JOHNS HOPKINS ALL CHILDREN'S HOSPITAL

Syncope
Clinical Pathway

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All Children's Hospital

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Updated: September 2021
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Rationale:

This protocol was developed by a consensus group of JHACH emergency medicine physicians, cardiologists, neurologists and resident physicians to standardize the management of patients ages 8-22 years presenting with a pre-syncopal or syncopal episode. It addresses the following clinical questions or problems:

1. How does one differentiate benign causes of syncope from life-threatening causes of transient loss of consciousness?
2. What is the utility of EKG and orthostatic vitals?
3. When should you consult Cardiology?
4. When should you consult Neurology?
5. When should you consider admission for further evaluation?
6. When should you provide either Cardiology or Neurology outpatient follow-up?

Background

Syncope is a symptom of various disease processes and is defined as a transient loss of consciousness that is abrupt and self-limited, with complete rapid recovery thereafter. In the pediatric population, syncope is a common presentation and thought to account for 3% of all pediatric visits to the Emergency Department[1]. Neurally-mediated syncope is even more common and thought to account for 60-80% of all syncopal cases in children[2]. It is estimated that another 10% of cases are secondary to cardiac disease, 5% due to misdiagnosed seizures and up to 17% of cases due to unexplained etiology. Given this variability and potential for life threatening etiology, evaluation of these patients is often inconsistent, time consuming and can require extensive resource utilization.

It is important to note that older definitions of syncope in some literature are broad and this term historically was used synonymously with transient loss of consciousness which would thereby include seizure, stroke and head trauma. The current definition of syncope is more deliberate and per the most current AHA/ACC definition, syncope is its own distinct entity that must be differentiated from these other forms of transient loss of consciousness[3].

The purpose of this guideline is to provide a standardized, evidence-based algorithm for managing pediatric syncope in the emergency department. The goal of this algorithm is to provide a stream-lined approach to syncope that allows for effective screening of life-threatening etiologies of transient loss of consciousness, limits unnecessary testing, reduces the patient's length of stay in the emergency department and therefore also reduces the patient's overall healthcare cost. The guideline includes patient ages from 8 years to 22 years as these are the included ages in the largest studies providing the best evidence[4].

Pathophysiology

Syncope is the result of a pathologic mechanism that leads to global cerebral hypoperfusion and subsequent insufficient blood flow to the reticular activating system (RAS). The RAS is a neuronal network in the brainstem that is responsible for supporting consciousness[4]. In order to maintain perfusion to the RAS, the body requires a functional cardiovascular system with adequate volume and systemic vascular resistance. The cessation of cerebral blood flow for 6-8 seconds can cause loss of consciousness, although it seems that some individuals may be more sensitive than others[5].

The specific mechanism leading to the cerebral hypoperfusion seen in syncope can vary. Neurally-mediated/reflex syncope, the classification vaso-vagal syncope belongs to, is the most frequent cause of loss of consciousness. This form of syncope is triggered by stimulation of peripheral receptors via sympathetic and vagal afferent fibers to the RAS, which ultimately leads to reduced efferent sympathetic activity, increased vagal tone and subsequent diminished cardiac output[4]. This pathway can also be triggered by neurohumoral agents from the cortex, limbic system and hypothalamus as seen in the case of emotion or fear. Orthostatic syncope differs from reflex syncope in that the autonomic nervous system fails to compensate in the setting of reduced blood flow during orthostatism. Lastly, cardiac syncope induces a reduction of cerebral perfusion due to impaired cardiac output[4].

It is also important to note that syncope itself, falls within the categorization of transient loss of consciousness (TLOC). There are multiple etiologies of TLOC (see table below) and it is important that syncope be differentiated from trauma, epileptic seizures, stroke, metabolic disorders, psychogenic disorders and rare miscellaneous disorders[5]. Typically, syncope is brief and lasts less than 20 seconds, although rarely it can last for minutes[6]. This can make distinguishing syncope from other causes of prolonged loss of consciousness difficult. However, by definition, syncope is accompanied by almost immediate restoration of behavior and orientation and this can help differentiate from other forms of loss of consciousness[5].

Table 1
Causes of Transient Loss of Consciousness

Syncope	Trauma	Neurologic	Metabolic	Psychogenic	Miscellaneous
• Reflex • Orthostatic • Cardiac	• Concussion • Intracranial hemorrhage	• Epilepsy • Stroke • TIA	• Hypoglycemia • Hypoxia • Hypocapnia • Pregnancy	• Psychogenic pseudo-syncope • Conversion disorder	• Cataplexy • Drop attacks

Table 2
Classification of Syncope

Reflex/Neurally-Mediated Syncope	Orthostatic Syncope	Cardiac Syncope
Vasovagal: • Emotional distress: fear, pain, phobia Situational: • Cough, Sneeze, Post-micturition, Post-prandial, Post-exercise Carotid Sinus Syncope	Autonomic Failure: • Diabetes, Uremia, Spinal Cord Injuries Drug Induced Orthostatic Hypotension: • Alcohol, Diuretics, Vasodilators, Antidepressants Volume Depletion: • Hemorrhage, Diarrhea, Vomiting, etc	Arrythmia: • Sinus Node Dysfunction • AV Node Block • Implanted device malfunction • Tachyarrhythmias: SVT, Channelopathy, Prolonged QTc Structural Diseases: • Valvular disease, cardiomyopathy, masses, pericardial disease, coronary artery anomalies Other: • PE, Aortic Dissection, Pulmonary HTN

Evaluation

The evaluation of any patient presenting with TLOC should begin with thorough history taking. It is important to determine whether the event reflects true syncope versus other forms of loss of consciousness. If the patient presents unconscious or is ill appearing, then appropriate resuscitative measures should be undertaken as per PALS guidelines[1]. The episode should be described in detail including patient's memory of the event, preceding symptoms, and subsequent symptoms. Injuries resulting from the episode should also be identified including closed head injury, laceration, factures, etc.

When taking the history, a cardiac and neurological history are vital in identifying red flag symptoms. For cardiac history, it is important to identify any personal or family cardiac history. This may include first- or second-degree relatives with sudden cardiac arrests before age 50, history of arrythmia, or sudden unexpected modes of death such as single car accidents or drowning of a strong swimmer. History of cardiac catheterization may result in scar tissue and result in delayed presentation of heart block and syncope later. Preceding symptoms of palpitations, syncope with exertion or chest pain, syncope in the setting of strong emotional or painful stimuli, and multiple episodes of syncope with increase in frequency are considered red flag symptoms and patient should be evaluated for arrhythmia and cardiac disease.

Neurological history is another important factor to consider when evaluating TLOC in pediatric patients. Seizures are an important and frequent cause of transient loss of consciousness. A history of convulsions, tonic or clonic activity, post-ictal state and family history of seizure disorders are important components of history to inquire about. Furthermore, a history of substance ingestion, and a thorough medication history can also elucidate causes of syncope.

Emergency Center Syncope Clinical Pathway

Inclusion Criteria:
8 years to 22 years

Exclusion Criteria:
- Ill appearing
- Needing sustained CPR
- Congenital heart disease
- Previous cardiac surgery
- Significant neurologic disease
- Significant co-morbidities

Cardiac Red Flags:
1. Palpitations preceding syncope?
2. Multiple episodes of syncope within a short period?
3. Exertional syncope?
4. Emotional syncope?
5. Syncope with chest pain?
6. Sudden cardiac death in family members (1st or 2nd Degree) <50 yrs of age?
7. History of familial arrhythmias?

Neurologic Red Flags:
1. Concern for seizure or post-ictal state?
2. Concern for severe secondary headache?
3. Valsalva induced syncope?

Green Light:
1. History consistent with reflex mediated syncope (see Table 1).
2. Normal EKG and negative urine HCG.
3. Normal exam and at baseline.

12 Lead EKG:
- Wolff-Parkinson-White Syndrome
- Hypertrophy (right or left ventricle)
- Pathologic abnormal repolarization (not non-specific changes) or pathologic Q waves
- PR interval prolongation (first degree AV block, PR > 200 msec)
- QT interval prolongation
- Bundle branch block
- Tachyarrhythmias
- AV interval block (second or third)
- >1 Premature Atrial Contractions or Premature Ventricular Contractions
- Sinus bradycardia < 50 bpm
- Epsilon wave in V1-V3

Patient presents to EC with syncope

1. 12 Lead EKG.
2. Orthostatic vital signs.
3. If post-pubertal female, obtain urine HCG.
— In Triage.

