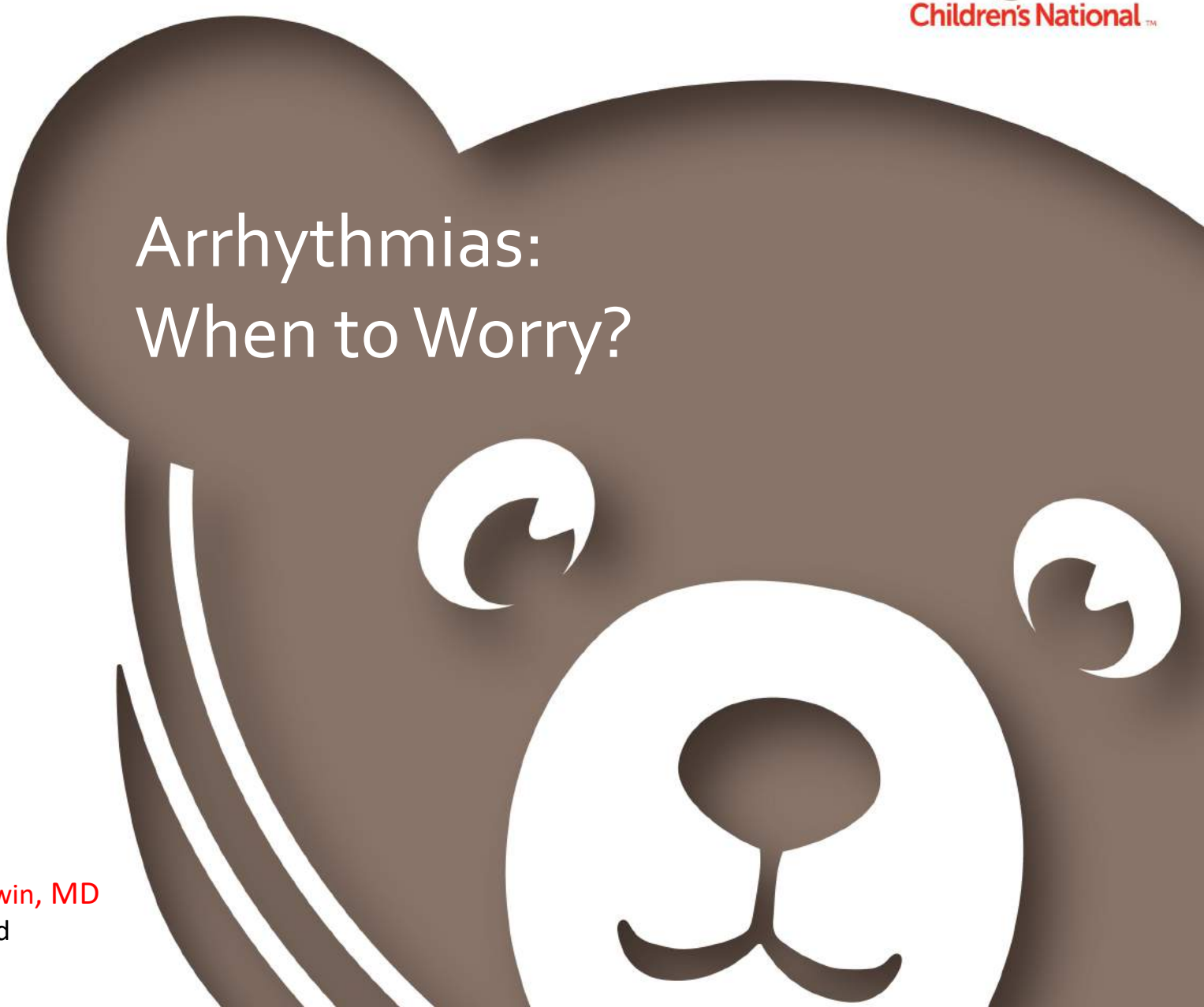
Future of Pediatrics
June 20, 2019

Children's National

Arrhythmias: When to Worry?



