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Syncope is the sudden loss of consciousness and postural tone with spontaneous recovery precipitated by cerebral dysfunction. Near-syncope or presyncope is the sensation or feeling that the individual is about to (or nearly) pass out. With near syncope the individual maintains postural tone and does not have a total loss of consciousness. Frequently, patients will confuse syncope or presyncope with vertigo or dizziness. This article will review pediatric syncope, highlighting the differences between adults and children, and critical diagnostic and therapeutic interventions.

— The Editor

Syncope in Pediatric Patients

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Introduction

Epidemiology. Syncope is a common complaint, not just in adults, but also in pediatric patients. Approximately 3% of all emergency department (ED) visits and about 2% of pediatric ED visits are because of syncope.[1,2] Additionally, syncope is the reason for 1%–6% of adult[2-4] and 1% of pediatric hospital admissions.[5] Most commonly, syncope occurs in the 15–19 age group, with at least one episode in up to half of all adolescents (lower range of 10%–20%, upper range of 50%),[6-8] which is similar to reported adult rates.[9] Syncope is less frequent in the young child or infant (≤ 5 years old).[6] Also, syncope has a gender predilection, occurring more often in females than males.[6]

Pathogenesis. No matter what the underlying etiology of syncope, the final common pathway is cerebral dysfunction, either globally affecting both cerebral hemispheres (more commonly) or the reticular activating system in the brainstem.

Impaired cerebral blood flow is most frequently caused by systemic hypotension. Cardiac output (CO) is a function of stroke volume (SV) and heart rate (HR) or CO = HR × SV. Obviously, any

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factor that decreases stroke volume (without an offsetting heart rate increase) or any hemodynamically significant dysrhythmia (whether tachycardia that impairs ventricular filling or severe bradycardia) will cause a decrease in CO and thereby a decrease in cerebral blood flow causing syncope. Less commonly, decreased cerebral blood flow may occur from cerebral vasoconstriction or regional hypoperfusion of the brain due to underlying central nervous system (CNS) illness or injury. Occasionally, syncope may occur when there is normal cerebral blood flow but decreased substrate delivery (e.g., glucose or oxygen) to the brain, as can occur with hypoglycemia, carbon monoxide poisoning, asphyxia, or profound anemia.

PHYSIOLOGIC RESPONSES

Physiologic Responses to Upright Position. The autonomic nervous system (ANS) generally plays a significant role in the pathophysiology of syncope. Cerebral perfusion is maintained by autoregulation and requires adequate cardiac output, which in turn is mediated by the autonomic nervous system via a complex series of cardiovascular system interactions including: heart rate, cardiac function (e.g., contractility, stroke volume), systemic vascular resistance, and the baroreceptors.

Upon standing, blood pools in the lower extremities, but several compensatory mechanisms — the "muscle pump" and reflex responses — work together so that a new steady state is achieved within 1-2 minutes. The reflex responses mediated by sympathetic nervous system activation of the baroreceptors causes increased HR and contractility (↑CO from both ↑HR and ↑SV) and peripheral vasoconstriction, which elevates PVR. Muscle contraction of the lower limbs (specifically, the leg and gluteal muscles) also decreases venous pressure, increasing the leg arterial-venous pressure difference, which promotes blood flow through the lower limb capillaries and veins back to the heart (the "muscle pump"). Immobility as occurs when standing still for a prolonged time, such as a soldier standing at attention, negates the effects of the muscle pump.[10,11]

With a prolonged period of standing, additional physiologic adaptions occur that involve the baroreceptors, neurohumoral responses, and alterations in total blood volume, vascular tone, and cardiac function. Increased circulating catecholamines (e.g., norepinephrine, epinephrine, and vasopressin) and activation of the renin-angiotensin-aldosterone system are part of the neurohumoral response to a prolonged upright posture.[10,11]

Neurocardiogenic Syncope

Neurocardiogenic syncope is also referred to as vasovagal, vasodepressor, neurally mediated, and reflex syncope. As the name implies, neurocardiogenic syncope involves the interaction of various autonomic nervous system reflexes, the central nervous system, and the cardiovascular system.[1,4,12-14] The Bezold-Harisch reflex is cited as the mechanism responsible for vasovagal syncope and has two components. There is "cardio-inhibitory syncope" due to a vagal (parasympathetic) mediated reflex causing bradycardia or even asystole, plus "vasodepressor syncope" from withdrawal of sympathetic input leading to a drop in PVR with venous pooling in the periphery leading to hypotension. "Mixed" vasodepressor syncope implies that both cardio-inhibitory (vagal effect) and vasodepressor (sympathetic withdrawal) components are present.

Vasovagal syncope can occur in heart transplant patients, suggesting that the Bezold-Harisch reflex or vagal stimulation plus sympathetic withdrawal as the only factor may be a somewhat simplistic explanation, and that other variables may also play a role.[10,11] Other postulated mechanisms are: neuro-endocrine mediated responses (possibly involving catecholamines, the renin-angiotensin-aldosterone system, antidiuretic hormone, serotonin, or other neuropeptides); dysfunctional central nervous system control, neuronhumoral mediated sympatho-inhibition, impaired peripheral vascular responses with altered peripheral vascular resistance, and "local" vascular control. An increase in serotonin concentration causing peripheral vasodilatation with a drop in systemic blood pressure is another suggested mechanism.[11] It may be that multiple different mechanism may be acting together to cause vasovagal syncope.[5]

Etiology of Syncope

There are many causes of syncope. *(See Table 1.)* As with all clinical presentations, the emergency physician must be able to distinguish the benign causes of syncope from the serious, life-threatening causes, and then initiate appropriate treatment. We will begin with the most common cause of syncope: vasovagal or neurocardiogenic; next with the most serious and life-threatening causes of syncope (cardiac, neurologic, and respiratory); and then with causes related to postural or orthostatic syncope, including abnormalities of

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the autonomic nervous system, such as the postural orthostatic tachycardia syndrome (POTS), which have been the focus of much recent research.

Neurocardiogenic Syncope

Vasovagal or neurocardiogenic syncope is the most common cause of syncope in children, accounting for well over half of all cases.[15,16] The classic case of vasovagal syncope is the individual who passes out during or immediately following an episode of pain, fear, or anger.

Situational Syncope. Situational syncope is also a reflex (neurally mediated) or vasovagal syncope that occurs during or immediately after a specific activity, such as coughing, swallowing, urination, or defecation. C-fiber mechanoreceptors are located throughout the body (e.g., in the lungs, esophagus, bladder, and rectum). Sudden activation of a substantial number of these mechanoreceptors transmits increased afferent neural traffic to the nucleus tractus solitarius in the medulla then to the ventral lateral medulla resulting in a paradoxical reflex bradycardia (from increased vagal tone) and decreased peripheral vascular resistance (from sympathetic withdrawal).

Carotid Sinus. The carotid sinus syndrome is another reflex (neurally mediated) syncope due to hypersensitivity of the carotid sinus causing vagal stimulation, which leads to bradycardia and/or a drop in blood pressure. Pressure on the carotid sinus, as occurs with a tight collar or head movement, may cause carotid sinus syncope.

Cardiovascular Syncope.

It is critical to diagnose the cardiovascular causes of syncope, since they can be associated with sudden death. Although there are many causes of cardiovascular syncope, the final common mechanism is a decrease in cardiac output causing a decrease in cerebral perfusion. *(See Table 2.)*

Dysrhythmias. Because cardiac syncope can be a harbinger for sudden death, the possibility should be considered in every patient with syncope. The number one etiology of dysrhythmias in pediatric patients is probably congenital heart disease.[17] Sudden cardiac death is associated with both noncorrected congenital heart disease patients and those who are status post corrective cardiac surgery. Sudden death from dysrhythmias is known to occur particularly in congenital heart disease patients status post corrective cardiac surgery for tetralogy of Fallot, transposition of the great vessels (after a Mustard or Senning procedure for a baffle), and single ventricle (after a Fontan procedure).[17-22] Patients with noncorrected congenital heart disease who are especially vulnerable to sudden death include patients with Eisenmenger's syndrome, Ebstein anomaly, atrioventricular canal, aortic stenosis, coronary artery abnormalities, and mitral valve prolapse.[17-22] Secondary or acquired cardiovascular disease can also be associated with sudden death from dysrhythmias and includes myocarditis/cardiomyopathies (for example, hypertrophic obstructive cardiomyopathy, primary/secondary myocarditis), pulmonary hypertension, and Kawasaki's disease.[17-26]

Sudden death from dysrhythmias can also occur in patients with a "structurally normal" heart, as with Wolff-Parkinson-White (WPW) syndrome, short or long QT syndromes, complete heart block, ventricular tachycardia, Brugada syndrome, and arrhythmogenic right ventricular dysplasia.[17-19,21,26-34]

The electrocardiogram (ECG) is invaluable in the diagnosis of cardiac diseases. For example, the classic findings with WPW are a short PR interval and a delta wave. Patients with WPW have an accessory pathway (Kent bundle). Re-entrant conduction down the atrioventricular node and up the accessory pathway can cause supraventricular tachycardia. Rapid conduction over the accessory pathway can lead to ventricular fibrillation and sudden death. The recommended treatment is an electrophysiologic study with ablation of the accessory pathway.

