Congenital Heart Disease: Today’s Approach to Diagnosis and Management.

Cardiology for the Non-Cardiologist, Banff, 2018
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Faculty Presenter Disclosure

Cardiology for the Non-Cardiologist
Faculty: Frank Dicke

Relationships with Financial Sponsors:
- Grants or Research Support: None
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- Patents: None
- Other: None
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Potential for conflicts of interest: None
Mitigating Potential Bias

- While we have received unrestricted educational grants from several pharmaceutical companies, most presentations have no mention of specific products and are unrelated to the supporting companies or their products. No specific presentations will be supported or sponsored by a specific company.

- Information on specific products will be presented in the context of an unbiased overview of all products related to treating patients.

- All scientific research related to, reported or used in this CME activity in support or justification of patient care recommendations conforms to the generally accepted standards.

- Clinical medicine is based in evidence that is accepted within the profession.
Latest Clinic News:

Our office requires all scheduled appointments confirmed 48 hours prior to the appointment date.

All appointments not confirmed 48 hours prior to the appointment will be cancelled.

Our Services:

- Pediatric Cardiology
- Consultation
- Echocardiogram
- ECG
- Holter Monitor
- Stress Test
Learning Objectives

• Learning about the diagnosis of pediatric congenital cardiac conditions
• Understanding management of congenital heart disease in the primary care office.
1. Malformations of the heart where:
   a) Blood goes where it shouldn’t
   b) Blood doesn’t go where it should

2. Resulting physiology
   a) Asymptomatic
   b) Cyanosis
   c) Congestive Heart Failure
   d) Shock
   e) Sudden death/Syncope
What is the second most common type of CHD?

A. VSD
B. PDA
C. AVSD
D. ASD
E. Coarctation
What is the second most common type of CHD?

A. VSD
B. PDA
C. AVSD
D. ASD
E. Coarctation
Figure 1

Locations of heart malformations that are usually identified in infancy, and estimated prevalence based upon the CONCOR database (van der Born, 2012 #1149). Numbers indicate the birth prevalence per million live births. Abbreviations: CoA, Coarctation of the Aorta; AS, aortic stenosis; ASD, atrial septal defect; AVSD, atroventricular septal defect; Ebstein, Ebstein anomaly; HLH, hypoplastic left heart; MA, mitral atresia; PDA, patent ductus arteriosus; PS, pulmonary stenosis; PTA, persistent truncus arteriosus; TA, tricuspid atresia; TGA, transposition of the great arteries; TOF, tetralogy of Fallot; VSD, ventricular septal defect; SV, Single Ventricle.
Presentations

• Pink
• Blue
• Grey
• White
• Black

• Asymptomatic
• Cyanosis
• CHF
• Shock
• Dead or dead-like
<table>
<thead>
<tr>
<th>Color</th>
<th>Presentation</th>
<th>Clinical Findings</th>
</tr>
</thead>
</table>
| Pink   | Asymptomatic | - Murmur  
- Arrhythmia                               |
| Blue   | Cyanosis     | - Mixing lesions  
- 5 “T’s”, HLHS                         |
| Grey   | CHF          | - Shunts – VSD/AVSD/PDA  
- Pump failure                               |
| White  | Shock        | - Obstructive lesions  
- AS, Coarct, HLHS                         |
| Black  | Dead/Dead-like | - Syncope  
- ALTE/sudden death                      |
Case 1

- 2 day old baby in the nursery with increasing cyanosis unresponsive to oxygen
- O/E mild tachypnea, no distress, sats 75%, normal S1, single S2, no murmur, normal pulses, good perfusion
- CXR shows very clear lung fields and heart is mildly enlarged
Presentation?

A. Pink ➢ Asymptomatic
B. Blue ➢ Cyanosis
C. Grey ➢ CHF
D. White ➢ Shock
E. Black ➢ Dead or dead-like
### Presentation?

<table>
<thead>
<tr>
<th>Option</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>A. Pink</td>
<td>➢ Asymptomatic</td>
</tr>
<tr>
<td>B. Blue</td>
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</tbody>
</table>
### Case 1 - Cyanosis

<table>
<thead>
<tr>
<th>Non – cardiac</th>
<th>Cardiac</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sepsis</td>
<td>Tetralogy of Fallot</td>
</tr>
<tr>
<td>Respiratory</td>
<td>Transposition</td>
</tr>
<tr>
<td>pneumothorax</td>
<td>Tricuspid (and/or pulmonary) Atresia</td>
</tr>
<tr>
<td>TTN</td>
<td>Truncus Arteriosus</td>
</tr>
<tr>
<td>aspiration</td>
<td>TAPVD</td>
</tr>
<tr>
<td></td>
<td>HLHS</td>
</tr>
</tbody>
</table>
Case 1 - Cyanosis