Elizabeth (Libby) Sherwin, MD
Pediatric Cardiologist and Electrophysiologist

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

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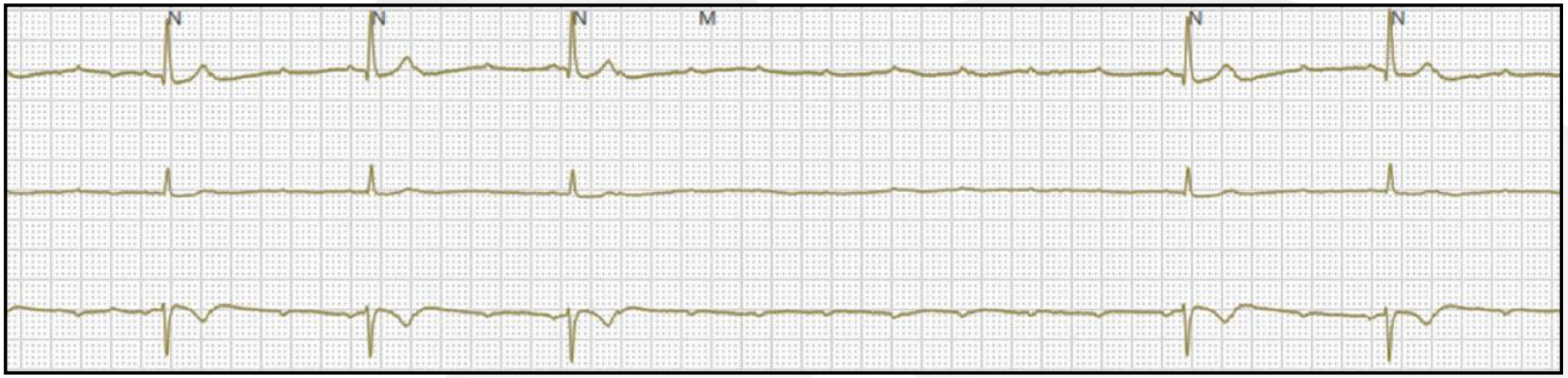
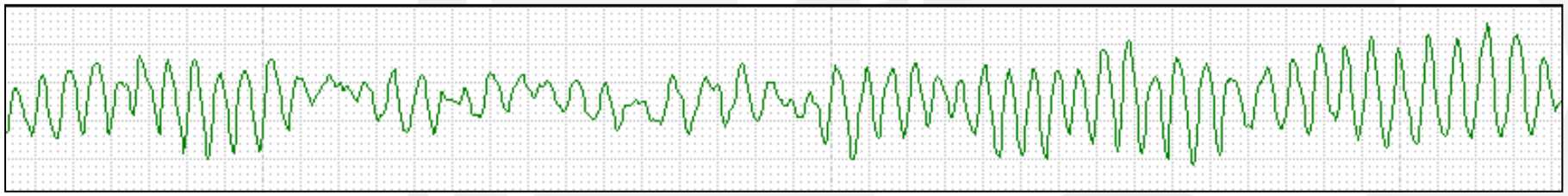
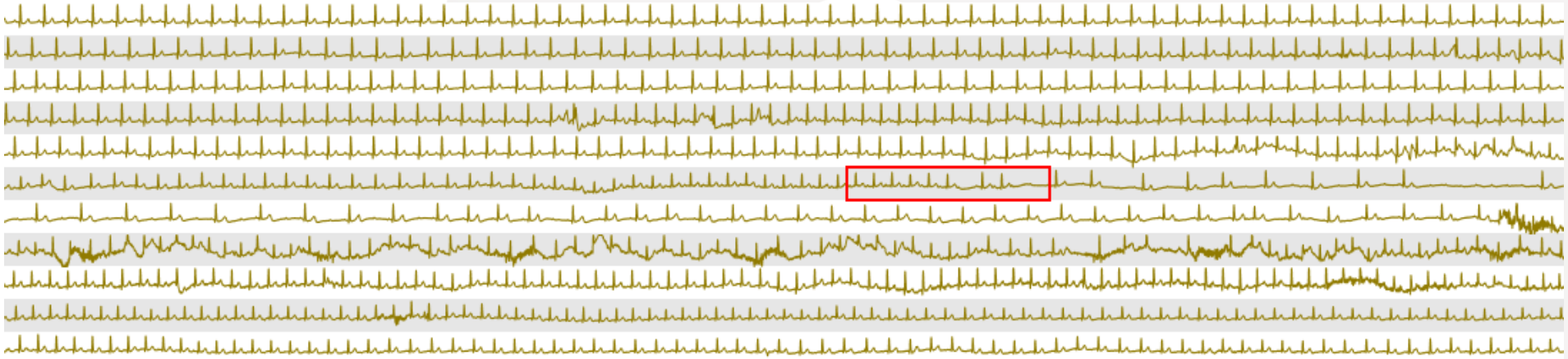




Context is key



Vasovagal syncope

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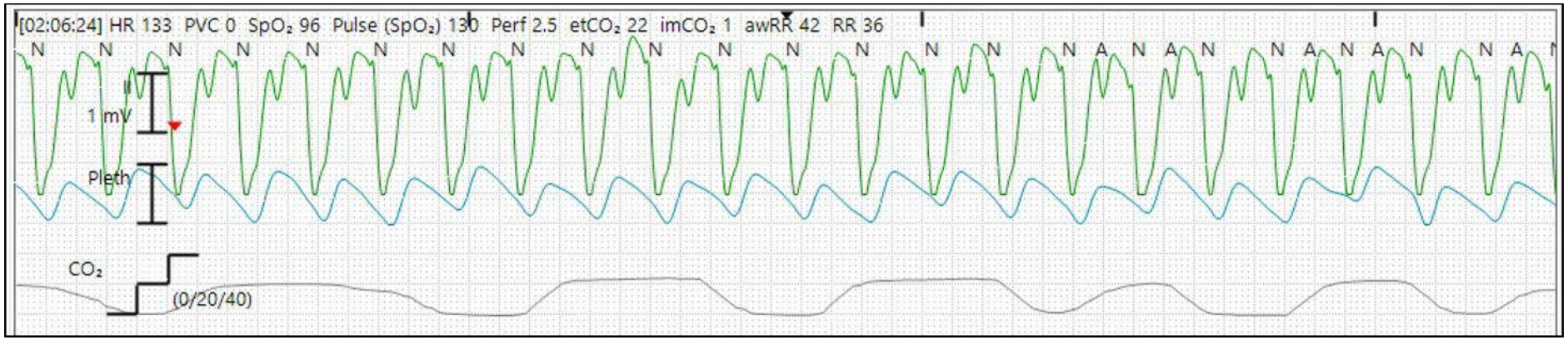