1. History & Physical Exam.
2. Cardiorespiratory monitoring.
3. Routine laboratory testing not recommended.

Abnormal EKG, Red Flags or Abnormal Cardiac Exam? — Yes → Consult Cardiology to assess further workup and disposition.

No

Abnormal Red Flags or abnormal Neuro Exam? — Yes → Consult Neurology to assess further workup and disposition.

No

Is there orthostatic intolerance or abnormal orthostatic vitals? — Yes → 1. Administer supportive care. 2. 10 to 20 ml/kg of IV or oral fluid resuscitation for symptomatic hypovolemia.

No

Symptoms have resolved and exam reassuring. — Yes → Discharge home with syncope handout and have discussion including adequate oral intake, avoiding provoking events and recognizing vasovagal events if applicable.

No

Consider further testing, longer observation or consultation with Cardiology or Neurology for admission.

Orthostatic Vitals:
1. Patient should lay supine and flat for 5 minutes. Obtain blood pressure.
2. Patient should be seated upright for 1 minute. Obtain blood pressure.
3. Patient should stand steadily without movement for 3 minutes. Obtain blood pressure.

Orthostatic Hypotension[3]:
Sustained reduction of systolic blood pressure of at least 20 mmHg or diastolic blood pressure of 10 mmHg within 3 minutes of standing.

Orthostatic Tachycardia[1,3]:
Sustained heart rate increase of $\geq$30 beats/min ($\geq$40 beats/min for ages 12-19) within 10 minutes of standing. In the setting of orthostatic intolerance, this may be suggestive of Positional Orthostatic Tachycardia Syndrome. Further assessment, ideally done by Cardiology, is **required** to make this diagnosis.
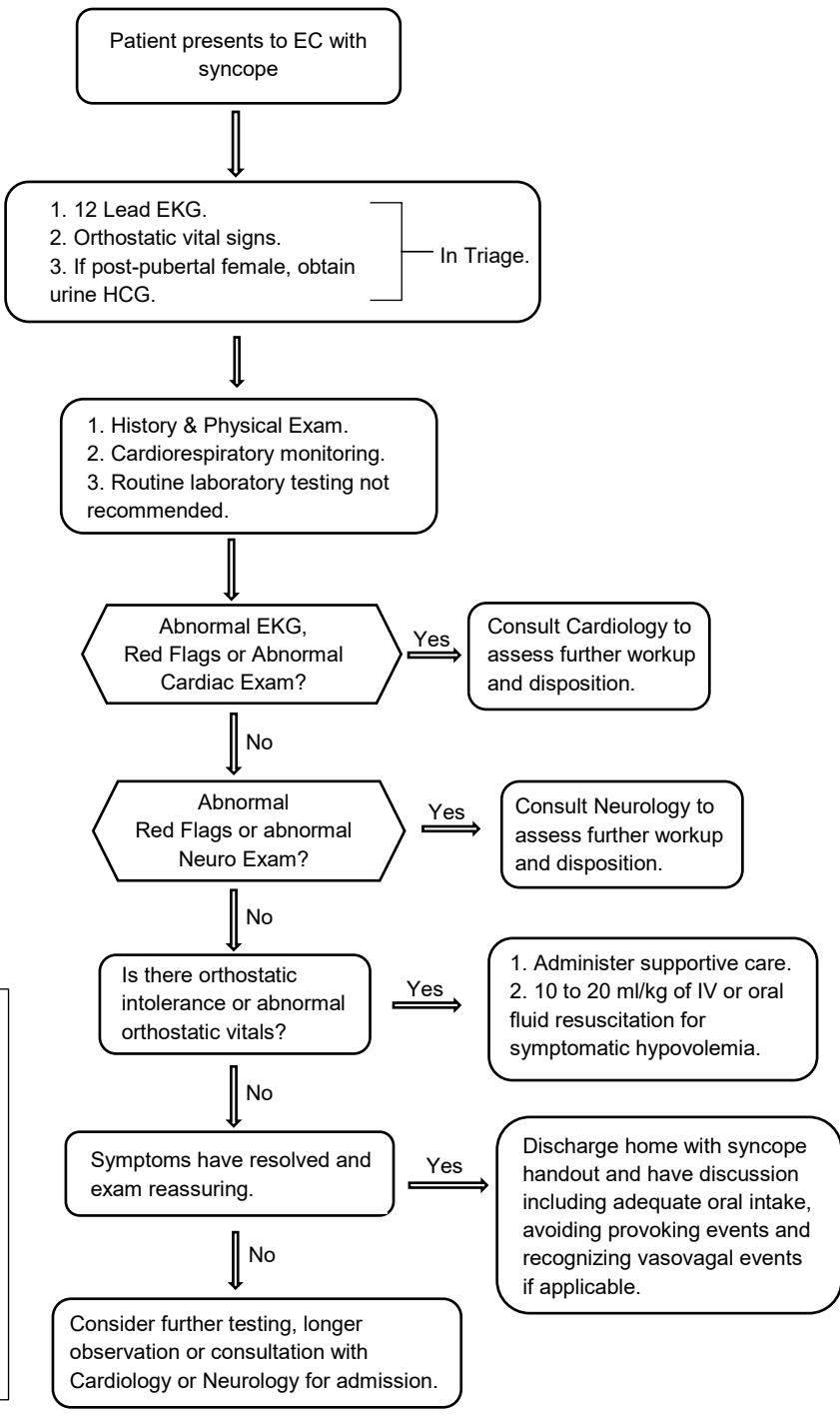
Emergency Center Management

Electrocardiography (EKG):

Per AHA/ACC guidelines, routine EKG is recommended for all pediatric patients who present with syncopal episode[3]. Per Ritter et al, a history of exercise induced syncope, positive family history, abnormal physical exam, or abnormal ECG had 96% sensitivity in identifying cardiac etiology of syncope in a retrospective study including 480 previously healthy children.

Laboratory Studies:

Per AHA/ACC guidelines, routine blood tests are not indicated in patients who present with syncope[3]. There is limited pediatric data regarding the role of laboratory studies in syncope. Per adult data, irregularities in blood sugar, electrolytes and hemoglobin were found to be the underlying cause of syncope in only 2-3% of all adult patients including those with seizures[7]. This testing was confirmatory of clinical suspicion in almost all cases. Bleeding as a cause of syncope was diagnosed clinically and confirmed with CBC or fecal occult blood test[7].

Orthostatic Vital Signs:

Orthostatic intolerance (OI) is a common clinical feature in children, especially adolescents. Much of the literature regarding pediatric OI is based on utilizing the adult definition of orthostatic vital sign abnormalities as formulated by an expert consensus panel[8]. One pediatric study including 300 high school aged children in the US revealed that over 33% had symptoms of orthostatic intolerance and 25% met vital sign criteria for postural orthostatic tachycardia syndrome[9]. It also indicated that adolescents with OI had statistically significant higher heart rate changes at 2 and 5 minutes in the upright position than those without OI. This study, amongst others, have noted that postural vital sign changes in children often reveal heart rate changes that are greater than what is accepted in adults. However, there is poor sensitivity and specificity for a "cut-off" for a heart rate that predicts OI. Of note, the expert consensus panel does acknowledge this greater variation and sets a larger heart rate increment for patients 12-19 years of age[8]. Adult emergency department studies have also indicated poor sensitivity of positive orthostatic vitals in scenarios of hypovolemia, including dehydration and blood loss, leading to questioning of the utility of obtaining orthostatic vitals in the emergency department[10]. However, given the known physiologic response differences in the pediatric population and correlation of OI with higher upright orthostatic heart rates, our guideline will recommend continuing to obtain orthostatic vitals using the format as stated in the algorithm. Of note, the formal diagnosis of Positional Orthostatic Tachycardia Syndrome should not be made in the emergency department as more elements are needed for this diagnosis.

Echocardiography:

Per AHA/ACC practice guidelines, routine echocardiography is not recommended in syncope management and only indicated when there is concern for structural heart disease[3].

Routine screening with echocardiography is not indicated in healthy pediatric patients who present with syncope. In a retrospective study of 322 previously healthy pediatric patients who presented with syncope, the overall sensitivity and positive predictive value of echocardiography for detecting a cardiac related syncopal event was 18% and 11% respectively[11].

