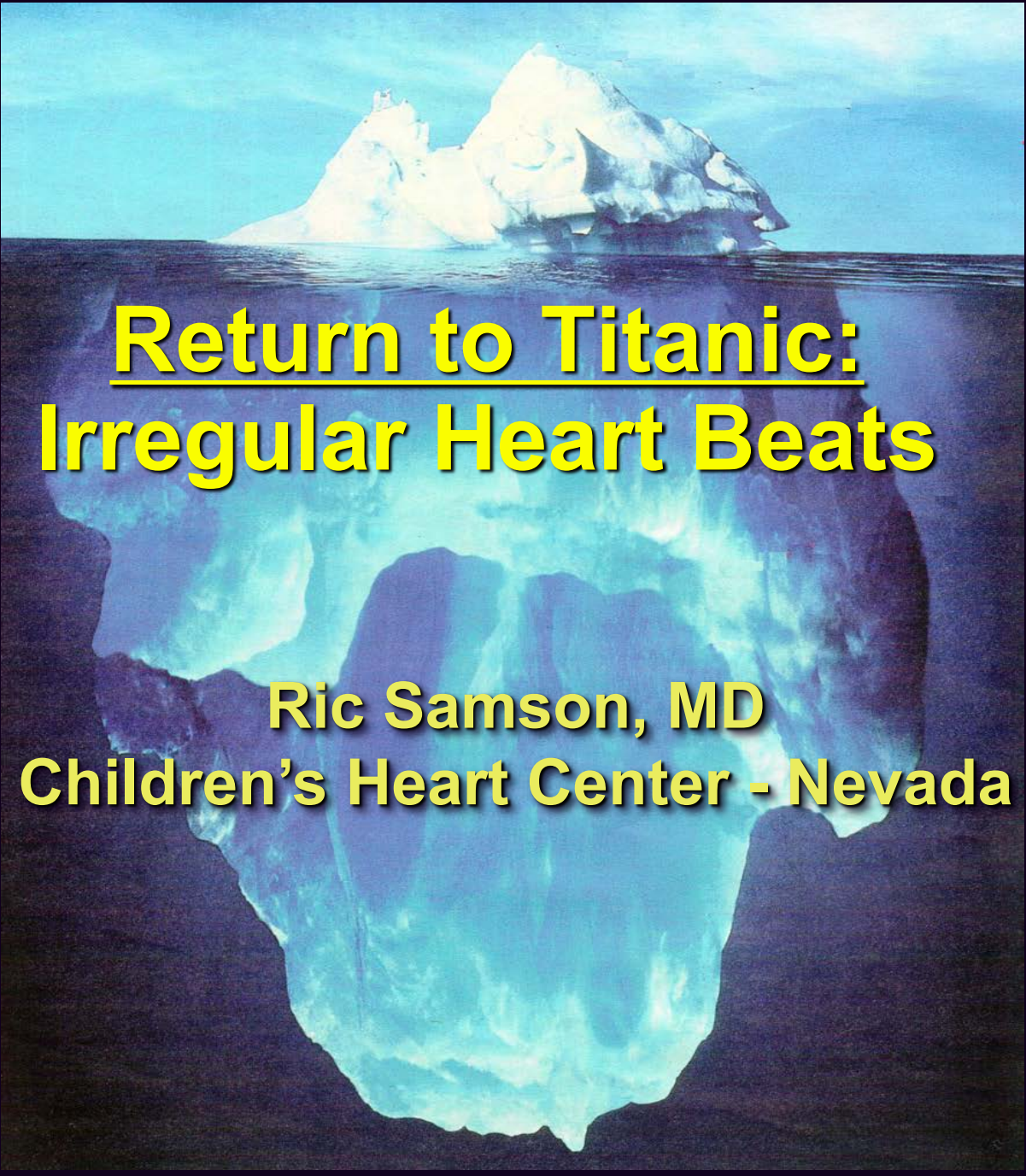


A photograph of an iceberg floating in the ocean. The tip of the iceberg is visible above the water line, while the much larger, submerged portion is visible below. The sky is a pale blue, and the water is a dark blue. The text is overlaid on the image.

Return to Titanic: Irregular Heart Beats

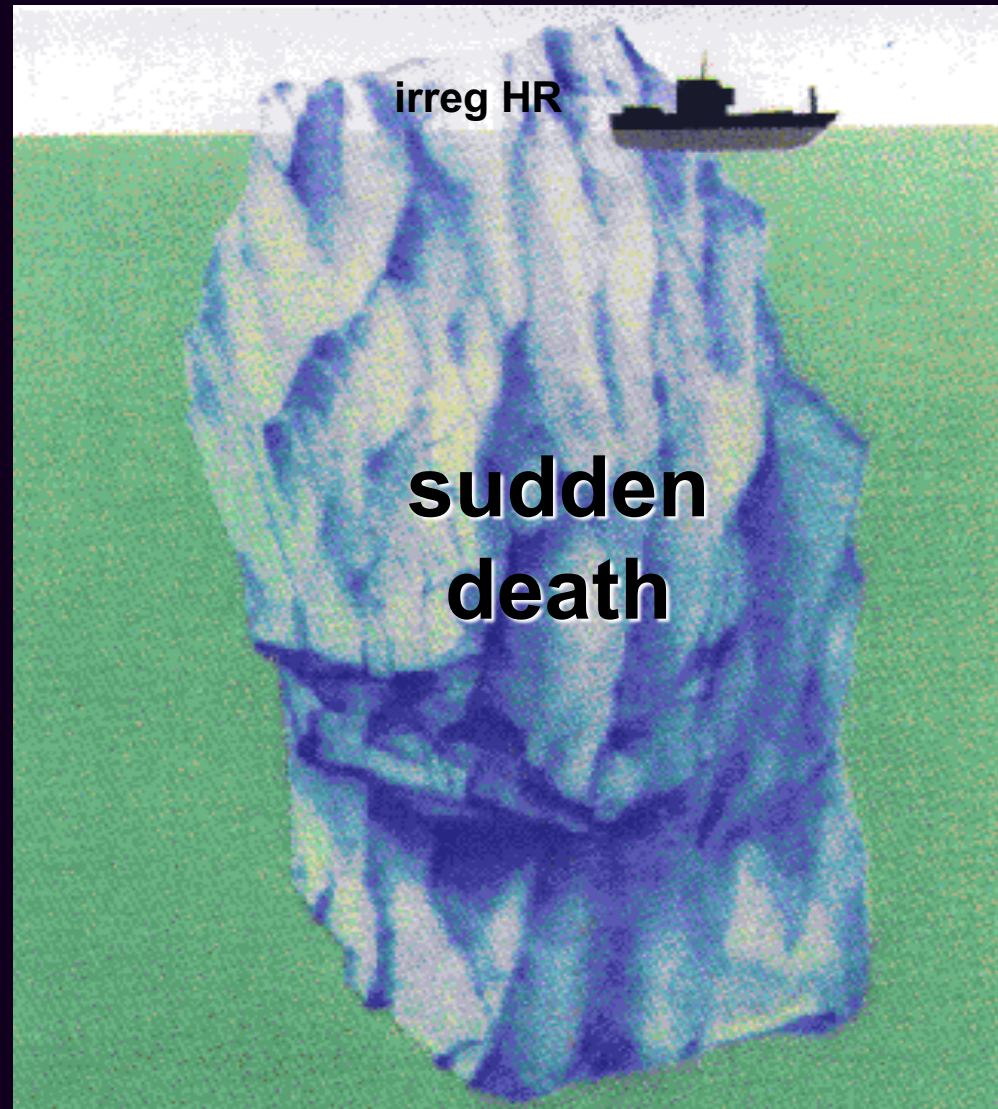
Ric Samson, MD
Children's Heart Center - Nevada

