Arrhythmias in Pediatrics: Objectives

1. Review and refresh the basics of arrhythmias
2. Have a healthy relationship with arrhythmias:
   – Awareness
   – Curiosity
   – Respect
   – Balance of confidence and knowledge with a healthy dose of concern
3. Breathe easier knowing when and how to refer & manage patients
Context is key

Vasovagal syncope
Context: ECG strips leading up to that moment: ST and T wave changes during sinus rhythm
Arrhythmias: Electrical Heart Disease

- **Benign ectopy:**
  - Premature atrial contractions (PAC)
  - Premature junctional contractions (PJC)
  - Premature ventricular contractions (PVC)

- **Tachyarrhythmias:**
  - Supraventricular tachycardia (SVT)
  - Ventricular arrhythmias

- **Bradycardia:**
  - Sinus bradycardia
  - Heart block

**MANY (MOST) ARE BENIGN OR HEMODYNAMICALLY WELL TOLERATED (AT LEAST IN THE SHORT TERM)**
Classification of Arrhythmias

• Sporadic
  – SVT, some ventricular tachycardia

• Congenital heart defects
  - Pressure or volume overload
  - Post-operative
  - Late arrhythmias related to scars from suture lines

• Familial = inherited = genetic
  – Long QT syndrome (LQTS)
  – Brugada syndrome
  – Catecholaminergic polymorphic ventricular tachycardia (CPVT)

• Muscle disorders (Structural ± inherited)
  – Hypertrophic, arrhythmogenic, and other cardiomyopathies

• Other: Acquired, Infectious, Autoimmune
  – Myocarditis, Chagas, Lupus

* Examples only! Not a complete list
Why worry? Implications of Arrhythmias

• Symptoms
  – May lead to disruption to daily activities
  – Dizziness or loss of consciousness → injury

• Ventricular dysfunction
  – If incessant and/or unrecognized

• Sports participation and physical activity
  – Many pediatric arrhythmias require no restrictions
  – Children often self-limit
    • By symptoms or due to worry
  – Some sports present unique risks
    • Swimming

• Risk of collapse or sudden death
  – Rare in children
  – May be related to exercise...may not be (diagnosis-specific)
“Sudden Death” on the Pitch

By Report By Christopher C. ...

Last updated Jun 18, 2019

A Tampa Hillsborough

Now, the
CARDIAC ARREST VS. HEART ATTACK

People often use these terms interchangeably, but they are not the same.

**WHAT IS CARDIAC ARREST?**

CARDIAC ARREST occurs when the heart malfunctions and stops beating unexpectedly.

Cardiac arrest is triggered by an electrical malfunction in the heart that causes an irregular heartbeat (arrhythmia). With its pumping action disrupted, the heart cannot pump blood to the brain, lungs and other organs.

**WHAT HAPPENS**

Seconds later, a person becomes unresponsive, is not breathing or is only gasping. Death occurs within minutes if the victim does not receive treatment.

**WHAT TO DO**

CALL 9-1-1 Cardiac arrest can be reversible in some victims if it’s treated within a few minutes. First, call 9-1-1 and start CPR right away. Then, if an Automated External Defibrillator (AED) is available, use it as soon as possible. If two people are available to help, one should begin CPR immediately while the other calls 9-1-1 and finds an AED.

**WHAT IS THE LINK?**

Cardiac arrest is a LEADING CAUSE OF DEATH.

Nearly 360,000 out-of-hospital cardiac arrests occur annually in the United States.

Fast action can save lives.

**WHAT IS A HEART ATTACK?**

A HEART ATTACK occurs when blood flow to the heart is blocked.

A blocked artery prevents oxygen-rich blood from reaching a section of the heart. If the blocked artery is not reopened quickly, the part of the heart normally nourished by that artery begins to die.

**WHAT HAPPENS**

Symptoms of a heart attack may be immediate and may include intense discomfort in the chest or other areas of the upper body, shortness of breath, cold sweats, and/or nausea/vomiting. More often, though, symptoms start slowly and persist for hours, days or weeks before a heart attack. Unlike with cardiac arrest, the heart usually does not stop beating during a heart attack. The longer the person goes without treatment, the greater the damage.

