



# NCC Pediatrics Continuity Clinic Curriculum: Syncope *Faculty Guide*

**Goals & Objectives:** *To obtain a thorough history of pre-syncopal/syncopal event(s) to understand the etiology, evaluation, and management. (Based on 2010 ABP content specs)*

- Plan appropriate evaluation & management of a pre-syncopal/syncopal episode
- Know when to refer to subspecialty (cardiology, neurology)
- Recognize the cardiac causes of syncope

## **Pre-Meeting Preparation:**

- “Syncope” (*PIR, 2000*)
- “Consult with the Specialist” (*PIR, 2003*)—\*Syncope section only\*
- Algorithm for Evaluation of Syncope; History Taking 101

## **Conference Agenda:**

- Review Syncope Quiz
- Complete Syncope Cases
- **Round-Table Discussion:** Discuss patient cases or personal experience with syncope.

**Post-Conference:** Board Review Q&A

## **Extra-Credit:**

- [AHA/ACCF Scientific Statement on the evaluation of syncope](#) (*Circulation 2006*)
- [“Management and therapy of vasovagal syncope: A review”](#) (*World J Cardiology 2010*)
- [“Postural orthostatic tachycardia syndrome”](#) (*Postgrad Medical J 2007*) (seen in algorithm)

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# Syncope

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## OBJECTIVES

After completing this article, readers should be able to:

1. Describe the steps involved in identifying the correct diagnosis of syncope.
2. Identify the only mandatory screening test in syncope.
3. Describe the ramifications of syncope with exercise.
4. Identify the most common diagnosis in syncope and why it is dangerous.
5. List the differential diagnosis and treatment of syncope.

## Definition

Syncope is a transient loss of consciousness and muscle tone. Loss of cerebral oxygenation and perfusion is the usual mechanism. It often is benign but may cause injury (15% of cases). Syncope can be caused by serious cardiac disease, which should be suspected whenever syncope occurs with exercise.

## Epidemiology

Up to 3% of emergency department visits by adults and 6% of hospital admissions are due to syncope. Among children, only 0.125% of emergency department visits are due to syncope. Nevertheless, 47% of college students report having fainted, and 15% of children suffer from syncope before the end of adolescence. The lay public realizes that healthy children who faint frequently are free of serious disease and may not require emergency medical attention. About 75% of children who faint have neurocardiac (vasovagal) syncope due to neurally mediated hypotension and bradycardia; most others have seizures, migraine, or cardiac disease.

## Pathogenesis

The causes of syncope can be categorized as cardiac, noncardiac, and neurocardiac (Table).

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## CARDIAC SYNCOPE

Cardiac syncope is due to outflow obstruction (aortic stenosis, hypertrophic cardiomyopathy), myocardial dysfunction (cardiomyopathy, carditis, ischemia), or arrhythmias (ventricular tachycardia, long Q-T syndromes, Wolff-Parkinson-White syndrome). Cardiac disease is suggested when syncope accompanies exercise. Cardiac syncope is potentially fatal and always deserves careful evaluation and treatment.

## NONCARDIAC SYNCOPE

This form of syncope includes many entities, some of which are distinguished easily by a careful history and are not true syncope. Seizures often manifest unusual eye or limb movements, may be prolonged, and—unlike syncope—usually are followed by postictal stupor. Seizures may result from cardiac or neurocardiac syncope if cerebral perfusion or oxygenation is sufficiently reduced, which is especially likely if the child is held upright during the syncopal episode. Epileptic seizures may cause cardiac disturbances, usually tachycardia. Many patients who have epilepsy have normal results on electroencephalography (EEG); diagnosis is made best by history.

Breathholding spells begin in infancy and resolve by school age. The history is stereotypical. The spells always begin with pain or anger, followed in order by a brief cry, holding of the breath (usually with the mouth open and a distressed expression), cyanosis or pallor (the latter if bradycardia occurs),

possibly a loss of consciousness, and finally perhaps a brief tonic or clonic seizure. If the history is not perfectly typical, epilepsy or cardiac syncope should be suspected and these diagnoses pursued. If the history is typical, iron deficiency anemia, which is a possible cause, should be ruled out, and the parents should be reassured that breathholding is benign and will resolve. Anti-epileptic medications should not be prescribed to breathholders unless the resulting seizure is lengthy. Seizures related to breathholding usually are brief and hypoxic and do not require treatment with antiepileptic medication. Breathholding is not a manipulative behavior. Its only sequel is a later propensity to neurocardiac syncope in 17% of patients. The best treatment is to keep the child horizontal and wait.

Migraine may cause syncope because of the pain or occasionally directly (brainstem migraine). The diagnosis can be made based on a history of associated severe headache with nausea, vomiting, photophobia, and relief by sleep. Family history is positive for migraine in 75% of cases.

Orthostatic hypotension is defined as a 20-mm Hg drop in systolic blood pressure upon assuming an upright posture. Patients should be monitored for at least 2 minutes upright with serial blood pressure measurements. Pregnancy should be considered as a cause in women of childbearing age. Dehydration, prolonged bedrest, drugs, and neuropathies may be predisposing factors.

Hyperventilation may produce sufficient cerebral vasoconstriction to cause syncope. An associated Valsalva maneuver or chest compression can accentuate the cerebral hypoperfusion caused by hyperventilation.

Situational syncope can be caused by cough, micturition, defecation, neck stretching, hair grooming, venipuncture, or even swallowing in certain individuals. The diagnosis is based on the history.

Narcolepsy may mimic syncope. Those who have narcolepsy go to

**TABLE 1. Etiologies of Syncope**

**Cardiac Syncope**

- Outflow Obstruction
  - Valvular aortic stenosis
  - Pulmonary hypertension
  - Hypertrophic cardiomyopathy
  - Eisenmenger syndrome
- Myocardial Dysfunction
  - Dilated cardiomyopathy
  - Myocarditis
  - Neuromuscular disease (eg, Duchenne dystrophy)
  - Kawasaki disease
  - Anomalous coronary artery
- Arrhythmias
  - Long Q-T syndromes
  - Ventricular tachycardia
  - Arrhythmogenic right ventricular dysplasia
  - Supraventricular tachycardia (Wolff-Parkinson-White)
  - Sinus node dysfunction
  - Atrioventricular block

