



Pediatric Cardiac Conditions in Sport

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Conference

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Disclosures

- No financial interests or relationships to disclose
- I will not discuss off label medication use

Learning objectives

1. Compare global approaches to pre-participation screening.
2. Recognize warning signs of cardiac disease in pediatric athletes.
3. Describe key cardiac causes of sudden death in young athletes.
4. Understand difference between pediatric and adult athlete with sudden cardiac death

Case 1

- 16-year-old junior national level competitive swimmer
- No cardiac symptoms.
- No family history of sudden death or cardiomyopathy.
- However no formal assessment/screening

Case 1

- Obtains athletic scholarship to swim at UConn where he undergoes screening
- Why the difference in approach?



Sudden cardiac death in child athletes

- Rare - 1.9 per 100,000 athlete-years
- Leading cause of death in sports participation
- Often *first manifestation* of heart disease
- Determinants of risk
 - Higher risk in males
 - Black athletes
 - Participating in high-intensity sports (basketball/football)



Preparticipation screening

- AHA/ACC and ESC recommend preparticipation cardiac screening to identify high-risk athletes
 - Goal = Detect silent cardiovascular disease
 - No consensus on *how* cardiac screening should be performed
- AHA suggests 14-point history and physical examination (class 1)
 - Low sensitivity
 - Most athletes with serious cardiac diseases are asymptomatic with no abnormal physical signs
- ECG?
 - Channelopathies (long QTS and WPW) and most cardiomyopathies have abnormal ECGs!

Tiered approach to SCD screening

1. History and physical examination (low sensitivity 10-20%)
2. ECG
 - Increases sensitivity for channelopathies, accessory pathways and cardiomyopathy
 - Not recommended for all in US/Can due to cost, false positives and need for expert interpretation
 - Reserved for abnormal H&P and higher risk patients
3. Echocardiography - if H&P and/or ECG suggest possible structural cause
4. Other tests
 - Exercise testing
 - Ambulatory monitoring
 - Genetic testing

TABLE 4 Adapted From the AHA's Recommended 14-point Screen for Cardiovascular Disease

| AHA's 14-Point PPE |
|--|
| Personal history |
| 1. Chest pain, discomfort, tightness, or pressure related to exertion |
| 2. Unexplained syncope or near-syncope not felt to be vasovagal or neurocardiogenic in origin |
| 3. Excessive and unexplained dyspnea or fatigue or palpitations associated with exercise |
| 4. Previous recognition of a heart murmur |
| 5. Elevated systemic blood pressure |
| 6. Previous restriction from participation in sports |
| 7. Previous testing for the heart, ordered by a physician |
| 8. Family history of premature death (sudden and unexpected or otherwise) before 50 y of age attributable to heart disease in ≥ 1 relative |
| 9. Disability from heart disease in close relative <50 y of age |
| 10. Hypertrophic or dilated cardiomyopathy, LQTS, or other ion channelopathies, Marfan syndrome, or clinically significant arrhythmias; specific knowledge of genetic cardiac conditions in family members |
| Physical Examination |
| 11. Heart murmur, not felt to be innocent |
| 12. Femoral pulses to exclude aortic coarctation |
| 13. Physical stigmata of Marfan syndrome |
| 14. Brachial artery blood pressure (sitting position), preferably taken in both arms |

Global preparticipation screening differences

| Region/Society | Screening Modality | ECG Universal? | Key Points/Limitations |
|---------------------|-----------------------------------|----------------|--|
| US (AHA/ACC/AAP) | History & Physical (14-point/AAP) | No | ECG for select cases; focus on cost/resource; PPE every 2–3 yrs |
| Canada | History & Physical | No | Similar to US; ECG for select cases |
| Europe (ESC) | History, Physical, ECG | Yes (<35 yrs) | ECG improves detection; requires expertise; more false positives |
| International/Other | Varies | Varies | Emergency action plans universally recommended |

Brief review of evidence

Table 5. Statistical Performance of the AHA 14-Point Evaluation and ECG ([Table view](#))

| | AHA 14-Point (95% CI) | ECG (95% CI) |
|---------------------------|-----------------------|-------------------|
| Sensitivity | 18.8% (4.1–45.7) | 87.5% (61.7–98.5) |
| Specificity | 75.1% (73.7–76.5) | 97.5% (97.0–98.0) |
| Positive predictive value | 0.3% (0.1–0.9) | 13.6% (10.7–17.2) |
| Negative predictive value | 99.5% (99.4–99.6) | 99.9% (96.9–98.0) |
| Accuracy | 74.9% (73.4–76.3) | 97.5% (96.9–97.9) |

AHA indicates American Heart Association.

