



Pediatric Cardiac Emergencies

Jacob Wannemacher MD
7/22/23



Overview

- Pediatric chest pain
- Pediatric Syncope
- Pediatric EKG's
- When not to give Epinephrine



Chest pain in children

- Musculoskeletal or idiopathic > 50%
- Psychiatric, GI, pulmonary causes common
- Sickle cell related chest pain more common than cardiac
- Cardiac cause only 1% or less of cases



Cardiac Causes of chest pain

- Pericarditis
- Myocarditis
- Endocarditis
- Aortic Dissection
- Ischemia
- Cardiomyopathy
- Arrhythmia
- Valvular disease



High risk history features for cardiac cause

- Associated with exertion, syncope, seizure-like activity, palpitations, drug use, or dyspnea
- Ischemic qualities: substernal pressure, radiation to shoulders, neck, back, arms
- Associated diaphoresis
- Special populations: congenital heart disease, Marfan's or other connective tissue disease, Turner syndrome, glycogen storage disorders, muscular dystrophy, hyperlipidemia
- Risk factors for PE: OCP's, pregnancy, cancer, surgery, thrombophilia
- Family history of early heart attack or sudden death < age 50
- Family history of first degree relative with ICD, hypertrophic cardiomyopathy or known arrhythmic condition



High Risk Exam Features for Cardiac Cause

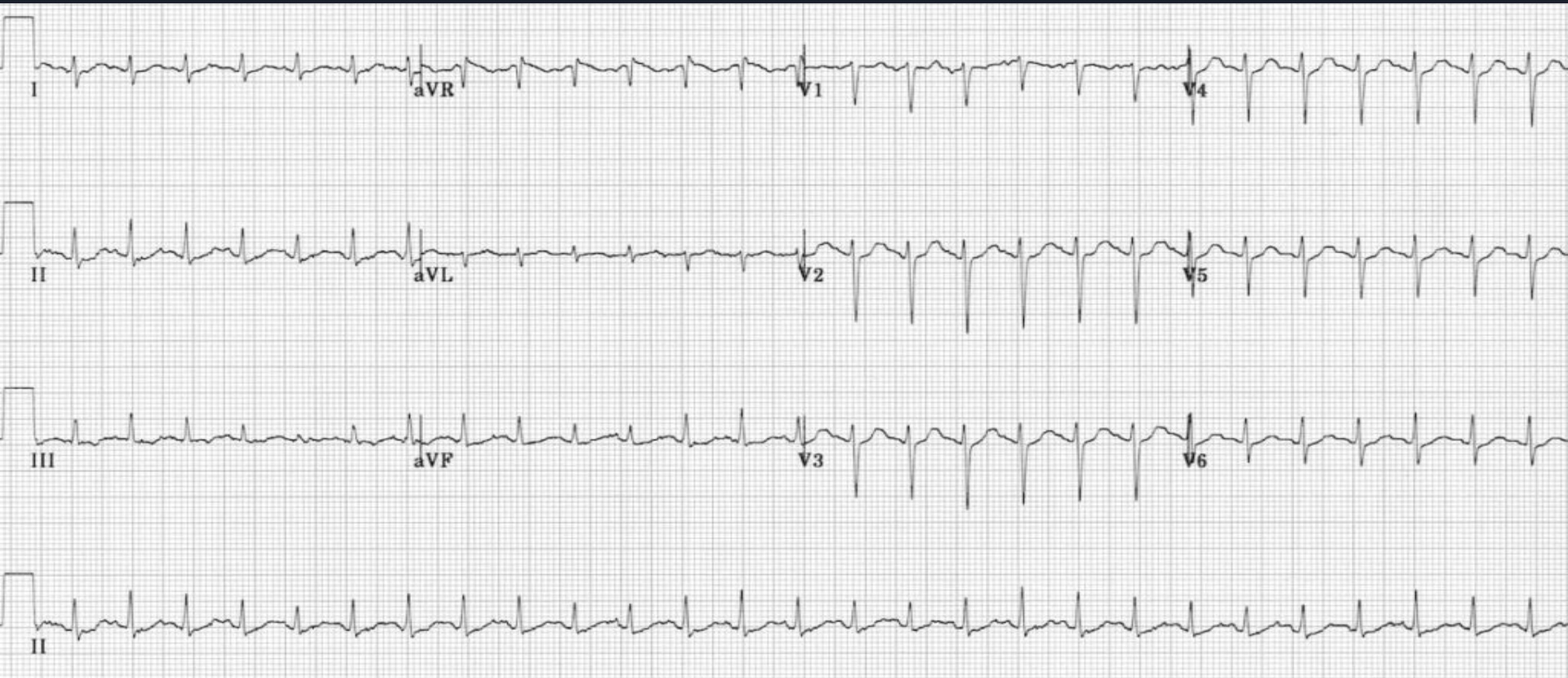
- Tachycardia out of proportion
- Decreased or differential pulses
- Diaphoresis
- Marfanoid features
- Pathologic murmur, gallop, friction rub, precordial heave
- Rales or wheezing
- Hepatomegaly, volume overload, orthopnea
- Poor perfusion



Workup

- Per history and physical
- No need for EKG if well appearing, no high risk features and likely alternative cause
- CXR only if history and exam suggest need
- If ill appearing: cardiac monitor, EKG, CXR, labs, +/- echo
 - Additional workup per presentation: PE, dissection, etc

16 y.o with chest pain, shortness of breath





Myocarditis

- Common cause of dilated cardiomyopathy in children
- Usually viral induced
 - Can be rickettsial, inflammatory cause (Kawasaki, JIA, lupus), hypersensitivity (eosinophilic) from meds, malignancy, etc
- Often after or during nonspecific flu-like illness.
 - Viral prodrome usually 1-4 weeks prior.
 - About 50% have fever at time of diagnosis



Myocarditis Presentation

- Wide variety
- May mimic flu-like symptoms or gastroenteritis
 - +/- Fever
 - Vomiting or abdominal pain, usually without diarrhea
- May mimic viral respiratory illness or asthma
 - Wheeze, rales, tachypnea
 - Chest pain variable
- CHF, poor perfusion unresponsive to fluids, cardiogenic shock, arrhythmia
- Sudden cardiac death in athletes



Myocarditis Diagnosis

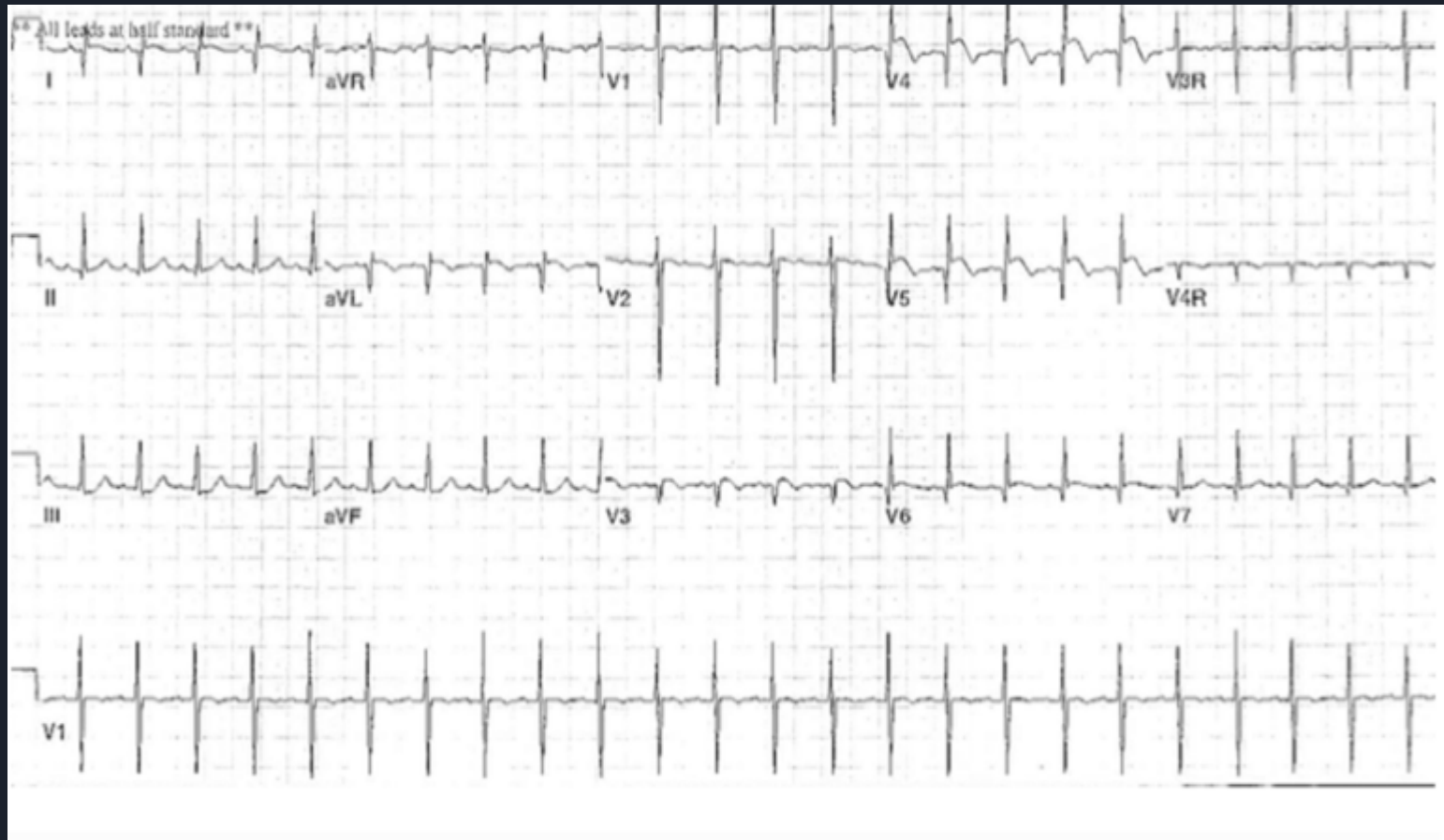
- Abnormal EKG is common, especially sinus tachycardia
 - Nonspecific ST segment changes, long QT, low voltage, LVH or strain pattern, arrhythmia, bundle branch block or heart block
- Troponin limited sensitivity, AST limited sensitivity, BNP limited sensitivity. Leukocytosis/inflammatory markers nonspecific
- Echocardiography
- Cardiac MRI or biopsy