The diagnostic hallmark of the long QT syndrome is QT interval prolongation on the ECG. Abnormal ventricular depolarization causes a prolongation of the refractory period, which can cause torsades de pointes that can degenerate into ventricular fibrillation and sudden death.[26-28]

Secondary causes of QT prolongation may be drugs/toxins, electrolyte abnormalities, and increased intracranial pressure. A prolonged QT interval may result from hypokalemia, hypocalcemia, psychotropics (notably tricyclic antidepressants), phenothiazines, antihistamines, and promotility drugs (e.g., cisapride), particularly with concomitant administration of other drugs (e.g., erythromycin).

With the short QT syndrome, there is a high risk of sudden death, even in infancy. Patients with the short QT syndrome have a structurally normal heart. Generally, the QT interval is ≤ 320 msec and the QTc ≤ 340 msec.[29] The clinical presentation of patients with a short or long QT syndrome, as with any of the tachyarrhythmias, includes syncope, palpitations, and sudden death.[26-29]

The bradyarrhythmias, which include sick sinus syndrome and complete heart block, can also cause syncope and sudden death. The sick sinus syndrome and a high degree AV block may present with the Stoke-Adams syndrome. Signs and symptoms of the Stokes-Adams syndrome include: a slow or absent pulse, vertigo, syncope, seizures, and Cheyne-Stokes respiration.

Congenital heart block may be part of a structural heart defect, occurring most commonly with congenital defects of the atrioventricular septum, L-transposition of the great vessels, or ventricular inversion. Congenital heart block may also occur as an isolated condition in structurally normal hearts; in infants whose mothers have systemic lupus erythematosis. Acquired causes of atrioventricular heart block are probably more common than congenital heart block. The most common etiology of acquired heart block is a postoperative complication of cardiac surgery. Rheumatologic/ immunologic disease, infections, and neuromuscular disorders are other causes of acquired heart block. Lyme disease and acute rheumatic fever are examples of rheumatologic/immunologic diseases that can cause atrioventricular block. Neuromuscular diseases that can cause heart block include muscular dystrophy, myotonic dystrophy, and Kearns-Sayre disease. Heart block may also be a complication of an acute infection such as endocarditis or diphtheria.

Sinus node dysfunction—atrial dysrhythmias, specifically atrial fibrillation or atrial flutter—may occur in post-operative cardiac surgery patients. The hemodynamic compromise that occurs with combined sinus node dysfunction and atrial flutter/fibrillation with a rapid ventricular response is particularly dangerous and may lead to syncope and sudden death.

Table 1. Etiology of Syncope in the Pediatric Patient

CARDIOVASCULAR: CONGENITAL OR ACQUIRED

Cardiac

- Dysrhythmias
- Myocardial disorders
- Valvular disease
- Pericardial disease
- Congenital heart disease
- Pulmonary hypertension
- Cardiac tumors (primary or metastatic): Atrial myxoma, rhabdomyosarcoma

Vascular

- Aortic disease: Severe coarctation, aortic dissection
- Coronary artery disease / disorders
- Disease / disorders of great vessels (other than aorta)
  - Subclavian steal
  - Thoracic outlet

NEUROLOGIC

- Central nervous system disorder
  - Hemorrhage
  - Stroke, transient ischemic attack
  - Migraine headaches
- Peripheral nervous system
  - Neuropathies

RESPIRATORY

- Hypoxia from respiratory failure or pulmonary disease
- Pulmonary emboli (can cause syncope by mechanism other than hypoxia)
- Cough (post-tussive)
- Breath-holding: Cyanotic, pallid, mixed (likely from autonomic dysregulation)

Disorder of Oxygen Transport

- Carbon monoxide poisoning
- Cyanide poisoning
- Other toxins
- Severe anemia

METABOLIC / ENDOCRINE

- Hypoglycemia

DRUGS / DRUG OVERDOSE

- Poisons/toxins *(see also* Disorders of Oxygen Transport, *above)*
- Drugs of abuse: Cocaine, opiates, alcohol, others
- Prescription drug abuse (intentional): Benzodiazepine, opioids, others
- Prescription drugs (unintentional): Antihypertensives, cardiac especially vasodilators (calcium channel blockers, beta blockers, nitrates), psychiatric drugs especially tricyclics, phenothiazines
- Nonprescription
- Over-the-counter drugs/herbals/vitamins

HYPOVOLEMIC / HEMORRHAGIC

- Decreased intravascular volume (from blood or volume loss)

POSTURAL (ORTHOSTATIC)

- Postural orthostatic tachycardia syndrome (POTS)
- Dysautonomic syndromes: Primary or secondary autonomic failure

NEUROCARDIOGENIC

- Vasovagal
- Situational
- Carotid sinus syncope

PSYCHIATRIC

- Anorexia nervosa, bulimia
- Hyperventilation (anxiety, panic disorders)

Dysrhythmias in patients status post cardiac surgery generally occur in the early postoperative period (within hours, days, or weeks) but may occur months and even years after surgical repair, probably from scarring and myocardial damage. Ventricular arrhythmias (e.g., ventricular tachycardia/fibrillation) may occur in patients with any type of myocardial disease.

Commotio cordis (concussion of the heart) is an unusual cause of sudden death. This lethal sports injury has been reported in baseball and hockey players.[35] Blunt chest trauma occurring during the electrically vulnerable period of the cardiac cycle leads to ventricular dysrhythmias (e.g., ventricular tachycardia/fibrillation) that result in sudden death.[35]

Myocardial Disease. Cardiac syncope in patients with myocardial disease may be due not only to dysrhythmias but also to poor cardiac output from impaired contractility and/or inadequate stroke volume. There are many causes of myocardial disease/injury, including: the various cardiomyopathies (viral-most common, but also drugs/toxins including cocaine, alcohol, inherited-hypertrophic obstructive cardiomyopathy [HOCM]), myocarditis (primary or secondary as from Lyme disease or acute rheumatic fever), myocardial

Table 2. Cardiovascular Causes of Pediatric Syncope

ARRHYTHMIAS: CONGENITAL OR ACQUIRED

- **Tachyarrhythmias**
  - o Wolff-Parkinson-White syndrome
  - o Ventricular tachycardia
  - o Ventricular fibrillation
  - o Arrhythmogenic right ventricular dysplasia
  - o Torsades de pointes
- **Device Malfunction**
  - o Pacemaker malfunction
  - o AICD malfunction
- **Specific Congenital Heart Defects**
  - o High-risk patients: Ebstein anomaly, tetralogy of Fallot, others
- **Post-operative Congenital Heart Disease**
- **QT Abnormalities**
  - o Long QT syndromes: Congenital Romano-Ward syndrome, Jervell and Lange-Nielsen syndrome
  - o Acquired: Medications causing QT prolongation: Psychotropics (tricyclic antidepressants, phenothiazines, promotility drugs [cisapride] — especially in combination with other drugs [e.g., erythromycin, ketoconazole])
  - o Short QT syndrome

STRUCTURAL DISORDERS: MYOCARDIAL DYSFUNCTION / DISEASE

- **Primary**
  - o Myocarditis
  - o Dilated cardiomyopathy
  - o Idiopathic hypertrophic subaortic stenosis (IHSS)
  - o Arrhythmogenic right ventricular dysplasia (*see also* Arrhythmias)
- **Secondary (Acquired)**
  - o Infections:
    - – *Viral:* Coxsackie, others
    - – *Parasitic:* Chagas disease
  - o Immunologic, vasculitis, rheumatologic diseases: Lyme disease, amyloidosis, sarcoidosis
  - o Generalized neuromuscular diseases: Muscular dystrophy

CORONARY ARTERY DISEASE

- **Congenital:** Anomalous coronary artery
- **Acquired:** Kawasaki disease

VALVULAR

- **Severe / Critical Aortic Stenosis**
- **Severe / Critical / Mitral / Pulmonic / Tricuspid Valve Disorder** (stenosis or regurgitation)
- **Prosthetic Valve Dysfunction**

VASCULAR

- **Aorta:** *Aortic dissection*, secondary to hypertension, atherosclerosis, connective tissue disorders (Marfan disease, Ehlers-Danlos syndrome)
- **Other Great Vessel Abnormalities**
  - o Subclavian steal syndrome
  - o Thoracic outlet syndrome

OUTFLOW OBSTRUCTION TO SYSTEMIC BLOOD FLOW

- **Severe Coarctation**
- **Cardiac Tumor Mass** (atrial myxoma)
- *Also*: **Hypertrophic Obstructive Cardiomyopathy, Critical Aortic Stenosis**

PERICARDIAL DISEASE

- **Pericarditis / Pericardial Tamponade**

CONGENITAL HEART DEFECTS

- **Cyanotic / Acyanotic Congenital Heart Disease**
- **Eisenmenger Syndrome**

PULMONARY HYPERTENSION

- **Primary** (idiopathic)
- **Secondary**

ischemia/infarct (congenital: anomalous coronary artery; or acquired: Kawasaki disease), and diseases that destroy the myocardium secondary to a rheumatologic/ immunologic process (such as sarcoidosis or amyloidosis), or due to a generalized neuromuscular disorder (e.g., muscular dystrophy, myotonic dystrophy), from an infection (Chagas disease), or a nutritional deficiency (beriberi).

Individuals with hypertrophic cardiomyopathy have an increased risk of death from several causes, including sudden cardiac death, heart failure, and stroke. Patients with a dilated cardiomyopathy who have a low ejection fraction have a high risk of syncope and sudden cardiac death.