- AHA scientific statement (2019) suggests reevaluation of 14-point evaluation
- High number of false positive results with poor sensitivity and very low PPV
- ECG outperforms AHA 14-point questionnaire by all measures of statistical performance when interpreted by experienced clinicians

No screening is perfect!

Implement emergency action plans, AED access and CPR training

Case 1 – Healthy athlete

- Now 17-year-old NCAA collegiate athlete SHOULD undergo SCD screening
- Negative 14-point history and physical screen
- No ECG implemented

Case 2

- 16-year-old basketball player collapses during practice
- No prodrome, quick recovery
- No resuscitation needed



Etiologies of sudden cardiac death in youth

1. Structural abnormalities

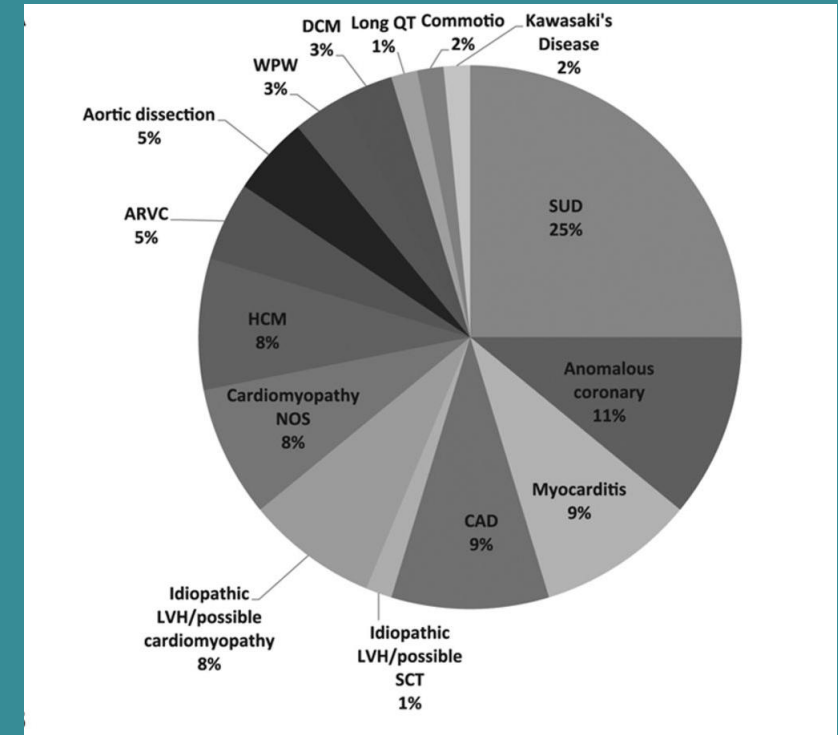
- Hypertrophic cardiomyopathy
- Arrhythmogenic cardiomyopathy
- Dilated cardiomyopathy
- Myocarditis
- Idiopathic left ventricular hypertrophy
- Coronary artery anomalies

2. Electrical cardiac abnormalities

- Long QT syndrome
- Brugada syndrome
- Catecholaminergic polymorphic ventricular tachycardia
- Wolff-Parkinson-White syndrome

3. Other

- Commotio cordis
- Valvular heart disease – Mitral valve prolapse, bicuspid aortic valve
- Aortic dissection