Myocarditis Treatment

- Treat as CHF and hope for spontaneous recovery
 - May need ECMO, LVAD, or Impella
- When due to MIS-C, IVIG, steroids, aspirin may be used.
 - Mixed data on IVIG and steroids for other causes as well.
- Athletes with myocarditis will need to be cleared by cardiology

9 week old with colic and poor feeding





ALCAPA

- Anomalous Left Coronary Artery arising from the Pulmonary Artery
- Typically at 2-3 months as pulmonary vascular resistance falls
- Anterolateral infarct pattern
 - Q waves in I, aVL, V5 and V6. ST and T wave changes, loss of normal R wave progression.
- Colic can be angina
- CHF symptoms
- May present as sudden cardiac arrest or SIDS



ALCAPA

- Treatment is surgical
- 90% die in 1st year of life if untreated
- If good collateral circulation from RCA may survive
 - Present later in life with angina, CHF, arrhythmias
- Anomalous Aortic Origin of Coronary Artery (AAOCA) presents later in childhood



Aortic Dissection

- Rare in children
- Consider in connective tissue disease (Marfan's), aortic root dilation (Turner's, homocysteinemia) or late pregnancy
 - Or after procedures such as balloon dilation of coarctation of aorta
- Chest pain "plus":
 - Limb ischemia or pulse deficit
 - Neuro deficit or symptoms
 - AKI
 - Mesenteric ischemia
 - Aortic regurgitation, may cause sudden CHF symptoms
 - Cardiac tamponade (causes hypotension)
 - Coronary ischemia (dissection can extend into coronary ostium), aspirin contraindicated
 - Syncope (usually due to tamponade)



Aortic Dissection

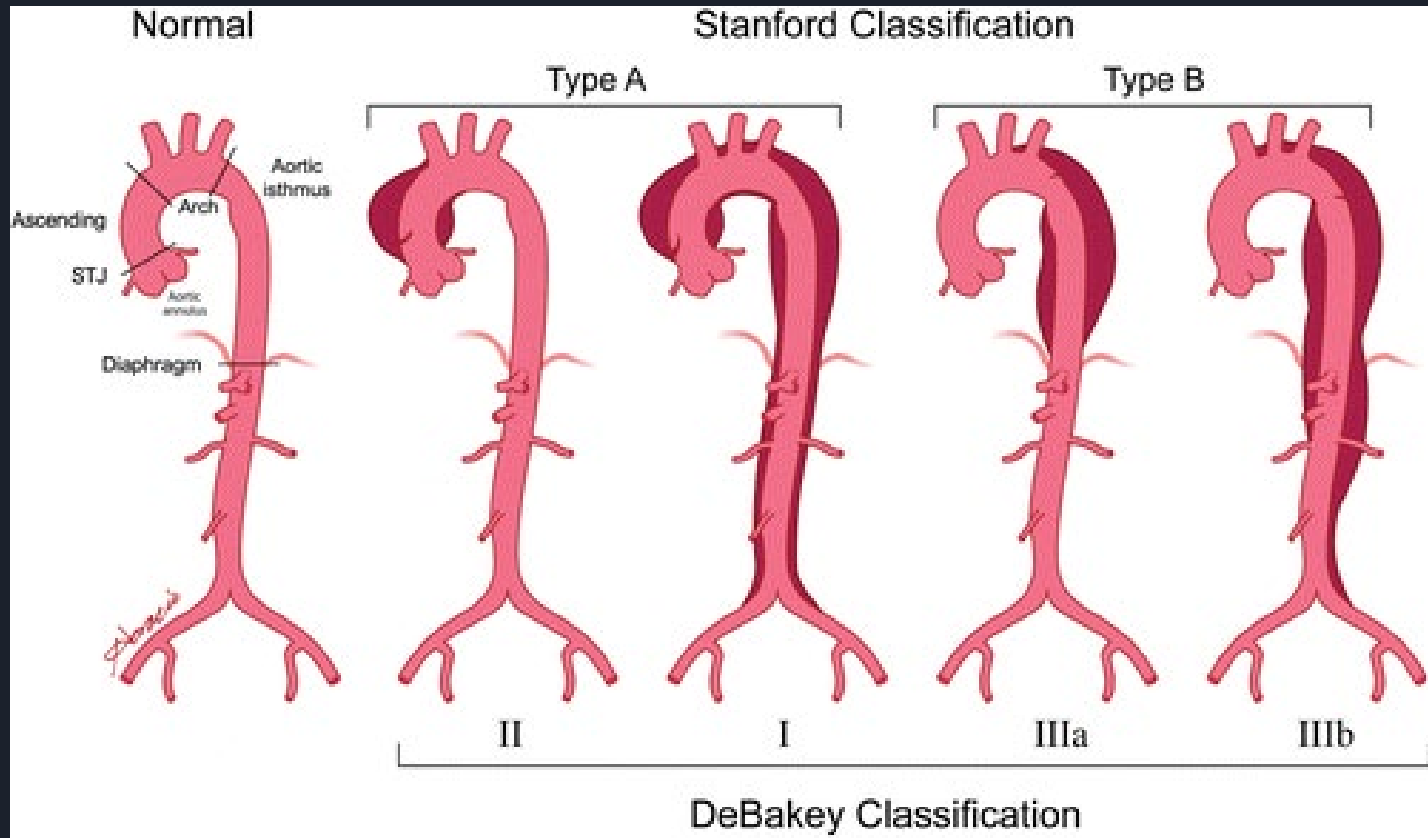
- Pain is not always classic. Sharp more than tearing. May be migratory and intermittent.
 - Not always hypertensive
 - Differential BP or pulses infrequently present
 - Mediastinal widening on CXR not sensitive
 - May have abdominal or back pain



Aortic Dissection

- Diagnosis:
 - Clinical
 - Negative D dimer can help rule out in low risk adults
 - CXR
 - CTA aorta of chest/abdomen if stable enough
 - TTE or TEE in OR if unstable
- Treatment: Esmolol, pain control, BP control after beta blockade with IV antihypertensives (target normal BP)
 - Consult CT surgery. Stanford type A dissection requires immediate surgical intervention
 - Stanford B may be medically managed

Aortic Dissection





Pediatric Syncope

- 20% of adolescents experience syncope
 - 80% are vasovagal
 - 10% neurologic such as seizure
 - 2-3% cardiac cause
- Evaluation:
 - Thorough cardiac, pulmonary, and neuro exam
 - Up to 40% of euvoletic adolescents may have abnormal orthostatic vitals
 - PMH, PSH
 - Medications
 - Family History



Myoclonus in Syncope

- Myoclonus can occur due to cerebral hypoxia
 - Shorter duration (< 1 minute) and less rhythmic than seizures
 - No post-ictal period or other seizure characteristics