The Titanic

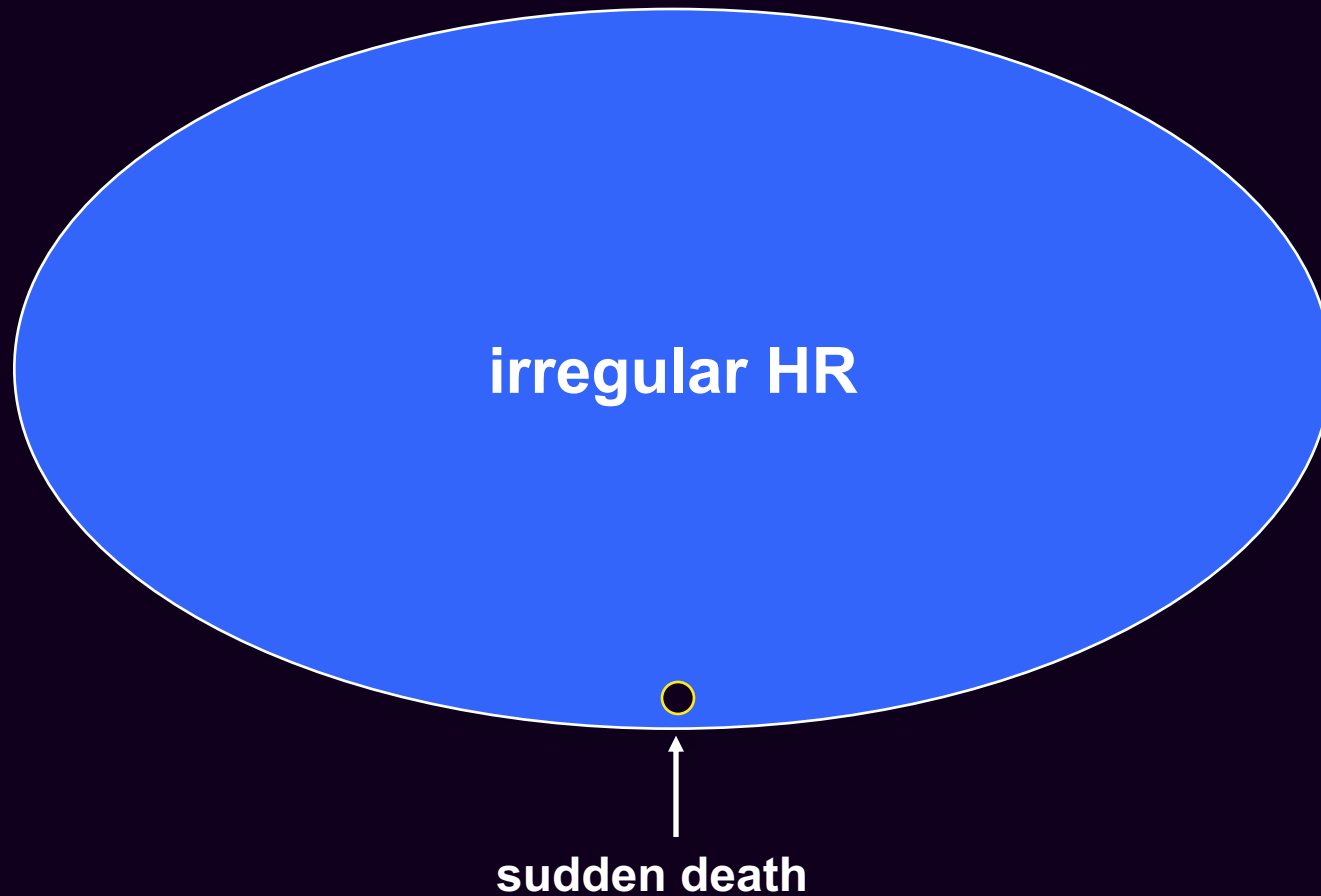


Irregular Heart Beats vs. Sudden Death

The Dilemma



Irregular HR vs. Sudden Death



Definitions

- **ectopy**: a departure of the heart rhythm from normal sinus rhythm
- **sudden death**: an unexpected death from natural causes which can occur instantaneously or within 24 hours of the patient's initial symptoms

Incidence of Sudden Death

- **Adults**
 - competitive athletes: 4 per million per year
 - recreational athletes under 30: 10-25 per million per year
 - US Air Force recruits - 1 per 3 million hours of exercise (1 per 735,00 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-traumatic deaths in people < 35
- < 20% exhibit cardiac symptoms prior to death

Patient Presentation #1

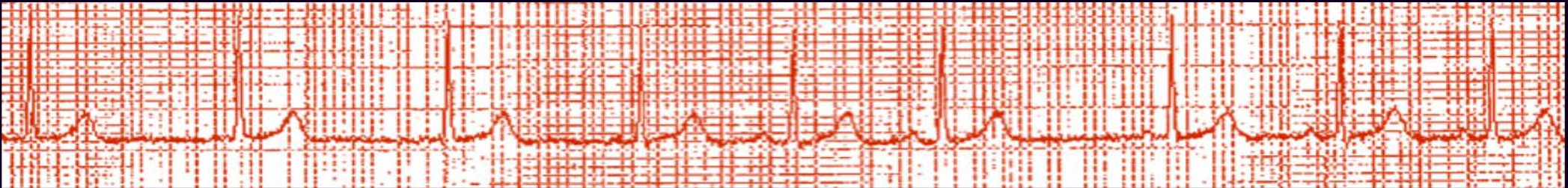
- 3 year old toddler seen in clinic for well child check is noted on exam to have an irregular heart rate
- Asymptomatic, completely healthy
- No significant PMH, FH
- HR 100 irregular, RR 20 O2 sat 98%
- Physical exam otherwise unremarkable

Sinus Arrhythmia



- **Physiologic variation in HR related respiration**
 - Speeds up during inspiration, slows down during expiration
- **Normal finding in a healthy child or young adult**
- **P waves and QRS complexes all identical**

Wandering atrial pacemaker



- **Physiologic variation in HR related respiration**
 - Secondary pacemakers within the atria
- **Also a normal finding in a healthy infant or child**
- **P waves vary slightly; QRS complex identical**

Patient Presentation #2

- **12 hour old newborn is noted to have an irregular heart rate**
- **uncomplicated pregnancy, labor and delivery**
- **HR 120 irregular, sats 98%**
- **Physical exam otherwise normal**