**Appropriate Evaluations:** Electrocardiography, cardiac echocardiography, chest radiography, creatine phosphokinase

**Noncardiac Syncope**

- Seizures
- Migraine
- Orthostatic hypotension
- Narcolepsy/cataplexy
- Familial dysautonomia
- Gastroesophageal reflux
- Spinal cord disease (autonomic instability)
- Metabolic disease (diabetes, hypoglycemia, endocrine)
- Situational (cough, micturition, neck stretching, hair grooming, venipuncture)
- Breathholding spells
- Toxins/drugs
- Hyperventilation
- Hysteria
- Fever (febrile delirium)
- Carotid sinus hypersensitivity

**Appropriate Evaluations:** Electroencephalography, blood glucose, toxic screen

**Neurocardiac Syncope**

**Appropriate Evaluation:** Electrocardiography

sleep abruptly. Cataplexy (the abrupt intrusion of rapid eye movement sleep into waking) may be mistaken for syncope if a history of excessive daytime sleepiness is not sought. Cataplexy may produce loss of consciousness in response to emotional reactions such as laughter or anger. Narcolepsy is diagnosed by the sleep latency test. Narcolepsy is common in children, a fact that was not appreciated until recently.

Metabolic causes of syncope are

rare, but hypoglycemia and electrolyte abnormalities should be considered because they could have serious consequences.

Hysterical syncope may be difficult to diagnose, but it should be suspected when the episode is prolonged, there is no change in vital signs or appearance, it does not raise concern in the patient, or the patient's recall or responsiveness during the event suggests that consciousness has been maintained.

**NEUROCARDIAC SYNCOPE (VASOVAGAL)**

This is the most common variety of syncope, and it can be diagnosed with a tilt table test, although this test is not perfectly sensitive or specific. Approximately 25% of cases are precipitated by acute illness and anemia or by such noxious stimuli as pain, fear, exhaustion, hunger, prolonged standing, or crowded/poorly ventilated rooms.

There usually are the following prodromal symptoms: nausea/vomiting, pallor, lightheadedness/vertigo, visual disturbances, sweating, and shortness of breath. After syncope, the patient may be fatigued, lightheaded, anxious, or nauseous or may complain of headache. The typical mental alertness following the episode is helpful in distinguishing syncope from seizures.

The pathophysiology of neurocardiac syncope is relatively well understood. Peripheral venous pooling leads to decreased ventricular filling and increased circulating catecholamines. The ventricle responds with vigorous contractions that stimulate mechanoreceptors (like hypertension) and produces a paradoxical withdrawal of sympathetic activity that causes hypotension, bradycardia, or both.

**Clinical Aspects**

The history is the key to diagnosis of syncope, and it must be obtained carefully both from the patient and from an eyewitness because the patient will be unreliable for observations during unconsciousness. Several points demand specific questioning:

- The *situation and antecedents* of the episode, which may identify precipitants (exercise suggests cardiac syncope)
- The *onset* of the episode, which may include epileptic activity that characterizes the event as a seizure (lateral eye movements, sensory hallucinations, focal or generalized motor activity)
- The *duration* of the episode (time expands with excitement, and comparison of the event with a common timed activity such as TV commercials will help to pro-

vide a clear picture of the duration)

- *Loss of consciousness*, which is presumed from unresponsiveness to the environment (voice, pain) and from amnesia, injury, or incontinence
- *A postictal state* of confusion, which suggests a seizure
- *Palpitations*, which suggest cardiac disease

The past history may reveal associated disease predisposing to syncope. Because many causes of syncope are familial, it is important to inquire carefully about a family history of syncope, sudden or early death, epilepsy and neurologic disease, heart disease, and deafness (Q-T syndromes may have familial deafness).

A general physical and neurologic examination must include measurement of vital signs and a cardiac evaluation. Approximately 90% of orthostatic hypotension occurs within 2 minutes of standing upright; such a finding provides an important clue to the diagnosis.

Electrocardiography is recommended for every case of unexplained syncope. Life-threatening cardiac disease is a first concern.

Other laboratory tests should be ordered according to findings from the history and physical examination. An EEG, video-EEG monitoring (if episodes are frequent), neuroimaging, or neurologic referral may help with suspected seizures. A multiple sleep latency test is used to diagnose narcolepsy. Determination of blood glucose or electrolyte concentrations or endocrinologic studies may be useful in selected cases. If cardiac disease is suspected, Holter or loop monitoring, chest radiography, echocardiography, exercise stress testing, or even invasive electrophysiologic testing may be pursued.

The tilt table test has made the diagnosis of neurocardiac syncope a positive one rather than simply one of exclusion. Protocols vary, but the patient is tilted upright for a time sufficient to reproduce symptoms and changes in cardiovascular function (hypotension or bradycardia). The tilt table test is not always specific or sensitive, and intravenous infusions of isoproterenol and other drugs may be confusing noxious stimuli. Nevertheless, this has become the gold standard for diagnosing neurocardiac syncope.

### Management

Treatment of syncope is directed at the specific causative entity. Cardiac disease may require antiarrhythmics or surgery. Seizures may require anticonvulsants, with the choice depending on the exact type of seizure. Breathholding spells, which can be upsetting, necessitate reassurance of parents. A variety of acute and prophylactic medications are available for migraine. Hysteria in childhood merits psychiatric evaluation and the consideration of abuse. The sleepiness of narcolepsy is treated with stimulants (eg, methylphenidate hydrochloride 20 mg every morning) and brief naps; cataplexy is treated with tricyclic antidepressants (eg, imipramine 25 to 75 mg q hs). For situational syncope, the inciting stimulus should be avoided.

Neurocardiac syncope often can be managed with simple suggestions, such as lying down before losing consciousness, wearing elastic hose to prevent venous pooling in the legs, increasing salt and water intake, eating regularly, avoiding noxious stimuli that precipitate syncope, and intermittently contracting

leg muscles when standing to increase venous return. Avoiding alcohol, beta-blockers, tricyclics, and isoproterenol may lessen the likelihood of neurocardiac syncope. Standard drug therapy includes mineralocorticoids (hydroflurocortisone 0.1 mg bid), atenolol (1 to 2 mg/kg per day), and pseudoephedrine (4 mg/kg per day qid). In truly refractory cases, cardiac pacing may be considered.

### Prognosis

The outlook for syncope depends on the specific diagnosis. Neurocardiac syncope recurs in two thirds of cases, and syncope of other causes has a 90% or greater recurrence rate. Treatment often is effective once the condition is diagnosed. The outlook for many childhood epilepsies is benign. Migraine and narcolepsy usually are controlled by medication. Death can occur unexpectedly in cardiac syncope, and vigorous diagnostic evaluations are warranted whenever palpitations or exercise are associated with syncope.