Vasovagal syncope

- Can be triggered by reduced preload (ie. orthostatic), excess vagal stimulation
 - Bradycardia, vasodilation, hypotension
- Usually less than 1 minute, may be up to 5 minutes
 - Nausea and fatigue can persist
- Prodrome: warmth, nausea, lightheadedness, visual grayout or tunnel vision
- Triggers: emotional upset, sight of blood, pain, hot or crowded conditions, prolonged recumbence or standing
- Contributing factors: anemia, dehydration, pregnancy, medications
- Less common in children under 6 years old
- Consider dangerous causes of orthostatic syncope (ie. ruptured ectopic pregnancy, severe dehydration, GI bleed, etc)
- Treat with increased fluid and salt intake, frequent episodes may need midodrine



Situational syncope

- Subtype of vasovagal syncope
- Urination, defecation, coughing, swallowing, stretching, neck extension, neck pressure, hair grooming
- Overactive valsalva or carotid sinus activation
- Breath holding spells are subtype



High Risk Factors for Syncope

- Exertional event
- Triggered by sudden sympathetic response (loud noise, swimming, anger, surprise)
- Absence of prodrome or situational syncopal factors
- Family history of sudden death under age 50, sensorineural hearing loss, long QT
- Occurring while lying down
- Prolonged LOC > 5 minutes (more likely seizure or psychogenic)
- Associated chest pain or palpitations
- Recurrent episodes
- Concerning exam findings
- Use of medications that alter cardiac conduction



Syncope Workup

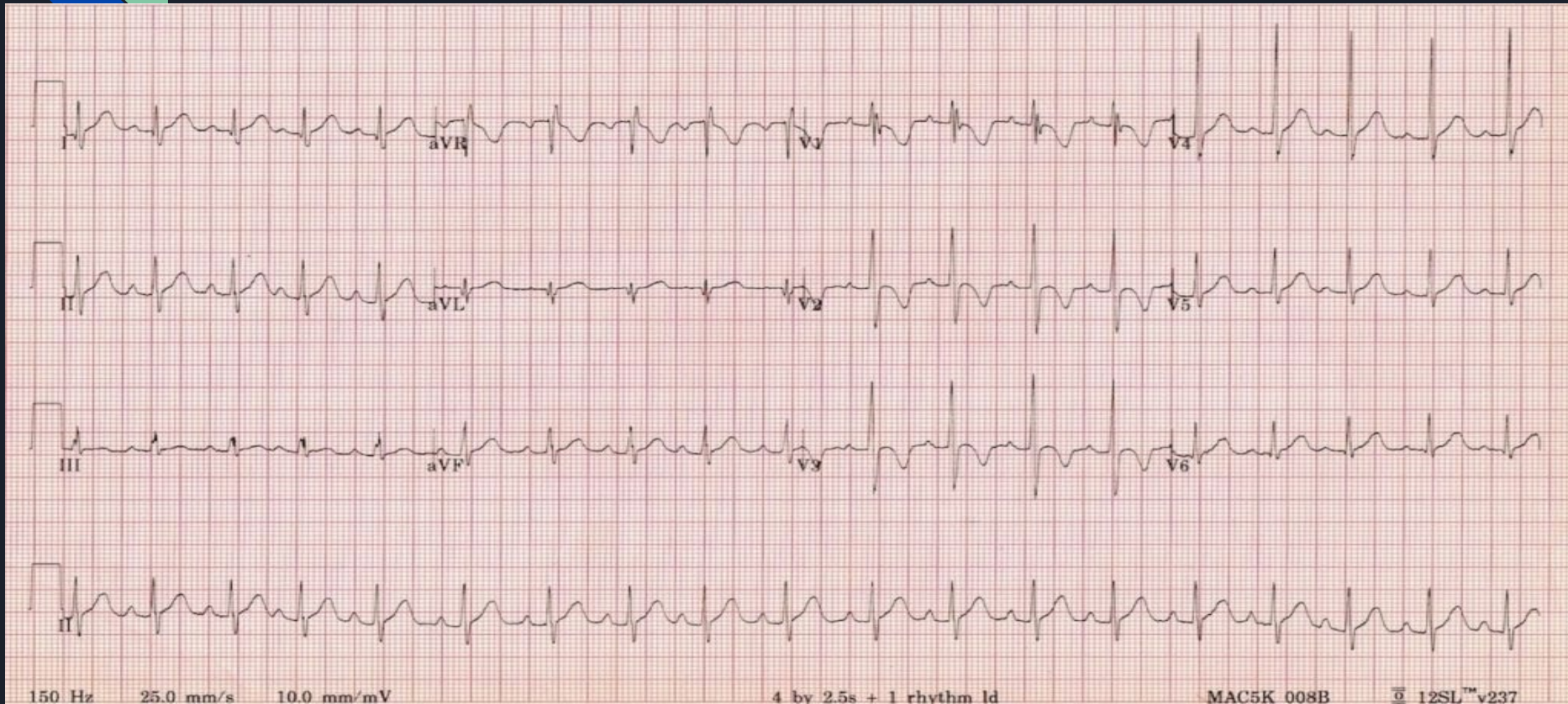
- EKG
 - Normal EKG does not rule out cardiac cause in high risk patient
 - Seattle criteria
- Urine pregnancy test in age-appropriate females
- Labs or other studies in selected cases
 - Chemistries including magnesium
 - Hemoglobin
 - Glucose
 - Thyroid function studies in tachyarrhythmias
 - Alcohol and UDS
 - Cardiac monitor
 - Echo
 - Holter or other additional testing per cardiology



Disposition

- Most high risk patients with normal EKG who are asymptomatic can follow up with cardiology promptly as outpatient.
 - Avoid exertion.
- Abnormal EKG should be discussed with cardiology for dispo
- Documented arrhythmia or ongoing symptoms should admit

