Syncope, chest pain and murmurs in children

The role of Standardised Clinical Assessment & Management Plans (SCAMPS)

Educational and Training Session – GP forum
Cardiology OPD waiting list (not yet scheduled)

<table>
<thead>
<tr>
<th>PAS code</th>
<th>n</th>
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<tbody>
<tr>
<td>murmur</td>
<td>957</td>
<td>52%</td>
</tr>
<tr>
<td>'other'</td>
<td>438</td>
<td>24%</td>
</tr>
<tr>
<td>Family history</td>
<td>138</td>
<td>8%</td>
</tr>
<tr>
<td>Chest pain</td>
<td>125</td>
<td>7%</td>
</tr>
<tr>
<td>Palpitations</td>
<td>80</td>
<td>4%</td>
</tr>
<tr>
<td>Dizziness</td>
<td>69</td>
<td>4%</td>
</tr>
<tr>
<td>Sickle cell disease</td>
<td>24</td>
<td>1%</td>
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</tbody>
</table>

- **murmur**: 957 (52%)
- **‘other’**: 438 (24%)
- **Family history**: 138 (8%)
- **Chest pain**: 125 (7%)
- **Palpitations**: 80 (4%)
- **Dizziness**: 69 (4%)
- **Sickle cell disease**: 24 (1%)
Murmurs in childhood
Referral sources for murmurs from generic referral pool; 100 alphabetical, consecutively filed referral letters A-C (awaiting an apt)
New murmur referrals (Jan – Nov 2014)
By county

murmur referral by age

0-1 year, 653, 48%
1-3 years, 363, 27%
4-12 years, 300, 22%
12 years+, 45, 3%
The length of time patients are waiting to receive an OPD apt for a *murmur*

- 0-3 months: 243, 26%
- 3-6 months: 206, 22%
- 6-12 months: 375, 39%
- 12-24 months: 123, 13%
Prevalence of congenital heart disease

- VSD: 28%
- ASD: 13%
- TofF: 10%
- PDA: 12%
- Valvar PS: 10%
- Coarctation: 9%
- Valvar AS: 6%
- d-TGA: 5%
- TAPVD: 3%
- Tri atresia: 2%
- HLHS: 1%
- PTA: 1%
Prevalence of congenital heart disease
(25-30% detected antenatally; 50% sent home from neonatal wards without CHD dx)

- History: poor feeding, diaphoretic, tachypnea, grey colour, cold
- Murmur (harsh, 3/6, persistent)
- Femoral pulses (92% of aortic coarctation have weak pulses by 5 days)
- 4-limb BP (>20 mm Hg difference in upper and lower limbs significant)
- Saturation check (50% sensitive for ‘major CHD’): >95% normal (foot)
- Split S2 (ASD)
- ECG: axis (0-180 normal), voltages, rsr’ V1
Appropriate use criteria for initial transthoracic echocardiography in out-patient work-up of murmurs
AHA/AAP/ACC/ASE/HRS/SOPE; 2014

• An attempt to respond to the need for the rational use of services in the delivery of high quality care.
• Guidelines are not intended to ignore ambiguity and nuance intrinsic to clinical decision-making.
• Target audience: paediatric cardiology, general paediatrics, family practice.
• Derived from a panel of 14-16 reviewers (not all echocardiographers)
• Guidelines attempt to combine EBM and practice experience
Algorithm for murmur w/up:

Positive history: Poor feeding / fatigue with feeds, diaphoresis, cold, grey colour; syndromes or major other organ malformations (incl inborn errors of metabolism) associated with CHD; Fam history of CHD, CM, SCD or SIDS; Kawasaki’s disease, Rheumatic fever; frequent respiratory infections; failure to thrive; maternal DM; preterm delivery; teratogenic exposure in-utero; chest pain with exertion;