Ambulatory Electrocardiography (AEKG):

Per AHA/ACC practice guidelines for ambulatory electrocardiography, the role of AEKG in young patients with syncope in the absence of structural or functional heart disease is limited[3]. This limited utility of AEKG is reflected in multiple studies that reviewed either unspecified syncope or presumed non-cardiac syncopal etiologies [12-14]. However, the AHA/ACC guidelines do provide a strong recommendation in obtaining AEKG in pediatric patients with exertional syncope when the cause is otherwise unknown[3].

Neuroimaging:

Per AHA/ACC practice guidelines, neuroimaging is not recommended in the routine evaluation of patients presenting with syncope. Similar recommendations are also made in pediatric studies involving syncope. Multiple retrospective studies have demonstrated low diagnostic yield and increased costs associated with obtaining a head CT in pediatric syncope without specific abnormal neurologic findings[13,15,16]. However, the presence of focal neurologic deficits in the context of syncope would be a specific indication for obtaining neuroimaging[3,15].

Electroencephalogram (EEG):

Per AHA/ACC practice guidelines, EEG is not recommended in the evaluation of patients with syncope unless there is concern for seizures. Other pediatric literature has reviewed how EEG is often obtained on children to potentially exclude syncope, but findings are rarely abnormal[2,17,18]. Patients who exhibit prolonged loss of consciousness, seizure-like activity and post-ictal confusion should have further evaluation with an EEG. Otherwise, the positive yield of EEG has been reported as being 1 in 300 studies[19].

References

1. Fant C, Cohen A. Syncope In Pediatric Patients: A Practical Approach To Differential Diagnosis And Management In The Emergency Department. *Pediatric emergency medicine practice*. 2017;14:1-28.

2. Moodley M. Clinical Approach to Syncope in Children. *Seminars in Pediatric Neurology*. 2013;20(1):12-17. doi:https://doi.org/10.1016/j.spen.2012.12.003

3. Shen Win-Kuang, Sheldon Robert S., Benditt David G., et al. 2017 ACC/AHA/HRS Guideline for the Evaluation and Management of Patients With Syncope: Executive Summary: A Report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines and the Heart Rhythm Society. *Circulation*. 2017;136(5):e25-e59.

4. Shanahan KH, Monuteaux MC, Brunson D, et al. Long-term Effects of an Evidence-based Guideline for Emergency Management of Pediatric Syncope. *Pediatric Quality & Safety*. 2020;5(6). doi: 10.1097/pq9.0000000000000361

5. Task Force for the Diagnosis and Management of Syncope, European Society of Cardiology (ESC), European Heart Rhythm Association (EHRA), et al. Guidelines for the diagnosis and management of syncope (version 2009). *Eur Heart J*. 2009;30(21):2631-2671. doi:10.1093/eurheartj/ehp298

6. Hoefnagels WAJ, Padberg GW, Overweg J, van der Velde EA, Roos RAC. Transient loss of consciousness: The value of the history for distinguishing seizure from syncope. *Journal of Neurology*. 1991;238(1):39-43. doi:10.1007/BF00319709

7. Kapoor WN. Syncope. *N Engl J Med*. 2000;343(25):1856-1862. doi:10.1056/NEJM200012213432507

8. Freeman R, Wieling W, Axelrod FB, et al. Consensus statement on the definition of orthostatic hypotension, neurally mediated syncope and the postural tachycardia syndrome. *Clinical Autonomic Research*. 2011;21(2):69-72. doi:10.1007/s10286-011-0119-5

9. Skinner JE, Driscoll SW, Porter CJ, et al. Orthostatic Heart Rate and Blood Pressure in Adolescents: Reference Ranges. *Journal of Child Neurology*. 2010;25(10):1210-1215. doi:10.1177/0883073809359539

10. McGee S, Abernethy III WB, Simel DL. Is This Patient Hypovolemic? *JAMA*. 1999;281(11):1022-1029. doi:10.1001/jama.281.11.1022

11. Ritter S, Tani LY, Etheridge SP, Williams RV, Craig JE, Minich LL. What Is the Yield of Screening Echocardiography in Pediatric Syncope? *Pediatrics*. 2000;105(5):e58. doi:10.1542/peds.105.5.e58

12. Hegazy RA, Lotfy WN. The value of Holter monitoring in the assessment of Pediatric patients. *Indian Pacing Electrophysiol J*. 2007;7(4):204-214.

13. Steinberg LA, Knilans TK. Syncope in children: Diagnostic tests have a high cost and low yield. *The Journal of Pediatrics*. 2005;146(3):355-358. doi:https://doi.org/10.1016/j.jpeds.2004.10.039

14. Fitchet A, Stirling M, Burnett G, Goode GK, Garratt CJ, Fitzpatrick AP. Holter monitoring vs tilt testing in the investigation of suspected vasovagal syncope. *Pacing and clinical electrophysiology : PACE*. 2003;26(7 Pt 1):1523-1527. doi:10.1046/j.1460-9592.2003.t01-1-00221.x

15. Massin MM, Bourguignont A, Coremans C, Comté L, Lepage P, Gérard P. Syncope in pediatric patients presenting to an emergency department. *The Journal of Pediatrics*. 2004;145(2):223-228. doi:https://doi.org/10.1016/j.jpeds.2004.01.048

16. Redd C, Thomas C, Willis M, Amos M, Anderson J. Cost of Unnecessary Testing in the Evaluation of Pediatric Syncope. *Pediatric Cardiology*. 2017;38(6):1115-1122. doi:10.1007/s00246-017-1625-6

17. McLeod KA. Syncope in childhood. *Arch Dis Child*. 2003;88(4):350. doi:10.1136/adc.88.4.350

18. Anderson JB, Willis M, Lancaster H, Leonard K, Thomas C. The Evaluation and Management of Pediatric Syncope. *Pediatric Neurology*. 2016;55:6-13. doi:https://doi.org/10.1016/j.pediatrneurol.2015.10.018

19. Lewis DA, Dhala A. SYNCOPE IN THE PEDIATRIC PATIENT: The Cardiologist's Perspective. *Pediatric Clinics of North America*. 1999;46(2):205-219. doi:10.1016/S0031-3955(05)70113-9