Ventricular tachycardia or ventricular fibrillation is the likely mechanism for sudden cardiac death occurring in young "apparently healthy" individuals, many of whom have underlying cardiovascular disease that is not diagnosed until after the event.

Cardiac Disorders with Obstruction to Flow. Another group of cardiac disorders that can present with syncope and sudden death involve obstruction to flow. Severe aortic stenosis (AS) may cause significant obstruction such that cardiac output (and coronary blood flow) is markedly impaired. Symptoms occurring with critical AS, severe mitral valvular disease, or marked coarctation of the aorta include chest pain, exertional dyspnea, and syncope especially with exercise. Impaired cardiac output is also the likely mechanism for the syncope occurring with "critical" or severe pulmonic stenosis. HOCM can also cause syncope by outlet track obstruction.

A sudden increase in pulmonary vascular resistance causing a decrease in blood flow leading to dysrhythmias is thought to be the mechanism of sudden death in patients with Eisenmenger syndrome.

Syncope and sudden death is also a complication of pulmonary hypertension, whether primary (idiopathic) or secondary (from congenital heart disease).[36]

Cardiac tumors are rare causes of cardiac syncope. Cardiac syncope can be due to impaired cardiac output, or blockage or obstruction to flow (as occurs with an atrial myxoma); while other cardiac tumors (such as rhabdomyosarcoma) infiltrate the myocardium causing myocardial disease with impaired contractility and decreased cardiac output. Inadequate cardiac output also occurs with pericardial effusion, which can lead to cardiac tamponade.

Vascular Causes of Syncope. Vascular causes are another major category of cardiac syncope, ranging from aortic dissection to subclavian steal and thoracic outlet syndrome.

Neurogenic Syncope

Syncope from neurologic disease is from cerebral hypoperfusion, either global/generalized or localized CNS hypoperfusion, or from cerebral vasoconstriction. Among the many disorders causing syncope from focal CNS hypoperfusion are CNS trauma, hydrocephalus, brain tumors (primary or metastatic), subarachnoid hemorrhage, any vasculitis affecting the cerebral vessels, and transient ischemia attack or stroke. Generally, other signs and symptoms, not just syncope, occur with neurogenic syncope.

Respiratory Syncope

Although respiratory distress/failure has many etiologies (for example, severe pneumonia or status asthmaticus), the common pathway for respiratory disease precipitating syncope is usually hypoxemia with decreased oxygen delivery to the CNS.

Hyperventilation syncope is due to cerebral arterial vasoconstriction secondary to hypocapnia from the hyperventilation. Usually, with hyperventilation syncope, there is a history of anxiety, fear, pain, or emotional distress and there may be associated signs/symptoms such as light-headedness, or dizziness, chest pain, dyspnea, paresthesias, and tachypnea.

Decreased substrate (e.g., oxygen/glucose) delivery to the CNS can also cause syncope. This is the case with inadequate glucose transported to the CNS with hypoglycemia, and decreased oxygen delivery to the CNS with carbon monoxide poisoning, asphyxia, cyanide poisoning, and other toxins. Pulmonary hypertension has also been associated with syncope and sudden death.[36]

In young children, especially those younger than 3 years of age, breath-holding spells should be considered. Breath-holding spells are common, occurring in about 4.6% of children.[37] Breath-holding spells usually begin between 6 and 18 months of age (with a range from the first week of life to 3 years), and end by 6 years of age in most (90%) of children.[38] There are two types of breath-holding spells: cyanotic and pallid.[37-39] Cyanotic spells are more common. Both spells are due to transient cerebral anoxia.

Typically, the cyanotic breath-holding spell is precipitated by the child/infant crying after being angered. The vigorous crying results in transient cerebral ischemia caused by cerebral arterial vasoconstriction from hypocapnia, hypoxemia from apnea, and decreased cardiac output from a prolonged Valsalva effect. The pallid breath-holding spell generally occurs after a painful or anxiety-producing incident, such as a minor fall. The transient cerebral ischemia occurring with the pallid breath-holding spell is due to an exaggerated vagal response to pain, causing prolonged bradycardia or asystole. Breath-holding spells are a benign entity with no major sequelae, although they are commonly misdiagnosed as seizures. An ECG should be considered in a pediatric patient with a breath-holding spell to evaluate for electrocardiographic abnormalities such as prolonged QT syndrome.

Orthostatis (Postural) Syncope

Orthostatic hypotension is a fairly common cause of syncope in children, and is due to a failure of normal physiologic compensatory mechanisms with postural changes.[16,40] Postural (orthostatic) hypotension with syncope is due to a fall in blood pressure upon assuming the upright posture caused by a decrease in blood volume and/or a loss of vasoconstriction reflexes in the vasculature of the lower extremities. The precipitants are rising suddenly from a recumbent position or prolonged standing.

Hypovolemia

Conditions causing hypovolemia that can lead to syncope often, but not always, are precipitated by postural changes including: hemorrhage, dehydration from vomiting or diarrhea or decreased oral intake, and adrenal insufficiency. Prolonged standing or bed rest can also be predisposing factors for syncope due to orthostatic hypotension.

Dysautonomic Syncope

Autonomic nervous system diseases, by destroying the autonomic pathways, can precipitate syncope due to postural hypotension.[41] Dysautonomic syndromes may be primary or secondary. Neurologic diseases or conditions that destroy autonomic pathways are referred to as primary disorders, while systemic diseases such as diabetes that destroy or disrupt autonomic pathways are considered secondary diseases.[42] Primary or secondary autonomic disorders may be central (affecting the brain or central nervous system) or peripheral (affecting either the peripheral autonomic nerves or the spinal nerves).

Neurologic disorders that can lead to a central form of dysautonomia include multiple sclerosis, CNS tumors (primary or secondary), and syringobulbia. Primary (neurologic) diseases (central or peripheral) causing dysautonomic syndromes are less common than secondary autonomic nervous system dysfunction. Certain inherited disorders, such as familial dysautonomia or dopamine-β-hydroxylase deficiency, are causes of peripheral secondary autonomic failure.[41,43-45] Physical deconditioning that can occur with a lengthy illness with bed rest or after prolonged weightlessness from a space flight can also cause postural syncope. Orthostatic syncope can occur after a spinal cord injury or sympathectomy, which eliminates the vasopressor reflexes, and in patients on certain medications, commonly antihypertensive and vasodilator drugs.[46] Some authors consider the reflex syncopes (e.g., vasovagal or neurocardiogenic syncope, situational syncopes, and carotid sinus hypersensitivity) as a category under autonomic disorders,[43,44] while other authors separate them and consider the orthostatic/dysautonomic disturbances of blood pressure control as one category and the neurally mediated

reflex disturbances of blood pressure/heart rate control, or vasovagal (neurocardiogenic) syncope as a second separate group.[40,46]

Postural Orthostatic Tachycardia Syndrome. POTS is a type of neurocardiogenic syncope characterized by an exaggerated heart rate increase in response to postural change. It is the most common form of orthostatic intolerance.[10] Upon standing, patients become tachycardic and may complain of dizziness, lightheadedness, blurred vision, fatigue, palpitations, and anxiety. Syncope occurs in about 40% of patients. Patients present at a fairly young age, typically as teenagers or young adults (age 14–45 years). It is thought to be due to abnormal baroreflex control with increased sympathetic activity. A low plasma volume may play a role,[42] and volume expansion (saline infusion or oral volume expansion), a high-salt diet, and fludrocortisones are come of the suggested therapies.[45]

Diagnostic Studies

In the evaluation of any patient with syncope, a history and physical examination that includes vital signs with pulse oximetry and orthostatic vital signs are the critical factors in making the diagnosis and in determining what ancillary diagnostic tests are indicated. The first question is "Is the transient LOC syncope or not? Or, is the LOC another disorder associated with LOC that is a mimicker of syncope (such as a seizure or hypoglycemia)?" If it is syncope, the major consideration is whether the syncope is due to a benign or life-threatening cause. In the patient with near syncope or pre-syncope, the syncopal symptoms abate before LOC occurs. However, the evaluation and management of the presyncope patient is basically the same as for the patient with syncope.

The history alone may yield the etiology of the syncope. When there is a "classic history," there may be no (or a limited) need for additional diagnostic studies, assuming there were no risk factors elicited from the history or physical examination. Examples of a pathognomic history include an adolescent with vasovagal syncope who fainted while having blood drawn for a routine physical or an infant with the typical features of a breath-holding episode. However, since individuals with underlying significant cardiac, neurologic, or other disease may also have a "benign" syncope such as a vasovagal syncope, some experts recommend limited testing such as an ECG, orthostatic vital signs, and a blood glucose in most or all patients.[15]

The history begins with details of the syncopal event: activity/position when event occurred, onset, prodrome, aura, reoccurrence, precipitating factors/situations, associated signs or symptoms (such as bowel or bladder incontinence), recovery time frame, and signs or symptoms, if any, during recovery (sleepiness, headache, or confusion may occur with a postictal period). The history should also include a review of systems and medications (ask about street drugs, especially cocaine, over-the-counter, and herbal drugs as well as prescription medications), allergies, social history, family history (especially of cardiovascular or neurologic disorders, sudden death, or myocardial infarction [MI] at an early age), social history (for example, any exposure to toxins, drugs of abuse including alcohol), unusual diet or eating disorders, past surgery (especially cardiac or neurosurgical procedures, even if remote), and past medical history, including known conditions/ diseases (diabetes, malignancy, pulmonary, cardiac, anemia, neurologic, psychiatric, other).

