



# EP Updates: ~~Bigger~~ Smaller and Better!

February 22, 2024

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Medical Director, Electrophysiology  
Lead, Cardiogenetics Inherited Arrhythmia Clinic

**Personal Disclosures: None**

**Talk Disclosures:**

**Brand name devices used**

**Though I love electrophysiology and always try to include too many things...just can't stop!**

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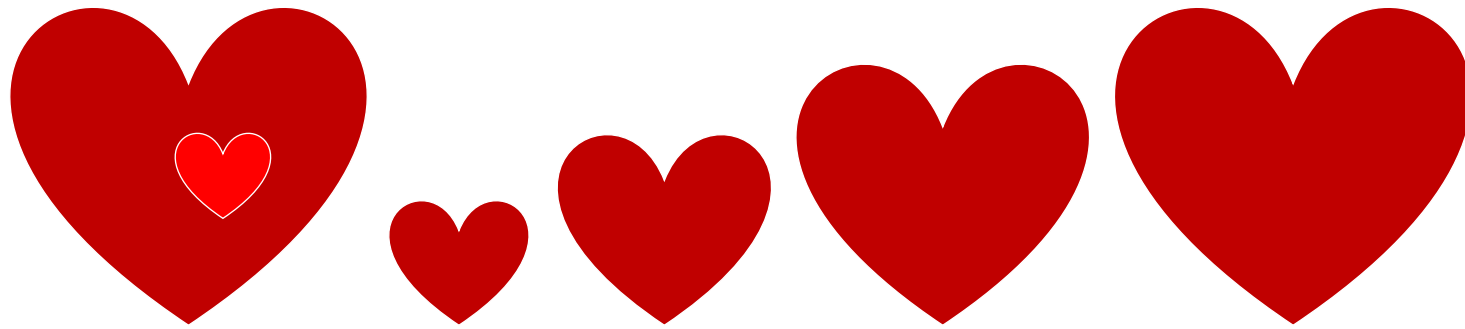


# Smaller and Better: Objectives

1. Review the spectrum of pediatric arrhythmia conditions
2. Understand advances in ablation therapy
3. Explore recent innovations in cardiac implantable electronic devices (CIED)

# **Pediatric Arrhythmias: A Brief Review**

# Pediatric and Congenital Arrhythmias



We manage arrhythmias from fetal life through adulthood!

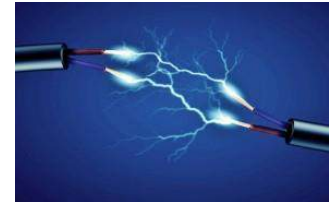
Cardiology clinics in 20 locations around DC, Maryland, Virginia

Electrophysiology clinics in Richmond, Fredericksburg, Fairfax, Arlington  
(as well as DC and Maryland)

# Buckets of Arrhythmias

## Sporadic

- Supraventricular tachycardia (SVT)
- Some ventricular tachycardia (VT)



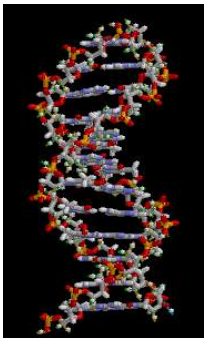
## Congenital heart defects

- Pressure or volume overload
- Late arrhythmias related to scars, suture lines



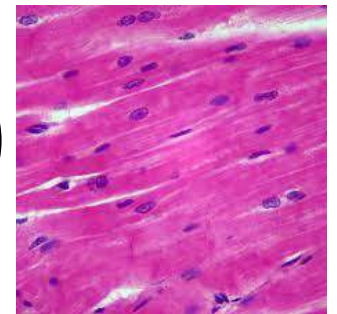
## Familial = inherited = genetic

- Long QT syndrome (LQTS)
- Brugada syndrome
- Catecholaminergic polymorphic ventricular tachycardia (CPVT)



## Muscle disorders (structural $\pm$ inherited conditions)

- Hypertrophic, arrhythmogenic, and other cardiomyopathies



## Other: Acquired, Infectious, Autoimmune

- COVID, myocarditis, Chagas, lupus

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

## Common Arrhythmias in Pediatrics

### Ectopy:

- Premature atrial contractions (PAC)
- Premature junctional contractions (PJC)
- Premature ventricular contractions (PVC)

### Tachyarrhythmias:

- **Supraventricular tachycardia (SVT)**
- Some ventricular arrhythmias: NSVT, Belhassen's VT

### Bradycardia:

- Sinus bradycardia, sinus pauses
- Heart block

# Most Worrisome Arrhythmias in Pediatrics

## **Incessant, unrecognized SVT**

→ *Heart failure due to tachycardia-induced cardiomyopathy*

## **Wolff-Parkinson-White (WPW) with rapidly conducting atrial fibrillation (“preexcited Afib”)**

→ *Sudden collapse (fainting, seizure), cardiac arrest/death*

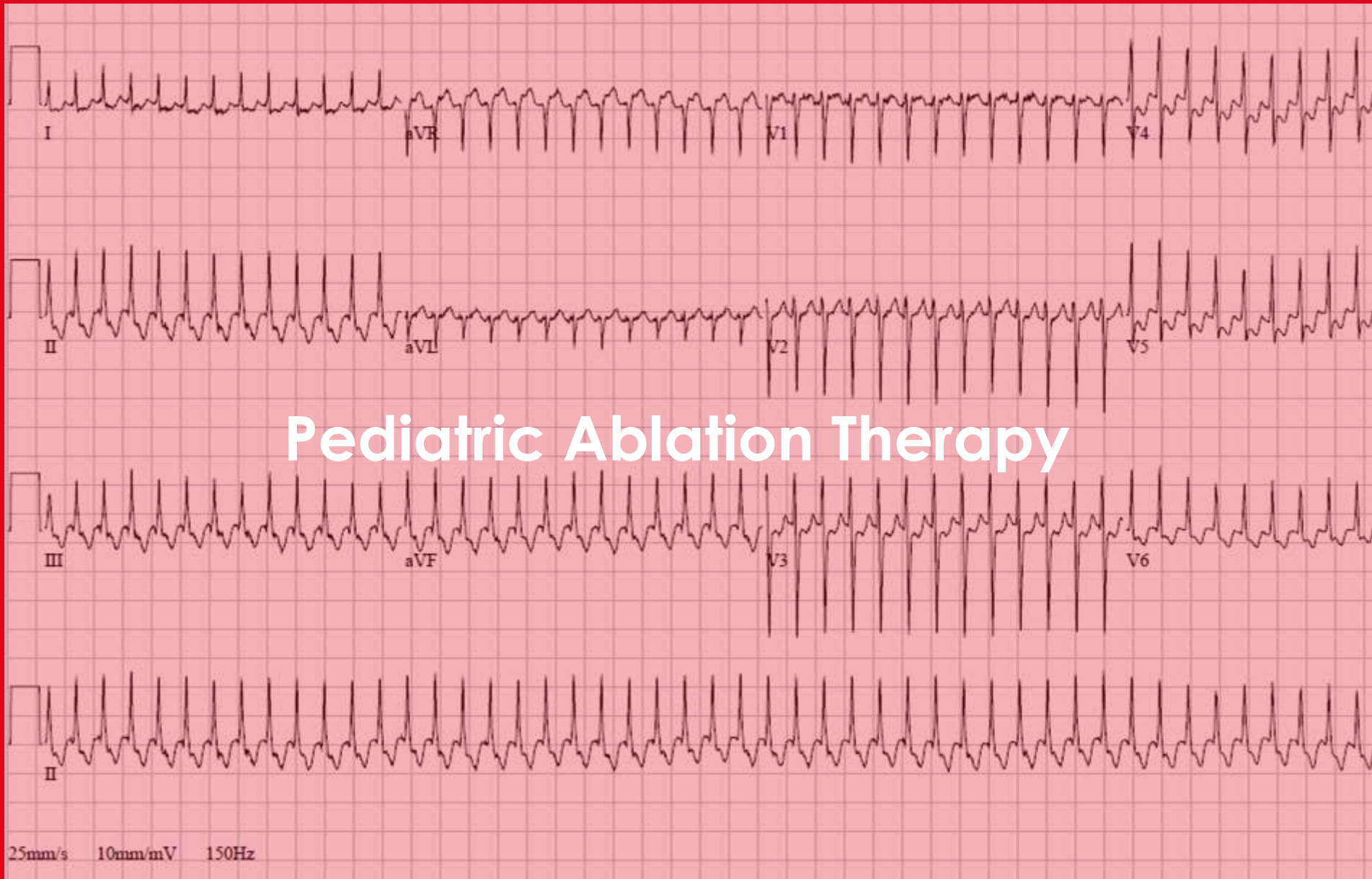
## **Polymorphic ventricular tachycardia (torsade de pointes)**

→ *Sudden collapse, cardiac arrest/death*



# Arrhythmia treatments

1. Observation
2. Antiarrhythmic medications
  - Pill-in-the-pocket
  - Preventative daily administration (one to three times per day)
3. Transcatheter ablation
  - SVT: success 92-98%
  - Atrial arrhythmias: success  $\geq 85\%$
  - Ventricular tachycardia: success ~80-90% acute success, with 15-20% recurrence
4. Cardiac devices: pacemakers, defibrillators
5. Surgical ablation with congenital heart disease
6. Cardiac sympathetic denervation



# SVT Ablation

- Catheter based procedure, typically performed under general anesthesia
- Developed in late 1980s in adult arrhythmias
- Pediatric RF (radiofrequency) ablation started in the early 1990s
- Cryoablation is freezing alternative to avoid AV node injury
- Nonfluoroscopic (no radiation) mapping and procedures evolved over the last 15 years
- Outpatient procedure ~2-4 hours, observation 4-6 hours post
- Recovery: 1-2 days out of school, 5 days off of sports to allow groin access sites to heal

## **PACES/HRS expert consensus statement on the use of catheter ablation in children and patients with congenital heart disease**

*Developed in partnership with the Pediatric and Congenital Electrophysiology Society (PACES) and the Heart Rhythm Society (HRS). Endorsed by the governing bodies of PACES, HRS, the American Academy of Pediatrics (AAP), the American Heart Association (AHA), and the Association for European Pediatric and Congenital Cardiology (AEPC)*

TASK FORCE CO-CHAIRS

Heart Rhythm June 2016

J. Philip Saul, MD, FHRS, FACC, FAHA, FAAP,<sup>1</sup> Ronald J. Kanter, MD, FHRS, FACC,<sup>2</sup>



Pediatric & Congenital Electrophysiology Society  
(PACES)

# Class I Indications for Ablation in Structurally Normal Hearts

## Narrow complex tachycardia, recurrent or persistent

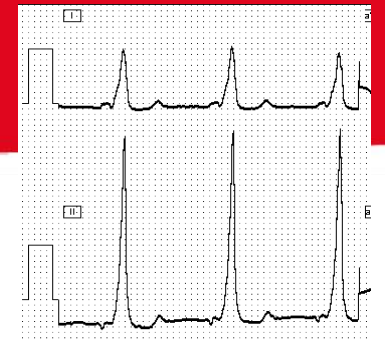
- With or without cardiac dysfunction
- Medications not effective or with intolerable side effects
- Family wishes to avoid chronic medications
- Unstable arrhythmias or requiring emergency medical care, electrical cardioversion

## WPW following cardiac arrest or syncope

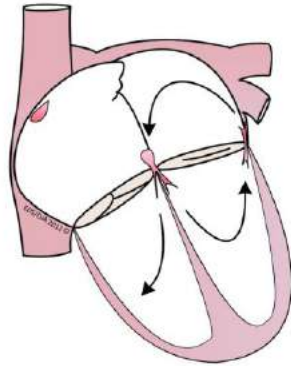
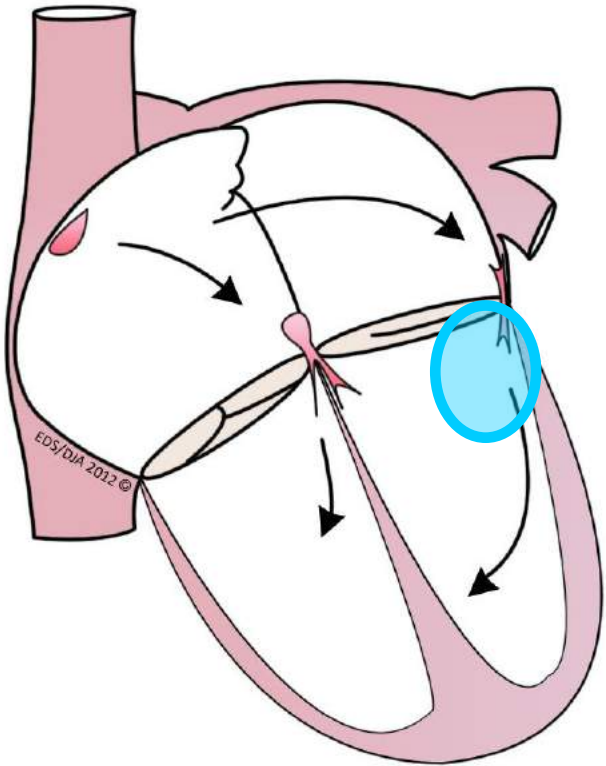
## Ventricular arrhythmias

- Frequent ectopy (PVCs) or VT *with ventricular dysfunction or hemodynamic compromise*
- Fascicular verapamil-sensitive reentrant VT, idiopathic outflow tract VT
- When medications are not effective, patient has intolerable side effects, or as alternative to medications

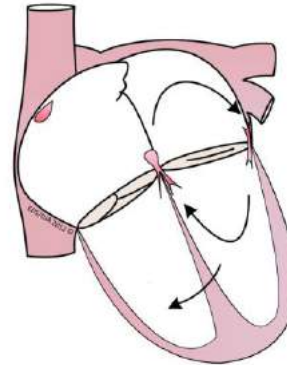
# A note on WPW...