ECG rhythm strip

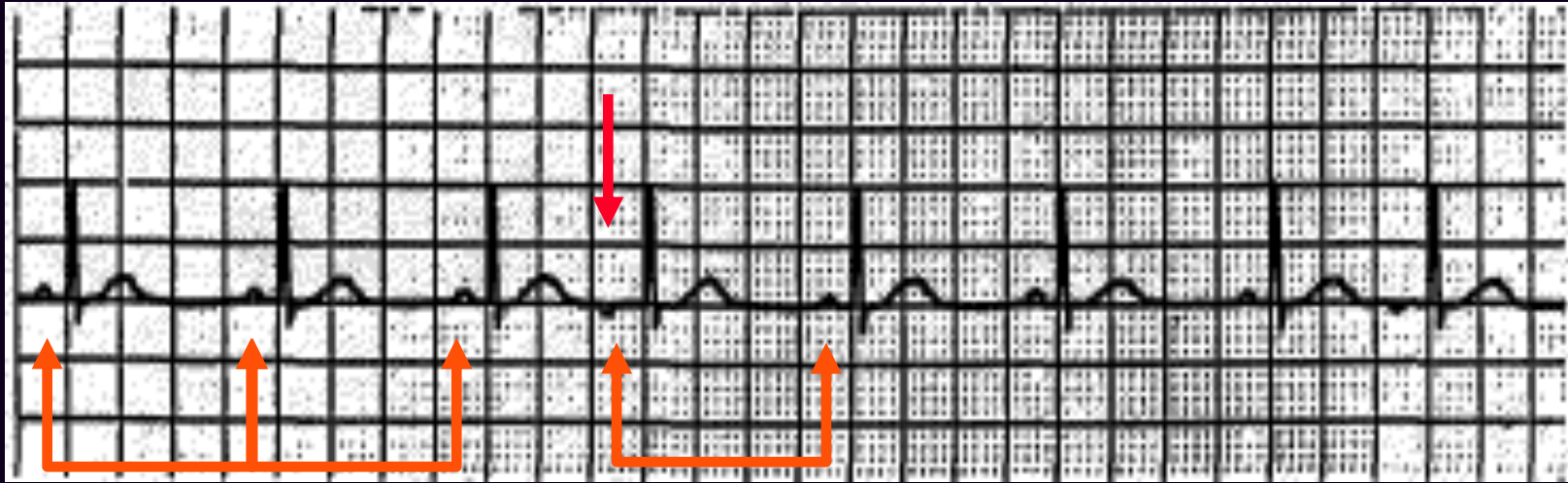


Premature Atrial Contractions

Premature Atrial Contractions

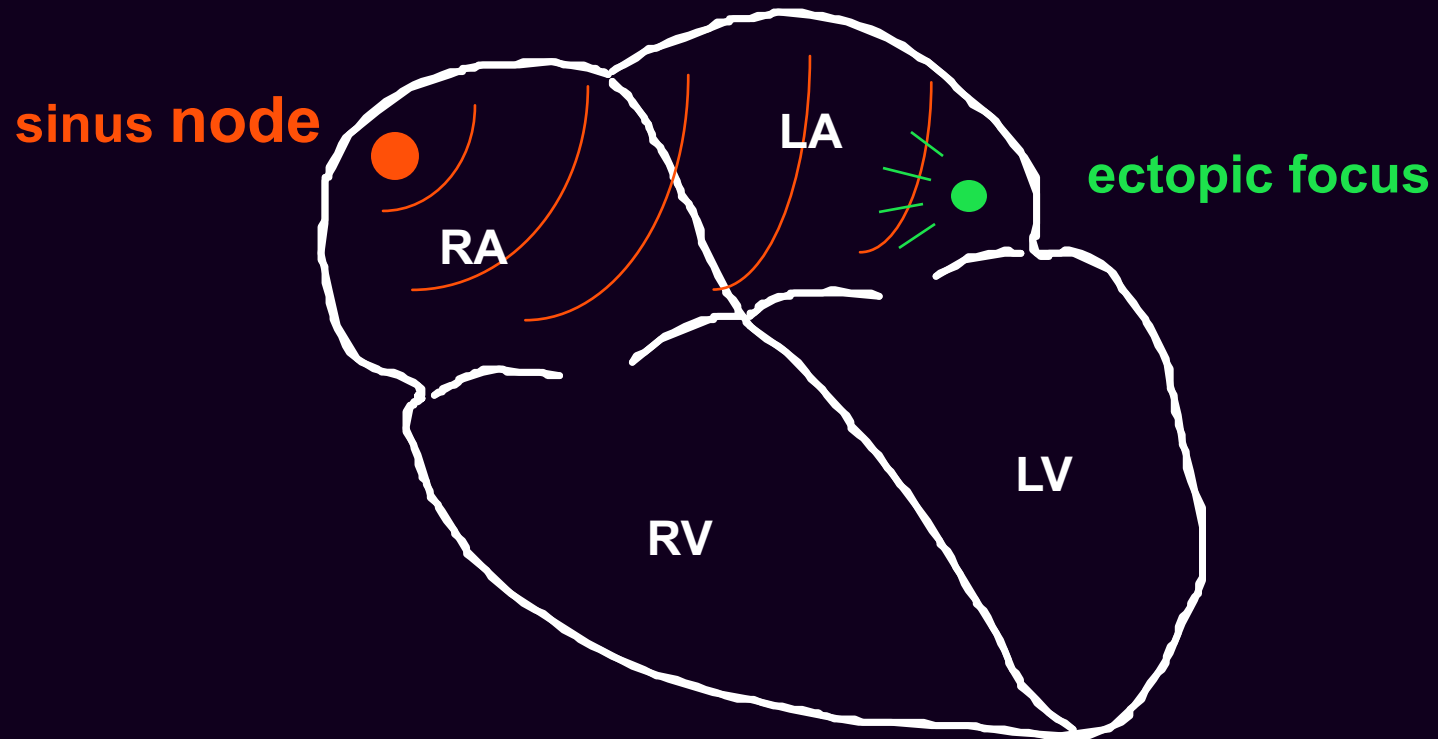
- early identifiable P wave
- P wave has different morphology from sinus P wave
- conducted
 - normally (normal QRS)
 - aberrantly (wide QRS)
 - non-conducted (no QRS)
- frequently “resets” the sinus node

Premature Atrial Contractions



- “resetting” of the sinus node
 - following PAC-P interval similar to sinus P-P interval

Premature Atrial Contraction



Premature Atrial Contractions

Causes

- **Acute**
 - mechanical - CVP line
 - metabolic - hypoxia, hypo/hyperkalemia, hypercalcemia
 - drugs – sympathomimetics, digitalis
- **Chronic**
 - s/p atrial surgery
 - atrial enlargement (ASD, AV valve regurgitation)
 - atrial tumors (myxomas)
 - normal heart

Premature Atrial Contractions

normal heart

- Nagashima et al, Pediatr Cardiol 8:103, 1987
- 360 children studied by 24° Holter
 - ages: 1 day, 1-11 mos, 4-6 yrs, 9-12 yrs, 13-15 yrs
- PACs seen in 51-77%
- incidence of 5 PACs or more per 24° was highest in 1 day olds (26%) and 13-15 year olds (23%)

Premature Atrial Contractions

Treatment/Prognosis

- Identify underlying causes
- Typically no specific anti-arrhythmic treatment required
- If associated with symptoms
 - β -blockers, other anti-arrhythmics
- Excellent Prognosis
 - Salice et al, Circ 68(III):395, 1983
 - » 6900 normal newborns prospectively studied by Holter
 - » 1 had PAC persisting beyond 1 year
 - » no development of symptoms, arrhythmias