Premature Atrial Contractions
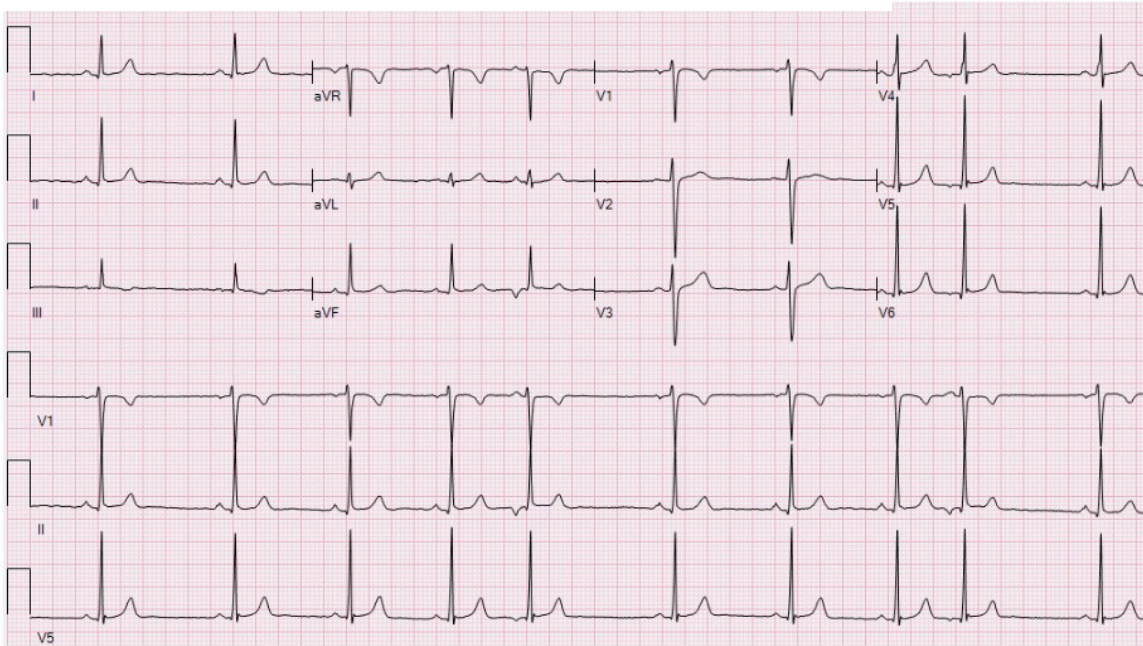
Premature Ventricular Contractions
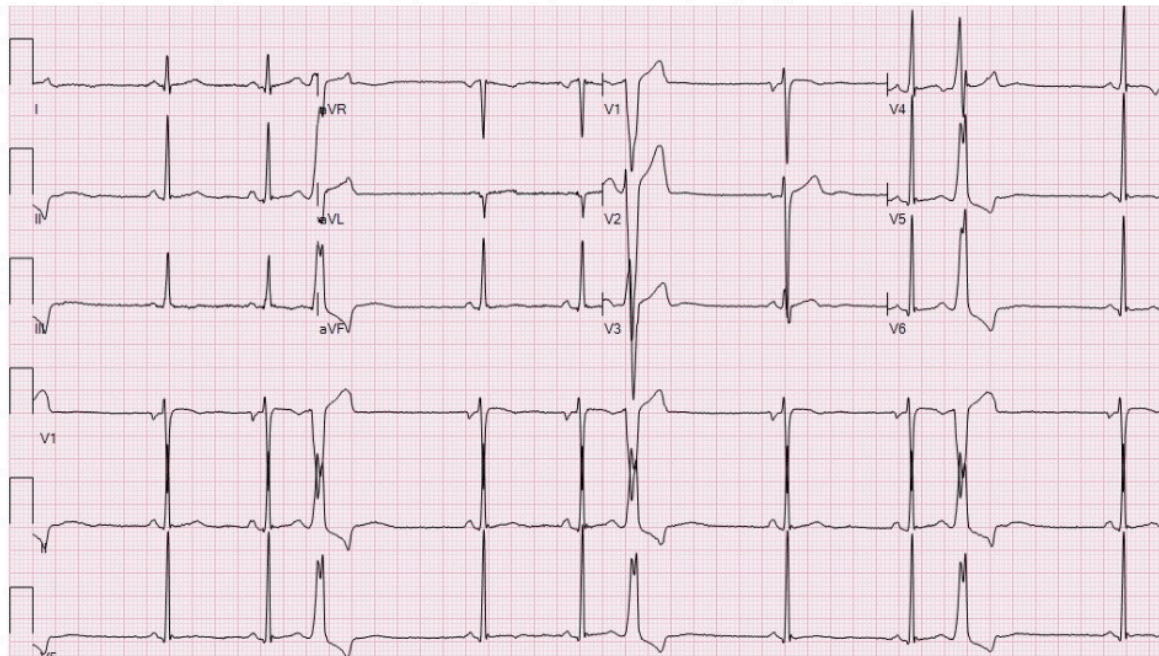
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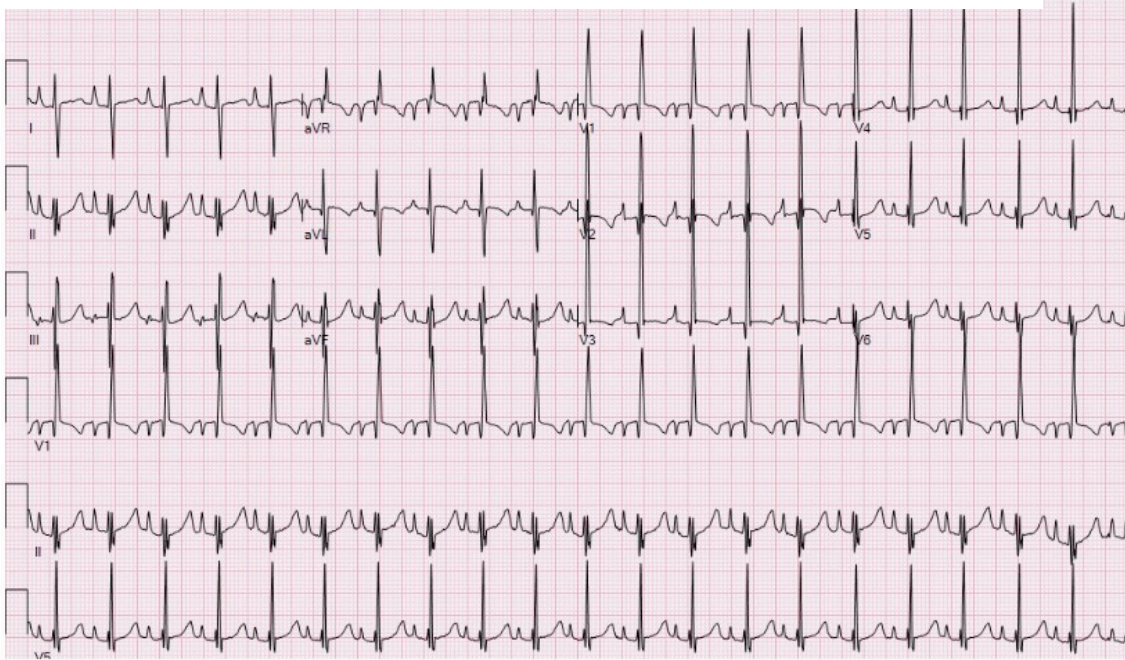
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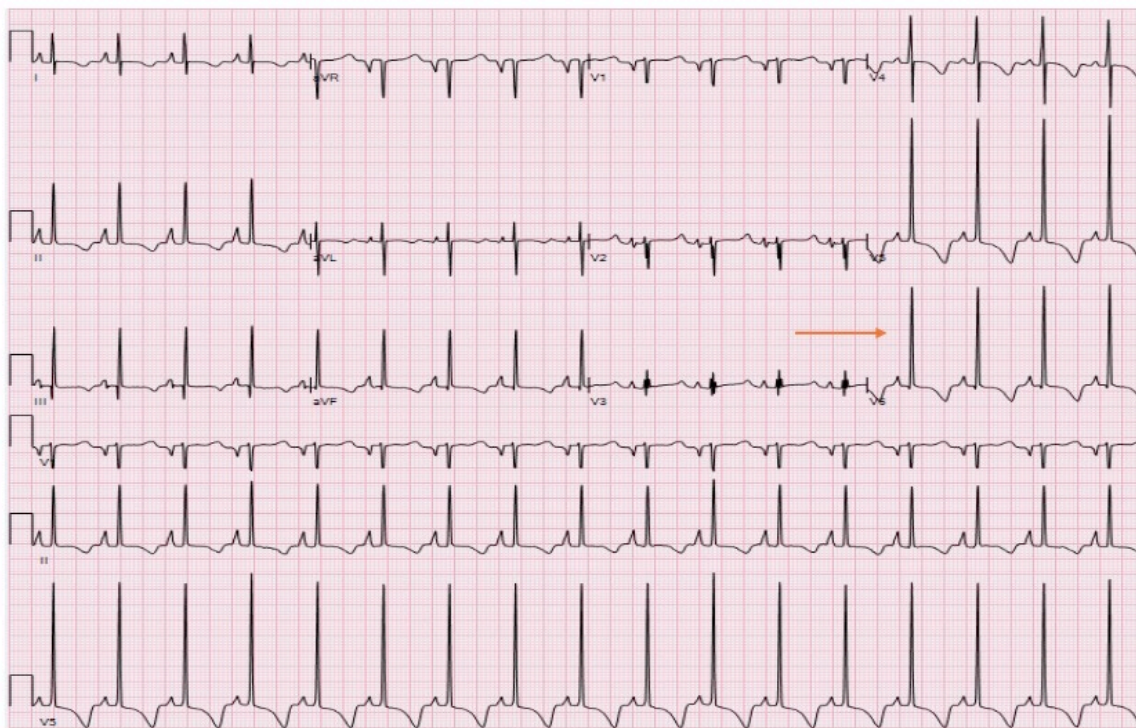
