AHA/ACCF Scientific Statement

AHA/ACCF Scientific Statement on the Evaluation of Syncope

From the American Heart Association Councils on Clinical Cardiology, Cardiovascular Nursing, Cardiovascular Disease in the Young, and Stroke, and the Quality of Care and Outcomes Research Interdisciplinary Working Group; and the American College of Cardiology Foundation

In Collaboration With the Heart Rhythm Society

Endorsed by the American Autonomic Society

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Syncope, a transient loss of consciousness, is a common clinical problem. The most common causes of syncope are cardiovascular in origin and are associated with a high rate of mortality in patients with underlying heart disease, transient myocardial ischemia, and other less common cardiac abnormalities.(1)

The primary purpose of the evaluation of the patient with syncope is to determine whether the patient is at increased risk for death. This involves identifying patients with underlying heart disease, myocardial ischemia, Wolff-Parkinson-White syndrome, and potentially life-threatening genetic diseases such as long-QT syndrome (LQTS), Brugada syndrome, and catecholaminergic polymorphic ventricular tachycardia. If these diagnoses can be excluded, the goal then becomes identification of the cause of syncope in an attempt to improve the quality of the patient's life and to prevent injury to the patient or others. The purpose of this statement is to summarize the data that direct the evaluation of the patient with syncope (Figure 1).

General Evaluation

In the general population, the most common cause of syncope is neurocardiogenic, followed by primary arrhythmias. Other

Syncope

↓

History, Physical Examination, Electrocardiogram

↓

Diagnostic for orthostatic hypotension or neurocardiogenic syncope

Unexplained Syncope

↓

Echocardiogram, exercise test, and ischemia evaluation

↓

If found, treat for structural heart disease and ischemia. For arrhythmia evaluation, consider electrophysiological testing if there is a history of a myocardial infarction. Consider implantable defibrillator if the left ventricular ejection fraction is ≤ 0.30, with or without a history of a myocardial infarction.

Normal

↓

Single, benign episode | Frequent episodes | Infrequent episodes

Evaluation complete | Correlate symptoms with rhythm with Holter or event monitor, or implantable loop recorder, as appropriate. | Implantable loop recorder

