

Heart Disease in Children – What the General Practitioner Needs to Know

Professor Orla Franklin
Paediatric Cardiologist

What I am not going to cover.....



What's on the Agenda?

Babies

Structural congenital heart disease

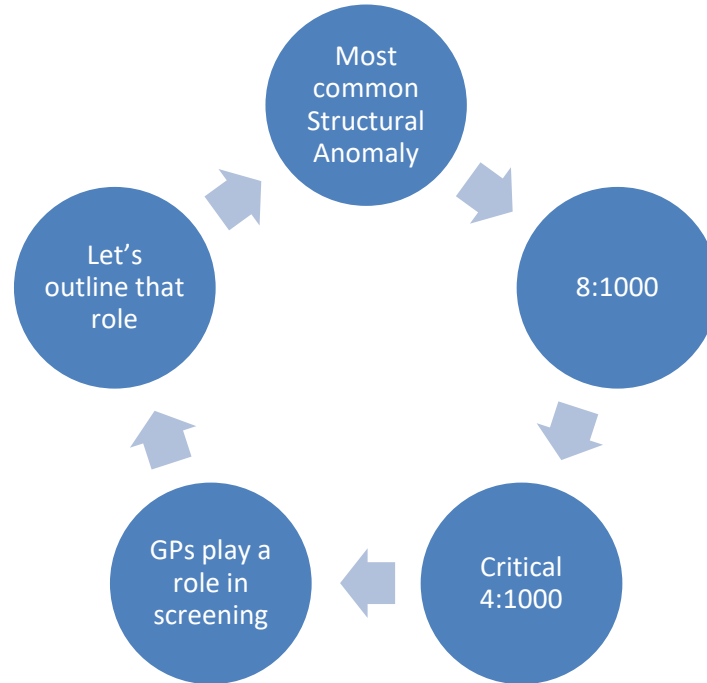
Older Children

Symptoms as a manifestation of
Previously undetected structural cardiac disease
Inherited cardiac disease
Cardiac Infection

Where are you vulnerable?

Where are you less exposed than you might think

Babies with Congenital Heart Disease



Screening for Structural CHD in Ireland

All women are offered a 20-week anatomy scan

All infants are examined by the neonatal team

Oxygen Saturations measured
prior to discharge

GP

Screening for CHD in Ireland



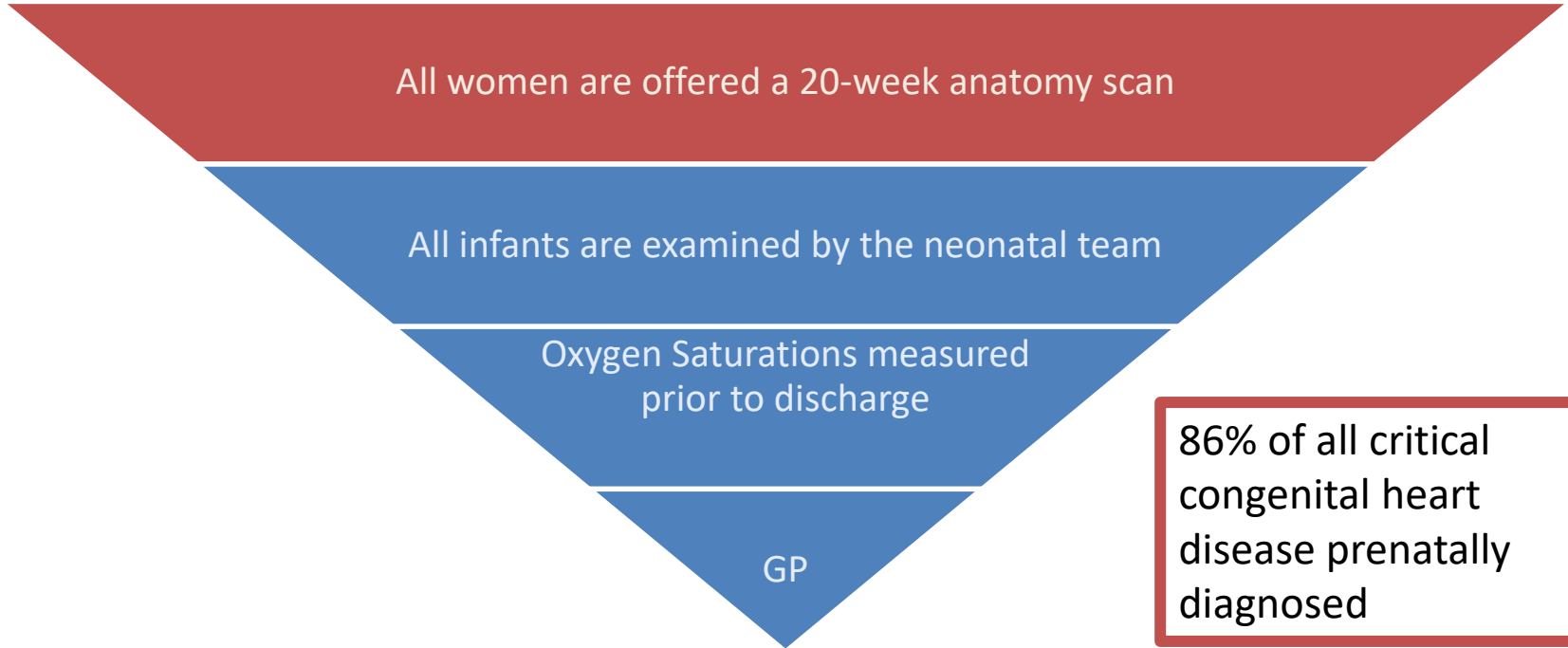
All women are offered a 20-week anatomy scan