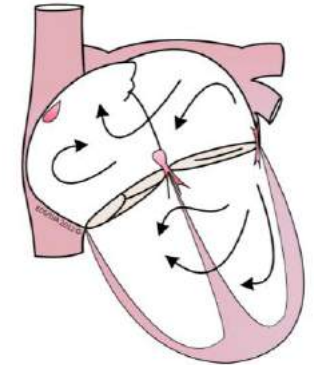
I have a very low *threshold* for referring any patient with WPW for EP study and ablation



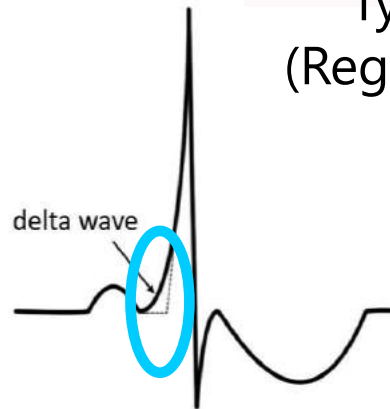
Typical SVT  
(Regular, narrow)



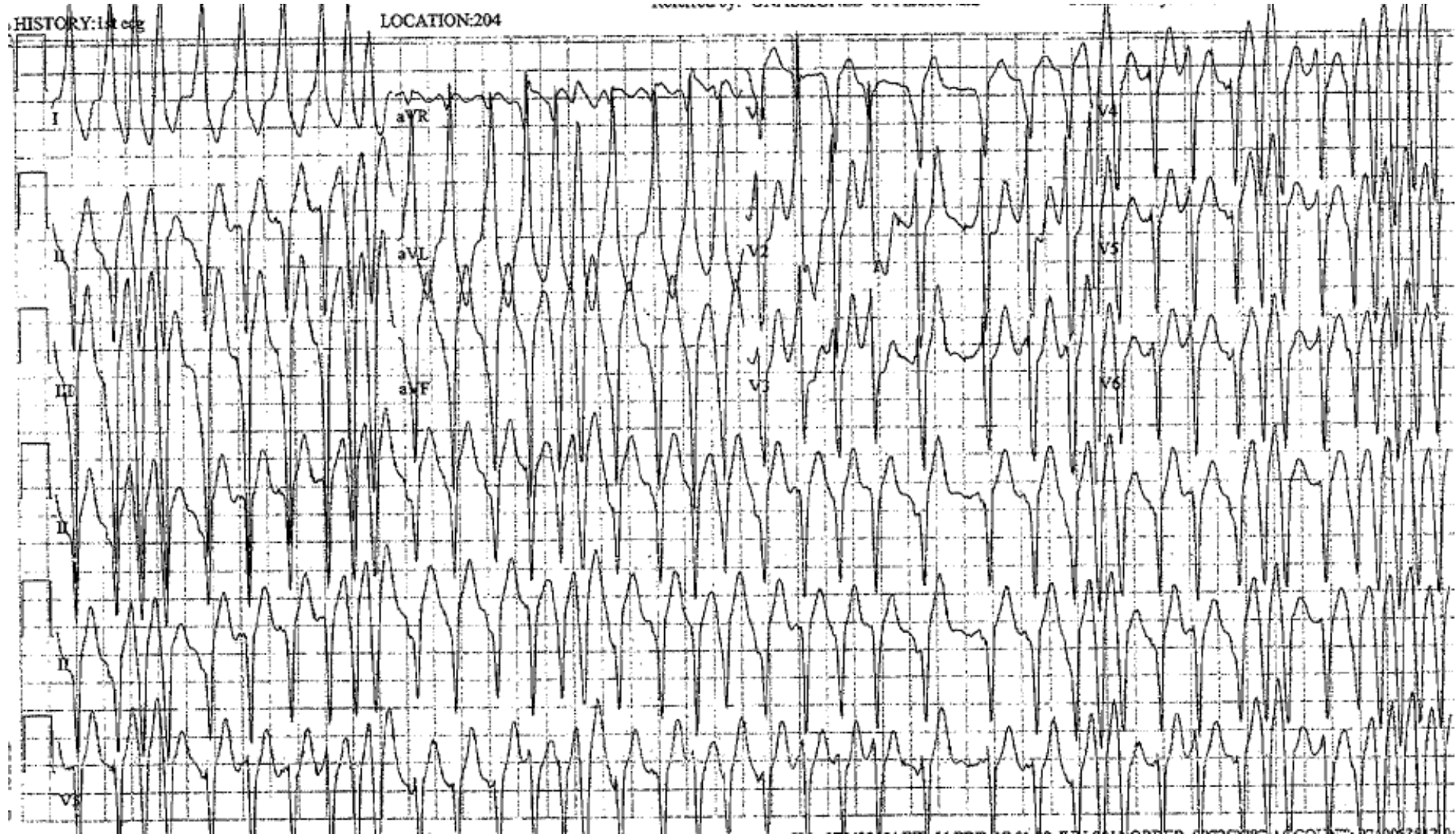
Antidromic SVT  
(Regular, wide)



Preexcited atrial  
fibrillation  
**(Irregular, wide)**

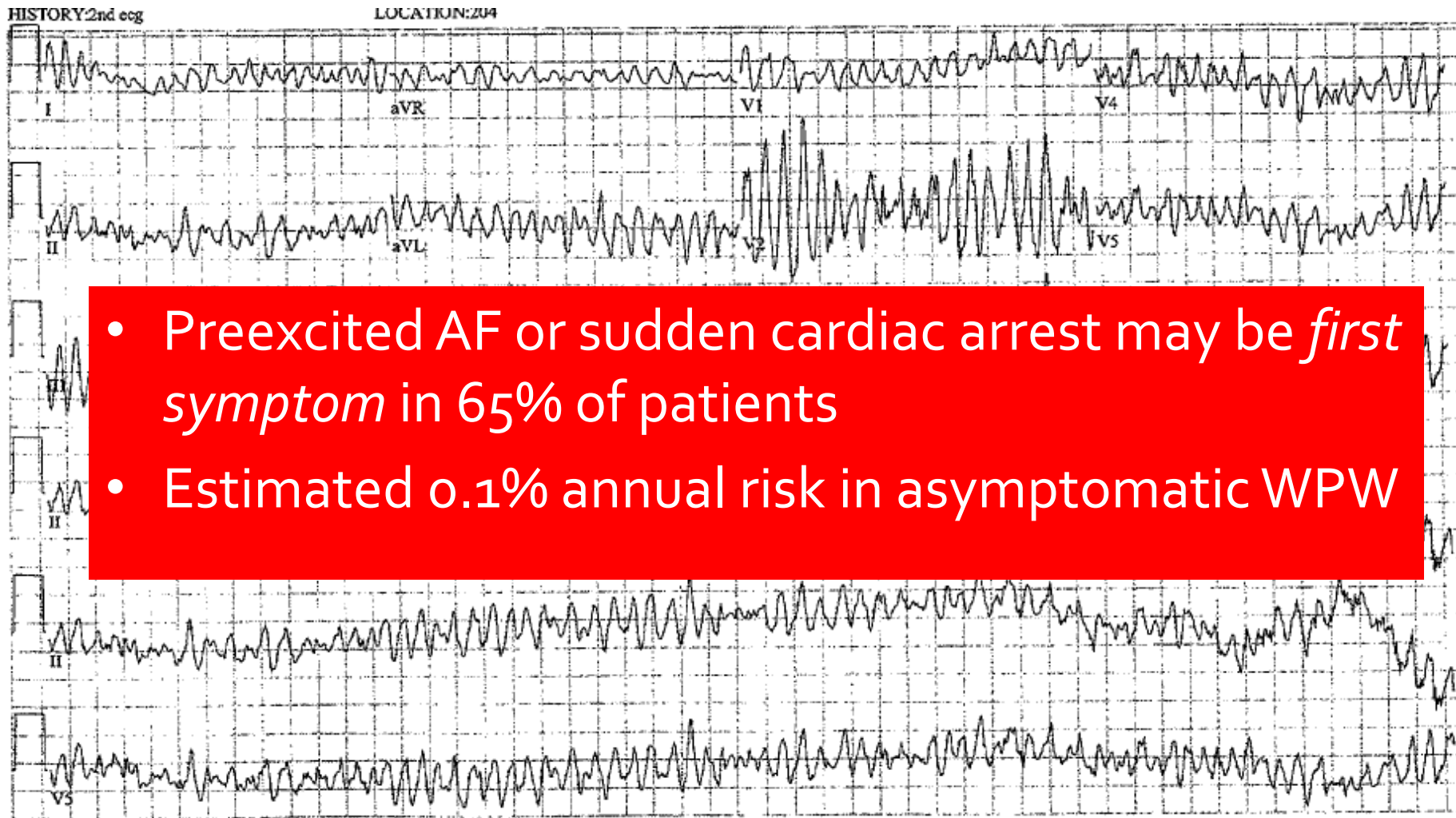


# Irregular wide complex tachycardia



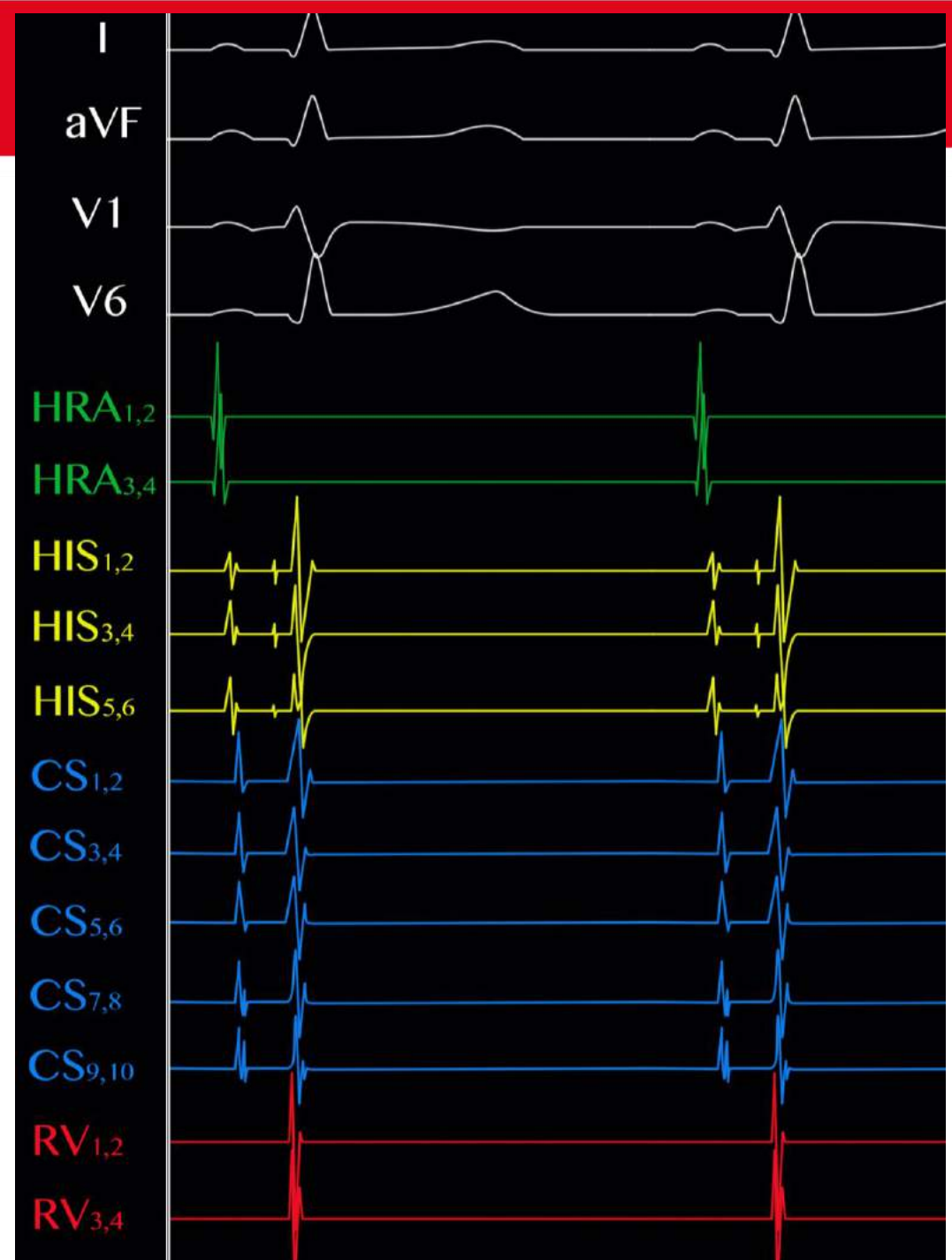
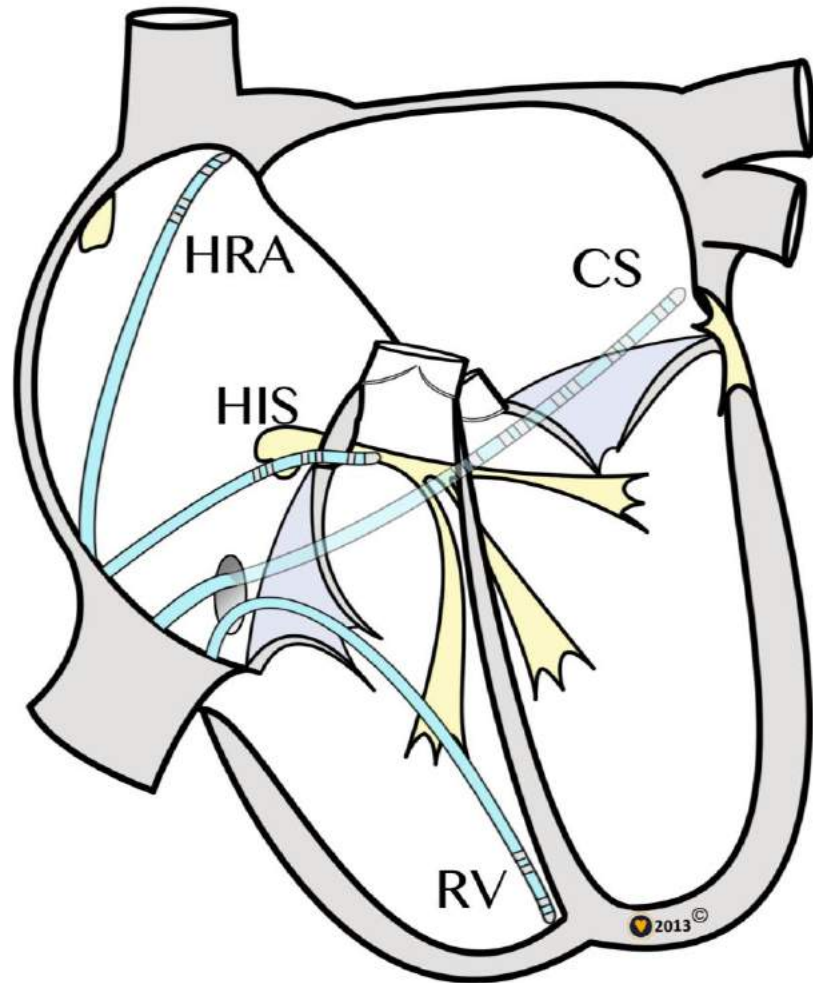
**Preexcited atrial fibrillation**

# Degeneration into ventricular fibrillation



**Mode of sudden cardiac arrest in WPW**

Catheters are placed in various positions inside the heart and produce electrograms







N/A	N/A	Loc	N/A	N/A
CL	LAT (ms)	Bi (mV)	Imp (C)	

200.0 mm/sec

2 sec

#	LAT	Tag	Type	Time	Comm
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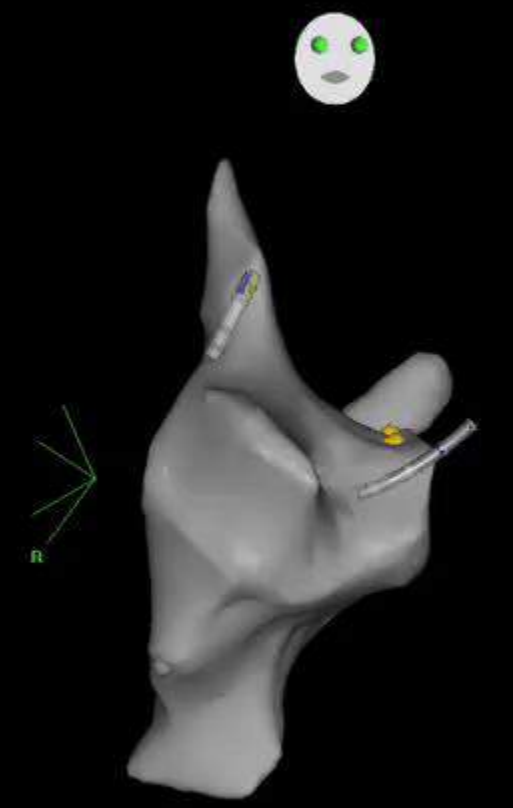
2-RA (7, 0)

LAT

1-IVC (0, 0)

LAT

Navigation and tool icons for the left panel, including a play button, a zoom slider, and various tool icons.

