

Sudden Cardiac Death in Pediatric Population

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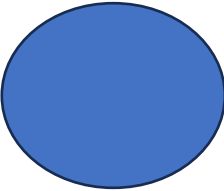
Definition

- Sudden unexplained death in an otherwise healthy person within 1 hour of symptom onset
- SCD in pediatric population
 - Devastating and tragic for both the family and the community
 - Dramatic in presentation
 - Highly publicized with news reports and social media coverage
 - Rare event

Incidence

- 0.7 to 6.4 per 100,000 patient-years
- Relative risk of SCD is 2.5 to 4.5 higher in athletes
- Disproportionate increased risk in male/black athletes
- Vast majority have no prior history of cardiac disease

SUBSET OF POPULATION
AT RISK OF SCD



SYMPTOMATIC
Exertional palpitations
Exertional chest pain



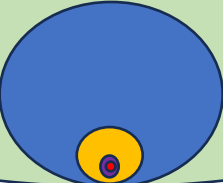
CARDIAC SYNCOPE



SCA
Resuscitated successfully on
many occasions



SCD

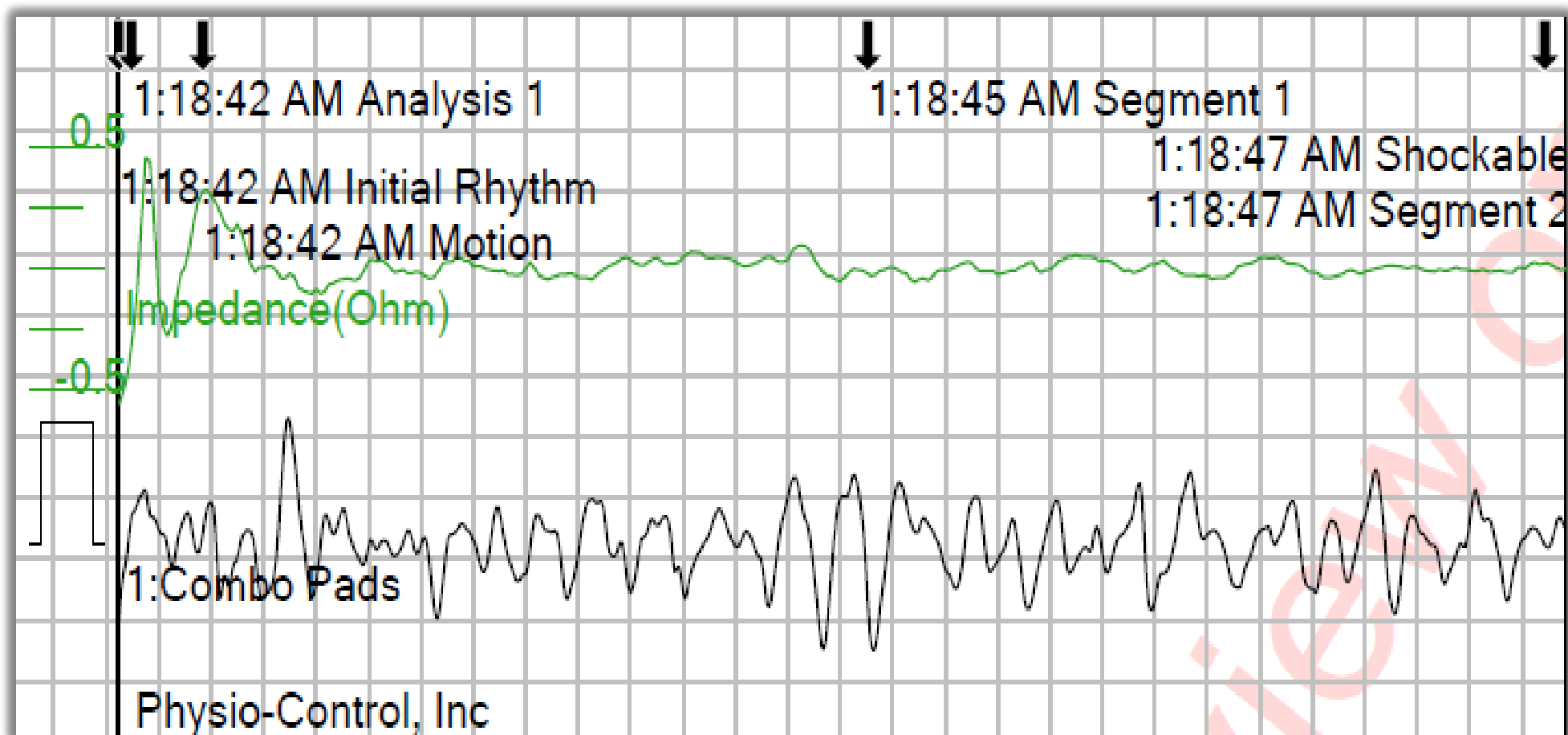


COMMON CAUSES OF SUDDEN CARDIAC DEATH IN A YOUNG PATIENT

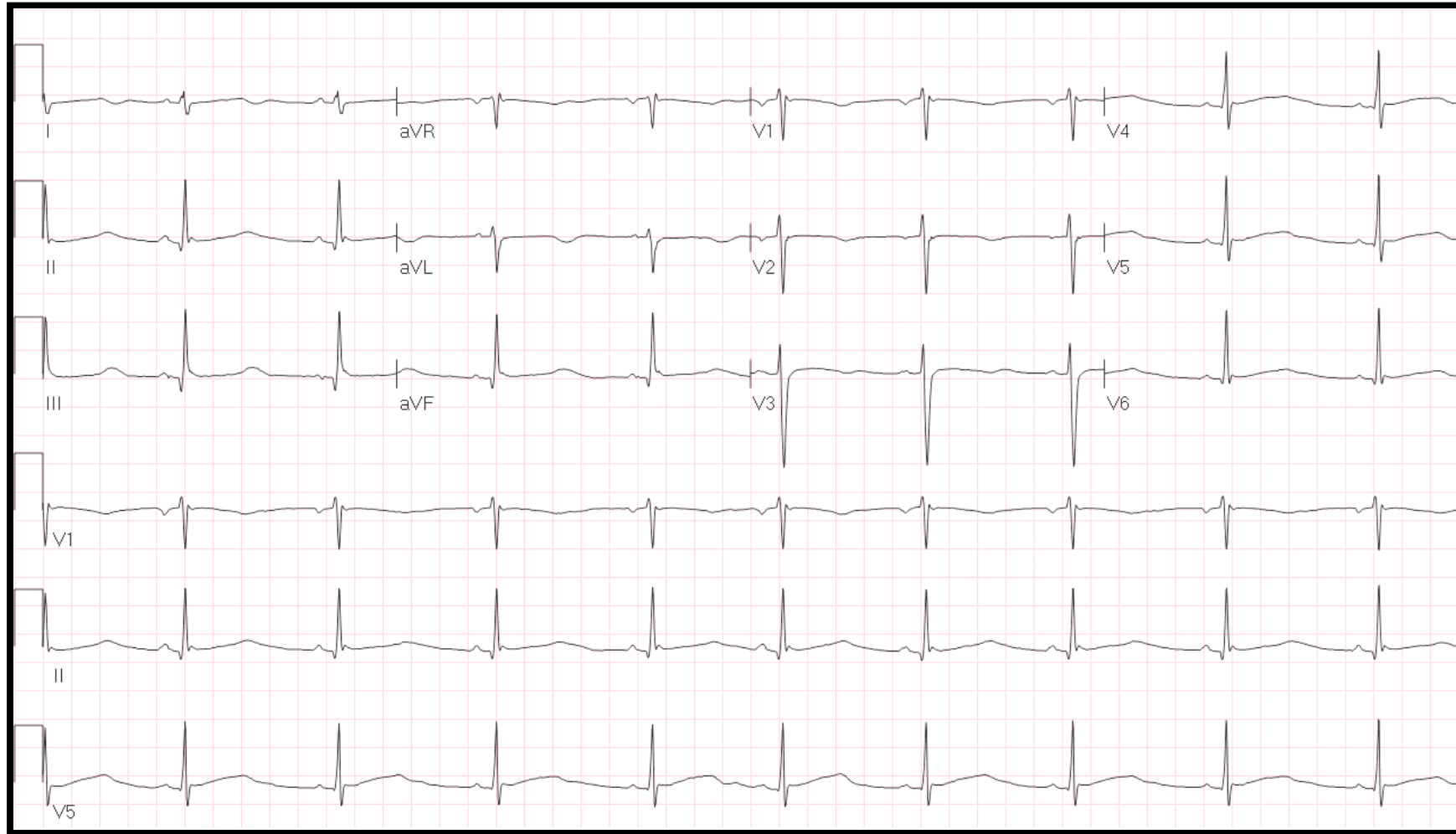
16-Year-Old Volleyball Player

- Was participating in trials and reported not feeling well
- Suddenly fell to the ground and appeared to be seizing
- She appeared dusky with no respiration and no pulse
- 911 was called and CPR started by the coaches
- EMS found her in ventricular fibrillation (V fib) and successfully defibrillated

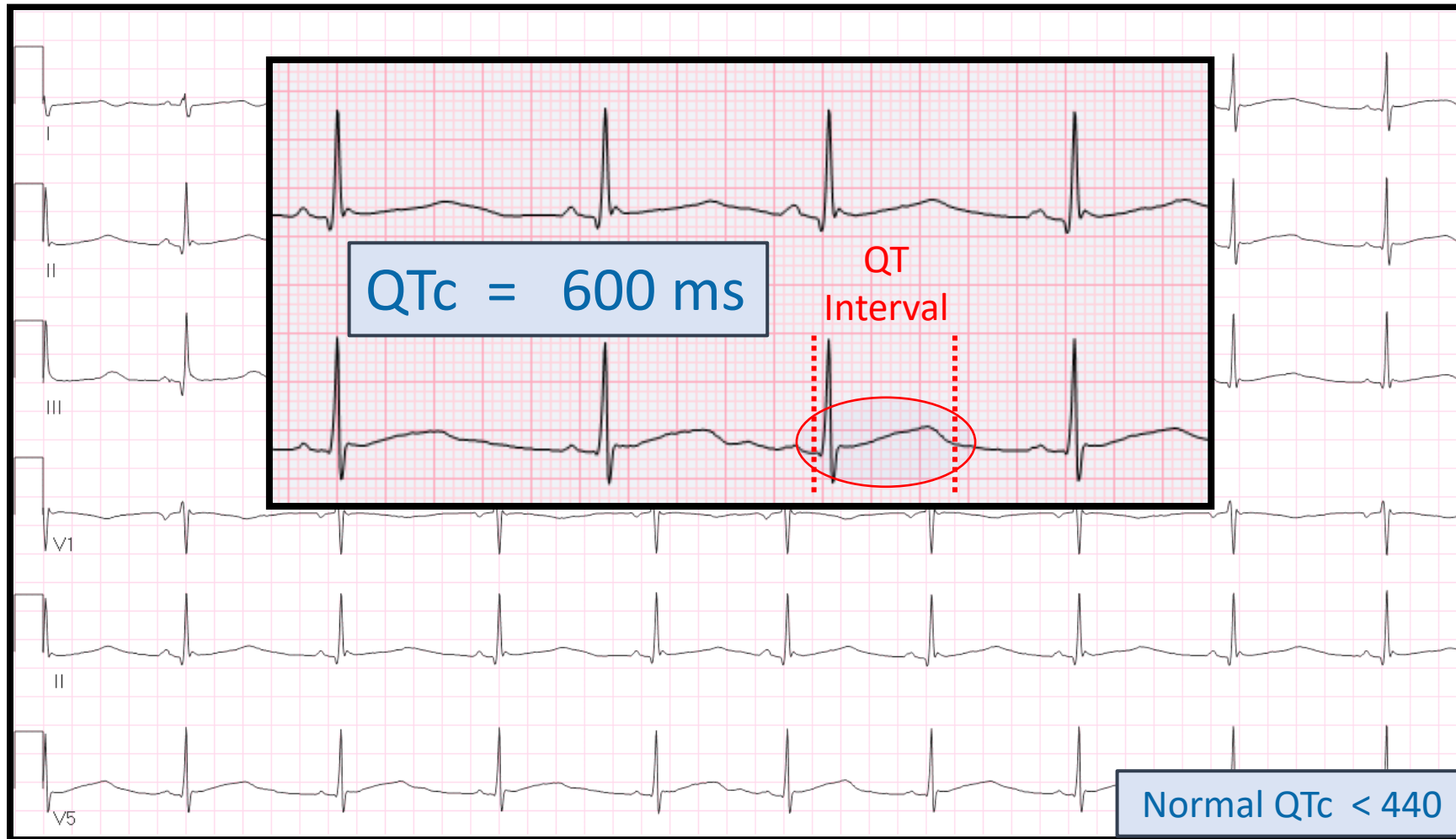
16-Year-Old Volleyball Player



ECG



ECG



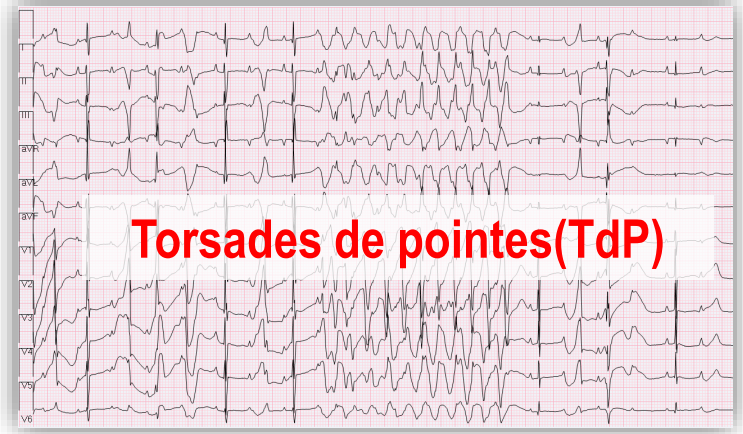
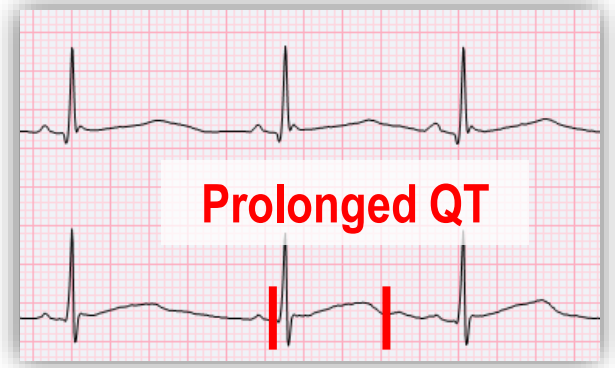
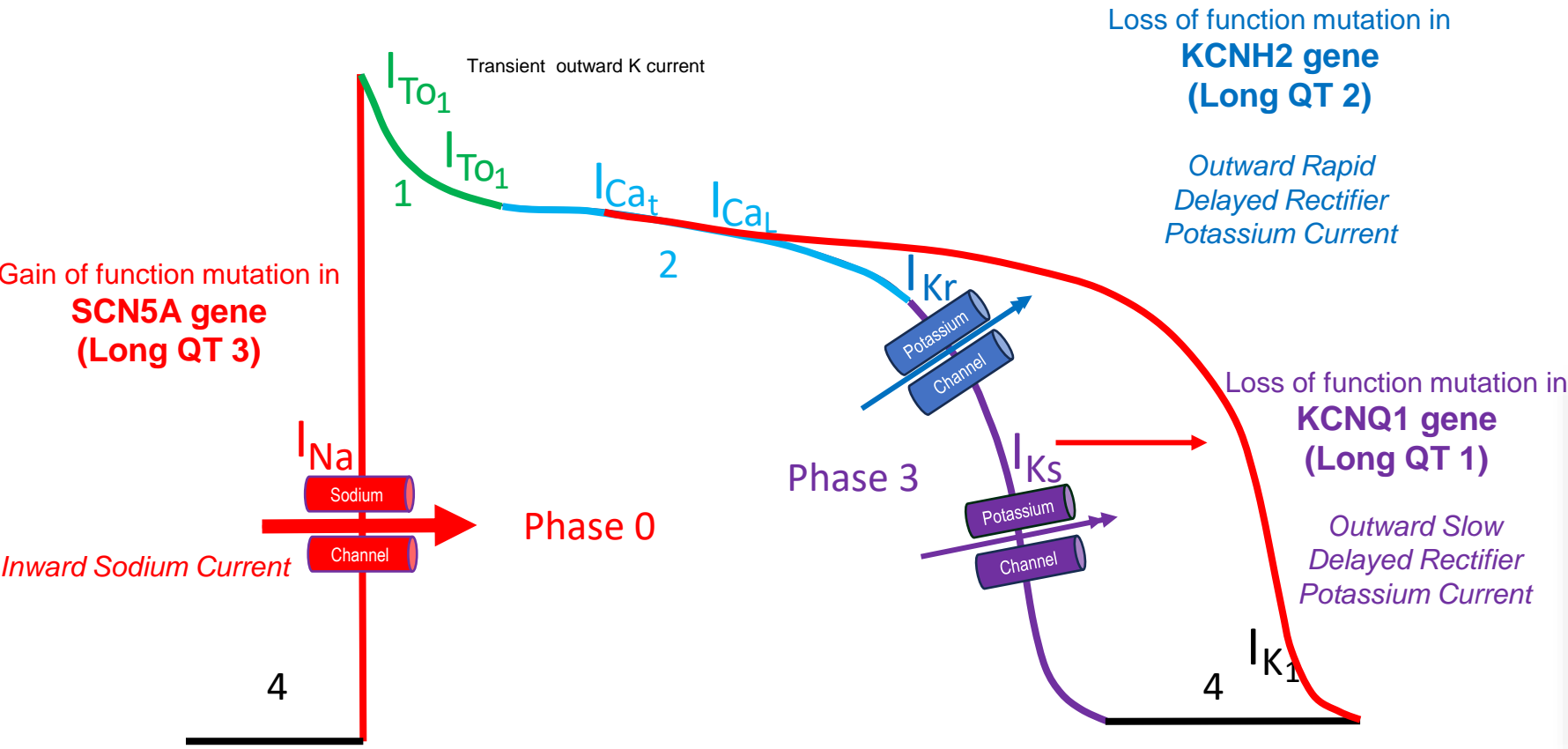
16-Year-Old Volleyball Player

- Interestingly she had similar episode the year prior
- That episode also occurred while playing volleyball
- However, she had regained consciousness spontaneously
- Taken to a local ED and diagnosed with vasovagal syncope
- ECG was performed and cardiology evaluation suggested
- No sports restrictions applied

Congenital Long QT Syndrome (LQTS) (Channelopathy)

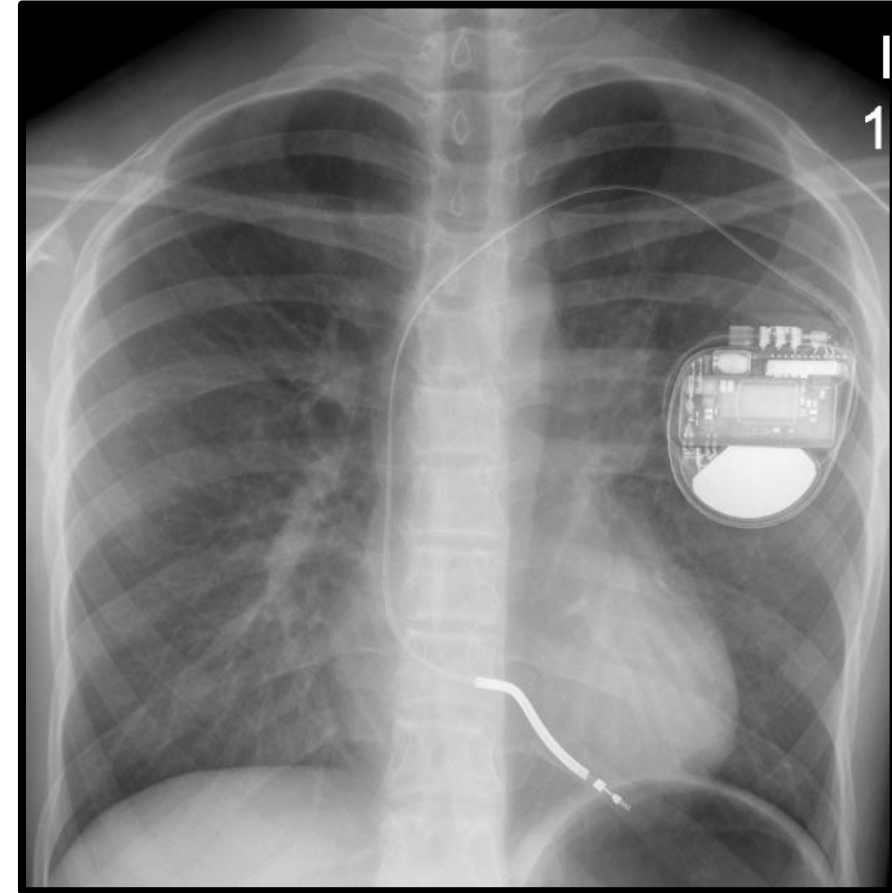
- LQTS is a potentially lethal, inherited ventricular arrhythmia syndrome
- Estimated to affect 1 in 2000 individuals
- Hallmark ECG feature is QT prolongation
- May present as: syncope, seizure or cardiac arrest
- Event triggers are mostly adrenergic arousal:
 - Rigorous physical activity
 - Intense emotions
 - Swimming (LQT1)
 - Sudden loud noise(LQT2)