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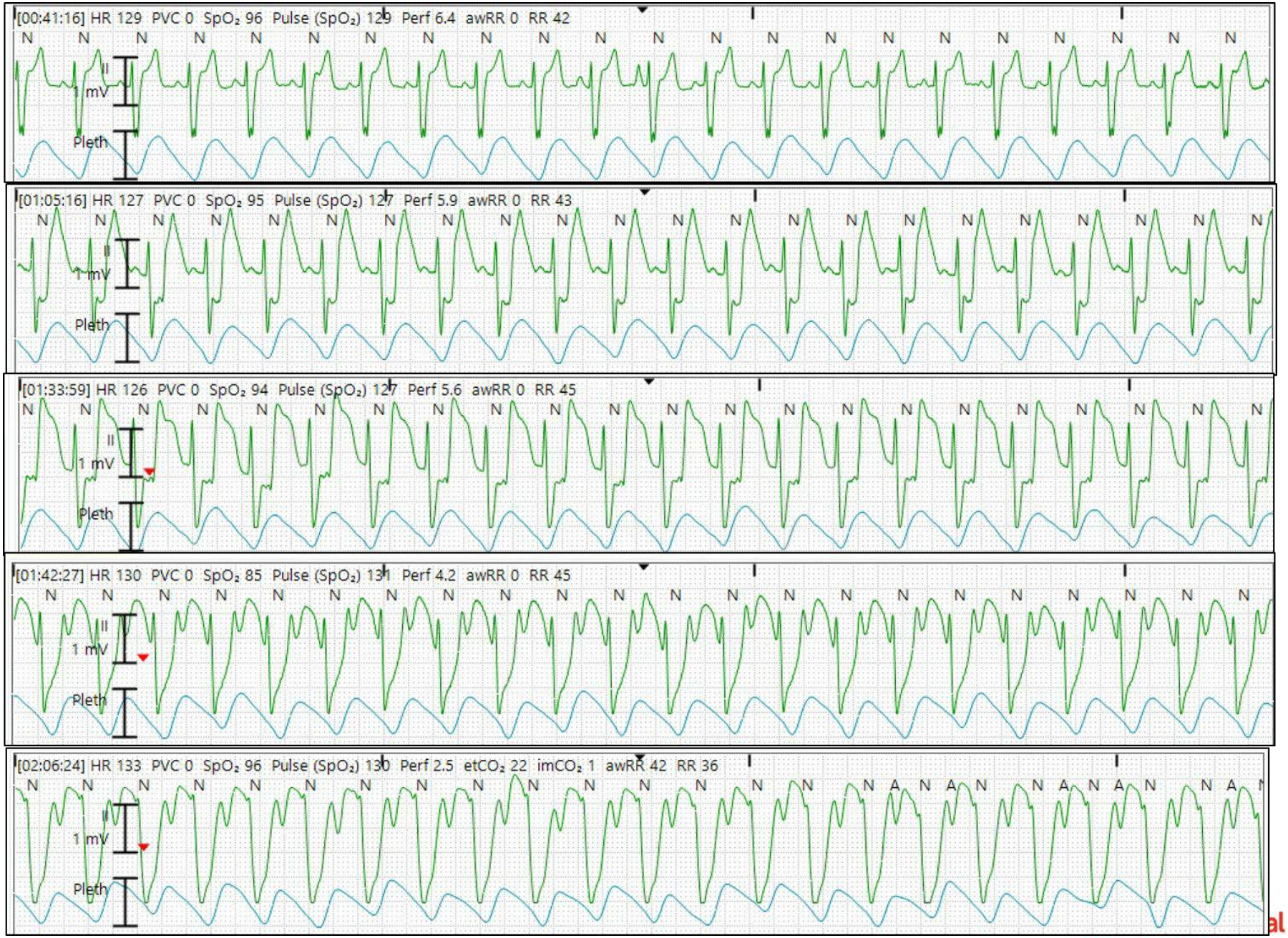
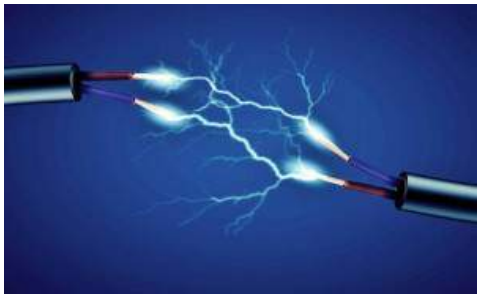
Context: ECG strips leading up to that moment: ST and T wave changes during sinus rhythm



Arrhy

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

- 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



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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

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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)

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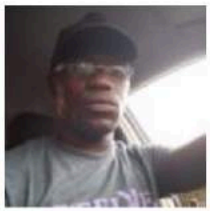
JUN 13, 2019

"SUDDEN DEATH" ON THE PITCH



By **Report By Christopher C. ...** — Last updated **Jun 18, 2019**

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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.

Cardiac arrest is an "ELECTRICAL" problem.

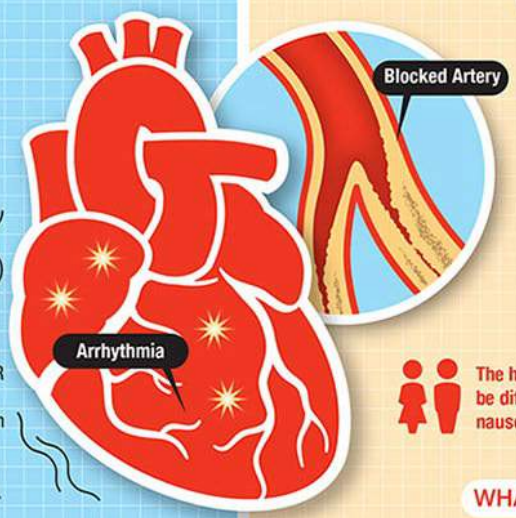
WHAT HAPPENS

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.



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 THE LINK?

Most heart attacks do not lead to cardiac arrest. But when cardiac arrest occurs, heart attack is a common cause. Other conditions may also disrupt the heart's rhythm and lead to cardiac arrest.

WHAT IS A HEART ATTACK?

A heart attack is a "CIRCULATION" problem.

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.**

The heart attack symptoms in women can be different than men (shortness of breath, nausea/vomiting, and back or jaw pain).

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.

American Heart Association. | CPR & First Aid

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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
Maron et al Am J. Med 2016 117:1339-1341

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WHEN SHOULD WE WORRY?

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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

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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

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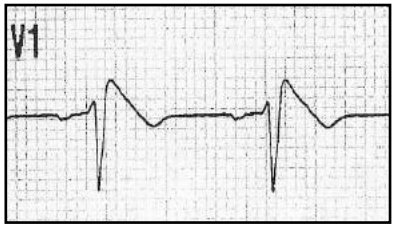
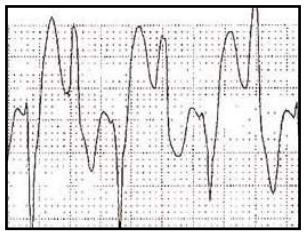
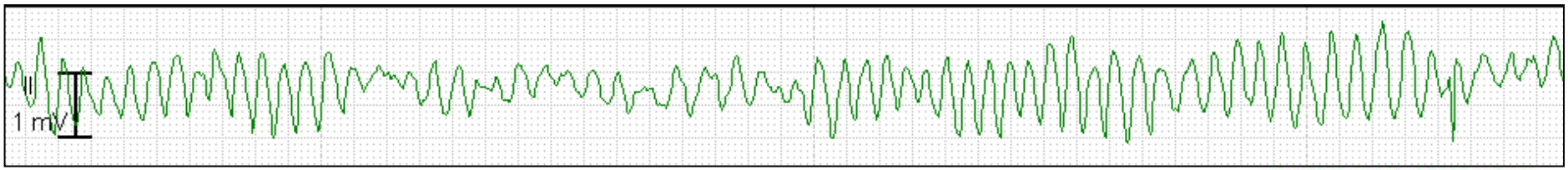
Sudden Death: Inherited Arrhythmia Syndromes