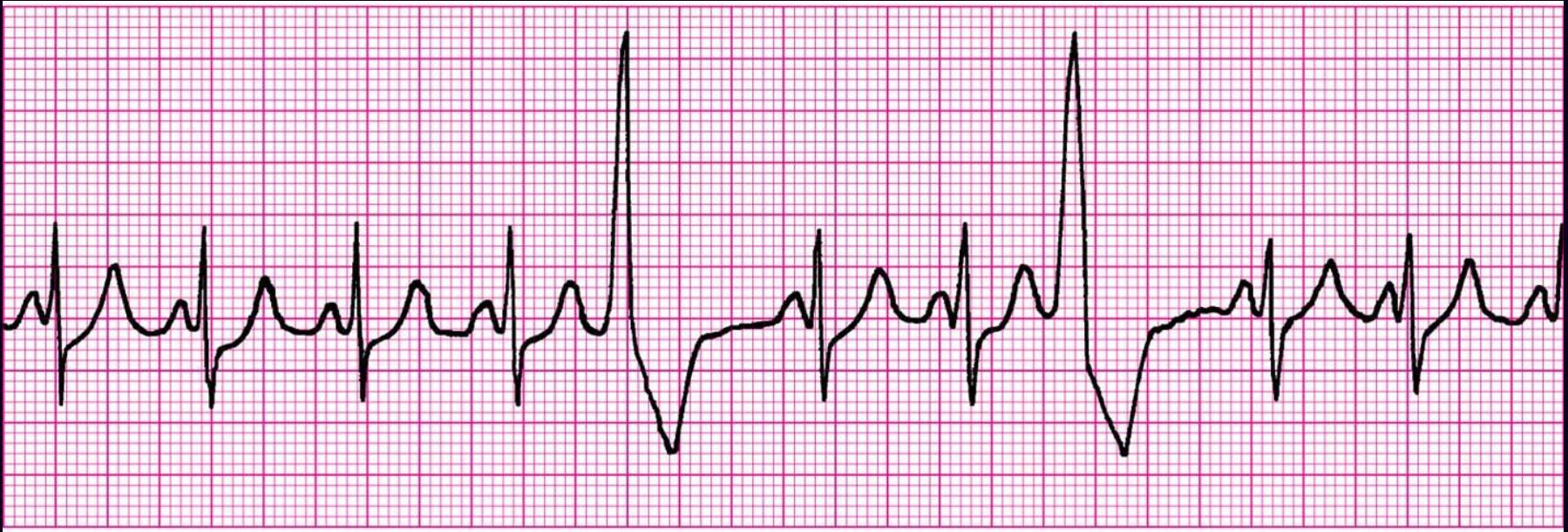
Patient Presentation #3

- **4 year old admitted for asthma exacerbation, receiving albuterol nebs**
- **noted to have irregular rhythm on monitor**
- **HR 120, O2 sat 99%**
- **Bilateral wheezes, o/w exam unremarkable**

Patient Presentation #4

- **12 yo male presents for routine pre-sports participation physical exam**
- **Healthy and asymptomatic**
- **Negative PMH, FH**
- **On auscultation is noted to have irregular HR**
- **PE otherwise normal**