Genetically Mediated: Mostly Autosomal Dominant

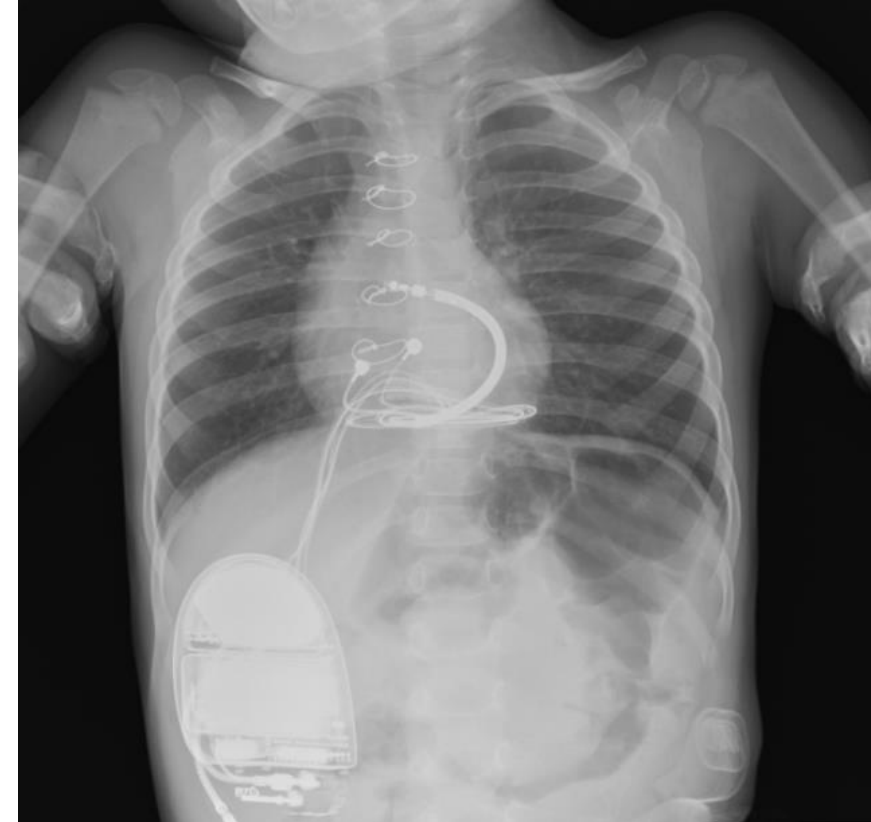
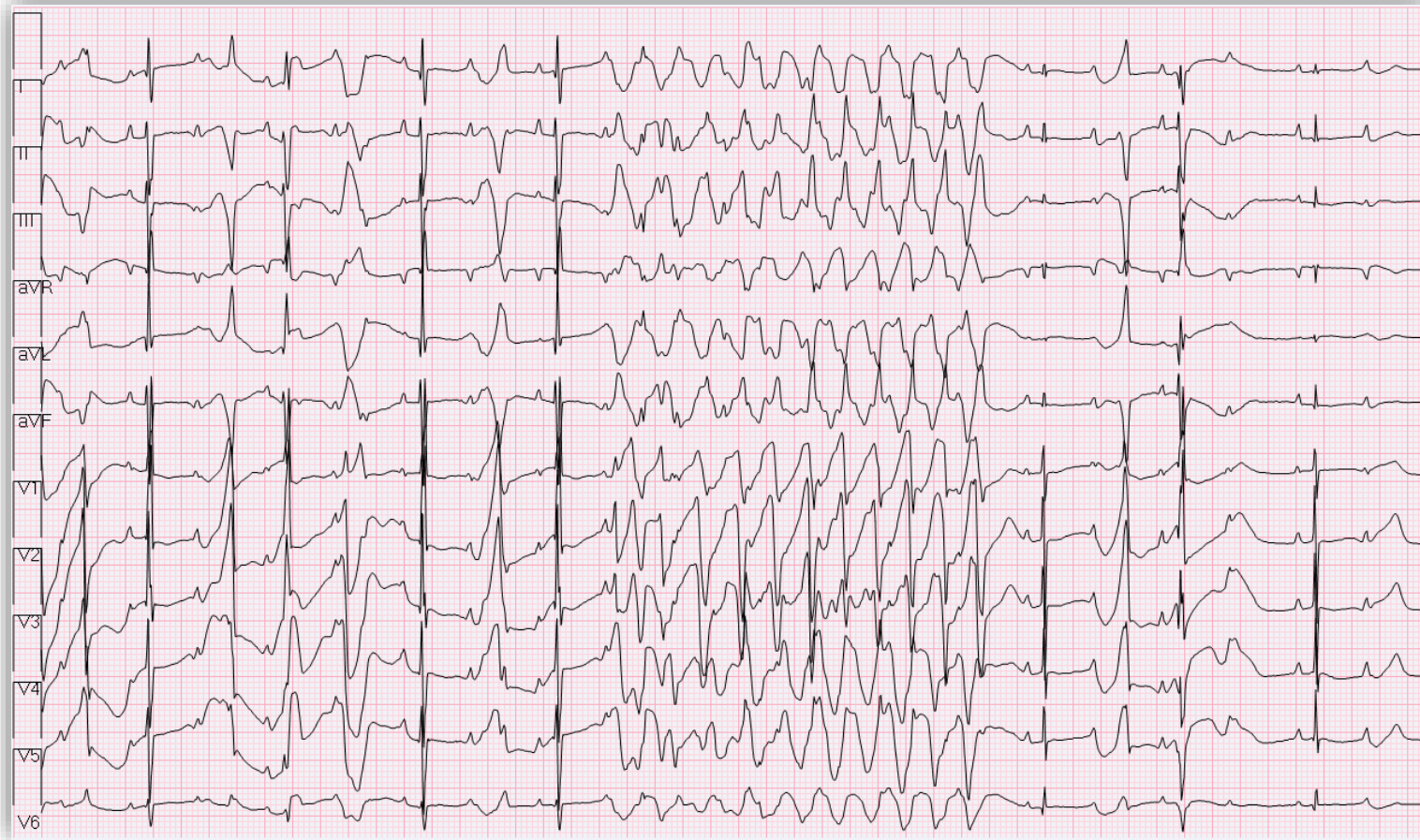


Treatment

- Beta-blockers
- Mexiletine(sodium channel blocker) for LQT3
- Avoidance of triggers
- Avoidance of QT prolonging medications
(qtdrugs.org/crediblemeds.org)
- Left sympathetic ganglionectomy
- ICD



Treatment



Siblings of 16-Year-Old Volleyball Player

- Long QT syndrome is a familial condition
- Evaluated the siblings
- Two siblings had the same mutation with prolonged QT
- Beta blocker started

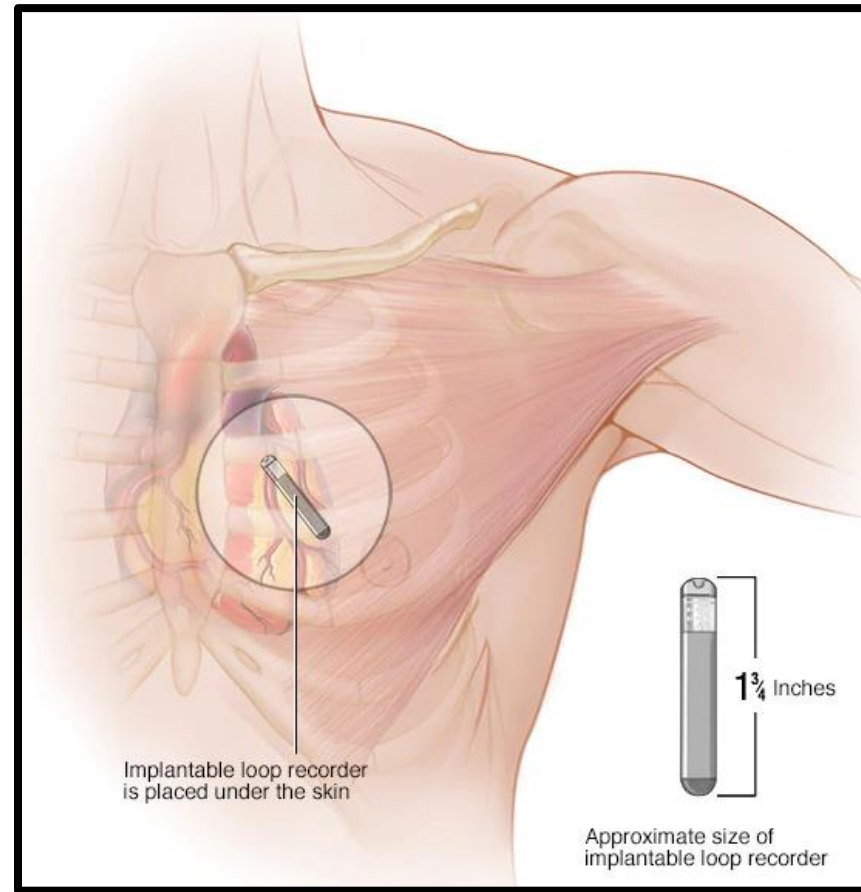
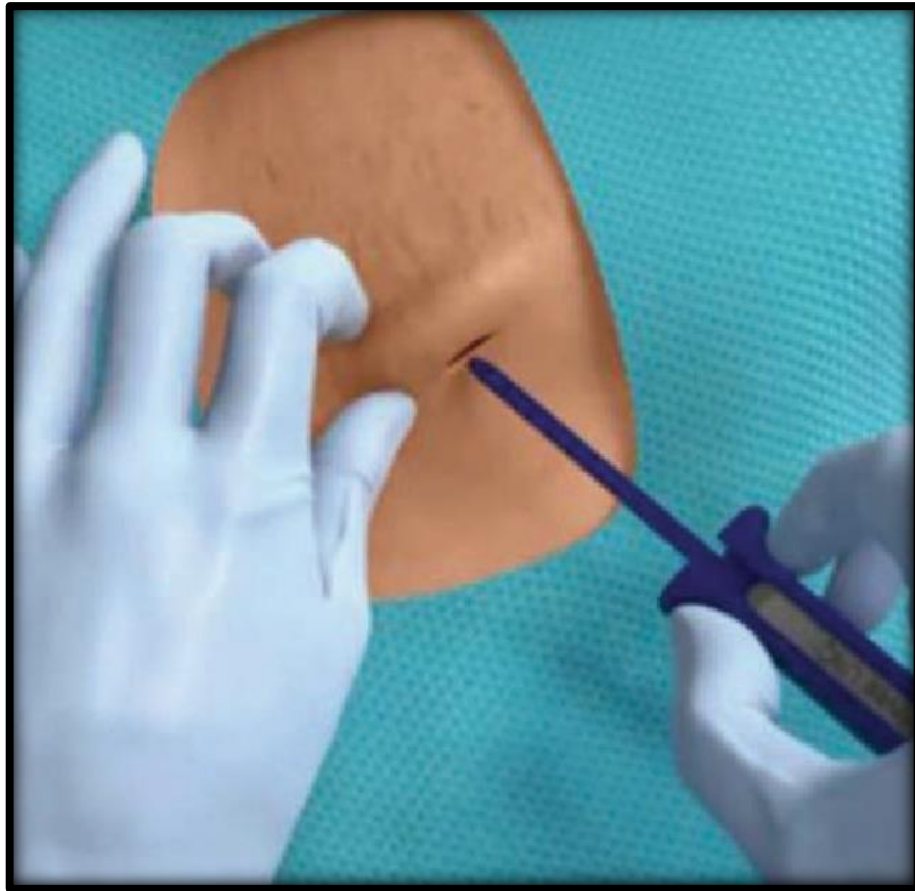
Sibling of 16-Year-Old Volleyball Player

- Past h/o syncope : while standing in the church
- 3 years after the diagnosis she had another episode of syncope
 - she got up to go to bathroom after a nap
 - felt lightheaded before passing out
 - and denies any palpitations
- Would you change your management?

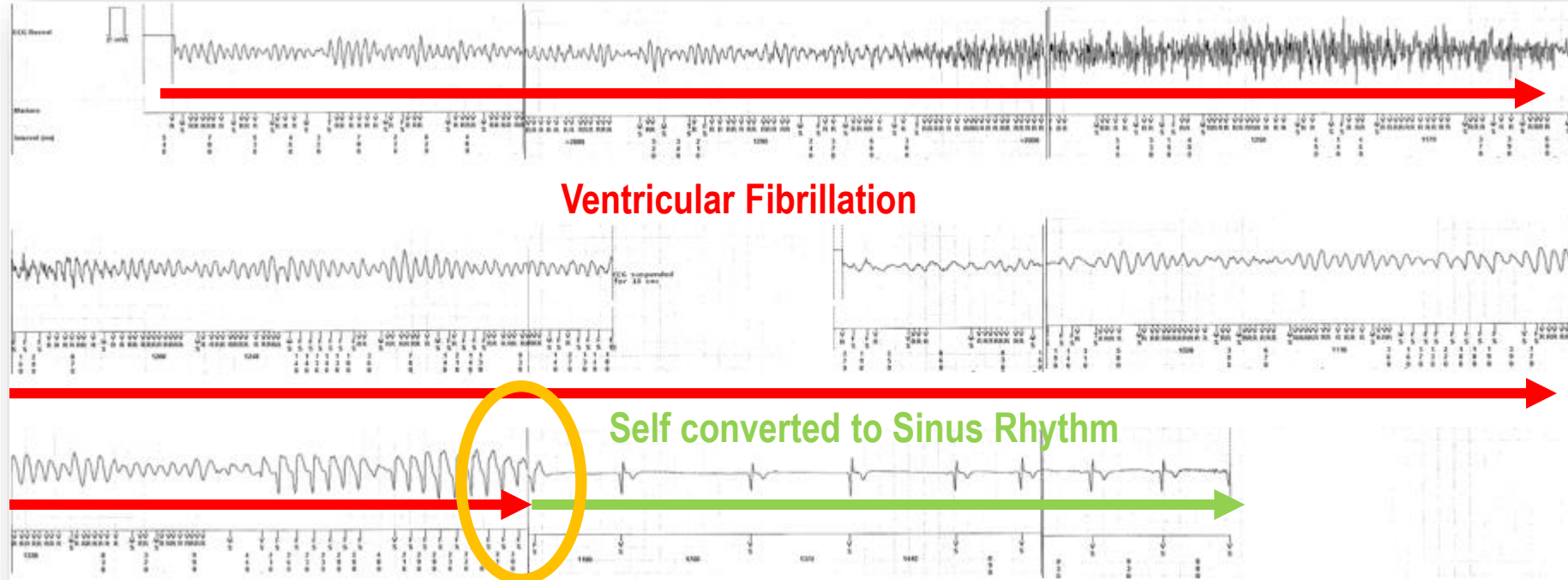
15-year-old with Seizures/Syncope

- Previously seen by neurology and referred to cardiology
- Episodes were rare: occurred once a year
- Triggered mostly by anxiety and while playing
- Normal cardiac evaluation, normal ECG and echo
- One family member died of heart attack at “21 y of age”
- History was suspicious : exercise/anxiety as the triggers
- Implantable loop recorder (ILR) was implanted

15-year-old with Seizures/Syncope



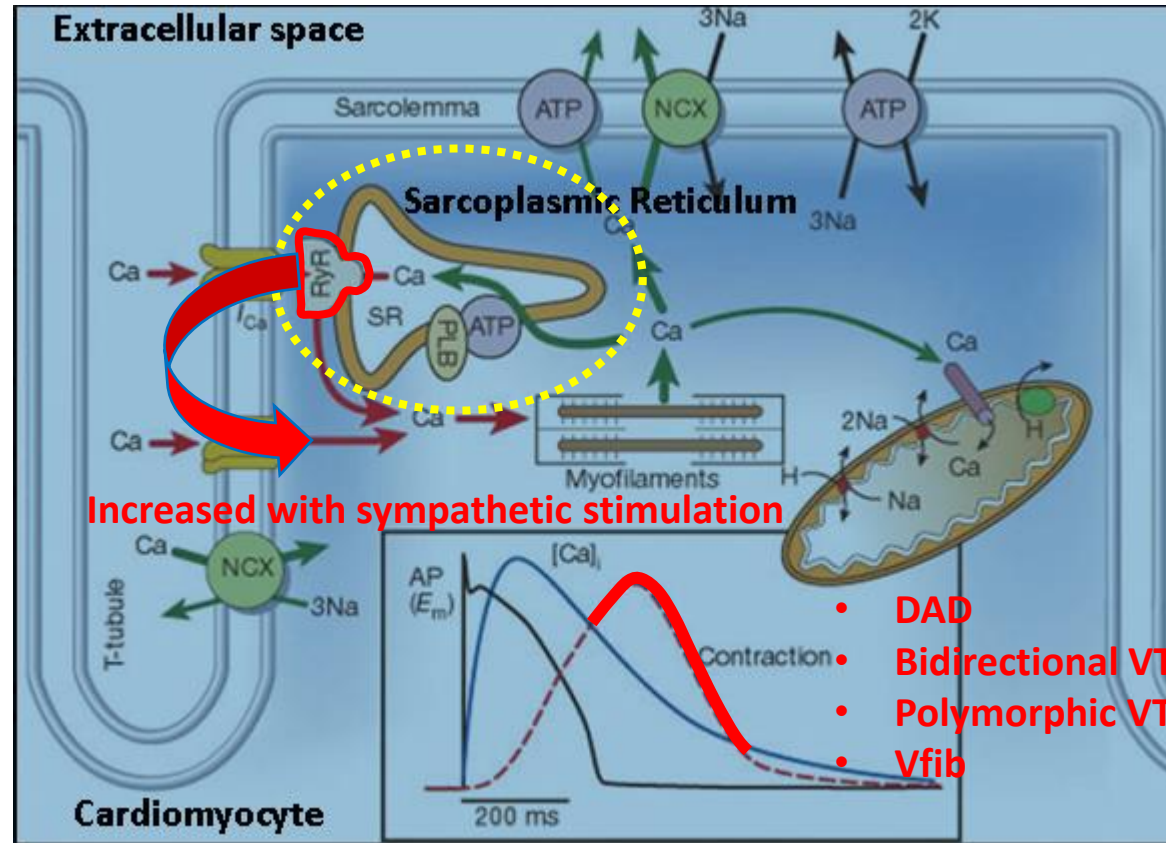
15-year-old with Seizure /Syncope



Catecholaminergic Polymorphic Ventricular Tachycardia (CPVT) :Channelopathy

- Genetically mediated: mostly autosomal dominant
- Ryanodine receptor mutation
- Syncope/SCA triggered by exercise or emotional stress
- Most events occur during first and second decade
- **Baseline ECG , echo and exam is normal**
- Stress test is important diagnostic tool
- Treatment : activity restriction, beta-blockers and flecainide
- ICD

Catecholaminergic Polymorphic Ventricular Tachycardia (CPVT) :Channelopathy



POSITIVE - LIKELY PATHOGENIC VARIANT

Result	Gene	Zygoty	Coding DNA	Variant	Ref. Seq.
PRESENT	RYR2	Heterozygous	c.13528 G>T	p.Ala4510Ser (A4510S)	NM_001035.2

14 y old who collapses on the field after scoring the winning goal in field hockey

- Resuscitated on the field by the coaches and nurses for 30 min before EMS arrived
- Shockable rhythm detected and AED used to convert to sinus rhythm

14 y old who collapses on the field after scoring the winning goal in field hockey