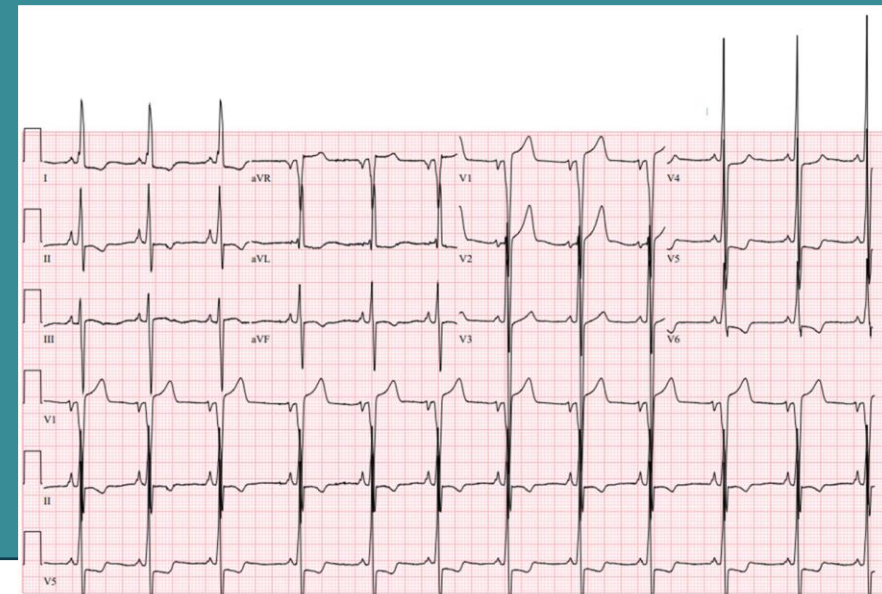


Structural causes of SCD in youth

1. Hypertrophic cardiomyopathy (HCM)
2. Arrhythmogenic cardiomyopathy
3. Dilated cardiomyopathy
4. Myocarditis
5. Idiopathic left ventricular hypertrophy
6. Coronary artery anomalies

Hypertrophic cardiomyopathy

- Leading inherited cause of SCD in adolescents (8+%)
- Prevalence 0.2% in general population
- Features:
 - Unexplained left ventricular hypertrophy in the absence of abnormal loading conditions
 - Typically asymmetric septal hypertrophy
 - Causes LV outflow tract obstruction, myocardial ischemia → ↑propensity to ventricular arrhythmias
- Exam – dynamic systolic murmur ↑ Valsalva
- ECG - abnormal >90%
 - LVH, ST-T wave changes, lateral T wave inversion, deep q waves, LAE

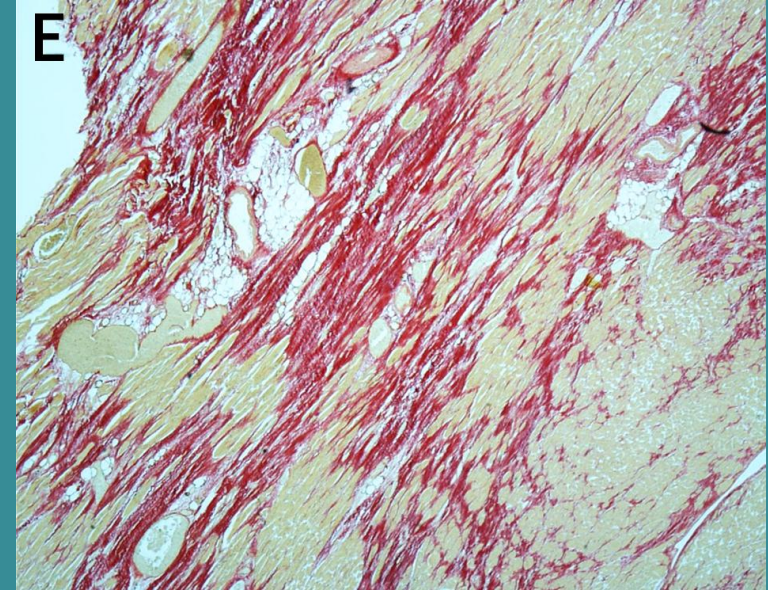


Athlete's heart vs HCM

- Benign, physiological cardiac remodeling from intensive training
 - Participation in vigorous exercise → modest increase in LVH + enlarged cavity size + normal ventricular function
 - No increased risk of SCD
- Epidemiology
 - Moderate to large BSA
 - Endurance athletes (rowing, cycling etc)
 - Black athletes
- Important to differentiate from cardiomyopathy
- Refer to Cardiologist! Possible need for deconditioning.