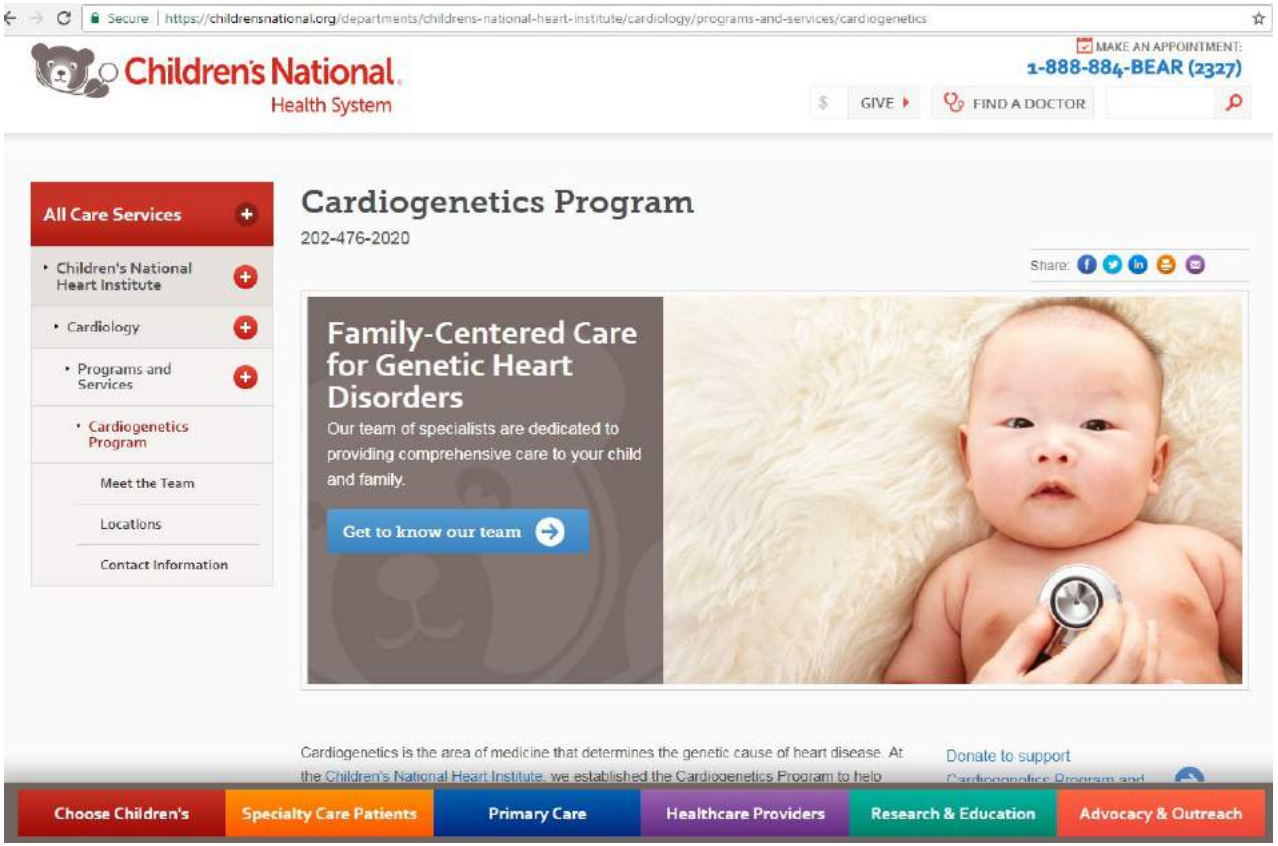
- Ion channelopathies
  - Long QT Syndrome (LQTS)
  - Brugada Syndrome
  - Catecholaminergic Polymorphic Ventricular Tachycardia (CPVT)
- Cardiomyopathy with high risk of arrhythmia
  - Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC)

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

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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)

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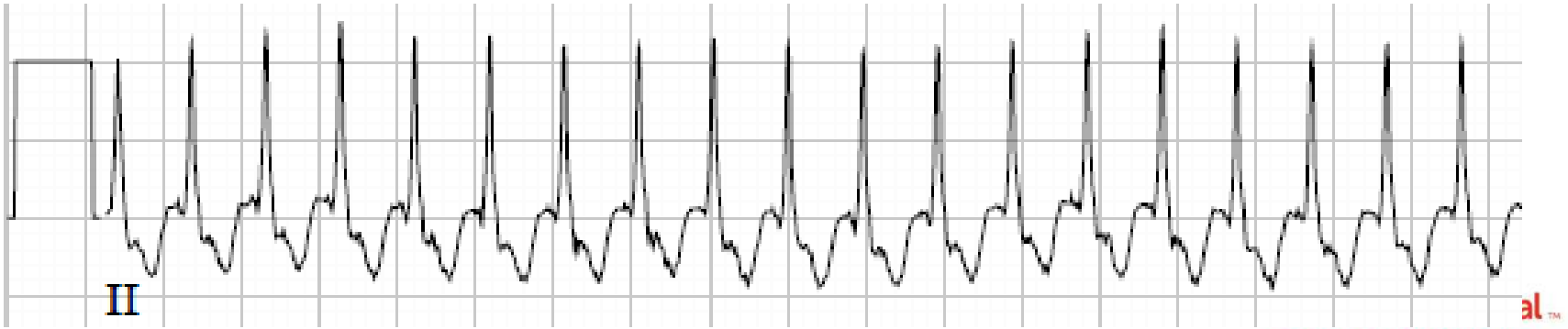
SVT: Supraventricular Tachycardia

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

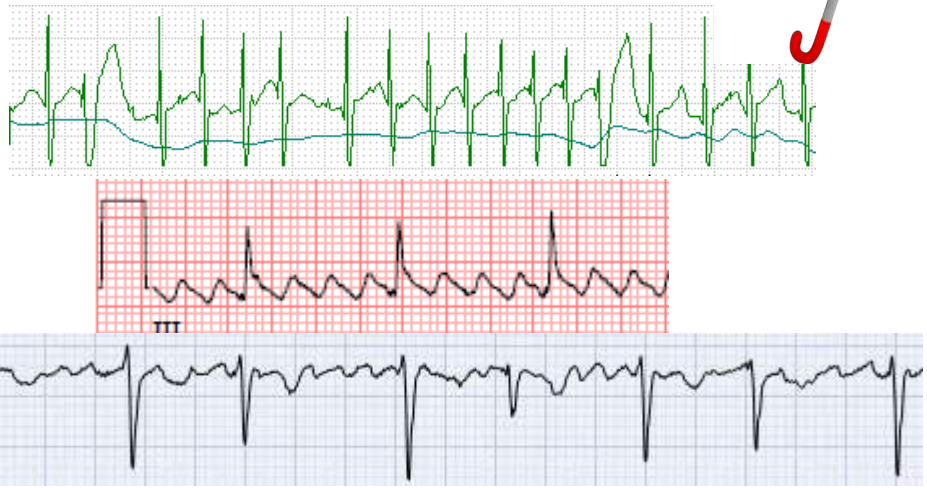
Often very well tolerated hemodynamically



