Objectives

- Cardiac-related Primary Care Issues:
  - Syncope
  - Sports clearance
  - ECG screening
    - Athletes
    - ADHD medications
  - Current management of SVT
  - Considerations in management of WPW
    - Symptomatic
    - Asymptomatic
  - Genetic influence in the management of Channelopathies
    - Long QT
    - CPVT

CASE

14 year old female soccer player

Syncope

- Returned home from soccer tournament
- Played two games in one day
- Felt dizzy in shower, no palpitations
- Out of shower, passed out in bathroom

History

- No family history of sudden cardiac death
- No previous syncope
CASE
14 year old female soccer player
Syncope
• Playing soccer
• During play, running down field → syncope
• History
  • No family history of SCD
  • No history of syncope

CASE
14 year old female soccer player
Syncope
• Middle of a soccer game
  • Played the whole first half
• Sat down for half-time coaching talk
• Stood up to run to field → syncope
• History
  • No family history of SCD
  • No history of syncope

Syncope
• Definition – transient global cerebral hypoperfusion
  • Rapid onset
  • Short duration
  • Spontaneous complete recovery
  • Vast majority of syncope in children is vaso-vagal.
    • Typical history of vaso-vagal, normal exam, and normal ECG often sufficient

Syncope in a Pediatric Population

- Retrospective, single center with large EP referral
- 87 consecutive patients referred for syncope
  - Vaso-vagal: 86%
    - History/exam/ECG sufficient for dx: 71%
  - Neurologic: 13%
    - Conversion disorder, migraine, seizures, other behavioral
    - Headache, lack of pre-syncope prodrome
  - Catecholaminergic Polymorphic VT: 1%

Vasovagal – Reflex – Neurally Mediated

- Diagnosis – cardiovascular reflexes to control circulation are intermittently inappropriate → bradycardia and hypotension.
- Treatment
  - Education and reassurance
  - Minimize caffeine
  - Hyperhydration (clear urine)
  - Increased salt (sports drinks helpful)
  - Maneuvers to terminate episode
    - supine positioning
    - Physical counterpressure maneuvers (leg crossing, hand grip)
  - Florinef/midodrine for refractory cases, though limited evidence

Indications for further evaluation

- Family history premature SCD (< age 30 years)
- Known/suspected heart disease
- Event triggers (loud noise, emotional stress)
- Exertional syncope
- Syncope without prodrome, while supine, preceded by chest pain or palpitations.
Sports Screening and Screening ECGs

Sports Clearance
2007 AHA Guidelines

- Patient history
  - Exertional chest pain
  - Unexplained syncope
  - Exertional dyspnea/tachypnea
  - Heart murmur
  - HTN
- Physical exam
  - Heart murmur
  - Diminished femoral pulses
  - UE blood pressure
  - Marfanoid stigmata
- Family History
  - Premature SCD <50 years
  - Disability from heart disease in relative ages <50 yrs
  - Specific diagnoses (LQTS, HCM, ARVC)
- Parental verification required
- Refer for 1 or more findings
- Cardiac screening should not be restricted to athletes

KHSAA Sports Screening Form

HISTORY
1. Have you ever passed out during exercise?
2. Have you ever been dizzy during or after exercise?
3. Have you ever had chest pain during or after exercise?
4. Have you ever had high blood pressure?
5. Have you ever been told you have a heart murmur?
6. Have you ever had racing of your heart?
7. Has anyone in your family died of heart problems before 50?

PHYSICAL
- Consider location of exam
- Rhythm (regular/irregular)
- Murmur (biphasic)
- Murmur (standstill)
- Pulses
- Marfanoid features

MISSING
- Family history detail
Italian Pre-participation Screening ECG

- 1982 – State Law by Ministry of Health
- Initiation of national pre-participation screening and medical clearance program
  - Annual ECG
  - Annual history and physical
  - Endorsed by International Olympic Committee
  - Endorsed by European Society of Cardiology
- Disqualification of athletes targets those with probable disease, not just those with confirmed disease

Pelliccia et al. Bethesda Conference #3 and the European Society of Cardiology Consensus Recommendations Revisited. JACC 2008

Athletic ECG screening

Comparison of pre- and post-screening SCD rates / 100,000 person years
- Pink – Italian ECG screening initiated 1982 (**evidence for ESC recs)
- Green – Israel ECG/exercise screening initiated 1997
- Yellow – Mayo Clinic, no ECG screening

Steinvil et al. Mandatory ECG screening of athletes. JACC 2011

Accuracy of Screening ECGs

On-line survey, 18 ECGs, pediatric cardiologists
  - 8 normal
  - 10 abnormal (HCM x 4, LQT, WPW x 2, PAH, myocarditis x 2

Hill et al. Accuracy of Interpretation of Preparticipation Screening ECGs. J Peds 2011
ECG screening in U.S.