Normal Pediatric EKG



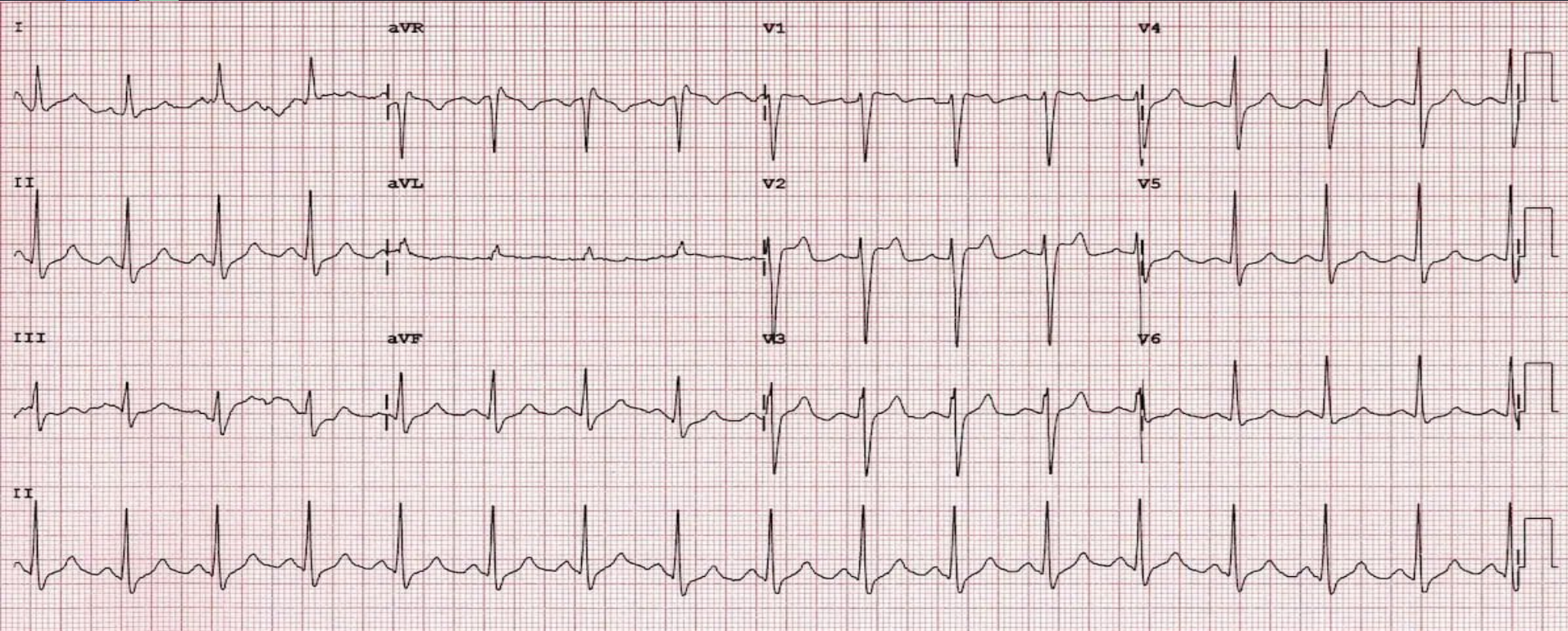


Normal Pediatric EKG

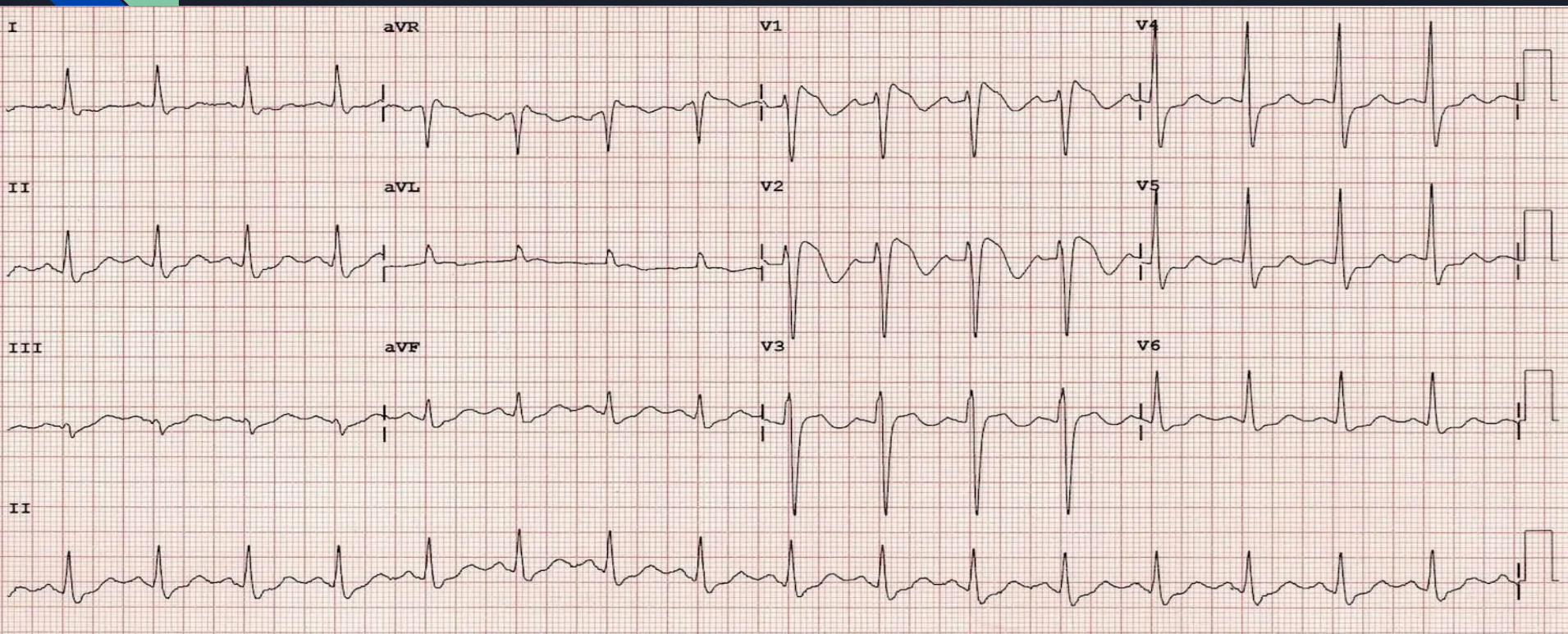
- Normal HR, intervals, and signs of hypertrophy depend on age
- Inverted T wave in V1-3 is normal
- Small Q waves in inferior and lateral precordial leads can be normal
- Right axis is normal in newborns
- Isolated monomorphic PVCs or PACs are common
- Benign early repolarization is usually in kids above age 10, more common in teenage athletes and males
- Look at V1 and V6 to determine LVH and RVH.
 - RSR' can be normal in V1 but larger R' suggests RVH
 - Q wave in V1 also suggests RVH.
 - Pure R wave in V1 also abnormal after 6 months of age.

Abnormal ECG Finding	Definition
T-wave inversion	>1 mm in depth in two or more leads V2–V6, II and aVF, or I and aVL (excludes III, aVR, and V1)
ST-segment depression	≥0.5 mm in depth in two or more leads
Pathologic Q waves	>3 mm in depth or >40 ms in duration in two or more leads (except for III and aVR)
Complete left bundle branch block	QRS ≥120 ms, predominantly negative QRS complex in lead V1 (QS or rS), and upright monophasic R wave in leads I and V6
Intraventricular conduction delay	Any QRS duration >140 ms
Left axis deviation	−30° to −90°
Left atrial enlargement	Prolonged P wave duration of >120 ms in leads I or II with negative portion of the P wave ≥1 mm in depth and ≥40 ms in duration in lead V1
Right ventricular hypertrophy pattern	R–V1 + S–V5 >10.5 mm AND right axis deviation >120°
Ventricular pre-excitation	PR interval <120 ms with a delta wave (slurred upstroke in the QRS complex) and wide QRS (>120 ms)
Long QT interval ^a	QTc ≥470 ms (male) QTc ≥480 ms (female) QTc ≥500 ms (marked QT prolongation)
Short QT interval ^a	QTc <320 ms
Brugada-like ECG pattern	High take-off and downsloping ST-segment elevation followed by a negative T wave in ≥2 leads in V1–V3
Profound sinus bradycardia	<30 bpm or sinus pauses ≥3 s
Atrial tachyarrhythmias	Supraventricular tachycardia, atrial-fibrillation, atrial flutter
Premature ventricular contractions	≥2 per 10-s tracing
Ventricular arrhythmias	Couplets, triplets, and nonsustained ventricular tachycardia