All infants are examined by the neonatal team

Oxygen Saturations measured
prior to discharge

GP

Screening for CHD in Ireland



Prenatal Diagnosis

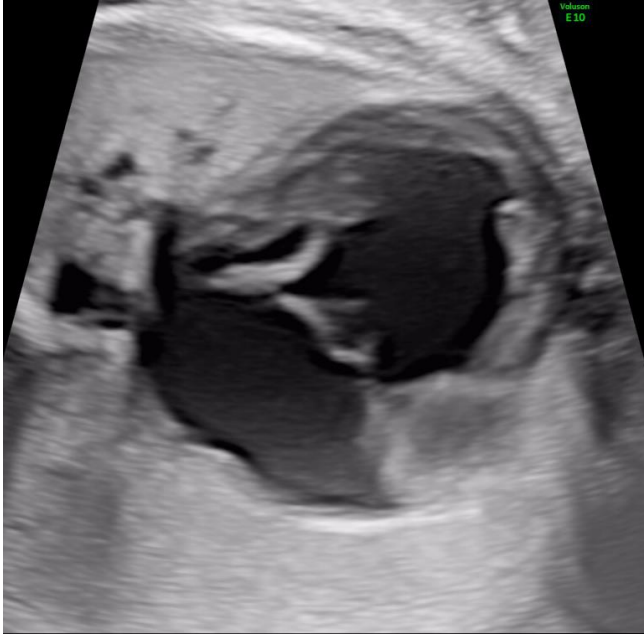


Four Chamber View

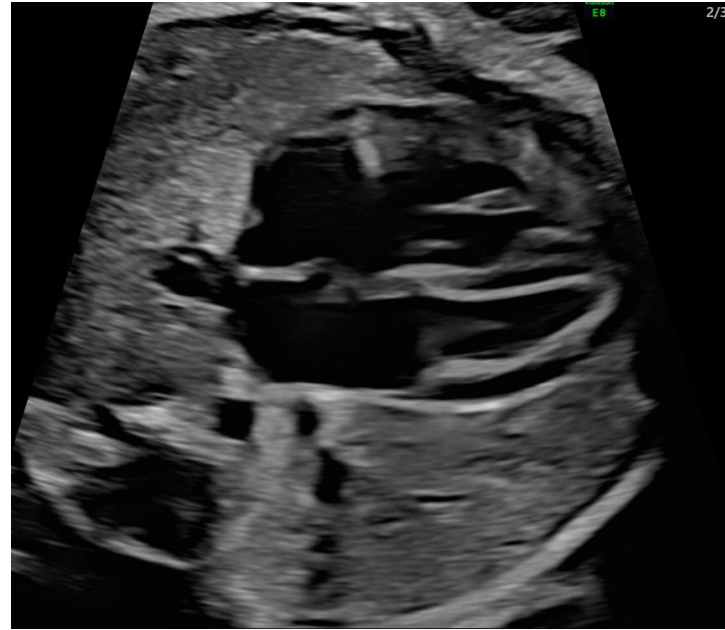


Outflow Tract View

Prenatal Diagnosis



HLHD



TGA

Screening for CHD in Ireland

All women are offered a 20-week anatomy scan

All infants are examined by the neonatal team

Oxygen Saturations measured
prior to discharge

GP

Murmur Harsh
Slow to feed
Short of breath
Cyanosed
Abnormal Pulses

Extracardiac features
Dysmorphic
Chromosome anomaly

Screening for CHD in Ireland

All women are offered a 20-week anatomy scan

All infants are examined by the neonatal team

Oxygen Saturations measured
prior to discharge

GP

Screening for CHD in Ireland

All women are offered a 20-week anatomy scan

All infants are examined by the neonatal team

39,821 babies screened
Physical examination
+pulse Oximetry
82% of babies
100% of babies with
a duct dependent
pulmonary circulation