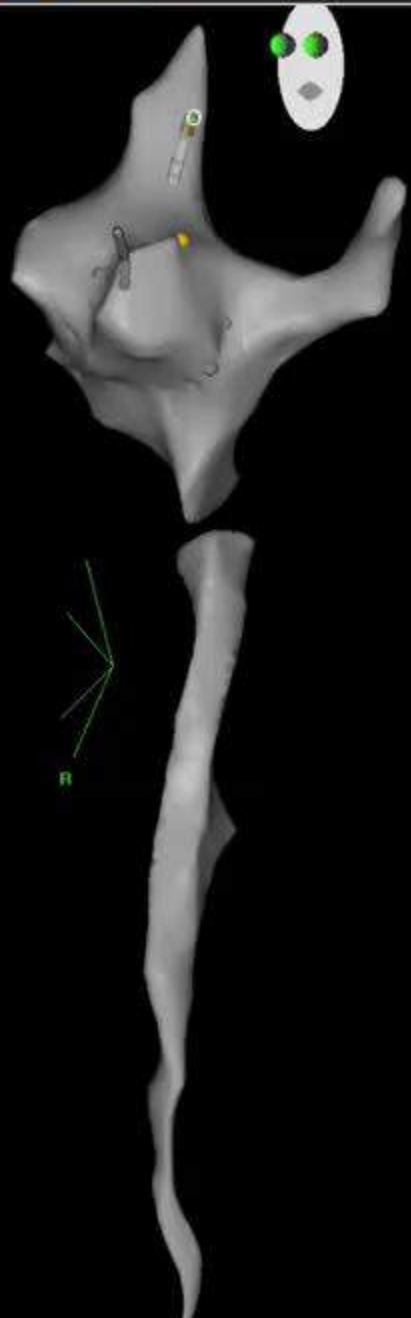


RAO view control panel with a heart icon and a vertical slider. The slider is set to 1.60.

Acquire

CL	LAT	Bi	Imp
837	-130	0.71	N/A

AP PA LAO RAO LL RL INF SUP

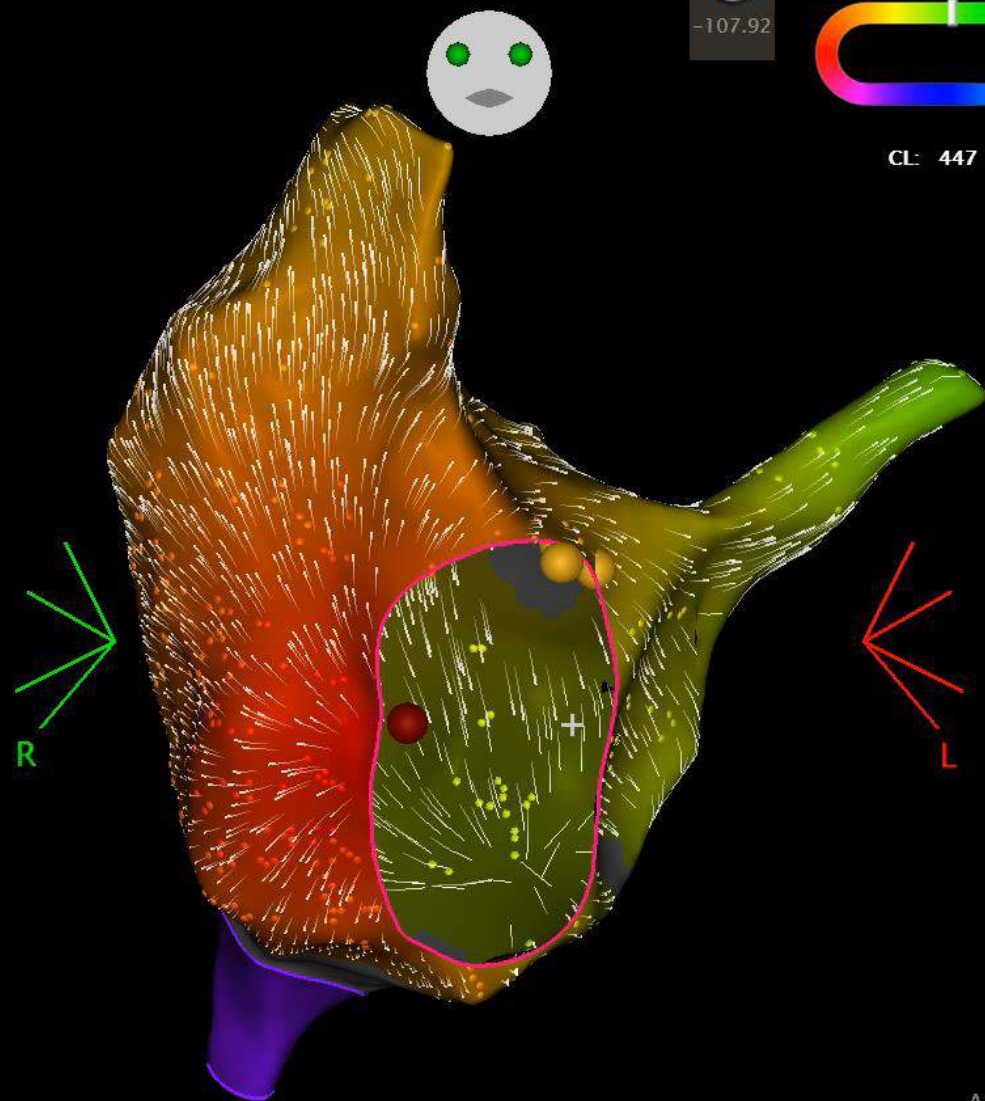


LAO view control panel with a heart icon and a vertical slider. The slider is set to 2.94.

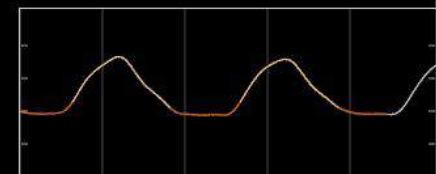
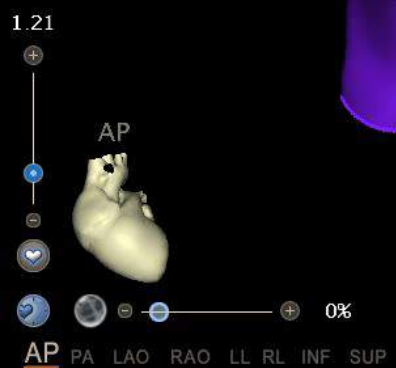
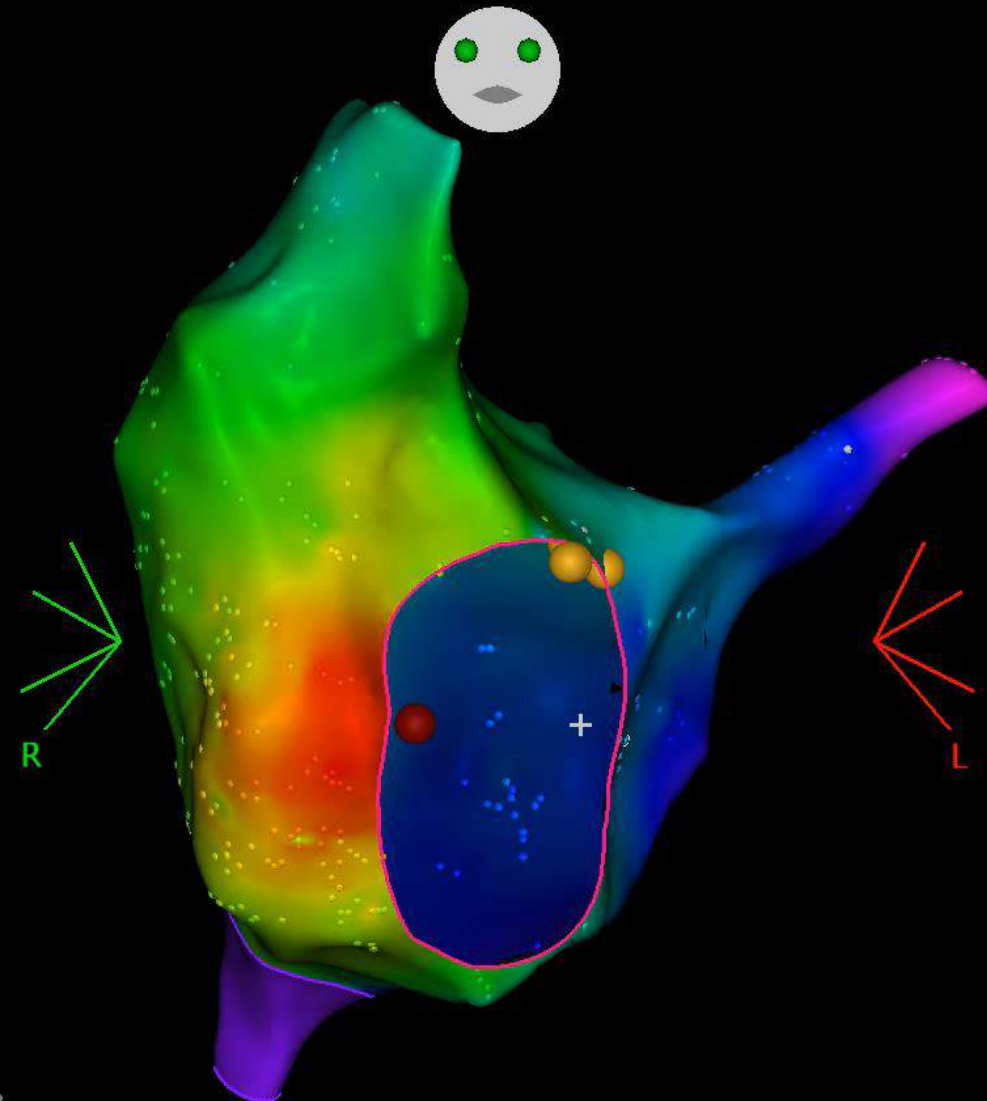
AP PA LAO RAO LL RL INF SUP

None

1-RAi (598, 0) Resp



1-RAi (598, 0) Resp



# When to consider ablation

- Hemodynamically unstable arrhythmias
- Significant arrhythmia burden and symptoms
  - Interfering with life
  - Resulting in ventricular dysfunction
- Arrhythmias refractory to medications
- Patient adverse reaction to medication, or aversion to chronic medications
- Possible job disqualification:
  - Pilot, police, military
- Wolff Parkinson White syndrome
  - Sudden cardiac arrest, preexcited atrial fibrillation, or suspicious syncope
  - High risk WPW pathway
  - Any patient with WPW.....

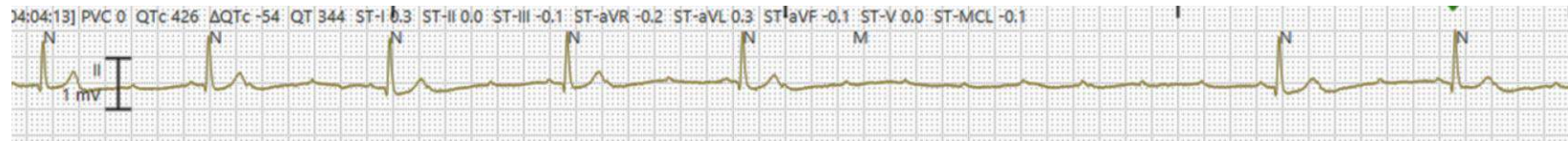


**Can be first line of therapy!**  
**Elective procedures in patients > 15-20 kg**  
**Urgent indication: any age/size**

# Parasympathetic Ablation



- Syncope may be due to transient sinus pause or transient complete heart block
  - Mediated by excessive parasympathetic activation



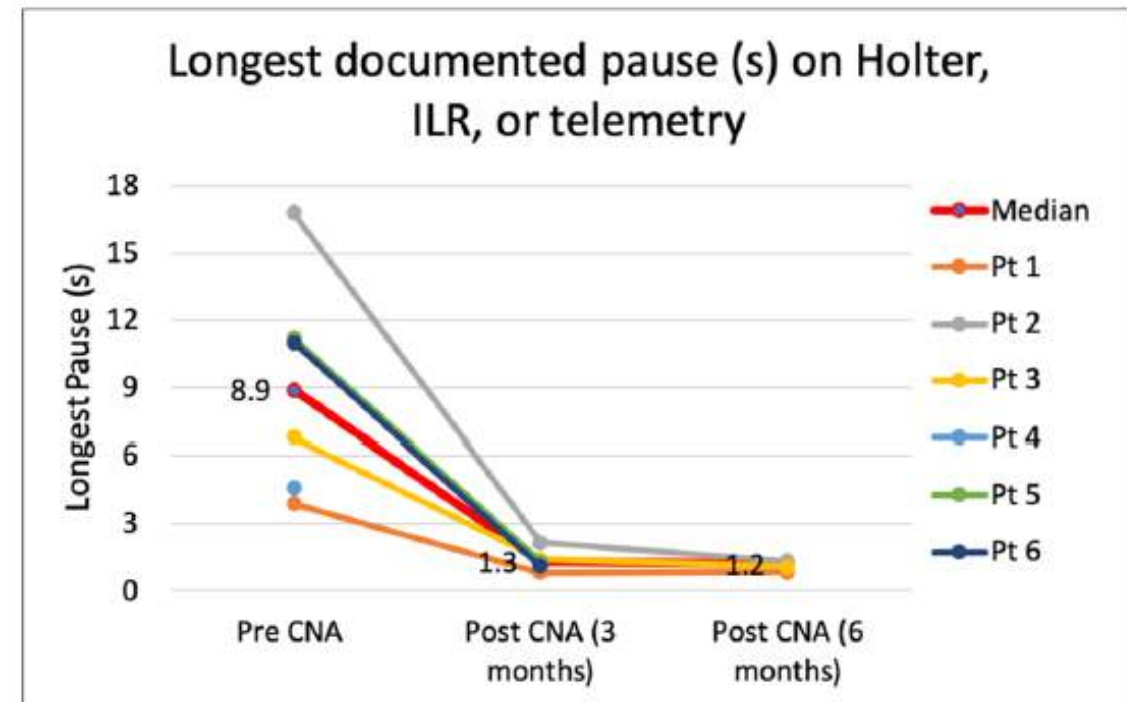
- Standard treatments
  - Vasovagal treatments with lifestyle modifications: fluids, avoiding triggers, salt and electrolytes, exercise
  - Medications: fludrocortisone, midodrine; glycopyrrolate for pauses
  - Definitive treatment: permanent pacemakers
- Evolving alternative: cardioneuroablation
  - Well-described (yet not commonly done) in adult electrophysiology for prevention of bradycardia-induced atrial fibrillation, syncope
  - Parasympathetic fat pad ablation

# Cardioneuroablation for pediatric patients with functional sinus node dysfunction and paroxysmal atrioventricular block

Nak Hyun Choi MD<sup>1,2</sup> | Jeff Hong MD<sup>1</sup> | Jeffrey P. Moak MD<sup>1</sup> November 2023