Sinus rhythm with symptoms | Arrhythmia with symptoms

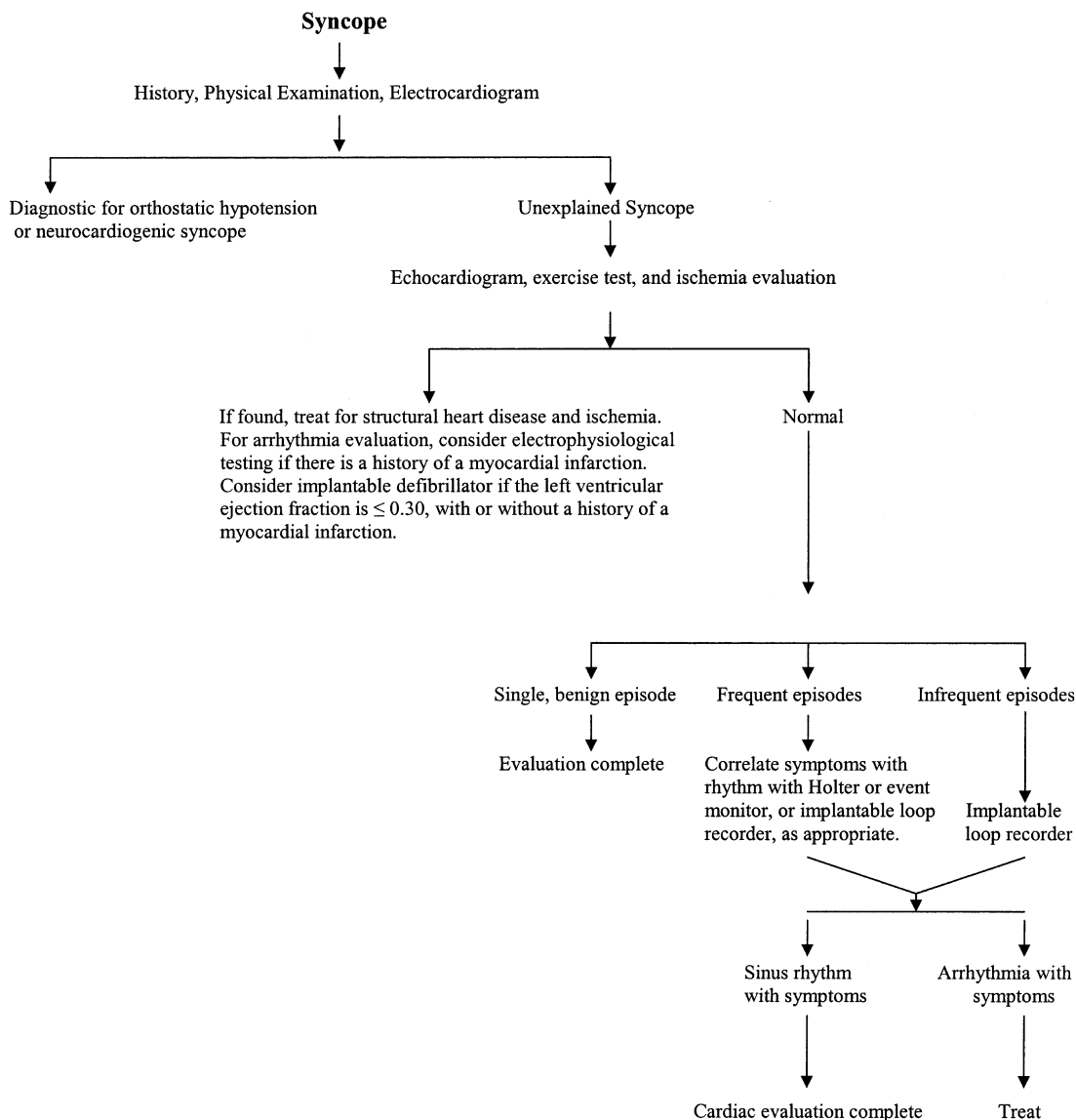Cardiac evaluation complete | Treat

Figure 1. Flow chart for the diagnostic approach to the patient with syncope.

names for neurocardiogenic syncope include neurally mediated, vasodepressor, and vasovagal syncope. The causes of syncope are highly age dependent.(2) Pediatric and young patients are most likely to have neurocardiogenic syncope, conversion reactions (psychiatric causes), and primary arrhythmic causes such as the LQTS and Wolff-Parkinson-White syndrome. In middle age, neurocardiogenic syncope remains the most frequent cause of syncope. Syncope related to other forms of neurocardiogenic syncope such as deglutition, micturition, defecation, and cough, as well as orthostasis and panic disorders, is more common in the middle-aged or elderly patients than in younger patients. In contrast to younger patients, elderly patients have a higher frequency of syncope caused by obstructions to cardiac output, eg, aortic stenosis and pulmonary embolus, and arrhythmias resulting from underlying heart disease.

History and Physical Examination

The approach to the patient with syncope begins with a meticulous history.(1,3,4) In most patients, the cause of

syncope can be determined with great accuracy from a careful history and physical examination, although the mechanism of syncope remains unexplained in 40% of episodes.(3) The process of evaluating the history, physical examination, and ECG should also include assessment of the medication list for agents associated with proarrhythmia, eg, Class IA and IC antiarrhythmic drugs.

Various aspects of the history help to establish the diagnosis. The observations of onlookers are important. The occurrence of tonic-clonic, seizurelike activity is associated with both cardiac and neurological causes of syncope.(5) Episodes of neurocardiogenic syncope are typically associated with postepisode fatigue or weakness, whereas the absence of a prodrome is consistent with cardiac arrhythmia or, less commonly, a central neurodegenerative disorder with autonomic failure such as Parkinson disease.(6) Auras, premonitions, postictal confusion, and focal neurological signs and symptoms suggest a neurological cause. Transient ischemic attacks rarely result in syncope. However, patients with

basilar artery or severe bilateral carotid artery disease may have syncope that usually is associated with focal neurological symptoms. A history of myocardial infarction with or without left ventricular dysfunction or repaired congenital heart disease raises the possibility of ventricular arrhythmias. A history of head trauma in a younger person without underlying heart disease may suggest a neurological origin, whereas syncope precipitated by neck turning, particularly in the elderly, raises the possibility of carotid sinus hypersensitivity. This can be assessed at the bedside with carotid sinus massage in patients in the supine and/or upright positions. However, this test should not be performed in patients with a recent transient ischemic attack or stroke, or ipsilateral to significant carotid artery stenosis or carotid artery bruit. The patient should be asked specifically if there is a positive family history for unexpected sudden cardiac death.

The history is also useful for identifying aggravating and alleviating factors. For example, the addition of a new drug, especially an antiarrhythmic or antihypertensive agent, raises the possibilities of proarrhythmia and orthostasis, respectively. In the elderly, phenothiazine and tricyclic drugs predispose to orthostasis. The possible role of over-the-counter medications and supplements, eg, ephedra-containing preparations, should be addressed.

