

The background is a movie poster for the film 'Titanic'. It features a close-up of Leonardo DiCaprio and Kate Winslet looking at each other, with the ship's deck and railing in the foreground. The text 'NOTHING ON EARTH COULD COME BETWEEN THEM.' is at the top. The names 'LEONARDO DiCAPRIO' and 'KATE WINSLET' are on either side of the ship's railing. The word 'TITANIC' is written in large letters across the ship's hull.

**Syncope and Sudden
Death:
Back to Titanic**

LEONARDO DiCAPRIO

KATE WINSLET

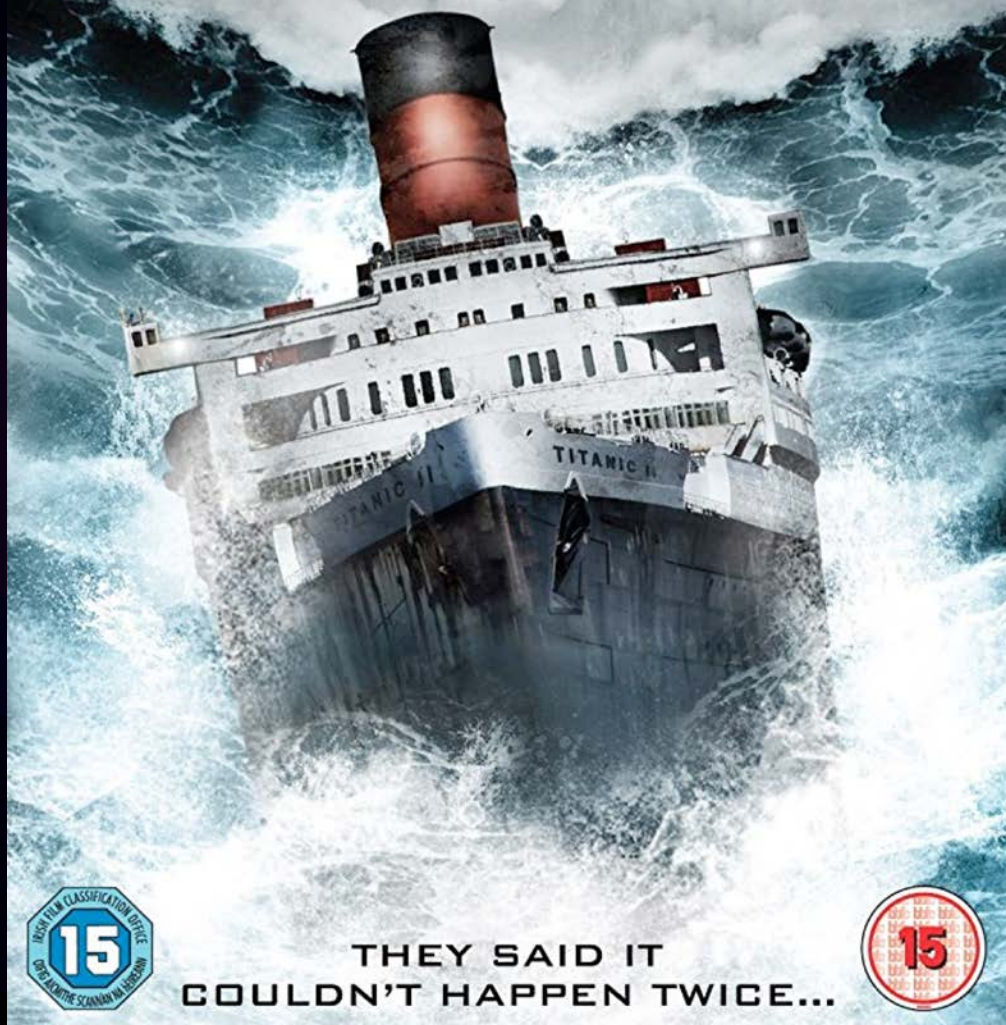
TITANIC

**Ric Samson, MD
Childrens Heart Center Nevada**

The Titanic



TITANIC 2

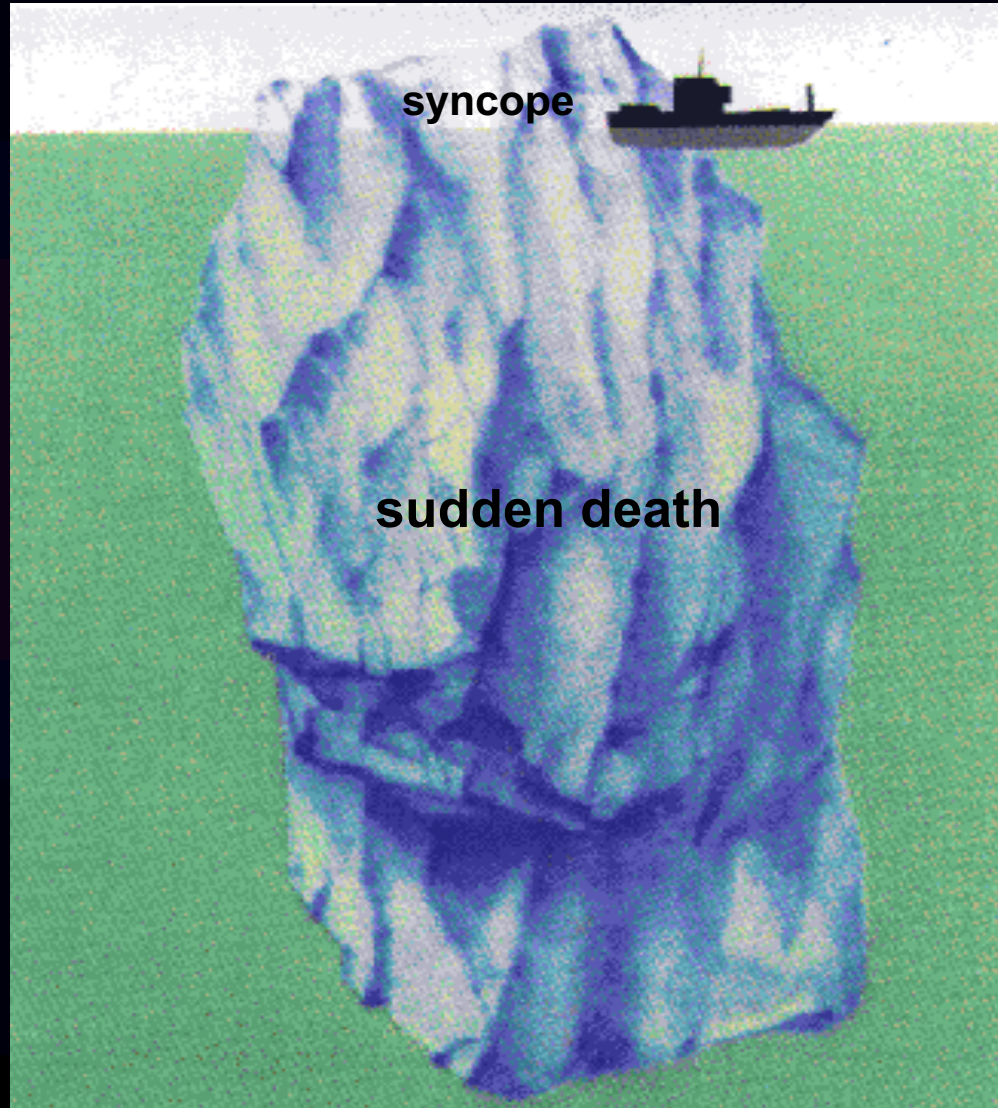


THEY SAID IT
COULDN'T HAPPEN TWICE...