### SUGGESTED READING

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## CONSULTATION WITH THE SPECIALIST

# Palpitations, Syncope, and Sudden Cardiac Death in Children: Who's at Risk?

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**Objectives** After completing this article, readers should be able to:

1. Clarify the definition and terminology of palpitations, syncope, and sudden cardiac death (SCD).
2. Differentiate the relatively benign forms of palpitations and syncope from those that are associated with an increased risk of SCD.
3. Characterize the evaluation for palpitations of cardiac origin.
4. Describe the management of neurally mediated syncope.
5. Discuss the structural heart diseases predisposing to sudden death.
6. Define the guidelines for participation in competitive athletics for children who have common cardiac problems.

## Case Reports

### Patient 1

*A 12-year-old female, who has a history of palpitations and exertional syncope, fainted while swimming. Her brother was near her and pulled her out of the water. The girl immediately regained consciousness and was found to have normal perfusion. The family history was significant for unexplained sudden death of the girl's older brother. Findings on physical examination of the patient were normal. Results of chest radiography were normal, but electrocardiography (ECG) revealed a sinus rhythm, with a corrected Q-T interval of 0.52 seconds.*

### Patient 2

*A 16-year-old cross country runner had experienced several episodes of syncope. Each time she was caught prior to*

*falling and was able to resume activities after some rest. Findings on her past medical history and family history were not significant. Physical examination results were normal. Chest radiography and ECG findings were normal, but a tilt table test showed onset of near-syncope accompanied by a significant decrease in the heart rate and blood pressure. The patient was started on a regimen of increased fluid intake and salt tablets prior to exercise. She has had no recurrences of syncope.*

## Overview

Palpitations and syncope are frequent presenting complaints to the pediatrician. Although mostly benign, these worrisome symptoms may be the prodrome of significant cardiac events. The devastating result could be brain damage or SCD. It is important to differentiate the relatively benign forms of palpitations and syncope from those that are associated with an increased risk of SCD.

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## Palpitations

Palpitations are a subjective sensation of unduly strong, rapid, or irregular heart beats that may be related to cardiac arrhythmias. They also can be due to physiologic causes such as sinus tachycardia associated with anxiety or exercise or pathologic causes such as atrial fibrillation associated with hyperthyroidism and ventricular tachycardia in the long Q-T syndrome. Patients who have had palpitations should be evaluated carefully to rule out any significant arrhythmia before being allowed to participate in any competitive sports.

A complete history is necessary to understand the nature of the palpitations. Inquiry should be made into the timing and circumstances that led to the palpitations; other associations with the onset of palpitations (eg, behavioral change); and a family history of syncope, palpitations, or SCD. A physical examination can rule out structural heart disease and systemic disorders. Although laboratory test results usually are normal, chest radiography is advised to rule out congenital heart disease and lung disease. ECG is needed to evaluate P waves, the presence of premature atrial beats or premature ventricular beats, and the Q-T interval. Commonly, these tests indicate a benign nature of the palpitations, leading to the great relief of those involved.

Asymptomatic patients who have premature atrial contractions or premature ventricular contractions, even if frequent, or short runs of unifocal ventricular tachycardia need not be referred to a cardiologist. Only reassurance is needed for the patient and family. If a cardiac origin of the palpitations is suspected, an extended evaluation usually is undertaken in collaboration with a pediatric cardiologist (Table 1).

## Table 1. Evaluation for Palpitations of Cardiac Origin

### Initial Evaluation

- History and physical examination
- Chest radiography
- Electrocardiography

### Advanced Evaluation (in association with a pediatric cardiologist)

- Echocardiography
- Holter monitoring
- Exercise stress test
- Electrophysiologic testing

## Syncope

Syncope is a temporary loss of consciousness due to generalized cerebral ischemia that usually is followed by rapid and complete recovery. In rare instances, anoxic seizures may result. Syncope may be preceded by palpitations, lightheadedness, dizziness, weakness, pallor, nausea, cold sweat, blurred vision, or hearing loss. Prompt relief from all symptoms usually occurs after lying down. Syncope may result from impaired response of the autonomic nervous system or from cardiac structural defects, especially those obstructing blood flow or from cardiac arrhythmias. The relatively uncommon long Q-T syndrome is an especially worrisome cause of syncope. Noncardiac mechanisms, such as metabolic, neurologic, and psychologic disorders, also may cause syncope.

Neurally mediated syncope, also referred to as vasovagal or neurocardiogenic syncope, is the most common form of syncope in children. It often is associated with orthostatic intolerance. It also has been reported that chronic fatigue syndrome in adolescents may be related to orthostatic intolerance. In such cases, the mechanism of syncope is reflex-mediated and originates from a decreased systemic venous return that leads to decreased left ventricular

end diastolic volume. Increased mechanical contractility results in stimulation of cardiac vagal fibers and ultimately a paradoxical response of marked bradycardia, vasodilation, and hypotension. This chain of events is referred to as the Bezold-Jarisch reflex. Reflex syncope also may result from a hypersensitive autonomic response caused by different afferent input, such as micturition, swallowing, deglutition, coughing, sneezing, or defecation. Neurally mediated syncope may present in one of the three clinically recognized forms: cardioinhibitory (low blood pressure, bradycardia/asystole), vasodepressor (low blood pressure, no bradycardia), or mixed (low blood pressure, bradycardia).

Physical exhaustion, prolonged recumbency, conditions predisposing to peripheral vasodilation (exercise, hot weather), and pregnancy enhance the chance of having neurally mediated syncope, as do a variety of noxious stimuli, such as blood drawing or emotional stress. Affected patients have a decrease in blood pressure and heart rate with tilt table testing similar to that observed in vasodepressor syncope. Neurally mediated syncope rarely is associated with sudden death.

Because certain causes of syncope may be related to life-threatening

conditions, a detailed evaluation should be undertaken whenever patients present with syncope. Those who have a family history of syncope, sudden death, myocardial disease, or arrhythmias and those who have exercise-associated syncope are at particularly high risk of SCD. Electrocardiography should be a part of the initial evaluation for all patients who present with syncope. A cardiology consultation is indicated when the cause of recurrent syncope cannot be identified, cardiac or arrhythmogenic syncope is identified, or a pacemaker is indicated for severe and recurrent syncope unresponsive to medical management. Further evaluation by the pediatric cardiologist often includes event monitoring, exercise stress testing, and tilt table testing.

