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. Antiepileptic 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 paradoxic 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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PIR QUIZ

Quiz also available online at www.pedsinreview.org.

9. 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.

10. You are evaluating a 13-year-old girl who fainted at school. Which of the following histories would raise your suspicion *most* for a vasovagal cause for the syncope?
   A. She fainted while playing soccer.
   B. She felt a bitemporal headache shortly before the episode.
   C. She was unconscious for approximately 5 minutes.
   D. The episode was followed by a prolonged period of disorientation and sleepiness.
   E. The episode was precipitated by a frightening event.

11. A patient presents to the emergency department after fainting at school. An eyewitness reports that the patient arose from her chair, started to walk across the room, and suddenly fell forward. Her arms and legs jerked for a few seconds, but then she became conscious and alert. The patient reports feeling a little dizzy before falling. Which of the following evaluations would be *most* indicated?
   A. Computed tomography of the brain.
   B. Echocardiography.
   C. Electrocardiography.
   D. Orthostatic blood pressure measurement.
   E. Tilt table testing.

12. A 13-year-old boy reports to your office after fainting at football practice. He was unconscious for 3 to 4 minutes, during which time jerking movements of the legs were noted. Afterwards, he was drowsy and disoriented for several minutes. He has no memory of the event. Family history is pertinent for a grandmother who died of heart disease at age 80. Of the following, the *most* likely cause of the episode is:
   A. Complicated migraine.
   B. Epilepsy.
   C. Hypoglycemia.
   D. Narcolepsy.
   E. Prolonged Q-T syndrome.

13. Which of the following statements about neurocardiac syncope is *true*?
   A. A history of breathholding spells early in childhood is common.
   B. A positive tilt table test is required for diagnosis.
   C. It is considered to be a manipulative behavior.
   D. It is the least common cause of syncope.
   E. Medication usually is required for adequate management.

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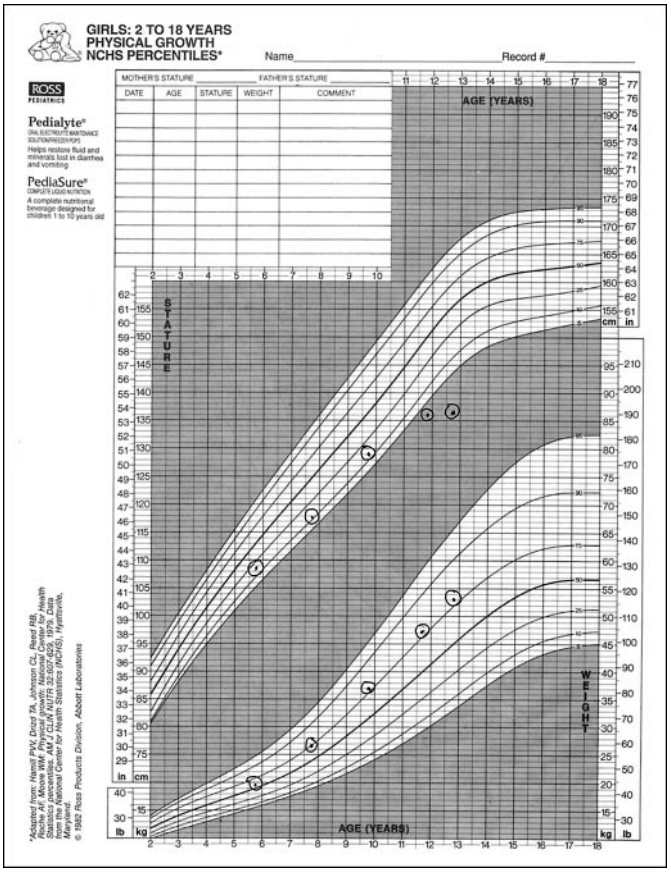

FIGURE 2. Growth chart showing decrease in height growth that was caused by kyphosis.

palliative. Although there was no evidence of metastasis, 3 months later there had been remarkable interval growth of the tumor. Radiation was employed in an attempt to delay further surgery. As with this patient, myxopapillary ependymomas tend to require multiple operations and later in life can lead to paraplegia; they also have the potential to metastasize. Chemotherapy has not been of significant benefit.

THE SPECTRUM OF LESIONS

Spinal tumors are classified as either extramedullary or intramedullary. The patterns of symptoms and signs associated with these two kinds of tumors differ (Table). Of the intraspinal tumors, the most common are gliomas (ependymoma, astrocytoma, and medulloblastoma), followed by neuroblastomas, extradural sarcomas, dermoid cysts, teratomas, lipomas, and intramedullary cysts. Ependymomas account for almost 25% of spinal tumors and can be described further as well-differentiated cellular or well-differentiated myxopapillary tumors.

Intraspinal tumors are rare and occur only 20% as often as intracranial tumors. The initial symptoms usually are manifested as musculoskeletal complaints, and the paucity of symptoms can make diagnosis difficult. The most common presenting symptoms include weakness of the legs, back pain, torticollis, urinary incontinence, abdominal pain, weakness or pain in an extremity, rectal sphincter laxity, and sensory disturbances. The back pain is persistent and often hard for the patient to describe accurately. It is accentuated by physical activity, such as sneezing, coughing, straining, neck flexing, and straight leg raising. Slow-growing intraspinal tumors can be present for long periods of time without causing bone destruction, which reduces the effectiveness of plain radiographs in detecting them.

LESSON FOR THE CLINICIAN

The misleading clue in this case was the back injury, which allowed the parents to explain her pain and delay pursuing further medical evaluation. The pain was so long-standing that it became a way of life for this family, thus minimizing their reaction. Pain out of proportion to the severity of an injury always must heighten the clinician's suspicion of an underlying disorder. Amid all the chronic complaints of pain seen in the office, the occult serious disorder presents a never-ending challenge. *(Sharisse M. Arnold, MD, Harvard University Medical School, Children's Hospital, Boston, MA)*

DEPARTMENT OF CORRECTIONS

Erratum

The correct answer to PIR Quiz Question #9 in the June issue is B. The answer A that is listed in the Answer Key is incorrect.