Syncope vs. Sudden Death

The Dilemma



Syncope vs. Sudden Death



Definitions

- **syncope:** a brief sudden loss of consciousness and muscle tone secondary to ischemia, or inadequate substrate delivery to the brain
- **sudden death:** an unexpected death from natural causes which can occur instantaneously or within 24 hours of the patient's initial symptoms

Incidence Syncope

- **Adults**
 - **1 - 6% of hospital admissions**
 - **3% of emergency room visits**
- **Children**
 - **15% experience a syncopal episode during childhood**

Incidence Sudden Death

- **Adults – seemingly healthy**
 - competitive athletes: 4 per million per year
 - recreational athletes <30: 10-25 per million per year
 - US Air Force recruits - 1 per 3 million hours of exercise (1 per 735,000 per year)
- **Children**
 - Mayo clinic: ages 1-22 in Olmstead County, MN:
 - » 1.3 per 100,000 per year
 - Gajewski, Annals Ped Cardiol 2010
 - » 0.6-6.2 per 100,000
- **30% of all non-trauma deaths in people < 35**
- **<20% have cardiac symptoms prior to death**

Headlines - Sudden Death



Hank Gathers

abc NEWS VIDEO LIVE SHOWS

Wes Leonard: Michigan High School Basketball Star Dies After Game-Winning Shot



Patient Presentation #1

- 12 year old boy presents with transient loss of consciousness following blood draw
- episode preceded by nausea, feeling of warmth
- no previous cardiac symptoms
- past medical history unremarkable
- family history negative for cardiac disease
- physical exam normal

Autonomic Syncope

Vasovagal

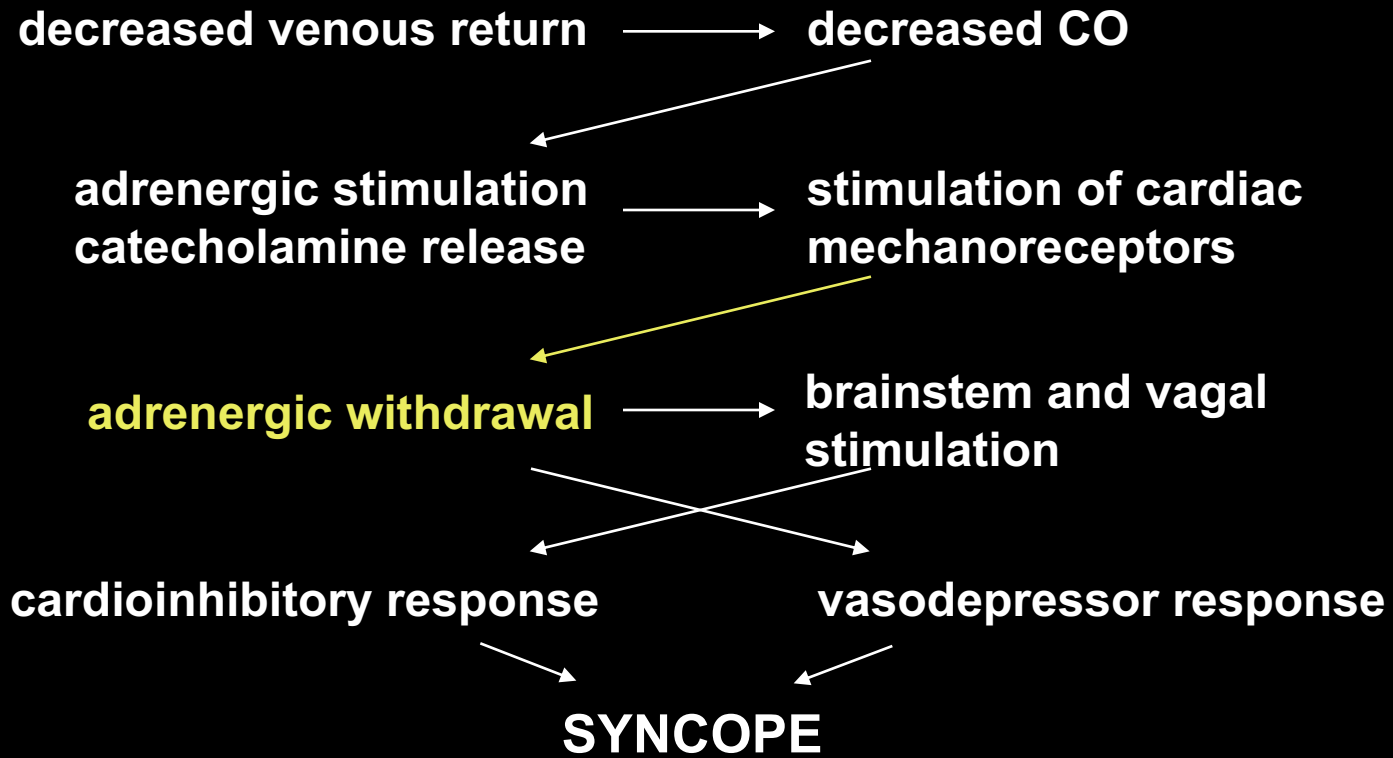
- **stimulus induced**
 - blood, pain, objectionable sights
 - nausea, anxiety, flushing, warmth
- **others:**
 - tussive
 - swallowing
 - defecation
 - breath holding spells
 - carotid hypersensitivity

Patient Presentation #2

- 16 yo female presents with syncopal episode while singing in the church choir
- 3-4 similar episodes in the past year
- typically occur in an upright position, e.g., in choir or while standing in line
- no palpitations or chest pain
- not associated with exercise
- physical exam normal

Neurocardiogenic syncope

- associated with upright posture
- exaggerated manifestation of a normal physiologic reflex (Bezold-Jarisch reflex)



Pathophysiology of neurocardiogenic syncope

- **Supine to upright**
 - Venous pooling in legs > decreased venous return > decreased cardiac output > lower BP > Compensatory release of intrinsic catecholamines
 - Catechols > increased vasoconstriction, HR, **contractility**
 - **Increased contractility sensed by cardiac β mechanoreceptors elicits paradoxical reflex**
 - **Unmyelinated C fibers within myocardium**
 - » **WHOA! Heart is beating too hard > Brainstem vagal stimulus**
 - » **Bradycardia (cardioinhibitory response)**
 - » **Vasodilation (vasodepressor response)**
 - **SYNCOPE**
 - Return to supine increases venous return, reverses process