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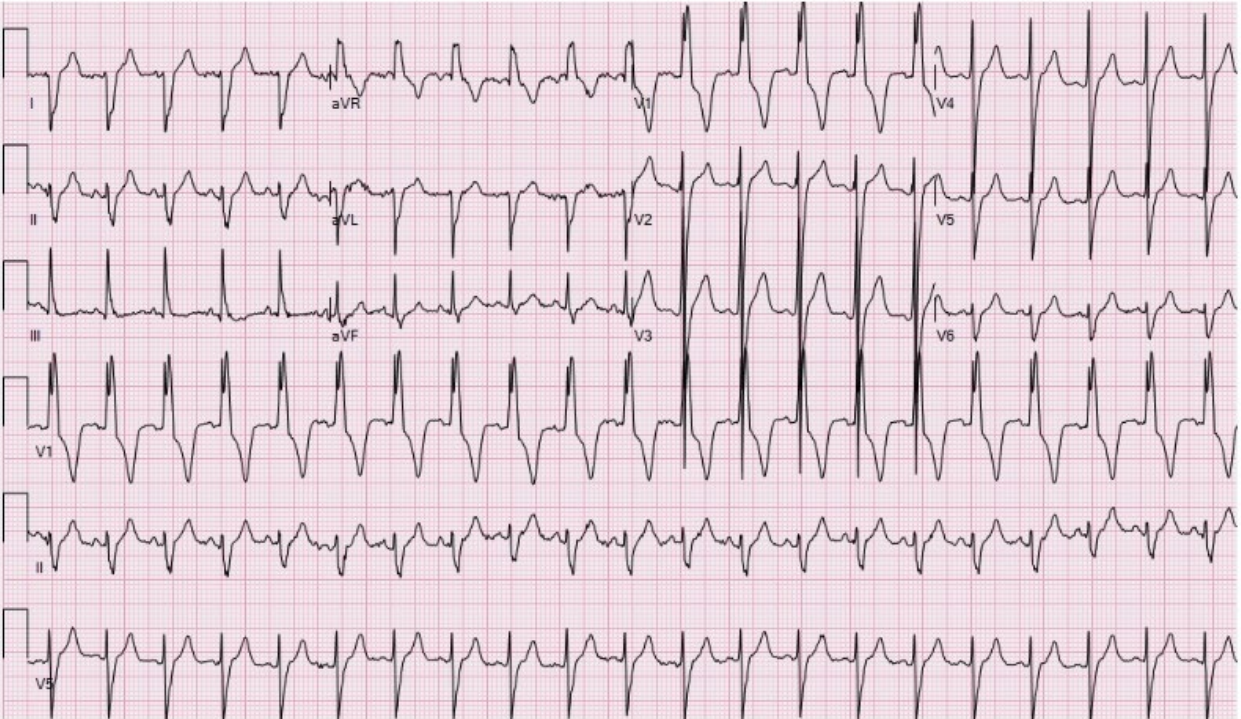
First Degree AV Block
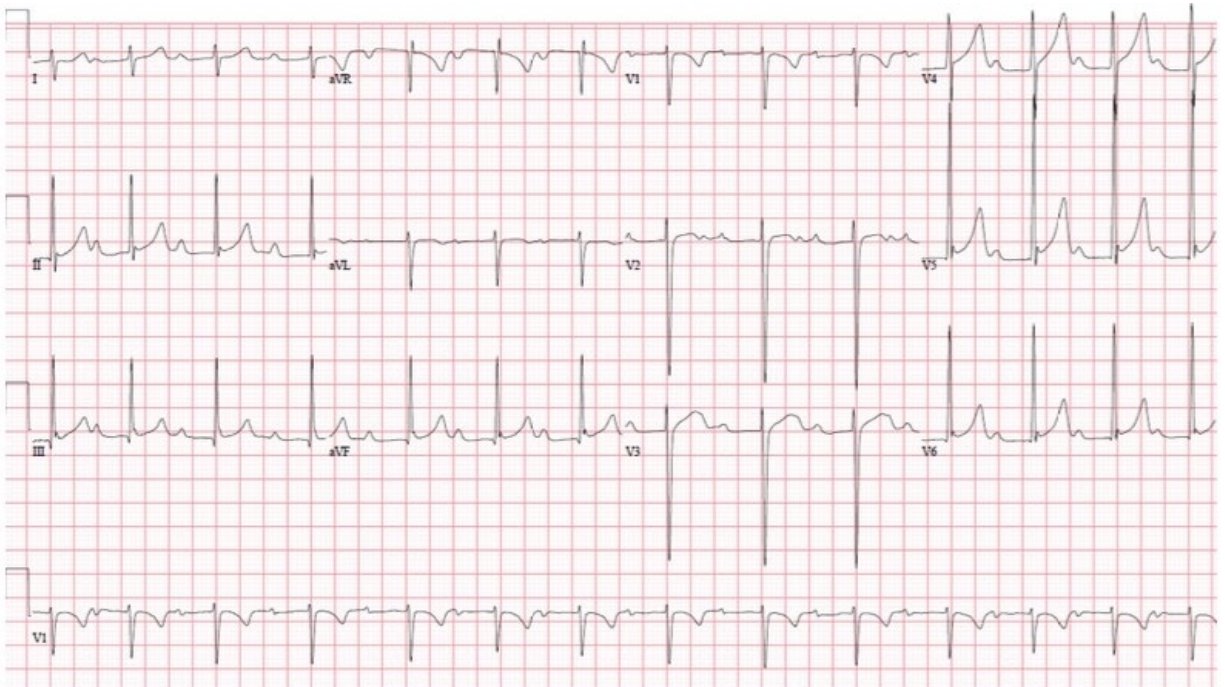
Second Degree AV Block ~ Mobitz Type 1
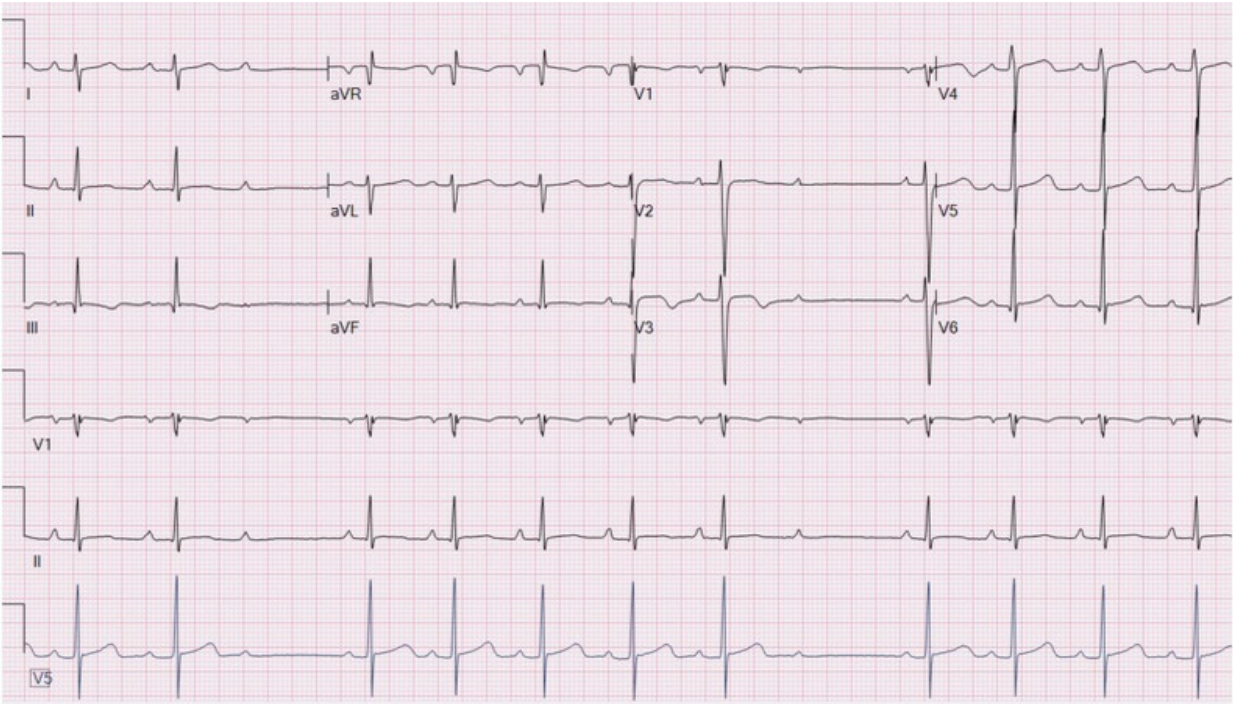
Second Degree AV Block ~ Mobitz Type 2
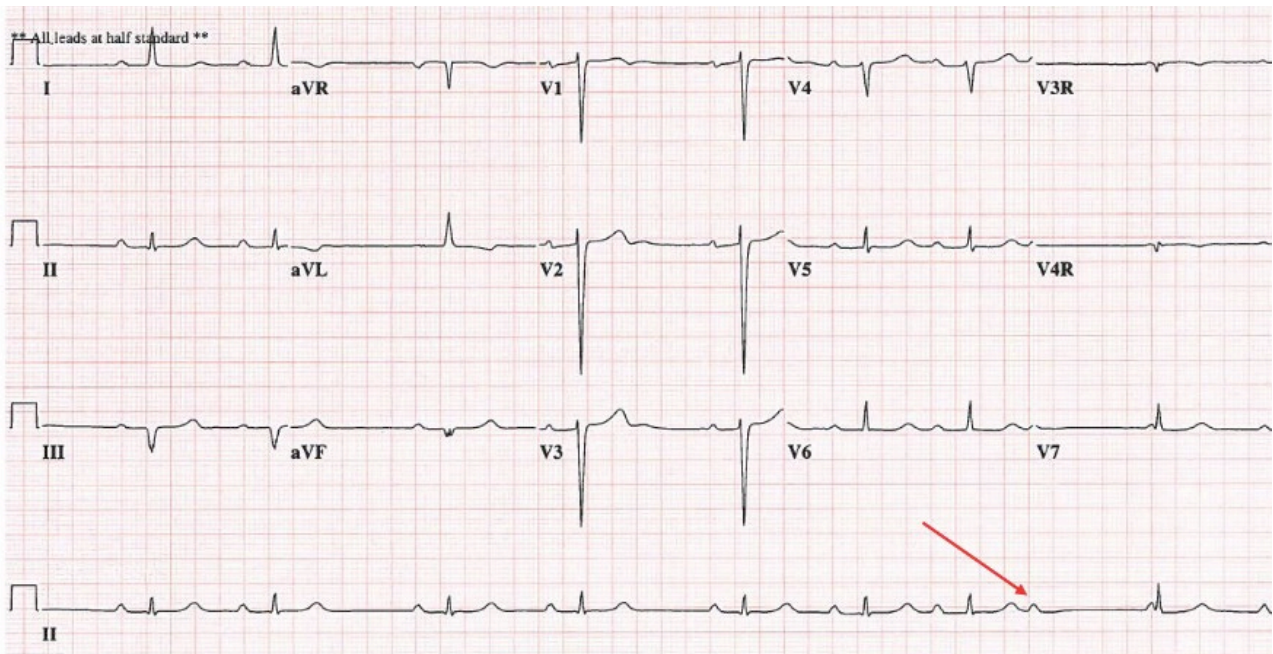