Tilt table testing may be indicated under certain situations in which the cause of syncope is not clear. The American College of Cardiology published guidelines in 1996 on the indications for tilt table testing. It is generally agreed that tilt table testing should be reserved for patients who have recurrent syncope or for high-risk patients after a single syncopal episode. However, adolescents who describe prodromes of lightheadedness, nausea, and sweating before syncope most likely have neurally mediated syncope. These prodromes strongly suggest that tilt table testing is not necessary. The tilt table test consists of placing the patient in a head-up tilted position after a short period of lying prone. The tilt angle is between 60 and 80 degrees on a table that has a footplate and safety straps. The appropriate end point is induction of syncope or presyncope associated with intolerable hypotension and resulting in an inability to maintain postural tone.

Fluid therapy (Table 2) often is effective for neurally mediated syn-

## Table 2. Management of Neurally Mediated Syncope

### Evaluation

History and physical examination: negative for other causes of syncope  
Electrocardiography: Normal, including QTc

### Therapy

Trial of fluid therapy: 1 8-oz glass of any type of fluid on arising, at meals, and between meals (approximately q2 to 4 h) until the evening meal. Two 8-oz glasses of fluid such as "Gatorade™" prior to athletic participation, including practice.

### Outcome

Approximately 90% will respond and need no referral

### Referral

Refer nonresponders to a pediatric cardiologist

cope, especially of the vasodepressor type, and should be the primary mode of intervention in such patients. Other therapies that have been described include beta-blockers, volume expansion (with salt tablets or fludrocortisone), pseudoephedrine, disopyramide, and newer medications such as midodrine. These can be used individually or in combination. For patients in whom the neurally mediated syncope is frequent or sufficiently severe to cause anoxic seizures and unresponsive to conventional medical treatment, implantation of a pacemaker may be warranted.

## Sudden Cardiac Death

SCD is any natural death that occurs due to cardiac causes within minutes to 24 hours after the onset of symptoms. Cardiac deaths have been classified as arrhythmic deaths or due to circulatory collapse. SCD in children is relatively rare. There are about 600 SCDs in children annually compared with 7,000 to 10,000 deaths from sudden infant death syndrome and 300,000 to 400,000 SCD deaths in adults. The prevalence of SCD increases with age, accounting for 19% of sudden deaths in children between

1 and 13 years of age and 30% between 14 and 21 years.

The risk of SCD may be slightly higher in athletes involved in strenuous training. In a population of high school athletes in Minnesota, Maron and associates reported the risk of SCD to be approximately 1 in 200,000 per year and higher in male athletes. This low occurrence of SCD in competitive sports makes structuring of cost-effective broad-based participation screening guidelines for high school and college athletes difficult. In addition, the range of causes of sudden death on the athletic field may include causes for which it is impossible to screen. In rare instances, an athlete who has a structurally normal heart and no underlying pathology may suffer blunt trauma to the chest that causes a ventricular dysrhythmia and SCD. Labeled as commutio cordis, it is believed to be induced by the abrupt, blunt blow to an electrically vulnerable phase of electrical excitability within the myocardium. Athletes who have syncope or near-syncope warrant a more thorough evaluation to determine the cause. Current recommendations for disqualification from competitive athletics are based

**Table 3. Guidelines for Participation in Competitive Athletics for Common Cardiac Problems**

Type of Cardiac Defect	Athletic Limitation
<b>Tetralogy of Fallot</b>	
Excellent result	No restrictions
Suboptimal result	Low-intensity competitive sports
<b>Transposition of the Great Arteries</b>	
Postoperative Mustard or Senning	Low and moderate static, low-dynamic sports
Arterial switch operation	No restrictions
<b>Postoperative Fontan Operation</b>	Low-intensity competitive sports
<b>Hypertrophic Cardiomyopathy</b>	Low-intensity competitive sports
<b>Myocarditis</b>	No competitive sports for 6 mo
<b>Congenital Coronary Anomalies</b>	No competitive sports until repaired
<b>Kawasaki disease</b>	
No coronary involvement	No restrictions
Minor residual abnormalities	Low static, low/moderate dynamic sports
Persistent coronary artery aneurysm or stenosis	Low-intensity competitive sports
Intermittent myocardial ischemia	No competitive sports
<b>Aortic Stenosis</b>	
Mild	No restrictions
Moderate	Low-intensity competitive sports
Severe	No competitive sports
<b>Wolff-Parkinson-White Syndrome</b>	No restrictions
<b>Long QT Syndrome</b>	No competitive sports
<b>Pulmonary Hypertension</b>	
Mild (<40 mm Hg peak pressure)	No restrictions
Moderate/severe (>40 mm Hg peak pressure)	No competitive sports
<b>Marfan syndrome</b>	
No aortic root dilation or mitral regurgitation	Low and moderate static, low-dynamic sports
Aortic root dilation	Low-intensity competitive sports

on the guidelines from the 26th Bethesda Conference (Table 3).

### Structural Heart Diseases Predisposing to Sudden Death

The incidence of SCD in patients who have congenital heart disease is particularly high after repair of certain lesions. Following tetralogy of Fallot repair, patients have a 2% to 5% incidence of SCD, with ventricular arrhythmias being the most likely cause. Other surgically repaired lesions associated with a high incidence of arrhythmias and SCD include the Mustard or Senning

operation for transposition of the great arteries and the Fontan operation for a single ventricle physiology. The latter lesions include hypoplastic left ventricles in which prior Norwood operations have been undertaken. Patients in these categories usually are limited to low-intensity competitive sports such as golf, billiards, and bowling, although certain individuals who have excellent surgical results and normal exercise test results may be allowed to participate in more dynamic sports. Atrial arrhythmias tend to dominate in these lesions. About 40% of SCDs in pediatric patients occur in those who have

unoperated congenital or acquired heart disease. Several subgroups of such heart disease have been identified.