16 y.o. M with family hx of early sudden death



Same 16 y.o. M with syncope during febrile illness





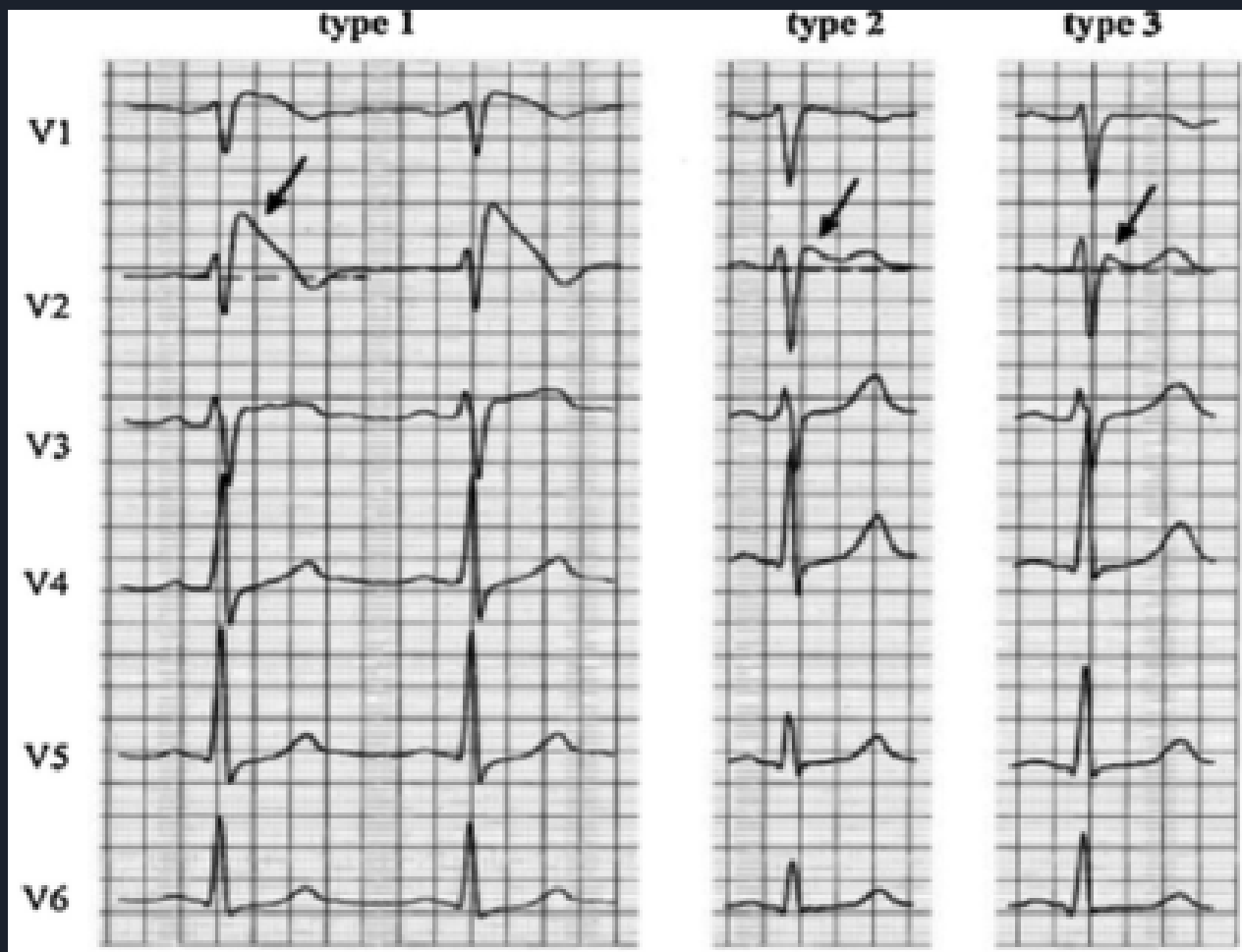
Brugada Syndrome

- Prevalence 1:2000 but most often presents in adulthood
- 3 types of EKG appearance, only type 1 is diagnostic
- EKG findings not always present, may be more present with fever, alcohol, electrolyte abnormality, sodium channel blockers.
- EKG plus clinical symptoms
 - Documented Vfib or polymorphic Vtach
 - Syncope
 - Coved type EKG in family members
 - Nocturnal agonal respiration
 - Inducibility of Vtach on programmed electrical stimulation
 - Sodium channel blockade induced changes on EKG
- Mortality of 10% per year if undiagnosed

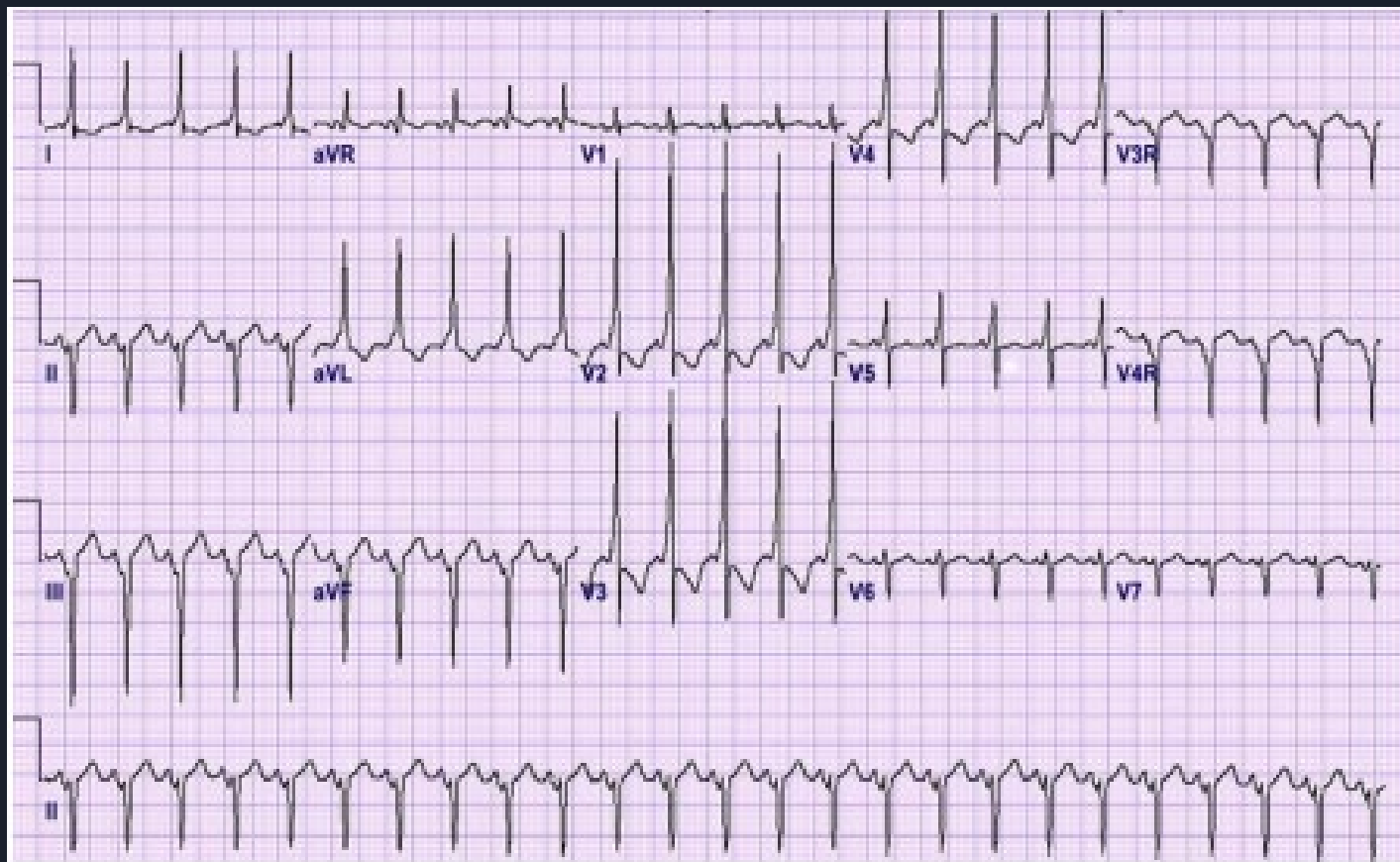


Brugada Syndrome

- Treated with implantable ICD, sometimes quinidine
- Treat fevers
- Avoid triggering medications:
 - sodium channel blockers
 - Some psychotropics
 - Antihistamines
 - Anesthetic/analgesics
 - Alcohol intoxication



13 y.o. F with dizziness





WPW

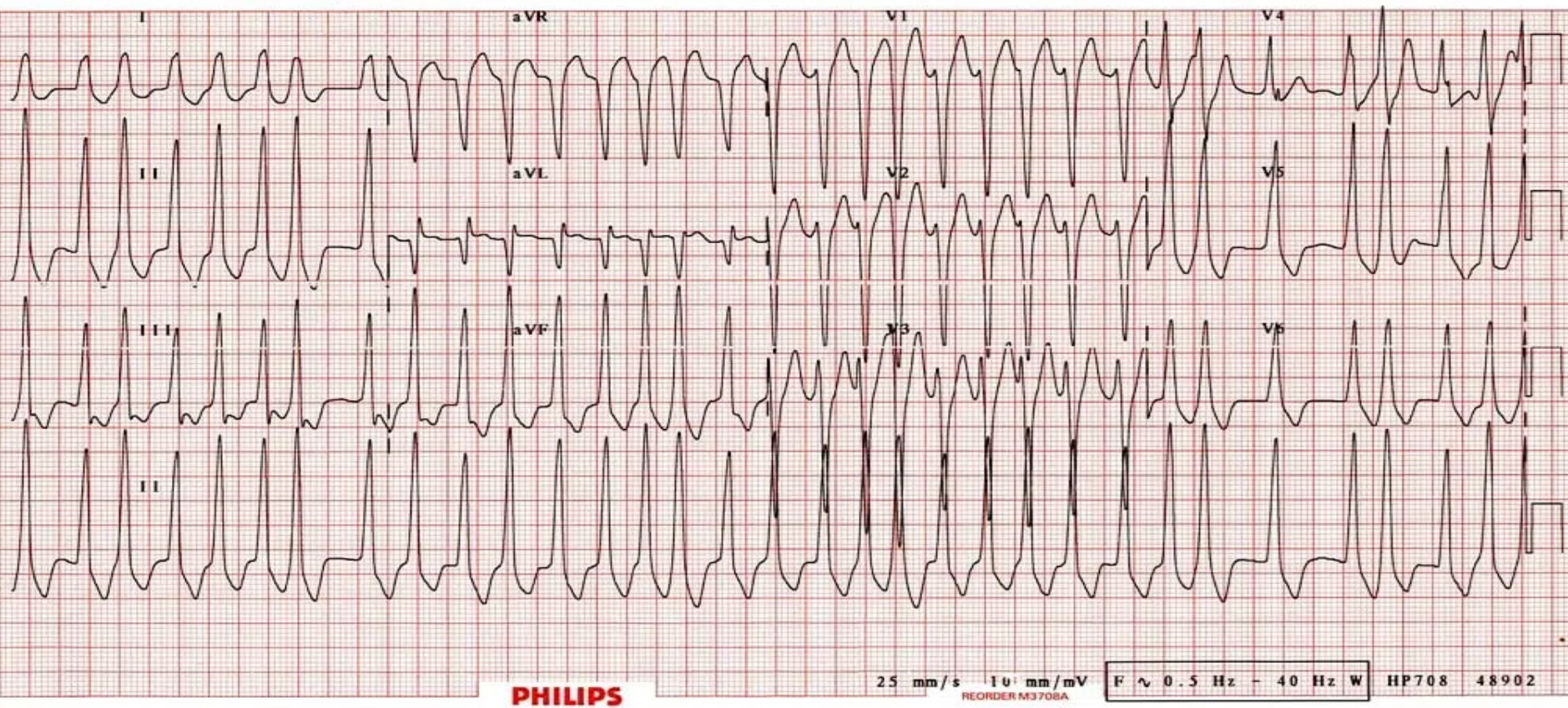
- Short PR, delta wave, QRS > 110 ms
- May have “pseudo-infarction pattern” with negative delta wave mimicking Q wave
- Discordant ST segment and T wave changes (may look like LV strain)
- Findings may be subtle when in sinus rhythm
- More prominent after a tachydysrhythmia resolves
- Verapamil, digoxin, and flecainide contraindicated