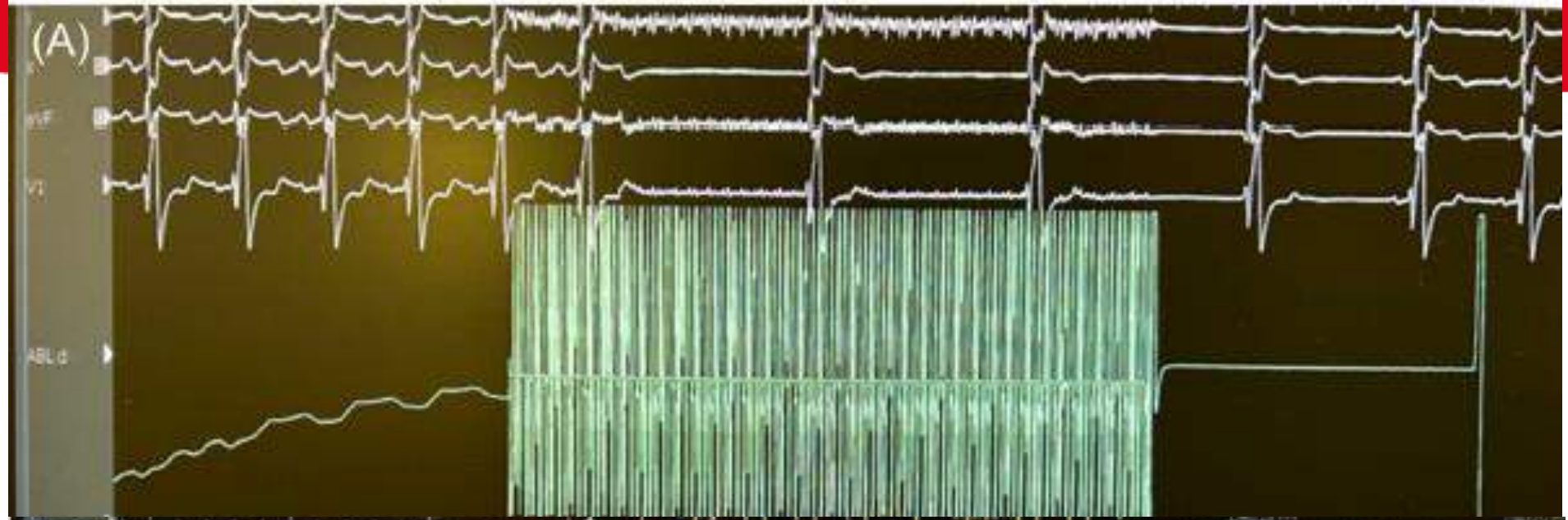
Selectively ablate the epicardial ganglionated plexi (GP) from the endocardial surface at the fat pad sites where the parasympathetic innervation enters the heart

- Six patients, median age 18.9 years (range 12.3–20.9 years)
  - 2 patients had prolonged sinus pauses (median 8.9 seconds)
  - 2 had paroxysmal AV block
  - 2 had both pauses and brief AV block
  - 4 patients had syncope
- No patients had syncope post-CNA.

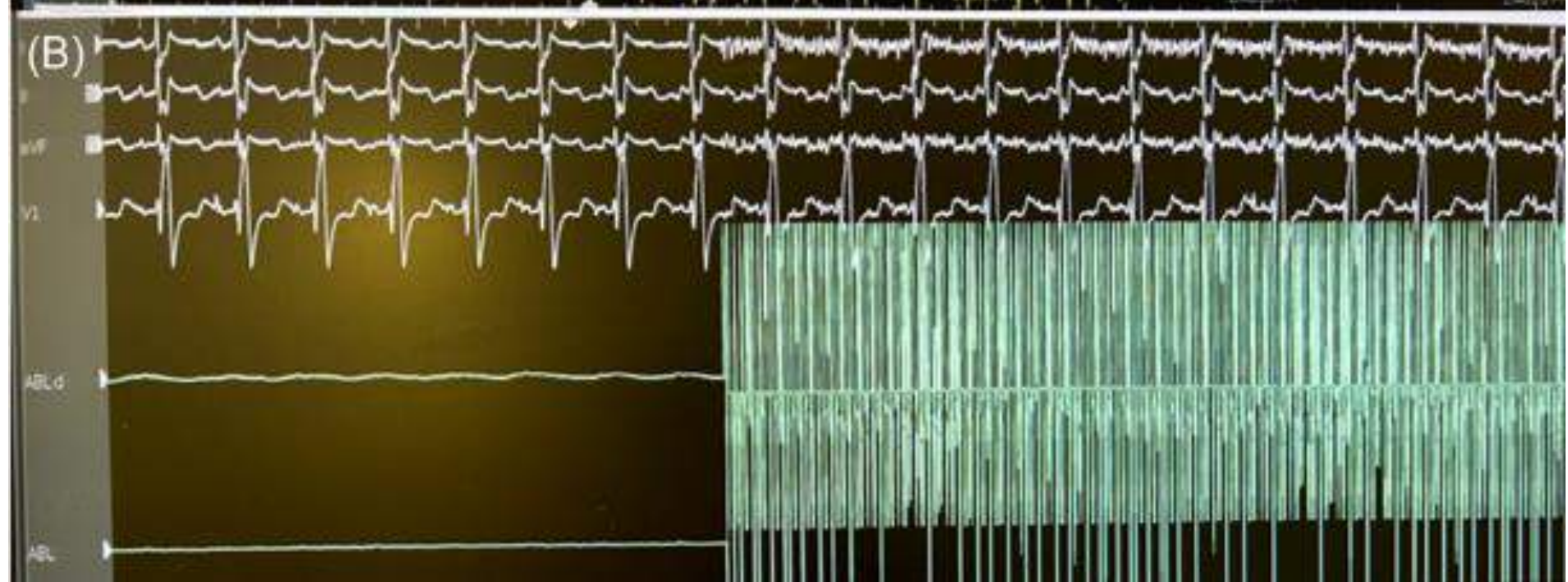


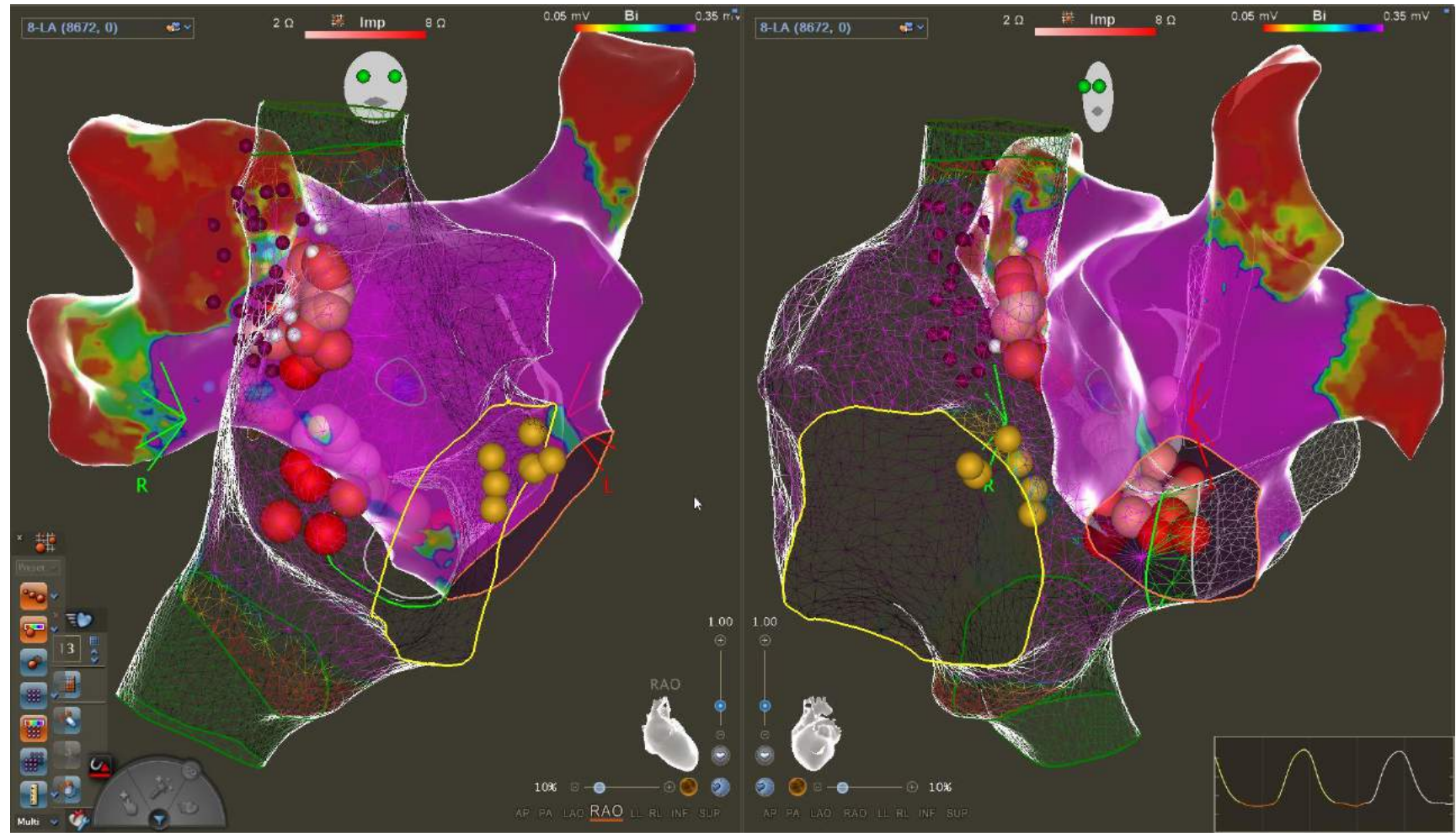
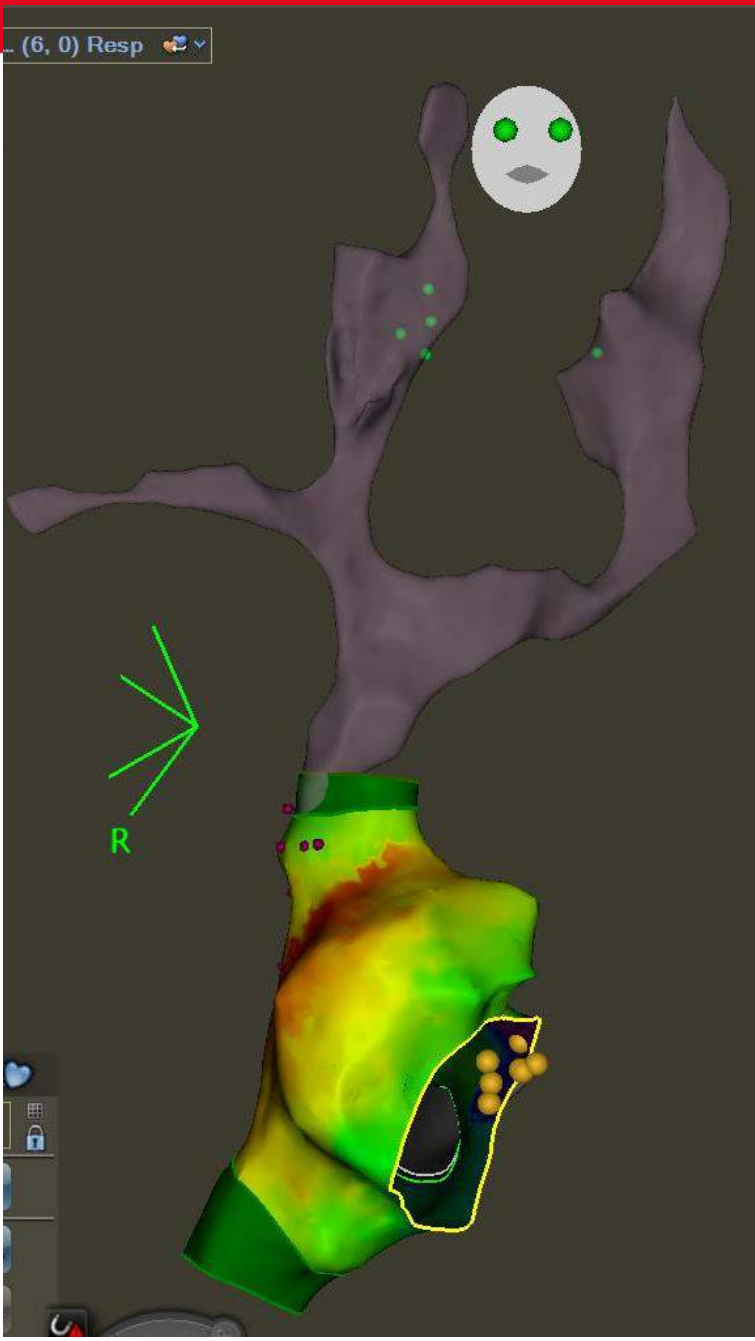
**FIGURE 5** Longest documented pause(s) on Holter monitor, implantable loop recorder, or telemetry pre- and postcardioneuroablation. ILR, implanted loop recorder.

**Pre-ablation**



**Post-ablation**



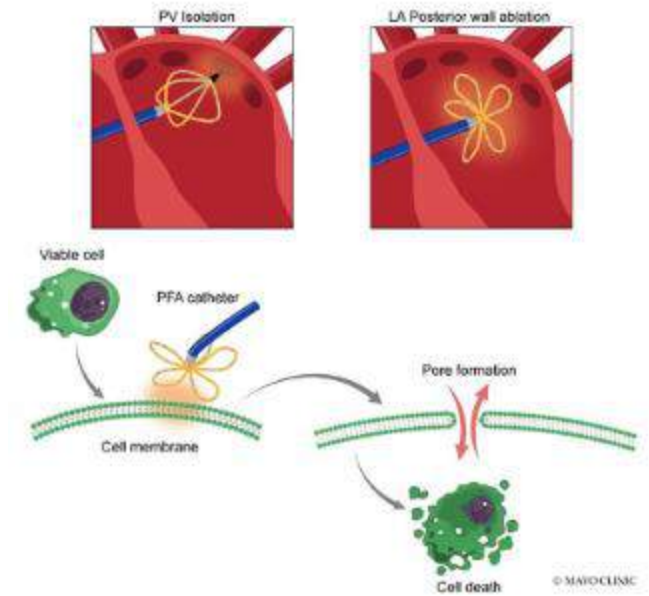






# New kid on the block: Pulse Field Ablation (PFA)

- Microsecond high-voltage electrical fields
- Irreversible electroporation → destabilizes cell membranes → cellular necrosis
- Goal is to eliminate arrhythmias while minimizing damage to tissue outside of the targeted myocardium
- Increasingly used for atrial fibrillation ablation since first clinical approval in 2021






## Pulsed Field or Conventional Thermal Ablation for Paroxysmal Atrial Fibrillation

Vivek Y. Reddy, M.D., Edward P. Gerstenfeld, M.D., Andrea Natale, M.D., William Whang, M.D., Frank A. Cuoco, M.D., Chinmay Patel, M.D., Stavros E. Mountantonakis, M.D., Douglas N. Gibson, M.D., John D. Harding, M.D., Christopher R. Ellis, M.D., Kenneth A. Ellenbogen, M.D., David B. DeLurgio, M.D., et al., for the ADVENT Investigators\*