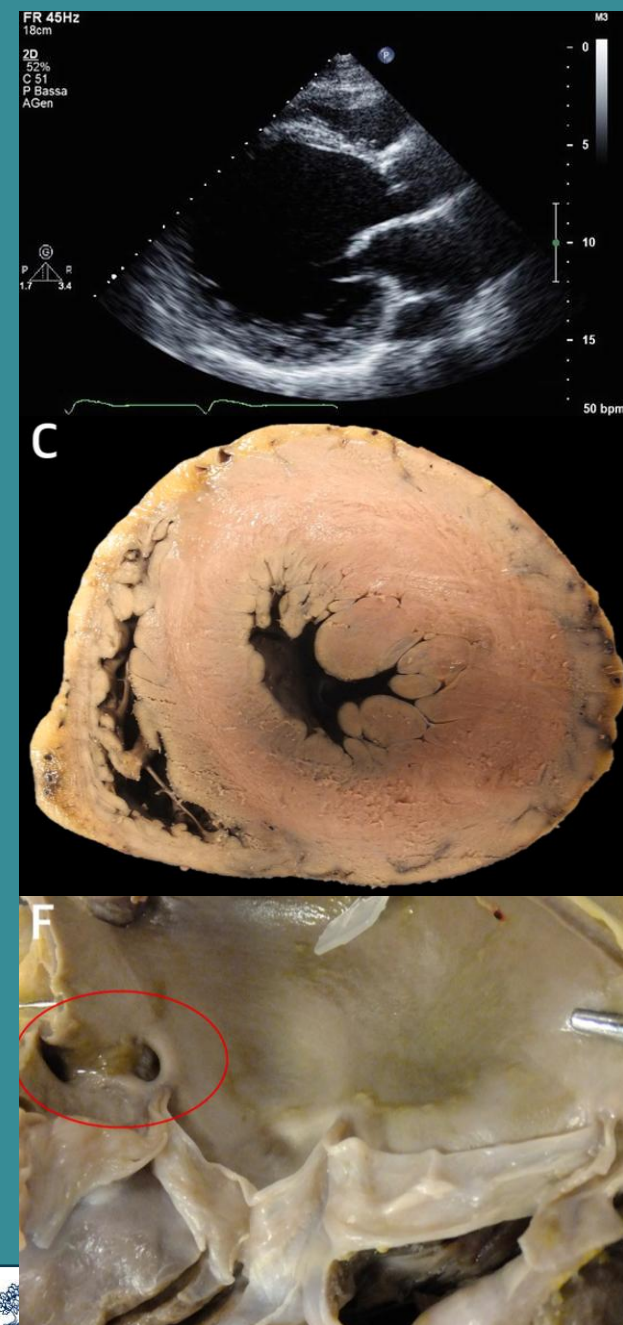
Arrhythmogenic cardiomyopathy (ARVC)

- Progressive fibrofatty replacement of myocardium
- ↑↑ risk of fatal arrhythmias
- 1:1000 in general population
- Major cause of SCD in athletes (5%)
- Predominant etiology in athletes who die during exercise → avoid vigorous exercise