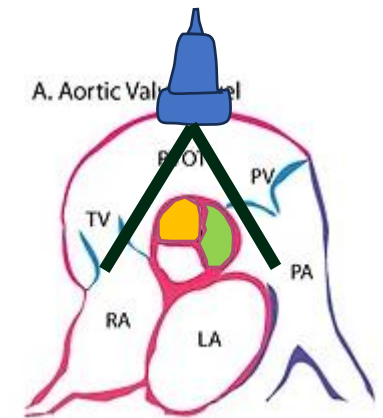
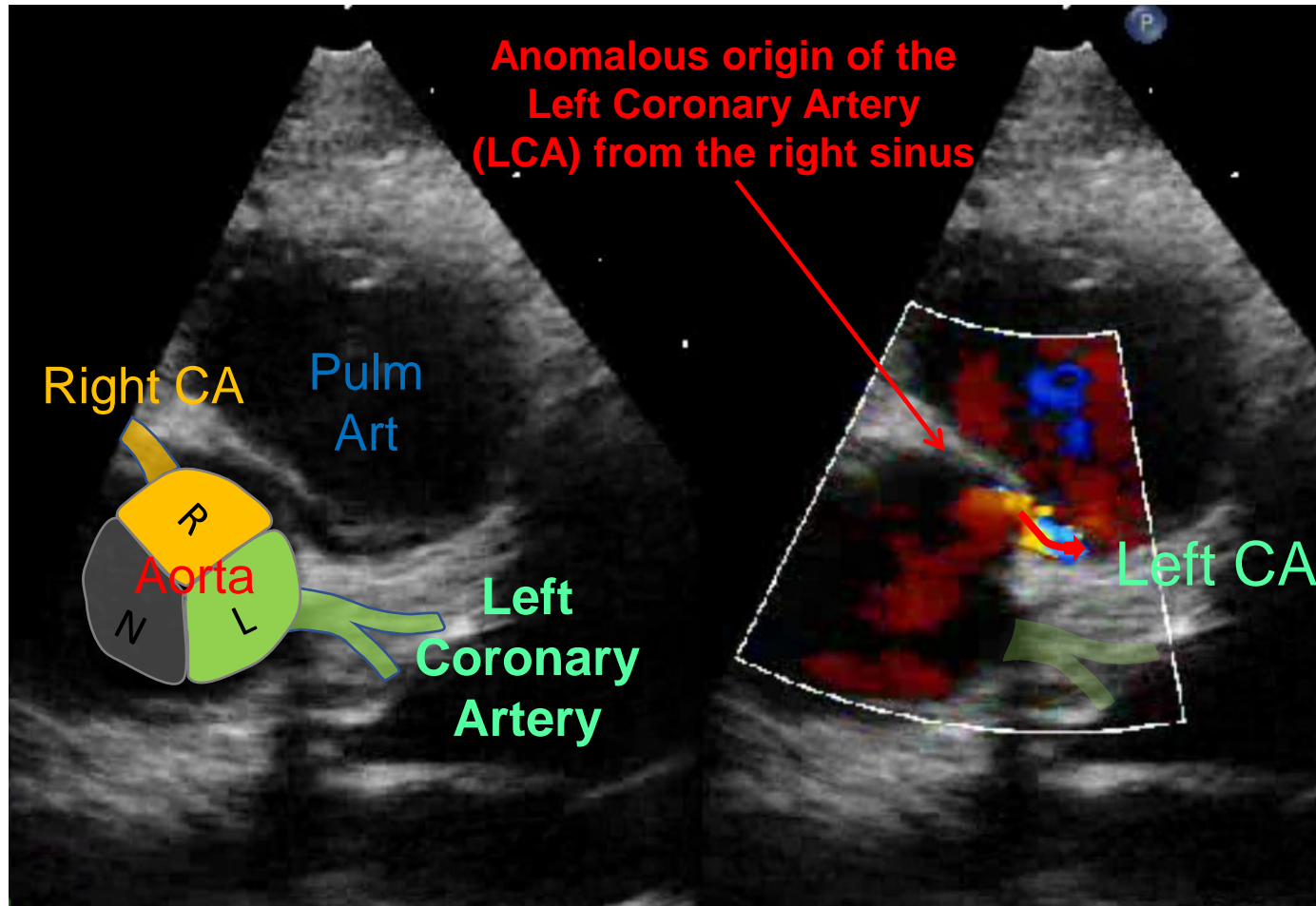
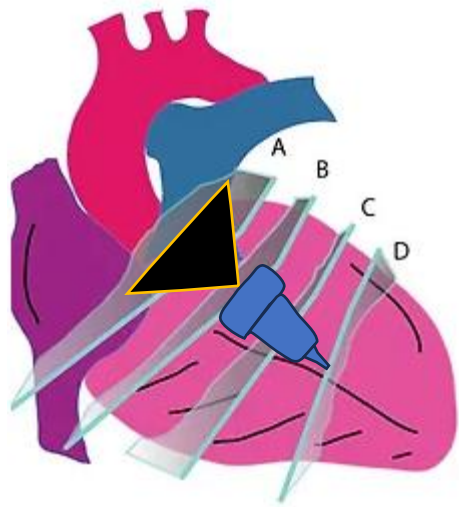


14 y old field hockey player

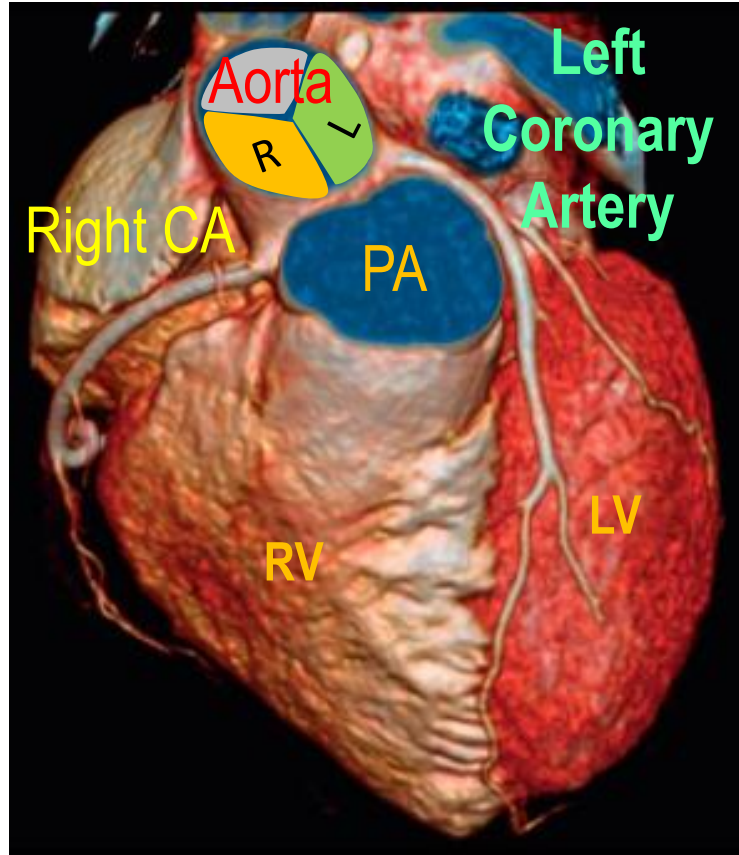
- Fortunately, she recovers well without any neurological insult
- On detailed questioning she admits to having symptoms of chest discomfort and near fainting while playing for the past few months
- Echocardiogram revealed.....

Anomalous origin of Coronary Artery from wrong Sinus of Valsalva as the cause of SCA

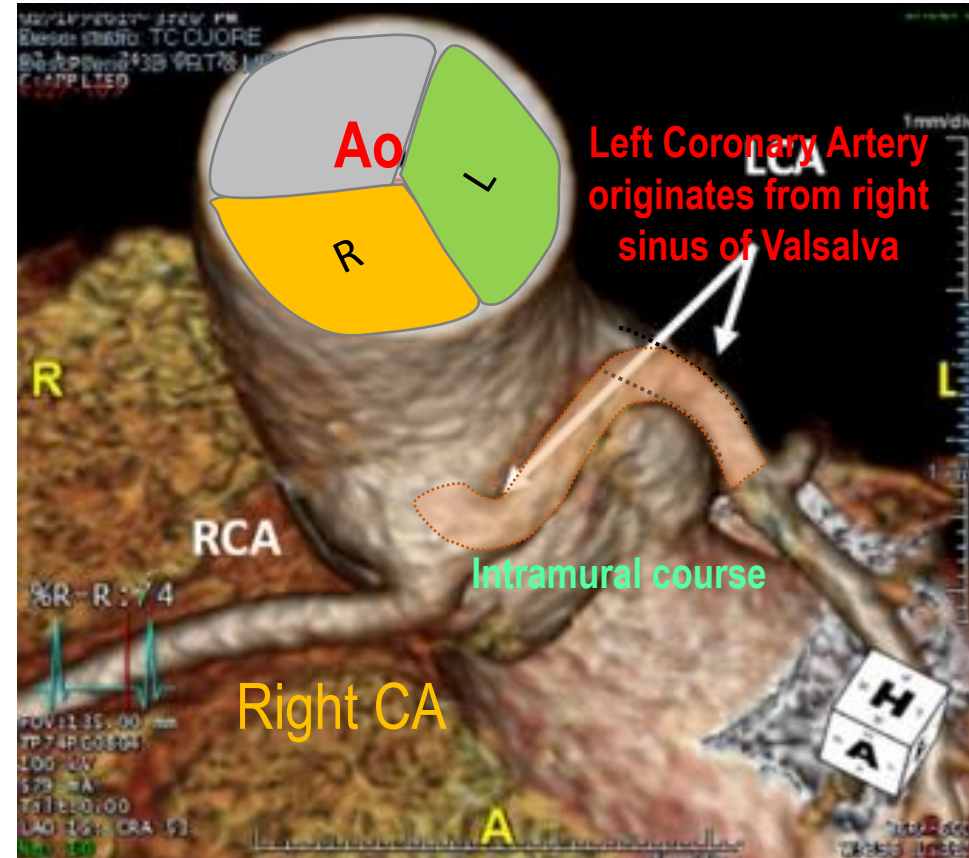
Parasternal Short Axis Views



Normal



Anomalous Origin of the Left CA



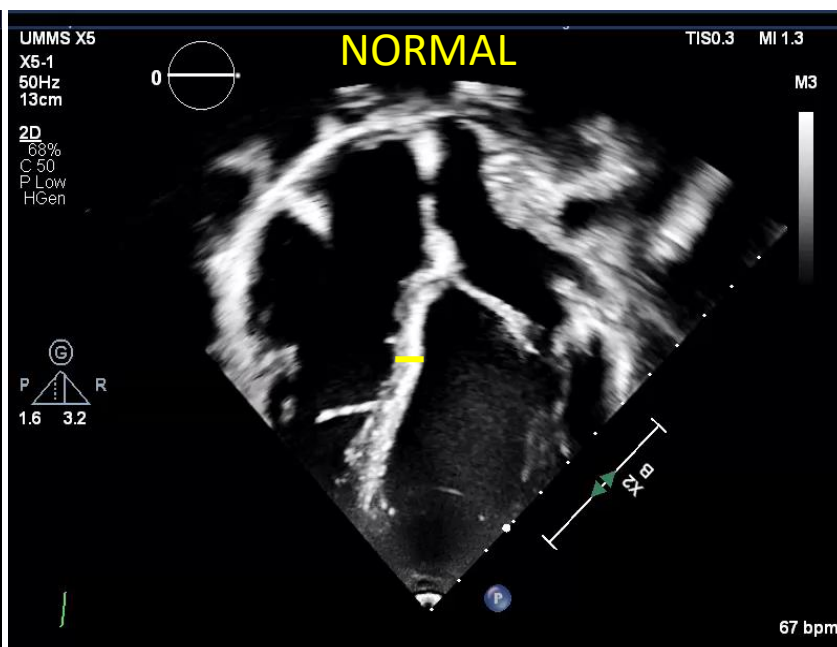
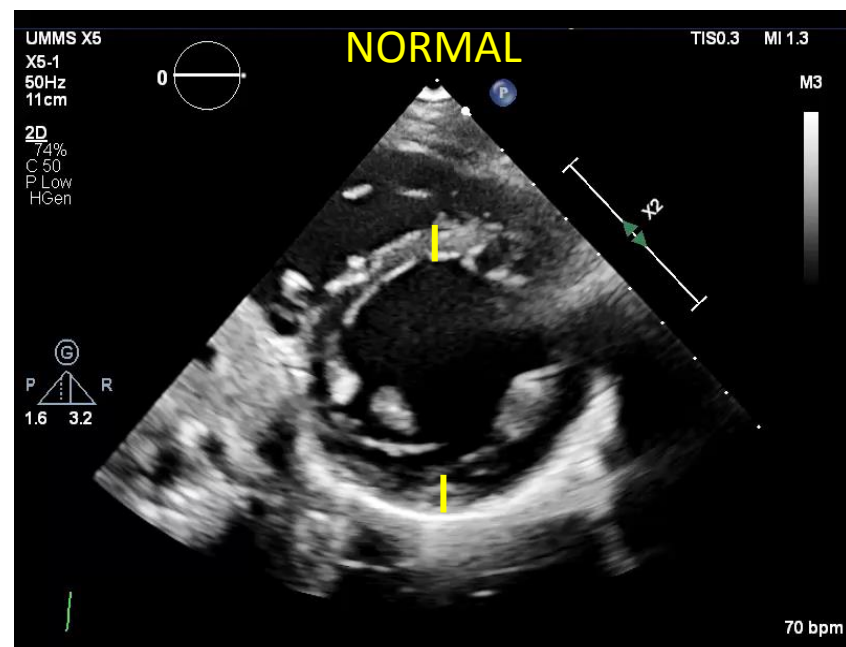
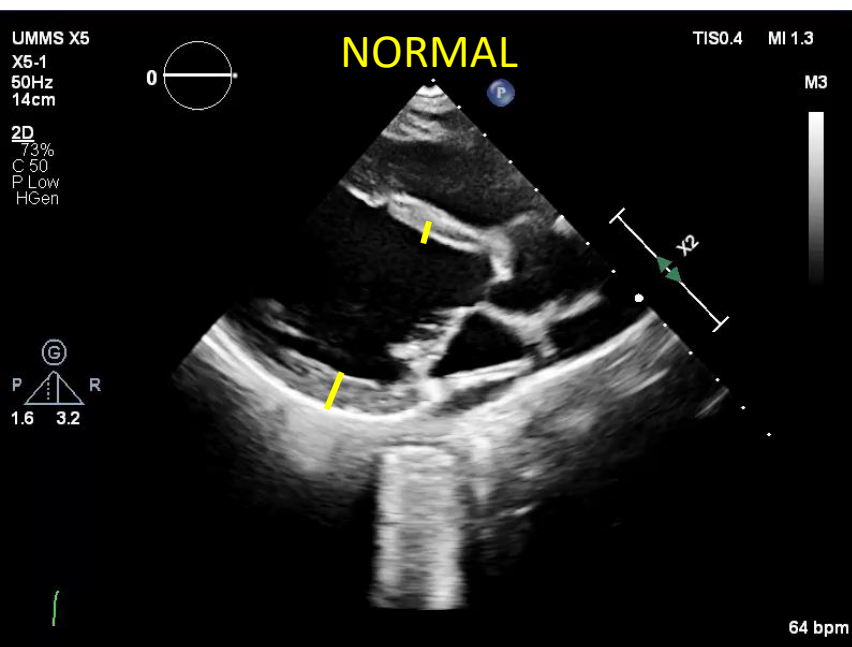
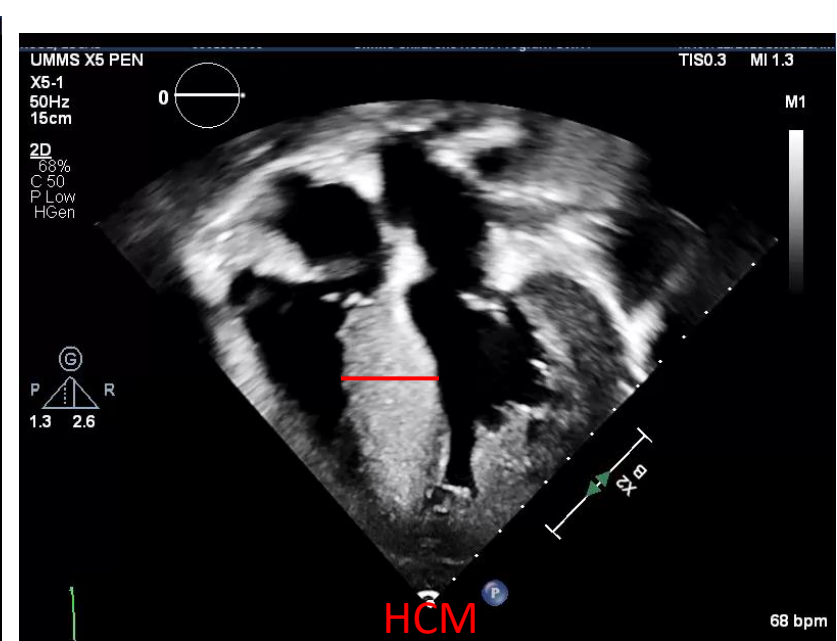
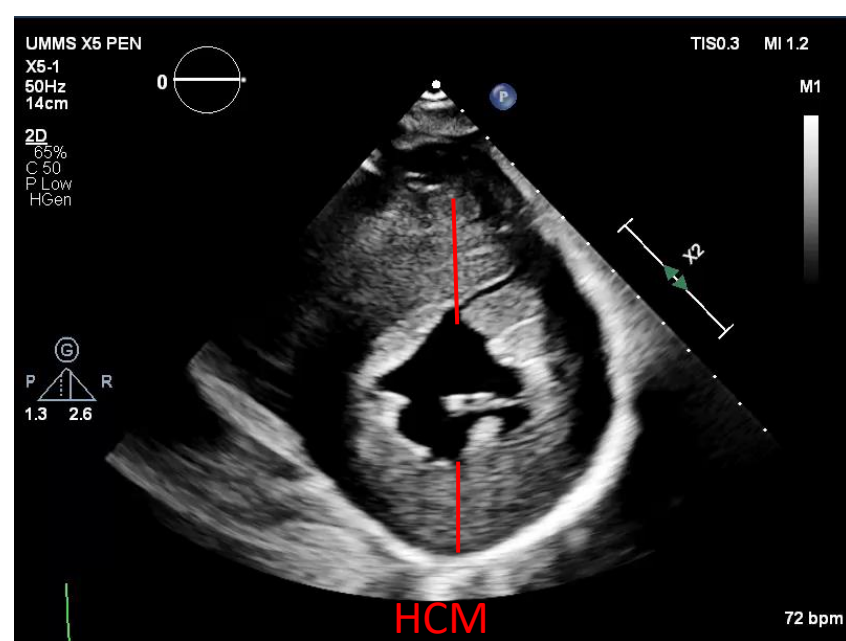
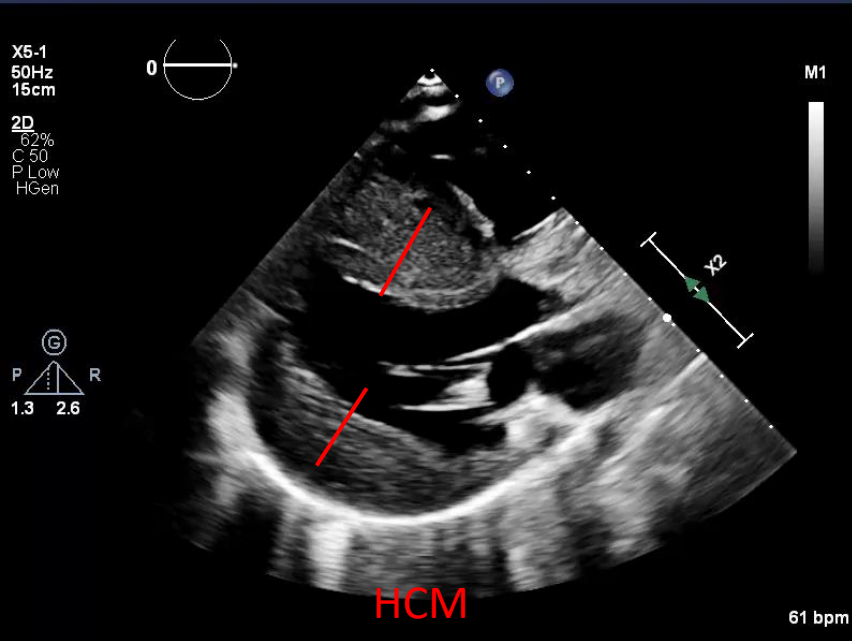
3D Reconstruction of Cardiac CT scan