The physical examination is also invaluable in excluding other causes of LOC from syncope and in determining the etiology of the syncope. Although the vital signs (including the pulse oxygen saturation), cardiovascular, neurologic, and pulmonary examination are critical in the evaluation, the remainder of the physical examination should not be overlooked. An unusual facies or appearance may be a clue to a syndrome, such as Marfan's or Down's syndrome, that is associated with cardiac or neurologic disease. Signs of dehydration suggest orthostatic (postural) syncope from volume depletion. Consider neuromuscular disease with possible cardiac involvement and cardiac syncope if there is musculoskeletal weakness or abnormal muscle mass or tone. Abnormal lung findings (e.g., rales, wheezing) suggest pulmonary (asthma, pneumonia) or cardiac disease (congestive heart failure [CHF]). The extremity examination may reveal edema (from CHF, renal, hepatic, other diseases) or calf pain and swelling from a deep vein thrombosis with the risk of pulmonary emboli. The skin should be examined for color (cyanotic, pale, jaundiced), capillary fill (normal or delayed due to decreased volume or blood loss), appearance, ecchymosis, signs of trauma, stigmata of underlying disease (café-au-lait spots, ash leaf, etc., as seen with neurologic disease), or signs of infection (e.g., cellulitis, petechiae with meningococcemia).

The vital signs should be noted. An irregular rhythm and/or abnormal heart rate (bradycardia or tachycardia) is suggestive of cardiac syncope from a dysrhythmia. Any differences in blood pressures or pulses between the extremities may denote a coarctation of the aorta, a dissecting aneurysm, or subclavian steal. With any abnormal cardiac finding: murmur, click, S3, S4, loud S2, abnormal S2 split, or pericardial friction rub; the possibility of cardiac syncope must be ruled out. Of course, any neurologic abnormality or focal neurologic finding is a red flag for syncope due to a neurologic cause.

Some form of postural syncope, whether from volume or blood loss or autonomic dysfunction, is the likely cause of syncope in patients with abnormal orthostatic vital signs, supine tachycardia, or dizziness upon assuming an upright posture.[47] Syncope from severe anemia or bleeding is suggested by pallor. Gastrointestinal bleeding and syncope secondary to blood loss should be considered when there is a positive hemoccult test of the stool, hematochezia, hematemesis, or melena.

In females of childbearing age, consider a pregnancy test since bleeding during pregnancy or an ectopic pregnancy can present with a chief complaint of syncope. Since hypoglycemia frequently presents with syncope or near syncope, an Accu-Chek® or serum glucose is another simple, readily available, inexpensive test that should be considered, particularly in diabetics.

An ECG should be considered in syncope patients of any age, although the yield may be low.[48,49] An ECG is easily obtained, painless, noninvasive, and readily available; and furthermore, may be diagnostic. For all these reasons, some experts recommend an ECG in any syncope patient, irrespective of the history, because the "benign" syncope (such as vasovagal) is so common that it can occur in patients with significant underlying cardiovascular disease.[15,50-54]

According to the American College of Cardiology, after the history and physical examination, the ECG is the first diagnostic test in evaluating a patient with syncope.[55] In adults, the history, physical examination, and an ECG will establish the diagnosis in up to 69% of syncope patients.[56] Whenever cardiac syncope is a possibility, obtain an ECG; check the QT interval and evaluate the PR interval and QRS complex for evidence of WPW syndrome as well as for any tachycardias, bradycardias, dysrhythmias, and additional ECG abnormalities. Remember that acute MI is rare but is possible even in children and infants, and can occur with a congenital anomalous coronary artery or Kawasaki disease.

An abnormal QT (long QT syndrome or short QT syndrome) is associated with syncope and sudden death. Assuming a stable sinus rhythm, a QTc < 80% of predicted is consistent with the short QT syndrome and a QTc > 0.44 is an abnormally prolonged QT consistent with the long QT syndrome. The corrected QTc is calculated using Bazett's correction where the corrected QTc = $QT/\sqrt{RR}$.[57] Additional evaluation such as an echocardiogram, Holter monitoring, pediatric cardiology consult, or even hospital admission may be indicated if cardiovascular syncope is suspected.[58]

A head CT scan may be warranted in certain patients, especially if neurologic signs or symptoms are present or new onset seizures are suspected, although routine CT scans are generally not indicated.

Standing upright or tilting a "normal" individual on a tilt table allows some blood to accumulate in the lower extremities with a mild diminution in cardiac output and sometimes a slight transient drop in systolic blood pressure. In patients with abnormal vasomotor reflexes or a decreased blood volume, standing (or tilt-table testing) can cause an abrupt and sustained drop in blood pressure, causing syncope. In patients in whom dysautonomic syndromes are a possibility, referral for tilt-testing should be considered. There has been increased use of tilt-table testing in both adult and pediatric syncope patients, particularly those with "unexplained" syncope.[59-65] However, a recent study has found that blood volume derangements are common in syncope patients and are not identified by tilt-table testing.[66] They recommend blood volume measurements should be added to tilt-table testings, especially in adults, when evaluating the 24% to 37% of patients in whom the cause of syncope is unknown even after standard diagnostic evaluation and testing.[66] Whether this finding (e.g., blood volume derangements) in adults applies to pediatric patients warrants investigation.

Other frequently obtained diagnostic tests in addition to the ECG are a fingerstick blood sugar or blood glucose, a qualitative pregnancy test in females of childbearing age, and a hematocrit. This is followed by a focused approach to diagnostic testing based on the initial history and physical examination.[8,51,52]

Differential Diagnosis of Syncope

Not only is there an extensive list of causes of syncope, there are also numerous entities that have similar characteristics with syncope. Differentiating these various disorders from syncope can be difficult. Approximately one out of eight adult patients (12.3%) presenting to an emergency department with a transient loss of consciousness had a diagnosis other than syncope.[4] Some of the patients had an LOC (most commonly due to an underlying neurologic or metabolic disorder), or a condition that mimicked LOC. The neurologic diseases that presented with LOC were transient ischemia attacks, seizures, and migraines. In addition to hypoglycemia (which was the number one disorder presenting with LOC), other "metabolic" causes of LOC were hypoxia and hyperventilation. Among the conditions mimicking a LOC were psychiatric diagnoses (e.g., somatization disorders) and drop attacks.[4] In another study, an even higher incidence (29%) of patients presenting to an ED with a transient LOC did not have syncope but instead were diagnosed with seizures.[1]

Many neurologic diseases and metabolic disorders can present with a transient LOC. *(See enclosed* Rapid Access Management *card for table "Differential Diagnosis of Syncope in Pediatric Patients.")* However, most of these CNS disorders will have other neurologic signs or symptoms such as a focal deficit and infrequently present with isolated syncope.

Although patients with neurocardiogenic or vasovagal syncope may have momentary jerking of extremities (myoclonic jerks) similar to the tonic-clonic movements occurring with generalized seizures — and conversely, the "drop attack" of an akinetic seizures may be similar to a syncopal episode, various features in the history and physical examination are useful in distinguishing syncope from seizures. With syncope, there is no aura and the LOC is very brief, with rapid recovery to normal alertness/mental status within seconds without any postictal confusion, sleepiness, or headache. The presence of any of the following are consistent with a seizure not syncope: focal neurologic signs (by history or examination), an aura preceding the LOC, post-episode confusion/somnolence/headache, incontinence of bladder and/or bowel, tongue-biting, and prolonged recovery to "usual" normal state.

If it is unclear whether syncope or a seizure occurred, a low serum bicarbonate and/or an elevated prolactin level are suggestive of a seizure and may help in making the diagnosis. The serum prolactin may be useful to help differentiate a seizure from a psychogenic nonepileptic seizure in older children and adults.[67] As with patients with breath-holding spells or syncope, an ECG has also been recommended in patients with seizures to exclude any ECG abnormalities such as prolonged QT syndrome.[50]

Metabolic disorders — most frequently hypoglycemia, but also organ dysfunction/failure (such as liver failure, or kidney failure), inherited metabolic disorders, toxins (including carbon monoxide poisoning), and medications (illegal drugs, alcohol, herbal, and prescribed medicines) — may occasionally present with an altered mental status.

The patient with hyperventilation and syncope usually has a history of tachypnea, often with paresthesias and sometimes carpopedal spasm. Psychiatric disorders that may have a LOC include conversion disorders, malingering, and Munchausen's syndrome.[68] Characteristics of "hysterical syncope" include secondary gain, occurrence before an audience, attention-seeking behavior, lack of injuries from the "syncopal" episode, and frequently, occurrence with histrionic gestures or moaning.

Infants and young children have several nonepileptic paroxysmal disorders that can be confused with syncope. In addition to cyanotic or pallid breath-holding spells; other disorders that may mimic syncope include apnea, jitteriness, benign neonatal sleep myoclonus,

hyperekplexia (startle disease or stiff baby syndrome), benign myoclonus of infancy, shuddering attacks, Sandifer syndrome (intermittent paroxysmal spells of generalized stiffening and opisthotonic posturing secondary to gastroesophageal reflux), benign toricollis, dystonia, and rhythmic movement disorders (e.g., head rolling, body rocking, head banging).