Oxygen Saturations measured
prior to discharge

GP

5 Cases Missed
All had left heart obstruction
Coarctation is the Achilles
Heel of all modalities of fetal
and infant screening for CHD

Screening for CHD in Ireland

All women are offered a 20-week anatomy scan

All infants are examined by the neonatal team

Oxygen Saturations measured
prior to discharge

GP

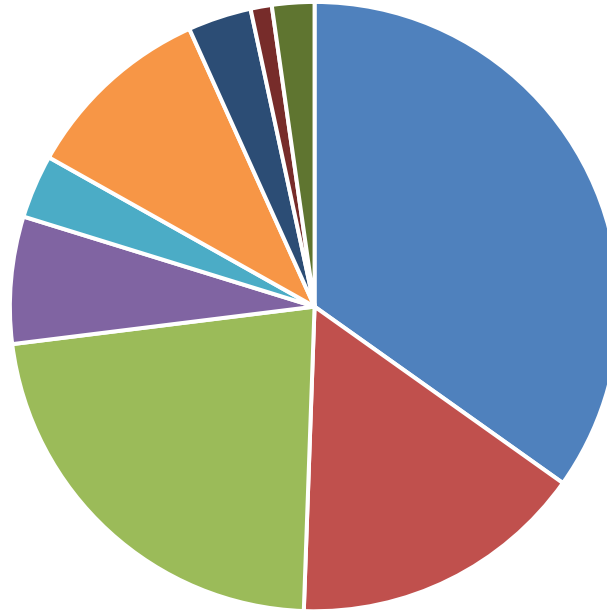
Shunt Lesions

For E.g. VSD

As the pulmonary vascular resistance falls blood will flood the lungs producing a detectable murmur and symptomatic cardiac failure

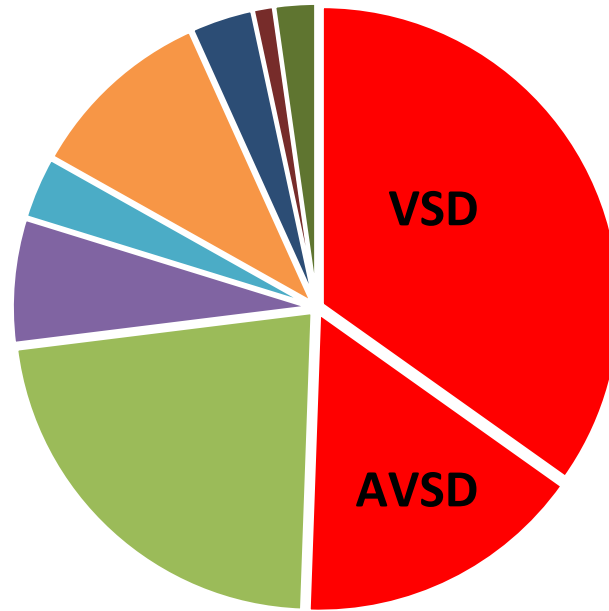
Left heart obstruction account for 75% off all duct dependant lesions detected postnatally are left heart

Postnatal diagnosis of CHD



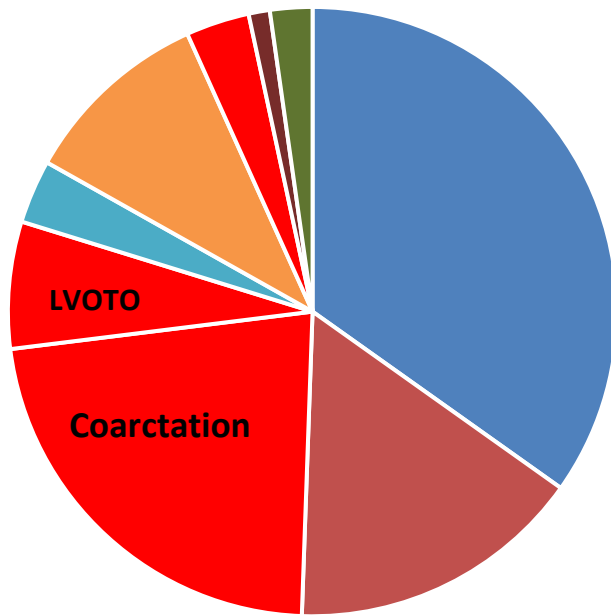
■ VSD ■ AVSD ■ Coarctation ■ LV outlet obstruction ■ RV outlet obstruction ■ Conotruncal ■ TAPVD ■ TGA ■ ASD