SVT: Supraventricular Tachycardia

– Atrial tachycardia:

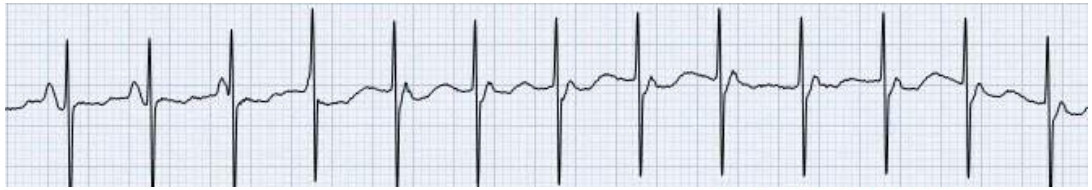
  - Ectopic atrial tachycardia
  - Atrial flutter
  - Atrial fibrillation



❖ **AV reentrant tachycardia**:

  - **Accessory pathway mediated**
  - **AVNRT**

– Junctional tachycardia



EAT from left atrial appendage

Tachycardia-induced cardiomyopathy

- Incessant arrhythmias → ventricular dysfunction
  - Atrial tachycardia present "≥ 90%" of the time
  - PVCs or ventricular arrhythmia "≥ 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

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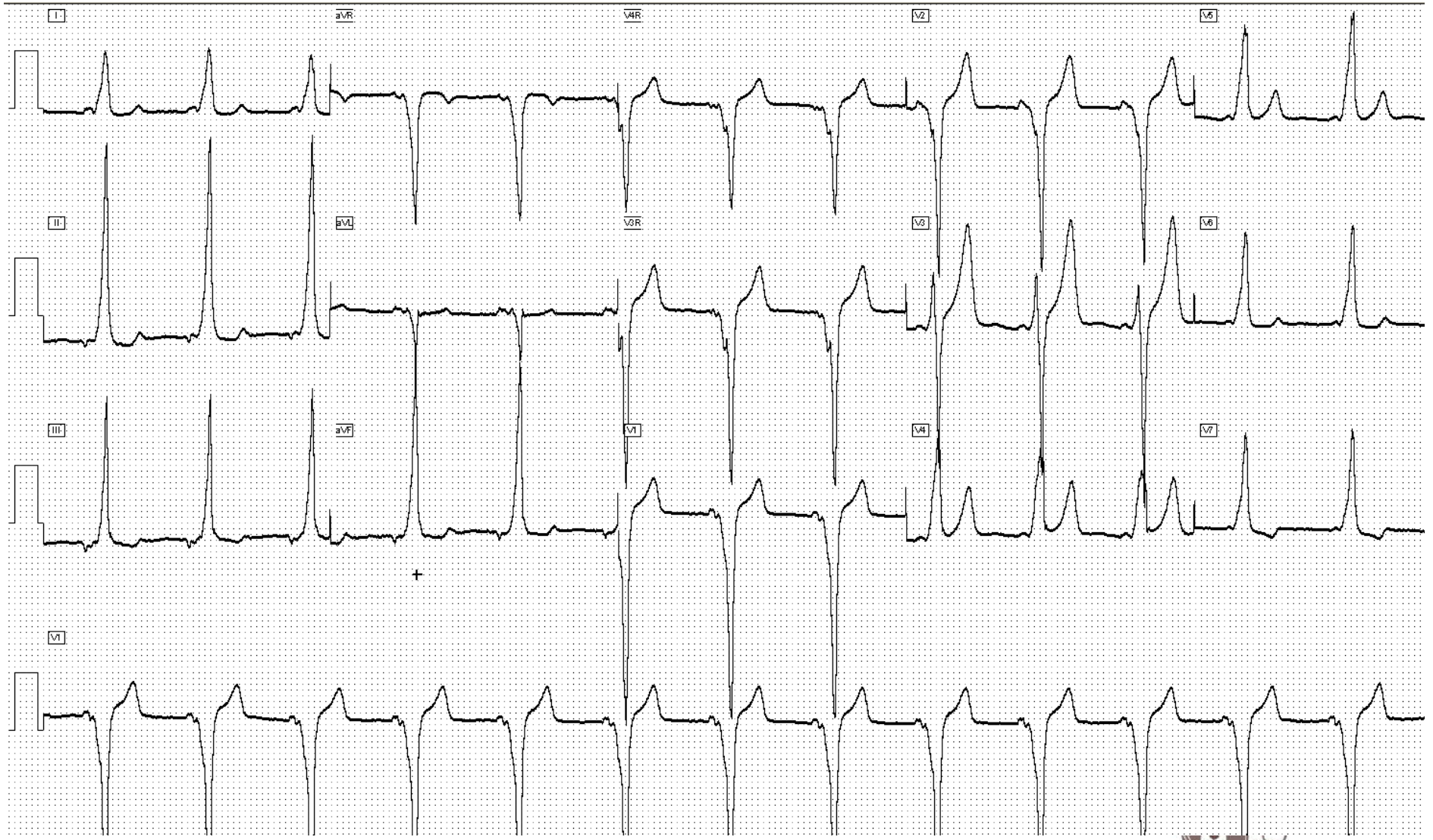
Wolff-Parkinson-White