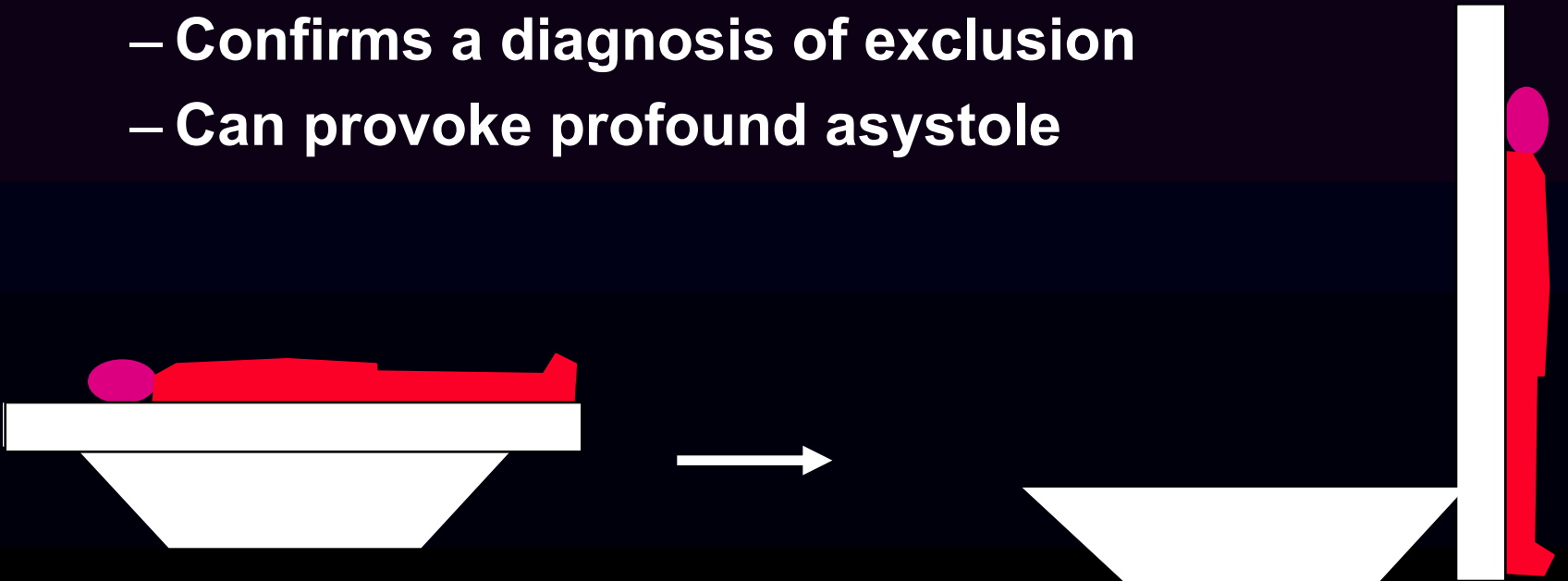
Neurocardiogenic Syncope

- not life-threatening
- diagnosis of exclusion; must rule out life-threatening conditions first
- confirmed by tilt-test
- neurocardiogenic syncope may be associated with the catecholamine release from physical exertion
- vasodepressor syncope can be seen after exertion due to continued vasodilation and decreased cardiac output
- cardioinhibitory response can be associated with profound asystole

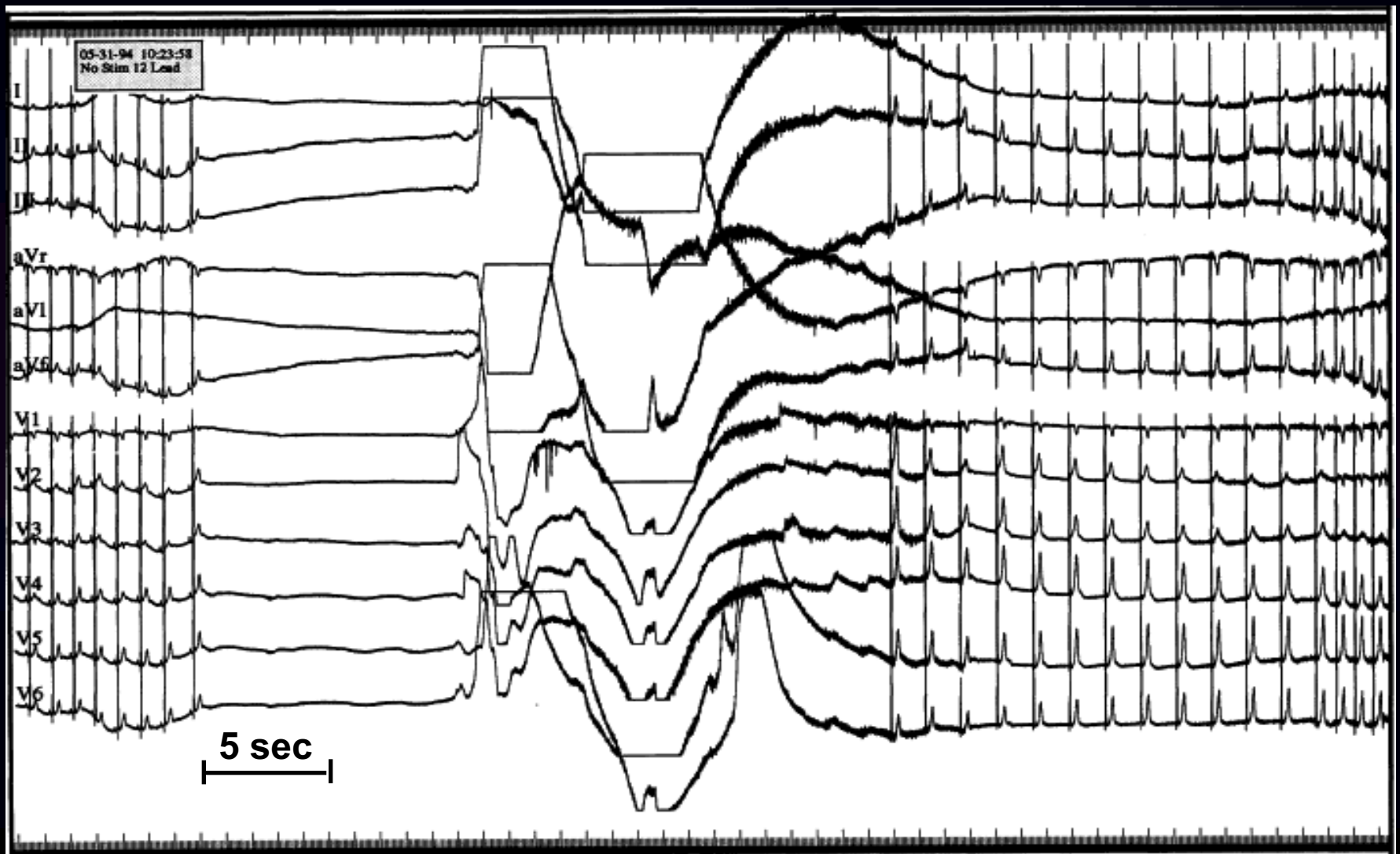
Neurocardiogenic Syncope

Tilt Table Test

- Tilt table test -
 - high sensitivity
 - low specificity - many false positives
 - » as high as 30% positive in asymptomatic people
 - Confirms a diagnosis of exclusion
 - Can provoke profound asystole



Cardioinhibitory Syncope



Neurocardiogenic syncope:

Treatment

- **Increased fluid and salt intake**
- **β blockers to block increase in contractility**
 - Atenolol, metoprolol, propranolol
- **Mineralocorticoids to cause fluid retention**
 - Fludrocortisone
- **Vasoconstrictors to block vasodilation**
 - Midodrine, pseudoephedrine
- **Anti-cholinergics to block vagal response**
 - Atropine, glycopyrronium (Robinul), disopyramide