Postnatal diagnosis of CHD – Most Common Lesions



■ VSD ■ AVSD ■ Coarctation ■ LV outlet obstruction ■ RV outlet obstruction ■ Conotruncal ■ TAPVD ■ TGA ■ ASD

Postnatal diagnosis of CHD – Presenting Lesions in Shocked Neonates



■ VSD ■ AVSD ■ Coarctation ■ LV outlet obstruction ■ RV outlet obstruction ■ Conotruncal ■ TAPVD ■ TGA ■ ASD

When a GP writes a letter

GP and Paediatrician referral letters triaged as low risk

- 96% are discharged after the first OPD review
- 2% are followed for a minor structural lesion e.g. small VSD, Bicuspid Aortic Valve
- 2% have a significant lesion that required intervention

Structural Congenital Heart Disease in Infants -

Prenatal screening programme for CHD in Ireland

- 86% of all critical CHD

All cases of CHD requiring intervention in the first year of life

- 2/3 are prenatally diagnosed
- 1/3 are postnatally diagnosed
- VSD and AVSD make up more than 50% of all postnatally diagnosed cases
- Coarctation / Left heart obstruction make up 75% of all duct dependent (critical) postnatally diagnosed cases

Of GP and Paediatrician referral letters triaged as low risk

- 96% are discharged after the first OPD review
- 2% are followed for a minor structural lesion e.g. small VSD, Bicuspid Aortic Valve
- 2% have a significant lesion that required intervention

Finding the Needle in the Haystack

Navigating 'Cardiac' Symptoms in Teenagers

Chest Pain

Palpitations

Syncope

Finding the Needle in the Haystack

Navigating 'Cardiac' Symptoms in Teenagers

Chest Pain

Palpitations

Syncope

The problem...

Patients and parents vastly overestimate the prevalence of cardiac causes of chest pain



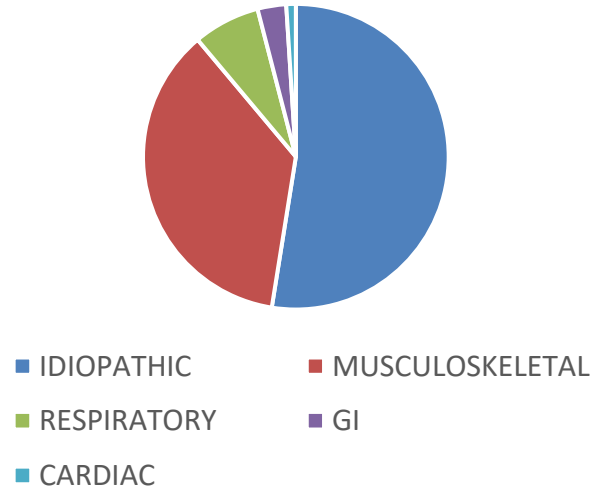
THIS IS MODERN MEDICINE

Chest pain in children is largely benign

Cardiac causes

- Anomalous coronary origins
- Cardiomyopathy
- Pulmonary hypertension
- Myocarditis
- Pericarditis

Causes of Chest Pain in Children



What should you do.....

History

- Family history, Exercise Intolerance, Exertional chest pain, Syncope/exertional syncope, Seizures, Asthma, Kawasaki Disease

Examination

- Murmur, Gallop, Rub, Heave, Thrill
- Body Habitus – Conn Tissue Disorders

ECG

- LVH, RVH, +/- Strain, ST/ T changes, Q waves
- Pre-excitation, non-sinus rhythm.

What should you do.....