During the evaluation of syncope, a careful physical examination is second only to the history. Orthostatic hypotension, autonomic dysfunction, and sometimes organic heart disease can be identified by measuring blood pressure and pulse rate in the upper and lower extremities and in the supine and upright positions. Carotid bruits raise the question of impaired cerebral blood flow and underlying coronary artery disease. The physical examination can also suggest the presence of pulmonary hypertension, left ventricular dysfunction, valvular heart disease, or other forms of organic heart disease. Abnormalities of cognition and speech, visual fields, motor strength, sensation, tremor, and gait disturbance suggest an underlying neurological disorder.

ECG, Echocardiogram, and Ischemia Evaluation

The ECG provides important information about the rhythm and atrioventricular (AV) conduction. Sinus bradycardia, a prolonged PR interval, or bundle-branch block raises the possibility of symptomatic sick sinus syndrome or intermittent complete AV block. Examination of the QRS complex may identify the presence of a delta wave, signifying the presence of an accessory pathway and Wolff-Parkinson-White syndrome. Genetic diseases of cardiac channels such as the LQTS and Brugada syndrome can be identified on the surface ECG and can cause syncope and life-threatening ventricular arrhythmias.(7) The ECG may suggest arrhythmogenic right ventricular dysplasia/cardiomyopathy (ARVD/C), in which case a cardiac MRI may aid in establishing the diagnosis. Ventricular ectopy or nonsustained ventricular tachycardia in a patient with underlying heart disease raises the possibility of an arrhythmic origin of syncope.

An echocardiogram is a helpful screening test if the history, physical examination, and ECG do not provide a diagnosis or if underlying heart disease is suspected. The echocardiogram is an excellent way to identify underlying heart disease, including valvular disease. It can also suggest pulmonary embolism if pulmonary hypertension or right ventricular enlargement is present. The most common cause of sudden death in athletes is hypertrophic cardiomyopathy, which is readily indicated by echocardiography.(8) The second most common cause of sudden death in the young is the presence of an anomalous coronary artery.(8) In young and thin individuals, the coronary ostia may be identified by a transthoracic echocardiogram, and if not visualized, the presence of an anomalous coronary artery may be further evaluated with a transesophageal echocardiogram, cardiac MRI or CT, or other imaging modality.

An evaluation for ischemia is appropriate in patients at risk for or with a history of coronary artery disease. Exercise testing should be performed in the patient with unexplained syncope, especially if the episode was exercise related. Exercise testing provides the opportunity to monitor pulse and blood pressure. In patients less than 40 years of age, a drop in blood pressure or failure of blood pressure to rise with exercise raises the question of hypertrophic obstructive cardiomyopathy or left main coronary artery disease(9); in the elderly patient, it may be a manifestation of autonomic failure. Exercise testing also screens for catecholaminergic polymorphic ventricular tachycardia.

Syncope in the Patient With a Normal Evaluation

In the absence of underlying heart disease, syncope is not associated with excess mortality. The main risk is related to physical harm that may occur if the patient has recurrent syncope. In this setting, the intensity of the workup to establish a diagnosis is determined by the "malignancy" of the episode. For the purposes of this statement, a malignant episode of syncope is defined as an episode of syncope that occurs with little or no warning and results in a significant injury or property damage, eg, a car accident.

When the general workup is normal, the origin of syncope can be extremely challenging to ascertain. Although many life-threatening clinical entities are less likely in the presence of a normal evaluation, the possibility of neurocardiogenic syncope, carotid sinus hypersensitivity, paroxysmal bradyarrhythmias, supraventricular tachycardia, ventricular tachycardia, and myriad noncardiac causes of syncope remains.(10) The gold standard for diagnosing an arrhythmic cause of syncope is ECG documentation of the rhythm disturbance at the time of symptoms. However, it is rare to have documentation of the cardiac rhythm from the initial episode of syncope.

Noninvasive ECG Monitoring

The type and duration of ambulatory ECG monitoring is dictated by the frequency of symptoms. A Holter monitor is appropriate for episodes that occur at least every day. Event monitoring is ideal for episodes that occur at least once a month. An implantable loop monitor allows the correlation of symptoms with the cardiac rhythm in patients in whom the symptoms are infrequent.