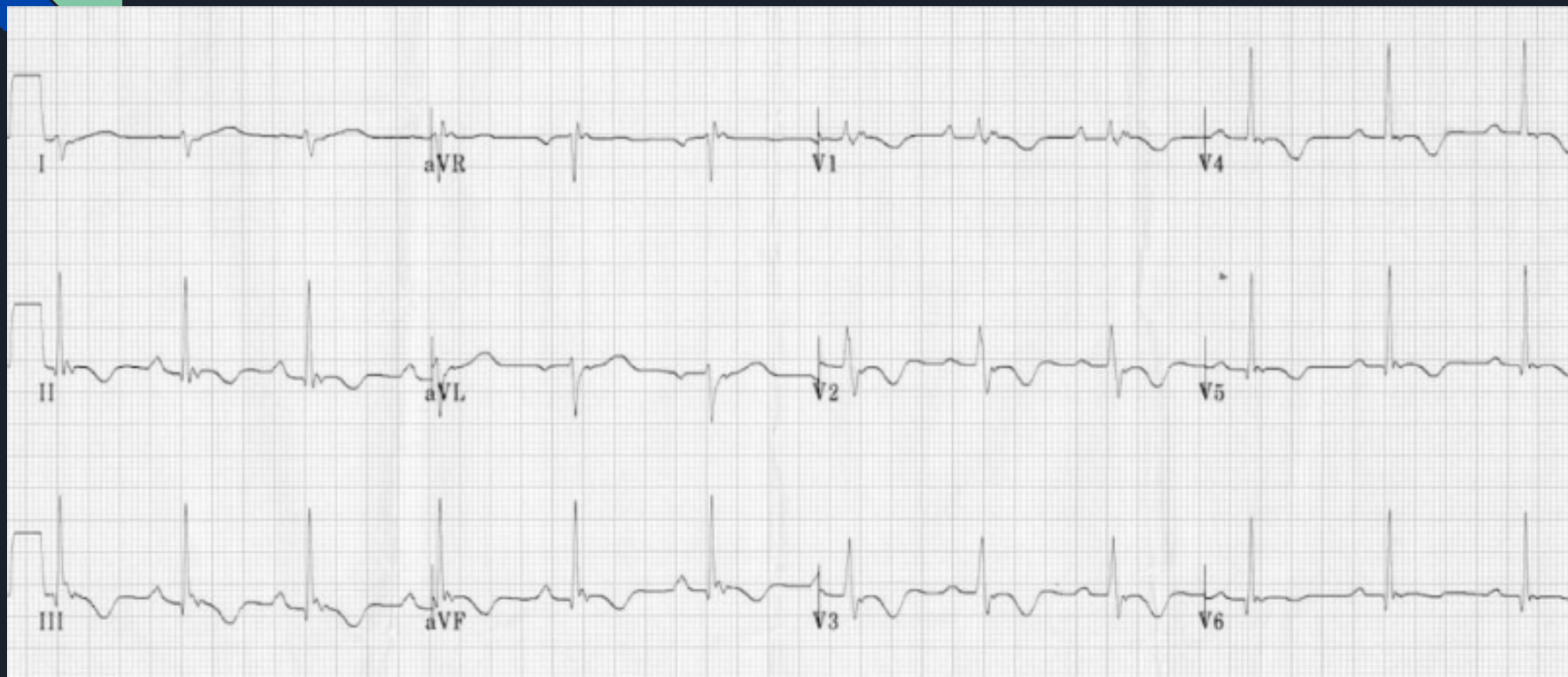
WPW Treatment

- In orthodromic AVRT (narrow QRS), can treat as typical SVT
 - Vagal maneuvers
 - Use of adenosine may induce afib/flutter with RVR
 - Can use beta blockers
- In antidromic AVRT (wide QRS)
 - Looks similar to Vtach, safest thing is to treat like Vtach
 - Procainamide if stable
 - Synchronized cardioversion if unstable
- Afib is often wide complex with varying QRS morphologies, very fast
 - Avoid nodal blocking agents as they may precipitate Vfib
 - Can use procainamide if stable
 - Synchronized cardioversion if unstable
- Definitive treatment is radiofrequency ablation.

Afib in WPW



18 year old with syncope





Arrhythmogenic Right Ventricular Cardiomyopathy

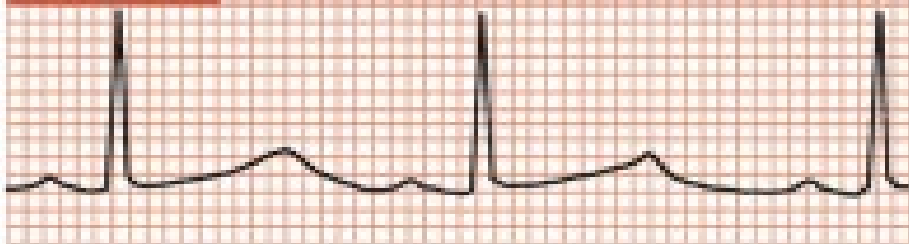
- Progressive fibrofatty replacement of cardiac muscle
- 1:2000 to 1:5000 incidence
- Autosomal dominant with incomplete penetration
- Usually presents in teenage years or young adulthood
- Causes focal structural heart changes, but EKG changes may be seen first
 - Echo and cardiac MRI have limited sensitivity
- T wave inversions in V1-3 at age > 14, epsilon wave in V1-3, terminal QRS prolongation, PVCs, and Vtach with LBBB morphology
- Syncope, palpitations, sudden cardiac death, or heart failure
- Treatment: ICD, beta blockers, heart failure management



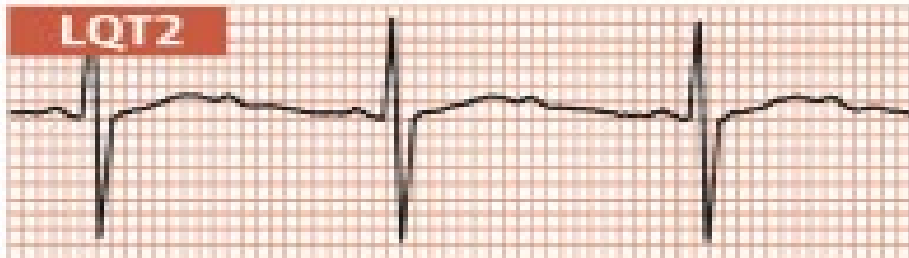
Long QT syndrome

- Congenital or medication induced
 - Romano Ward: autosomal dominant
 - Jervell Lange-Nielsen: autosomal recessive, sensorineural hearing loss
 - Congenital prevalence is about 1:2000
 - Mostly voltage gated potassium channel abnormalities
 - May also have epilepsy
- QTc over 450ms is significantly prolonged
 - May be intermittently present
 - Infants under 6 months have longer normal QTc, < 490 ms
 - 460ms in older females
- Usually no arrhythmia until QTc around 500ms or above. Above 480ms on repeat EKGs is highly suggestive
- T wave alternans or notching may be suggestive
- Bigeminy with long QT can degenerate into torsades rapidly