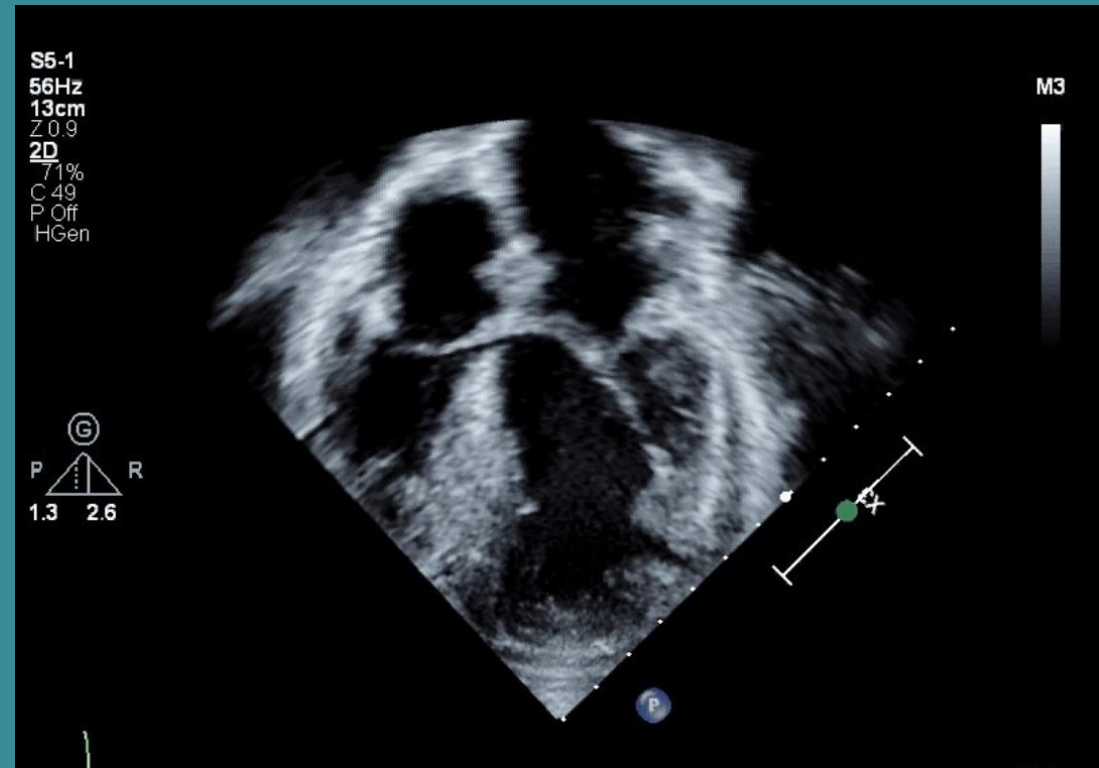
Other structural causes of SCD

1. Dilated cardiomyopathy – rare 1-3%
 - LV dilation + systolic dysfunction
 - Risk of serious ventricular arrhythmias
2. Myocarditis – 9%
 - Inflammatory disease of myocardium, often viral related
 - Risk of fatal arrhythmias
3. Idiopathic left ventricular hypertrophy – 8%
 - Novel entity increasingly reported up to 30% of SCD
 - Not related to athletic training, possibly early HCM
4. Anomalous coronaries - 11%
 - Origins of coronaries (often left) from wrong sinus often with intraarterial/intramural course



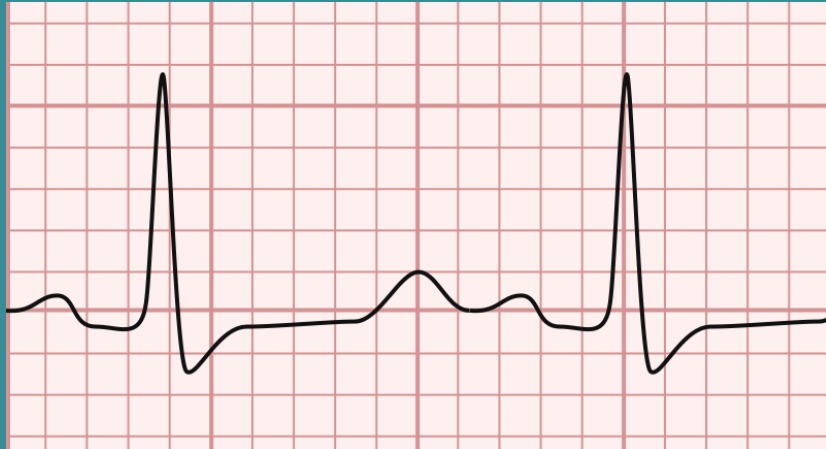
Case 2 – Hypertrophic Cardiomyopathy

- History
 - No previous symptoms or events
 - No family history of sudden death or cardiac disease
 - No previous testing
- Physical examination - Systolic murmur ↑ with Valsalva
- ECG – LVH with deep Q waves
- Echo
 - Asymmetric septal hypertrophy (1.8cm)
 - LVOT gradient 40 mmHg