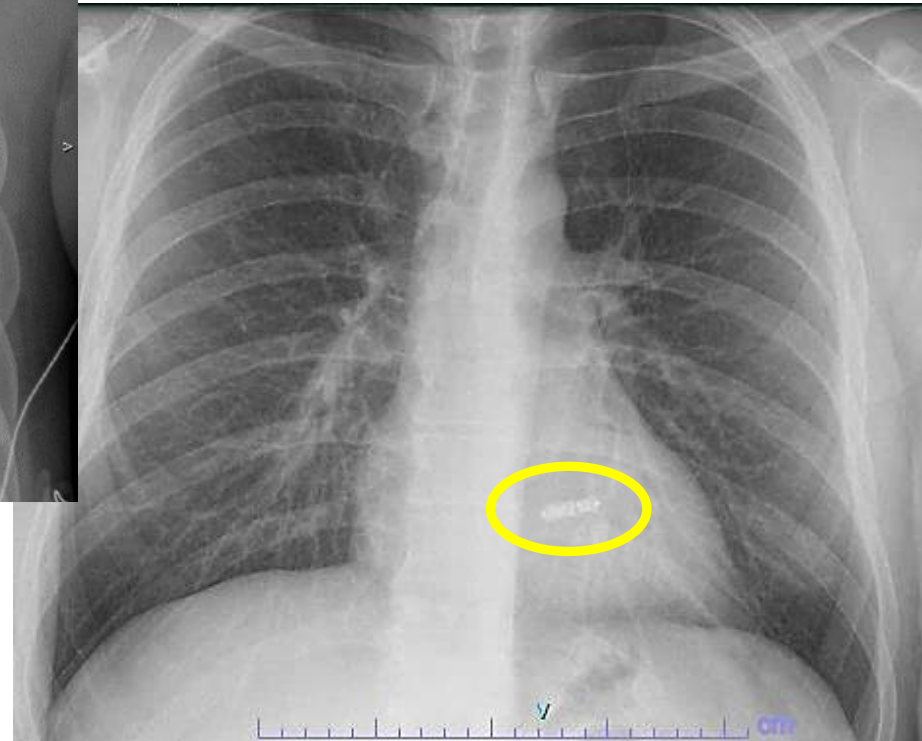
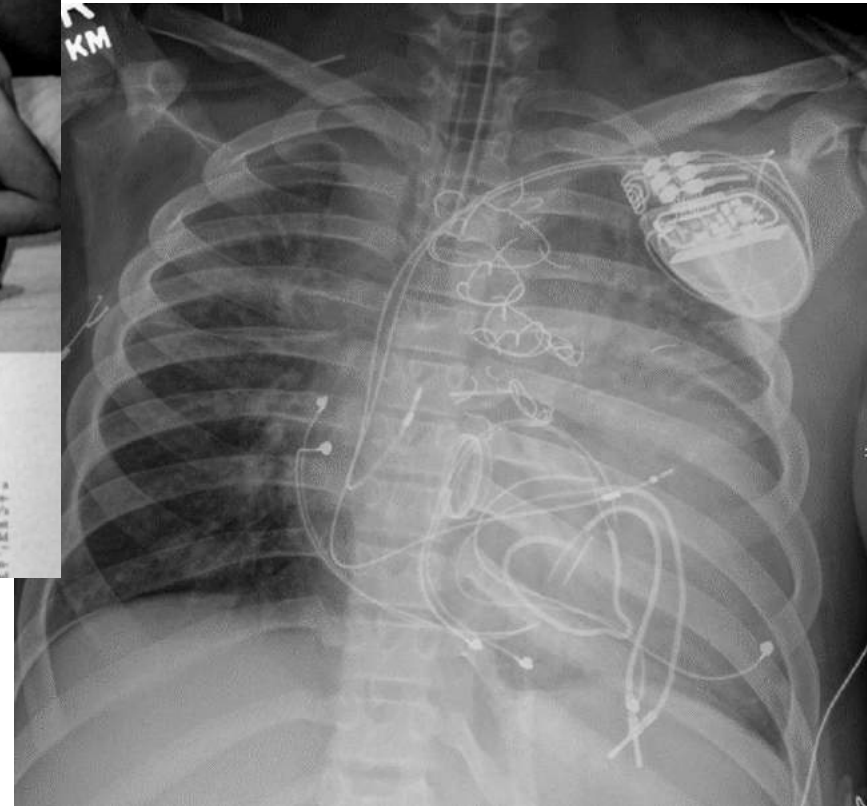
Front. Cardiovasc. Med.. 23 October 2023

## The promise of pulsed field ablation and the challenges ahead

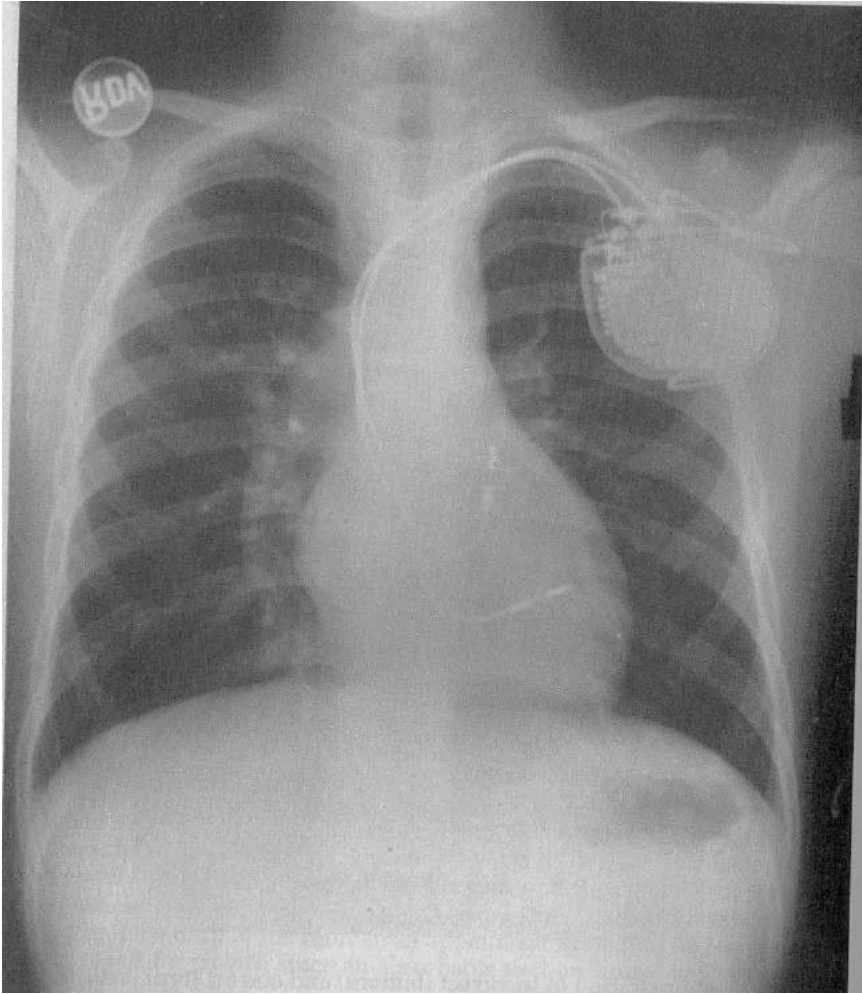
 Shruti Krishna Iyengar  Sumedh Iyengar  Komandoor Srivathsan\*

# **Cardiac Implantable Electronic Device (CIED) Advances**

# Evolution in Pacing

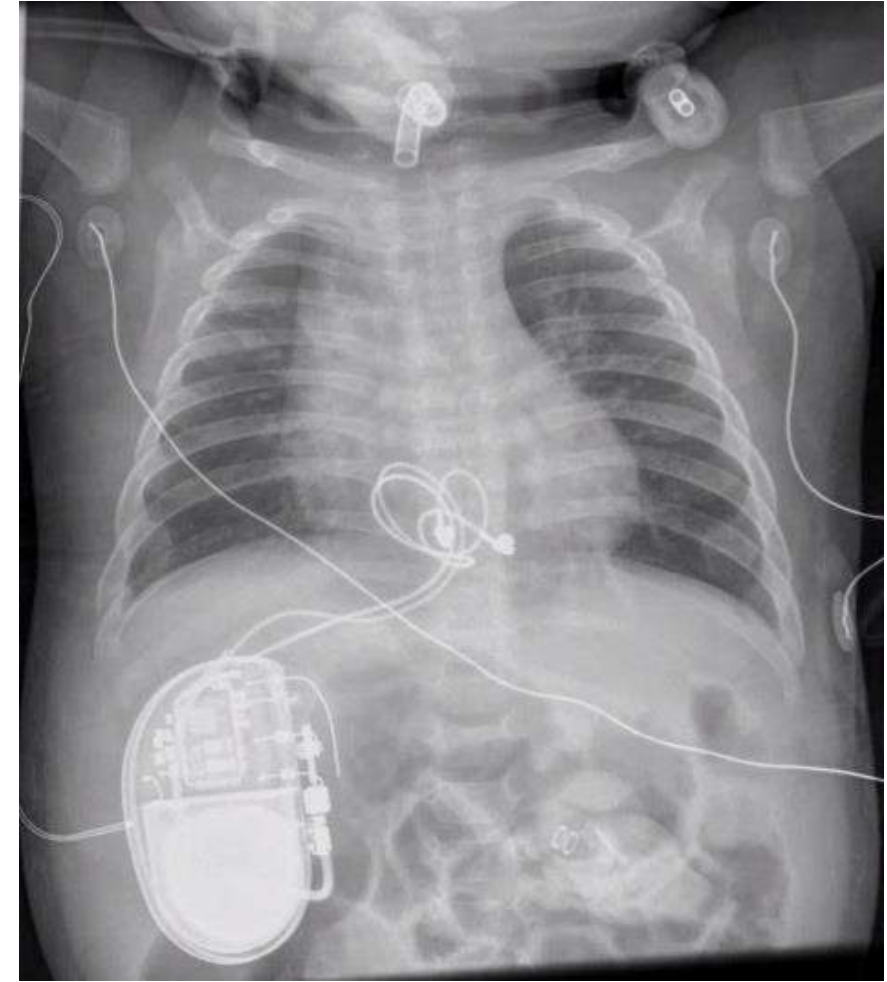


## Transvenous Pacemakers

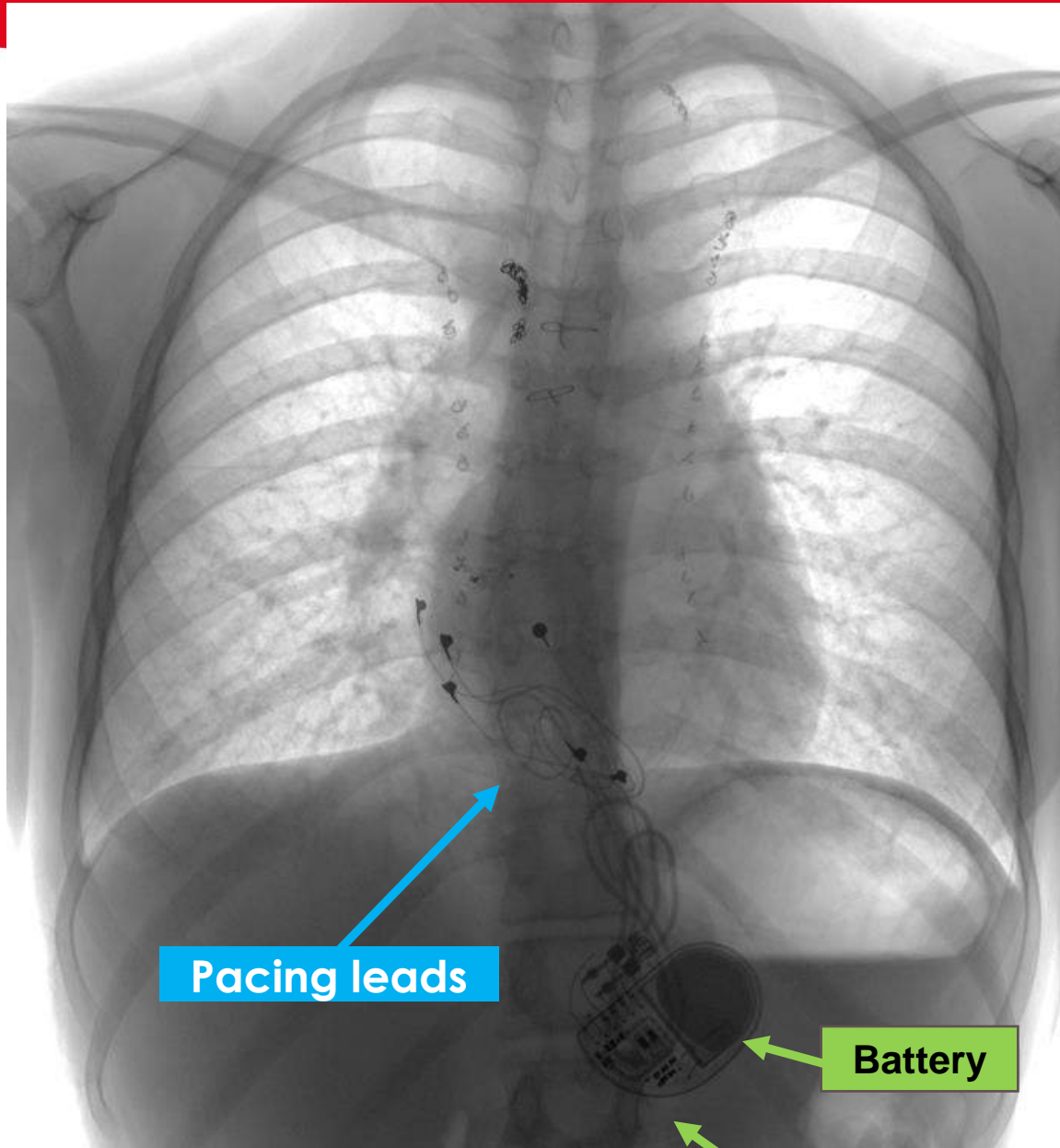


- Adults
- Teenagers

## Epicardial Pacemakers



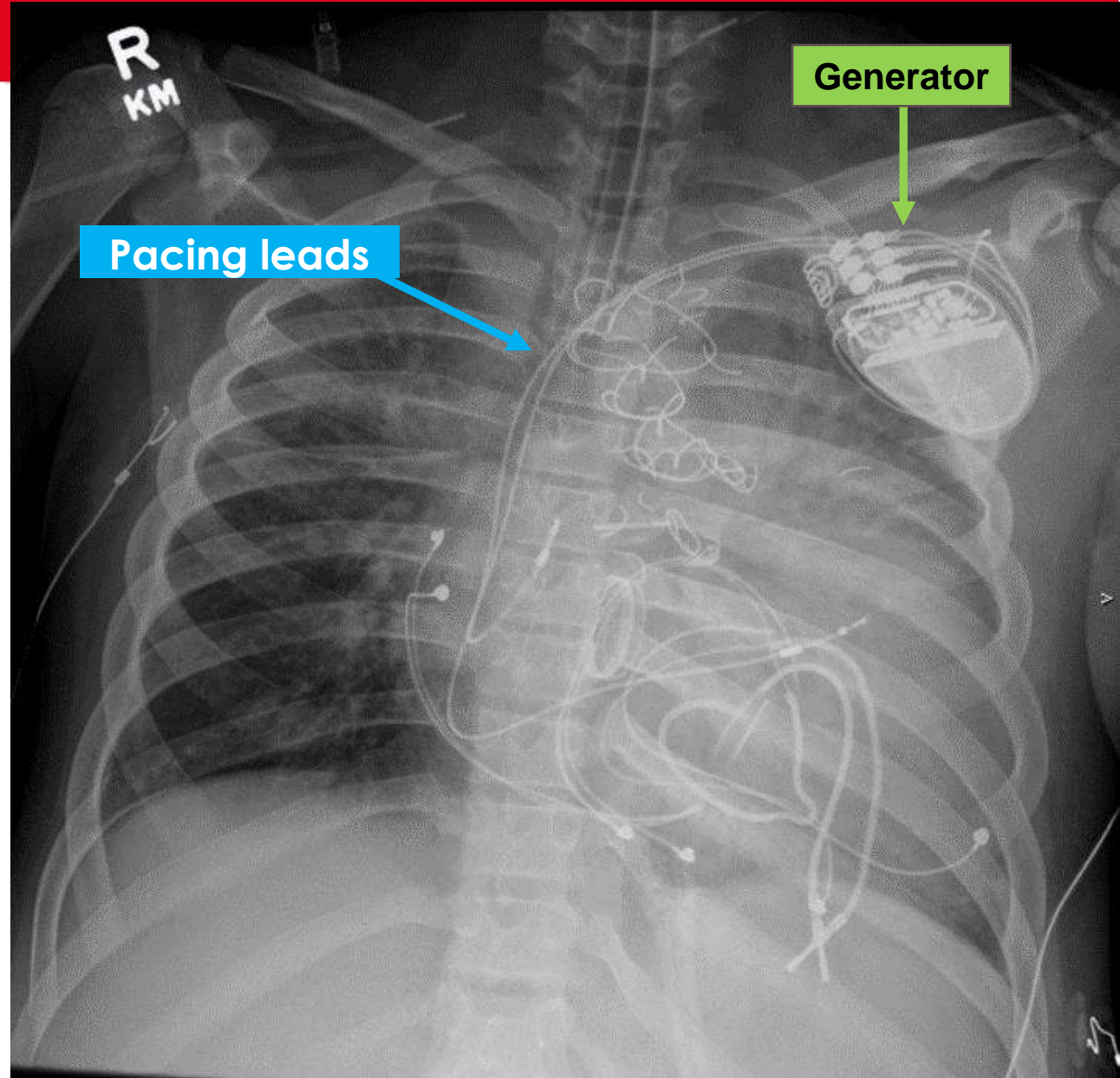
- Infants, younger kids
- Single ventricle anatomy