The differential diagnosis of transient LOC includes seizures, syncope, breath-holding spells, mild head injury, migraines, and psychosomatic disorders.

Management and Disposition

The history, physical examination, and ECG determine the need for additional diagnostic testing, consultations, and admission. Although the majority of causes of syncope in pediatric patients are benign, some etiologies can be life-threatening. Syncope may be a clue to a cardiovascular condition associated a with a high risk of sudden death. Patients with suspected cardiac syncope warrant diagnostic evaluation, consultation, and generally, admission. Patients with orthostatic syncope secondary to dehydration or volume depletion need rehydration in the ED and can then be discharged. Young children and infants with typical breath-holding spells and patients of any age with vasovagal syncope can be discharged home. Patients with syncope secondary to dysautonomic syncope and some patients with unknown syncope require referral for additional evaluation.

Acute Illnesses Presenting as Syncope

A significant number of pediatric ED syncope patients have an underlying medical condition. In one study of pediatric ED patients, 41% had an "underlying disease, predominantly dehydration, seizures, and anemia." In two studies of adult ED patients with syncope, the incidence of underlying disease was nearly one-quarter of the patients: 22.3% (neurologic/cardiac/drug-metabolic) in one report and 21.5% (gastrointestinal bleeding, postural hypotension, cardiac, CNS, metabolic/drug) in another study.[12,13]

Etiology of Syncope

After initial evaluation, the etiology of the syncopal episode may be unknown in as many as half (40%–50%) the patients, whether adult or pediatric. Series of patients with syncope have found the "unknown" category to be from 27.5%–50% for pediatric patients.[15,68,70] After extensive testing and specialty evaluation, 27.5% of children with syncope had no identifiable etiology despite complete cardiology evaluation,[69] 36.2% (after both pediatric cardiology and neurology evaluation)[70] versus 50% for a pediatric ED evaluation with limited testing.[15] Of those patients in whom a diagnosis was made, the history and physical examination was sufficient for the diagnosis in 77.6% of pediatric patients.[16]

When the diagnosis is made after initial ED evaluation, vasovagal syncope is by far the most common etiology in up to half of the pediatric and adult patients (pediatric ED patients: 50%–54%, and pediatric referral clinic after extensive testing 25.7%– 32.5%).[15,16,69,70]

In pediatric ED patients, the second most common cause of syncope was orthostatic hypotension (20%) due to either dehydration (15%) or anemia (5%), followed by neurologic diagnoses (17.5%) that included atypical seizures (7.5%), migraines (5%), and minor head trauma (5%).[71] After unknown (37.6%) and vasovagal (37.1%) syncope, the next most common etiologies of syncope were neurogenic (e.g., new onset seizure) (8.8%), or orthostatic (7.6%), followed by cardiac (4.1%), and psychogenic (0.6%).[14]

Prognosis of Syncope

The prognosis ultimately depends on the etiology of the syncopal episode. In adults, cardiovascular causes have the worst prognosis, with one year mortality rates as high as one in three (33%)[1,12,72,73] versus a 6%–12% mortality rate for noncardiovascular causes.[1,72,74] Although similar longitudinal studies have not been done for pediatric patients, the overall consensus is the same: cardiovascular syncope has the worst prognosis, with a significant increased risk of sudden death.[5,18,20,21]

Syncope and Sudden Death

Although the etiology of syncope is benign in the majority of patients, syncope may be the harbinger of sudden death. Pediatric and adult patients with syncope secondary to cardiovascular disease have an increased increased risk of sudden death.[22,72,73,75] One fourth (25%) of children and adolescents with sudden death had syncope or presyncope, with two thirds of the syncope/presyncope episodes associated with exercise.[6] Thus, a history of syncope occurring with exercise should be a red flag for potential underlying significant cardiovascular disease. *(See Table 2.)*

Longitudinal cohort studies of pediatric syncope and sudden death are limited by the lower incidence of life-threatening cardiovascular arrhythmias and the more extensive list of causes of sudden cardiac death in infants and children, unlike adults in whom atherosclerotic coronary artery disease (CAD) is the principal cause of sudden cardiac death. However, certain cardiac diagnoses that are most commonly associated with sudden death have been identified.[17-25,76-78]

In one study of pediatric survivors of sudden cardiac death, all of the survivors had known cardiac disease prior to their presenting episode of sudden death.[19] The diverse causes of sudden cardiac death in pediatric patients can be classified into several categories: congenital or acquired structural heart disease, "primary" dysrhythmias, pulmonary hypertension, and trauma (e.g., commotio cordis). Patients with significant congenital heart disease have a higher incidence of sudden cardiac death and/or dysrhythmias. Patients with Eisenmenger syndrome have a 10%–47% incidence of sudden death.[18] In tetralogy of Fallot, the incidence of ventricular tachycardia is 10% and of sudden cardiac death is 2.25%.[18] Other congenital heart defects and their incidence of sudden cardiac death are: Ebstein's anomaly 2.5%–20.0%, arterioventricular canal and ventricular septal defect 5.8%, D-transposition after intra-arterial repair 2.8%, status post Fontan repair 3%, aortic stenosis 1%, and Kawasaki syndrome 1%.[18] Patient's status post surgery for congenital heart disease and patients with abnormalities of the coronary arteries have an increased risk of sudden cardiac death.[18] Overall, the incidence of sudden death in the pediatric population is relatively infrequent,[21] with reported incidences of 1.3 cases per 100,000 patient years with about one third of these "definitely" cardiac related and another one fourth "probably" cardiac related.[22] In a 23-year cardiovascular reg-

istry at a given hospital, only 50 cases of sudden death in the age range of 7–35 years were noted.[76] Hypertrophic obstructive cardiomyopathy, acute myocarditis, and dilated cardiomyopathy are also associated with sudden death.[19-23] Patients with pulmonary hypertension, primary or secondary (as in Eisenmenger syndrome), have a significant risk of sudden death.[18-22] Acute dissecting aortic aneurysm has been linked with sudden death.[77] Cardiovascular abnormalities are common in patients with Marfan's syndrome with 30%–60% of pediatric patients with Marfan's syndrome having some cardiovascular anomaly. Sudden cardiac death from acute aortic dissection leading to aortic rupture is known to occur in patients with Marfan's syndrome.

Patients with a "structurally" normal heart who have an electrophysiological abnormality compromise the second group of patients at risk of sudden death. This group includes those with the long QT syndrome,[26-28] WPW syndrome,[18,19,21,23] primary ventricular tachycardia (catecholaminergic polymorphic ventricular tachycardia, exercise-related ventricular tachycardia),[30-32] Brugada syndrome,[33] short QT syndrome,[29] arrhythmogenic "right" ventricular cardiomyopathy/dysplasia,[34] and complete heart block.[19,21] The incidence of sudden death is reportedly as high as 9% for patients with long QT syndrome (mean five-year follow-up),[28] and 9.5% for catecholaminergic polymorphic ventricular tachycardia (seven-year follow-up).[30] Arrhythmogenic right ventricular dysplasia is a form of cardiomyopathy, which may be familial and is associated with recurrent ventricular arrhythmias and sudden death.

Risk Stratification of Syncope

In any age patient, some variables appear to be indicators of an underlying significant etiology for the syncope.[85] These include any of the following: any history of dysrhythmias, exertional syncope, congenital heart disease, CAD, CHF, associated symptoms (e.g. chest pain, dyspnea), systemic diseases which can affect the heart, a positive family history for: sudden death, acute coronary syndrome at an early age, or any physical examination findings of heart disease (congestive heart disease or congenital or acquired heart disease), and an abnormal ECG. *(See enclosed* Rapid Access Management *card for table, "History and Physical Examination Findings Suggestive of Serious Etiology of Syncope.")*

Large, prospective studies have not been done in infants and children. The consensus, however, is that a diagnosis of benign vasovagal syncope is less likely in the youngest patients (age ≤ 5 years) than in older pediatric patients, especially adolescents.[79] Breath-holding spells, seizures, or dysrhythmias should be in the differential diagnosis of syncope in infants and toddlers.[79]

Cost Effective Evaluation. In the majority of pediatric and adult syncope patients, the etiology is determined by the initial history and physical examination and ECG supplemented by a few basic ancillary studies; for example, Accu-Chek® or serum glucose, hematocrit, and serum pregnancy test in females in the childbearing age.[15,16] A routine CT scan in every patient with syncope is not indicated. It is not cost effective and exposes the patient to unnecessary radiation with the long-term risk of malignancy. Diagnostic testing should be focused and based on indicators in the history or physical examination as recommended by the American College of Emergency Physicians clinical policy on syncope.[48] The same principles seem logical for pediatric patients with syncope, although there is no pediatric-specific policy on syncope.

Summary

Syncope, the sudden loss of consciousness and postural tone with spontaneous recovery due to cerebral dysfunction, is common in childhood, occurring in up to half of all adolescents. Most etiologies of syncope are benign, although some causes are life-threatening and syncope can be the harbinger of sudden death. The history and physical examination and an ECG along with a focused diagnostic evaluation based on the initial history and physical examination will determine the etiology of the syncope in many patients.