- Usefulness limited by lack of physician workforce
- Cost estimate in US – Assume 10 million athletes (high school/middle school)
  - Physical/ECG - $750 million
  - 15% estimated to need additional testing (1.5 million athletes): cardiac eval/echo – $750 million
  - Annual cost est $1.5 billion + admin costs
  - Cost of preventing one death $3.4 million
- NHLBI working group: ECG screening in athletes to determine norms


Stimulant Medications

- AHA scientific statement 2008: universal ECG screening for stimulant meds for ADHD
- Single center, retrospective and survey analysis.
  - Compared ECG ordering practices one year prior to and after AHA statement.
  - Excluded patients with known cardiac disease or arrhythmias.
  - Single questions email survey to community pediatricians
- Results:
  - 24/372 (6.4%) ECG abnormalities (ventricular hypertrophy, PVCs, PACs, LAD, mild QTc prolong, AEL, T wave abnormality).
  - 18 further evaluation: 17 no cardiac abnormalities
  - None restricted from stimulant use


Stimulant Medications

- Retrospective cohort
  - Automated data from 4 health plans
- Inclusion:
  - Use of ADHD drug during study period
  - Age 2 – 24 years
  - Absence of life-threatening illness
  - Included patients with congenital heart disease
  - Control subjects matched by age and gender
  - 1,200,438 children/young adults age 2 – 24 years
- Results:
  - 81 serious CV events (SCD, MI, stroke) (3.1/100,000 person-yrs)
  - No increased risk of serious CV events in users of ADHD drugs

Cooper et al. ADHD Drugs and Serious Cardiovascular Events in Children and Young Adults. N Engl J Med 2011.
Current Management of SVT

ECG
1 week old with irritability

2 year old
Fussy

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Supraventricular Tachycardia

• Incidence in pediatric population as high as 1:250
• Rarely life-threatening
• Significant impact on quality of life

Symptoms
• Palpitations
• Chest pain in younger children
• Associated dizziness or shortness of breath

Diagnostic testing
• Echo
• ECG/Holter/Event Monitor

Supraventricular Tachycardia in Pediatrics

• Atrioventricular reentry tachycardia (AVRT)
• AV node reentry tachycardia (AVRNT)
• Focal atrial tachycardia
Accessory pathway mediated tachycardia

Supraventricular Tachycardia Management
- Watchful waiting
- Infrequent episodes, reliably terminated with Valsalva
- Medical therapy
  - Beta-blocker
  - Flecainide
- Ablation – technological advancements
  - Advanced mapping systems – Minimize fluoroscopy use
  - Success rate: 90-95%
  - Risk of adverse event: 0.1-0.5%
- Considerations in management
  - Frequency and control of episodes
  - Age/size
  - Comorbidities (ADHD, asthma)

Ablation of SVT in Children Safety of RF ablation <15kgs
- Multicenter, retrospective
- Inclusion: Patients <20kg, EP study for SVT Jan 94 – Jan 03
  - < 15kg vs 15.1 – 20kg
  - Results (N=69)
    - Higher incidence of structural heart disease <15kg
    - No difference in mechanism of tachycardia
    - No difference in procedure time or RF applications/time
    - No difference in complications
    - No difference in short-term or long-term success


Ablation Therapy for ADHD

- Single center, retrospective study
- Inclusion:
  - Patients under age 21 years with prior dx of ADHD.
  - EPS 2002 – 2009, SVT or WPW (N=695)
- Results (N = 20)
  - 11 WPW, 7 AVNRT, 2 concealed APs
  - 18 on ADHD medications (10 on two or more)
  - 10 on cardiac medications (beta-blocker, CCB, digoxin)
  - 17 underwent catheter ablation
  - 4 patients with increased ADHD therapy post-ablation.


Ablation of SVT in Children

Quality of Life

- Single center
- age 5 – 18 years
- Re-entry SVT
- Pediatric Quality of Life Inventory cardiac model questionnaire administered prior to and 6mos post-ablation.