Traditionally, ambulatory monitoring is carried out for 24 to 48 hours with a Holter monitor. The short duration of the recording limits the diagnostic yield. Event recorders allow

ambulatory monitoring for 30 to 60 days. Patient triggering stores the ECG from 1 to 4 minutes before and 30 to 60 seconds after activation. The major limitation of the device is the complexity of its use, which results in patient errors with acquisition and transmission of data.(11) The introduction of continuously recording monitors that have both patient-activated and automatic triggers appears to improve the diagnostic yield of event monitors.(12)

Implantable loop recorders inserted subcutaneously are capable of recording bipolar ECG signals for approximately 14 months. The patient may use an activator to record the rhythm at the time of symptoms, and the device automatically records bradycardia and tachycardia. In patients with unexplained syncope, use of an implantable loop recorder for 1 year yielded diagnostic information in more than 90% of patients.(12) This approach is more likely to identify the mechanism of syncope than is a conventional approach that uses Holter or event monitors and electrophysiological testing and is cost-effective.(13)

Analysis of T-wave alternans with signal-averaged ECG and assessment of heart rate variability are noninvasive methods for identifying patients at increased risk for sudden cardiac death. The routine use of these tools in patients with syncope and a negative initial evaluation is not established and currently is not indicated.

Tilt Table Test

Tilt table testing is used as an aid in establishing the diagnosis of neurocardiogenic syncope. However, serious questions about the sensitivity, specificity, diagnostic yield, and day-to-day reproducibility of tilt table testing exist.(14–17) The reported sensitivity and specificity of tilt table testing depend on the technique used. The sensitivity ranges from 26% to 80%, and the specificity is approximately 90%. In patients with a negative evaluation, ie, no evidence of ischemia and a structurally normal heart, the pretest probability that the diagnosis is neurocardiogenic syncope is high, so head-up tilt table testing contributes little to establishing the diagnosis. For example, in a patient of any age with an otherwise normal evaluation who has a negative tilt table test, the most likely diagnosis is still neurocardiogenic syncope. In patients with a malignant episode of syncope, it may be more important to rule out other causes of syncope such as bradyarrhythmias, supraventricular tachycardia, and ventricular tachycardia than it is to perform a tilt table test. This is especially true because the risk for recurrent syncope in the patient with a normal cardiac evaluation and syncope is similar in patients with a positive or negative tilt table test.

Electrophysiological Study

Electrophysiological testing involves placement of transvenous catheters within the heart to assess sinus node function, AV conduction, and susceptibility to supraventricular and ventricular tachycardias. In patients with a normal evaluation for syncope, the yield of electrophysiological testing is approximately 3%.(14) The sensitivity of electrophysiological testing for the detection of bradyarrhythmias is low.(15) Because of the low yield of electrophysiological testing in patients without underlying heart disease, this test is not routinely recommended.(16,17) However, given the low

risk of electrophysiological testing(18) and the high risk of recurrent syncope with potential harm to the patient, the risk-to-benefit ratio may favor electrophysiological testing in patients with a malignant episode of syncope.

Syncope in the Patient With Coronary Artery Disease

The risk of death in a patient with syncope and coronary artery disease is directly proportional to the severity of left ventricular dysfunction. In the patient with coronary artery disease, recurrent episodes of ischemia and arrhythmia diagnoses such as bradycardia and ventricular tachycardia should be considered.

The major goal of the evaluation of syncope in the patient with coronary artery disease is to identify a potentially life-threatening diagnosis. This necessitates an evaluation for ischemia, underlying heart disease, and arrhythmias. If these patients require revascularization, the arrhythmia evaluation is still needed because, when present, the substrate for ventricular tachycardia and lethal ventricular arrhythmias is not ameliorated by revascularization. However, ventricular tachycardia or ventricular fibrillation that occurs in the setting of an acute ST-segment–elevation myocardial infarction may not require a specific arrhythmia evaluation, particularly if the left ventricular ejection fraction is well preserved.

Electrophysiological Study

After an ischemia evaluation, the patient with coronary artery disease and syncope should undergo electrophysiological testing. Electrophysiological testing is a useful method to assess sinus node function and AV conduction; however, it is a much better test for identifying the presence of ventricular tachyarrhythmias and the risk of death in the patient with coronary artery disease and syncope. Independent of the left ventricular ejection fraction, patients with syncope, coronary artery disease, and inducible monomorphic ventricular tachycardia during electrophysiological testing should be treated with an implantable defibrillator.

Among patients with coronary artery disease, syncope, and mild-to-moderate left ventricular dysfunction, ie, left ventricular ejection fraction >0.35, inducible ventricular tachycardia during electrophysiological testing is relatively unlikely. Despite the low yield, electrophysiological testing is appropriate in this setting because of the significant implications of inducible ventricular tachycardia. After a negative electrophysiological test in this patient population, the evaluation is usually complete. However, when a definitive diagnosis is required, then an event monitor or an implantable loop monitor can be used.