Pathology on exam: tachycardia, tachypnea, sats <95%, poor colour, peripherally cold, active precordium, thrill, heave, displaced apex; hepatomegaly, systolic murmur grade 3/6+, diastolic murmur, rub, click, gallop, weak pulses

Definition of innocent murmur: soft, short duration, varies with positioning (‘sensitive’), systolic, single (no clicks or gallops), sweet (not harsh sounding), small (does not radiate)
Positive history:

• Poor feeding / fatigue with feeds, diaphoresis, ‘cold’, grey colour
• Syndromes or major other organ malformations (incl inborn errors of metabolism) associated with CHD;
• Fam history of CHD, CM, SCD or SIDS;
• Kawasaki’s disease, Rheumatic fever;
• Frequent respiratory infections; failure to thrive;
• maternal DM; preterm delivery; teratogenic exposure in-utero;
• Chest pain with exertion;
Pathology on exam:

• Tachycardia, tachypnea, sats <95%, poor colour, peripherally cold

• active precordium, thrill, heave, hepatomegaly, systolic murmur grade 3/6+, diastolic murmur, rub, click, gallop, weak pulses

• Definition of innocent murmur (seven S’s): soft, short duration, varies with positioning (‘sensitive’), systolic, single (no clicks or gallops), sweet (not harsh sounding), small (does not radiate)
Online resources of auscultation skills

- The Auscultation Assistant
  Web site: http://www.wilkes.med.ucla.edu/inex.htm

- Blaufuss Medical Multimedia Laboratories
  Web site: http://www.blaufuss.org

- Heart Sounds and Murmurs
  Web site: http://www.dundee.ac.uk/medther/Cardiology/hsmur.html

- Johns Hopkins University Cardiac Auscultatory Recording Database
  Web site: http://www.murmurlab.com/card6/ (registration required)

- Texas Heart Institute
  Web site: http://www.texasheart.org/education/cme/explore/events/eventdetail_5469.cfm

- University of Michigan Heart Sound and Murmur Library
  Web site: http://www.med.umich.edu/lrc/psb/heartsounds/index.htm

- University of Washington Department of Medicine
  Demonstrations: Heart Sounds and Murmurs
  Web site: http://depts.washington.edu/physdx/heart/demo.html
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<th>TagID</th>
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<tr>
<td>11620</td>
<td>1052</td>
<td>1203</td>
<td>19y3m</td>
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<td>Normal exam.</td>
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<td>11621</td>
<td>77</td>
<td>97</td>
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<td>F</td>
<td>Split S1 (normal finding, best heard at LL5B).</td>
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<tr>
<td>11622</td>
<td>1149</td>
<td>1116</td>
<td>3y7m</td>
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<td>Innocent murmur (Stoll's, LHSB).</td>
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<td>535</td>
<td>500</td>
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<td>Innocent murmur (pulmonary flow, LUSB).</td>
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<td>11624</td>
<td>485</td>
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<td>11625</td>
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<td>Abnormal diastolic sound ($3$-$4$ gallop).</td>
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<td>10y4m</td>
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<td>Abnormal diastolic sound (opening snap).</td>
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<tr>
<td>11639</td>
<td>780</td>
<td>374</td>
<td>13y11m</td>
<td>F</td>
<td>Friction rub (LMBB).</td>
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</tbody>
</table>

1 thru 20 of 20

Sorted by tagID
ECG interpretation made easy

..or ask our help in cardiology.
The masqueraders

If in doubt, refer to cardiology

My soft echo policy includes:

- Infants (higher yield of pathology)
- Significant other congenital malformations
- Any ECG abnormality
- Flow murmurs over the upper sternal edge (outflow tract murmur of ? ASD... ASD’s being quite common and with subtle exam findings)
Clinical vignette: L.N.