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Pediatric EP Study

- Outpatient procedure
- General anesthesia
- Bilateral femoral venous access
- Rare femoral arterial access
- EP study protocol
  - Diagnostic baseline testing
  - Mapping/ablation
  - Post-ablation testing
- Left sided ablations via transeptal access
- Fluoroscopy minimized
- Duration: 2-4 hours
ECG 16 year old with chest pain

ECG 12 year old with syncope at camp

Wolff-Parkinson-White
Wolff-Parkinson-White

- 1 – 4.8/1000 patients
- Asymptomatic WPW
  - Pre-excitation discovered as incidental finding
- Symptomatic WPW
  - Documented SVT
  - Palpitations
  - Syncope
  - Sudden cardiac death
- Management based on ECG finding and symptoms
  - Risk stratification for risk of AF and Sudden Cardiac Death
    - Lifetime risk estimated 3-4%, though unclear
    - Treatment of SVT (AVRT)

ECG

13 year old with exertional dizziness
Asymptomatic WPW
Non-invasive Risk Stratification

- Staged approach to risk stratification:
  - Exercise testing (N=127).
  - Transesophageal EP study if no AP block with exercise (N=66).
  - High risk by TEEPS referred for ablation (N=17)
- Results
  - Invasive EP study N=56 (high risk asymptomatic, clinical tachycardia, syncope, pre-ASD repair, SCD)

Asymptomtaic WPW
Invasive Risk Stratification

- 184 children, age 8-12 years
- 1995 – 2005
- Asymptomatic pre-excitation on ECG
- EP testing, followed as outpatients on no medications
- 133 remained asymptomatic
- 51 arrhythmic event
  - 19 potentially life-threatening
- Risk factors for potentially life-threatening arrhythmic events
  - Inducibility of SVT
  - Accessory pathway conduction properties
  - Multiple accessory pathways

PACES/HRS/AHA/ABP
Asymptomatic WPW
2012 Concensus

- Age 8-21
- Conduction of Accessory pathway – When does the AP stop conducting?
  - Holter
  - Exercise testing
  - Invasive assessment
    - Trans-esophageal EP study
    - Intracardiac EP study
Inherited Channelopathies
Ion channel disturbances

- One of the causes of SIDS
- Genetic heterogeneity
- Variable penetrance
- Present with syncope, seizure, sudden death
- Most common
  - LQT syndrome
  - CPVT
  - Brugada
  - Short QT syndrome

Channelopathies Diagnosis Not Always Obvious
16 year old Cardiac arrest in school

Neonate Bradycardia and seizures
### Long QT Syndrome

#### LQT1
- Broad T wave
- Syncope or sudden death with exercise
- Treatment with beta-blocker

#### LQT2
- Syncope or sudden death with sudden auditory stimuli or strong emotion

#### LQT3
- Abnormal bradycardia
- Sudden death during sleep
- Treatment with mexiletine

#### LQTS Management
- Avoid QT-prolonging drugs
  - [http://www.qtdrugs.org/](http://www.qtdrugs.org/)
- Avoid high intensity sports
- Beta-blocker therapy
- Risk stratification
  - Age/gender
  - Symptoms
  - QT interval
  - Genotype
- Left cardiac sympathetic denervation

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LQTS ICD therapy

- History of SCD
- Documented torsades de pointes
- Syncope despite beta-blocker
- LQT3
  - Flecaïnide or Mexilitine
- Excessive QT prolongation (>550ms)
- Woman with LQT2 and QT >500

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Catecholaminergic Polymorphic Ventricular Tachycardia

- RyR2 - cardiac ryanodine receptor gene
- Autosomal dominant
- Exercise induced syncope, seizures, and SCD
- Normal resting 12 lead ECG
- Dx made based on symptoms
- Bidirectional ventricular tachycardia
- Treatment
  - Beta-blocker
  - ICD

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Summary

- Primary care frequently encountered issues
  - Syncope – majority in pediatric population is benign
  - 2007 AHA recommendations for sports screening are inclusive
  - ECG screening for stimulant use is low yield
  - Preparticipation screening ECGs are not inclusive
  - SVT in children can be safely ablated in the current age.
  - SVT should not affect a child’s lifestyle.
  - Symptomatic WPW should be ablated.
  - Asymptomatic WPW should be referred to pediatric EP for risk stratification.
- Management of channelopathies
  - LQT management is largely guided by genotype
  - CPVT can often be treated with beta-blocker therapy
UK Pediatric Cardiology Services

- Weekly add-on clinics
- UK Cardiac Sports Clearance clinics
  - (June/July/August)
- Telemedicine clinics
  - Weekly

University of Kentucky Pediatric Cardiology

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