References

1. Day SC, Cook EF, Funkestein H, et al. Evaluation and outcome of emergency room patients with transient loss of consciousness. *Am J Med* 1982;73:15-23.
2. Gendleman HE, Linzer M, Gabelman M, et al. Syncope in a general hospital patient population. *NY State J Med* 1983;1161-1165.
3. Kapoor WN, Karph M, Wieand S, et al. A prospective evaluation and follow-up of patients with syncope. *N Engl J Med* 1983;309:197-204.
4. Blanc JJ, L'Her C, Touiza A, et al. Prospective evaluation and outcome of patients admitted for syncope over a one-year period. *Eur Heart J* 2002;23: 815-820.
5. Owens TR. Sudden cardiac death: Is your patient at risk? *J Resp Dis* 1998;19: 384-396.
6. Driscoll DJ, Jacobsen SJ, Porter CJ, et al. Syncope in children and adolescents: A population based study of incidence and outcome. *Circulation* 1997;94:1-54.
7. Narchi H. The child who passes out. *Pediatr Rev* 2000;21:384-388.
8. Steinberg LA, Knilans TK. Syncope in children: Diagnostic tests have a high cost and low yield. *J Pediatr* 2005;146:355-358.
9. Miller TH, Kruse JE. Evaluation of syncope. *Am Fam Phys* 2005;72:1492-1500.
10. Stewart JM. Chronic orthostatic intolerance and the Postural Tachycardia Syndrome (POTS). *J Pediatr* 2004;145:725-730.
11. Morillo CA, Ellenbogen KA, Pava LF. Pathophysiologic basis for vasodepressor syncope. In: Klein GF (ed): *Syncope*. *Cardiol Clin* 1997;15:233-250.
12. Eagle KA, Black HR, Cook EF, et al. Evaluation of prognostic classifications for patients with syncope. *Am J Med* 1985;79:455-450.
13. Twidale N, Waddy H, Tonkin A. Diagnostic evaluation and outcome in patients presenting to a casualty department with syncope or presyncope. *Austral Clin Rev* 1986; 6:137-141.
14. Martin GJ, Adams SL, Martin HG, et al. Prospective evaluation of syncope. *Ann Emerg Med* 1984;13:499-504.
15. Pratt JL, Fleisher GR. Syncope in children and adolescents. *Pediatr Emerg Care* 1989;5:80-82.
16. Lerman-Sagie T, Lerman P, Mukamel M, et al. A prospective evaluation of pediatric patients with syncope. *Clin Pediatr* 1994;66-70.
17. Wren C. Cardiac causes for syncope or sudden death. *Arch Dis Child* 1999;81: 289-291.
18. Berger S, Utech L, Hazinski MF. Sudden death in children and adolescents. *Pediatr Clin N Am* 2004;51:1653-1677.
19. Silka MJ, Kron J, Walance CF, et al. Pediatric survivors of sudden cardiac death: Effective end-points of testing and therapy. *Circulation* 1990;82:341-349.
20. Alexander ME, Berul CI. Ventricular arrhythmias: When to worry. *Pediatr Cardiol* 2000;21:532-541.

21. Klitzner TS. Sudden cardiac death in children. *Circulation* 1990;82:629-632.
22. Driscoll DJ, Edwards WD. Sudden unexpected death in children and adolescents. *Am J Cardiol* 1985;5:118B-121B.
23. Wang Y, Scheinman MM, Cien WW, et al. Patients with supraventricular tachycardia presenting with aborted sudden death: Incidence, mechanism, and long-term followup. *JACC* 1991;18:1711-1719.
24. Maron BJ, Poliac LC, Roberts WO. Risk for sudden cardiac death associated with marathon running. *J Am Coll Cardiol* 1996;28:428-431.
25. Maron BJ, Epstein SE, Roberts WO. Causes of sudden death in competitive athletes. *J Am Coll Cardiol* 1986;7:204-214.
26. Moss AJ, Schwartz PJ, Crampton RS, et al. The long QT syndrome. Prospective longitudinal study of 328 families. *Circulation* 1991;84:1136-1144.
27. Khositseth A, Martinez MW, Driscoll DJ, et al. Syncope in children and adolescents and the congenital long QT syndrome. *Am J Cardiol* 2003;92:746-749.
28. Garson A, Dick M, Fournier A, et al. The long QT syndrome in children: An international study of 287 patients. *Circulation* 1992;87:1866-1872.
29. Giustetto C, DiMOnte F, Wolpert C, et al. Short QT syndrome: Clinical findings and diagnostic-therapeutic implications. *Eur Heart J* 2006; 27: 2440-2447.
30. Leenhardt A, Lucet V, Denjoy I, et al. Catecholaminergic polymorphic ventricular tachycardia in children. *Circulation* 1995;91:1512-1519.
31. DeRosa G, Delogu A, Piatra M, et al. Catecholaminergic polymorphic ventricular tachycardic: Successful emergency treatment with intravenous propranolol. *Ped Emerg Care* 2004;20:175-177.
32. Bricker JT, Traweek MS, Smith RT, et al. Exercise-related ventricular tachycardia in children. *Am Heart J* 1986;112:186-188.
33. Brugada J, Brugada R, Brugada P. Right bundle-branch block and ST-segment elevation in leads V1 through V3. *Circulation* 1998;97:457-460.
34. Buja G. Arrhythmogenic right ventricular cardiomyopathy/dysplasia: Risk stratification and therapy. *Prog Cardiovasc Dis* 2008;282-293.
35. Wang JN, Tsai YC, Chen SL, et al. Dangerous impact – commotion cordis. *Cardiology* 2000;93:124-126.
36. Tulloh R. Management and therapeutic options in pediatric pulmonary hypertension. *Expert Rev Cardiovascular Therapy* 2006;4(3):361-374.
37. Lombrosco CT, Lerman P. Breath-holding spells (cyanotic and pallid infantile syncope). *Pediatr* 1967; 39: 563-581.
38. DiMario FJ Jr. Prospective study of children with cyanotic and pallid breath-holding spells. *Pediatrics* 2001;107 265-269.
39. Breningstall GN. Breath-holding spells. *Pediatr Neurol* 1996;14:91-97.
40. Manolis AS. Evaluation of patients with syncope: Focus of age related differences. Part I – Etiologies. *Am Coll Cardiol Curr J Rev* 1994;3:13-16.
41. Grubb BP, Kosinski DJ. Syncope resulting from autonomic insufficiency syndromes associated with orthostatic intolerance. *Med Clin N Amer* 2001;85:457-472.
42. Kaufmann H. Syncope: A neurologist's viewpoint. In: Klein GJ (ed). *Syncope*. *Cardiology Clin* 1997;15:177-194.
43. Weiner LH, Williams O. Syncope and orthostatic intolerance. *Med Clin N Amer* 2003;87:835-865.
44. Grubb BP, Kosinski D. Dysautonomic and reflex syncope syndromes. *Card Clin* 1997;15:257-268.
45. Stewart JM. Orthostatic intolerance in pediatrics. *J Pediat* 2002;140:404-411.
46. Benditt DG, Lurie KG, Fabian WH. Clinical approach to diagnosis of syncope. *Cardiology Clin* 1997;15:165-176.
47. Sinert R, Spektor M. Clinical assessment of hypovolemia. *Ann Emerg Med* 2005;45:327-329.
48. Huff JS, Decker WW, Quinn JV, et al. Clinical policy: Critical issues in the evaluation and management of adult patients presenting to the emergency department with syncope. *Ann Emerg Med* 2007;49:431-444.
49. Dovgalyuk J, Holstege C, Mattu A, et al. The electrocardiogram in the patient with syncope. *Am J Emerg Med* 2007;25:688-701.
50. Hannon DW, Knilans TK. Syncope in children and adolescents. *Curr Probl Pediatr* 1993;358-384.
51. Gordon TA, Moodie DS, Passalacqua M, et al. A retrospective analysis of the cost-effective workup of syncope in children. *Cleve Clin Q* 1987;54:391-394.
52. Sapin SO. Autonomic syncope in pediatrics: A practice-oriented approach to classification, pathophysiology, diagnosis, and management. *Clin Pediatr* 2004; 43:17-23.
53. Schnipper JL, Kapoor WN. Diagnostic evaluation and management of patients with syncope. *Med Clin N Amer* 2001;85:423-452.
54. Hayes OU. Evaluation of syncope in the emergency department. *Emerg Med Clin N Amer* 1998;16:601-615.
55. Strickberger SA, Benson DW, Biaggioni I, et al. Alta/ACCF scientific statement on the evaluation of syncope. *J Am Coll Cardiol* 2006;47:473-484.
56. Sarasin FP, Louis-Simonet M, Carballo D, et al. Prospective evaluation of patients with syncope: A population based study. *Am J Med* 2001;111:177-184.
57. Garson A Jr. How to measure the QT interval – what is normal? *Am J Cardiol* 1993;72:14B-16B.
58. Graham TP, Gessner IH, Friedman WF, et al. Recommendations for use of laboratory studies for pediatric patients with suspected or proven heart disease. A statement of the Council on Cardiovascular Disease in the young of the American Heart Association. *Circulation* 1986;74:443A-450A.
59. Heaven DJ, Sutton R. Syncope. *Crit Care Med* 2000;28(10) (suppl):N116-N120.
60. Galetta F, Franzoni F, Roman Femia F, et al. Responses to tilt test in young and elderly patients with syncope of unknown origin. *Biomedicine Pharmacotherapy* 2004;58:443-446.
61. Alehan D, Celiker A, Ozme S. Head-up tilt test: A highly sensitive, specific test for children with unexplained syncope. *Pediatr Cardiol* 1996;17: 86-90.
62. Fuchs SM, Jaffe DM. Evaluation of the "tilt" test in children. *Ann Emerg Med* 1987;16:386-390.
63. Thilenius OG, Quinones JA, Husayni TS, et al. Tilt test for diagnosis of unexplained syncope in pediatric patients. *Pediatr* 1991;87:334-338.
64. Pongiglione G, Fish FA. Strasburger JF, et al. Heart rate and blood pressure response to upright tile in young patients with unexplained syncope. *J Am Coll Cardiol* 1990;16:165-170.
65. Ross BA, Hughes S, Anderson E, et al. Abnormal responses to orthostatic testing in children and adolescents with recurrent unexplained syncope. *Am Heart J* 1991;122:748-754.
66. Fouad-Tarazi F, Calcatti J, Christian R, et al. Blood volume measurements as a tool in diagnosing syncope. *Am J Med Sci* 2007; 334(1): 53-56
67. Chen DK, So YT, Fisher RS. Use of serum prolactin in diagnosing epileptic seizures. *Neurology* 2005;65:668-675.
68. Hoorntze TM, Langerak W. Munchausen's syndrome by proxy identified with an implantable electrocardiographic recorder. *NEJM* 1999;341:1478-1479.
69. Ozme S, Alehan D, Yalaz K, et al. Causes of syncope in children: A prospective study. *Int J Card* 1993;40:358-362.
70. Kilic A, Ozer S, Turanli G, et al. Dysrhythmia as a cause of syncope in children without neurological or cardiac morphological abnormalities. *Pediatrics International* 2002;44:358-362.
71. Morillo CA, Eckbergh DL, Ellenbogen KA, et al. Vagal and sympathetic mechanisms in patients with orthostatic vasovagal syncope. *Circulation* 1997;96: 2509-2513.