Anomalous Origin of the Coronary Arteries

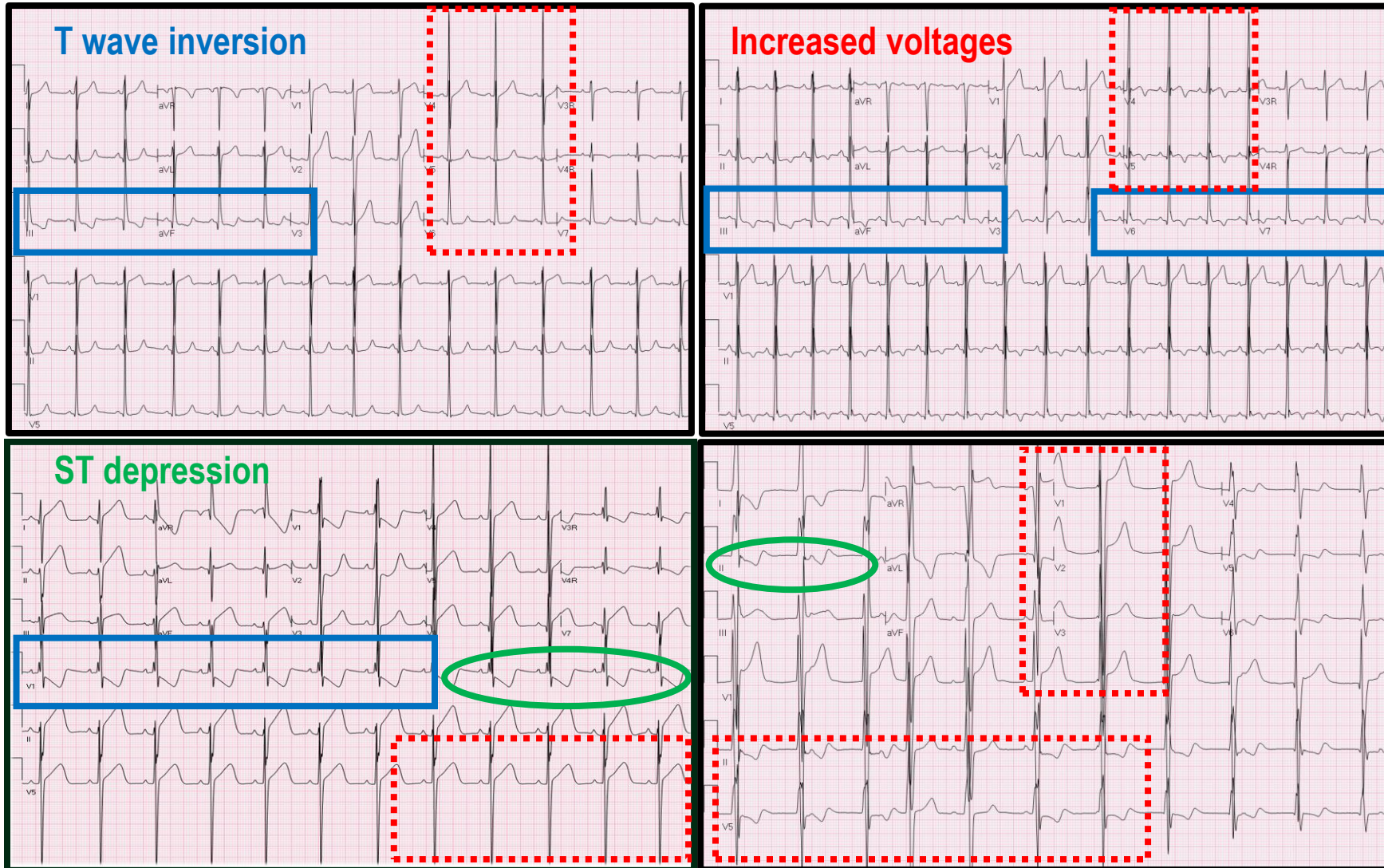
- Very rare: prevalence ~ 0.2 %
- This condition is not familial
- Likely culprits
 - Aberrant origin of LCA from right coronary cusp
 - Aberrant origin of RCA from left coronary cusp
- Hallmark: exertional symptoms i.e., syncope with exercise
- Baseline ECG is normal
- Echocardiogram could be diagnostic
- Surgical repair is definitive treatment

Hypertrophic Cardiomyopathy (HCM)

- 3-year-old initially evaluated for murmur
- Echocardiogram was performed : diagnosed with HCM
- He was playing hide and seek with his brother
- Grandfather found him unconscious in the backyard
- Family members had medical background and started CPR
- EMS arrived on time and defibrillated for Vfib



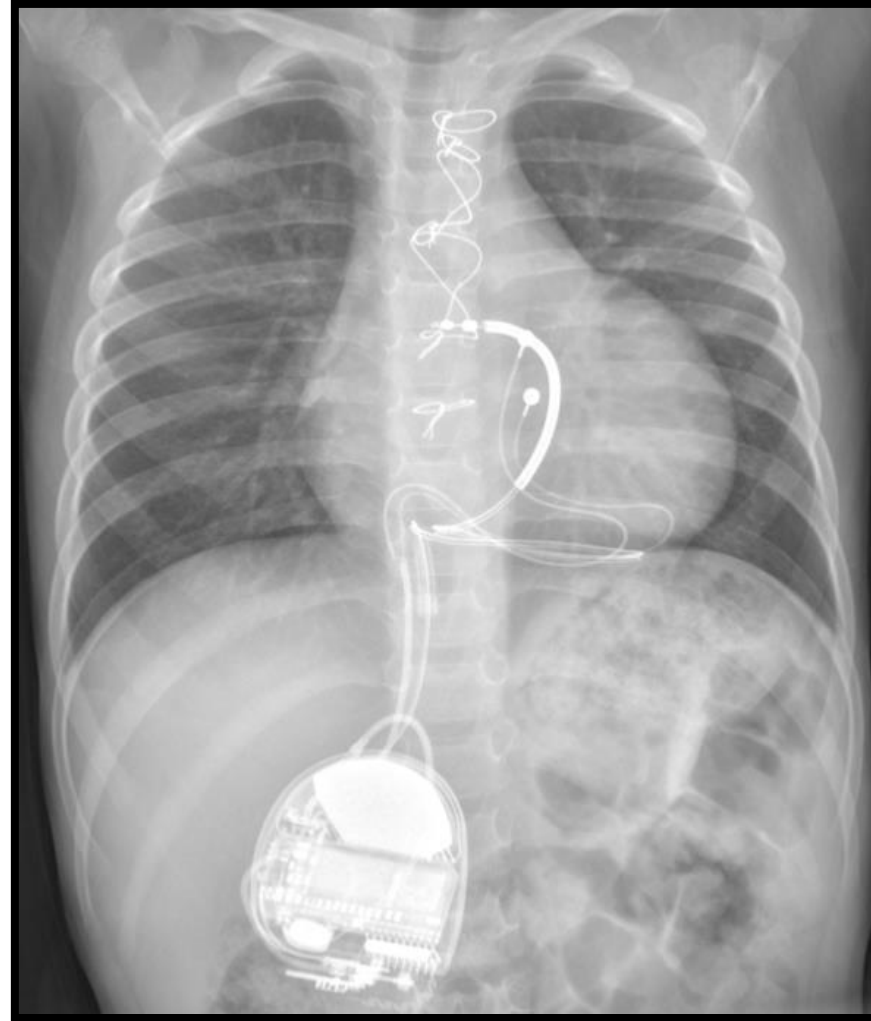
ECG is abnormal in > 90 % of patients in HCM



Hypertrophic Cardiomyopathy (HCM)

- One of the most common inherited heart disease (Autosomal Dominant)
- Prevalence of 1:300 to 1:500
- One of the most common cause of SCD in people < 30 y of age
- Most common cause of SCD in competitive athletes
- Overall annual incidence of SCD ~ 1%/year
- Highest risk subset of HCM patient have 5% -10% annual risk of SCD
- ICD is the only recommended therapy in the high-risk subset

Hypertrophic Cardiomyopathy (HCM)

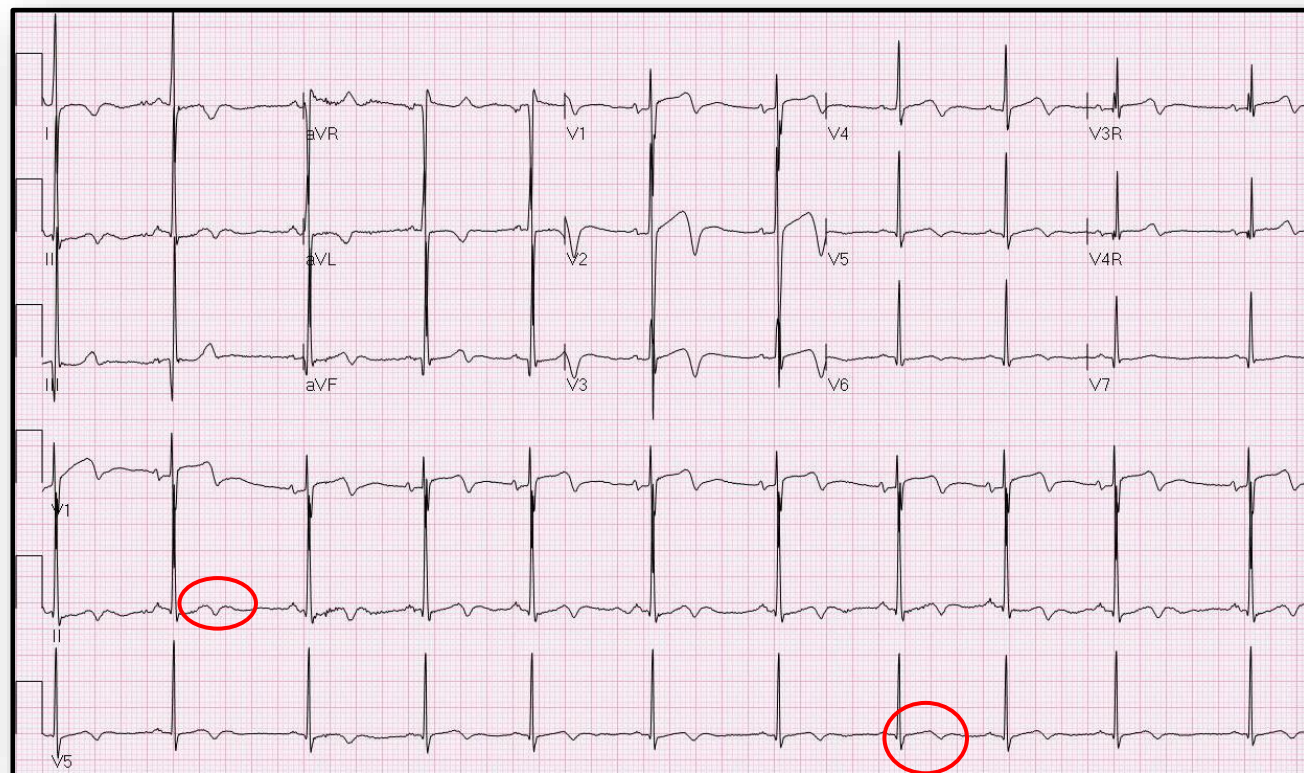


10 y old evaluated for syncope during soccer game

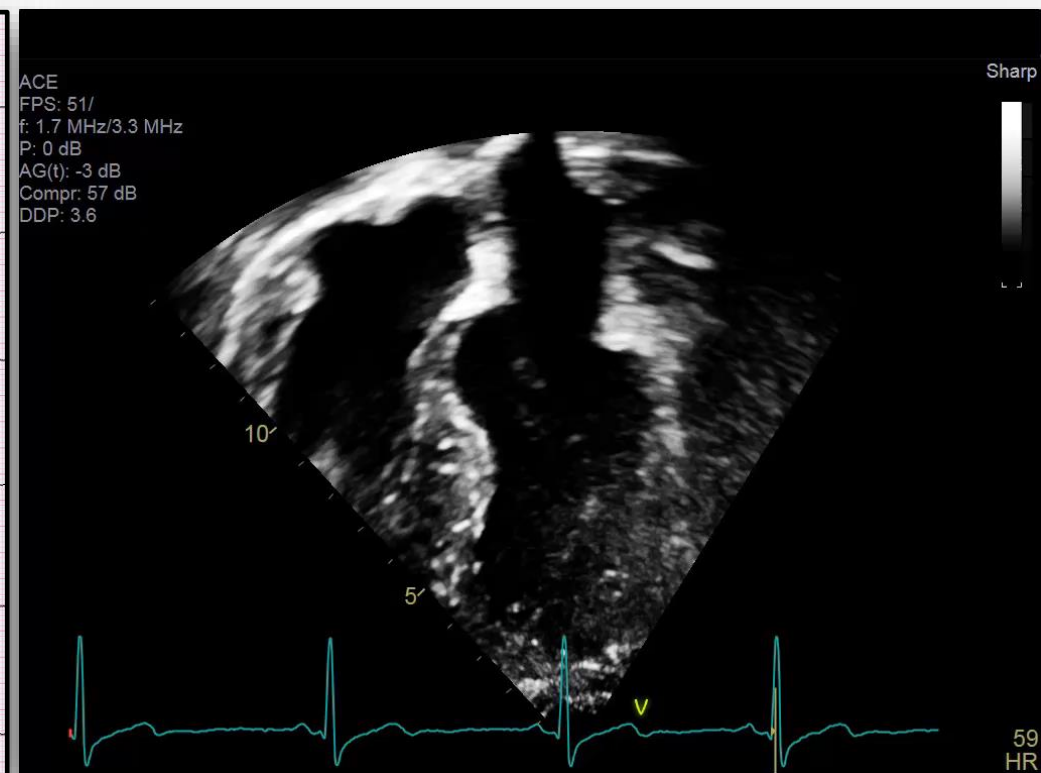
- Just finished drinking water during a scheduled break
- As he was getting ready to go back to play, someone saw him falling backwards
- Regained consciousness right away
- Recalls his vision going dark
- Denies any chest pain or palpitations before passing out
- Interestingly he had gone to the school nurse a week prior for dizziness
- History of maternal uncle dying suddenly at a young age

10 y old evaluated for syncope during soccer game

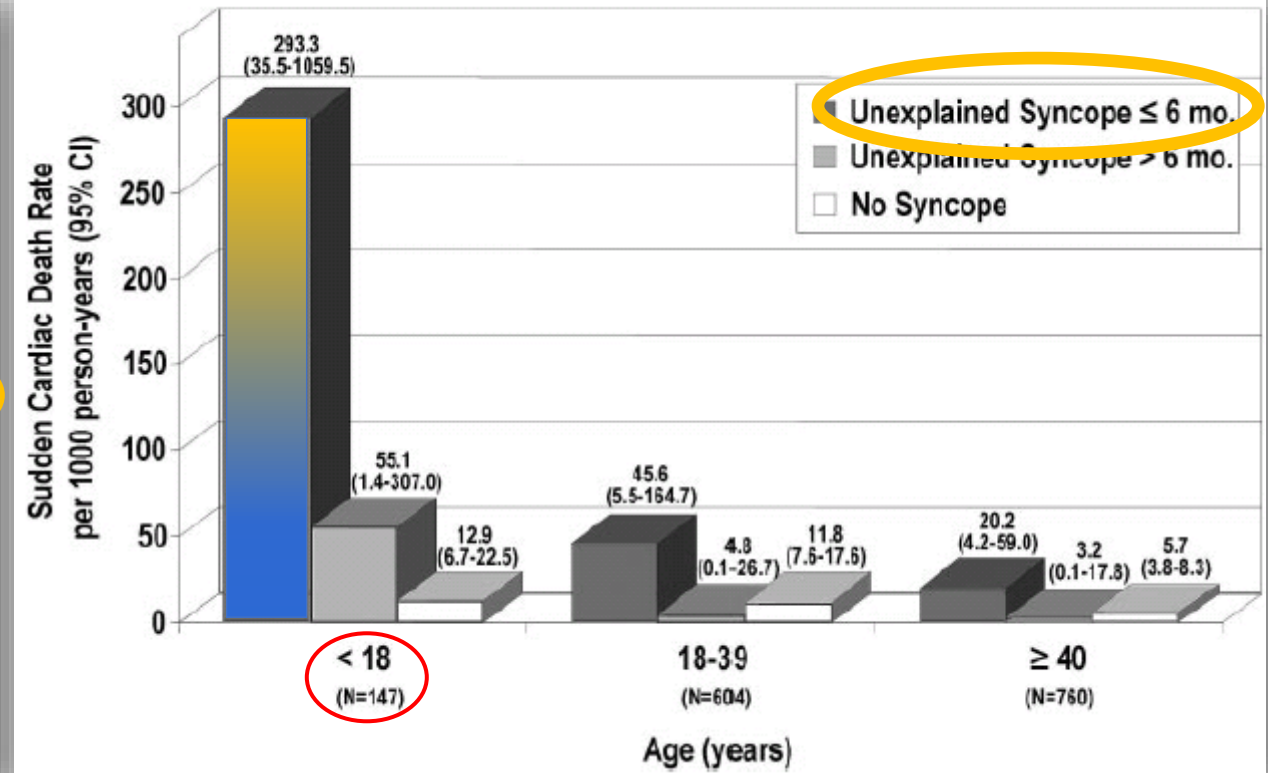
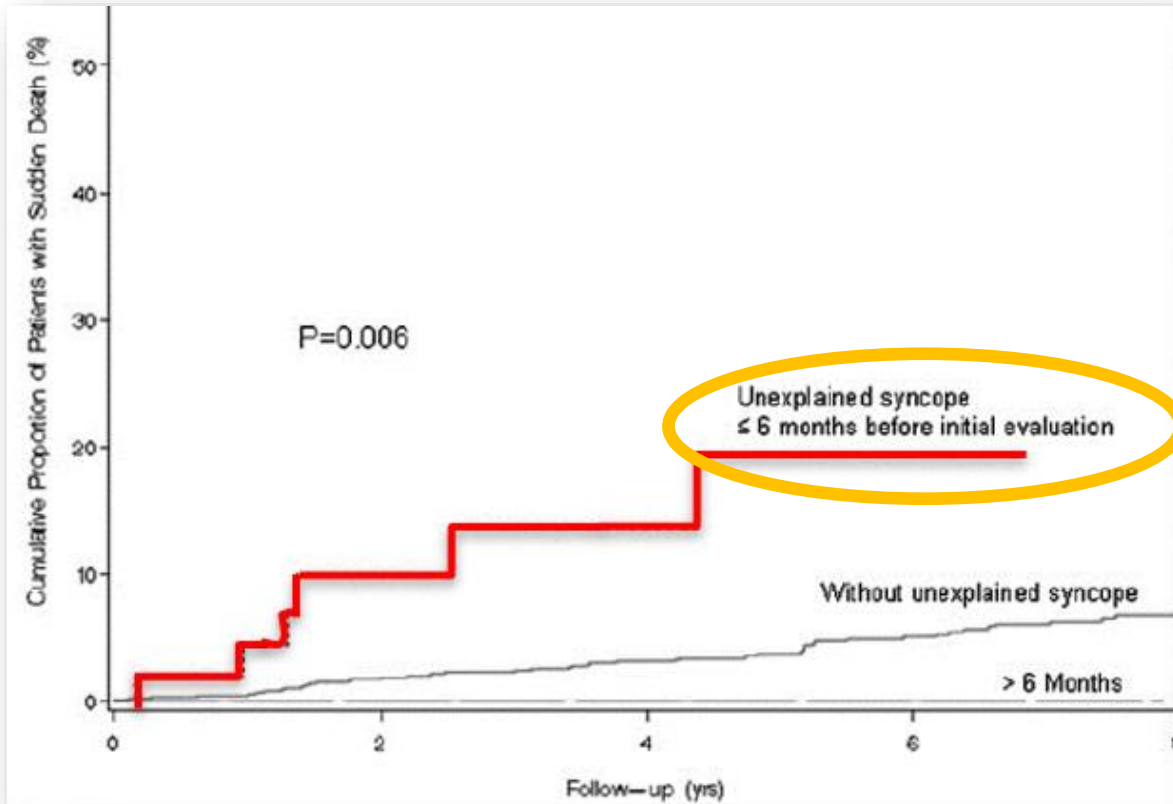
ECG



ECHO



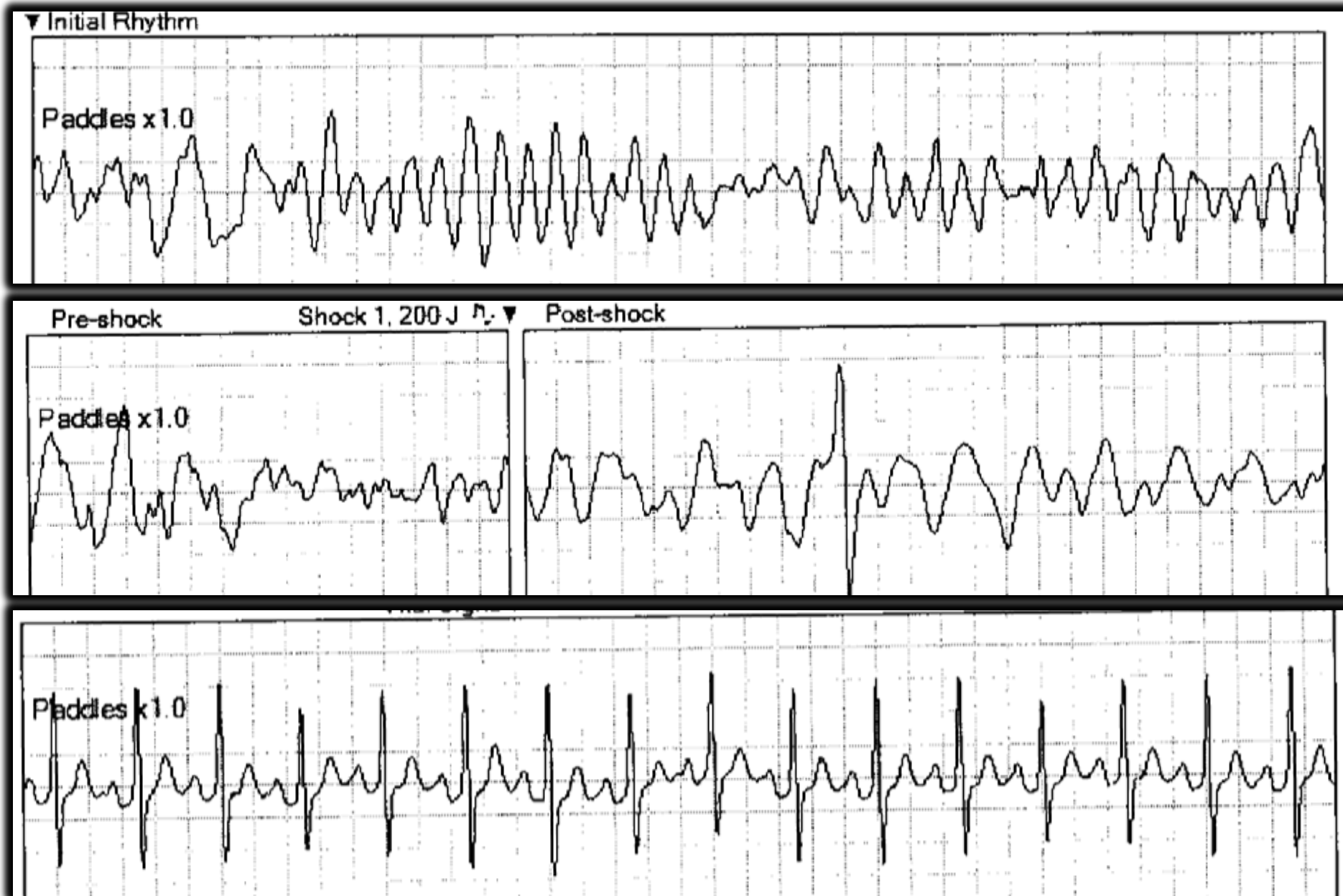
“Recent Unexplained Syncope” & Risk of SCD in HCM