**WHAT TO DO**

CALL 9-1-1 Even if you’re not sure it’s a heart attack, call 9-1-1 or your emergency response number. Every minute matters! It’s best to call EMS to get to the emergency room right away. Emergency medical services staff can begin treatment when they arrive—up to an hour sooner than if someone gets to the hospital by car. EMS staff are also trained to revive someone whose heart has stopped. Patients with chest pain who arrive by ambulance usually receive faster treatment at the hospital, too.
Sudden Arrhythmic Death Syndrome (SADS) Facts

• Estimated 10% of SIDS

• Pediatric sudden death
  – 80/100,000 infants <1
  – 3/100,000 children >1

• 0.6/100,000 high school students
  – Conflicting data on athletes vs. nonathletes
  – Minority are arrhythmic

• SADS: 350,000 arrhythmic sudden deaths/year in US
  – 4,000 in people <35 years old (CDC 2002)

https://www.sads.org
Chugh et al Heart Rhythm 2009
Priori et al Heart Rhythm 2013 10:1932-63
Toresdahl et al Heart Rhythm 2014 11(7):1190-4
WHEN SHOULD WE WORRY?
## Symptoms & History

### Act!
- Atypical syncope*
- Atypical seizure*
- Concerning family history

### Caution!
- Chest pain*
- Harsh murmur or other concerning exam findings
- Known congenital heart disease

### Breathe!
- Isolated palpitations without dizziness, chest pain or presyncope
- No personal or family history
- Normal exam

* Not always cardiac or concerning; context and history are key
When to think about an arrhythmia

- Palpitations
  - At rest
  - Sudden start/stop
- Younger patients: chest pain
- Fainting – atypical
  - Without any warning
  - Mid exertion
  - Preceded by palpitations
- Near drowning (in a capable swimmer)
- Atypical seizures
  - Not responsive to antiepileptic medications
  - Brought on by emotion, startle
When to think about an arrhythmia

• Child with sudden cardiac arrest (SCA)
  – Aborted SCA (i.e. survived)
  – Post-mortem
    • No obvious cause or known diagnosis

• Family member with pacemaker, ICD, or documented malignant arrhythmias

• Family member with SCA at young age
  – <40-50 years
  – Structurally normal heart or negative autopsy
Sudden Death: Inherited Arrhythmia Syndromes

- **Ion channelopathies**
  - Long QT Syndrome (LQTS)
  - Brugada Syndrome
  - Catecholaminergic Polymorphic Ventricular Tachycardia (CPVT)

- **Cardiomyopathy with high risk of arrhythmia**
  - Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC)

Risk of polymorphic VT, ventricular fibrillation
Children’s National Cardiogenetics Clinics

1. Inherited Cardiomyopathy clinic
2. Inherited Arrhythmia clinic

- Electrophysiologist
- Geneticist
- EP Nurse
- Genetic counsellor
- Coordination with adult IA providers

202-476-3135
Rhythms

Worry!
- Cardiac arrest
- Ventricular fibrillation
- Polymorphic VT
- Prolonged pauses

Caution!
- WPW
- Heart block
- Sinus pause
- Slow SVT
- Incessant arrhythmia
- Ventricular tachycardia

Breathe!
- Most SVT
- Belhassen’s VT
- Isolated ectopy
- Sinus tachycardia (POTS)
SVT: Supraventricular Tachycardia

- Typically in structurally normal hearts
- Estimated up to 1:250 children
- Most common arrhythmia in pediatrics

Often very well tolerated hemodynamically
SVT: Supraventricular Tachycardia

- Atrial tachycardia:
  - Ectopic atrial tachycardia
  - Atrial flutter
  - Atrial fibrillation

- AV reentrant tachycardia:
  - Accessory pathway mediated
  - AVNRT

- Junctional tachycardia
EAT from left atrial appendage

Negative P waves in I

Negative P waves in aVL
Tachycardia-induced cardiomyopathy

• Incessant arrhythmias $\rightarrow$ ventricular dysfunction
  – Atrial tachycardia present “$\geq 90\%$” of the time
  – PVCs or ventricular arrhythmia “$\geq 24\%$” of the time

• With conversion to normal sinus rhythm, ventricular function in the vast majority of cases
  – Electrical cardioversion
  – Medications
  – Ablation

• Consider thromboembolic risk with incessant arrhythmias as well
AV Reentrant Tachycardia in WPW: typical “SVT”

Orthodromic AVRT

Antidromic AVRT
Irregular wide complex tachycardia

Preexcited atrial fibrillation
Degeneration into ventricular fibrillation

- Preexcited AF or sudden cardiac arrest may be *first symptom*
- Estimated 0.1% annual risk in asx WPW