- A, B, C’s
- Includes oxygen
- Hyperoxic Test
- Prostaglandin Therapy
- Other investigation/referral
• Measures partial pressure of oxygen in arterial blood
• Important to distinguish pre-ductal from post-ductal samples
• Response to inspired oxygen helps differentiate cardiac from non-cardiac causes for cyanosis
Prostaglandin Therapy

• Potent vasodilator that has a particularly strong influence on ductal patency

• Be Prepared!
  • Apnea
  • Hypotension

• May make things worse
  • TAPVD – obstructed
  • TGA/HLHS – restrictive PFO
Office Management

- Pre-op: maintaining acceptable saturation levels, potential acute management of desaturation (Tet spell), nutrition
- Post-op: maintaining acceptable saturation
  - Oxygen
  - Hydration
  - Anti-coagulation
  - Nutrition
Tet Spell
Case 2

- 2 week old baby with 2 day history of poor feeding, lethargy, pallor
- O/E pale, unresponsive, cool peripherally with no palpable pulses or blood pressure attainable by BP cuff
- Tachycardia at 190 bpm, normal heart sounds, soft murmur, sats 89% improve slowly with oxygen
- CXR big heart, increased lung markings
Presentation?

A. Pink ➢ Asymptomatic
B. Blue ➢ Cyanosis
C. Grey ➢ CHF
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E. Black ➢ Dead or dead-like
Presentation?

A. Pink
B. Blue
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D. White
E. Black

➢ Asymptomatic
➢ Cyanosis
➢ CHF
➢ Shock
➢ Dead or dead-like
Case 2 - Shock

Non Cardiac
- Sepsis
- Dehydration
- Metabolic disorder
- Hemorrhage

Cardiac
- Coarctation of Aorta
- Aortic valve stenosis
- HLHS
- Other
  - Cardiomyopathy
  - Myocarditis
Case 2 - Shock

- A, B, C’s
- IV access and volume supplementation
- Inotropic support
- Hyperoxic test/PGE2 therapy may be appropriate
- Other investigation/referral
Office Management

- Stabilize and transfer to tertiary care
- Acute intervention usually required
Case 3

• 28 week gestation baby with increasing respiratory distress, desaturation and apnea
• O/E tachypnea, tachycardia, hepatomegaly, bounding precordium with loud systolic murmur, bounding pulses
• CXR cardiomegaly and increased lung markings
Presentation?

A. Pink ➢ Asymptomatic
B. Blue ➢ Cyanosis
C. Grey ➢ CHF
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E. Black ➢ Dead or dead-like
Presentation?

A. Pink ➢ Asymptomatic
B. Blue ➢ Cyanosis
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Case 3 - CHF

Non Cardiac
- Sepsis
- RDS
- Apnea
- Seizures

Cardiac
- Shunt lesions
  - PDA
  - Large VSD/AVSD
  - A-V fistulae
- Arrhythmia
- Pump failure
Shunt Lesions

• VSD, PDA, AVSD, A-V malformation
• Typically present at 2-3 months of age when the pulmonary vascular resistance starts to drop
• Usually as feeding intolerance, tachypnea or failure to thrive
• Tachypnea, tachycardia and hepatomegaly
• Large heart on CXR
Case 3 - CHF

- A, B, C’s
- IV access with diuresis
- Intubation and ventilatory support may be required
- Inotropes may be needed to support BP
- Other investigation/referral
Office Management

- Pre-op: maintaining growth and weight gain by managing CHF and optimizing nutrition
  - Diuretics/fluid restriction
  - Ng feeding
  - Supplemental calories
  - Aiming for weight gain of 20-40g/day
  - Surgical repair over 5kg, sooner if sub-optimal weight gain
• Previously completely well 12 yr old boy drops dead on the basketball court
• O/E vital signs absent, nothing else remarkable
• EMS rhythm strip - asystole
Case 4 – Dead…or nearly so

Non Cardiac
• See cardiac

Cardiac
• HOCM
• Arrhythmia
  • Long QT
• Aortic dissection
  • Marfans
• Ischemic event
  • Cocaine/ecstasy
  • hyperlipidemia
Causes of Sudden Death in 387 Young Athletes

- **HCM (26%)**
- **Commotio Cordis (20%)**
- **Coronary artery anomalies (14%)**
  - LVH – Indeterminant (7%)
  - Myocarditis (5%)
  - Ruptured Ao Aneurysm (Marfan) (3%)
  - ARVC (3%)
  - Tunneled coronary (3%)
  - AS (2.5%)
  - CAD (2.5%)
  - DCM (2%, 9pts)
  - Myxomatous MV (2%)
  - Asthma (2%)
  - Heat Stroke (1.5%)
  - Drug Abuse (1%)
  - LQTS (1%)
  - Sarcoidosis (1%)
  - Trauma (1%)
  - Other (1.5%)
  - Ruptured cerebral art. (1%)
Who needs to be screened?