Patient Presentation #3

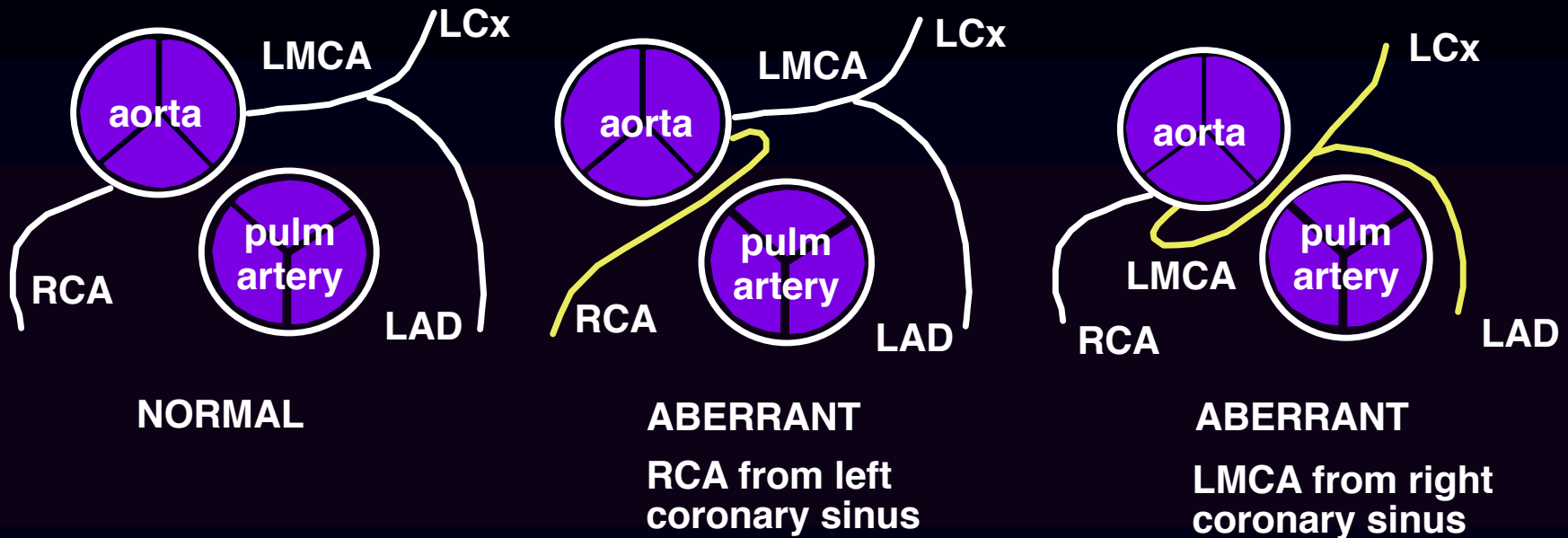
- **16 year old female - previously healthy competitive swimmer**
- **while swimming in event, became syncopal, sinking to bottom of pool**
- **required CPR, brought to ER**
- **PMH negative**
- **FH history negative for syncope, sudden death**
- **Physical exam normal**

Causes of Sudden Cardiac Death in Children

Table 1 :Causes of sudden death[8]

Causes	Relative incidence (%)
Hypertrophic cardiomyopathy	36
Increased cardiac mass	10
Coronary artery anomalies	24
Marfan's syndrome	6
Congenital heart disease	5
Myocarditis	3
Dilated cardiomyopathy	3
Arrhythmogenic right ventricular dysplasia	3
Ischemic heart disease	2
Commotio cordis	<1

Coronary Artery Anomalies



- Most often an autopsy diagnosis
- Not always easily identified by echo
- Confirmed by cardiac CT and/or catheterization

Marfan's Syndrome



Flo Hyman

- characteristic body habitus
- skeletal, cardiac, ocular involvement
- progressive aortic root dilation and risk of dissection
- autosomal dominant: + fam history
- intrinsic defect in connective tissue
- genetic defect traced to the fibrillin gene located on chromosome 15

Other causes

- **Myocarditis – inflammation of myocardium related to a viral infection**
 - Often reversible if given IVIG
- **Dilated cardiomyopathy – enlarged heart with poor ventricular function**
 - Some forms associated with genetic mutations
- **Commotio cordis – impact striking chest over precordium at a precise instant during cardiac cycle (vulnerable period) and inducing VF**
 - “very bad luck”

Syncope/Sudden Death CHD Causes

- **“non-life threatening” arrhythmias can become significant in patients with CHD**
 - **SVT in D-TGA (Mustard/Senning)**
 - **SVT in single ventricle (Fontan)**
 - **Bradycardia in CHD patients**
 - » **complete heart block**
 - » **sick sinus syndrome**

Syncope/Sudden Death: CHD Causes

- **obstruction to flow**
 - **aortic stenosis, pulmonary stenosis**
 - » **loud murmur**
 - **pulmonary hypertension**
 - » **loud P2 component of S2**
- **Tetralogy of Fallot**
 - » **hypercyanotic spells**
 - » **s/p repair - ventricular tachycardia**
- **Primary arrhythmias**

Primary arrhythmias (<1%)

- **Intrinsic abnormalities of cardiac conduction**

Arrhythmogenic right ventricular dysplasia

Long QT syndrome

Andersen–Tawil syndrome

Short QT syndrome

Brugada syndrome

Catecholaminergic polymorphic ventricular tachycardia

Wolff–Parkinson–White syndrome

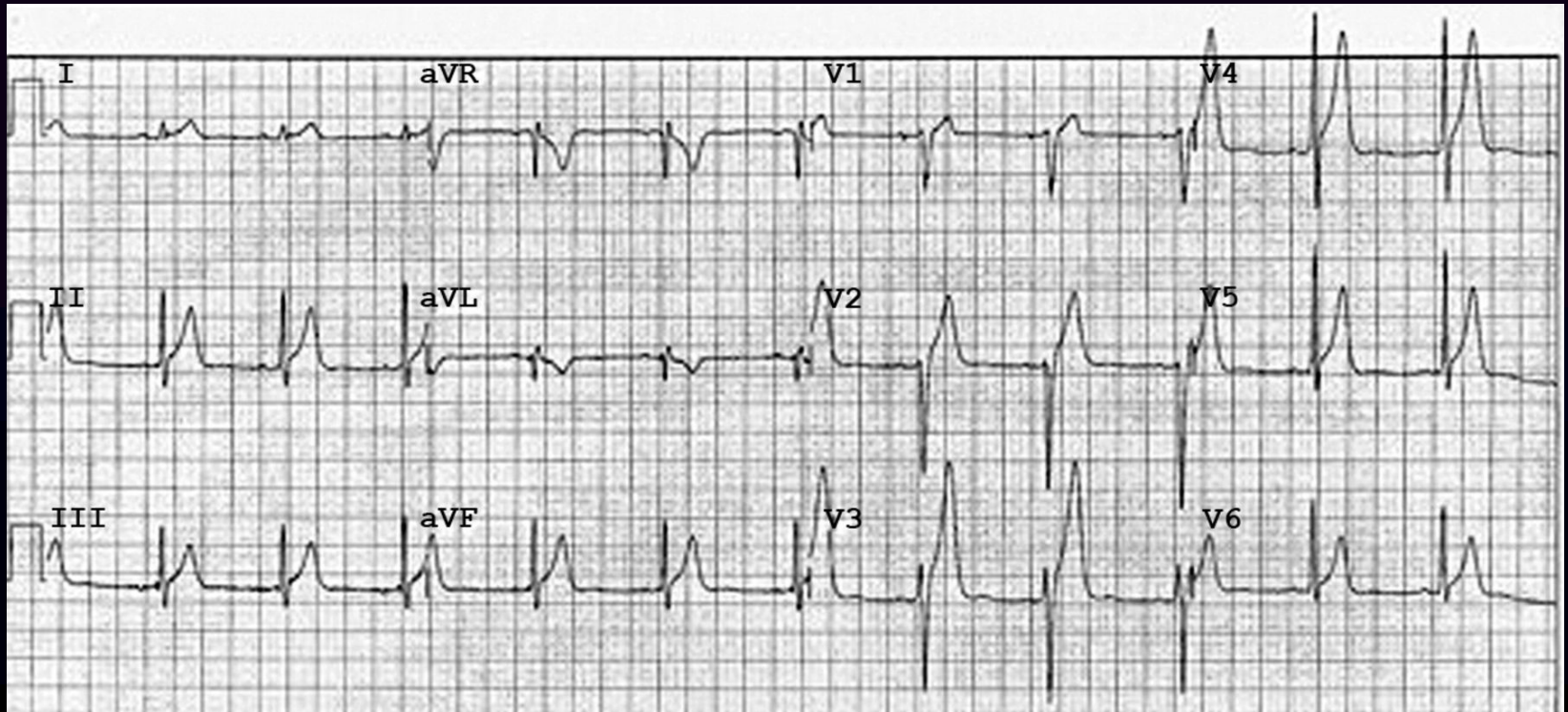
- **12-lead ECG is usually diagnostic in these conditions**
 - **CPVT is the exception; requires exercise testing to elicit**