ECG – irregular HR

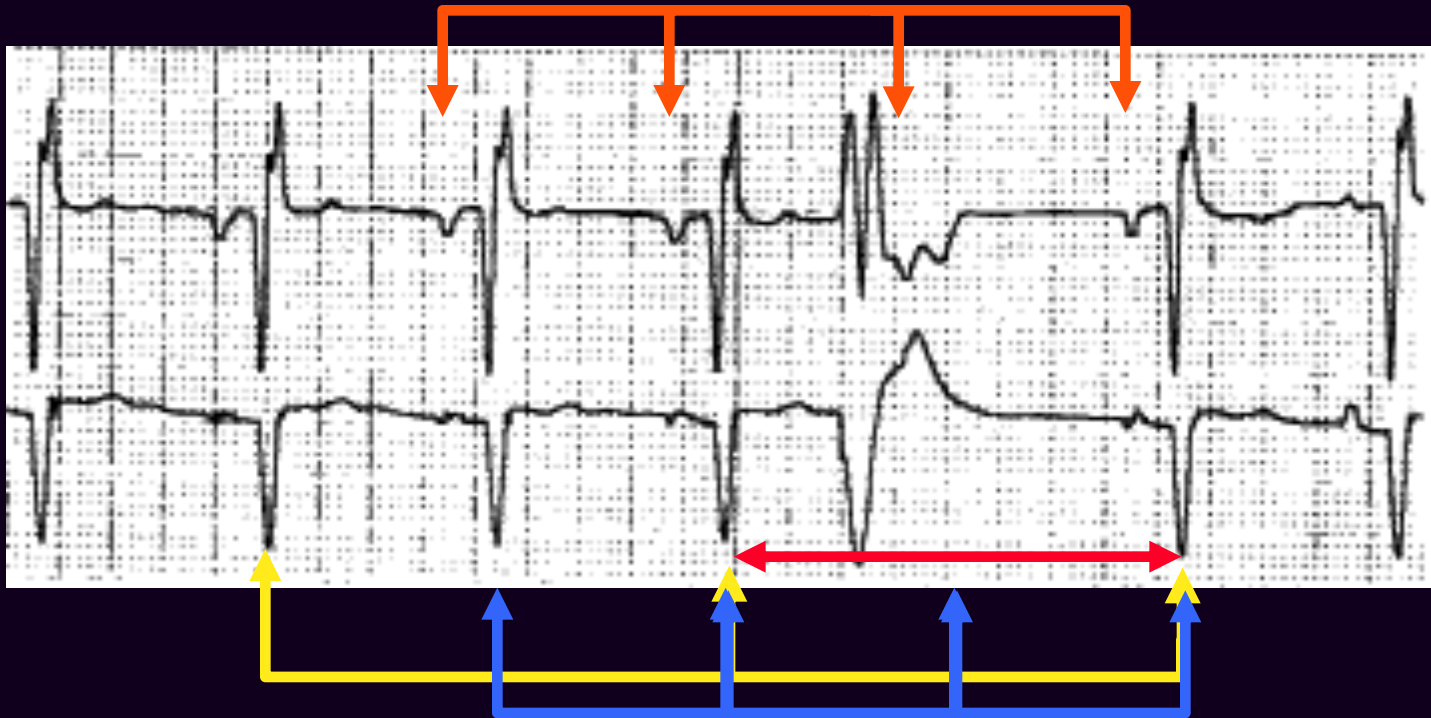


Premature Ventricular Contractions

Premature Ventricular Contraction

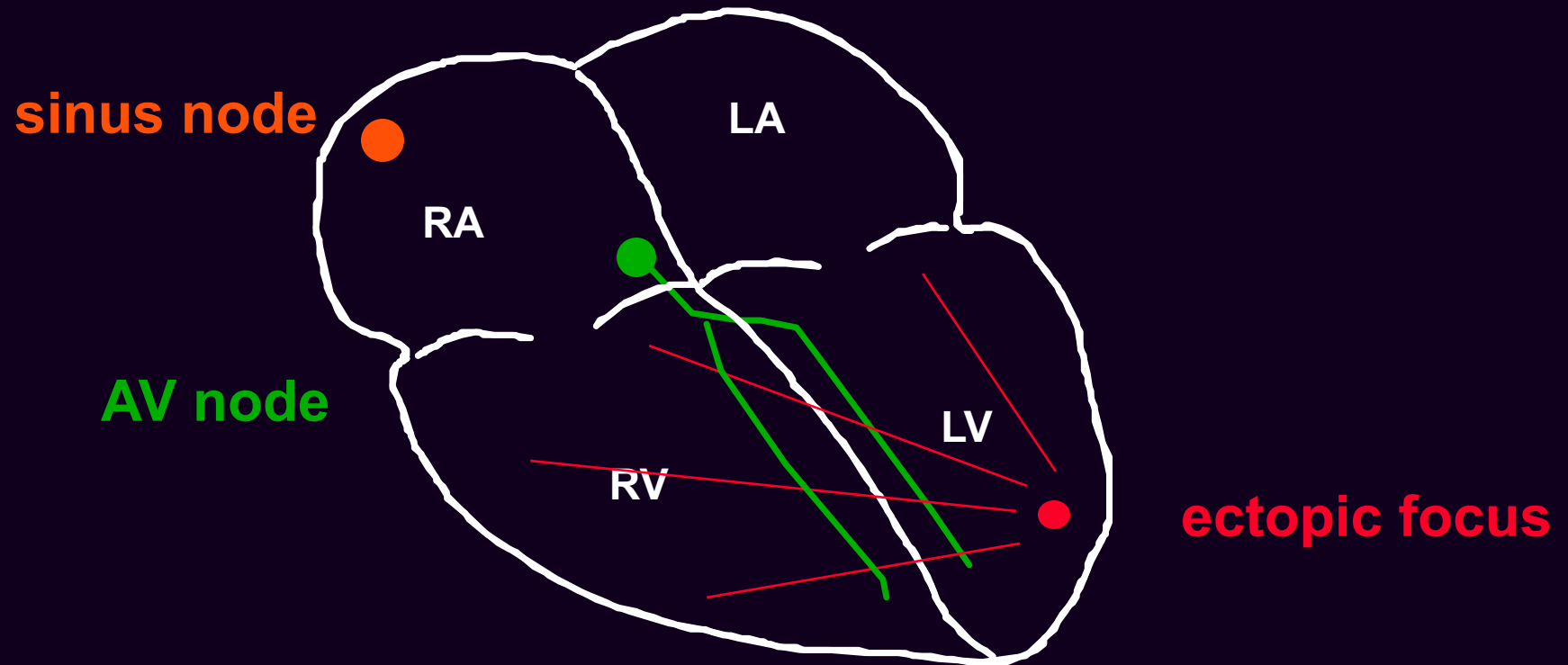
- Aberrant QRS, different from QRS in sinus rhythm
- No identifiable P wave
- Compensatory pause
- Fusion beats
- Other terms
 - bigeminy, trigeminy
 - couplets, triplets
 - non-sustained ventricular tachycardia

Premature Ventricular Contraction



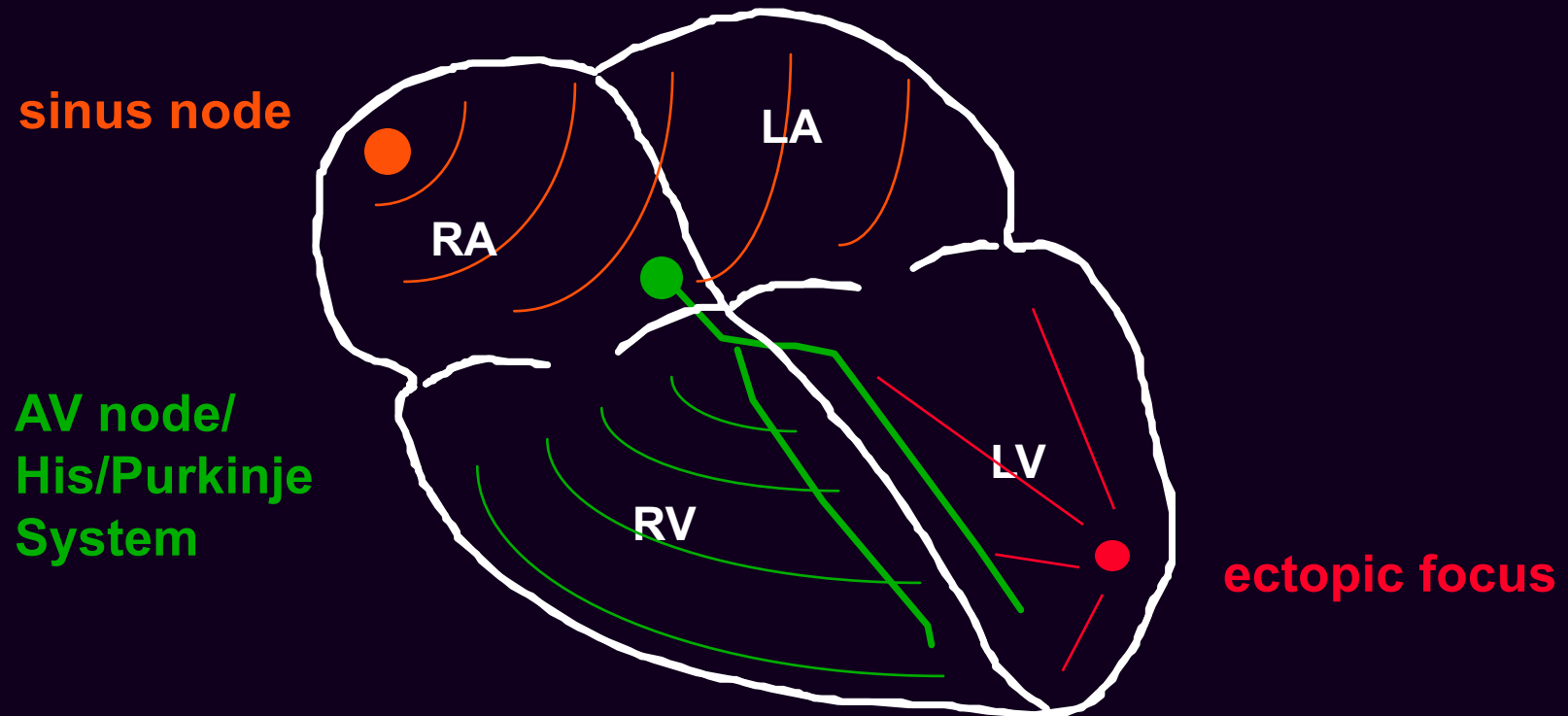
- **compensatory pause**
 - helpful but not diagnostic
 - interval between previous and following sinus beats = 2 x sinus interval

Premature Ventricular Contraction



Premature Ventricular Contractions

- fusion beats - not so premature PVCs
 - part of ventricle depolarized from PVC, the rest by normal His-Purkinje System



Premature Ventricular Contraction Causes

- **Acute**
 - mechanical - CVP line
 - metabolic
 - » hypoxia
 - » electrolyte abnormalities
 - drugs
 - » sympathomimetics
 - » digoxin, anti-arrhythmics

Premature Ventricular Contraction Causes

- **Chronic**
 - **normal heart**
 - » **Southall et al, Br Heart J 43:14, 1980**
Southall et al, Br Heart J 45:281, 1981
 - **18% of normal newborns by Holter**
 - **26% in older children, esp. teenagers**

Premature Ventricular Contraction Treatment

- Identify any underlying cause
- Determine whether PVCs are suppressed by exercise
- If exam is normal and patient is asymptomatic, no treatment necessary
- If symptomatic, β -blocker or anti-arrhythmic

Premature Ventricular Contraction

Prognosis: normal heart

- **Southall et al, Pediatrics 68:58, 1981**
Nagashima et al, Pediatr Cardiol 8:103, 1987
 - PVCs resolved by 8-12 weeks of age in all infants
- **Jacobsen et al, J Pediatr 92:36, 1978**
 - 17 asymptomatic children with nl hearts and frequent PVC followed up to 13.8 yrs
 - » 4 with bigeminy, 3 with trigeminy
 - PVCs resolved in 9/17
 - all remained asymptomatic