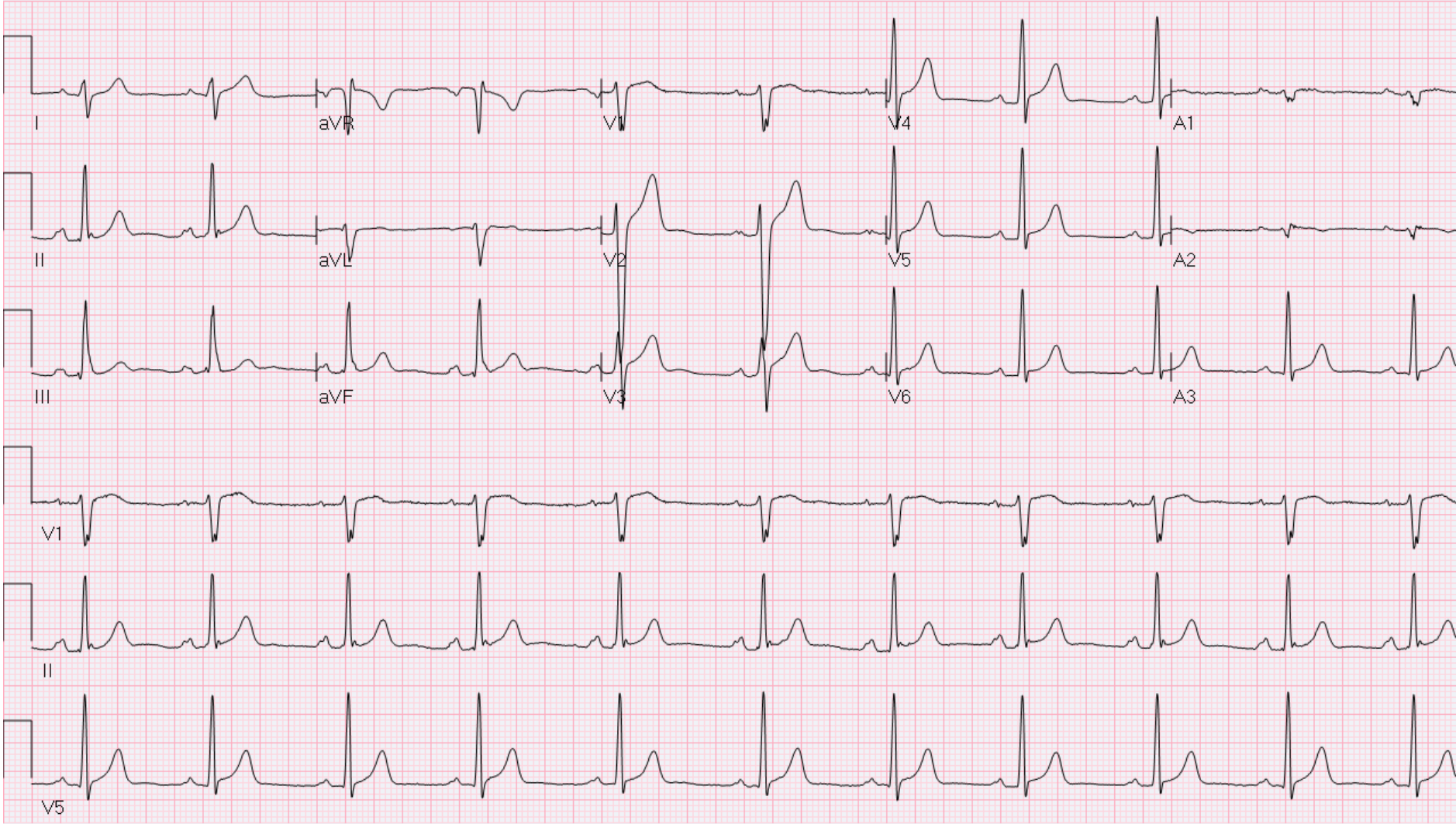
16-year-old teenager who collapsed while playing basketball

- 16-year-old male fell while playing basketball, striking his face/head on the ground
- He got back up, seemed dazed, and again fell to ground
- Assisted by his friend, taken inside and 911 called
- CPR started by EMS
- Found to be in ventricular fibrillation, defibrillated
- Resumed spontaneous respirations, and regained consciousness
- No previously known cardiac conditions

EMS RHYTHM TRACINGS



INITIAL ECG



Genetic Testing



RESULT: POSITIVE

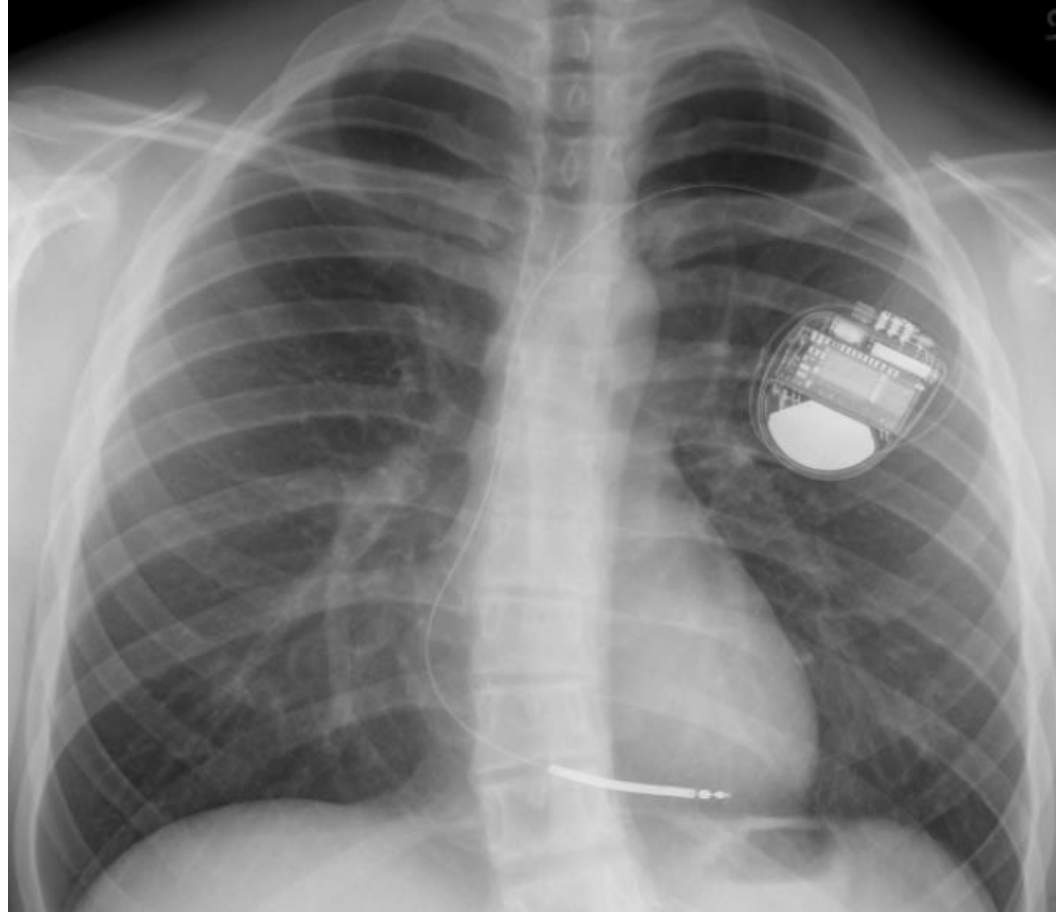
One Pathogenic variant identified in SCN5A. SCN5A is associated with a spectrum of autosomal dominant cardiac conditions.

GENE	VARIANT	ZYGOSITY	VARIANT CLASSIFICATION
SCN5A	c.3761del (p.Met1254Serfs*26)	heterozygous	PATHOGENIC

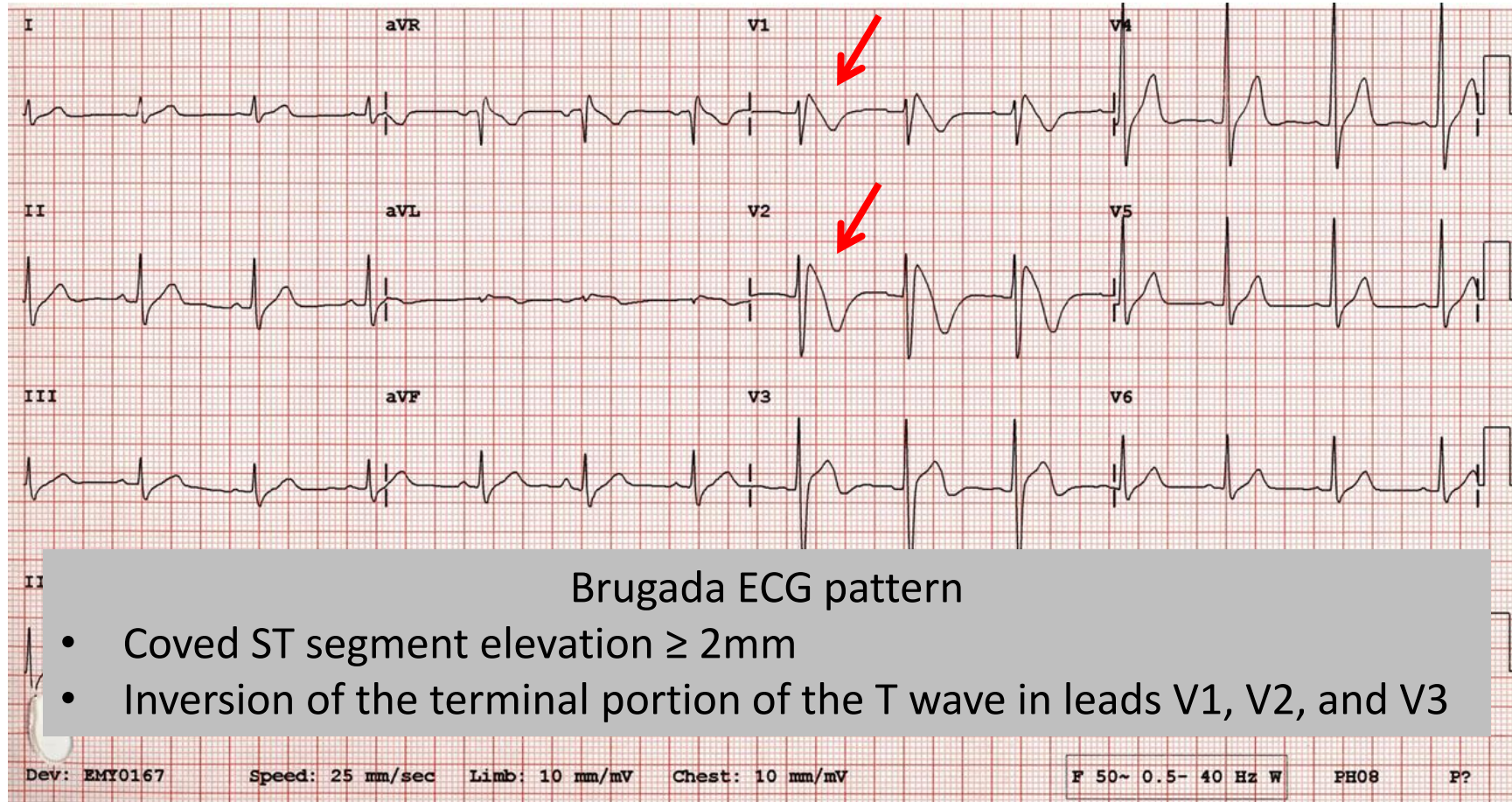
About this test

This diagnostic test evaluates 39 gene(s) for variants (genetic changes) that are associated with genetic disorders. Diagnostic genetic testing, when combined with family history and other medical results, may provide information to clarify individual risk, support a clinical diagnosis, and assist with the development of a personalized treatment and management strategy.

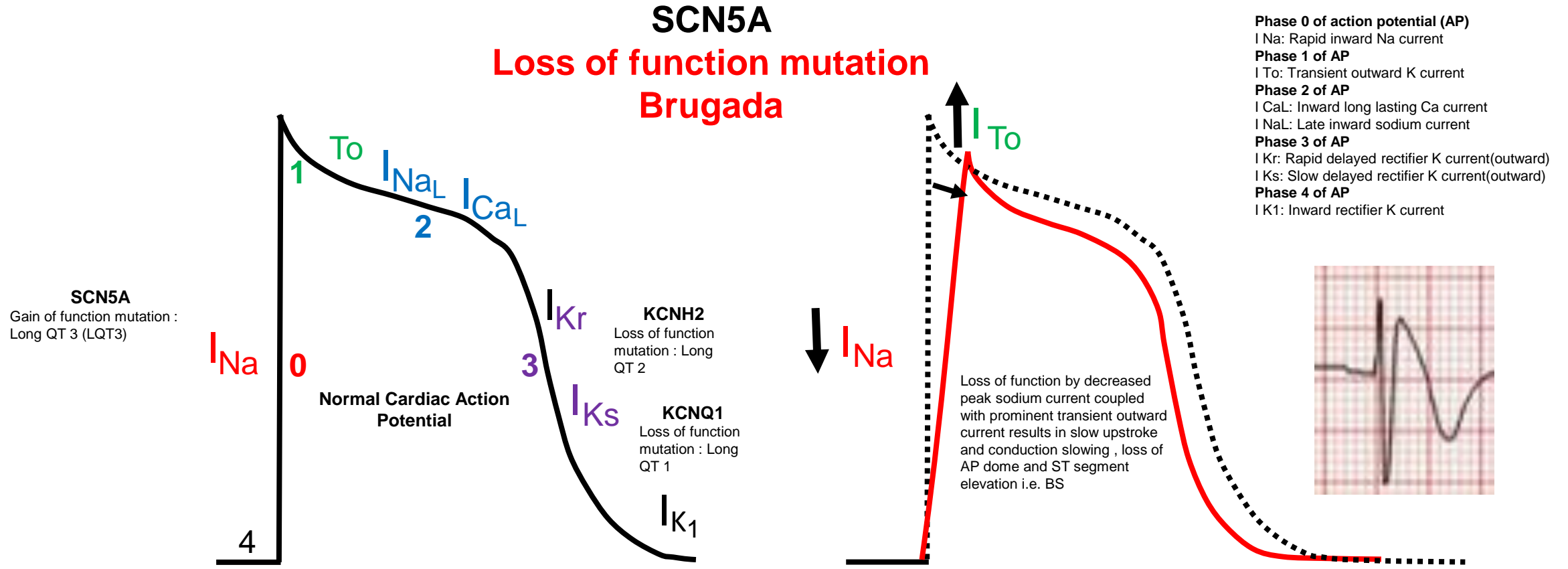
ICD FOR SECONDARY PREVENTION



Brugada Syndrome



Brugada Syndrome



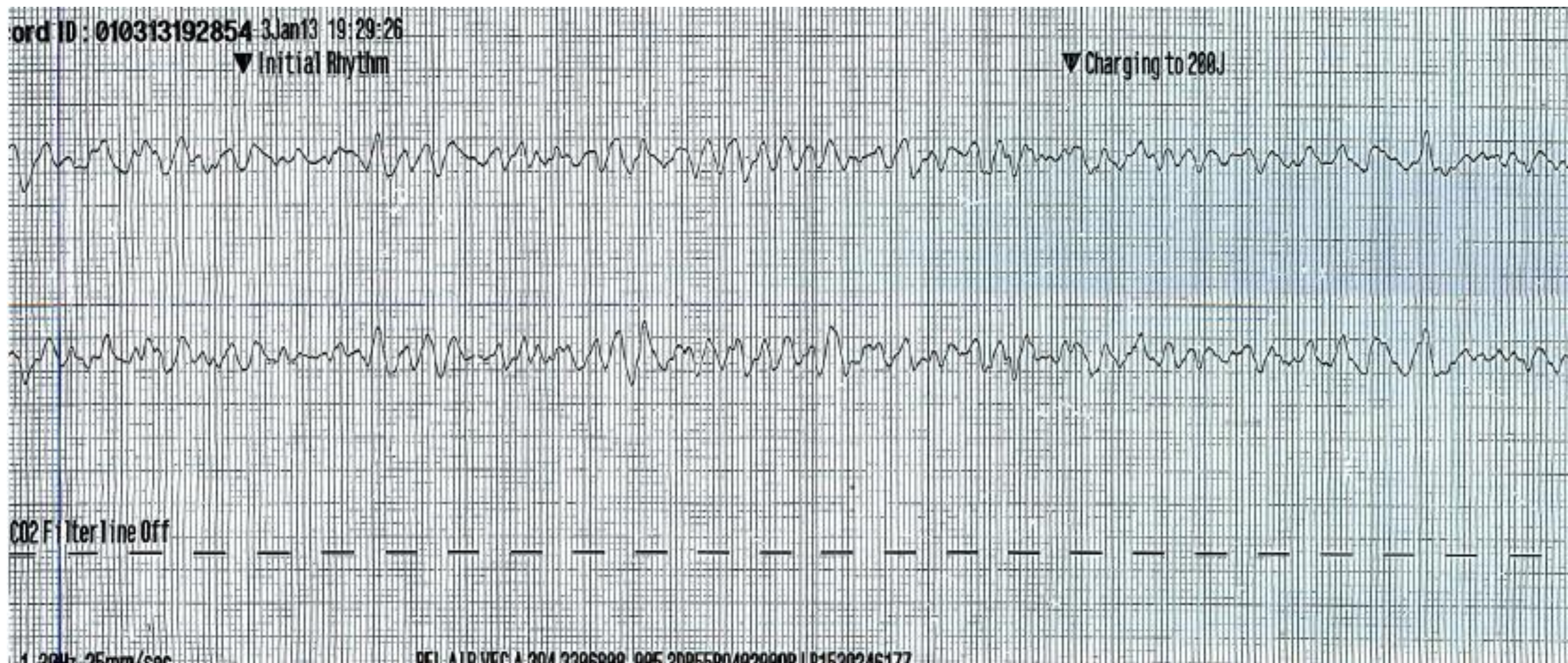
Brugada Syndrome

- Endemic in East and Southeast Asia..SUNDS
- Arrhythmias can occur at any age(2-77y)
- 4% of all SCD, 20% of SCD in structurally normal heart
- Palpitations, dizziness, syncope, nocturnal agonal respiration
- Fast polymorphic VT degenerating into VF
- Occurs at rest /sleep when vagal tone is augmented
- Febrile illness is also a trigger

A 17-y old female teenager who fell while playing basketball

- Previously healthy 17 y old female playing competitive basketball in school
- Came out of the game and fell on the ground soon after she sat on the bench
- Pulseless and pale → coaches started CPR
- AED showed ventricular fibrillation and shock delivered