Third Degree (Complete) AV Block
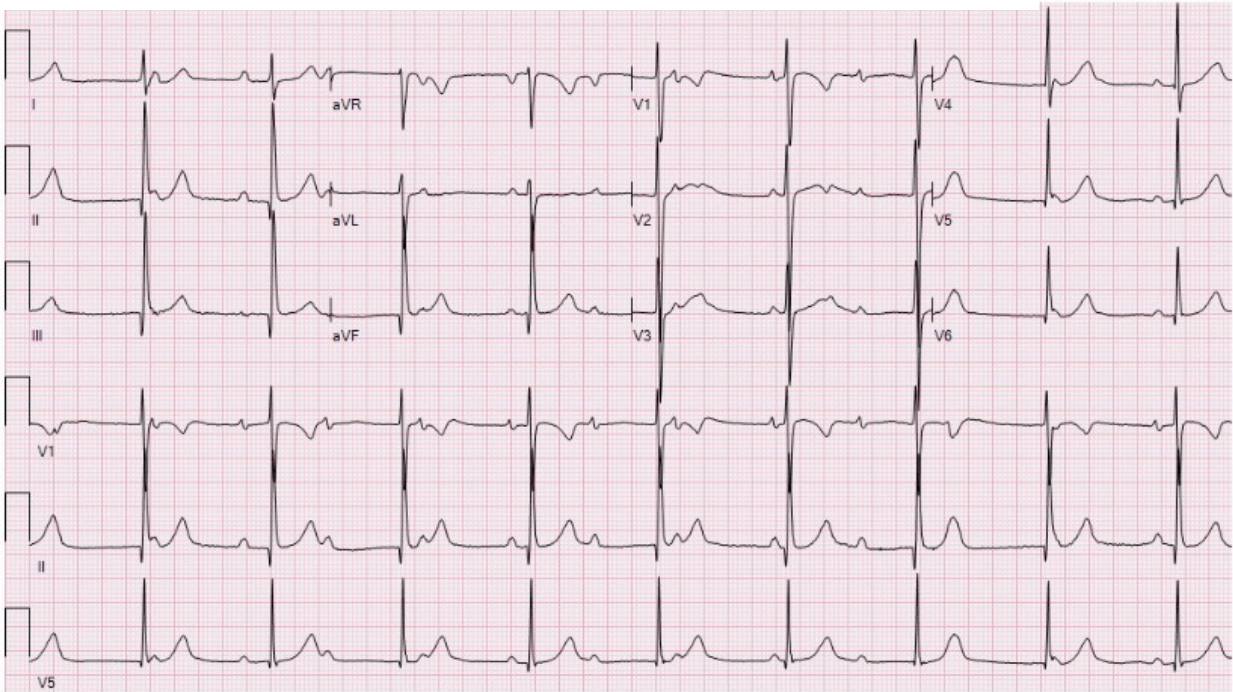
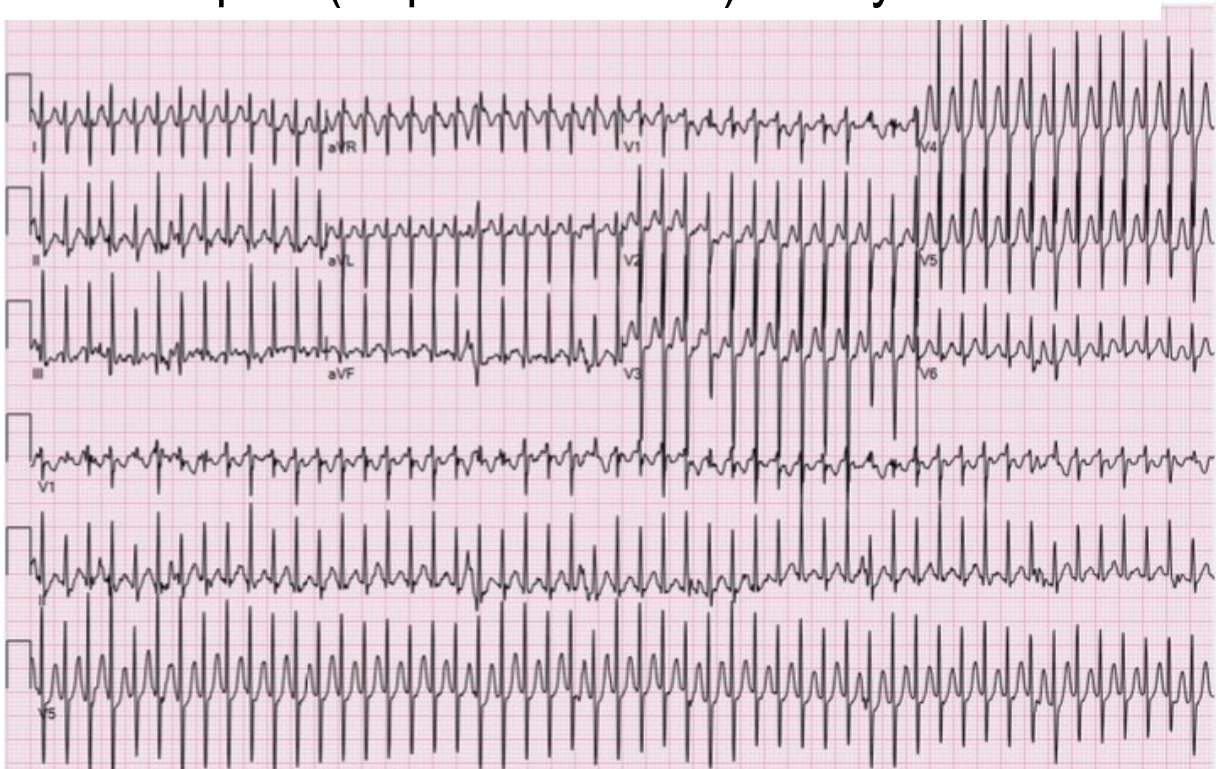## Ischemia –ST segment depression in inferolateral leads with reciprocal ST segment elevation
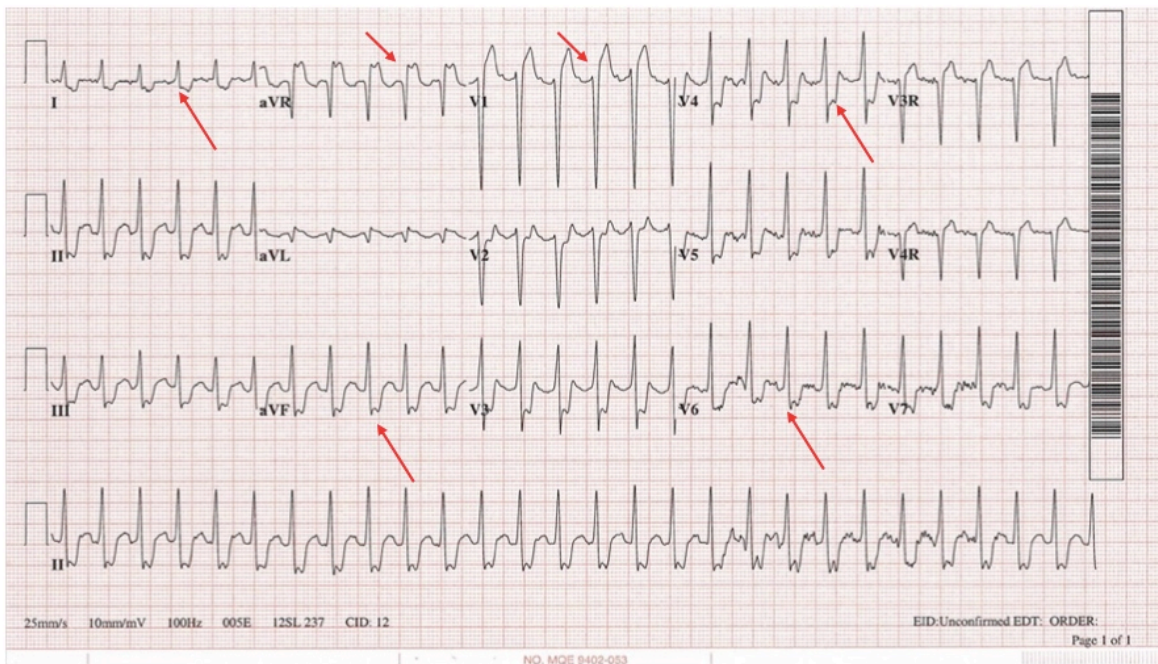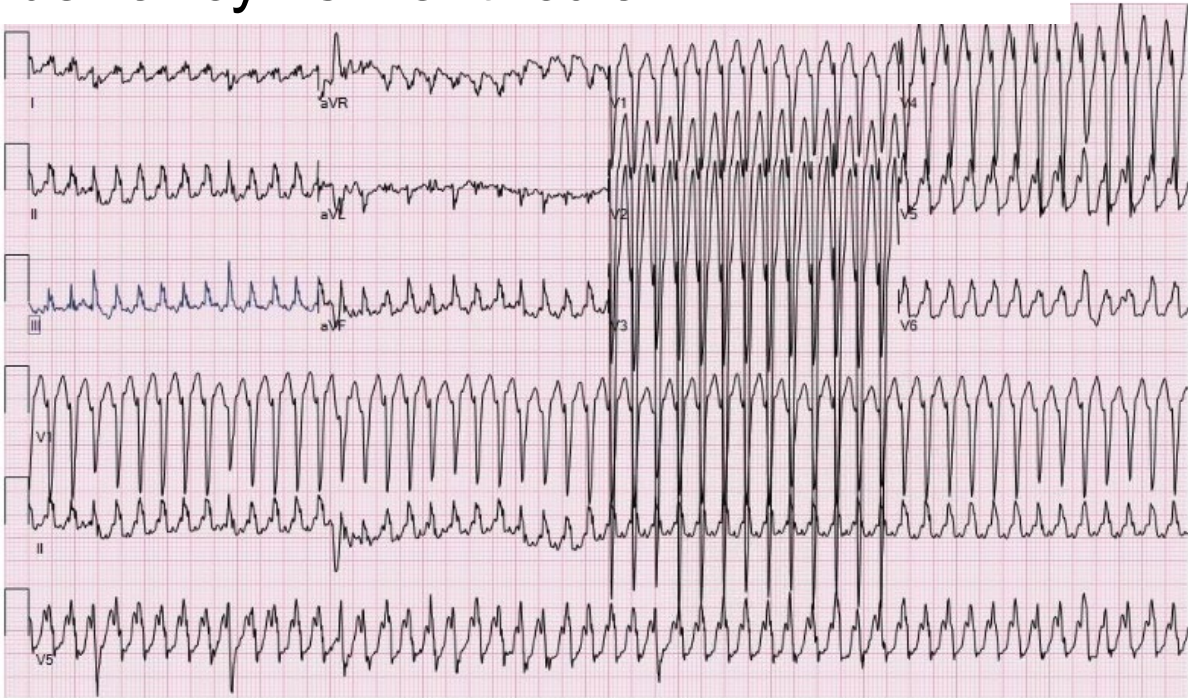
Wolfe Parkinson White Syndrome