In the interest of finding a specific diagnosis, the patient with coronary artery disease and syncope may undergo a diagnostic electrophysiological test if the left ventricular ejection fraction is <0.35. However, even in the absence of syncope, the patient with coronary artery disease and a left ventricular ejection fraction <0.35 has a substantial survival benefit when treated with an implantable defibrillator.(19,20) Therefore, the patient with syncope and severe ischemic cardiomyopathy is an appropriate candidate for an implantable defibrillator regardless of the result of electrophysiolog-

ical testing.(19–21) Comprehensive recommendations for implantable defibrillator therapy can be found in the ACC/ AHA/NASPE 2002 guideline update for implantation of cardiac pacemakers and antiarrhythmia devices.(21)

Nonischemic Dilated Cardiomyopathy

Syncope is associated with increased mortality among patients with nonischemic dilated cardiomyopathy (NIDCM).(22) The likely reason for the increased mortality is that episodes of syncope in these patients often are caused by self-terminating episodes of ventricular tachycardia that, if recurring, can lead to cardiac arrest. The frequency of syncope in this group of patients is poorly defined. In the US Carvedilol study, which evaluated patients with refractory heart failure from any origin, 33% of patients had dizziness, but only 0.3% of patients had syncope over a 6-month period.(23)

The differential diagnosis of syncope in patients with NIDCM includes bradycardia, tachycardia, orthostatic hypotension, and pulmonary embolism. Although other causes of syncope may still occur in these patients, the presence of myocardial dysfunction increases the probability of an arrhythmic origin. Vigorous ventricular contraction is thought to be a critical component of the vagal response in patients with neurocardiogenic syncope. Therefore, it is unclear whether patients with ventricular dysfunction can develop vasodepressor syncope, although they have an abnormal vasodilatory response to a reduction in preload.(24) Heart failure drug therapy, eg, ACE inhibitors and β-blockers, can further aggravate the abnormal baroreflexes in patients with a cardiomyopathy by causing vasodilatation, volume depletion, and sinus node dysfunction. For these reasons, an abnormal head-up tilt table test alone should not lead to a diagnosis of vasodepressor syncope in a patient with NIDCM.

Electrophysiological Study

Electrophysiological testing is frequently used to determine whether there is an arrhythmic basis for syncope, but it is less useful for patients with NIDCM than for patients with a prior myocardial infarction.(25) Ventricular stimulation also appears to have a limited role in the evaluation of these patients after an episode of syncope. A study of patients with advanced heart failure and syncope that included a large number of patients with NIDCM found that the 1-year risk of sudden death was 45% regardless of the cause of syncope.(26) These results suggest that electrophysiological testing in patients with NIDCM and syncope has a low negative predictive value. There is no consistent evidence to support the use of noninvasive testing to further risk-stratify a patient with unexplained syncope and a dilated cardiomyopathy.

NIDCM, Unexplained Syncope, and Negative Electrophysiological Study

Data are limited with regard to the appropriate therapy for patients with NIDCM and syncope when a complete evaluation is unrevealing. There is no evidence to support the empirical use of antiarrhythmic drugs in these patients. Implantable defibrillator therapy, however, may be reasonable. A study of 14 patients with NIDCM, unexplained syncope, and a negative electrophysiological test who were