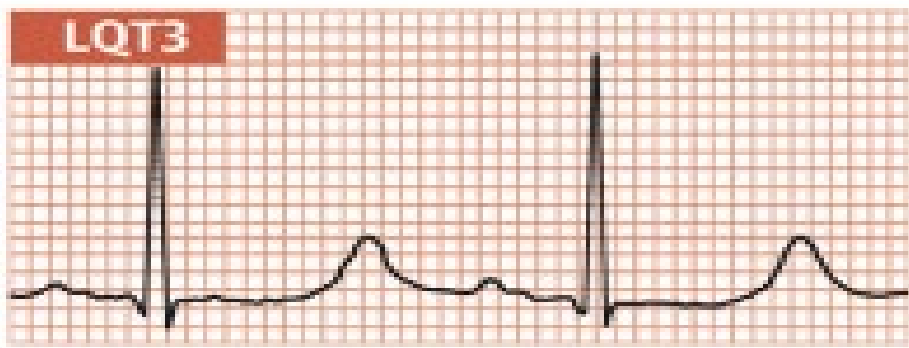
LQT1



LQT2



LQT3





Long QT Syndrome

- Treat with beta blockers, may need ICD
- Note: short QT syndrome: $QTc < 340$ ms
- Avoid QT prolonging meds
 - Ipratropium instead of albuterol
 - Antiarrhythmics class 1A, 1C, III
 - Zofran
 - Propofol
 - Tacrolimus
 - Azole antifungals
 - Methadone
 - Loperamide
 - Triptans
 - Antipsychotics
 - Antidepressants

HR 90 PVC 9 RESP 22 T1 38.3

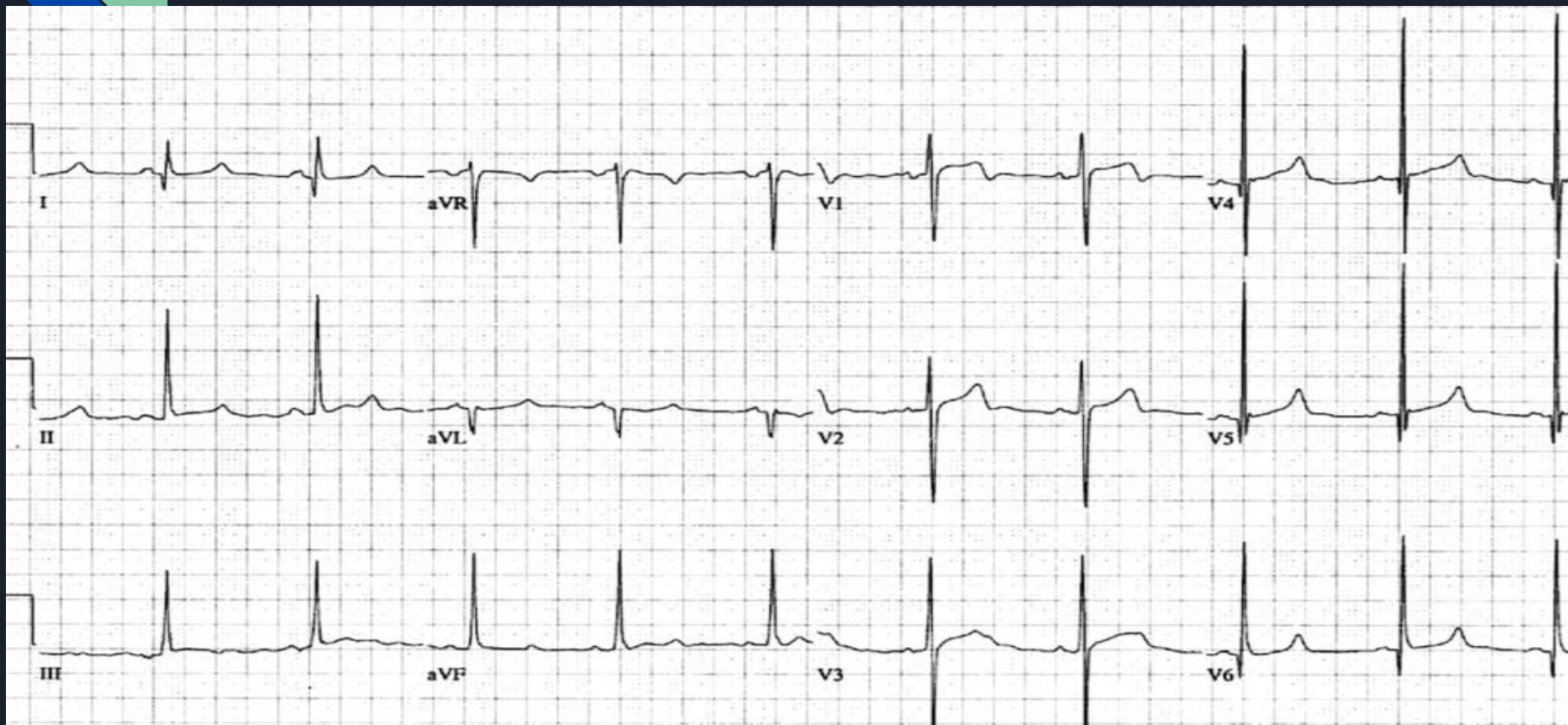


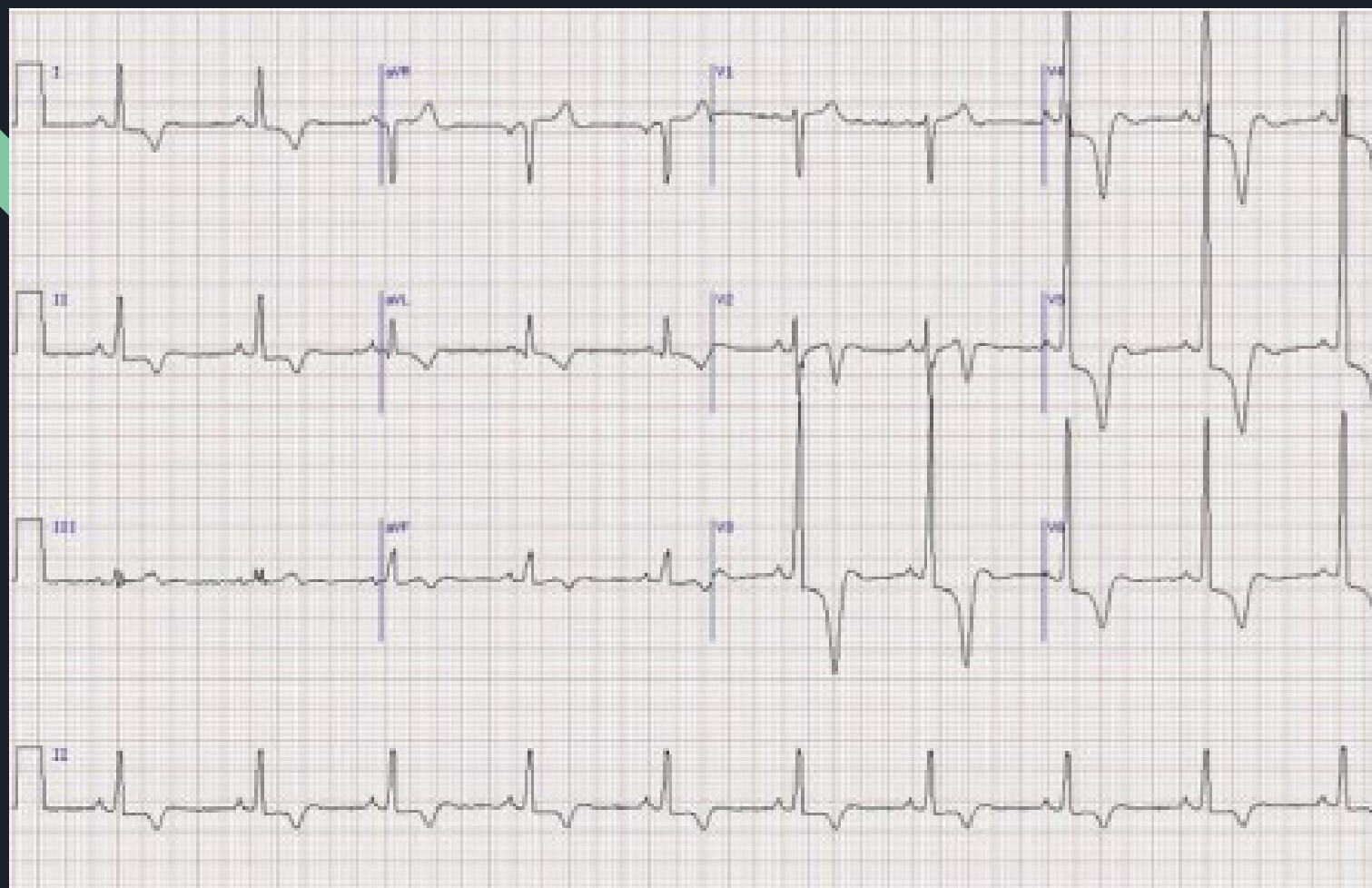


Torsades treatment

- Magnesium 50mg/kg over 10 minutes up to 2g
- Cardiovert if unstable
- Do not leave the room!
- Amiodarone may not be indicated
- If recurrent likely needs overdrive pacing with dopamine or isoproterenol
- Differential diagnosis of torsades:
 - Coarse Vfib
 - WPW with afib and varying morphology
 - Polymorphic Vtach with normal QT interval: MI, takotsubo's, or catecholaminergic ventricular tachycardia