- Delta wave noted with arrows
- 2 beats are not pre-excited for contrast
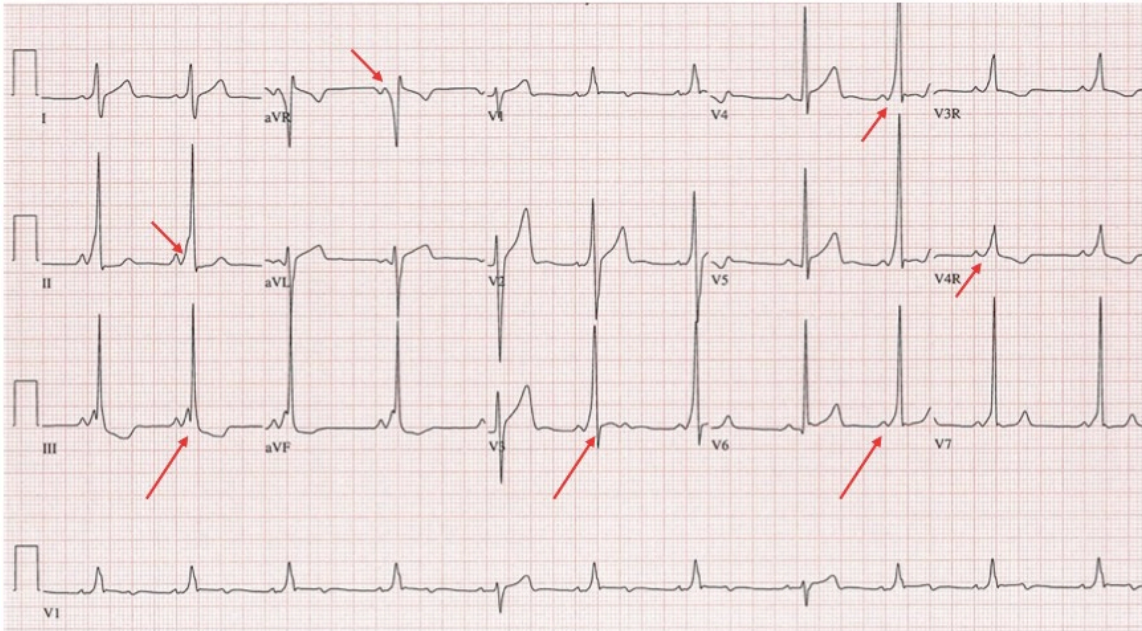
Narrow Complex (Supraventricular) Tachycardia