Premature Ventricular Contractions

- **Less Benign**
 - s/p heart surgery
 - myocarditis
 - tumors (tuberous sclerosis)
 - multiform PVCs
 - couplets, triplets or non-sustained ventricular tachycardia

Premature Ventricular Contraction

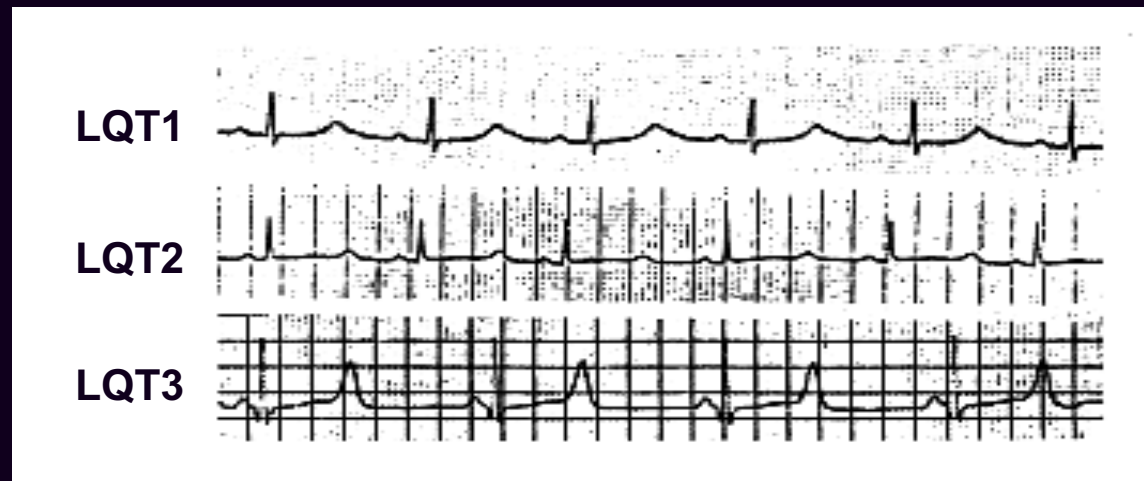
- Potentially Life Threatening
 - PVCs in the setting of
 - » Long QT syndrome
 - » Hypertrophic Cardiomyopathy
 - Maron et al, Am J Cardiol 48:532, 1981
 - presence of ventricular ectopy in patients with HCM was a strong predictor of sudden death

Long QT syndrome

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

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

- Phoon, Pediatrics 100 (3): 439, 1997
- if QT interval is $< 1/2$ of R-R interval, QTc < 0.45
 - not valid if HR < 70 bpm

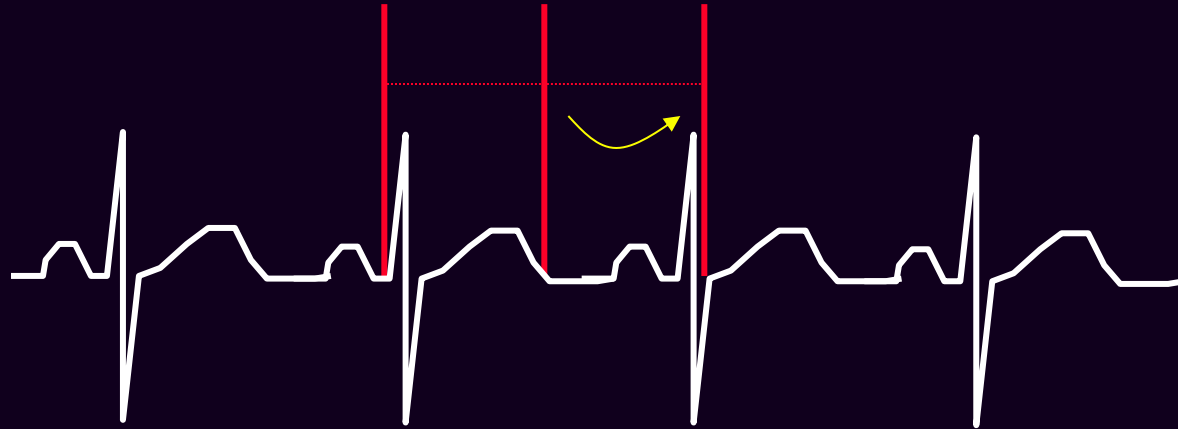


Corrected QT interval: short cut

QTc OK, (< 0.45)