- PC: murmur
- 13 yo, boxer (waiting list x 18 months)
- h/o FTT
- PMedHx: Addison’s disease
- Fam Hx: A fib only (no CHD)
- O/E: 2-3/6 ESM ULSE, faintly radiating posteriorly
Chest pain

SCAMP (Standardized Clinical Assessment and Management Plan): Evidence-based decision-making in clinical practice
vignette

• 18 year old male with 2 week history of exertional chest pain.
• Chest pain, described as a crushing central chest discomfort, coincided with the start of intensive football training
• Pain occurred only with high level of exertion and forced the patient to stop
• Normal physical exam.
Defining the patients with chest pain in OPD

- Median age: 13 years (7-21)
- Male and female (50% each)
- Exertional chest pain present in 37%
- Exertional dyspnea in 31% (we all get some of this!)
- Dizziness or lightheadedness in 14% (not uncommon in healthy albeit unfit people)
Defining the patients with chest pain in OPD

• (+) HISTORY - 1% (n=4) positive family history – 3 SCD and 1 HCM in sibling

• (+) EXAM - 4% (n=16) had abnormal clinical findings: pathological murmur, systolic click, friction rub, gallop, abnormal second heart sound

• (+) ECG - 6% had abnormal EKG: LVH by voltage criteria, abnormal ST segments (pericarditis), WPW, noted atrial or ventricular ectopy, QRS axis deviation
What not to miss as a cardiologist ... the 1.2% of patients with CP in OPD

- Prospective review (406 patients in a year):
  - Pericarditis (2/406)
  - Arrhythmia (3/406): SVT (2), NSVT (1) ... OPD monitors [note: all had ‘palps’]

- Retrospective review (10 years worth of CP in OPD):
  - Coronary anomalies (the majority presenting with exercise-induced CP by history) (32/41)
  - Pericarditis (characteristic CP, ? rub, abnormal ST segments) (4/41)
  - Hypertrophic cardiomyopathy (Fam hx, abnormal ECG) (3/41)
  - Myocarditis (ventricular ectopy) (1/41)
  - Pulmonary hypertension (progressive reduction in mild exertional tolerance) (1/41)

- Rarely, other cardiac etiologies: Aortic dissection, Pulmonary embolus

Battery of tests ordered by cardiologists for chest pain in children:

• EKG (100%)
• Echocardiography (43%) .. Especially with exertion
• Exercise stress tests (28%) .. No proven diagnostic yield in pediatric CP – even in patients KNOWN to subsequently have SCD from coronary anomalies (0/263)\(^{1\&2}\) (I’m guilty of organizing EST for patient / physician re-assurance!)
• 24-hour Holter monitors (7%) ..yield only if CP + palpitations\(^3\)
• King of Heart monitors (10%) ..yield only if CP + palpitations\(^3\)

More experienced physicians order less tests in patients with chest pain!
SCAMP model - bch

- Proposed SCAMP (as applied to patients in the study)
What 98.8% of paediatric chest pain really is

- Musculoskeletal (costochondritis, ‘precordial catch syndrome’, ‘slipping rib syndrome’) (37%)
- Pleuritic / pulmonary (asthma, bronchitis, PTX) (7%)
- Gastrointestinal (GER, esophagitis, gastritis) (2.9%)
- Psychogenic / anxiety (0.9%)
- Drug-related (0.1%)
- Unknown / idiopathic (52%)

“I wear two hats...1. to rule out cardiac pathology and 2. to reassure”

...remember: the chest pain is real for the patient so acknowledging this (along with re-assurance) is important.
What happens to those patients with non-cardiac Chest pain on follow-up?

No cardiac deaths on median follow-up of 4.4 years (17,886 cumulative patient years)!

• 3,700 patients discharged from cardiology OPD with a diagnosis of non-cardiac CP
• 18% re-present to the A&E with further CP
• 7% were re-referred for an additional cardiac opinion

• 2 suicides
Vignette resolution

• EKG and PE normal.
• Echocardiography demonstrated Left MCA arising from right sinus of Valsalva.
• Patient referred for surgical correction.
Patients Presenting with Chest Pain

Obtain patient, family history, and cardiac exam

Abnormal Patient History± or Cardiac Exam†

Yes

Does the patient have Palpitations with Chest Pain* and/or positive family history?