treated with an implantable defibrillator found that 50% of the patients received appropriate defibrillator therapy during 2 years of follow-up.(27) Furthermore, recurrent syncope and presyncope were due primarily to ventricular tachyarrhythmias, and the incidence of appropriate shocks was similar to that of patients with NIDCM and a history of cardiac arrest.(27) A similar study found a high proportion of appropriate defibrillator therapies in a group of patients with NIDCM and syncope who had no inducible arrhythmias.(28) At least 2 additional studies that reported the outcome of patients with NIDCM treated with an implantable defibrillator for secondary prevention of sudden cardiac death included patients who presented with syncope.(29,30) In 1 study, 10 patients presented with syncope resulting from ventricular tachycardia.(29) In the other study, 6 of 49 patients presented with syncope.(30) About half of the patients in each study received an appropriate shock during follow-up.
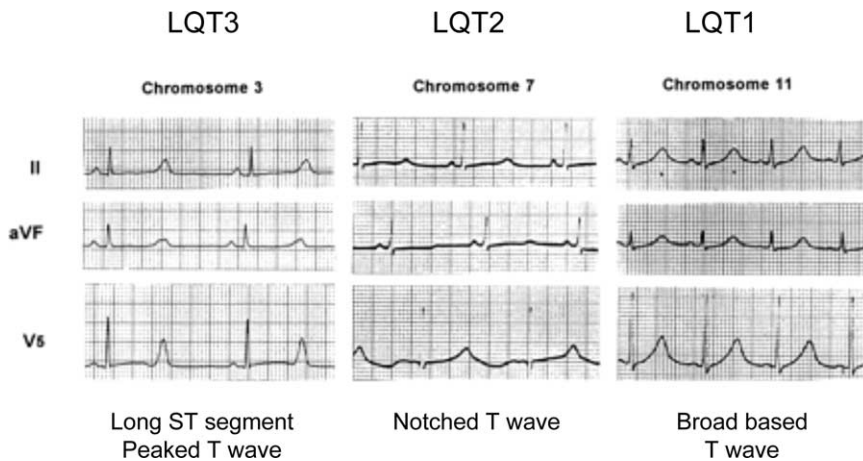
Several recent large trials have demonstrated the efficacy of defibrillator therapy in patients with advanced heart disease without prior syncope. The SCD-HeFT trial included a large number of patients with NIDCM and congestive heart failure. This study demonstrated a survival advantage with defibrillator therapy.(20) In the DEFINITE trial, which enrolled only patients with NIDCM, defibrillator therapy significantly reduced the risk of sudden cardiac death and demonstrated a nearly significant reduction in overall mortality.(31) Because syncope probably further increases the risk of sudden cardiac death in patients with NIDCM, ICD therapy is appropriate in this setting.

Syncope in Other Forms of Structural Heart Disease

Hypertrophic Cardiomyopathy

Hypertrophic cardiomyopathy is a relatively frequent (1 in 500 individuals), genetically determined myocardial disease with a variable prognosis.(32–34) The diagnosis of hypertrophic cardiomyopathy is confirmed with echocardiography, which demonstrates a hypertrophied, nondilated left ventricle in the absence of secondary causes of hypertrophy. Although the risk of sudden cardiac death was initially overestimated because of referral bias, hypertrophic cardiomyopathy remains an important cause of sudden death, particularly in young patients. The annual risk of sudden death in unselected patients with hypertrophic cardiomyopathy is estimated to be 0.6% to 1%.(33,34) There is often a striking discordance between the risk of sudden cardiac death, echocardiographic findings, and the presence of symptoms.

Syncope is a major risk factor for subsequent sudden cardiac death in hypertrophic cardiomyopathy (relative risk ≈5), particularly if it is repetitive or occurs with exertion.(32–34) However, in addition to self-terminating ventricular arrhythmias, many other mechanisms can cause syncope in hypertrophic cardiomyopathy, including supraventricular arrhythmias, severe outflow-tract obstruction, bradyarrhythmias, decreased blood pressure in response to exercise, and neurocardiogenic syncope.(32) The presence or absence of other sudden cardiac death risk factors such as family history of sudden death, frequent nonsustained ventricular tachycardia, or marked hypertrophy may help in the determination of risk. Most authorities agree that electrophysiologi-

LQT3   LQT2   LQT1

Chromosome 3   Chromosome 7   Chromosome 11

Long ST segment   Notched T wave   Broad based
Peaked T wave   T wave

Figure 2. Different patterns of QT prolongation in LQTS. Morphology of the QT segment and T wave may be different in different genetic subsets of the LQTS, although there is significant individual variation. Reproduced with permission from Moss et al.(64)

cal testing plays a minimal role in risk stratification. Specific genetic mutations have been identified that are associated with high risk of sudden cardiac death. However, the use of genotyping to determine sudden cardiac death risk is not readily available and is not routine.

Observational studies have demonstrated that implantable defibrillator therapy is effective in high-risk patients with hypertrophic cardiomyopathy.(35) Twelve percent of patients with hypertrophic cardiomyopathy who received an implantable defibrillator for primary prevention of sudden cardiac death received appropriate therapies during 3 years of follow-up. The incidence of appropriate and inappropriate implantable defibrillator shocks was 40% and 25%, respectively, in patients treated for secondary prevention.(35)

Arrhythmogenic Right Ventricular Dysplasia/Cardiomyopathy
ARVD/C is characterized by ventricular tachycardia and morphological abnormalities of the right ventricle caused by myocyte replacement by adipose tissue or fibrosis.(36) ARVD/C may be familial (30% to 50%), but sporadic forms occur and may represent a different disease process. The diagnosis of ARVD/C can be subtle, and the adoption of consensus criteria based on structural, functional, and ECG characteristics has been helpful.(37,38) Up to 20% of sudden cardiac deaths in patients <35 years of age may be secondary to ARVD/C. Sudden cardiac death may be the first manifestation of the disease process, but patients usually present with premature ventricular beats, syncope, or sustained ventricular tachycardia with a left bundle-branch block morphology.