Pacing leads

Battery

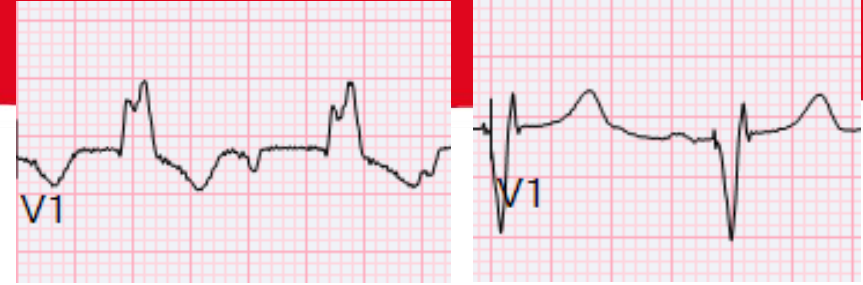
Circuitry



Pacing leads

Generator

# General indications for pacing



## Bradycardia

- Sinus node dysfunction
  - Pauses
- Accelerated junctional rhythm (Fontan)
- Heart block (high grade, complete)

*\*Significant and/or symptomatic*

## Ventricular dysfunction

- Cardiac resynchronization therapy (CRT)  
= biventricular pacing
  - Conduction system pacing  
= pacing in the ventricular septum to activate the His-Purkinje system (i.e. pace the left bundle branches)

## Special circumstances

- Incessant arrhythmia (“storms”)
- Severe LQTS with bradycardia or irregular sinus rate

# Choosing the type of pacemaker

- Anticipated need: short term or long term?
- What type of pacing is needed?
  - Atrial only? (Ex: slow sinus rates)
  - Ventricular only? (Ex: occasional pauses or improved heart block)
- How much pacing is needed?
  - 100% of the time? Rarely?
- How old/big is the patient?
  - Infant vs. grown child or adult
- What is the anatomy?
  - Single ventricle? Venous occlusions? Stenotic or artificial valves?

## Location

- Epicardial
- Transvenous

## Number of Leads

- One (AAI, VVI)
- Two (DDD)
- Three (CRT)

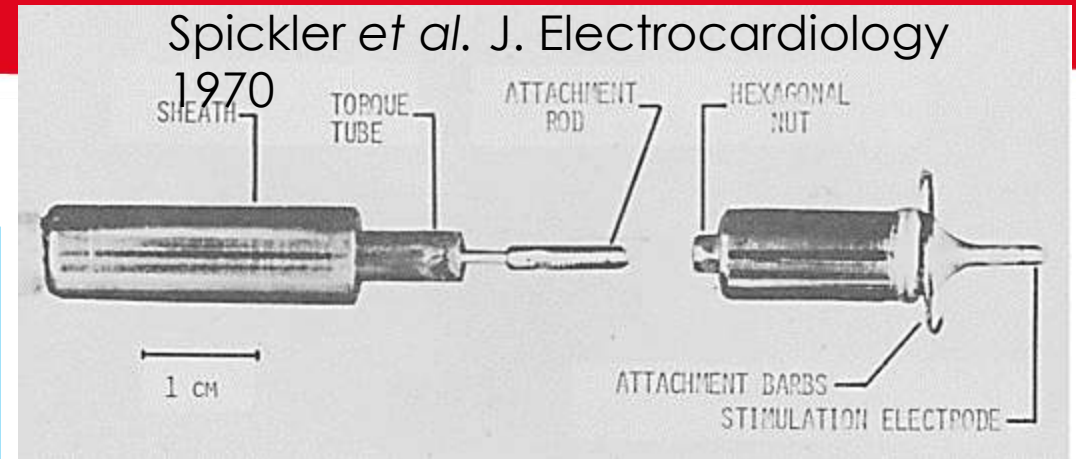
## Leadless!



# Leadless Pacing

## Conceptualized by 1970

- Animal model
- Limited battery technology



## First FDA approved TLP 2016: Micra™

- Ventricular pacing only



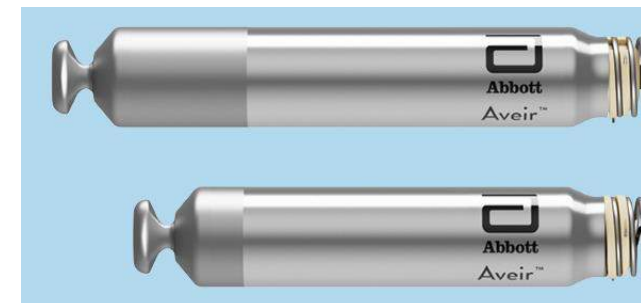
## Atrial mechanical sensing 2020:

### Micra™ AV

- VDD pacing for atrioventricular synchrony

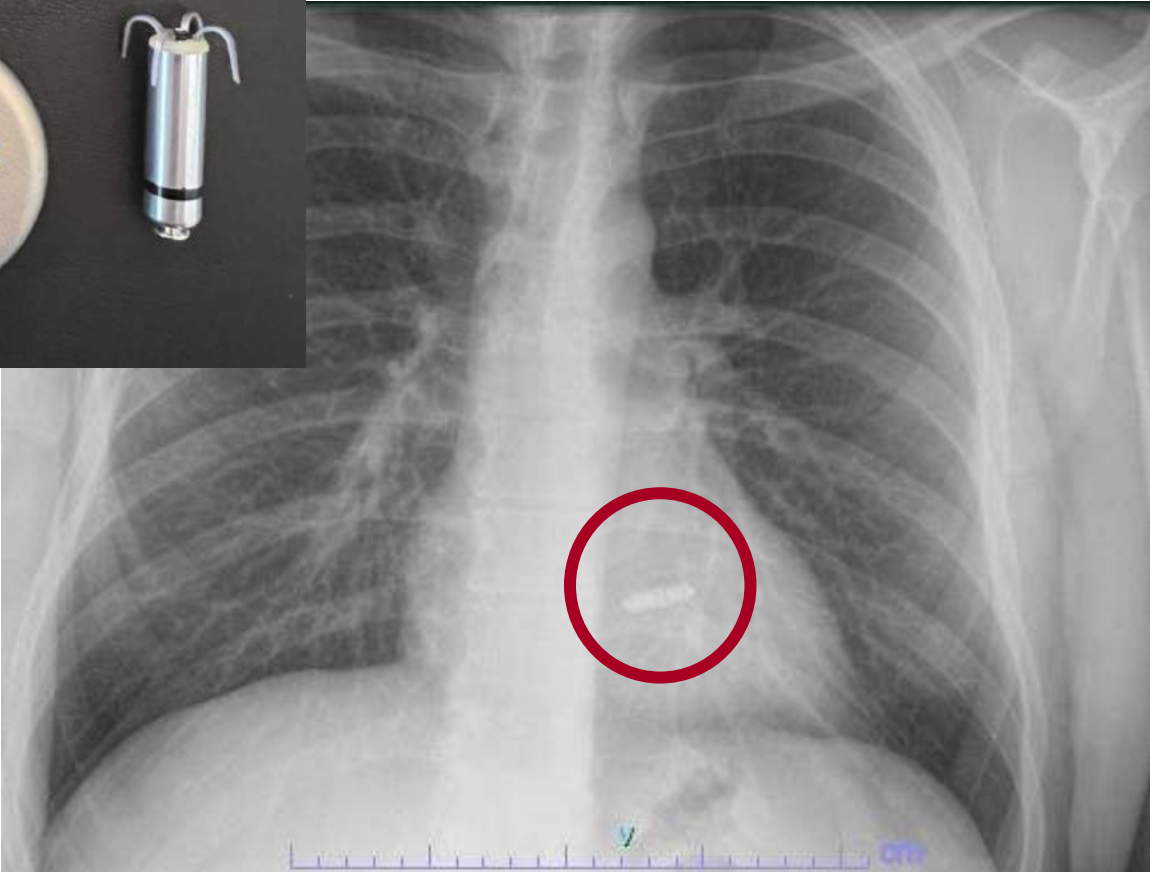
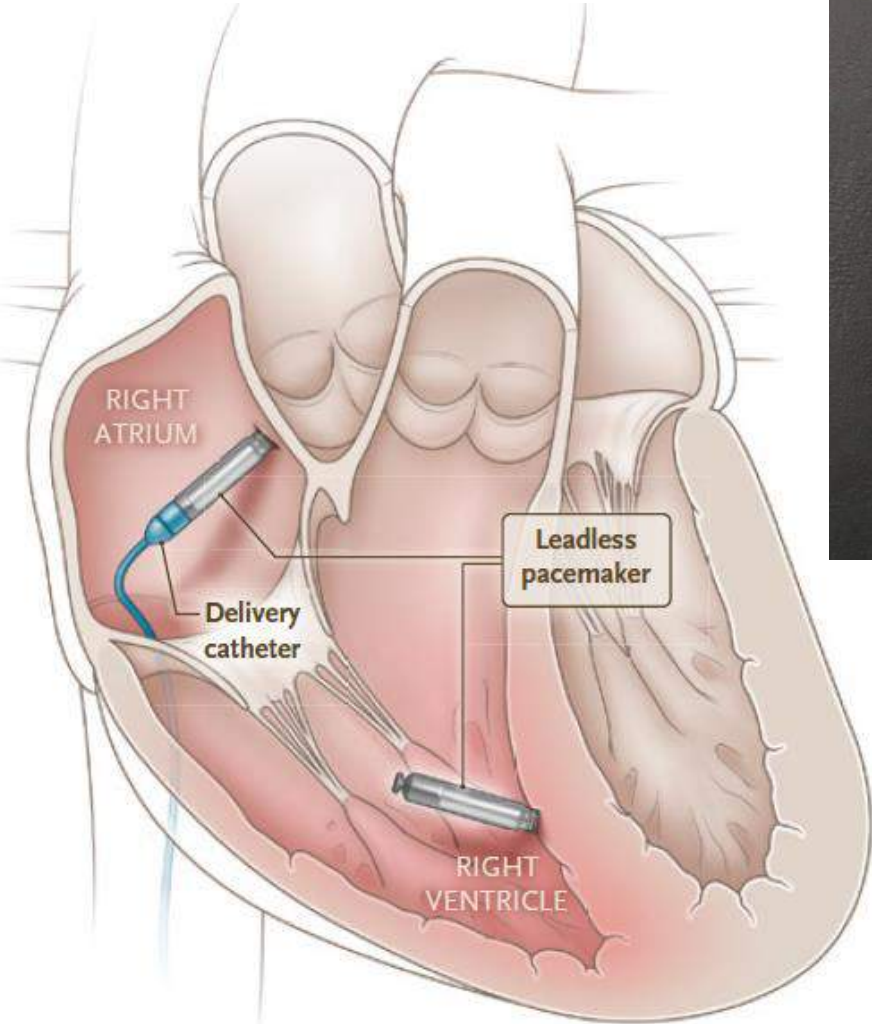
## Now FDA approved 10/2023: Aveir™ DR

- Dual chamber leadless pacing





# Leadless pacing





## CREATIVE CONCEPTS

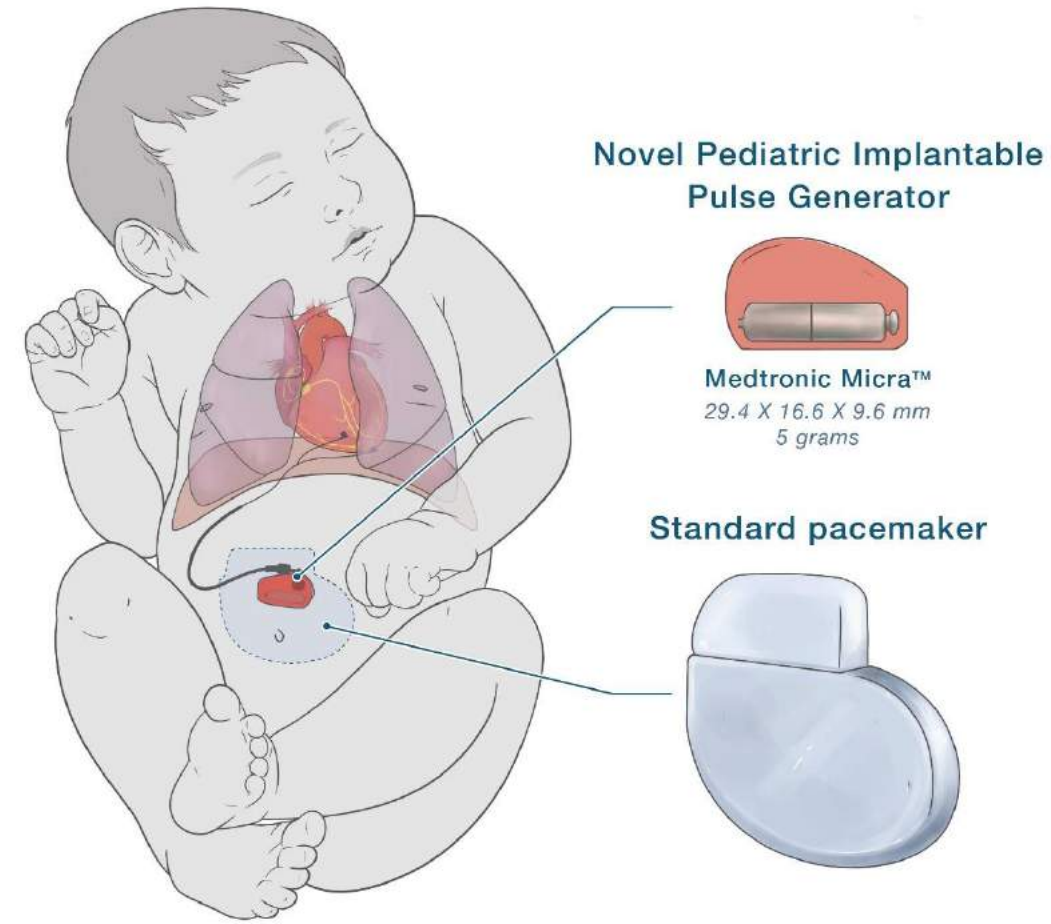
# Tiny pacemakers for tiny babies

Charles I. Berul, MD, FHRS,<sup>\*</sup> Soham Dasgupta, MD,<sup>†</sup> Marc D. LeGras, MD,<sup>‡</sup>  
S. Murfad Peer, MD,<sup>§</sup> Bahaaldin Alsoufi, MD,<sup>†</sup> Elizabeth D. Sherwin, MD,<sup>\*</sup>  
Manan Desai, MD,<sup>\*</sup> Can Yerebakan, MD,<sup>\*</sup> Christopher Johnsrude, MD<sup>†</sup>