Case 3

- 14-year-old rower collapses mid race
- ECG – QTc 520ms

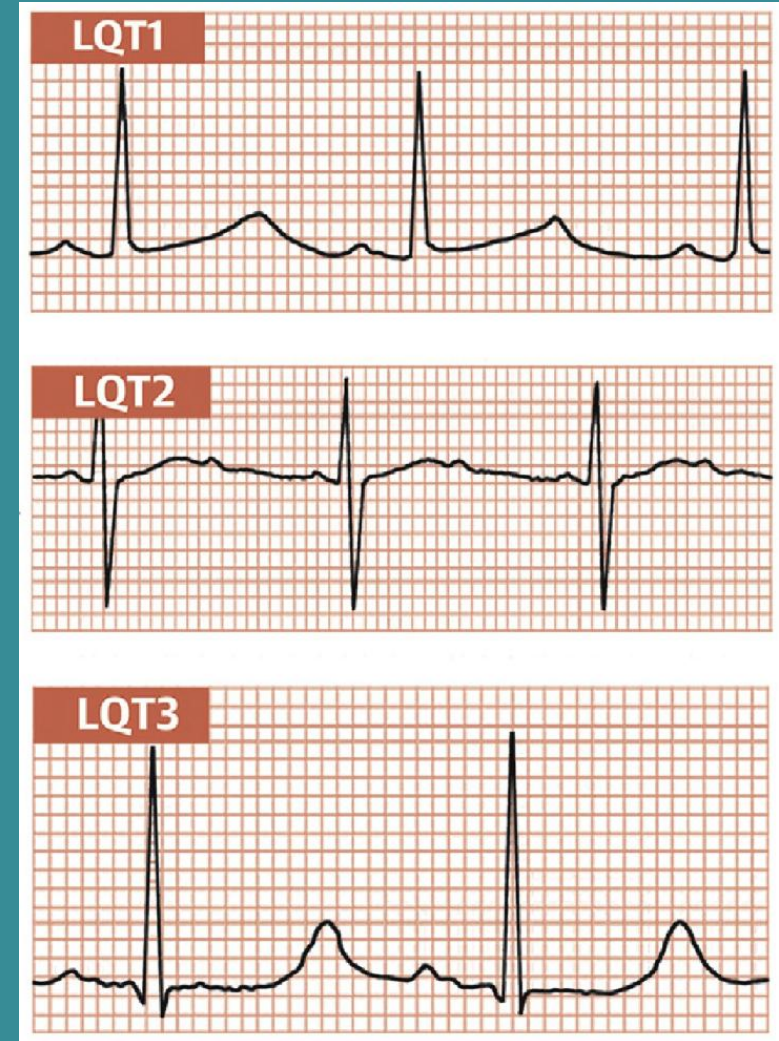


Electrical cardiac causes of sudden death

- Long QT syndrome
- Brugada syndrome
- CPVT
- WPW

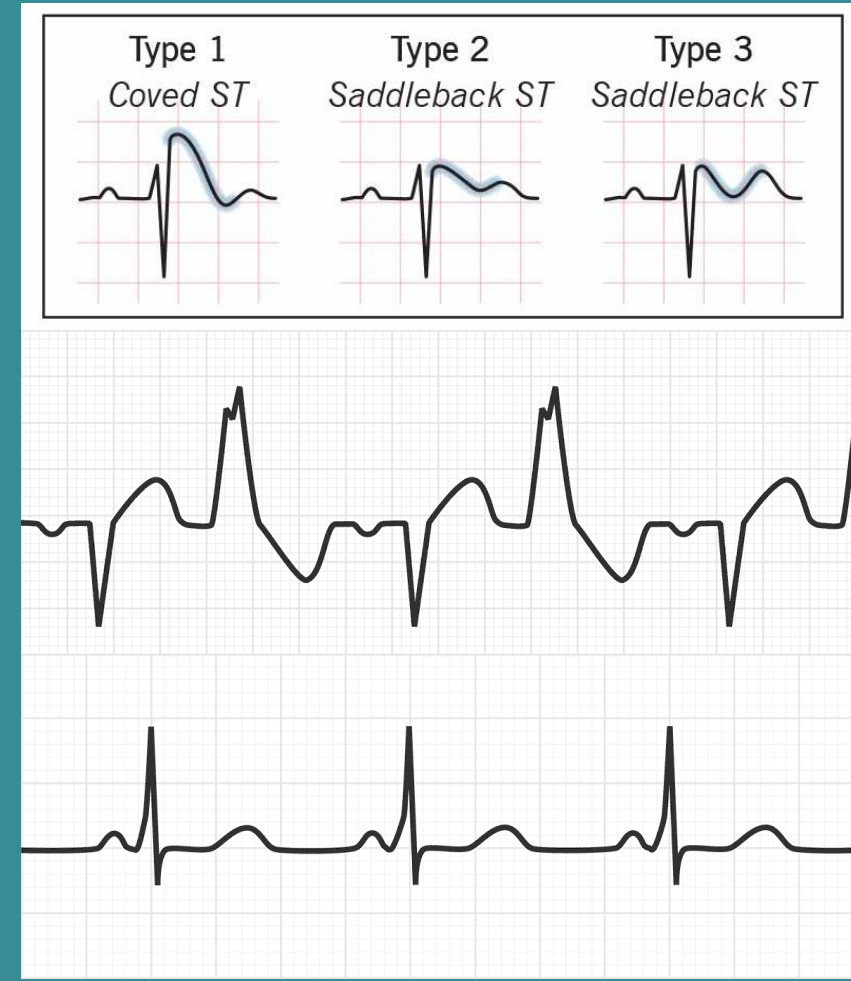
Long QT syndrome

- Genetic channelopathy characterized by prolonged ventricular repolarization
 - QTc >480ms female and >470ms male (postpubertal)
 - Highest risk when QTc >500ms
- Propensity to life-threatening ventricular arrhythmias
 - Especially during sport (3% SCD)
 - Triggered by several stimuli in athletes (Adrenergic surges related to emotional stress, electrolyte disturbances)
- Inheritance – primarily Autosomal dominant



Other electrical cardiac causes of SCD

- Brugada syndrome (<1%)
 - Inherited channelopathy affecting Na channels – Cove type ST elevation + RBB
 - Propensity for ventricular arrhythmias + SCD particularly when ↑ temperature
- Catecholaminergic polymorphic ventricular tachycardia (CPVT) (<1%)
 - Inherited arrhythmia
 - Stress-induced bidirectional VT
 - Often normal baseline ECG but bidirectional PVCs on exercise tests
- Wolff-Parkinson White syndrome (WPW) (3%)
 - Ventricular preexcitation + documented SVT
 - Risk of malignant arrhythmia is low