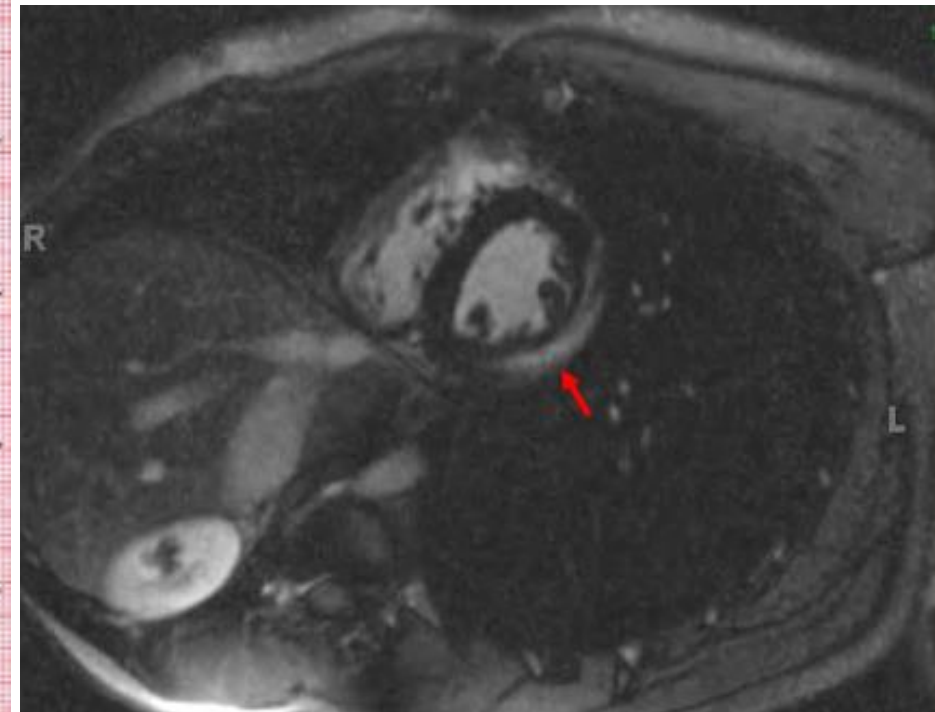
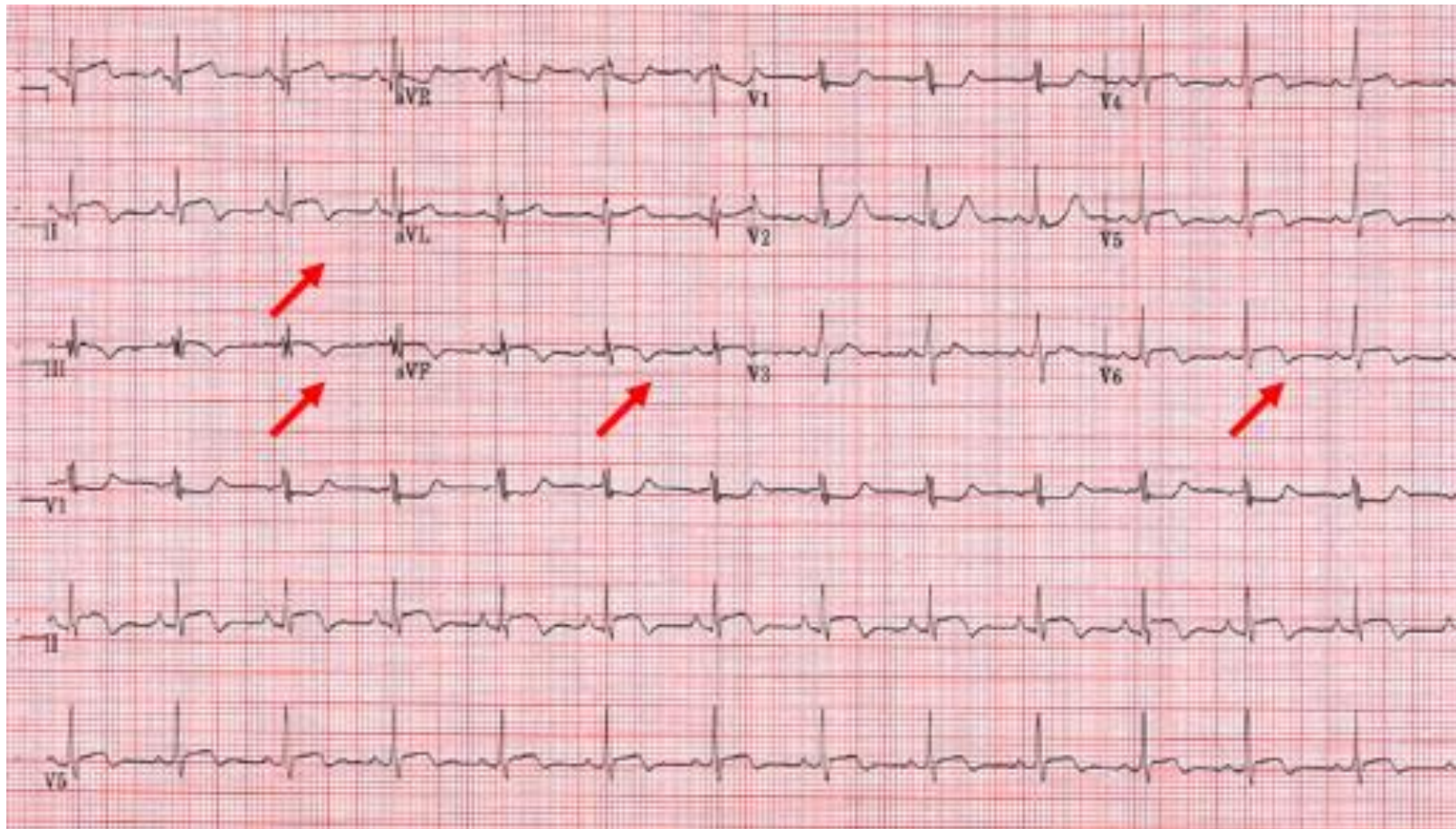
AED disclosure



A 17-y old female teenager who fell while playing basketball

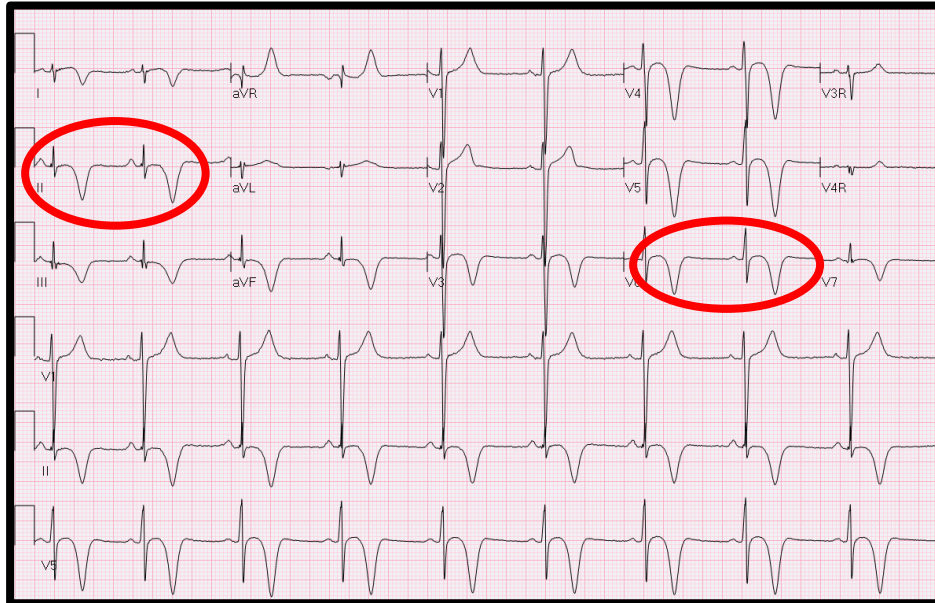
- Transferred to UMMC PICU
- Baseline ECG
 - ST and T wave abnormalities
 - Ventricular ectopy
- ECHO showed mildly reduced function
- Cardiac MRI : extensive subepicardial delayed enhancement
- ICD implanted

A 17-y old female teenager who fell while playing basketball

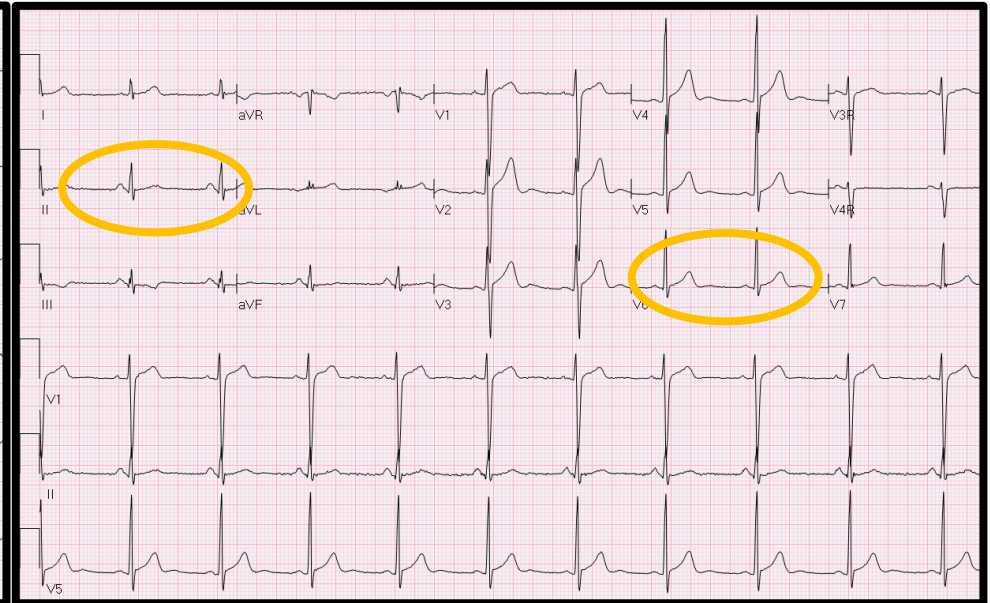


Myocarditis: ECG changes

ECG in myocarditis

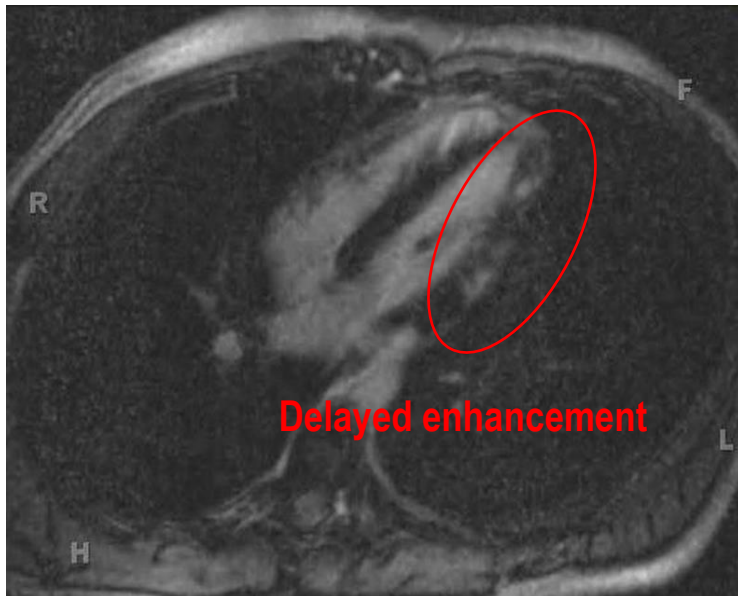
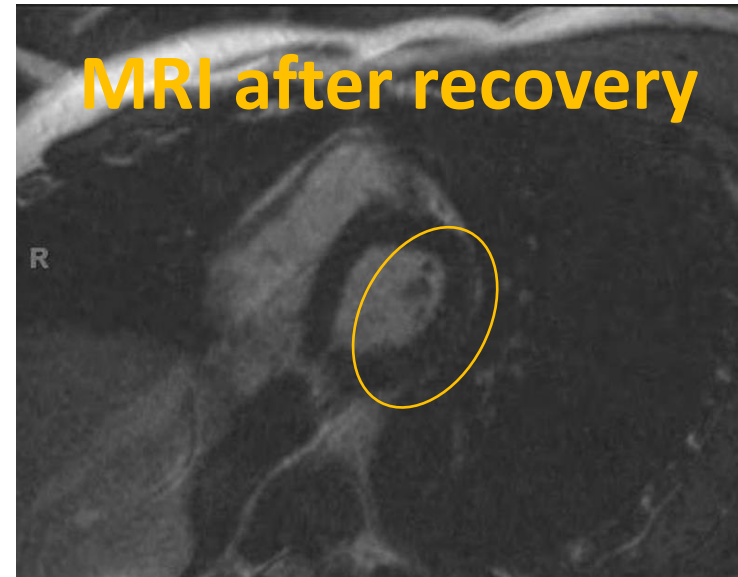
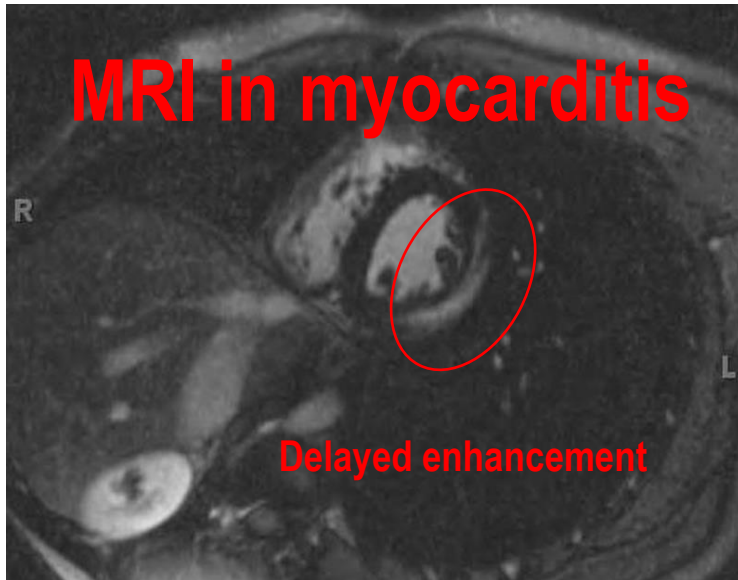


ECG after recovery



T wave inversion

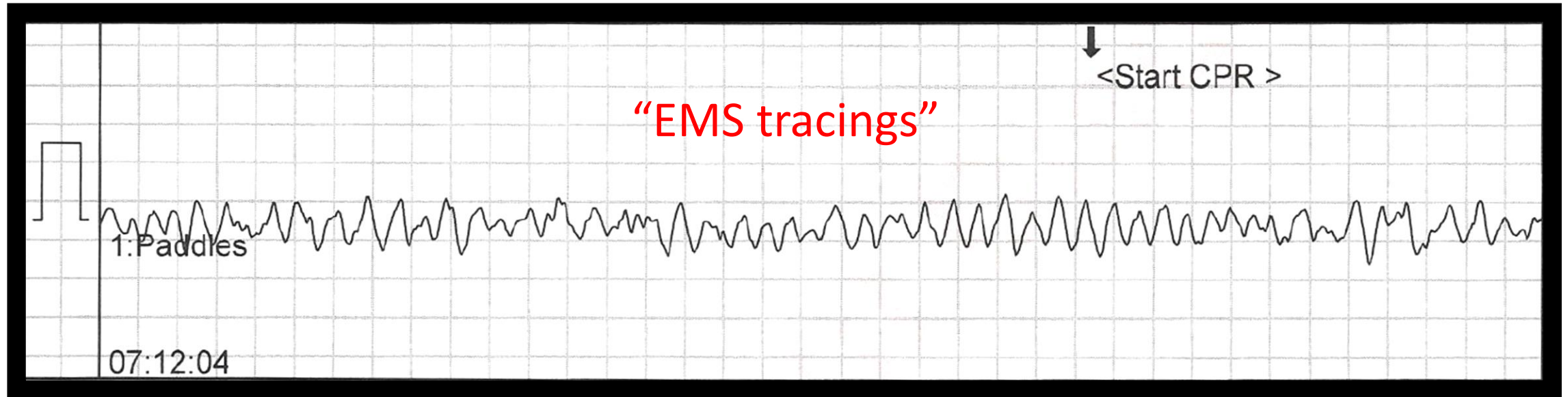
Myocarditis: Cardiac MRI



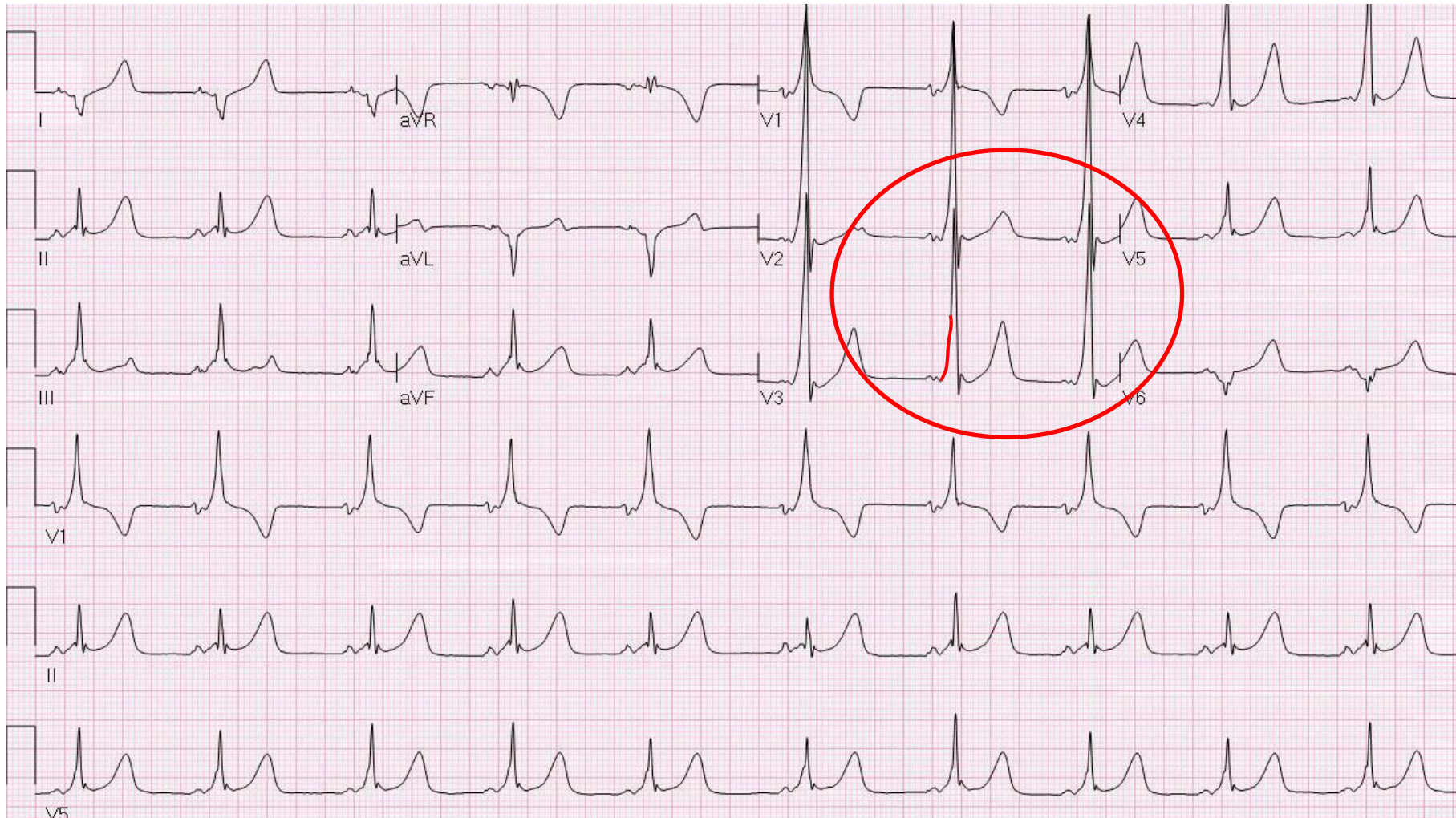
Football Player Who Collapsed At Home

- Woke up in the morning and felt his heart racing
- Parents thought that he had “pregame jitters” and asked him to get ready for the game
- As he was getting ready , family heard a loud noise and found him unconscious
- Family started chest compressions and called 911
- EMS repeatedly defibrillated until reached ER