72. Kapoor WN, Karph M, Wieand S, et al. A prospective evaluation and follow-up of patients with syncope. *Circulation* 1983;309:197-204.
73. Ammirati F, Colivicchi F, Santini M. Diagnosing syncope in clinical practice. *Eur Heart J* 2000; 21: 935-940.
74. Silverstein MD, Singer DE, Mulley AG, et al. Patients with syncope admitted to medical intensive care units. *JAMA* 1982; 248(10): 1185-1189.
75. Soteriades ES, Evans JC, Larson MG, et al. Incidence and prognosis of syncope. *NEJM* 2002;347:878-885.
76. Lambert EC, Menon VA, Wagner HR, et al. Sudden unexpected death from cardiovascular disease in children. *Am J Cardiol* 1974;34:89-96.
77. Topaz O, Edwards JE. Pathologic features of sudden death in children, adolescents, and young adults. *Chest* 1985;87:476-482.
78. Campbell RM. The treatment of cardiac causes of sudden death, syncope, and seizure. *Semin Pediatr Neurol* 2005;12:59-66.
79. Quinn JV, Stiell IG, McDermott DA, et al. Derivation of the San Francisco syncope rule to predict patients with short-term serious outcomes. *Ann Emerg Med* 2004;43:224-232.

CME QUESTIONS

11. Which of the following statements is true regarding syncope?
    A. Syncope usually requires extensive diagnostic studies, such as head CT scan.
    B. Up to 50% of adolescents will have at least one syncopal episode.
    C. Syncope occurs in 50% of ED visits.
    D. Syncope is characterized by a prolonged loss of consciousness and a prolonged recovery phase.
    E. Syncope is characterized by post syncope symptoms such as headache or sleepiness.

12. Which of the following is recommended by many experts in the evaluation of all syncope patients, regardless of age?
    A. Renal function tests (BUN, creatinine)
    B. Liver function tests
    C. ECG
    D. CT scan of head
    E. Serum ammonia

13. The most common cause of syncope in pediatric patients is:
    A. Cardiac causes
    B. Vasovagal causes
    C. Breath-holding spell
    D. POTS syndrome
    E. Pulmonary emboli

14. Which of the following statements is true regarding breath-holding spells?
    A. They can be life-threatening.
    B. Infants/children with breath-holding spells should be admitted.
    C. They occur most commonly in school-age children (ages 5-10 years).
    D. They may be due to an exaggerated sympathetic response causing tachycardia.
    E. The two types are cyanotic and pallid.

15. Which of the following statements is true of syncope?
    A. There are very few conditions that are confused with syncope.
    B. Seizures are often mistaken for syncopal events, and vice versa.
    C. Hypoglycemia is not in the differential diagnosis for syncope.
    D. Syncope is rarely confused with other disorders such as seizures.
    E. Respiratory disorders/conditions never cause syncope.

16. Which is statement is correct regarding vasovagal syncope?
    A. A parasympathetic mediated reflex causes tachycardia.
    B. A sympathetic mediated reflex causes bradycardia or even asystole.
    C. Increased sympathetic input causes increased peripheral vascular resistance (PVR) and hypotension.
    D. Withdrawal of sympathetic input decreases PVR with venous pooling in extremities and hypotension.
    E. Vasovagal syncope does not involve the autonomic nervous system or the central nervous system.

17. Which of the following is correct regarding vasovagal syncope?
    A. Vasovagal syncope does not occur in heart transplant patients.
    B. Vasovagal syncope may also involve local vascular control, or be mediated by hormones or abnormal CNS regulation.
    C. Vasovagal syncope does not involve the autonomic nervous system.
    D. Vasovagal syncope is the result of increased heart rate and increased stroke volume.
    E. Cerebral perfusion is maintained during vasovagal syncope.

18. Which of the following is correct regarding the physiologic changes occurring with assuming the upright posture?
    A. Blood pools in the lower extremities.
    B. Central venous volume increases.
    C. There is an increase in stimulation of the carotid/aortic arch baroreceptors.
    D. There is an increase in neural discharges to the medulla oblongata.
    E. Activation of the vagus increases peripheral vascular resistance.

19. Which of the following is correct regarding postural orthostatic tachycardia syndrome (POTS)?
    A. Patients become symptomatic when they lie down.
    B. POTS usually occurs in geriatric patients.
    C. Symptoms of POTS include dizziness, lightheadedness, blurred vision, and anxiety.
    D. POTS is thought to be due to increased parasympathetic (vagal) activity.
    E. There are no suggested treatments.

20. Which of the following is correct regarding the dysautonomic syndromes?
    A. They are never due to disorders affecting the central nervous system.
    B. They do not involve neurally mediated reflexes.
    C. They are most commonly due to primary autonomic disease.
    D. They may be due to peripheral forms of secondary autonomic failure such as diabetes, AIDS, or paraneoplastic disease.
    E. The most common causes are from acute central nervous system disorders.

Answers: 11. B, 12. C, 13. B, 14. E, 15. B, 16. D, 17. B, 18. A, 19. C, 20. D

In Future Issues:

Influenza Update

PEDIATRIC Emergency Medicine Reports
The Practical Journal of Pediatric Emergency Medicine

Pediatric Syncope

Etiology of Syncope in the Pediatric Patient

CARDIOVASCULAR: CONGENITAL OR ACQUIRED

Cardiac

- Dysrhythmias
- Myocardial disorders
- Valvular disease
- Pericardial disease
- Congenital heart disease
- Pulmonary hypertension
- Cardiac tumors (primary or metastatic): Atrial myxoma, rhabdomyosarcoma

Vascular

- Aortic disease: Severe coarctation, aortic dissection
- Coronary artery disease / disorders
- Disease / disorders of great vessels (other than aorta)
  - Subclavian steal
  - Thoracic outlet

NEUROLOGIC

- Central nervous system disorder
  - Hemorrhage
  - Stroke, transient ischemic attack
  - Migraine headaches
- Peripheral nervous system
  - Neuropathies

RESPIRATORY

- Hypoxia from respiratory failure or pulmonary disease
- Pulmonary emboli (can cause syncope by mechanism other than hypoxia)
- Cough (post-tussive)
- Breath-holding: Cyanotic, pallid, mixed (likely from autonomic dysregulation)

Disorder of Oxygen Transport

- Carbon monoxide poisoning
- Cyanide poisoning
- Other toxins
- Severe anemia

METABOLIC / ENDOCRINE

- Hypoglycemia

DRUGS / DRUG OVERDOSE

- Poisons/toxins *(see also* Disorders of Oxygen Transport, *above)*
- Drugs of abuse: Cocaine, opiates, alcohol, others
- Prescription drug abuse (intentional): Benzodiazepine, opioids, others
- Prescription drugs (unintentional): Antihypertensives, cardiac especially vasodilators (calcium channel blockers, beta blockers, nitrates), psychiatric drugs especially tricyclics, phenothiazines
- Nonprescription
- Over-the-counter drugs/herbals/vitamins

HYPOVOLEMIC / HEMORRHAGIC

- Decreased intravascular volume (from blood or volume loss)

POSTURAL (ORTHOSTATIC)

- Postural orthostatic tachycardia syndrome (POTS)
- Dysautonomic syndromes: Primary or secondary autonomic failure

NEUROCARDIOGENIC

- Vasovagal
- Situational
- Carotid sinus syncope

PSYCHIATRIC

- Anorexia nervosa, bulimia
- Hyperventilation (anxiety, panic disorders)

History and Physical Examination Findings Suggestive of Serious Etiology of Syncope