Case 3 – Long QT syndrome

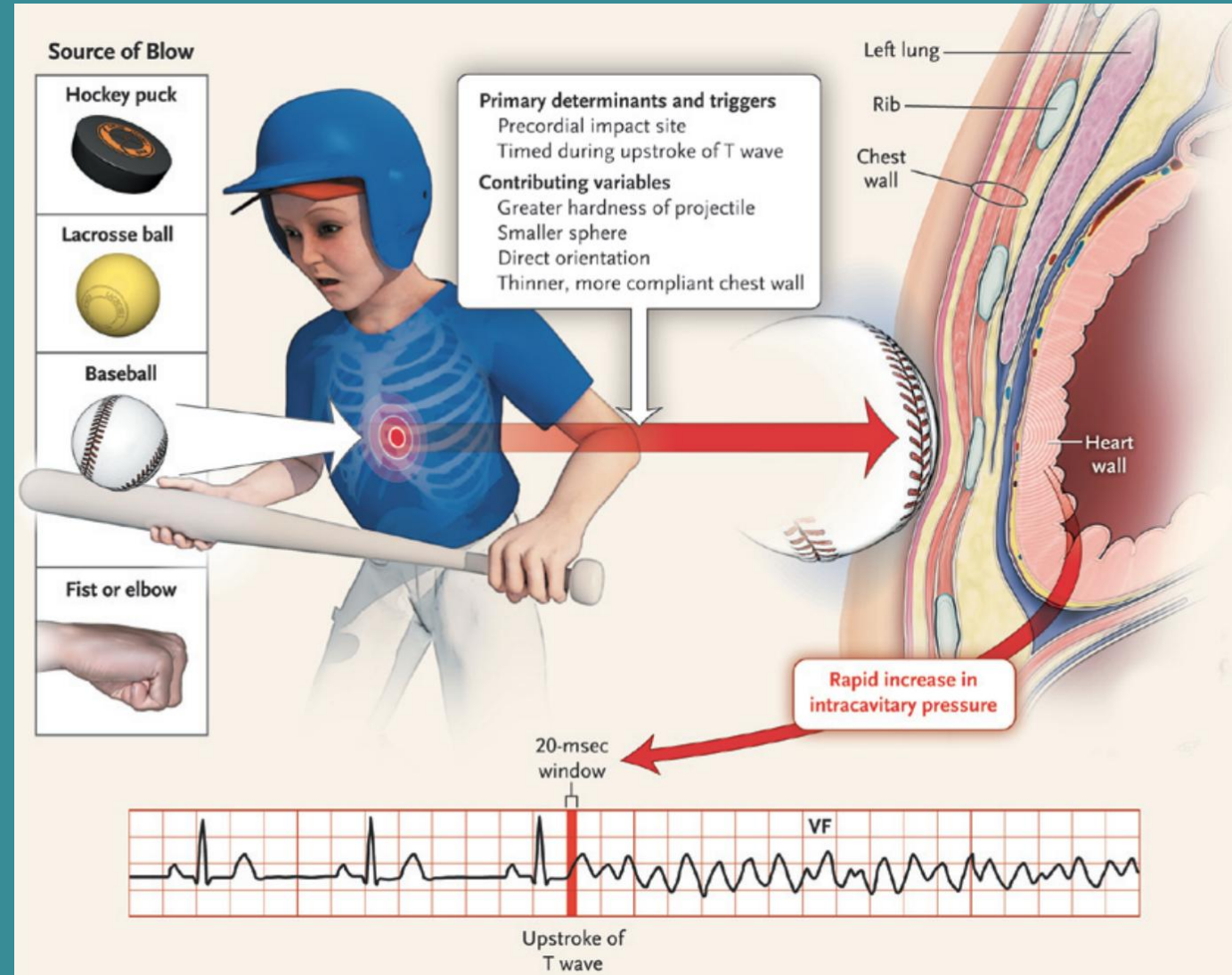
- History
 - No previous syncopal episodes or other cardiac symptoms
 - Family history of cousin who died suddenly while swimming
- Physical examination – Normal
- Echocardiogram – Normal
- Genetic testing – KCNQ1 mutation = LQT1

Other cardiac conditions causing SCD

- Commotio cordis
- Valvular heart disease
- Aortic dissection

Commotio cordis

- SCD related to sudden blow to the precordium (2%)
- More common in children/adolescents due to pliant chest walls
- Sudden blow at onset of T-wave → ventricular fibrillation



Other cardiac conditions causing SCD

- Valvular heart disease
 - Mitral valve prolapse (MVP)
 - Common condition but very rarely associated with malignancy arrhythmias
 - Higher risk of family history of SCD, ECG abnormalities (inferior T-wave inversion, QTc prolongation, ventricular extrasystoles)
 - Bicuspid aortic valve
 - Recent study present in 1% either attributed to aortic stenosis or as an incidental finding
- Aortic dissection
 - Rare cause of SCD in athletes (5%)
 - Static exercise - weight training may result in increased BP and possibly aortic dilation
 - Marfan syndrome may affect basketball/volleyball players resulting in aortic dilation

Child athletes are not small adult athletes

| | Children/young adults (<35 years) | Adults (>35 years) |
|-----------|---|---|
| Etiology | Inherited or congenital cardiac conditions | Primarily atherosclerotic coronary artery disease |
| Screening | Universal screening AHA 14-point screen +/- ECG | Targeted screening based on coronary risk assessment History, risk factors +/- ECG and stress testing |

Exercise in Young Athletes with Cardiac Disease

- Traditionally restrictive → Currently more liberal
- Promote **shared decision-making approach** – athlete, family and medical team
- **Individualized** by disease type and severity
- Must weigh benefits of sport and risk of SCD
- Encourage **safe inclusion** whenever possible

Key take aways

1. Most pediatric athletes are safe to play
2. Screening aims to identify risk
3. Recognize red flags early and refer appropriately
4. Promote safe participation through collaboration

Questions?

Email me at jon@lipikids.com

Referral information at www.lipikids.com