CXR:**
  - Cardiomegaly
  - Pulmonary plethora

- **Mx:**
- Small – significant % close spontaneously in first 2 years
- **Medical:**
  - Control HF, prevention of pulmonary vascular disease, maintenance of normal growth

- **Surgical indications:**
  - Failure of medical treatment, large defect in infant aged 6-12 months with reversible pulmonary hypertension, aortic regurgitation
Acyanotic HF: Patent Ductus Arteriosus

• Increased flow from the aorta to the pulmonary artery

• Clinical features:
  – Small: Asymptomatic
  – Large: Recurrent LRTIs, failure to thrive, eventual HF, endarteritis risk
  – Exertional dyspnoea
  – Bounding peripheral pulses
  – Significant shunt may cause pulmonary hypertension

• MURMURS
  – Continuous machinery murmur below left clavicle
Acyanotic HF: Patent Ductus Arteriosus

- **ECG:**
  - Left ventricular hypertrophy

- **CXR:**
  - Cardiomegaly
  - Pulmonary plethora

- **Mx:**
  - Small: may close spontaneously

- **Medical:**
  - Prostaglandin synthetase inhibitors (ibuprofen, aspirin), HF Mx

- **Surgical:**
  - Ligation / transcatheter occlusion
Acyanotic HF: Coarctation of Aorta

• Different for sick newborn / well child

• Clinical features:
  – Presentation may be abrupt and acute
    • Ductal closure precipitates circulatory collapse
    • Blockage proximal to duct (B), RV can supply blood to lower limbs
    • Duct closure can also worsen the coarctation itself (A or C)
  – Reduced / absent lower extremity pulses
  – BP discrepancy upper and lower limbs

• NO MURMURS
Cyanotic HF: Tetralogy of Fallot

- VSD
- Overriding aorta
- RV outflow tract obstruction
- RV hypertrophy

• Clinical features:
  - Severe cyanosis
    • May lead to MI, CVAs
  - Clubbing
  - Squatting on exercise

• Ejection systolic murmur
  - ULSE, pulmonic region
Cyanotic HF: Tetralogy of Fallot

- ECG
  - Normal at birth
  - RV hypertrophy when older

- CXR
  - Relatively small, “boot-shaped” heart
  - Pulmonary artery “bay”
  - Oligaemic lung fields (reduced vessels)

- Mx
- Surgical:
  - Neonates: Shunt from subclavian to pulmonary artery
  - 6 months of age: close VSD, relieve RV obstruction
- Hypercyanotic spells:
  - Sedation, analgesia, IV beta blocker, fluids, HCO3-
Cyanotic HF: Transposition of Great Arteries

• Coexistent ASD/VSD/PDA allows compatibility with life

• Clinical features:
  – Severe cyanosis
  – Spontaneous PDA closure reduces mixing

• MURMURS
• S2 single and loud
• VSD / PDA murmur
Cyanotic HF: Transposition of Great Arteries

• ECG:
  – Normal

• CXR:
  – “Egg on side”
  – Pulmonary plethora

• Mx:
• Medical:
  – Prostaglandin infusion – keep the PDA open
• Surgical:
  – Balloon atrial septostomy to produce ASD
  – Arterial switch procedure in neonatal period
Conclusion

• Keep calm!
• Work through the steps methodically
• Remember the basics of murmurs before tackling the question
• Remember the difference between acyanotic and cyanotic conditions
Flow List