Yes

Extroal Chest Pain

High Suspicion for alternate diagnosis? (i.e., asthma)

Yes

Trial appropriate therapy (i.e., bronchodilator)

Improvement?

Yes

Continue Management

No

Refer to Pediatric Cardiology

No

Refer to Pediatric Cardiology

Is the chest pain primarily with exertion or at rest?

Yes

Extremely low likelihood of cardiac chest pain

Reassurance

No

Chest Pain at Rest and/or reproducible on exam

Refer to Pediatric Cardiology

No

No

Patient febrile or acute onset symptoms

Refer to ED for eval for myocarditis/pericarditis for acute onset symptoms in setting of fever, rub, gallop, abnormal vital signs, or ill-appearance

No

Refer to Pediatric Cardiology

Normal Cardiac Exam and patient history

± chest pain with radiation to jaw, chin, back, left shoulder, increased pain while supine, history of drug use, kawasaki, cardiac surgery, or hypercoaguable state

† Abnormal Cardiac Exam = pathologic murmur, loud or single S2, gallop, rub, peripheral edema, hypoxia, ill appearance, significant tachycardia, tachypnea, or irregular rhythm

# Positive Family History = 1st degree relative or multiple family members with: cardiomyopathy, sudden cardiac death at age < 50, severe familial hyperlipidemia, pulmonary HTN, or known familial arrhythmias
syncope

Transient loss of consciousness and postural tone as a result of decreased cerebral blood flow with spontaneous recovery
SYNCOPE facts:

• 1% of A&E visits

• Risk factors for syncope: female, teenager, being human (one third of dutch medical students!)

• Tiny minority with a cardiac etiology: ‘mechanical obstruction’ (HOCM) or ‘electrical failure’ (non-perfusing rhythm)

• Vast majority are neurocardiogenic / vasovagal

• 10% others: metabolic (hypoglycemia), neurologic (seizures), psychogenic.
Case vignette 1 – OPD recently

• 12 yr old boy referred to OPD with two episodes of syncope – in choir and at white board in school. Stuffy hot room. Standing for 25 minutes. (ate lunch, slept well night before)

• Fam hx: none of relevance

• Prodrome: dizziness, blurred vision (no CP or palpitations)

• Duration of syncope: seconds

• Post event: nauseated, pale, weak afterwards.

• Normal exam, EKG, echocardiogram
Case vignette 2 – clinic last week

• 13 yr old girl presenting with h/o multiple syncopal events (“dozens”)
• Fam hx: Mother A fib (in her 40’s), 2 maternal aunts underwent ablation, maternal GM underwent ablation.
• Associated with palpitations “too fast to count” immediately before syncope. Events occurred “out of blue”. Out for seconds – recovers instantly.
• Two siblings have similar “seizure activity”

• Plan to insert implantable reveal device
Context is everything

- Prolonged standing, shower, rising suddenly from seated position, occasionally after intense exercise whilst standing
- RF: anemia, female, teenager, warm environment, fear or pain
- Symptoms: visual changes, nausea, lightheadedness, dizziness, diaphoresis.

- Sudden unprovoked syncope, exercise-triggered or by loud noise
- Concerning family history (or personal history of CHD)
- Ask about medications: unmasking a LQTS phenotype
Advice / therapies for vasovagal syncope

- Activate venous pumps (alternate feet, fold arms and legs), eat salt (opposite advice of normal), keep well hydrated, cut down caffeine
- Pre-empt the faint: lie down (not sit) with prodrome and STAY DOWN for several minutes.
- Fludrocortisone in more persistent vasovagal syncope
- Other drugs (e.g. beta-blockers) with limited clinical utility
Differential of **cardiac causes** of syncope