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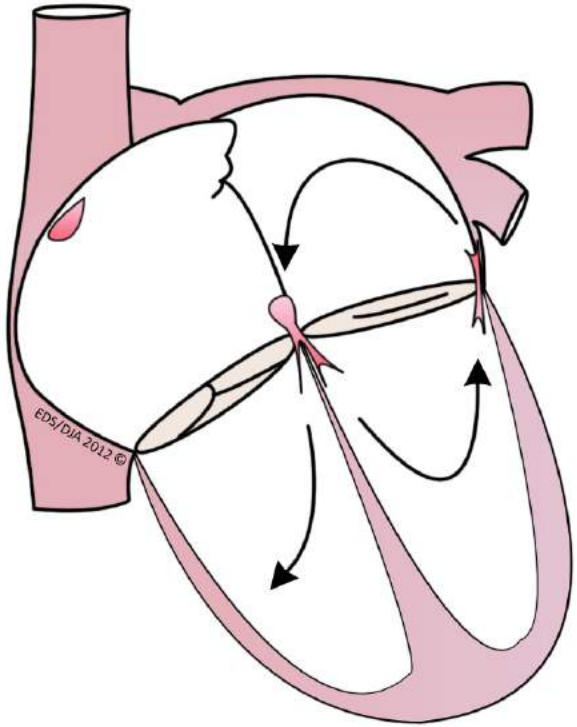
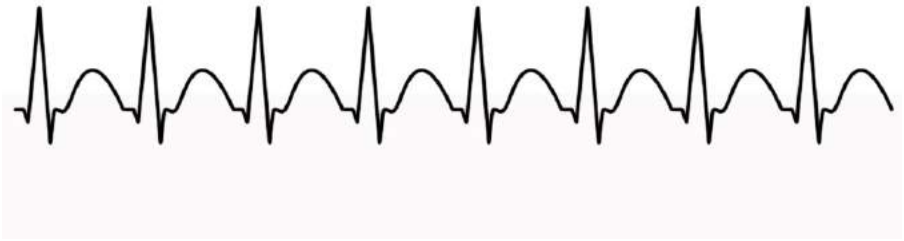
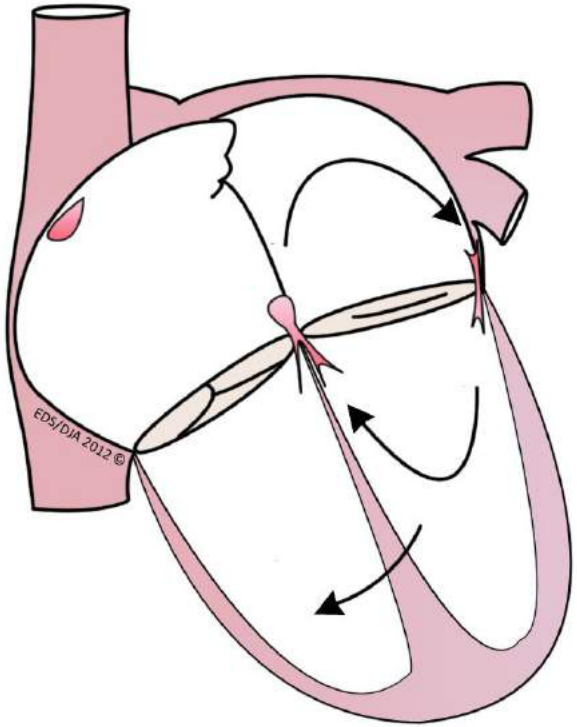
AV Reentrant Tachycardia in WPW: typical "SVT"





Orthodromic AVRT



Antidromic AVRT

Irregular wide complex tachycardia

Preexcited atrial fibrillation

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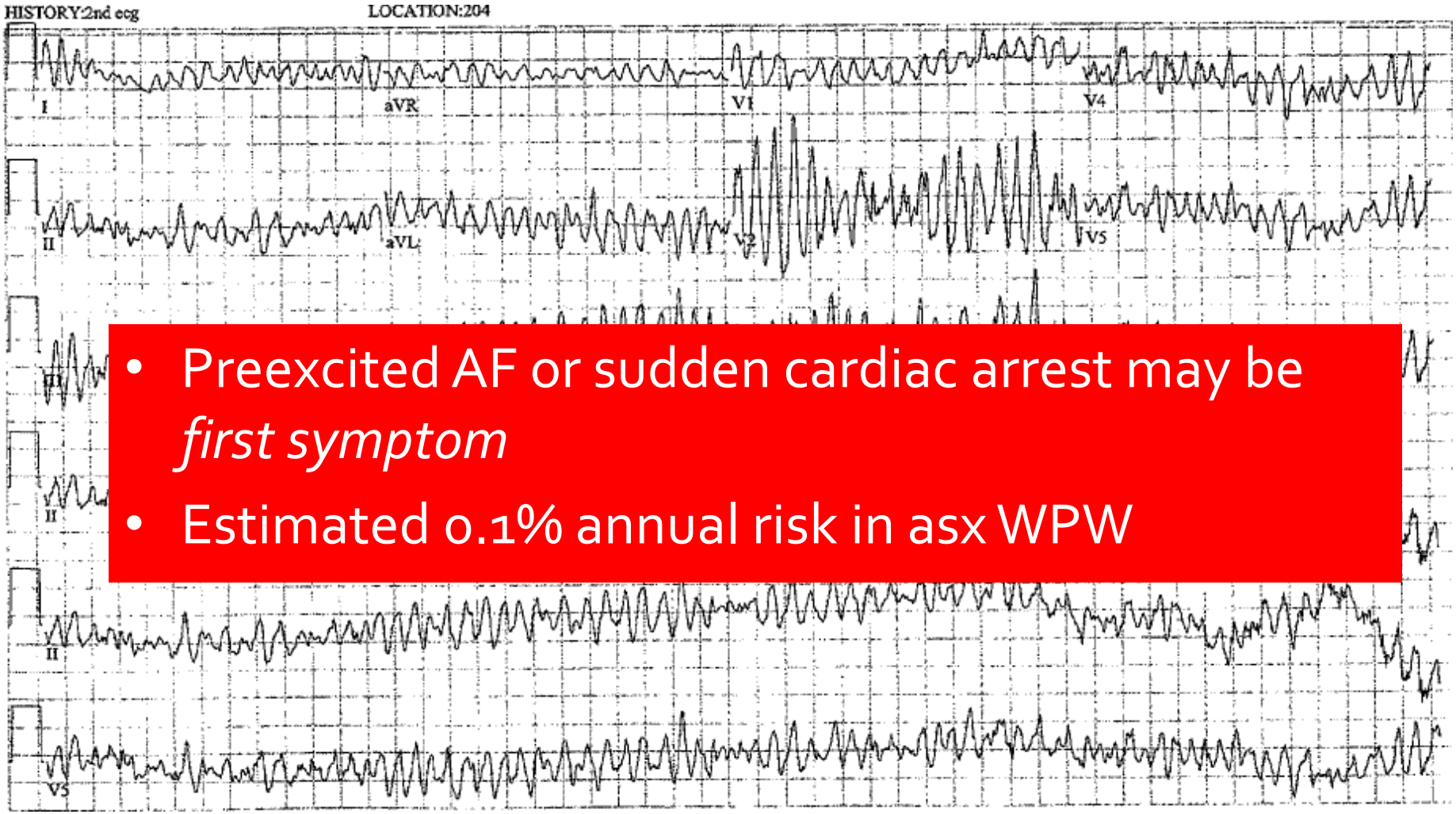
Degeneration into ventricular fibrillation