ECG “Markers of Sudden Death”

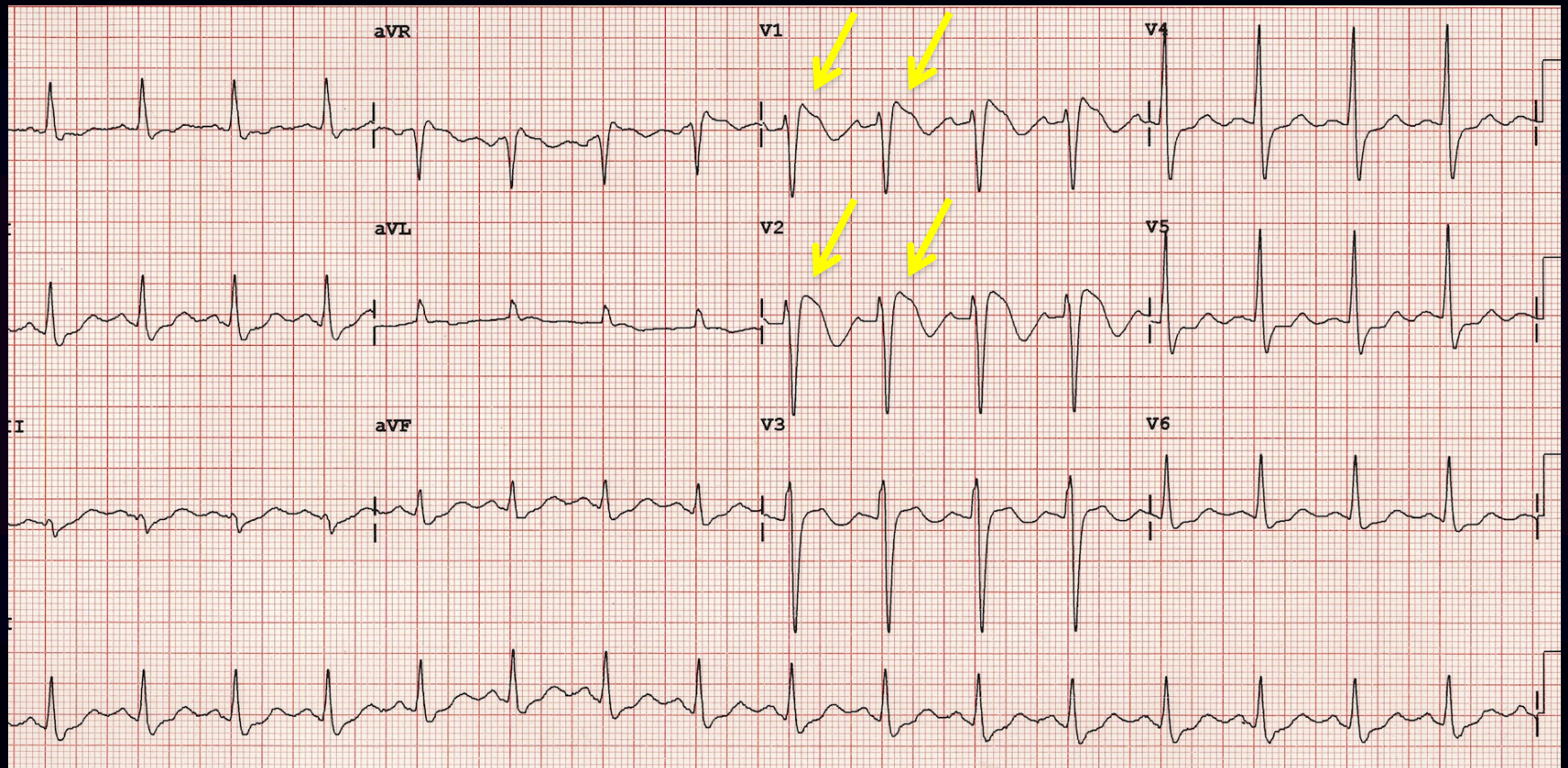
- **Hypertrophic cardiomyopathy**
 - LVH, deep Q waves in III, V6
- **Long QT syndrome – prolonged QTc interval**
 - Bazett’s formula: $QTc = QT / \sqrt{\text{preceding R-R interval}}$
- **Short QT syndrome – $QTc < 350$ ms**
- **Wolff-Parkinson-White syndrome**
 - Delta wave, “ventricular pre-excitation
- **Brugada syndrome**
 - RsR’ in V1, V2 with ST segment elevation
- **Arrhythmogenic right ventricular dysplasia/cardiomyopathy**
 - Epsilon wave

Short QT Syndrome

- QTc interval <350 ms
- Intrinsic abnormality of potassium channels
- Very rare, very lethal condition