1. **Profound bradycardia** / heart block (pacemaker indication)
2. **Non-perfusing rhythm**: VT, torsades-de-pointes (**LQTS**), VF, SVT (rarely); channelopathy as potential etiology of arrhythmia: LQTS, CPVT, Brugada syndrome,
3. **LV outflow tract obstruction** (HCM, severe AS)
4. **Pulmonary hypertension** (advanced)
The role of echocardiography in syncope w/up

• 6 years of data; 480 patients with syncope
• 22 patients identified with cardiac pathology: LQTS (14), arrhythmias (6), cardiomyopathy (2)
• Incidental finding of: 26 minor valve anomalies, 7 hemodynamically insignificant shunt lesions (e.g. PFO’s), 2 patients with mildly decreased LV function.

• All patients with significant (related to syncope) cardiac pathology were picked up by ECG

The role of echocardiography in syncope w/up: official guidelines

<table>
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<th>Indication</th>
<th>Appropriate Use Rating</th>
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<tr>
<td>17. Syncope with or without palpitations and with no recent ECG</td>
<td>R (3)</td>
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<td>18. Syncope with no other symptoms or signs of cardiovascular disease, a benign family history, and a normal ECG</td>
<td>R (2)</td>
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<td>19. Syncope with abnormal ECG</td>
<td>A (7)</td>
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<td>20. Syncope with family history of channelopathy</td>
<td>M (5)</td>
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<td>21. Syncope with family history at a young age (before the age of 50 years) of sudden cardiac arrest or death and/or pacemaker or implantable defibrillator placement</td>
<td>A (9)</td>
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<tr>
<td>22. Syncope with family history of cardiomyopathy</td>
<td>A (9)</td>
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<tr>
<td>23. Probable neurocardiogenic (vasovagal) syncope</td>
<td>R (2)</td>
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<td>24. Unexplained pre-syncope</td>
<td>M (4)</td>
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<td>25. Exertional syncope</td>
<td>A (9)</td>
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<tr>
<td>26. Unexplained post-exertional syncope</td>
<td>A (7)</td>
</tr>
<tr>
<td>27. Syncope or pre-syncope with a known non-cardiovascular cause</td>
<td>R (2)</td>
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The number in parenthesis next to the rating reflects the median score for that indication.

Abbreviations: A = Appropriate; M = May Be Appropriate; R = Rarely Appropriate; ECG = Electrocardiogram.
Implanted reveal devices
SCAMP

- **Abnormal Exam**
  - Pathological murmur, gallop, click, irregular rhythm, loud/single S2

- **Abnormal EKG**
  - Long QT, short QTc, brugada pattern, abnormal ST segments or T-waves, ectopy, heart block, WPW

- **Alarming Event History**
  - Exertional syncope, syncope in supine position, syncopal event precipitated by noise, by physical or emotional stress, or acute collapse with few premonitory symptoms

- **Positive Family History**
  - Known familial arrhythmia, sudden cardiac death, unexplained death or heart failure in 1st or 2nd degree relatives at age <50 years of age, cardiomyopathy in 1st or 2nd degree relatives.

- **Patients Presenting with Syncope**
  - Obtain event history, family history, dynamic physical exam, and ECG

  - Abnormal physical exam*, or ECG†?

    - **Yes**
      - Refer to Cardiology
    - **No**
      - Alarming event history# or positive family history #?

        - **Yes**
          - Recommend reassurance, fluids, salts, and antigravity counseling
        - **No**
          - Have the symptoms improved?

            - **Yes**
              - Continue supportive measures
            - **No**
              - Consider diagnosis of POTS. Consider empiric pharmacological therapy and cardiology referral
In Summary: Chest pain, syncope and murmurs

• Common and benign in the significant majority
• Can rarely be cardinal symptoms of rare and serious cardiac conditions (refer if any clinical concern)
• Practical guidelines available for all three (e.g. SCAMPs) that allow a medically-sound, evidence-based evaluation