History

- **Family history**, Exercise Intolerance, **Exertional chest pain**, **Syncope/exertional syncope**, **Seizures**, Asthma, Kawasaki Disease

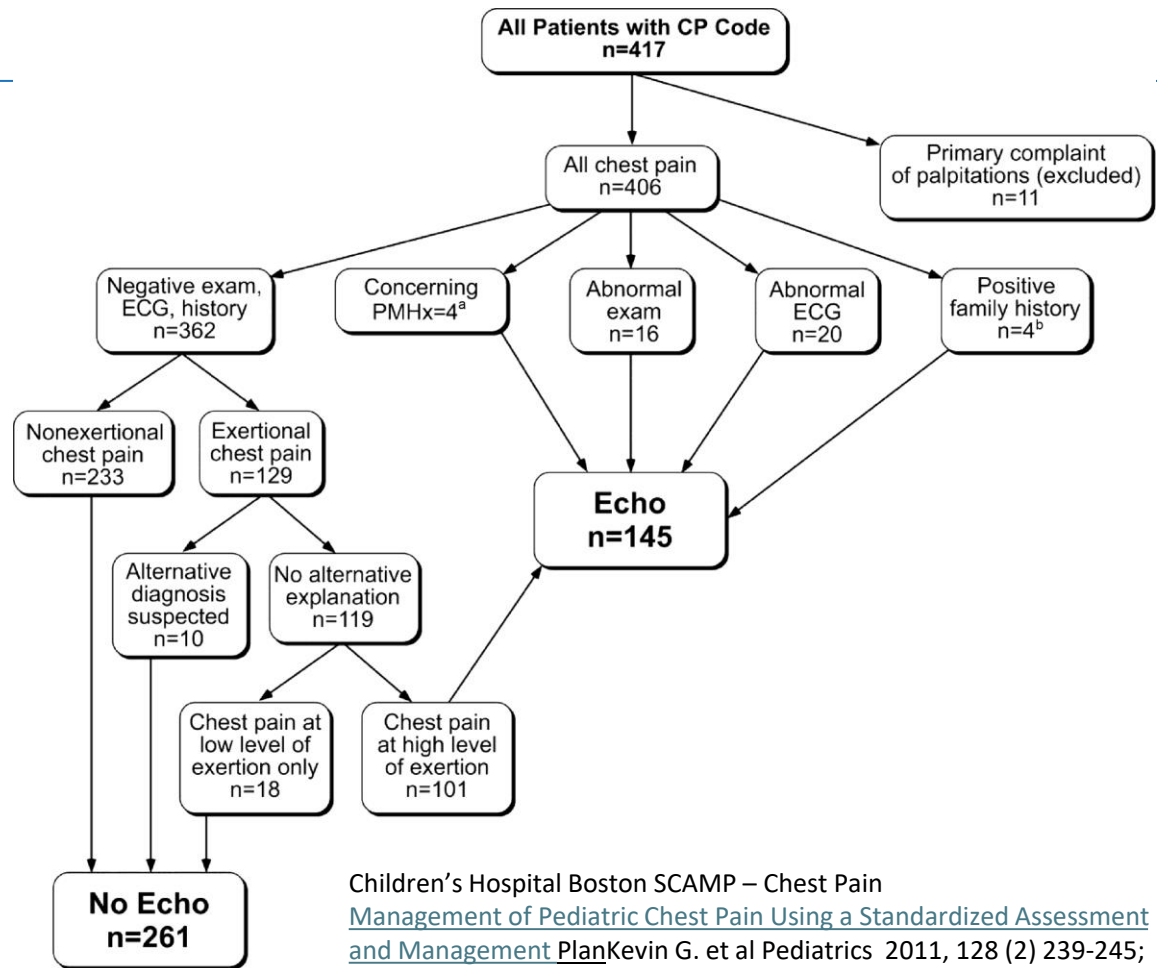
Examination

- Murmur, Gallop, Rub, Heave, Thrill
- Body Habitus – Conn Tissue Disorders

ECG

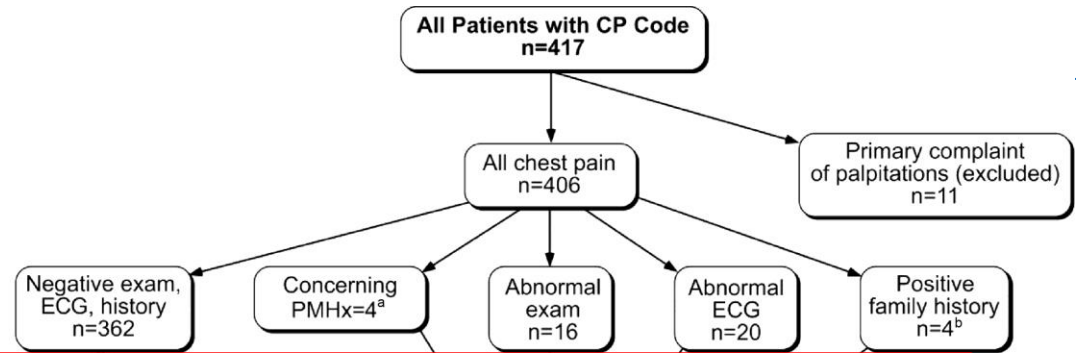
- LVH, RVH, +/- Strain, ST/ T changes, Q waves
- Pre-excitation, non-sinus rhythm.