Wide Complex Tachycardia (SVT w/ aberrancy vs Ventricular

Brugada Syndrome
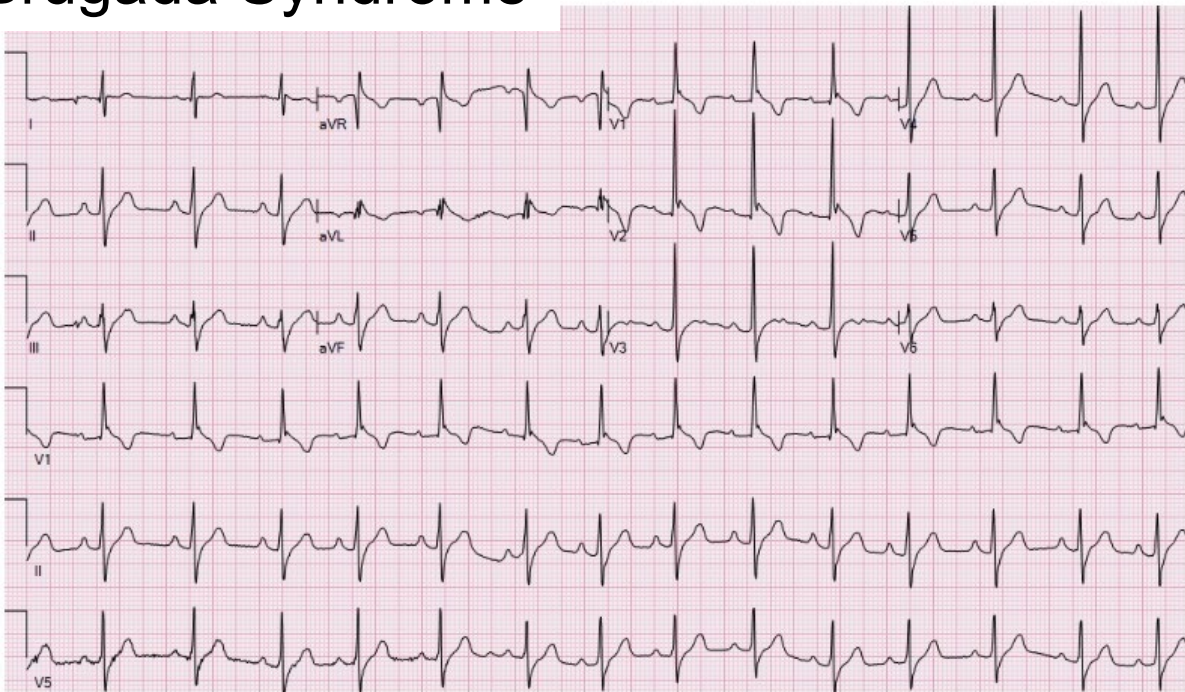
Prolonged QT Interval
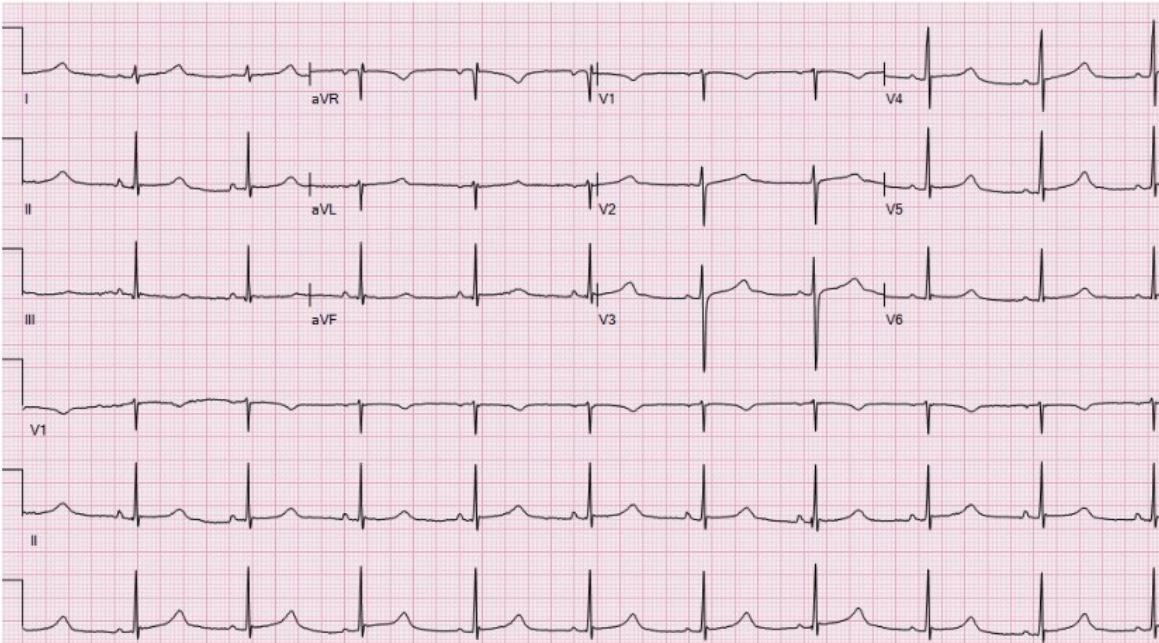
Arrhythmogenic Right Ventricular Dysplasia (ARVD)

- Epsilon Waves in V_1, V_2, V_3 (positive deflection at end of QRS complex, thin arrows)
- T wave inversions in V_1, V_2, V_3
- Prolonged S wave upstroke (thick arrow)

https://litfl.com/ecg-library/
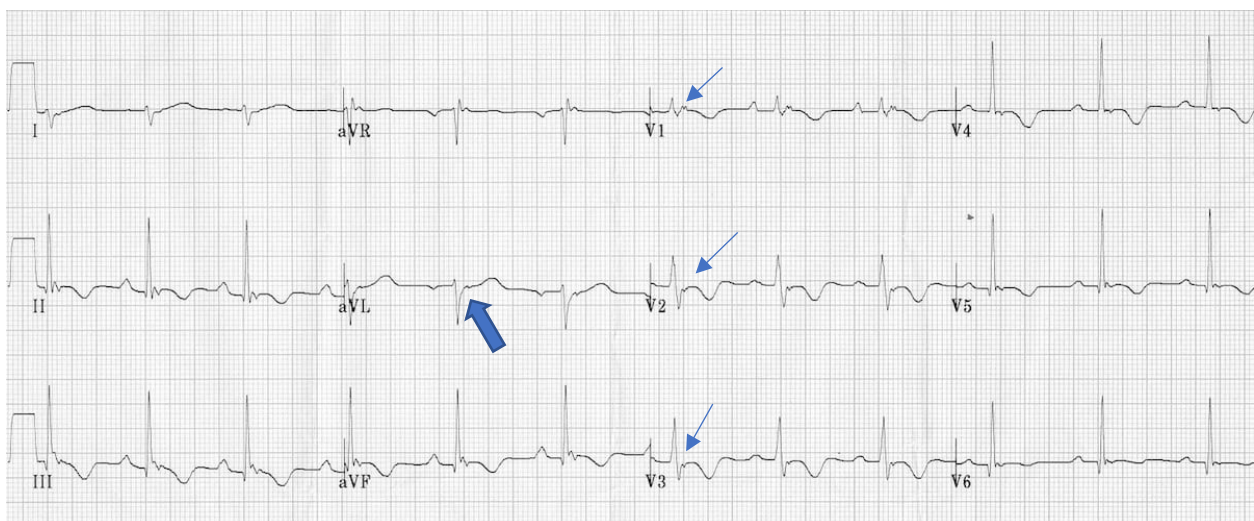
Outcome Measures:

- Length of Stay in EC
- Percentage of Labs Drawn
- Admission Rate

Documentation Recommendations:

- It is important to link the suspected etiology. The underlying diagnosis does not have to be confirmed to be linked; clinical suspicion is sufficient. For example, "Syncope secondary to suspected dehydration."
- If after evaluation there are multiple possible causes that cannot be excluded, you can still link those diagnosis. For example, "Syncope secondary to suspected dehydration versus hypokalemia."

Patient Class Recommendations:

- If a patient will be admitted for monitoring, please place them in observation if the etiology remains unknown and the patient is clinically stable.
- If the patient will be admitted to the ICU because of ongoing vital sign instability and/or the need for ongoing medical interventions to stabilize the patient, please order inpatient.

Disclaimer