Syncope is regarded as an ominous finding in ARVD/C. The utility of electrophysiological testing in these patients is not firmly established. Typically, sustained ventricular tachycardia is induced in patients who have presented clinically with ventricular tachycardia, but its role in risk stratification is unclear. Several recent series have demonstrated the efficacy of implantable defibrillator therapy in this group, noting annual rates of appropriate implantable defibrillator therapy of approximately 15% to 20%.(39,40)

Syncope Resulting From Inherited Cardiac Ion Channel Abnormalities
Inherited cardiac ion channel abnormalities can cause syncope and sudden death as a result of ventricular arrhythmias

in the absence of structural heart disease. The 2 most prevalent are the LQTS and the Brugada syndrome. The diagnosis of both conditions relies on a careful family history and analysis of the ECG (Figures 2 and 3). Family screening is important when an index patient is identified, although sporadic cases also occur.

Long-QT Syndrome
LQTS is characterized by prolongation of the QT interval with a QTc >450 ms. A genetic defect in either cardiac potassium (LQT1 and LQT2) or sodium (LQT3) channels results in delayed repolarization and QT prolongation.(41) LQTS generally is inherited with an autosomal dominant pattern variable penetrance. The risk of cardiac events depends on the specific genetic defect, gender, and age.(42) The most important nondemographic risk factor is the degree of QT prolongation. The lifetime risks of syncope or aborted or actual sudden death in LQTS patients with a QTc <440 ms, 460 to 500 ms, and >500 ms are approximately 5%, 20%, and 50%, respectively. Syncope is an ominous finding and is presumably secondary to an episode of torsade des pointes polymorphic ventricular tachycardia that terminates spontaneously. Treatment options include β-blockers and implantable defibrillators.(43) Other important interventions include restriction of strenuous or competitive exercise, avoidance of medicines that prolong the QT interval (a comprehensive listing is available at www.QTdrugs.org), and family screening.

Brugada Syndrome
The Brugada syndrome is a heritable disorder of the cardiac sodium channel resulting in ST elevation in the anterior precordial leads, ie, V_1 and V_2, and susceptibility to polymorphic ventricular tachycardia.(44) The distinctive ECG pattern is diagnostic (Figure 2), although the ECG is often dynamic. The diagnostic ECG findings may be intermittent, change over time, or be present only after provocative maneuvers, eg, procainamide infusion. Patients with Brugada syndrome who present with syncope have a 2-year risk of sudden cardiac death of approximately 30%(44); hence, implantable defibrillator therapy typically is recommended.

Evaluation of the Pediatric Patient With Syncope
The differential diagnosis and evaluation of syncope in pediatric patients are similar to those in adults. Because
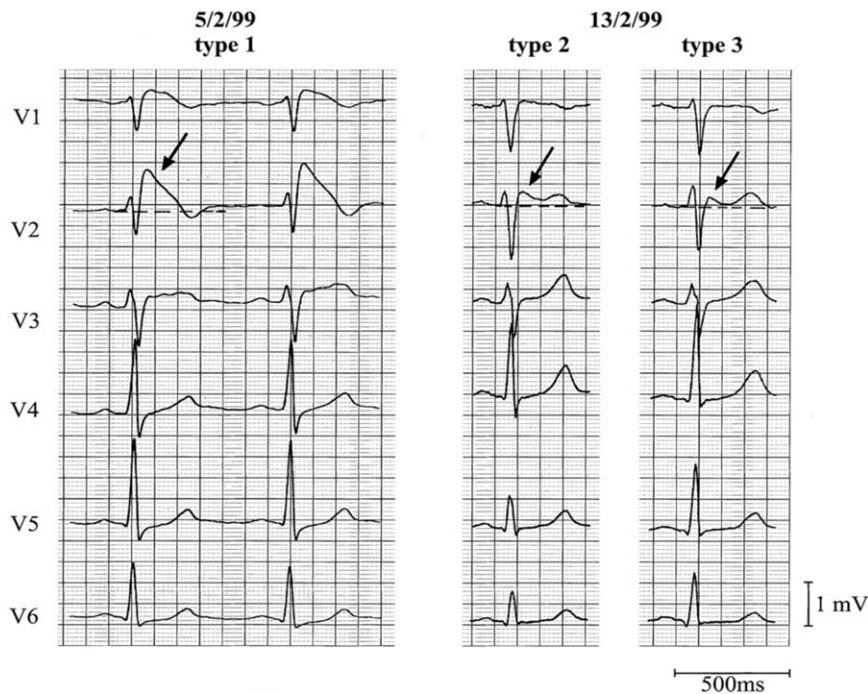
Figure 3. ECG changes in the Brugada syndrome. ST elevation occurs in the anterior precordial leads, leads V_1 and V_2. Type 1 (coved) ECGs with $\geq$1 mV of ST elevation have the most prognostic significance. ECG recordings may change over time, as in this example, and serial ECGs may be important. Reproduced with permission from Wilde et al.(65)