What do we do.....



Children's Hospital Boston SCAMP – Chest Pain
[Management of Pediatric Chest Pain Using a Standardized Assessment and Management Plan](#) Kevin G. et al Pediatrics 2011, 128 (2) 239-245;

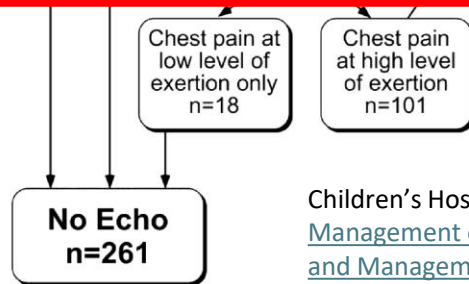
Chest Pain SCAMP



Holter and event monitors are unlikely to be helpful in the evaluation of chest pain in the absence of palpitations or syncope

Exercise Stress Tests do not have a role in the routine evaluation of paediatric chest pain

ECHO is the diagnostic test of first choice



Children's Hospital Boston SCAMP – Chest Pain
[Management of Pediatric Chest Pain Using a Standardized Assessment and Management Plan](#) Kevin G. et al Pediatrics 2011, 128 (2) 239-245;

Sudden Death in Athletes

1.3 cases per 100,000 population per year between 1 – 35 yoa

Risk Factors Male / Ethnicity/ Type of Sport:Basketball

Causes – Cardiomyopathy, Structural HD, Ion Channelopathy

40% of all SCD remains unexplained (in particular in the Young)

Sudden Death in Athletes

1.3 cases per 100,000

Risk Factors Male / Et

Causes – C

40% of all SCD remain

Heritability

Inherited

Structurally abnormal heart

- Cardiomyopathies
 - Hypertrophic cardiomyopathy
 - Arrhythmogenic cardiomyopathy
 - Dilated cardiomyopathy
- Coronary artery anomalies
- Valvular heart disease
- Aortic disease

Acquired

- Ischaemic heart disease
- Myocarditis

Structurally normal heart

- Channelopathies
 - Brugada syndrome
 - Long QT syndrome
 - Catechol-aminergic polymorphic ventricular tachycardia
- Commotio cordis
- Substance misuse
- Electrolyte imbalance

Sudden Death in Athletes

Common findings

- Sinus bradycardia
- First-degree atrioventricular block
- Incomplete right bundle branch block
- Early repolarisation
- Isolated QRS voltage for left ventricular hypertrophy

Pathological findings

- T-wave inversion
- ST-segment depression
- Pathological Q waves
- Left atrial enlargement
- Left-axis deviation/left anterior hemiblock
- Right-axis deviation/left posterior hemiblock
- Right ventricular hypertrophy
- Ventricular pre-excitation

ecg@childrenshealthireland.ie

- Brugada-like early repolarisation

Sudden Cardiac Death -Screening of Teenagers

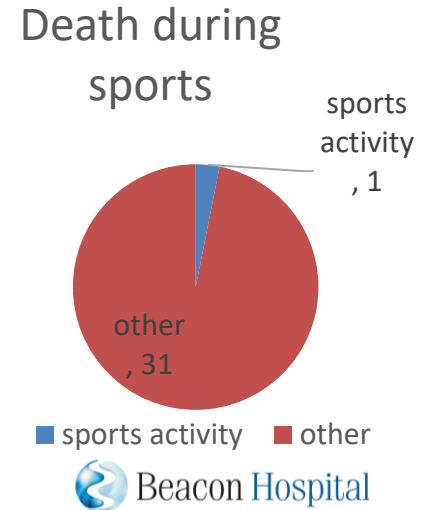
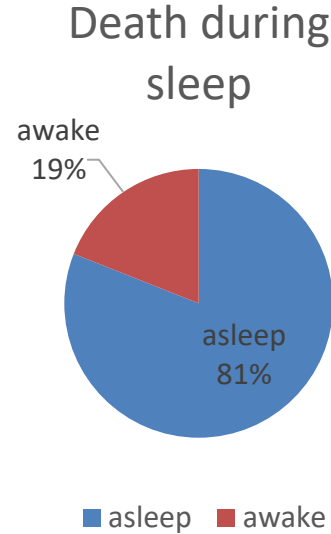
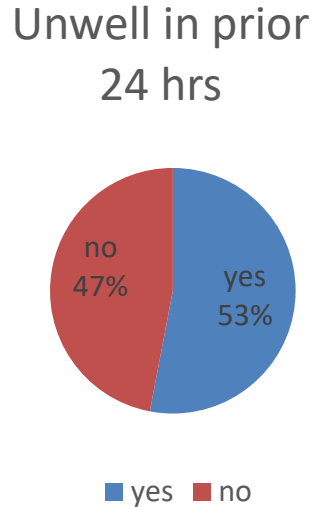
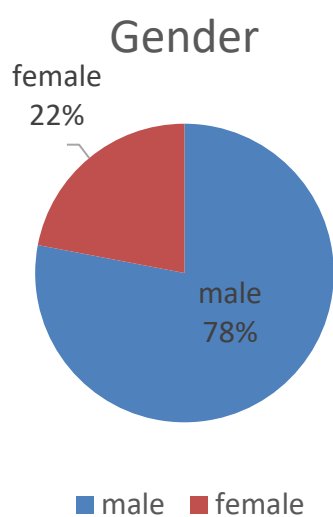
Controversy as to what constitutes screening