Mode of sudden cardiac arrest in WPW
Pediatric Ventricular Tachycardia

- PVCs: 15% of infants, 40% of adolescents, 60% in some CHD
- VT: incidence less well defined; 50% have CHD
  - Belhassen’s VT (idiopathic left ventricular tachycardia)
    - Narrow complex tachycardia like SVT, but with superior QRS axis
    - Usually well tolerated hemodynamically
    - Will not terminate with adenosine but is sensitive to calcium channel blockers
  - Right ventricular outflow tract (RVOT) VT
    - Nonsustained or sustained
    - Usually well tolerated hemodynamically
  - Scar related
    - Congenital heart disease, arrhythmogenic right ventricular cardiomyopathy (ARVC), myocarditis
Belhassen’s Ventricular Tachycardia
(a.k.a. fascicular VT, verapamil-sensitive VT)

Left axis deviation, iRBBB pattern
Pediatric Ventricular Tachycardia

• When to worry?
  – Syncope, near syncope
  – Poor hemodynamics
  – Presence of congenital heart disease
    • Tetralogy of Fallot
    • Single ventricle physiology (Fontan)
  – Cardiomyopathy or heart failure
  – Myocarditis
  – Concerning family history
Pediatric Bradycardia

- Sinus bradycardia
  - Increased vagal tone (athletes)
  - Eating disorders
  - Sinus node dysfunction
    - Postoperative congenital heart disease
    - Familial sick sinus syndrome

- AV block
  - Congenital
    - Lupus antibody mediated
    - Intrinsic to CHD (AV canal defect, L-looped ventricles)
  - Postoperative
  - Inherited
Pediatric Bradycardia

- When to worry?
  - Symptomatic
    - Fatigue
    - Dizziness
    - Syncope
  - Severe bradycardia
  - Postoperative heart block
    - If persists beyond 7-10 days, need pacemaker
  - Family history of bradycardia, arrhythmias (atrial fibrillation, pacemakers or defibrillators), syncope, sudden unexplained death
Medications

Protect

- Disease specific
  - LQTS
  - Brugada

Caution!

- Psychiatric medications
- Energy Drinks

Breathe!

- ADHD medications

www.crediblemeds.org
www.brugadadrugs.org
ADHD Medications

- Minimal cardiac risks
- Main population in whom to have caution:
  - Long QT syndrome (LQTS)

ADHD Drugs and Serious Cardiovascular Events in Children and Young Adults

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Attention Deficit Hyperactivity Disorder and Long-QT Syndrome: Risky Business

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• Thorough review for history of possible symptoms, including any chest pain, fainting, dizziness, shortness of breath, palpitations, and/or exercised induced complaints.
• Any past medical history of cardiac disease, unexplained faints, or seizures.
• Review the family history for any relatives with heart disease; including, heart muscle disease (cardiomyopathy), fainting, arrhythmia (ex. long QT), sudden unexplained death, sudden infant death syndrome, drowning, unexplained car accidents.
• Perform a thorough cardiovascular physical exam.
• Screen for genetic syndromes or other diseases that could affect the heart.
Exercise & Sports Participation

Protect

- Hypertrophic cardiomyopathy
- Long QT syndrome (type 1)
- CPVT
  - Catecholaminergic Polymorphic Ventricular Tachycardia

Caution!

- HCM
- LQTS
- Pacemakers
- Defibrillators

Breathe!

- SVT
- Most isolated ectopy
Safe Sports

• Severe restrictions are almost always unnecessary
• Moving away from restriction toward assuring safe sports
  – Awareness of symptoms
  – Ability to stop and rest if needed
  – Stay well hydrated
  – In case of life threatening conditions:
    • Presence of AED
    • No swimming alone, caution with diving into cold water
• Joint care between Pediatricians and Cardiologist

• Initial evaluation includes:
  – Documentation of arrhythmia
    • ECG, Holter, extended Holters, and loop recorders
    • Patience
  – Additional testing may be indicated:
    • Echocardiogram, exercise stress test
    • ± MRI, family screening, laboratory testing, EP study

• Treatment
General Treatment Options

1. Simple observation
2. Medications
   - Daily prophylactic/maintenance therapy
   - Pill-in-the-pocket therapy
3. EP study with ablation therapy
4. Pacing
5. Implantable cardioverter defibrillator (ICD)
6. Surgical therapy
   - Surgical ablation, Maze procedure, sympathectomy
Conclusions

• All types of arrhythmias are possible in pediatrics
  – Many are well tolerated
• SVT is the most common and is usually not life-threatening
• Red flags that raise concern:
  – Incessant arrhythmias
  – Presence of congenital heart disease
  – Wolff-Parkinson-White syndrome
  – Inherited arrhythmia syndromes
  – Concerning family history
Children’s National Electrophysiology Team

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Not sure if you need to worry? Call us
THANK YOU

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