- 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

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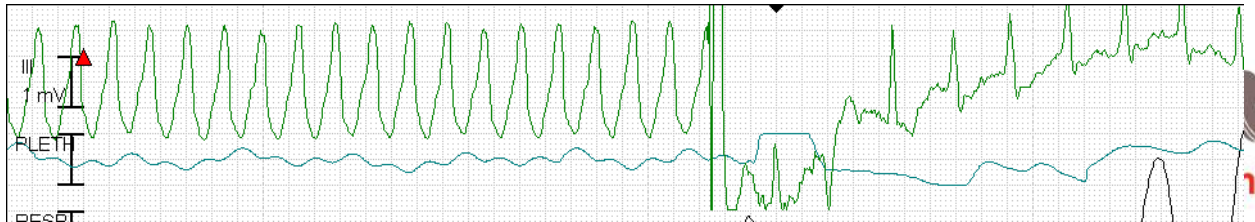
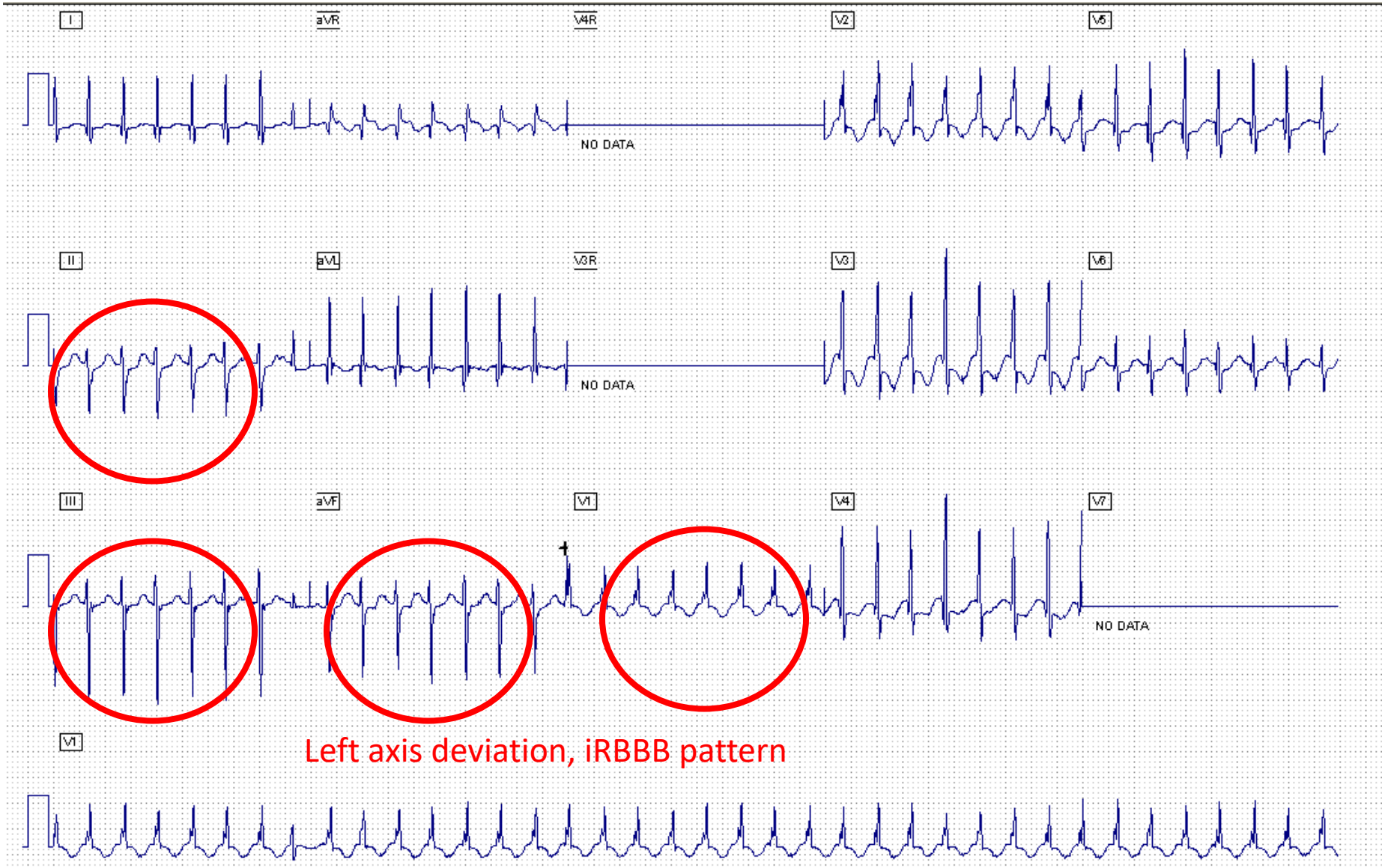
Pediatric Ventricular Tachycardia

- 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 (ARV(myocarditis



Belhassen's Ventricular Tachycardia (a.k.a. fascicular VT, verapamil-sensitive VT)



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

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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

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Pediatric Bradycardia

Circulation
JOURNAL OF THE AMERICAN HEART ASSOCIATION

American Heart Association

ACC/AHA Guidelines for Implantation of Cardiac Pacemakers and Antiarrhythmia Devices: Executive Summary : A Report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Committee on Pacemaker Implantation)

Gabriel Gregoratos, Melvin D. Cheitlin, Alicia Conill, Andrew E. Epstein, Christopher Fellows, T. Bruce Ferguson, Jr, Roger A. Freedman, Mark A. Hlatky, Gerald V. Naccarelli, Sanjeev Saksena, Robert C. Schlant and Michael J. Silka