### Cardiomyopathies and Myocarditis

Restrictive, hypertrophic, or dilated cardiomyopathies may predispose the patient to SCD. Of these, hypertrophic cardiomyopathy is the most common cause of SCD in adolescents in the United States. Patients may present with symptoms of chest pain, syncope, and palpitations associated with exercise or sudden death during exercise. Factors associated with an increased risk of SCD in patients who have hypertrophic cardiomyopathy include a strong family history of sudden death, clinical symptoms, a young age, presence of ventricular arrhythmia, and a thickened intraventricular septum. ECG may show left ventricular hypertrophy, ST-T wave changes, and deep and wide Q waves in the left precordial leads. Echocardiography is the principal diagnostic modality to judge the severity and progression of cardiomyopathy (Fig. 1). Even though hypertrophic cardiomyopathy is a common cause of SCD in athletes in the United States, it is a rare cause of SCD in athletes in Italy. This may be related to the much more aggressive screening, including echocardiography, and disqualification from sports by Italian law of patients who have hypertrophic cardiomyopathy.

Arrhythmogenic right ventricular dysplasia and right ventricular cardiomyopathy have been reported as leading causes of SCD in athletes in studies performed in Italy, but they apparently are less common in other geographic regions, including the United States. These lesions are associated with a high frequency of cardiovascular symptoms and complica-



tions. Patients may present with ventricular arrhythmias (45%), congestive heart failure (25%), heart murmur (10%), complete heart block (5%), or sudden death (5%). First-degree relatives of 30% of the patients are affected. ECG usually shows a left bundle-branch pattern. Because limited data are available regarding the risks of athletic participation for such patients, they are best advised to refrain from participation in any competitive sports.

SCD has been reported in 14% to 42% of patients who had acute and chronic myocarditis and died either at rest or during exercise. These patients may present with a wide range of symptoms from subtle findings such as persistently increased heart rate or low-grade ventricular ectopy to severe congestive heart failure with cardiomegaly and poor exercise tolerance (Fig. 2). Viruses have been identified as the most common causes of acute or chronic myocarditis.

### Coronary Artery Disease

Patients who have congenital or acquired coronary artery disease may present with SCD, with the disease being diagnosed at autopsy. The most common coronary artery anomaly leading to SCD is the left main coronary artery arising from the right sinus of valsalva. It may be difficult to recognize these patients prospectively because they usually are asymptomatic until the initiating event that is related to exercise. Routine 12-lead ECG and exercise stress testing are not much help in the diagnosis. A history of syncope, palpitations, or chest pain related to exercise is associated with an increased risk of anomalous origin of the coronary artery and warrants an echocardiogram to define the coronary arteries. It is reasoned that compression of the left coronary artery, which runs between the aorta and the pulmo-

nary artery, causes coronary insufficiency and acute ischemia. This, in turn, predisposes the patient to fatal arrhythmias. Occasionally, patients who have an anomalous origin of the left coronary artery from the pulmonary artery may present in infancy with congestive heart failure. They also may die suddenly, presumably of an ischemic arrhythmia or cardiogenic shock.

Acquired coronary artery disease usually is the result of Kawasaki disease. Kawasaki disease may present with SCD in up to 2% of untreated patients. Such deaths may be related to rupture of a large coronary artery aneurysm, acute myocarditis, or a large coronary artery thrombosis. Familial hyperbetalipoproteinemia is an atherosclerotic heart disease inherited in an autosomal-dominant pattern that may cause SCD in homozygotic adolescents.

### Valvular Heart Disease

Patients who have aortic valve disease, including aortic stenosis and chronic aortic regurgitation, may be asymptomatic or present with symptoms such as syncope, dyspnea, or chest pain. If palpitations are present, the patient may be suffering from arrhythmias associated with myocardial ischemia. Symptomatic patients usually have severe left ventricular obstruction and left ventricular hypertrophy. A high estimated valve gradient ( $>75$  torr) measured by echocardiography indicates a risk for SCD. Recommendations for athletic participation are based on the severity of the aortic stenosis.

Mitral valve prolapse (MVP) is a relatively common and benign finding in the pediatric population that is associated with an excellent prognosis. However, patients who have MVP and ventricular arrhythmias, mitral regurgitation, prolonged Q-T interval, history of syncope or pre-

syncope, and family history of sudden death should be considered at high risk to develop SCD. MVP also is seen frequently in association with Marfan syndrome and other connective tissue diseases. Isolated MVP or MVP in association with premature ventricular contractions does not require treatment. However, patients who have MVP and a history of syncope or complex ventricular arrhythmias, significant mitral regurgitation, or a family history of sudden death should restrict their activities to leisurely, noncompetitive sports.

### Arrhythmias and Long Q-T Syndrome

Primary arrhythmias not associated with congenital cardiac malformation occasionally are encountered in the pediatric population and rarely may lead to SCD. These include Wolf-Parkinson-White (WPW) syndrome, isolated sick sinus syndrome, congenital complete atrioventricular block, and ventricular and supraventricular tachycardias. Syncope and palpitations are common presenting complaints in patients who have supraventricular tachycardias from WPW syndrome. SCD is rare in these patients and may occur from rapid conduction via the accessory pathway, leading to ventricular fibrillation. Because digoxin may potentiate this conduction via the accessory pathway, its use in treating WPW syndrome is controversial.

Complete heart block usually presents in infancy; in such patients, SCD generally is related to extreme bradycardia and a tendency to develop ventricular arrhythmias. Sick sinus syndrome may manifest as marked sinus bradycardia, sinus arrest with slow junctional escape, tachycardia-bradycardia syndrome, or atrial fibrillation.

Patients who have long Q-T interval syndrome are at an increased risk

for ventricular arrhythmias and SCD. They usually present with episodes of syncope and a family history of syncope and sudden death. The long Q-T syndrome is inherited in an autosomal-dominant pattern, with female predominance. The risk of cardiac events is higher in males until puberty and higher in females during adulthood. Affected patients exhibit a prolonged Q-T interval and at times profound bradycardia and ST-T wave changes on ECG. Medical therapy is initiated with beta-blocking drugs such as propranolol or atenolol. Beta blockers are associated with a significant reduction in cardiac events among patients who have the long Q-T syndrome. However, syncope, aborted cardiac arrest, and long Q-T syndrome-related death continue to occur among patients receiving beta blockers, particularly those who were symptomatic before starting the therapy.

### Pulmonary Hypertension

Pulmonary hypertension is another significant risk factor for SCD. Among patients who die of SCD, 11% to 17% have pulmonary hypertension associated with heart disease. Another 4% have primary pulmonary hypertension. Syncopal episodes and arrhythmias in patients who have pulmonary hypertension usually sug-

gest a poor prognosis. Affected patients are at a high risk for SCD during pregnancy and delivery and during strenuous physical activities.