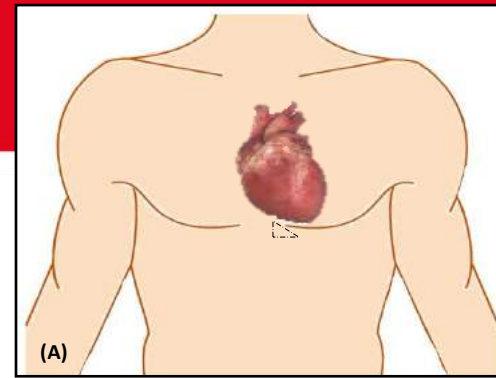
*From the <sup>\*</sup>Children's National Hospital, George Washington University School of Medicine, Washington, DC, <sup>†</sup>Norton Children's Hospital, University of Louisville, Louisville, Kentucky, <sup>‡</sup>Pediatric Cardiology Center of Oregon, Portland, Oregon, and <sup>§</sup>Masonic Children's Hospital, University of Minnesota, Minneapolis, Minnesota.*

## Adapting leadless pacemakers: Epicardial implantation

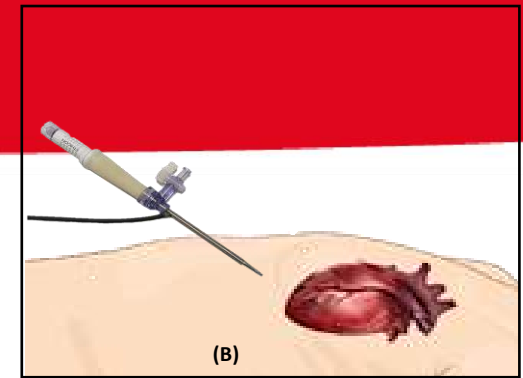
- Specially modified pediatric-sized **implantable pulse generator** (Pediatric IPG): Medtronic Micra leadless pacemaker is incorporated into a polymer header and connects to a standard epicardial pacing lead
- Compassionate use for smallest neonates



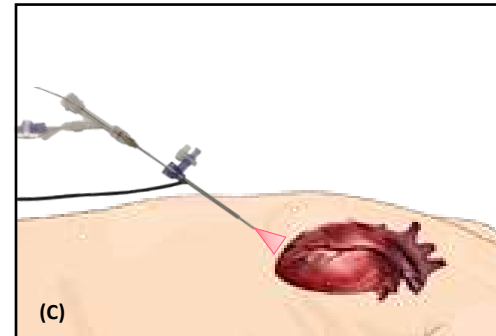
# Prototype Miniature Pacemaker: minimally invasive epicardial pacemaker



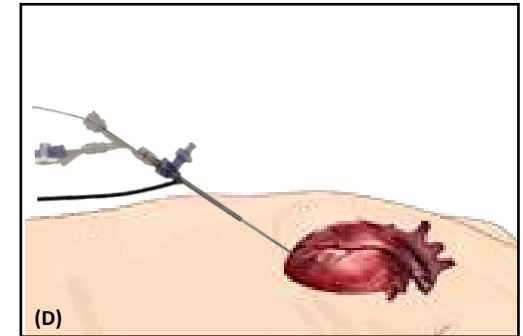
(A) Identify the subxiphoid region



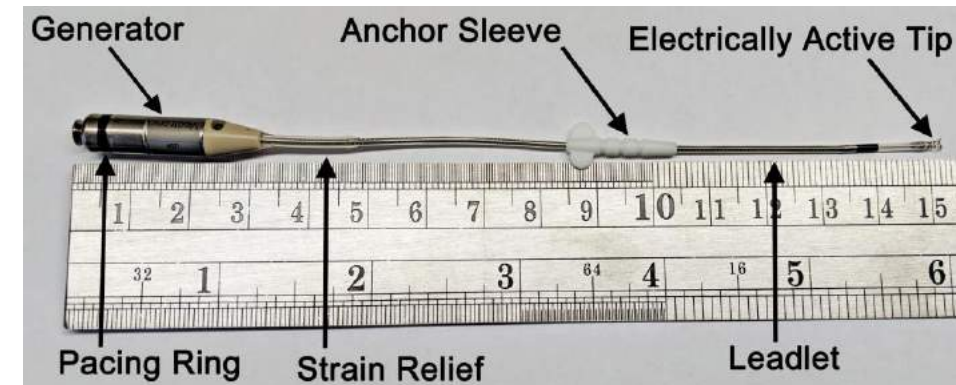
(B) Insert the illumination sheath



(C) Image the heart with PeriScope



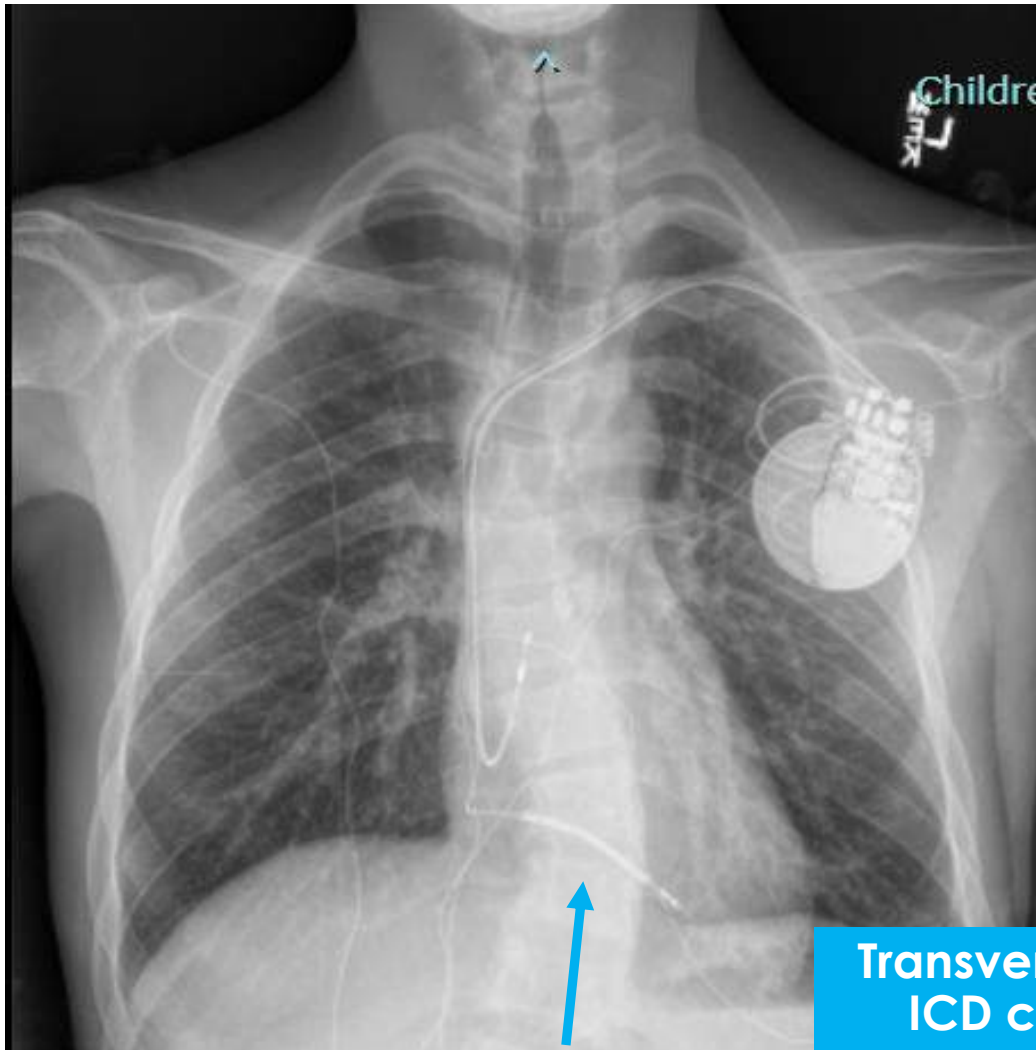
(D) Obtain pericardial access



Charles I. Berul, MD, FHR, CCDS, CEPS-P  
Pioneer in Cardiac Pacing and Electrophysiology Award Winner

TOGETHER WE ARE  HRS

# Implantable Cardioverter Defibrillators (ICD)

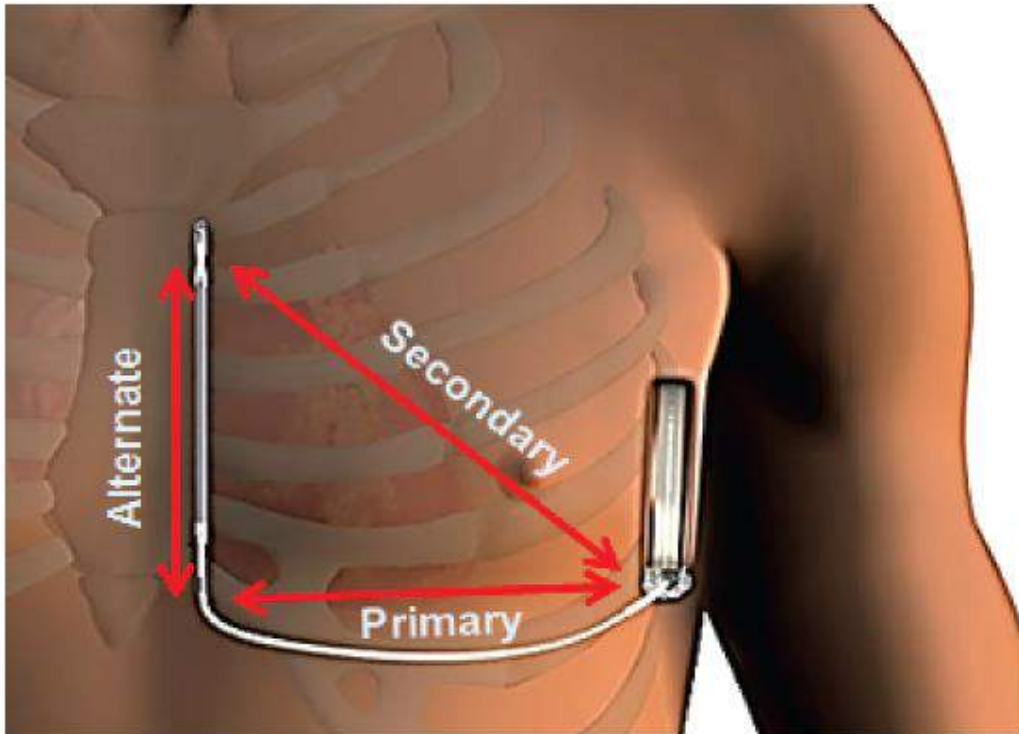


Transvenous  
ICD coil

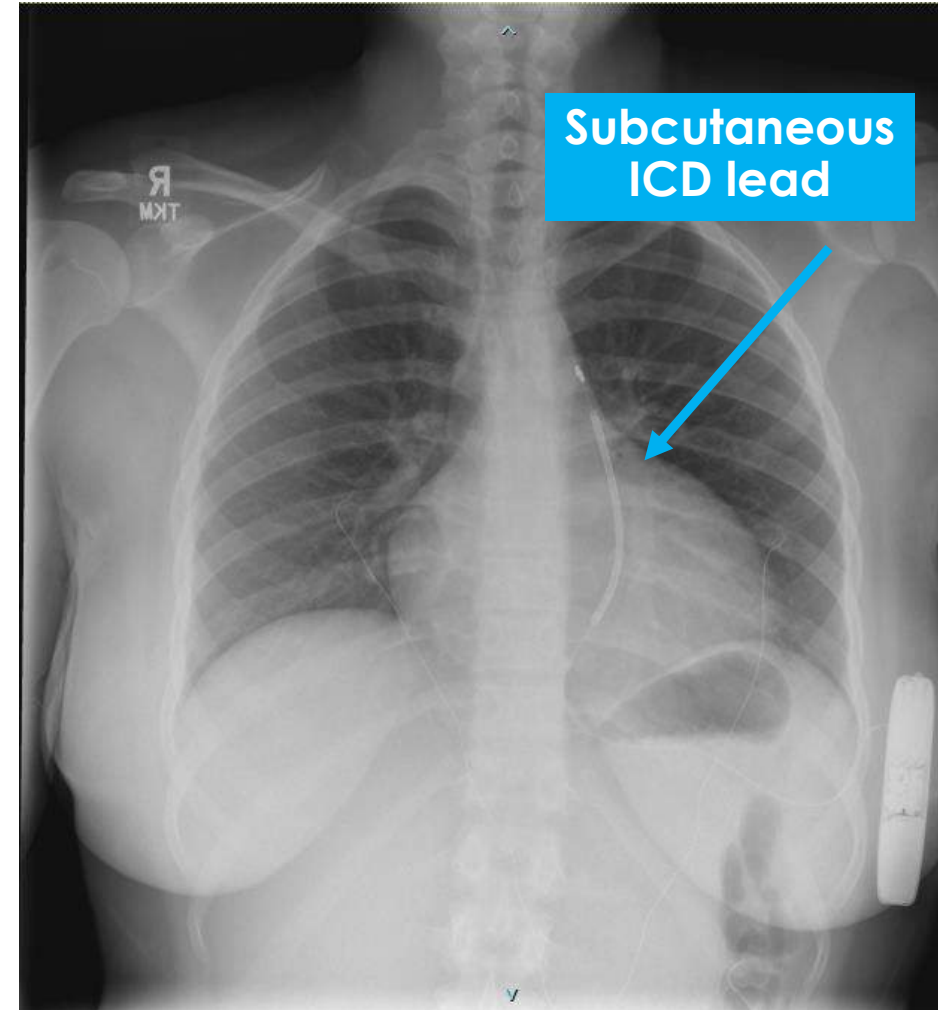
- Listen for rapid ventricular arrhythmias
- Deliver shock to terminate those arrhythmias and restore sinus rhythm
- ICDs also act as pacemaker