Indications for Permanent Pacing in Children and Adolescents

- 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

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Medications

Protect	Caution!	Breathe!
• Disease specific – LQTS – Brugada www.crediblemeds.org www.brugadadrugs.org	• Psychiatric medications • Energy Drinks	• ADHD medications

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ADHD Medications

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

The NEW ENGLAND JOURNAL of MEDICINE

ORIGINAL ARTICLE

ADHD Drugs and Serious Cardiovascular Events in Children and Young Adults

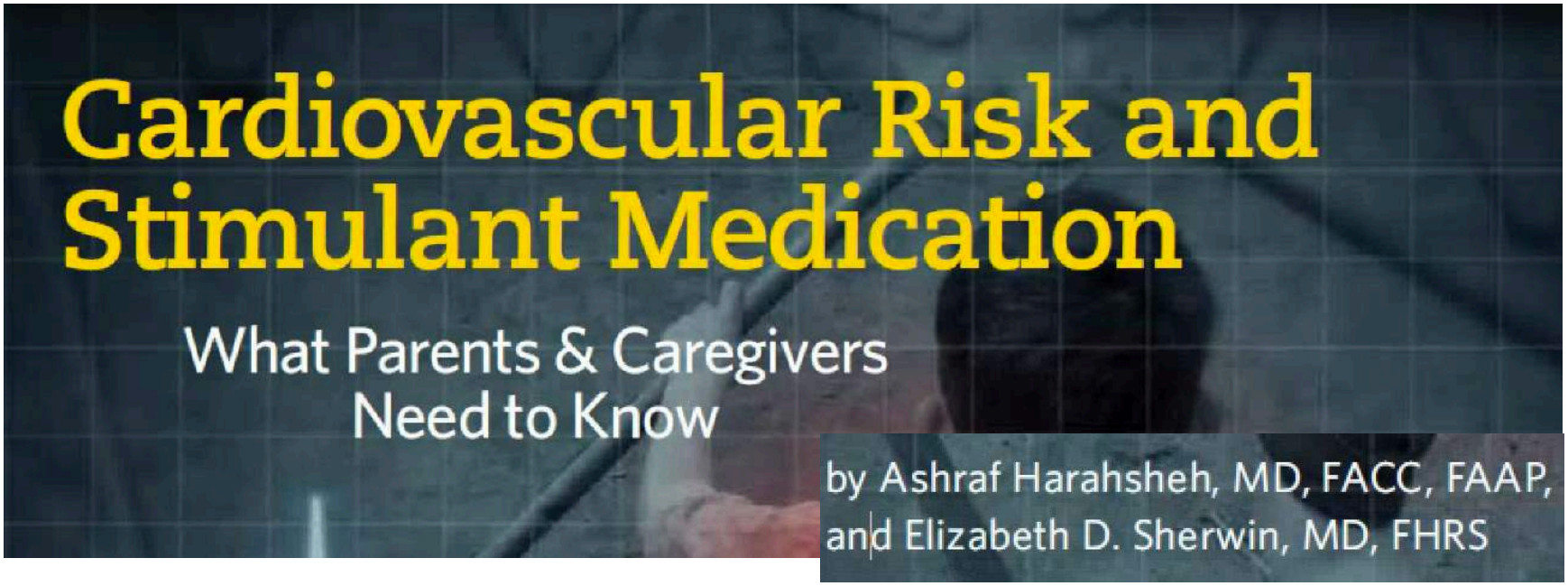
William O. Cooper, M.D., M.P.H., Laurel A. Habel, Ph.D., Colin M. Sox, M.D., K. Arnold Chan, M.D., Sc.D., Patrick G. Arbogast, Ph.D., T. Craig Cheetham, Pharm.D., Katherine T. Murray, M.D., Virginia P. Quinn, Ph.D., M.P.H., C. Michael Stein, M.B., Ch.B., S. Todd Callahan, M.D., M.P.H., Bruce H. Fireman, M.A., Frank A. Fish, M.D., Howard S. Kirshner, M.D., Anne O'Duffy, M.D., Frederick A. Connell, M.D., M.P.H., and Wayne A. Ray, Ph.D.

J Cardiovasc Electrophysiol. 2015 Oct; 26(10): 1045–1047.
Published online 2015 Aug 10. doi: 10.1111/jce.12744

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

Jonathan R. Kaltman, MD[1,2] and Charles I. Berul, MD[1]

National



- 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.

Attention magazine, copyright 2018 by CHADD, December 2018, vol. 25, no. 6, pages 13-15.

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

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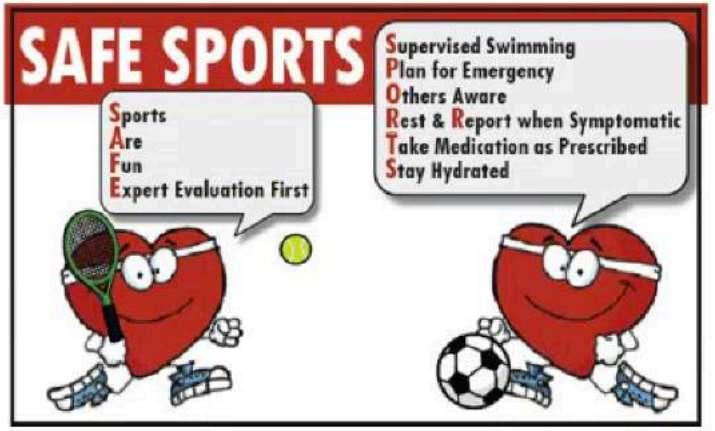
Safe Sports



Cheung *et al* CJC 2016

- 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

Arrhythmia Diagnosis & Management

- 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

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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

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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

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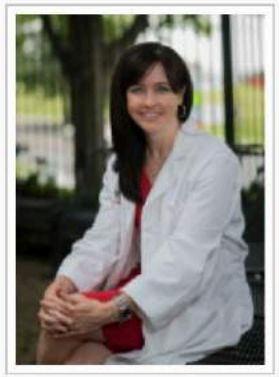
Children's National Electrophysiology Team

Charles Berul

Jeffrey Moak

Jonathan Kaltman



Elizabeth Sherwin

EP Nurses: Robin Fabian, Vicki Freedenberg, Carolyn Ramwell, Beth Jarosz

Not sure if you need to worry? Call us

THANK YOU

Elizabeth Sherwin, MD
edsherwin@childrensnational.org
www.ChildrensNational.org/HeartRhythm

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