• Is there a murmur?
  – No? Coarctation of aorta
• Is the murmur continuous?
  – Yes? Patent ductus arteriosus
• How is S2 described?
  – Fixed & split? R heart loading, ASD or AVSD
  – Single S2? Transposition of great arteries
  – No? VSD or Tetralogy of Fallot
• Differentiate VSD/ToF
  – Cyanosis v acyanosis
  – CXR – cardiomegaly v small “boot shaped” heart
  – ECG – A & V hypertrophy v no change / RV hypertrophy
<table>
<thead>
<tr>
<th></th>
<th>Murmur type</th>
<th>Place</th>
<th>Other weird sounds</th>
<th>CXR</th>
<th>ECG</th>
<th>Mx</th>
</tr>
</thead>
<tbody>
<tr>
<td>Still's</td>
<td>Soft, mid-systole</td>
<td>Left sternal border</td>
<td>-</td>
<td>Normal</td>
<td>Normal</td>
<td>None</td>
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<tr>
<td>Venous hum</td>
<td>Soft, throughout cardiac cycle</td>
<td>Beneath clavicles, bilateral</td>
<td>-</td>
<td>Normal</td>
<td>Normal</td>
<td>None</td>
</tr>
<tr>
<td>Neck bruit</td>
<td>Soft, ejection systolic</td>
<td>Above clavicle</td>
<td>-</td>
<td>Normal</td>
<td>Normal</td>
<td>None</td>
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<tr>
<td>ASD</td>
<td>Ejection systolic 2 or 3/6</td>
<td>Upper left sternal edge</td>
<td>Fixed and widely split 2nd heart sound</td>
<td>Cardio-megaly + enlarged pulm arteries + increased pulm vascular markings</td>
<td>Right axis deviation (due to RV hypertrophy) + RSR in V1 (RBBB)</td>
<td>If RV dilation Cardiac catheterisation 3-5 years</td>
</tr>
<tr>
<td>Small VSD</td>
<td>Loud, pansystolic</td>
<td>Lower left sternal edge</td>
<td>Quiet pulm 2nd sound</td>
<td>Normal</td>
<td>Normal</td>
<td>None</td>
</tr>
<tr>
<td>Large VSD (=/+ than aortic valve)</td>
<td>Mid-diastolic murmur</td>
<td>Apical</td>
<td>May be soft pansystolic murmur + loud pulm 2nd sound</td>
<td>Cardio-megaly + enlarged pulm arteries + increased pulm vascular markings + pulm oedema</td>
<td>Bi-ventricular hyper-trophy (by 2 months) + upright T wave in V1 (if RV hypertrophy due to pulm hypertension)</td>
<td>Diuretics (captopril) and calories Surgery at 3-6 months</td>
</tr>
<tr>
<td>AVSD</td>
<td>Pansystolic murmur</td>
<td>Apical</td>
<td>If large: cardiomegaly + increased pulmonary markings</td>
<td>Superior QRS axis (negative AVF)</td>
<td>Surgical 3 years</td>
<td>None</td>
</tr>
<tr>
<td>Condition</td>
<td>Murmur type</td>
<td>Place</td>
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<tr>
<td>PDA</td>
<td>Continuous (machinery) murmur</td>
<td>Beneath left clavicle</td>
<td>Bounding pulse</td>
<td>Normal (or may look like a large VSD)</td>
<td>Normal (or may look like a large VSD)</td>
<td>Cardiac catheter coil or occlusion at 1 year Surgical ligation (Prem: indomethacin / ibuprofen)</td>
</tr>
<tr>
<td>Coarctation</td>
<td>None</td>
<td>None</td>
<td>None</td>
<td>Cardiomegaly</td>
<td>Normal</td>
<td>Maintain duct patency (PG) Surgery ASAP</td>
</tr>
<tr>
<td>Tetralogy</td>
<td>Loud 3 to 6/6 harsh ejection systolic murmur</td>
<td>Left sternal edge</td>
<td></td>
<td>Small heart + up tilted apex (boot-shaped) + pulmonary artery bay (concavity) on left heart border + decreased pulmonary vascular markings</td>
<td>RV hypertrophy (upright T wave in V1) develops with age</td>
<td>Definitive treatment at 6 months (close VSD, relieve RV outflow obstruction) If cyanosed infant, may need shunt between subclavian and pulmonary arteries</td>
</tr>
<tr>
<td>Transposition</td>
<td>Usually no murmur (may be systolic murmur)</td>
<td>Normal</td>
<td>Loud and single 2nd heart sound</td>
<td>Egg on its side cardiac shadow + increased pulmonary vascular markings</td>
<td>Normal</td>
<td>Maintain duct patency (PG) or balloon atrial septoplasty Surgery in first few days of life</td>
</tr>
</tbody>
</table>
MCQs

Q1) A previously well 3-day-old becomes unresponsive and dusky on the neonatal unit. A CXR shows an “egg on side” appearance of the heart. Which of the following 3 are correct?

- A) There may be a machinery murmur below the left clavicle
- B) ECG will show RV hypertrophy
- C) There is a loud, single second heart sound
- D) There is a ejection systolic murmur at the ULSE
- E) Give an immediate prostaglandin infusion
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MCQs

• Q2) A 2-year old child is brought to the GP by his mother after having problems feeding, and not “putting on as much weight as his sister did”. Which of the 3 below are true of a ventricular septal defect?
  – A) Fixed, split second heart sound
  – B) Pansystolic murmur heard at the LLSE
  – C) Tricuspid diastolic murmur
  – D) Physiological splitting of S2
  – E) May require HF medication for feeding problems
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• Is the murmur continuous?
  – Yes? Patent ductus arteriosus
• How is S2 described?
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  – Single S2? Transposition of great arteries
  – No? VSD or Tetralogy of Fallot
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  – Cyanosis v acyanosis
  – CXR – cardiomegaly v small “boot shaped” heart
  – ECG – A & V hypertrophy v no change / RV hypertrophy
MCQs

• Q3) On a routine baby check, you notice that a 48 hour old baby has absent femoral pulses. Which three of these is correct?
  – A) There is a loud, late systolic murmur at the upper left sternal edge
  – B) Blood pressure may be higher in the right arm than the right leg
  – C) Closure of the patent ductus arteriosus may cause circulatory collapse
  – D) The patient may appear to have peripheral cyanosis
  – E) There are no murmurs present