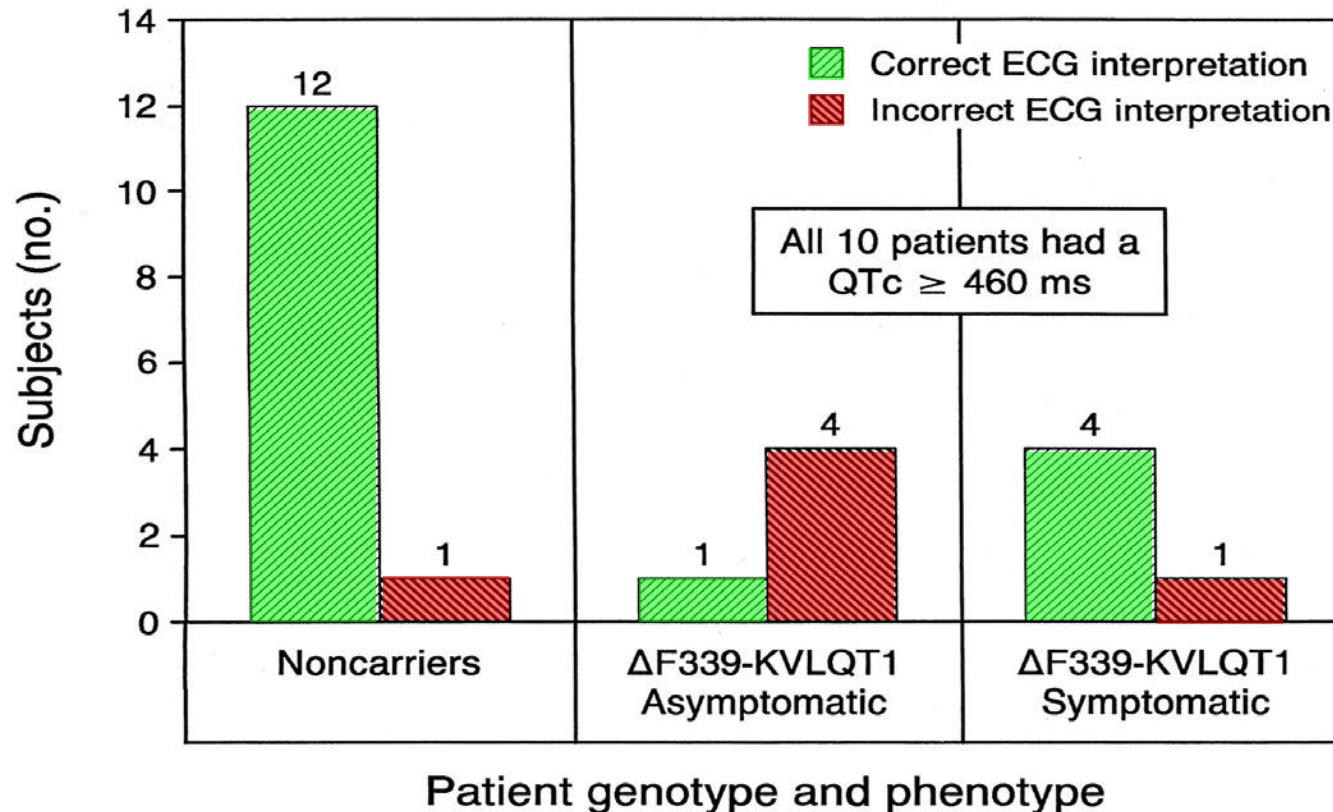


Must calculate QTc



Can't Always Trust the Computer

- Miller et al, Pediatrics 108:8;2001



**Family of LQTS
patients and
automated
ECG
interpretation**

Examples – erroneous interpretations

12-Aug-2006

Male

Room: OP

Vent. rate 127 bpm

PR interval 124 ms

QRS duration 76 ms

QT/QTc 322/467 ms

P-R-T axes 54 68 21

** * Pediatric ECG analysis * **

Normal sinus rhythm

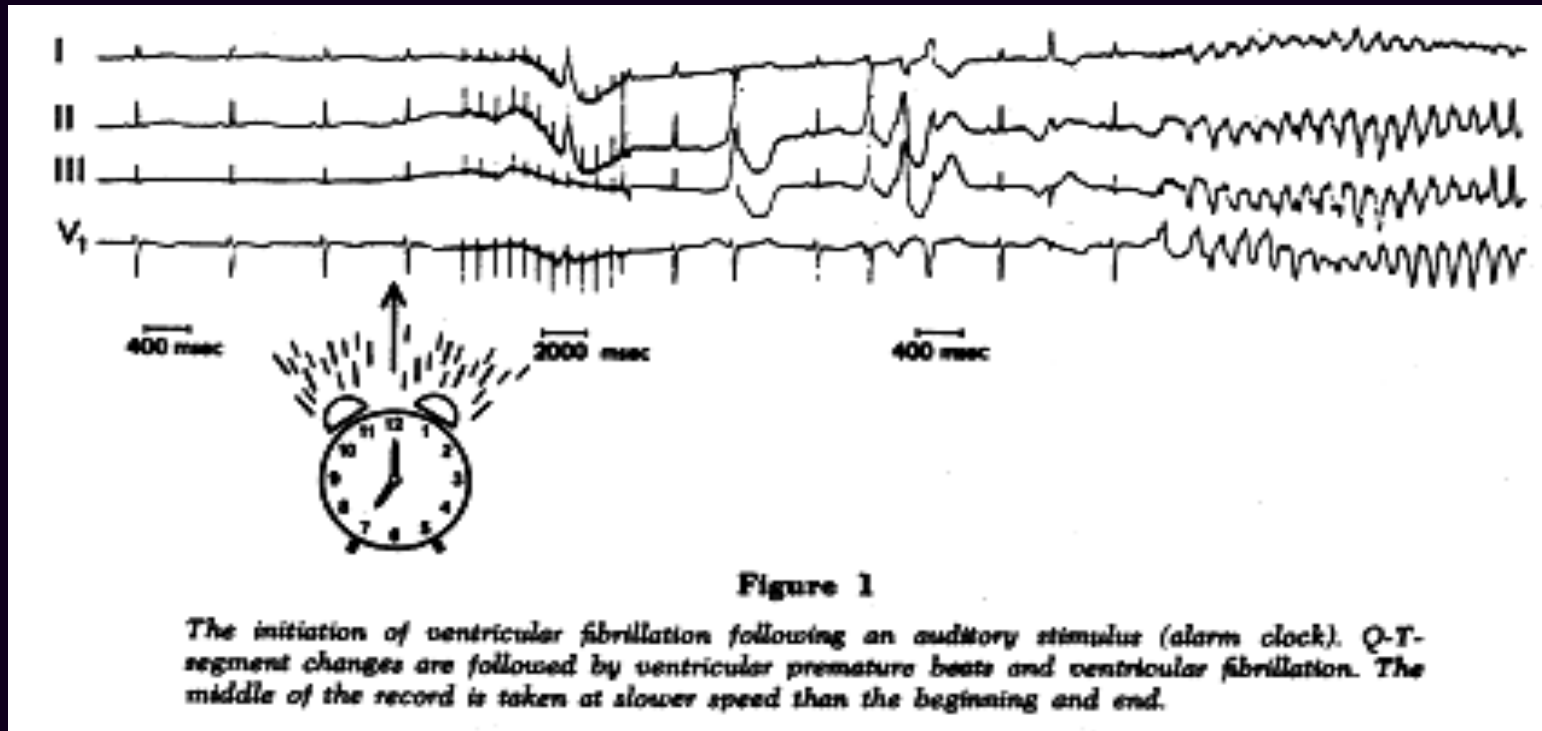
Normal ECG

What about the prolonged QT interval?!

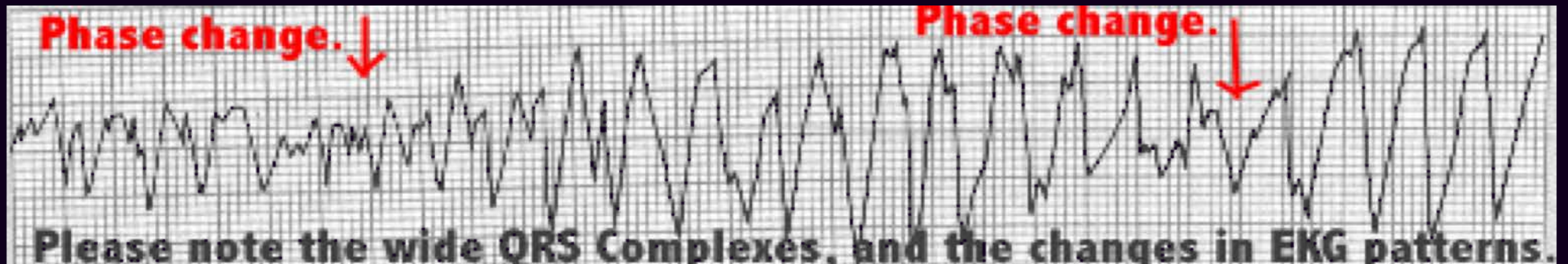


Long QT Syndrome

- Brought on by stress, anxiety, catecholamines



Torsade de Pointes



Long QT Syndrome

- **Congenital**

- Jervell-Lange-Nielsen - autosomal recessive
 - » associated with sensorineural hearing loss
- Romano-Ward - autosomal dominant
 - » associated with normal hearing
- Now characterized genetically as defects in cardiac ion channels (channelopathies)
 - » LQTS 1-15