### Marfan syndrome

Marfan syndrome is associated with a decreased life expectancy, with 30% to 60% of patients having cardiovascular anomalies. Rupture of the dilated aortic root is the most common cause of SCD in these patients. Mitral valve prolapse with insufficiency is also common. Beta-blocker therapy is recommended for patients who have valve disease.

### Conclusion

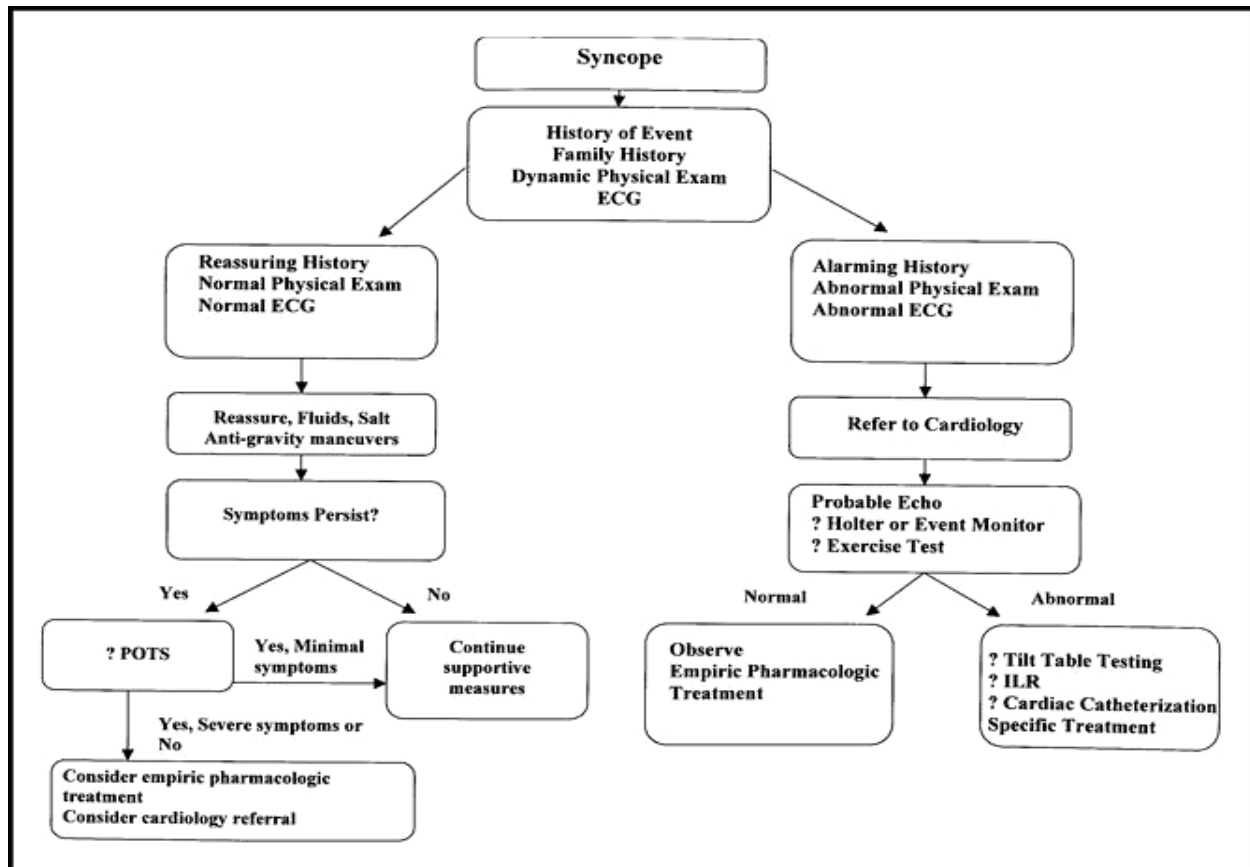
It is important for the pediatrician to understand the various cardiac and noncardiac causes of syncope. SCD in the young is rare. Often, judicious evaluation of the patient who has syncope can determine if he or she is predisposed to a higher risk of SCD. This, in turn, may reduce the risk of SCD substantially and obviate unnecessary restrictions for those in whom the symptoms are due to a benign or easily treatable cause.

### Suggested Reading

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## Algorithm for Syncope Evaluation



Reference: DiVasta AD and ME Alexander. *Fainting freshmen and sinking sophomores: cardiovascular issues of the adolescent.* Current Opinion in Pediatrics, 2004, 16: 350-356

## History Taking 101: Syncope

It is important to know if this is the first or a recurrent episode. Explain to the patient you are going to dissect the most recent episode for every detail in order to fully understand the episode. Ask specific questions and open-ended questions. Example questions:

### **For the patient**

- When was the last episode (exact date)?
- Do you remember the event? (prior and after)
- What were you doing when it occurred?
- What time of day did it occur?
- Were you changing positions? (Getting out of bed, standing up quickly, etc)
- When did you last eat prior to the event? What about drinking fluids (water, soda, etc)?
- Did you feel anything in particular prior to the event?
  - Palpitations? Chest pain? Sweaty? Blurred vision? Dizziness?
- Did you lose consciousness?
- How did you feel when you woke up? Were you dizzy? Any palpitations?
- Do you remember passing out?
- Did you urinate or defecate while unconscious?
- Was this episode similar to any previous episodes?
  - If no, how was it different? Different setting? Timing of day?
- Have you ever passed out while exercising?
- Do you ever feel like you are going to pass out but don't?
  - When does this occur?
  - How frequently?
  - What do you do to prevent yourself from passing out?

### **For the bystander/observer**

- What did they look like prior to passing out? Any pallor? Excessive sweating?
- Did they lose consciousness?
- How long were they unconscious for?
- While unconscious, did they have any large jerking movements? (Some patients will twitch but this is not seizure-like movements – you want to know was this a seizure or not)
- What was their mentation like when they woke up? Normal? Slurred speech? Incoherent? Catatonic?



## Syncope Quiz

1. Define syncope:

- Temporary loss of consciousness due to generalized cerebral hypoperfusion that usually is followed by rapid and complete recovery.
- A sudden, transient loss of consciousness associated with an inability to maintain postural tone that is not otherwise explainable as seizure, vertigo, coma or shock

2. Vasovagal syncope is the most common cause of syncope in pediatric patients, accounting for 75% all syncope cases. (*It is also known as neurally-mediated, simple faint*)

3. What are the important aspects in the history to help differentiate etiologies of syncope?

- “Warning symptoms” or a prodrome – seen often with vasovagal, extremely rare with cardiac
- Preceding event/situation – cardiac often occur with exertion; orthostatic with positional change
- Onset of episode
- Loss of consciousness
- Post-ictal state (catatonic)
- Jerking movements
- Frequency of episodes
- Personal or family history of cardiac problems (e.g. long QT, WPW, HCM, Brugada syndrome, Romano-Ward syndrome)
- Family history of sudden death, unexplained deaths, or drowning
- Do you get pre-syncope easily?