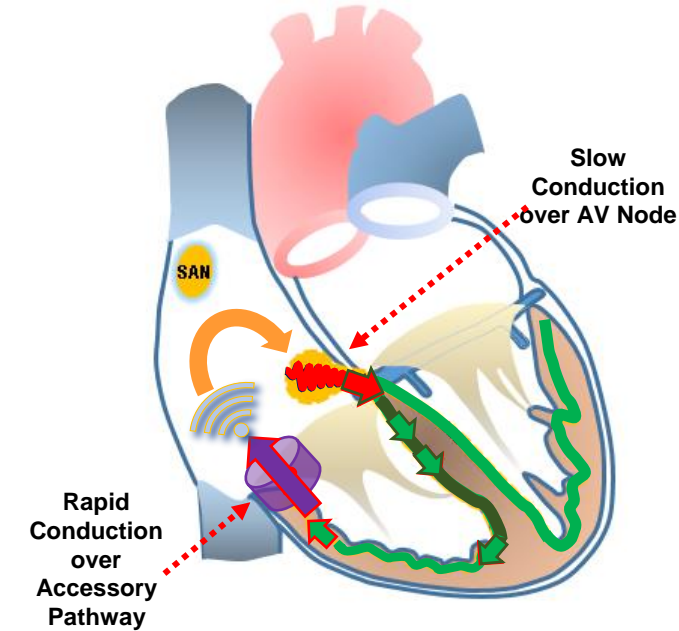
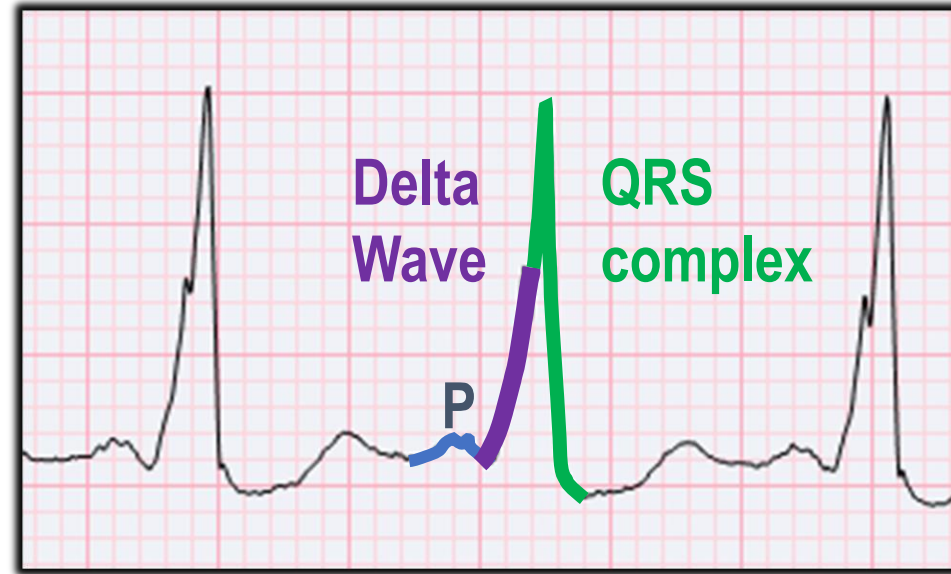
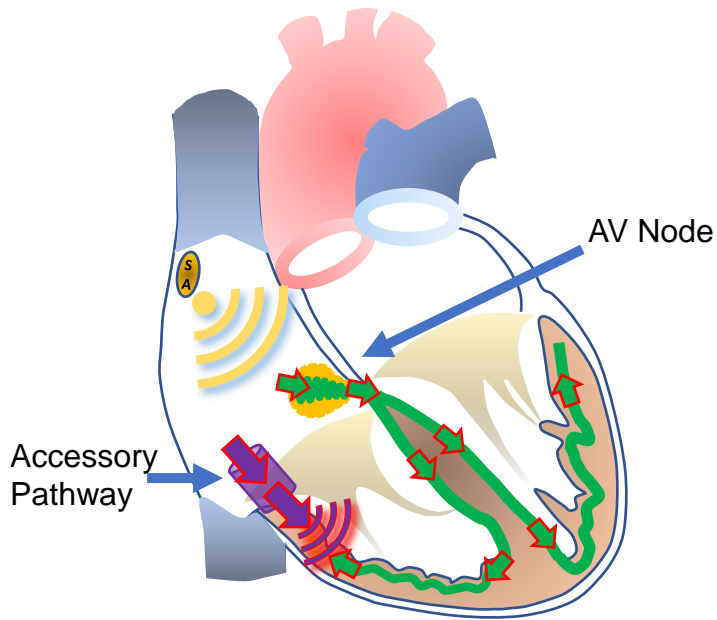
Football Player Who Collapsed At Home



Football player who collapsed at home “ECG in the Hospital”

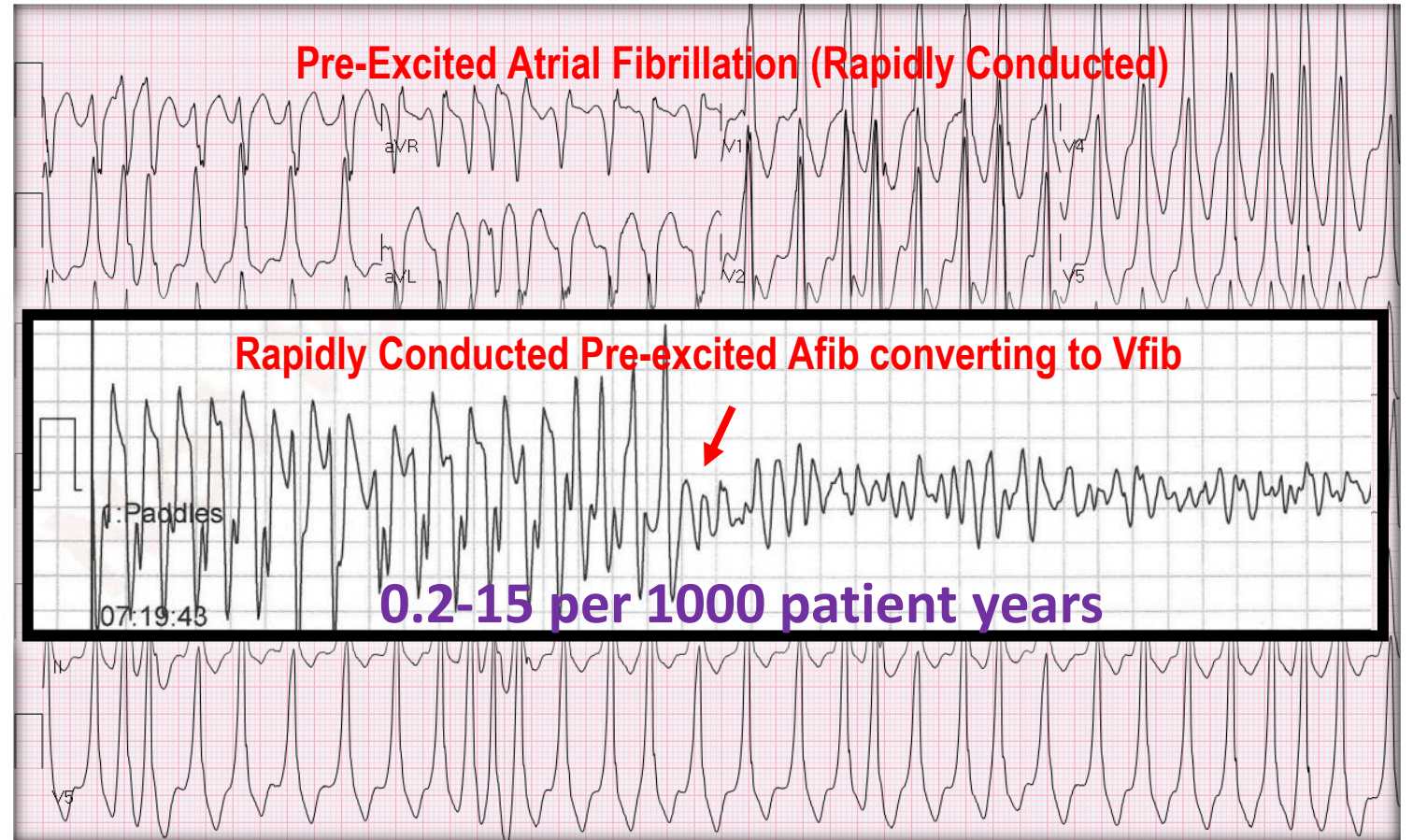
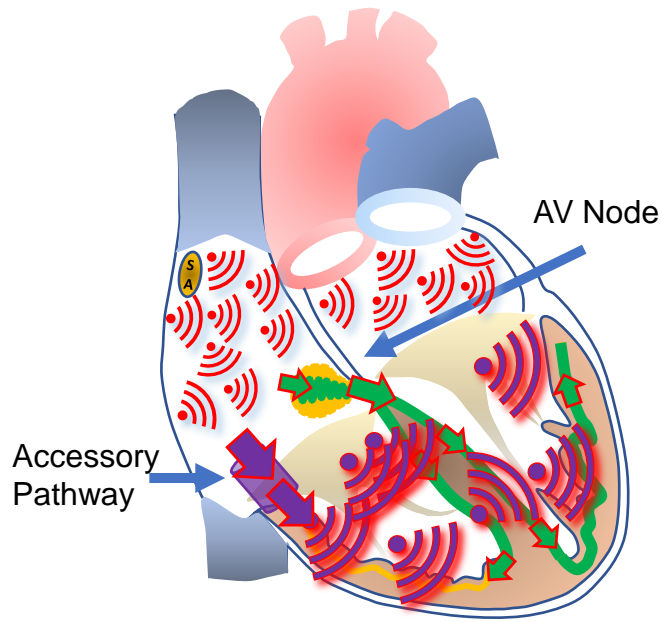


Wolff-Parkinson-White Syndrome (WPW)



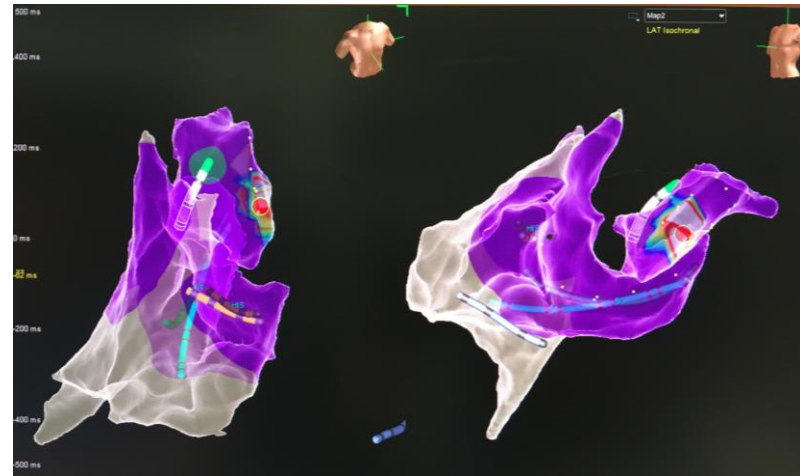
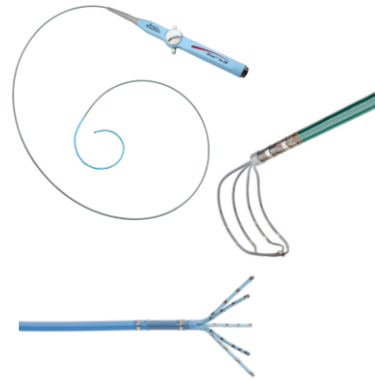
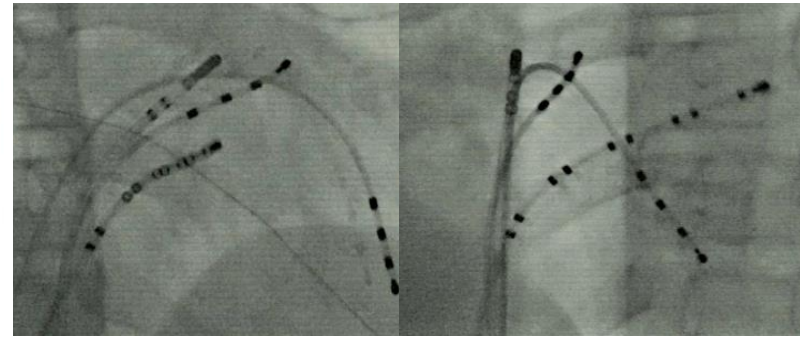
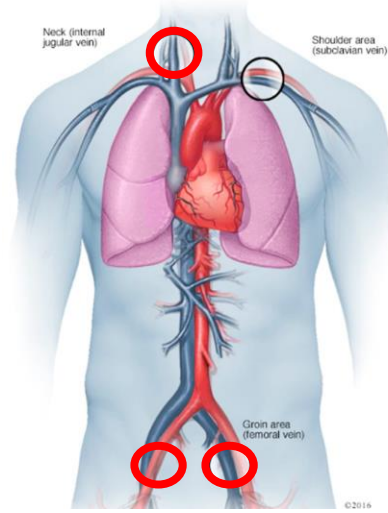
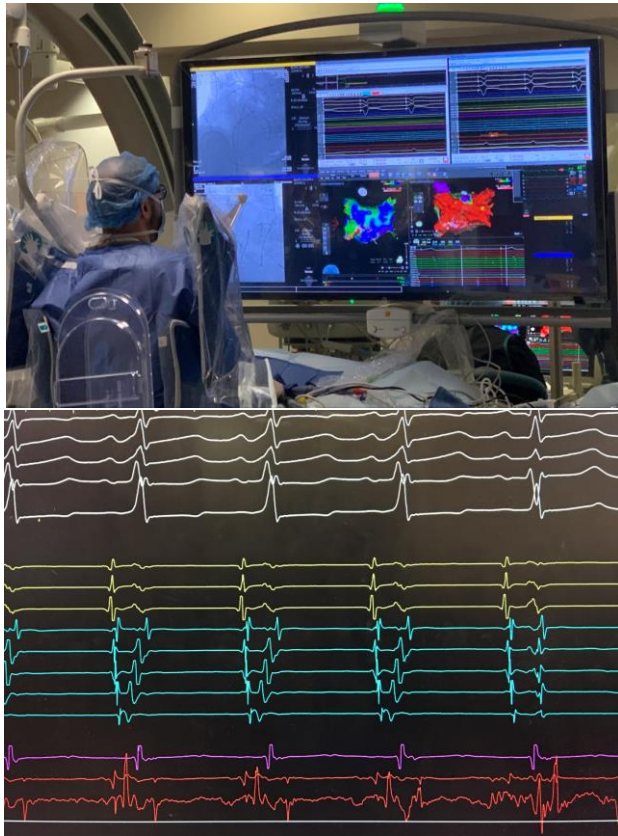
- Caused by an accessory pathway bridging the AV groove
- Can conduct from A to V and also V to A
- Prevalence : 1-3/1000 individuals and not familial in most cases
- Mostly presents as SVT but rarely can cause SCD

WPW + Atrial Fibrillation → Ventricular Fibrillation

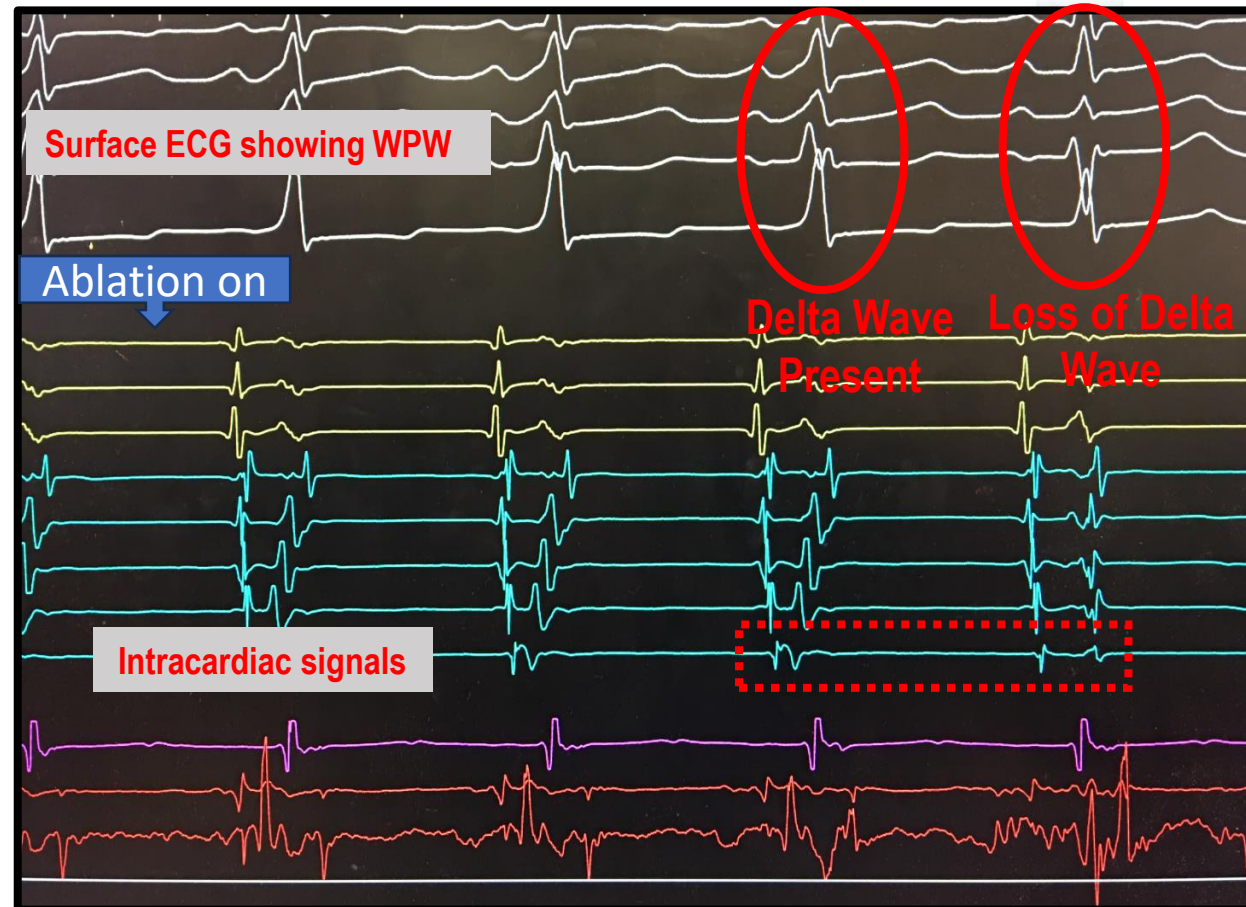


Catheter ablation is safe and effective and eliminates the risk of SCD

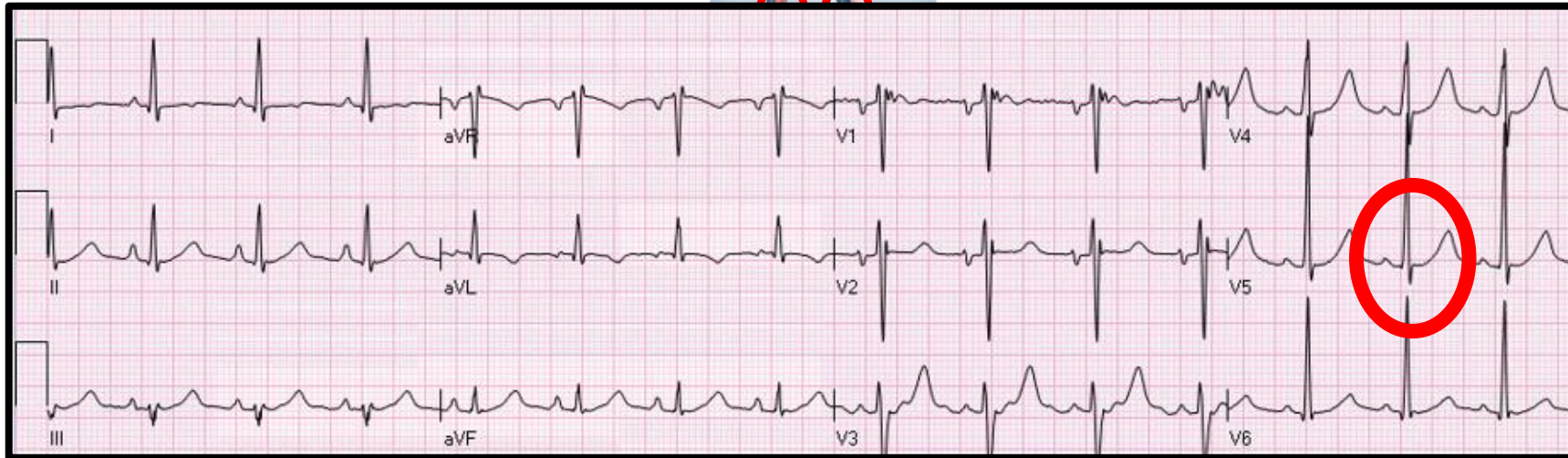
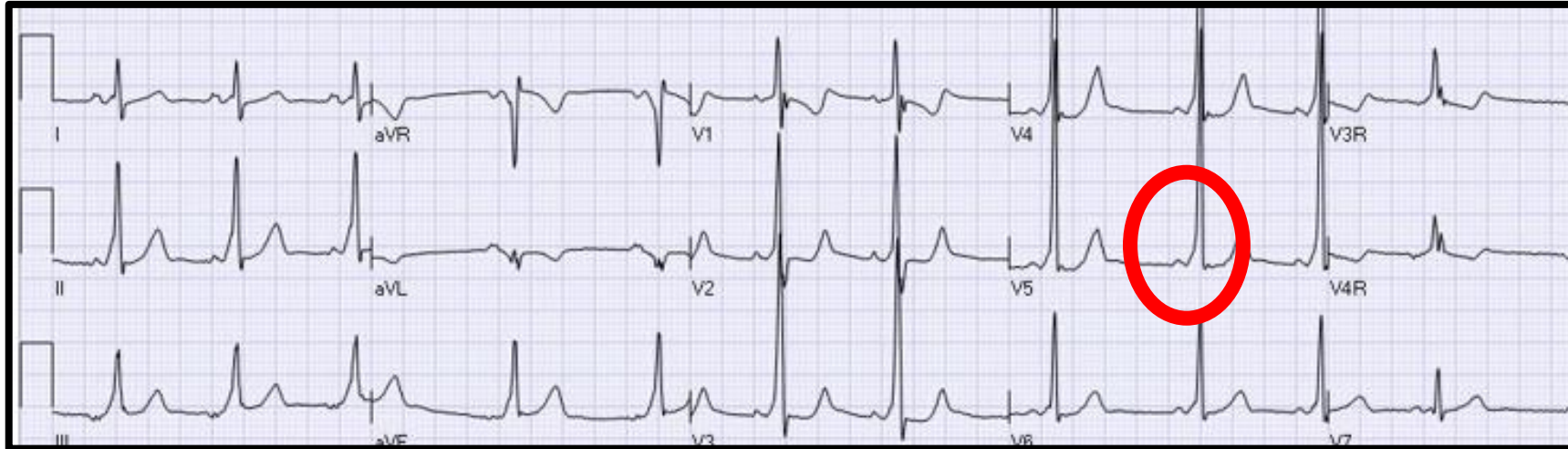
Electrophysiology Study (EP study) and Ablation is curative