# Evolving Defibrillators: the subcutaneous ICD (S-ICD)

Figure 1: S-ICD System

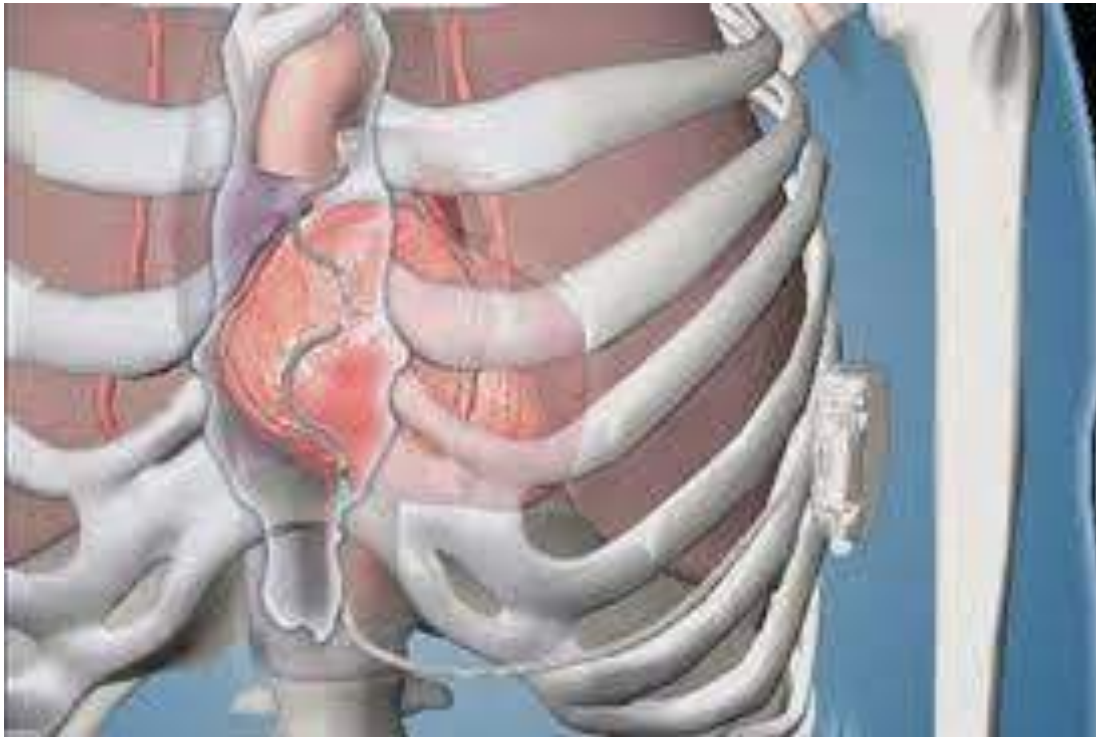


Anatomic location and sensing vectors of the subcutaneous implantable cardioverter-defibrillator system.





## Future Defibrillators: Extravascular Implantable Cardioverter-Defibrillator (EV-ICD)

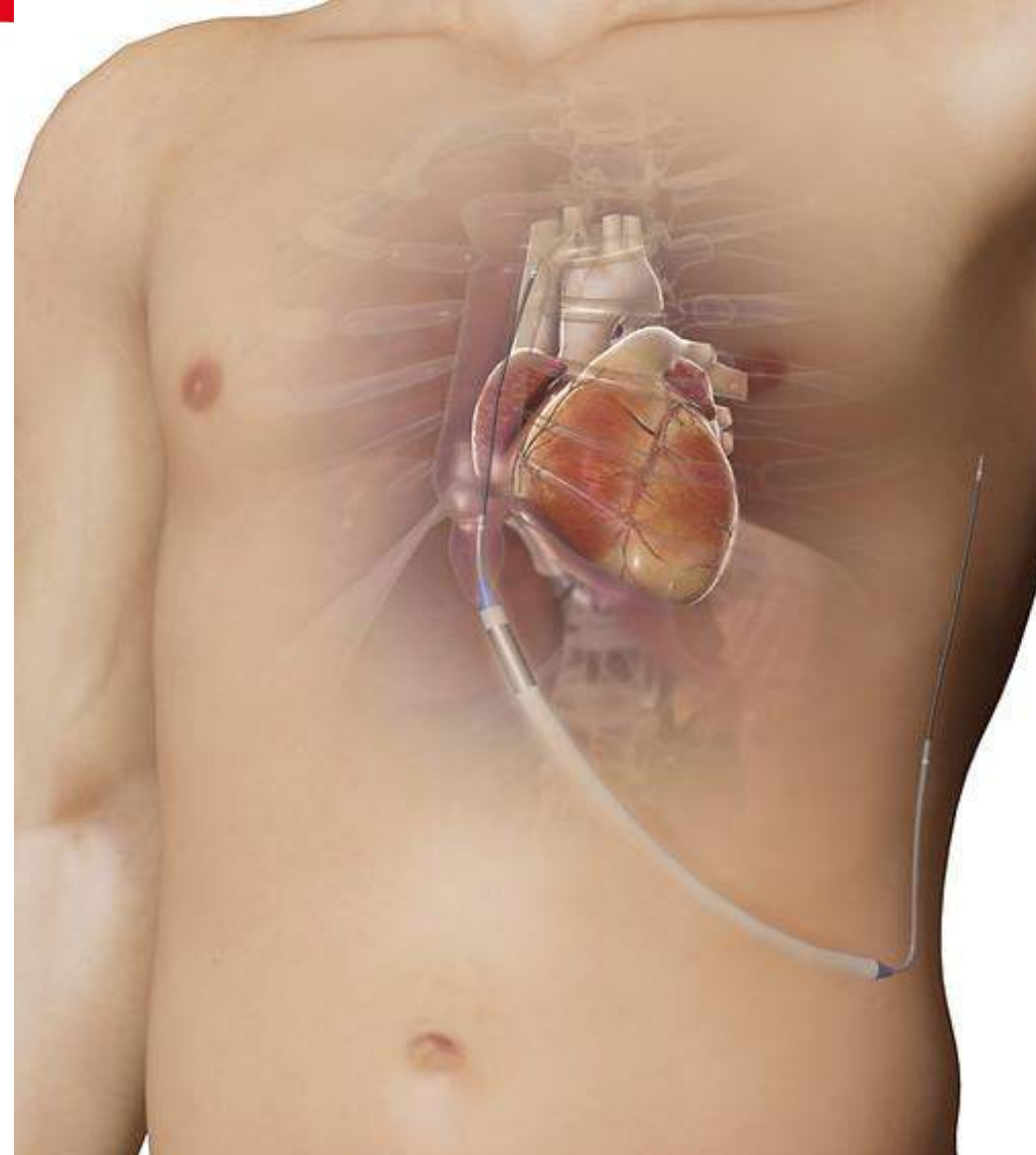


- Subcutaneous generator
- **Substernal** defibrillation lead
  - Lower energy needed
- Can provide some pacing (anti-tachycardia pacing)
- Longer battery longevity

# Future ICD??

## Implantable Subcutaneous String Defibrillator (ISSD)

- No generator
- Rechargeable battery
- Simple implant
- Still in development





## Summary

- Children may have any arrhythmia at any age
  - Many are well tolerated
  - Few will be life threatening
- Ablation is an excellent option for many arrhythmias
  - Generally  $\geq 15$ -20kg
  - Highly successful, low risk
- Cardiac pacing has evolved enormously in the last 10 years
  - Defibrillator innovation is slower but evolving
- Many options for the type of pacemaker
  - Temporary or permanent
  - Epicardial, transvenous, leadless permanent devices
  - Becoming smaller and smarter
  - In our lifetime, pacing and defibrillation may look very different!

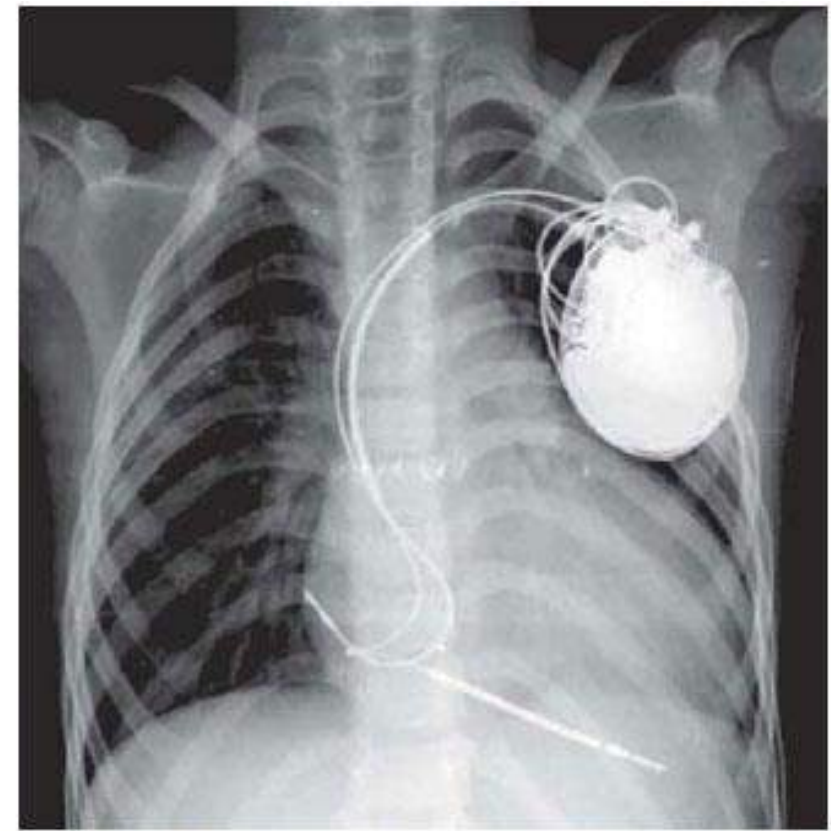


# Thank You!



Elizabeth Sherwin, MD  
edsherwin@childrensnational.org  
[www.ChildrensNational.org/HeartRhythm](http://www.ChildrensNational.org/HeartRhythm)

- Goals: treat and terminate **tachycardias**
- ICD = Internal cardioverter defibrillator
  - “AICD” is old term, was a specific company brand for Automated ICD
  - If an ICD is not automated, patient is in trouble....
- May be single chamber (ventricular) or dual chamber (with atrial pace/sense lead)
- All ICDs are also pacemakers
  - Pacing function called “brady” settings



*Fig. 4 – Chest radiography in the early postoperative period with the atrial and ventricular electrodes implanted*



# Children's National Hospital

- 323-bed acute care hospital
- 30+ outpatient locations
- Level I pediatric trauma center serving three states
- Nationally ranked in the top 10 by US News
- Critical care air and ground transport program
- 7<sup>th</sup> nationally for NIH pediatric research funding

# We Care for the Nation's Children



**240,390**  
Unique patients  
in 2019



**6,819** Employees



Contribute nearly  
**\$124 Million**  
In community benefit



**759**  
Physicians



**1,793**  
Nurses



**160**  
School-Based Nurses

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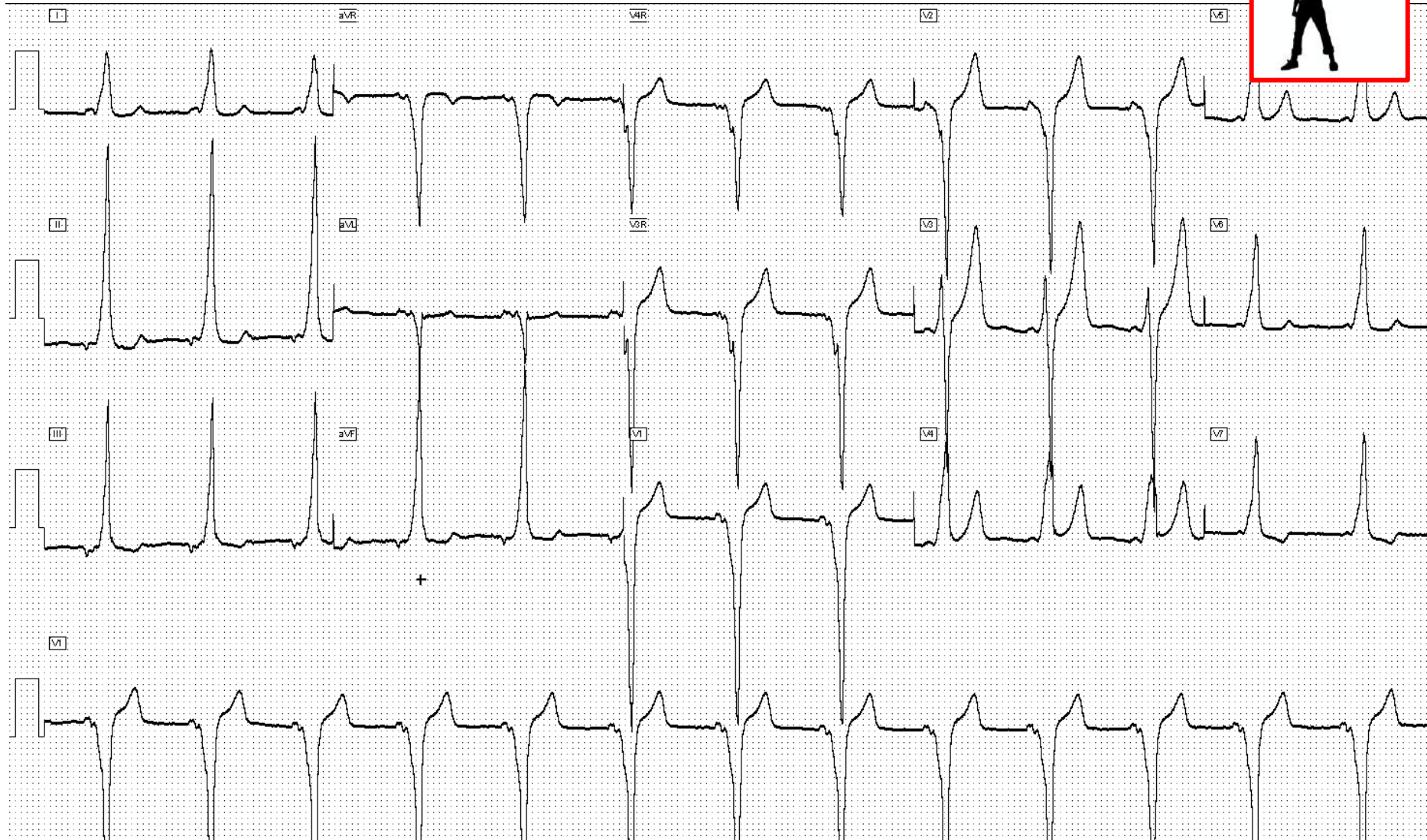


**160**  
School-Based Nurses

**Thank You!**



# Wolff-Parkinson-White





**Negative P waves in I**

**Negative P waves in aVL**



# Benefits of Leadless Pacing



## No generator pocket

- No pocket infection, erosion
- Reduces risk of bleeding in patients on anticoagulation
- Great battery longevity



## No leads

- Eliminates lead fracture
- No venous occlusion with chronic hardware in veins, maintains patency of vascular access to heart
- Reduced risk of infection, can be implanted during bacteremia (El-Chami *et al.* 2019)



## Simpler

- Easier post-implant recovery
- Improved cosmesis: no chest/abdominal incisions or visible/palpable generator
- Shorter procedure time for experienced implanters

# Leadless Pacing: Limitations in Pediatric & CHD

## Size matters

Large implant tool  
27Fr = 9 mm outer  
diameter (Micra™)

## AV Synchrony

Initially only VVI  
VDD with limited  
atrioventricular  
synchrony

## Long game

Long-term extraction  
Venous occlusion  
Effect of retained  
device over many  
decades?

## Anatomy

Vascular occlusions  
Lack of direct venous  
access to the heart  
Hypoplastic ventricles  
VSD patch, scars  
Smooth walled LV  
Intracardiac shunts