4. Name 5 non-cardiac causes of syncope?

- Vasovagal syncope
- Seizure
- Migraine
- Narcolepsy/Cataplexy
- Familial dysautonomia
- Gastroesophageal reflux
- Spinal cord disease (autonomic instability)
- Breath-holding spells
- Toxins/drugs
- Hyperventilation
- Hysteria
- Fever
- Carotid sinus hypersensitivity–  
*usually older/adult patients*
- Metabolic disease (e.g. diabetes, hypoglycemia, endocrine) – *rare for syncope to be sole presentation*

5. What are some potential cardiac etiologies for syncope?

1) **Outflow obstruction** → less system circulation and thus cerebral hypoperfusion. *On exam, would expect to find a murmur or other clinical findings.*

2) **Myocardial dysfunction** → less systemic circulation due to poor cardiac output. *On exam, would expect gallop or murmur.*

3) **Arrhythmia** → something benign can quickly become malignant (e.g. V-Fib, V-tach, Torsades de pointes). *On history, ask about palpitations before and after event. They may not be felt prior but can be felt afterwards. Also ask about chest pain.*

6. What are the red flags for syncope?

- Syncope with exertion or exercise
- Family history of syncope, sudden death
- Injury with syncope
- Other cardiac symptoms
- History of cardiac surgery
- Syncope with post-ictal state or catatonia
- Syncope without a prodrome
- Abnormal PE (e.g. murmur, neuro change)
- Abnormal EKG

## Syncope Cases

### Case 1

Caitlin is a 15yo female, currently a freshman in high school, who presents after an episode of syncope. She was at her locker before lunch when she fainted and fell to the ground. +LOC that lasted 10-15 seconds but no injuries obtained. The school nurse called home, and Mom brought her in immediately for evaluation.

**What additional information would you like to know?**

- Did she have any preceding symptoms (such as palpitations, dizziness, flushing, vision changes) before fainting?
- Were there any seizure-like movements or post-ictal state?
- Did she eat breakfast?
- Any recent stressors?
- Any personal or family history of cardiac problems?
- Previous syncopal or pre-syncopal episodes?
- How is her overall health? Does she appear healthy?
- Any risk factors for anorexia? Any signs of systemic disease?

Caitlin's friend witnessed the episode and denied any unusual movements while Caitlin was on the ground, and she reported that she was alert with normal mentation upon awakening. Caitlin remembers feeling flushed and dizzy prior to the event, but denies palpitations. She did eat lunch, but admits to not finishing it because she was nervous about a math exam that afternoon. Mom recalls she herself had a similar episode when she was younger when she had to have blood-work drawn. Caitlin also recalls feeling dizzy and warm and thought she was going to pass out a few weeks ago during the Pledge of Allegiance, but symptoms resolved with sitting. No cardiac family history. Mom states Caitlin is a top student and on the dance team; she is otherwise healthy with no medical problems.

**It is now time for your exam and work-up. What are some areas you would focus on? Any special tests?**

- Review VS and make sure BP and HR are appropriate (no dehydration or hypotension).
- Cardiac exam with dynamic testing for murmurs, checking pulses (including femoral).
- Neuro exam with reflexes and Romberg.
- ECG

VS: BP 110/75, HR 80, RR 14, Temp 98.8F, weight 50kg, height 150cm, BMI 22. Physical exam is unremarkable. ECG shows normal sinus rhythm with sinus arrhythmia.

**What is the likely cause of Caitlin's fainting? What supports this diagnosis?**

Vasovagal syncope. Her prodrome prior to fainting was classic for vasovagal syncope. No red flags on history or exam with normal EKG.

**What are some things she can do to prevent fainting in the future? (Hint: see extra credit)**

- Adequate hydration helps ~90% of cases – Dr. Burklow recommends 66oz/day ~2L (8-12oz AM/noon/PM/evening) plus more before sporting events
- Increasing salt in diet and avoiding caffeine
- Avoiding prolonged hot showers and hot, humid weather
- Recognize pre-syncope symptoms and avoid situations that may cause them (e.g. prolonged standing, getting up too quickly)
  - Make sure not to lock knees, know when to sit down, avoid fast position changes
  - Utilize postural techniques: isometric contractions of extremities, folding arms, crossing legs (helps return blood to the heart)
- Ensure good follow-up plan

**Other Management Tips:**

- If symptoms of prodrome begin, patient should sit or lie down until symptoms resolve. While sitting/lying down, can also try the postural techniques.
- If episodes are occurring frequently and starting to interfere with daily activities, consider referring to cardiology (who then may consider starting Fluorinef)\*\*