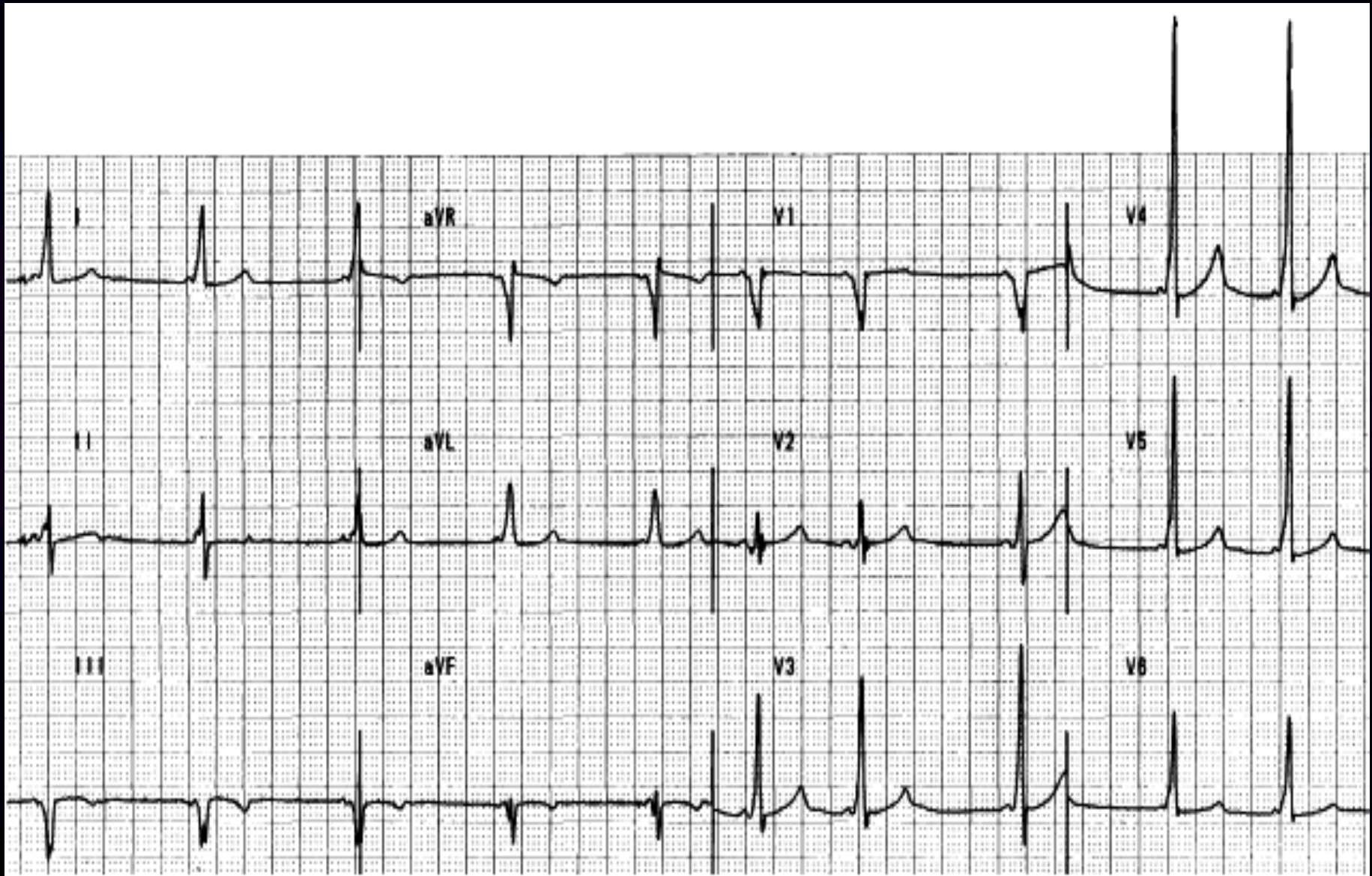
Brugada syndrome



RsR' in V1 and V2 with ST segment elevation

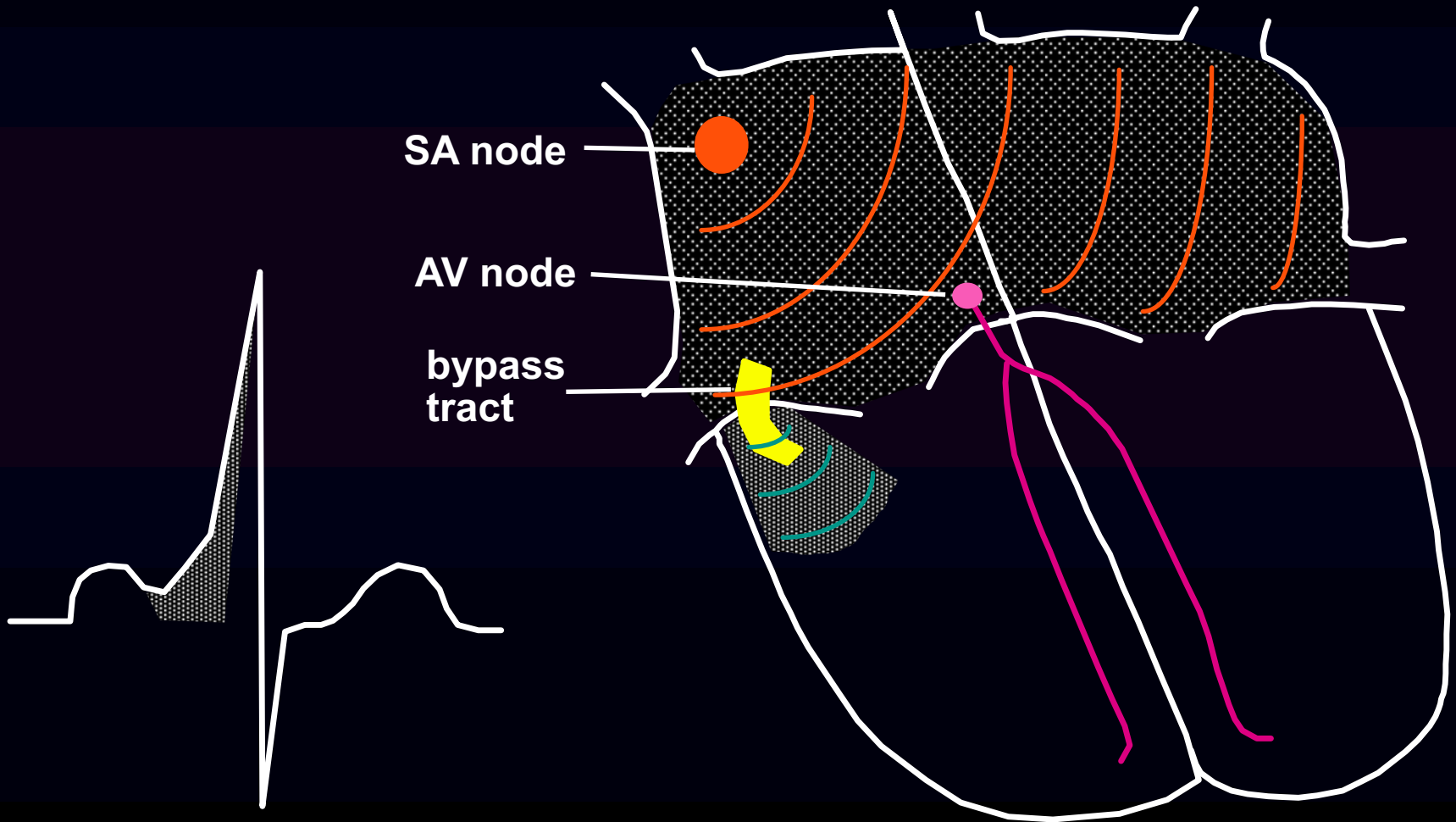
WPW - ECG

“Short PR interval with Bundle Branch Block”