A. All relatives?
B. Only relatives in the immediate family?
C. Kids only?
D. All first degree relatives?
E. No relatives?
Who needs to be screened?

A. All relatives?
B. Only relatives in the immediate family?
C. Kids only?
D. All first degree relatives?
E. No relatives?
Office Management

- Almost all causes have genetic implications
- All immediate (first degree) family members need to be screened for suspected inherited disease
- Appropriate treatment and activity restrictions need to be imposed on family members with suspected disease
ADHD and Cardiac Disease

- ADHD common childhood disorder 4-12%
- Prevalence in children with cardiac disease even higher 40+%
- FDA and Health Canada have both issued warnings of significant adverse events in children using psychotropic medications
- Events reported were generally similar in incidence to non-treated population
ADHD and Cardiac Disease

- A committee was struck to see if Adderall posed an additional risk compared to other ADHD meds (not compared to placebo).
- No increased risk was proven *however*, increased risk was not ruled out.
- Health Canada has since lifted the ban.
ADHD and Cardiac Disease

• 30 deaths have been reported in children between 1992 and 2003 with an estimated 2.5 million children on treatment

• Sudden cardiac death “SCD” is estimated at 0.8-6.2/100,000 (therefore expected deaths among 2.5 million children would be 20-150 children)

• HCM, cardiomyopathy, long QT, coronary anomalies and arrhythmias make up majority of causes of SCD
So, what do we do?
Pre-Treatment ECG Required?

A. True
B. False
Pre-Treatment ECG Required?

A. True
B. False
ADHD and Cardiac Disease

• Screening tool – CPS Guidelines
• These would all be reasons To seek further cardiac eval regardless of intent to start ADHD therapy
ADHD and Cardiac Disease

- Ongoing monitoring
  - Baseline ECG currently not routinely recommended
  - Review Hx/Px on current kids on meds if not already done
  - Referral to cardiologist if worrisome clinical findings present or cardiac symptoms develop
  - Patients with identified cardiac disease should be reviewed by a cardiologist and are not necessarily excluded from treatment
ADHD and Cardiac Disease

• Normal or Variant ECG findings
  • Sinus brady/tachy/arrhythmia
  • RV conduction delay without RVH/Right axis
  • Intraventricular conduction delay
  • Early repolarization
  • Non-specific ST-T wave changes
  • QTc≥450ms by computer, normal by hand
  • Borderline QTc of 440-450ms
ADHD and Cardiac Disease

• Worrisome Findings
  • LVH, RVH, left axis, right axis>8yrs, RBBB, LBBB
  • WPW
  • QTc>460ms
  • Atrial, junctional or ventricular tachyarrhythmias including PAC’s, PVC’s
  • 2nd, 3rd degree heart block
  • Abnormal T waves especially v5/v6
References

A joint position statement with the Canadian Paediatric Society, the Canadian Cardiovascular Society, and the Canadian Academy of Child and Adolescent Psychiatry

SA Bélanger MD PhD, AE Warren MD MSc, RM Hamilton MD, C Gray MD, RM Gow MB BS, S Sanatani MD, J-M Côté MD, J Lougheed MD, J LeBlanc MD MSc, S Martin MD, B Miles PhD C Psyc, C Mitchell MD, DA Gorman MD, M Weiss MD PhD, R Schachar MD, Mental Health and Developmental Disabilities Committee

Paediatr Child Health 2009;14(9):579-85
Thank you!

• In addition to congenital heart disease
  • Syncope, palpitations
  • Chest pain
  • Murmurs
  • Arrhythmias
  • Screening for CHD, Marfans, Down, Turner and other syndromes
  • Kawasaki, rheumatic fever and other acquired heart disease
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- Stress Test

Opening Hours:

Monday - 7:30am - 3:00pm
Tuesday - 7:30am - 3:00pm
Wednesday - 7:30am - 3:00pm
Thursday - 7:30am - 3:00pm
Friday - 7:30am - 12:00pm
Saturday - CLOSED
Sunday - CLOSED

Click here for referral documents.