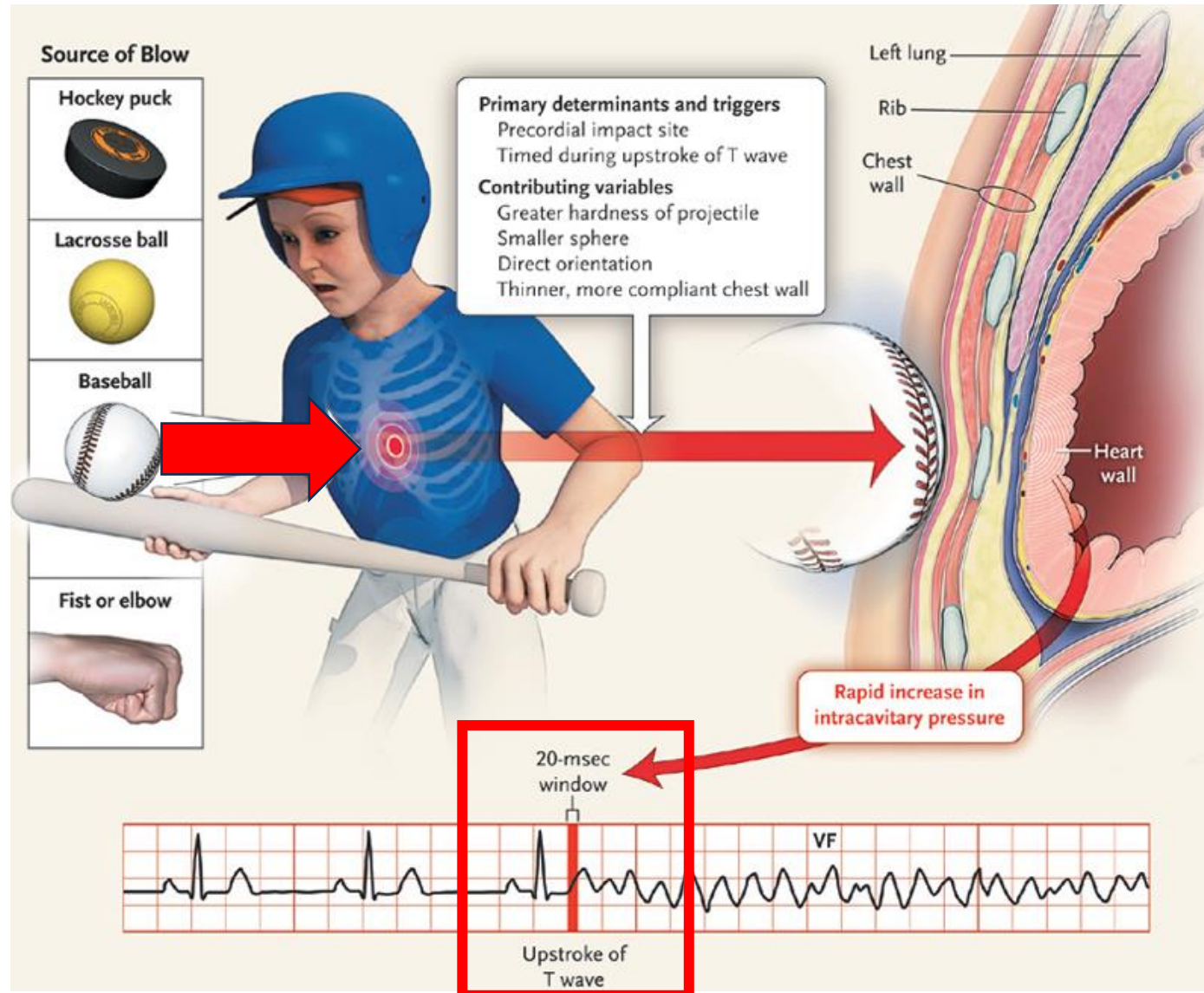
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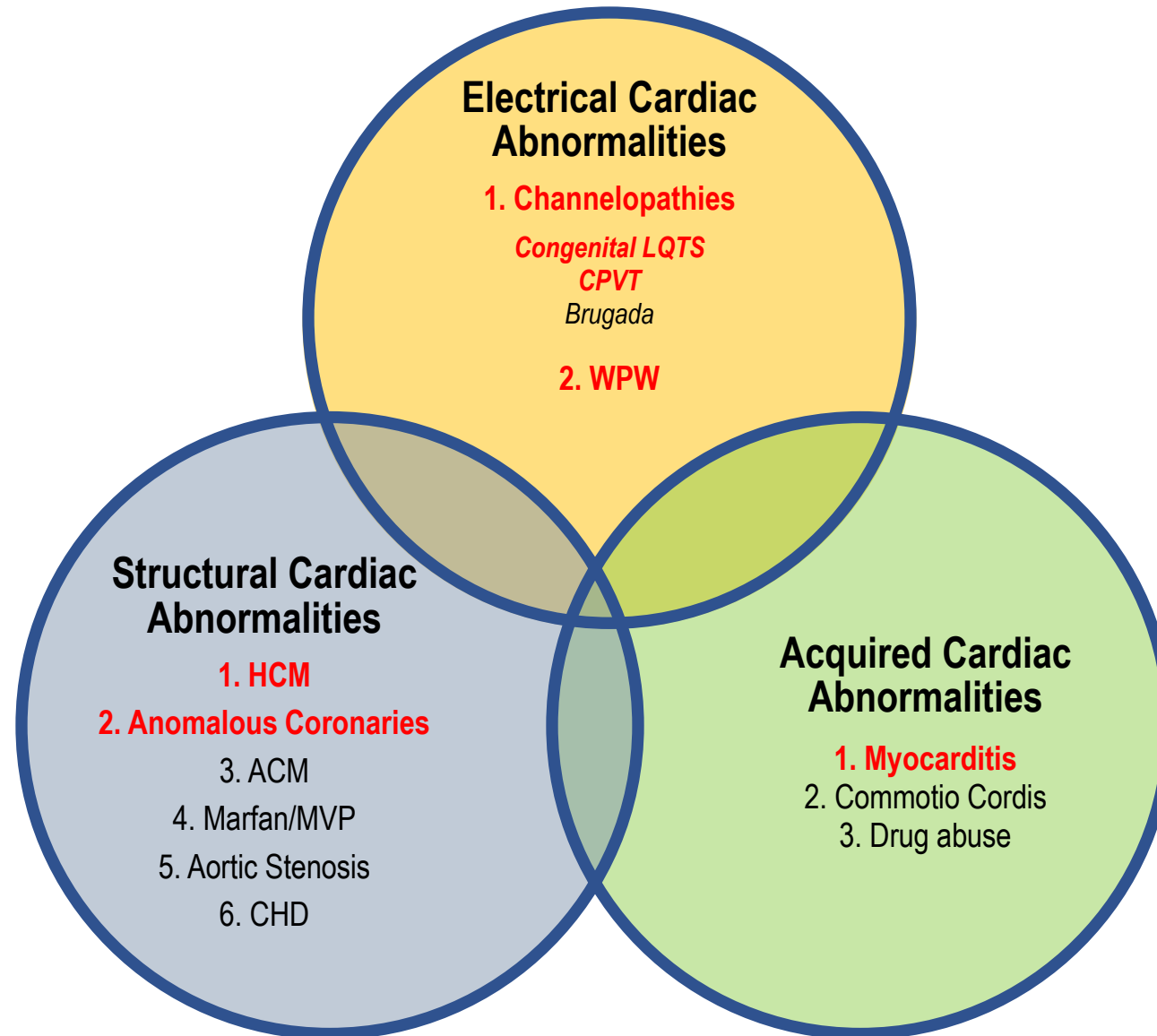
Electrophysiology Study (EP study) and Ablation is curative



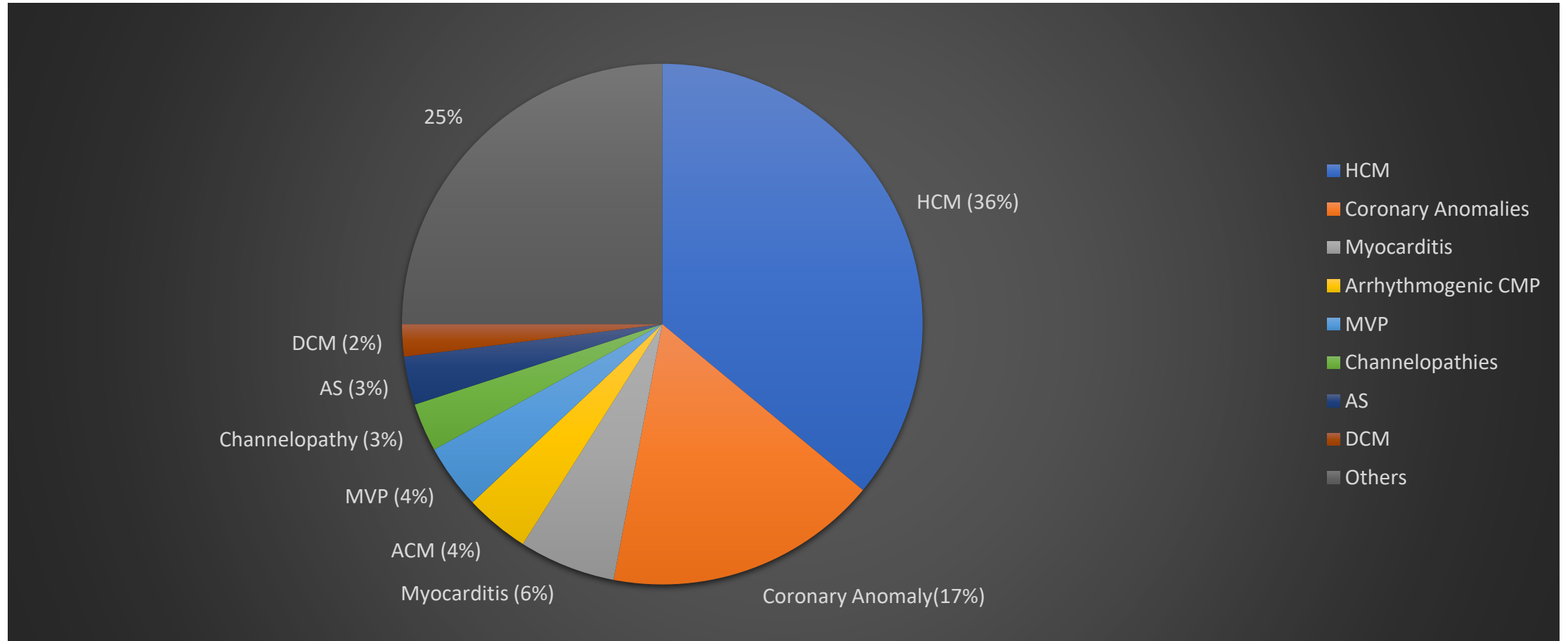
Comotio Cordis



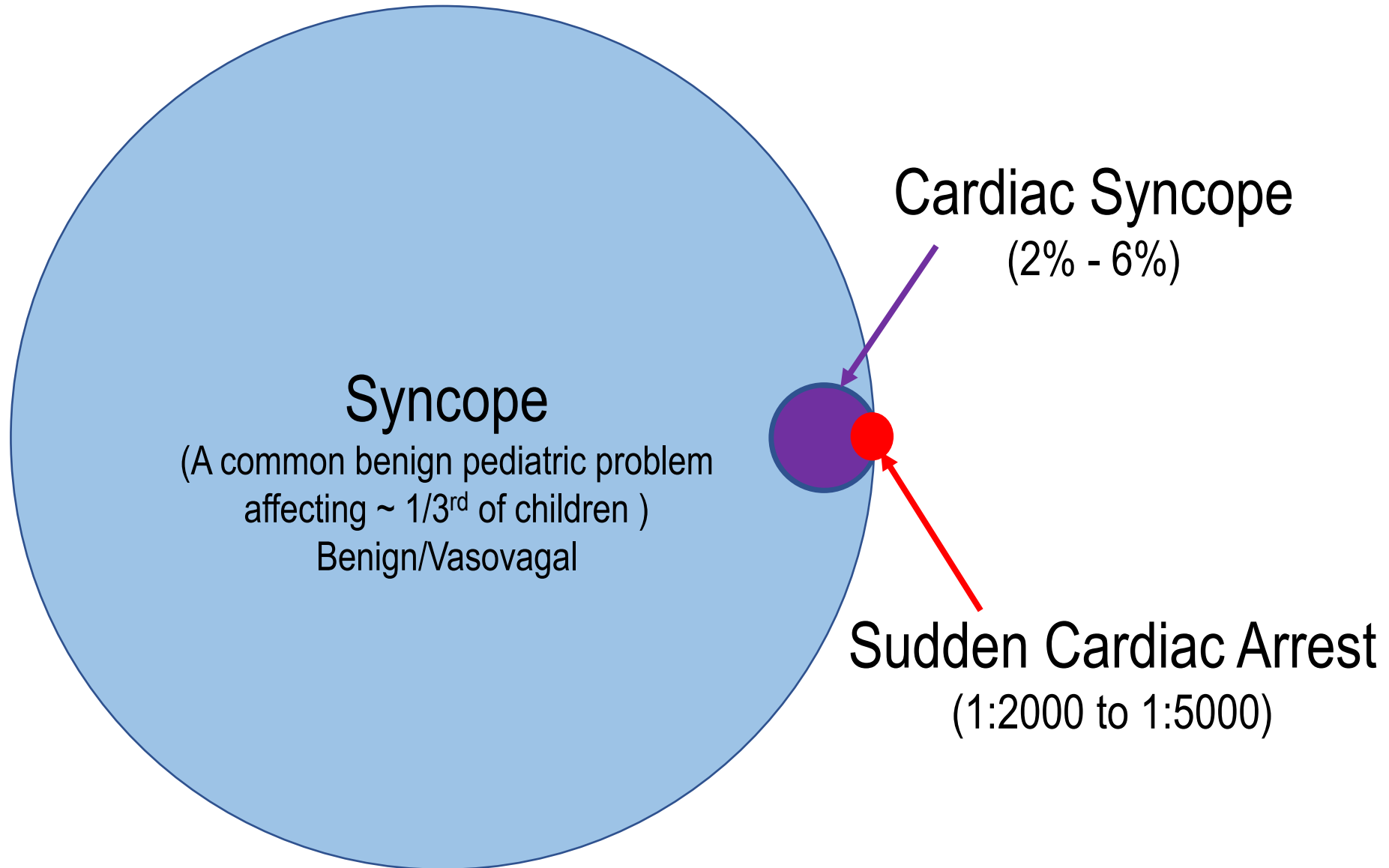
Causes of Sudden Cardiac Death in Young



Causes of SCD in Young Competitive Athletes in US



Syncope : Simple Faint or Near Miss?

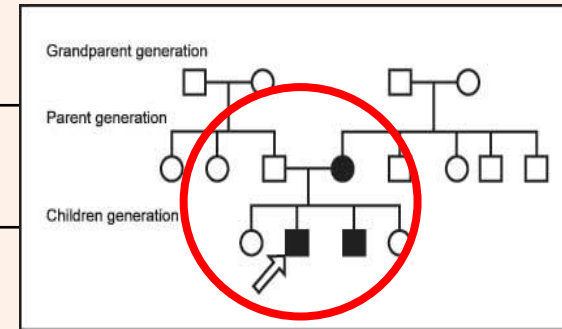


When to Suspect Cardiac Etiology

- Exertional
- While swimming
- Triggered by loud noise
- Triggered by extreme emotions
- History of cardiac disease
- Family history of SCD
- Abnormal examination
- Abnormal ECG
- Preceded by chest pain or palpitations
- Absence of prodromal symptoms

Family History Is Critical : Red Flags in Family History

1	Sudden unexpected unexplained death at a young age (<30 y)
2	Unexplained fainting spells or seizures
3	Unexplained driving or drowning accidents
4	SIDS , Congenital deafness
5	Heart attack before 50 (clarify “heart attack”)
6	Hereditary cardiomyopathies (HCM, DCM, ARVD)
7	Hereditary channelopathies (LQTS, Brugada, CPVT, short QTS)
8	Pacemaker or defibrillator implants at a young age



Pre-Participation Screening (PPS) Background

- PPS is the practice of screening athletes before participation in sports
- Primary goal is to detect silent predominantly genetic/congenital cardiovascular conditions that predispose the athlete to SCA/SCD

AHA recommendation regarding PPS in young competitive athlete: updated in 2014

- AHA recommends pre-participation screening involving 14 key elements
 - Targeted personal history
 - Targeted family history
 - Physical examination
- AHA does not recommend routine use of
 - ECG
 - Echocardiogram

The 14-Element AHA recommendations for PPS of Competitive Athletes

Personal history

1. Chest pain/discomfort/tightness/pressure related to exertion
2. Unexplained syncope/near-syncope†
3. Excessive and unexplained dyspnea/fatigue or palpitations, associated with exercise
4. Prior recognition of a heart murmur
5. Elevated systemic blood pressure
6. Prior restriction from participation in sports
7. Prior testing for the heart, ordered by a physician

Physical examination

11. Heart murmur‡
12. Femoral pulses to exclude aortic coarctation
13. Physical stigmata of Marfan syndrome
14. Brachial artery blood pressure (sitting position)§

Family history

8. Premature death (sudden and unexpected, or otherwise) before 50 y of age attributable to heart disease in ≥ 1 relative
9. Disability from heart disease in close relative < 50 y of age
10. Hypertrophic or dilated cardiomyopathy, long-QT syndrome, or other ion channelopathies, Marfan syndrome, or clinically significant arrhythmias; specific knowledge of genetic cardiac conditions in family members

What did we learn from
these case scenarios?

16-year-old Volleyball Player “Long QT Syndrome”

- She had exertional syncope the prior year
- Her ECG had shown “borderline” QT prolongation
- Physical examination: normal
- Cardiology evaluation was suggested
- No sports restrictions applied
- Family history could be helpful

Sibling of 16-year-old Volleyball Player “Long QT syndrome”

- She had past h/o vasovagal syncope
- This episodes also appears to be vasovagal, and she is compliant with her medications
- Otherwise, syncope despite medical management could be a consideration of additional therapies such as ICD

15-year-old with Seizure & Syncope “CPVT”

- He had exertional syncope the prior year
- Family h/o heart attack at 21 y of age is suspicious
- Physical examination: normal
- Baseline testing :normal
 - Normal ECG
 - Normal ECHO
- Stress test useful

14-year-old Field Hockey Player “Anomalous Coronaries”

- She had symptoms of chest discomfort and near fainting while playing for the past few months
- Family history not contributory
- Physical examination: normal
- Baseline ECG is normal
- Echo would likely pick up the diagnosis

3- year-old with previous diagnosis of “HCM”

- This is obvious case of aborted SCA
- However, what if this patient had regained consciousness spontaneously and then came for evaluation?
- I would have still treated it as aborted SCA given his past medical history of HCM unless until proven otherwise
- Or what if we did not have pre-existing diagnosis of HCM and he presented for the first time for unwitnessed / unexplained syncope?
- Given the young age at the time of syncope, I would still be concerned and refer to cardiology

10-year-old with syncope and murmur “New Diagnosis of HCM”

- Personal Hx: syncope was exertional
- Family h/o SCD at a young age
- Physical examination: murmur
- ECG would be abnormal in most cases
- Was his syncope vasovagal?

**16 y old teenager with SCA playing
basketball
“Brugada Syndrome”**

- This is obvious case of aborted SCA
- However, what if this patient had regained consciousness spontaneously and then came for evaluation?
- I would still be concerned as it happened while playing and without any prodromal symptoms
- Family history may be positive
- Physical examination: normal
- ECG with V1 and V2 in second intercostal space will be helpful

**17 y old female teenager with SCA while
playing basketball
“Myocarditis”**

- Personal history may be positive for
 - Recent h/o viral illness
 - Chest pain, palpitations, syncope
- Physical examination
 - Murmur, gallop
- Baseline ECG, echocardiogram and cardiac enzymes may be abnormal

Football player who collapsed at home

WPW

- Personal history may be positive for palpitations, syncope
- Family history mostly negative
- Screening ECG would have picked up the diagnosis even if asymptomatic
- Physical examination: normal
- SCA could be the initial presentation in WPW

Take Home Message

- Exertional symptoms are most worrisome for a cardiac etiology
- HCM is the most common cause of SCD in athletes
- Channelopathies
 - most common cause of cardiac syncope
 - commonly misdiagnosed as seizure disorder
- Obtaining detailed family history is critical
- ECG is the single most important investigation

Take Home Message

- Despite our best efforts at prevention and management it is unlikely that we can prevent every single case of SCA
- Assume SCA in any athlete/student who has collapsed, is not breathing normally or only gasping, and is unresponsive, no definite pulse felt
- Chain of survival is critical for secondary prevention
 - early EMS activation
 - early CPR/bystander CPR
 - early defibrillation/public access defibrillation
 - early post-resuscitation care in hospital