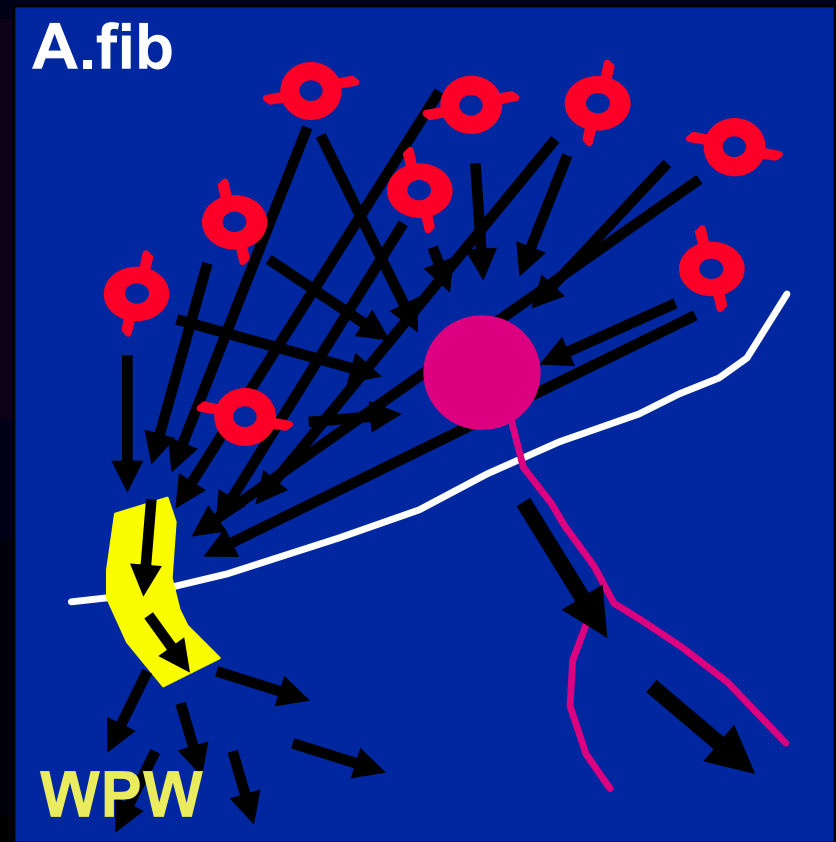
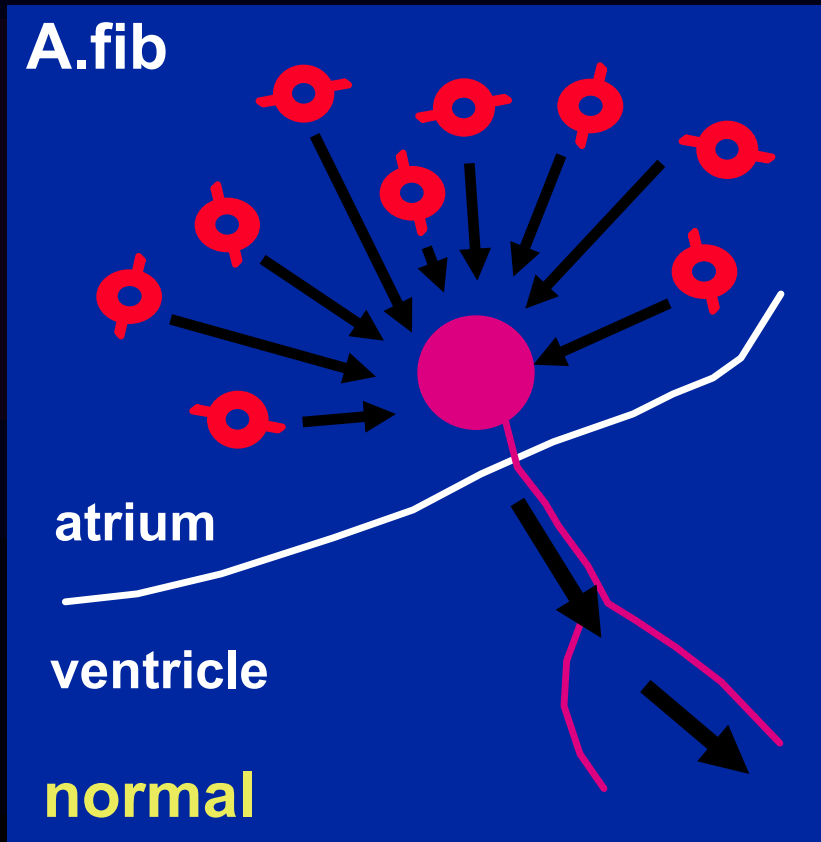
Wolff-Parkinson-White Syndrome