- **Acquired**

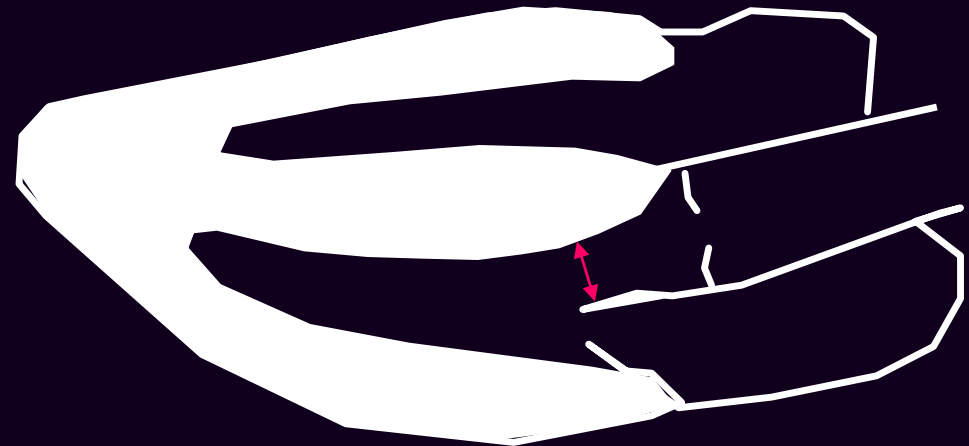
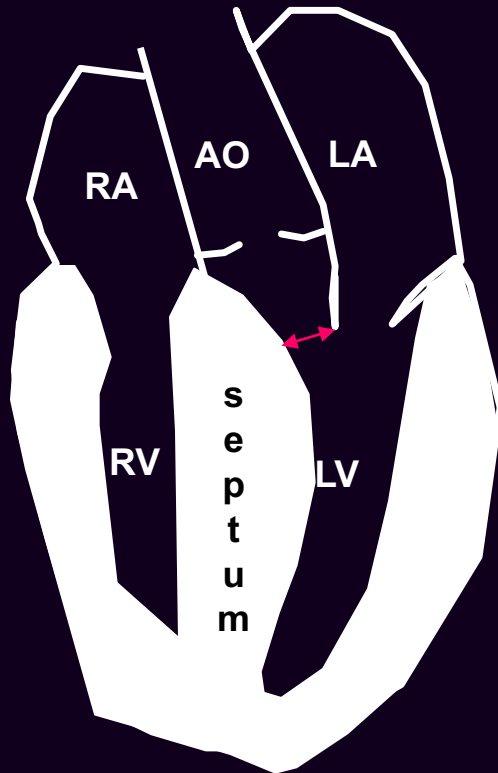
- medications
 - » tricyclic anti-depressants, ketoconazole
 - » erythromycin, TMP-SMX, famotidine
- <https://crediblemeds.org>

Hypertrophic Cardiomyopathy

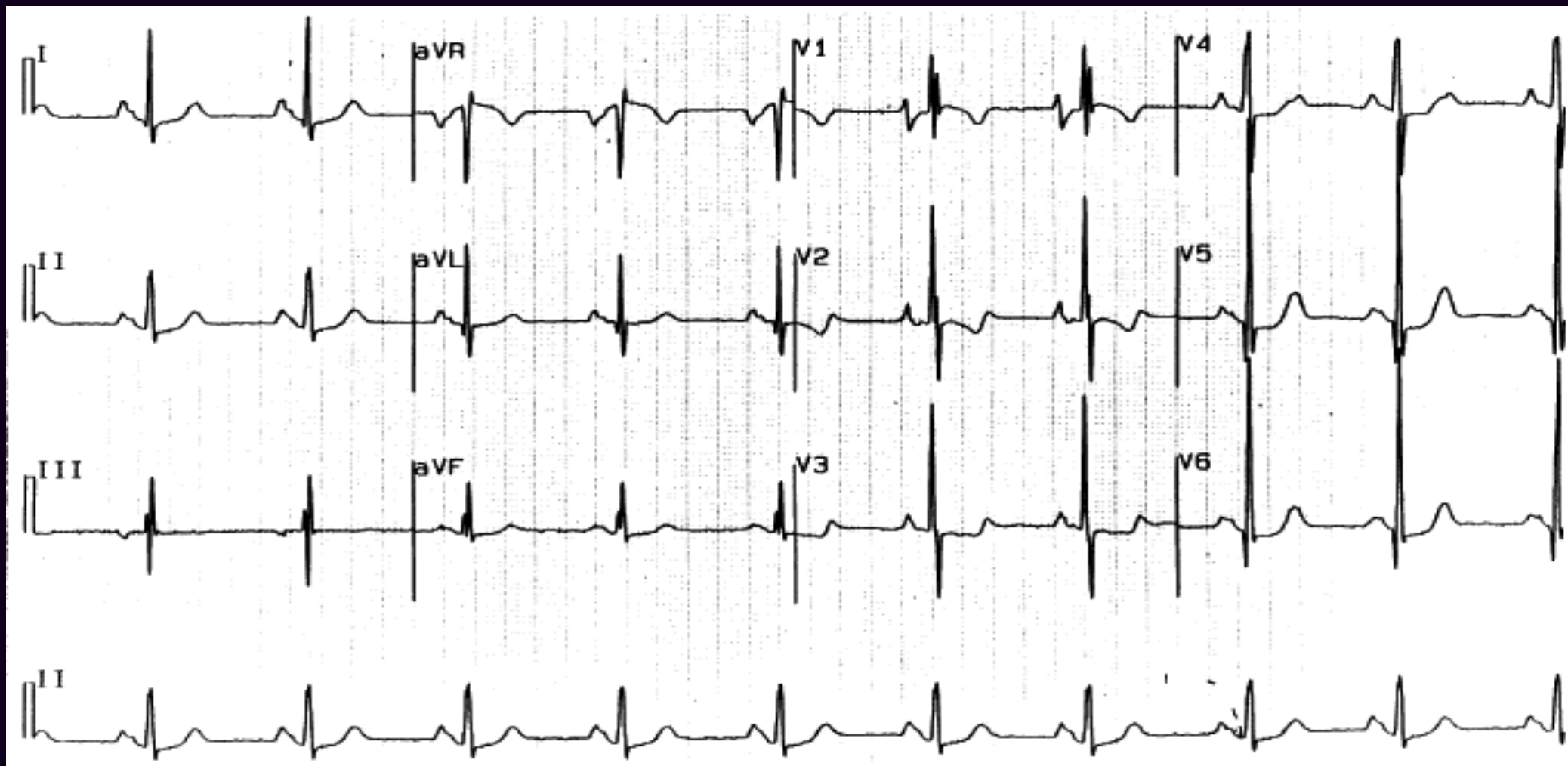
- abnormal hypertrophy of ventricular septum and LV free wall without extrinsic cause
- 60% inherited as autosomal dominant
- most common cause of sudden death among healthy young patients
- multiple genetic defects identified – involving sarcomeric proteins of heart
- physical exam – S4 louder when upright

Hypertrophic cardiomyopathy

- Murmur of HCM: louder when upright
 - Supine: increased venous return → increased end diastolic volume
 - Increased EDV → less stenosis, softer murmur



Hypertrophic Cardiomyopathy ECG



Risks factors for HCM

- **Genetics – family history of sudden death**
- **Severe septal hypertrophy**
- **Severe LV outflow obstruction**
- **Blunted BP response on exercise stress test**
- **Ventricular ectopy on holter**

Approach to the Child with Irregular Heart Rate

- **History**
 - **symptoms**
 - » **palpitations, syncope, CHF**
 - » **h/o cardiac surgery**
 - **medications**
 - » **bronchodilators**
 - **family history**
 - » **arrhythmias**
 - » **syncope, sudden death**

Approach to the Child with Irregular Heart Rate

- **Physical Exam**
 - **Cardiac Auscultation**
 - » r/o underlying heart defect
 - » ectopy suppressed with exercise
 - assess by performing 25 jumping jacks
- **ECG**
 - diagnose arrhythmia
 - r/o Long QT Syndrome, HCM
- **24° Holter**
 - longer period of recording
 - quantitate ectopy
 - document response of ectopy to increased HR

Who to Refer

- **suspicious symptoms**
 - palpitations, exercise intolerance, syncope
- **abnormal cardiac exam**
- **multiform PVCs**
- **ventricular couplets or worse**
- **ectopy not suppressed by exercise**
- **positive family history**