HISTORY	BENIGN	SERIOUS
Position	Occurs with change in position (from lying to sitting or standing, or sitting to standing) (implies orthostatic syncope)	Unrelated to position (occurs while recumbent or sitting)
Exercise	Not related to exercise	Occurs during exercise or exertion (occurs when CO fails to meet increased demands), ↑CO needed with exercise
Onset	Gradual	Rapid (suggests dysrhythmias)
Prodrome	With prodrome	None (suggests dysrhythmia with sudden onset)
Isolated vs. recurrent	Isolated single event	Recurrent over a short time frame (usually hours, days, or few weeks)
Injury: Secondary to syncope	No injury from syncope (no injuries occur with psychogenic syncope)	Injury (bruises, lacerations, fractures, etc.) from syncope
Associated symptoms: CV	No associated CV symptoms	Chest pain, shortness of breath, palpitations
Associated signs: Skin	No cyanosis, no pallor; or, short-lived, mild (transient) pallor	Severe pallor or cyanosis

FAMILY HISTORY	BENIGN	SERIOUS
Sudden death	No	Yes
Myocardial Infarction at an early age	No	Yes
Cardiomyopathy	No	Yes
Neuromuscular disorders	No	Yes
Congenital deafness	No	Yes (Consider Jervett-Lange-Nielsen Syndrome)
Marfan syndrome	No	Yes (Consider aortic dissection)
Autoimmune disease (maternal lupus)	No	Yes (Complete AV block in infant from maternal lupus)

PAST MEDICAL HISTORY	BENIGN	SERIOUS
Prior cardiac surgery	No	Yes (damage to conduction system may occur months, even years, later)
History of congestive heart failure	No	Yes (may have poor cardiac output and / or dysrhythmias)
Congenital heart disease	No	Yes
Acquired heart disease	No	Yes

VITAL SIGNS	BENIGN	SERIOUS
Pulse/heart rate Tachycardia or bradycardia	No	Yes
Regular	Yes	No
Respirations: Bradypnea	No	Yes (Consider serious CNS or respiratory disease)
Respirations: Tachypnea	Usually no, occasionally yes from hyperventilation secondary to pain / anxiety	Yes (Consider respiratory disease)
Blood pressure (low)	No	Yes (Consider hypovolemia or hemorrhage)
Blood pressure (high)	No	Yes (Consider hypertensive crisis / encephalopathy)
Positive orthostatic vital signs	No	Yes (Consider hypovolemia, hemorrhage, autonomic nervous system disorders)

PHYSICAL EXAMINATION	BENIGN	SERIOUS
General appearance	Normal	Abnormal; Unusual facies (may have syndrome with CV disease, such as Down's syndrome, Williams disease, Marfan syndrome)
Respiratory	Normal	Abnormal; Rales, wheezing suggest underlying CV or pulmonary disease
Mental status	Normal	Abnormal; may have inadequate CNS perfusion or CNS disease
Neurologic	Normal	Abnormal; especially focal abnormalities, suggest CNS disease / injury
Neck exam	Normal	JVD suggests CHF
Musculoskeletal	Normal	Abnormal; weakness, decreased strength, tone, or muscle mass imply neuromuscular disorder
Extremity	Normal	Abnormal; calf pain (consider deep vein thrombosis), edema (Is there CHF or other systemic disease?)
Dermatologic	Normal	Abnormal; decreased turgor / tenting or other signs of inherited disease; café-au-lait, von Recklinghausen's disease

Key: CO = cardiac output; CV = cardiovascular; CNS = central nervous system; JVD = jugular venous distention

Differential Diagnosis of Syncope in Pediatric Patients

NEUROLOGIC			CLINICAL FEATURES
Breath-holding Spells* — Three Categories			
	Incidence		
Cyanotic	52%–62%	Hold breath in expiration → apnea / cyanosis	Age 6–24 months, resolved by age 4–5 years, + family history 20%–35%, Key: inciting event: crying / emotional upset (cyanotic) or pain, fall, hit head (pallid) → loss of tone/apnea, ± seizure, ± posturing
Pallid	19%–28%	Apnea → pallor	Normal EEG, short time frame; common cause of infant syncope, if severe can cause LOC, seizure, posturing
Mixed or *unclassified*	19%–20%		Etiology: Autonomic dysregulation, treatment of iron deficiency anemia
Apnea*			Common, especially in premature infants, due to brain stem immaturity, resolves with age
Hyperekplexia* ("Stiff baby syndrome," "Startle disease")			Stiffness when awake, nocturnal myoclonus, exaggerated startle reflex, toddler sudden falls in response to surprise / stimuli / stress / emotion; rare genetic disease
Seizures			Postictal period, aura, bladder / bowel incontinence, automatisms
Migraine (Basilar)			Visual symptoms, aura, headache
Vertigo			No LOC, dizziness, or spinning sensation
Transient ischemia attack			Neurologic symptoms (weakness, aphasia, etc.) that resolve
Sleep disorders: Cataplexy, narcoplexy			
VASCULAR			
Aortic dissection			BP and / or pulse differences between arms
Subclavian steal			BP and / or pulse differences between arms, symptoms with arm exercise or arm movements
RESPIRATORY			
Hyperventilation			History of tachypnea ± paresthesias, ± carpopedal spasm
Pulmonary emboli			May cause syncope; Symptoms: dyspnea, chest pain; Diagnosis: spiral CT chest or VQ scan
Pulmonary hypertension			May cause syncope / sudden death, loud S2, ECG ± RVH; Symptoms: SOB, DOE, exercise tolerance; Diagnosis: echocardiogram
METABOLIC			
Hypoglycemia			May cause syncope; Associated symptoms: diaphoresis, ± history of DM / glucose disorders, ↓ oral intake, alcohol ingestion (especially in infants/young children); Diagnosis: check glucose
PSYCHIATRIC			
Hysteria (Conversion disorder) Factitious disorders: Malingering, Munchausen's syndrome			No associated neurologic / cardiovascular changes, no injury occurs, patient may describe event thus, no LOC, may have secondary gain / audience
Panic disorder / anxiety			Hyperventilation

* = Unique to pediatric population

Key: ECG = electrocardiogram; EEG = electroencephalogram; LOC = loss of consciousness; BP = blood pressure; CT = CAT scan; VQ = ventilation perfusion scan; RVH = right ventricular hypertrophy; SOB = shortness of breath; DOE = dyspnea on exertion; DM = diabetes mellitus

Cardiovascular Causes of Pediatric Syncope

ARRHYTHMIAS: CONGENITAL OR ACQUIRED

- **Tachyarrhythmias**
  - o Wolff-Parkinson-White syndrome
  - o Ventricular tachycardia
  - o Ventricular fibrillation
  - o Arrhythmogenic right ventricular dysplasia
  - o Torsades de pointes
- **Device Malfunction**
  - o Pacemaker malfunction
  - o AICD malfunction
- **Specific Congenital Heart Defects**
  - o High-risk patients: Ebstein anomaly, tetralogy of Fallot, others
- **Post-operative Congenital Heart Disease**
- **QT Abnormalities**
  - o Long QT syndromes: Congenital Romano-Ward syndrome, Jervell and Lange-Nielsen syndrome
  - o Acquired: Medications causing QT prolongation: Psychotropics (tricyclic antidepressants, phenothiazines, promotility drugs [cisapride] — especially in combination with other drugs [e.g., erythromycin, ketoconazole])
  - o Short QT syndrome

STRUCTURAL DISORDERS: MYOCARDIAL DYSFUNCTION / DISEASE

- **Primary**
  - o Myocarditis
  - o Dilated cardiomyopathy
  - o Idiopathic hypertrophic subaortic stenosis (IHSS)
  - o Arrhythmogenic right ventricular dysplasia (*see also* Arrhythmias)
- **Secondary (Acquired)**
  - o Infections:
    - – *Viral:* Coxsackie, others
    - – *Parasitic:* Chagas disease
  - o Immunologic, vasculitis, rheumatologic diseases: Lyme disease, amyloidosis, sarcoidosis
  - o Generalized neuromuscular diseases: Muscular dystrophy

CORONARY ARTERY DISEASE

- **Congenital:** Anomalous coronary artery
- **Acquired:** Kawasaki disease

VALVULAR

- **Severe / Critical Aortic Stenosis**
- **Severe / Critical / Mitral / Pulmonic / Tricuspid Valve Disorder** (stenosis or regurgitation)
- **Prosthetic Valve Dysfunction**

VASCULAR

- **Aorta:** *Aortic dissection*, secondary to hypertension, atherosclerosis, connective tissue disorders (Marfan disease, Ehlers-Danlos syndrome)
- **Other Great Vessel Abnormalities**
  - o Subclavian steal syndrome
  - o Thoracic outlet syndrome

OUTFLOW OBSTRUCTION TO SYSTEMIC BLOOD FLOW

- **Severe Coarctation**
- **Cardiac Tumor Mass** (atrial myxoma)
- *Also*: **Hypertrophic Obstructive Cardiomyopathy, Critical Aortic Stenosis**

PERICARDIAL DISEASE

- **Pericarditis / Pericardial Tamponade**

CONGENITAL HEART DEFECTS

- **Cyanotic / Acyanotic Congenital Heart Disease**
- **Eisenmenger Syndrome**

PULMONARY HYPERTENSION

- **Primary** (idiopathic)
- **Secondary**

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