Delta Wave

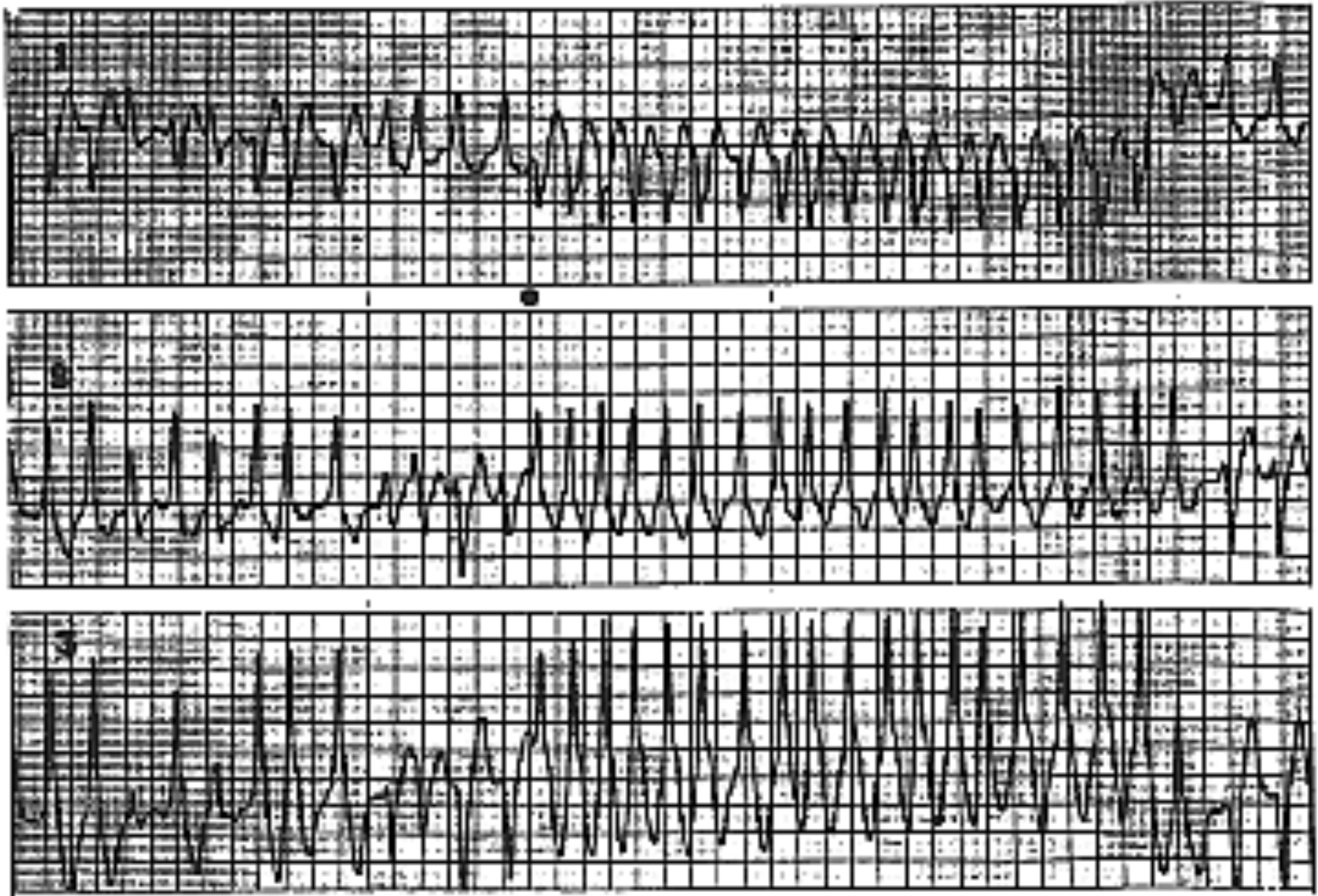


Wolff-Parkinson-White Syndrome

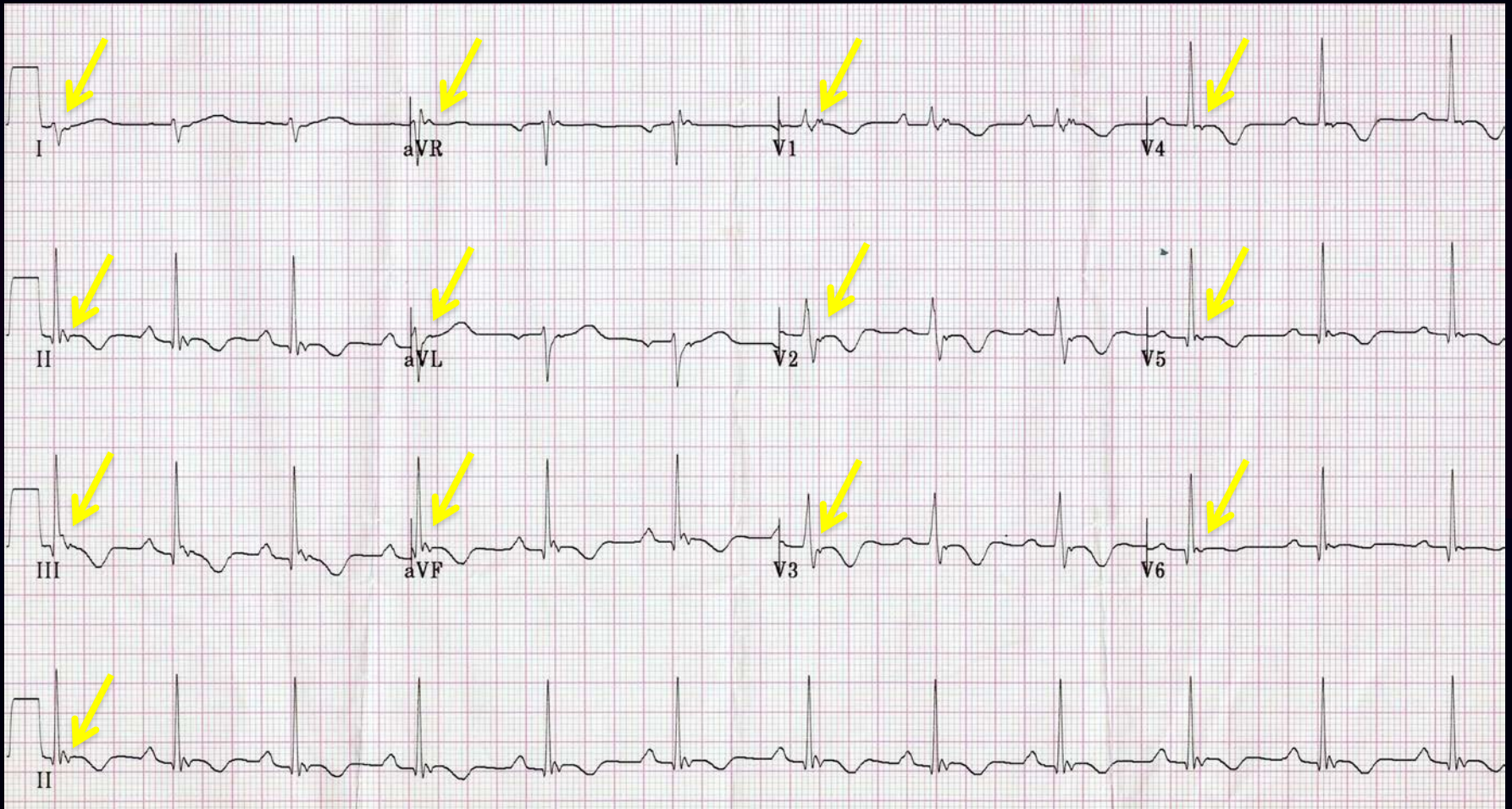
- < 1% risk of sudden death
- of those, 20% present with sudden death as first manifestation



WPW: A-fib leading to V-fib



Arrhythmogenic right ventricular cardiomyopathy



Epsilon wave – slurred terminal portion of QRS

Long QT Syndrome

- Torsade de Pointes ventricular tachycardia
- corrected QT interval > 0.45

$$QT_c = \frac{QT \text{ interval (sec)}}{\sqrt{\text{preceding R-R interval (sec)}}}$$

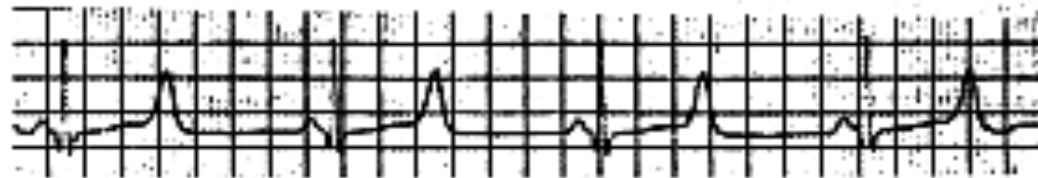
LQT1



LQT2



LQT3



Catecholaminergic polymorphic ventricular tachycardia

- **Presents with syncope associated with exercise or emotional stress**
- **Thus far, isolated to genetic mutations encoding for proteins which handle calcium within cardiac cells**
- **PE, echo, ECG and MRI are normal**
- **Diagnosed by exercise treadmill testing showing PVCs of at least 2 morphologies**
- **Genetic testing confirms about 50% of cases**

Syncope Evaluation: history, physical exam, ECG, CXR, and labs

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Catecholaminergic Polymorphic VT

Syncope vs. Sudden Death

Evaluation - History

- **stimulus, event or body position related**
- **exertion related**
- **associated symptoms**
 - **nausea, flushing, diaphoresis**
 - **dyspnea, palpitations, chest pain with exercise**
- **recurrent or infrequent**
 - **multiple episodes less likely to be serious**
- **associated with injuries**
- **hydration status/environmental**

Syncope vs. Sudden Death Evaluation - Physical Exam

- **Vital Signs**
 - orthostatic BPs
- **Cardiac Exam**
 - S2 physiologically split, P2 not loud
 - murmur
 - » aortic stenosis - loud SEM USB
 - » hypertrophic cardiomyopathy - SEM louder when upright
- **Neurologic Exam**
 - rule out seizures

Sudden Death vs. Syncope

Diagnostic Tests

- **ECG - highly sensitive, moderately specific**
- **24° Holter, event recorder - document heart rhythm at the time of symptoms**
- **echo - r/o CHD, cardiomyopathy**
- **exercise stress test - rarely diagnostic, but reassuring, a must for exertion-related syncope**
- **tilt test – to confirm neurocardiogenic syncope**
- **cath , EP study - when structural, rhythm abnormalities suspected**