- American Heart Association & American College of Cardiology -History and Examination
- European Society of Cardiology -History Examination and ECG

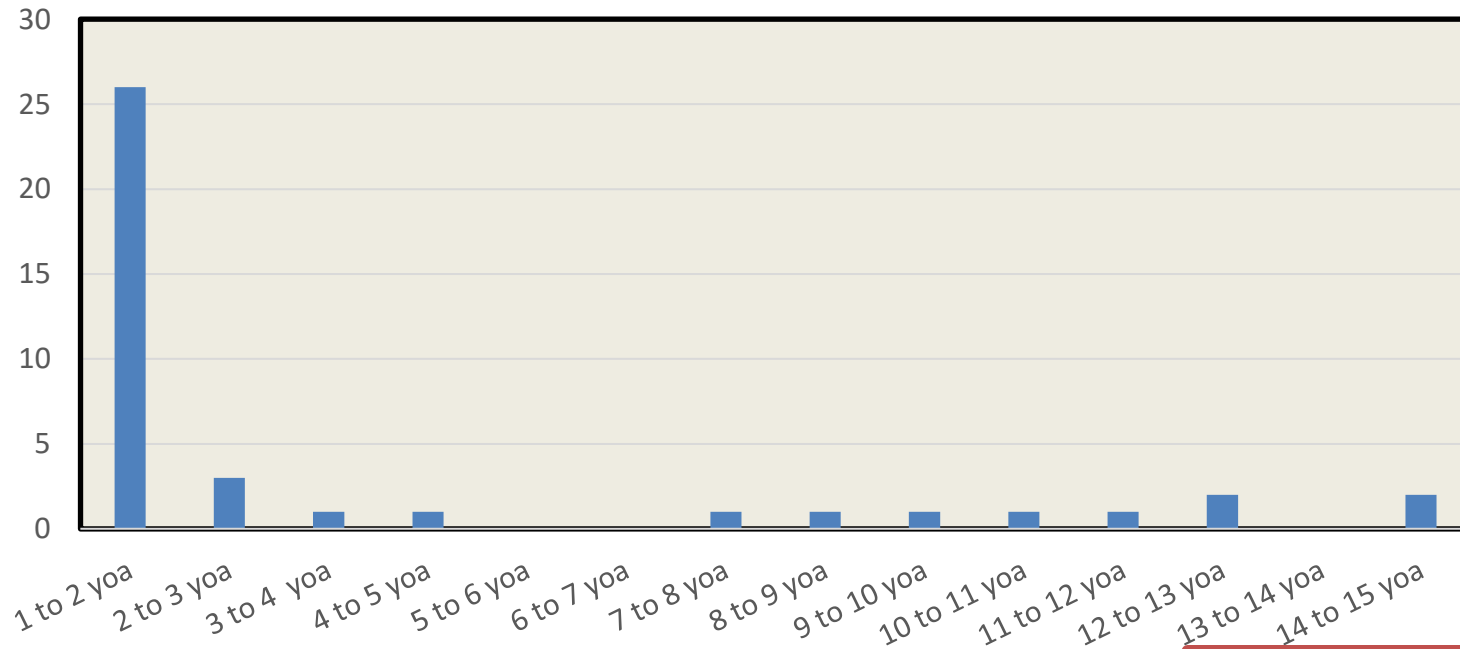
Mass cardiac screening in athletes remains controversial