15 y.o athlete with syncope







Hypertrophic Cardiomyopathy

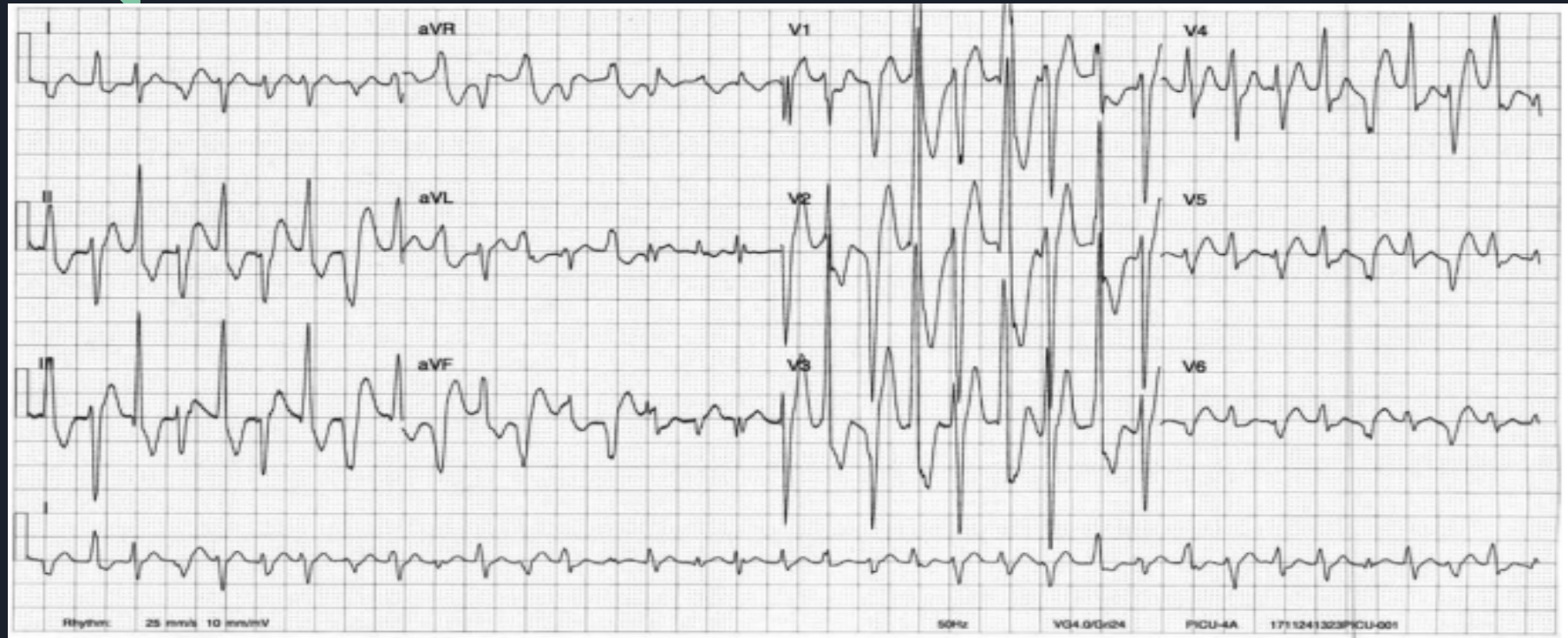
- Incidence of about 1 in 500
- Most common cause of sudden cardiac death in young people, mortality 1-2% per year
- Symptoms: exertional chest pain or exercise intolerance, syncope or presyncope, arrhythmia, CHF.
 - May be asymptomatic and found incidentally on EKG or CXR
- Dynamic LVOT obstruction may be present
 - Ejection murmur decreases with increased preload or increased afterload
- Diastolic dysfunction



Hypertrophic Cardiomyopathy

- EKG findings: LVH, dagger Q waves in lateral leads +/- inferior leads, nonspecific ST-T wave abnormalities
- Treatment:
 - Beta blockers improve diastolic dysfunction and LVOT obstruction.
 - Septal reduction surgery may be needed.
 - ICD if events of sustained vtach or cardiac arrest.

8 y.o. F unconscious while swimming





Catecholaminergic Polymorphic Ventricular Tachycardia

- Prevalence of 1 in 10,000
- Syncope or cardiac arrest triggered by exercise, water immersion or emotion. Most commonly age 4-14.
- Diagnose with exercise induced ventricular ectopy, bidirectional or polymorphic Vtach. Normal QT interval.
- Mutations in cardiac ryanodine receptor gene in 50-60%, can be de novo
- Suspect based on multimorphic ventricular ectopy between arrests
- Bidirectional Vtach is almost pathognomonic, though also described in Andersen Tawil syndrome and digoxin overdose



Catecholaminergic Polymorphic Ventricular Tachycardia

- Treatment of Vtach/Vfib storm is avoidance of catecholamines
- Initiation of fentanyl 10 mcg/kg and general anesthetics
- May require ECMO if persistently hemodynamically unstable
- Flecainide or beta blockers may help with rhythm control
- Treated long term with left heart sympathectomy and ICD
- Huffing inhalants can also cause Vtach where Epi is harmful
 - May treat with esmolol



Summary

- High index of suspicion for cardiac disease in respiratory illness
- Thoughtful resource utilization for chest pain and syncope presentations
- Strong familiarity with normal and abnormal pediatric EKG
- Bug your cardiologists! But not for everything.



Sources

1. Bellamy D, Nuthall G, Dalziel S, Skinner JR. Catecholaminergic Polymorphic Ventricular Tachycardia: The Cardiac Arrest Where Epinephrine Is Contraindicated. *Pediatr Crit Care Med*. 2019 Mar;20(3):262-268.
2. Maron et al. Management of Hypertrophic Cardiomyopathy: JACC State-of-the-Art Review. *J Am Coll Cardiol*. 2022 Feb 1;79(4):390-414.
3. Shah SR, Park K, Alweis R. Long QT Syndrome: A Comprehensive Review of the Literature and Current Evidence. *Curr Probl Cardiol*. 2019 Mar;44(3):92-106.
4. Krahn et al. Congenital Long QT Syndrome. *JACC Clin Electrophysiol*. 2022 May;8(5):687-706.
5. Bosman LP, Te Riele ASJM. Arrhythmogenic right ventricular cardiomyopathy: a focused update on diagnosis and risk stratification. *Heart*. 2022 Jan;108(2):90-97.
6. Krahn et al. Arrhythmogenic Right Ventricular Cardiomyopathy. *JACC Clin Electrophysiol*. 2022 Apr;8(4):533-553.
7. Zavala et al. Pediatric Syncope: A Systematic Review. *Pediatr Emerg Care*. 2020 Sep;36(9):442-445.
8. Schunk PC, Ruttan T. Pediatric Syncope: High-Risk Conditions and Reasonable Approach. *Emerg Med Clin North Am*. 2018 May;36(2):305-321.
9. Krahn AD, Behr ER, Hamilton R, Probst V, Laksman Z, Han HC. Brugada Syndrome. *JACC Clin Electrophysiol*. 2022 Mar;8(3):386-405.



Sources

10. Barbut G, Needleman JP. Pediatric Chest Pain. *Pediatr Rev.* 2020 Sep;41(9):469-480.
11. Williams JL, Jacobs HM, Lee S. Pediatric Myocarditis. *Cardiol Ther.* 2023 Jun;12(2):243-260.
12. Freedman et al. Pediatric Myocarditis: Emergency Department Clinical Findings and Diagnostic Evaluation. *Pediatrics* (2007) 120 (6): 1278–1285.
13. Law et al. Diagnosis and Management of Myocarditis in Children: A Scientific Statement From the American Heart Association. *Circulation.* 2021 Aug 10;144(6):e123-e135.
14. Tintinalli JE, Stapczynski J, Ma O, Yealy DM, Meckler GD, Cline DM. eds. *Tintinalli's Emergency Medicine: A Comprehensive Study Guide, 8e.* McGraw Hill; 2016.