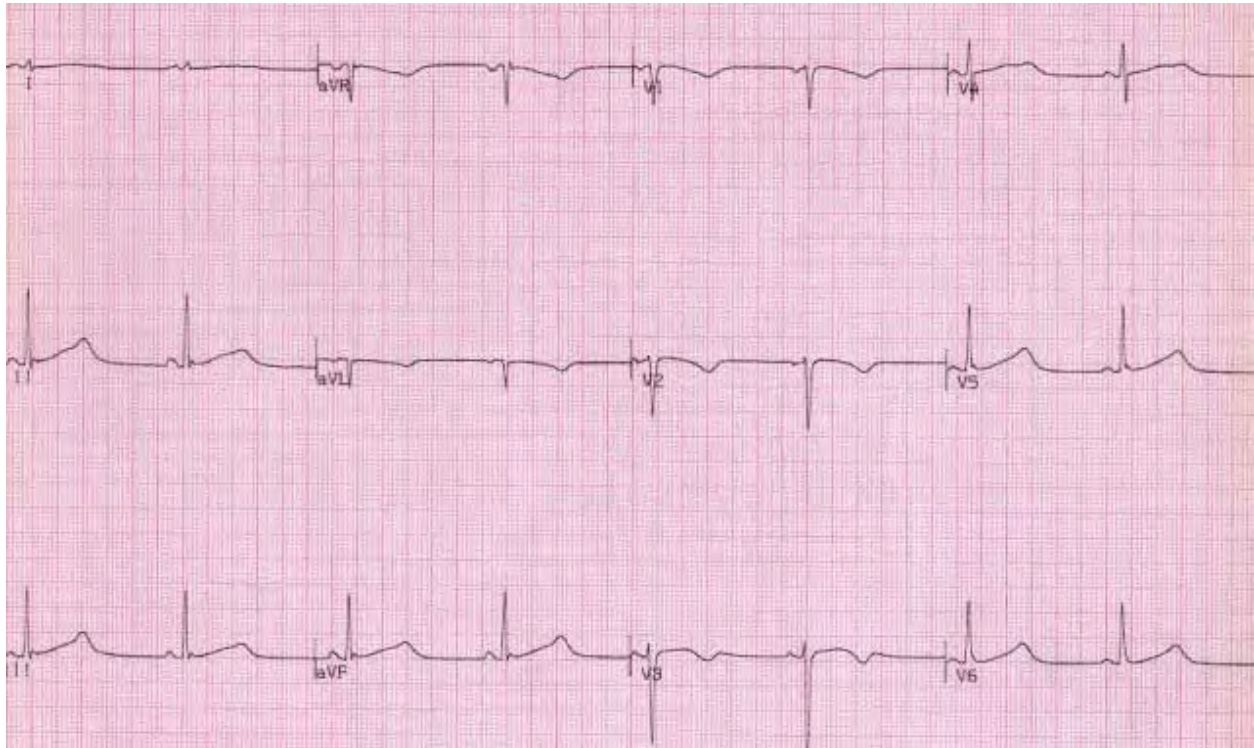
**Case 2**

Jack is a 12yo male who presents for his sports physical prior to trying out for a traveling soccer team. During your history-taking, he admits to fainting last week during a soccer practice. He thought it was due to possible dehydration so he never mentioned it to parents. When asked further about this episode, he recalls that he was running on the field then suddenly collapsed. He was unconscious for about a minute and “came to” when his teammates roused him. No previous episodes of fainting. Mom reports that her brother (Jack's uncle) died of drowning when he was a teenager.

**Are you concerned about Jack? Why or why not?**

Yes. Red Flags: syncope with exertion, family history of possible sudden cardiac death – both concerning for cardiac etiology of syncope.

VS and physical exam are unremarkable. You decide to obtain an ECG in clinic (shown below).  
**Interpret the ECG and calculate the QTc:**

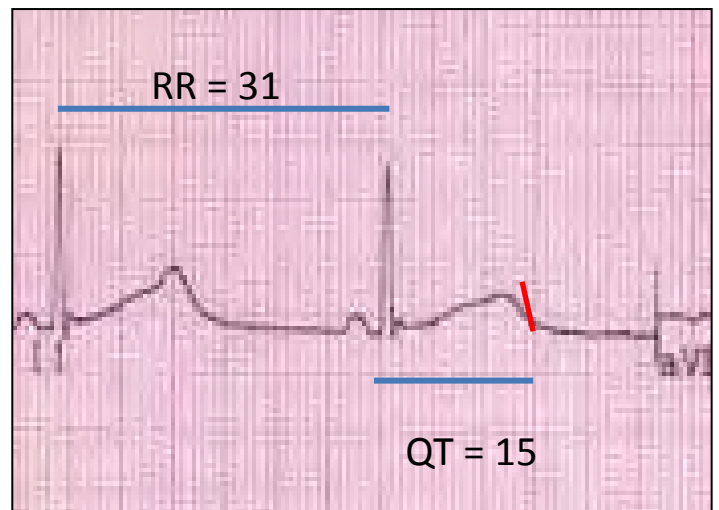


Normal axis, NSR, HR 50  
QTc = 538ms → **Prolonged QT**  
(likely congenital)

$$QTc = \frac{QT}{\sqrt{RR}} = \frac{0.6}{\sqrt{1.24}} \text{ (15 blocks)} = 0.538$$

**What is your next step for the patient? Can you clear him for sports participation?**  
ASAP Cardiology Referral.

No, you must restrict from sports (both practice and games) until cleared by cardiology.



There are various forms of prolonged QT. Some can present with syncope (+/- exertion), but many present as sudden cardiac death. It is always important to obtain an ECG when working up syncope and always calculate the QTc when reading over a ECG.



## Syncope Board Review

1. A 15-year-old girl is brought to your office after “passing out” while she was participating in a band program outside on an 80.0°F day. She recalls feeling lightheaded, then awakening surrounded by her bandmates. The reported duration of the episode was 1 minute. She has had one similar episode in the past. She has no underlying medical problems, and there is no family history of seizures or heart disease. Currently, her temperature is 37.3°C, heart rate is 84 beats/min, respiratory rate is 18 breaths/min, and blood pressure is 98/64 mm Hg. The remainder of her findings, including those of cardiovascular and neurologic examinations, are normal. Of the following, the MOST appropriate next step in her evaluation is:
  - A. Cardiac event monitoring
  - B. CT scan of the brain
  - C. Electrocardiography**
  - D. Electroencephalography
  - E. Tilt table testing

*Although this episode of syncope is likely due to vasovagal (with the hot weather contributing) you must always rule out serious causes of syncope such as an arrhythmia and get an ECG as part of your work-up.*

2. A 15-year-old girl presents after fainting at a concert on a hot day. She complained of palpitations, lightheadedness, and blurred vision prior to losing consciousness briefly. She has had two such episodes in the last month. There is no family history of sudden death. Physical examination reveals no abnormality. Electrocardiography and chest radiograph findings are normal. A tilt test results in dizziness and hypotension. Of the following, the most appropriate management is:
  - A. Calcium channel blocker
  - B. Digoxin
  - C. Increasing fluid intake throughout the day**
  - D. Insertion of a demand pacemaker
  - E. Trial of propranolol

*Again the prodrome and positive tilt test are consistent with vasovagal syncope. A tilt test is no longer commonly used. ABP prep questions stress the importance of identifying vasovagal syncope and red flags for cardiac-related syncope.*

3. Which of the following statements about syncope is *true*?
  - A. Cardiac disease is the most common cause
  - B. Electrocardiography is an essential part of the evaluation for unexplained syncope**
  - C. Fainting usually results in injury
  - D. It is more common in children younger than 10 years of age
  - E. Most children who have syncope have an underlying pathologic cause