- Mandatory preparticipation cardiac screening with ECG and exercise testing in Israel in 1997 did not reduce SCD events in competitive athletes. Steinvil et al

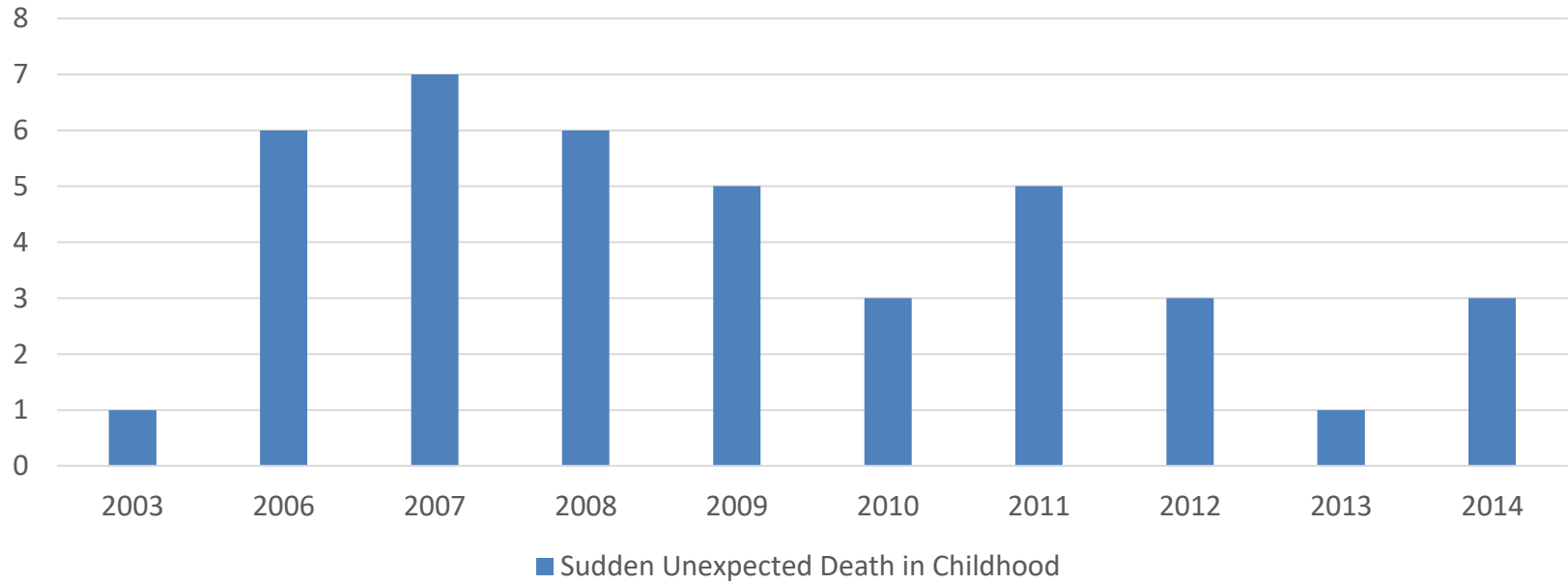
Detail -32 Irish Paediatric Autopsy Cases of Sudden Unexpected Death in Childhood



Age at time of sudden unexpected death in Childhood in Ireland (Ages 1-15 N=40)



Acknowledge Dr Ronan Margey,
Dr Terry Prendiville



Acknowledge Dr Ronan Margey
Dr Terry Prendiville

Sudden Unexpected Death in Childhood
(1-15 years of age) in Ireland [n=40]

CPVT -Sudden victory – sudden death

Electronic Gaming can be a precipitant

Proarrhythmic diagnosis -31% premorbid

Proarrhythmic diagnosis -54% postmortem

CPVT, Long QT, Congenital cardiac surgery, KD

What we do know for sure.....

Cardiopulmonary Resuscitation:

- Immediate availability of quality cardiopulmonary resuscitation (CPR) performed by bystanders and automatic external defibrillators (AEDs) are crucial elements of SCD prevention.
- It is the greatest determinant of survival after a cardiac arrest is the time from collapse to defibrillation, with survival rates declining from 7% to 10% per minute for every minute lost.'
- Delivery of appropriate CPR teaching programs in the general population may have a significant impact on outcomes

Availability of AEDs save lives

Headlines

To diagnose CHD at the postnatal check listen for a murmur

To save lives at the postnatal check feel the femoral pulses

1-2% of children with chest pain will have underlying cardiac pathology

Screening encourages safe continued participation

All sports facilities should have ready access to AED

Everybody should have basic CPR training

Thank You