underlying heart disease is less common in the young, syncope in pediatric patients is generally benign. The goal of the evaluation is to identify high-risk patients with underlying heart disease, which may include identifiable genetic abnormalities such as the LQTS, Brugada syndrome, or hypertrophic cardiomyopathy. The evaluation of the pediatric patient with syncope may extend to other family members when a genetic condition is identified.

The evaluation of syncope in the absence of underlying heart disease, ECG abnormalities, or positive family history is limited in scope and often ends with the exclusion of underlying heart disease. Syncope associated with high-intensity physical activity is a typical presentation of hypertrophic cardiomyopathy or catecholaminergic polymorphic ventricular tachycardia and generally is evaluated with an echocardiogram and an exercise stress test.

Syncope in a pediatric patient with a normal ECG and echocardiogram may be due to breathholding spells.(45,46) Breathholding spells resulting from emotional upset have been reported in 2% to 5% of well patients, and therapy is rarely required.(46) Neurocardiogenic syncope in the healthy child or adolescent is a common disturbance.

Bradycardia and Tachycardia
In pediatric patients, syncope caused by isolated bradycardia is relatively uncommon. In children, resting bradycardia may indicate drug ingestion, cardiac manifestations of anorexia nervosa, or central nervous system trauma. Transient but profound sinus pauses or sustained bradycardia may result in syncope due to a neurocardiogenic reflex. Most symptoms associated with sinus node dysfunction are due to an inadequate chronotropic response, but associated tachycardia should be sought. Syncope is uncommon in patients with first- and second-degree heart block, but the occurrence of complete AV block is a Class I indication for permanent

pacing.(23) Tachycardia may cause syncope in the ostensibly normal pediatric patient; palpitations are a usual accompanying symptom, but other nonspecific symptoms such as lightheadedness, chest pain, dyspnea, pallor, or nausea may be present.

Underlying Heart Disease
As in the adult patient, when underlying heart disease is present, syncope is potentially life-threatening for pediatric patients as compared with their normal counterparts.(47) The evaluation of syncope in this setting requires an understanding of the patient's cardiac anatomy, surgical history, and residual hemodynamic burden. Syncope resulting from hypercyanotic spells in children with untreated congenital heart disease such as tetralogy of Fallot is uncommon and should be considered an indication for surgical intervention. Complete AV block or ventricular tachycardia may account for syncope in patients with a history of surgery involving the ventricle(s), eg, tetralogy of Fallot or a ventricular septal defect. If ventricular tachycardia is suspected, then electrophysiological testing may be helpful. Patients with sustained ventricular tachycardia or symptomatic nonsustained ventricular tachycardia should be treated.

Hypertrophic cardiomyopathy is the most common cause of sudden cardiac death in adolescents.(48) Young age and syncope are risk factors for sudden cardiac death among these patients.(37,49,50) In pediatric patients with aortic stenosis, syncope typically occurs with exercise and is an ominous sign.(46) Syncope frequently is seen in children with primary pulmonary hypertension. Near-syncope or syncope has been reported as the presenting symptom in 13% of children with idiopathic dilated cardiomyopathy.(51)

Coronary artery anomalies are present in approximately 1% of the population. Although most children and adolescents with coronary artery anomalies are asymptomatic,

syncope or sudden cardiac death may be the presenting symptom.(52,53) Patients are most at risk when an anomalous coronary artery courses between the aorta and the pulmonary artery trunk.

Special Considerations in the Elderly

The annual incidence of a fall in the elderly is 30%. Up to 30% of falls in the elderly may be due to syncope.(54) The clinical presentation of syncope in the elderly is often variable and atypical. Marked clinical overlap exists between falls, orthostatic hypotension, and dizzy spells, which may all present as syncope.(54,55) Furthermore, syncope may be multifactorial in the elderly patient. Special considerations in the evaluation of syncope in the elderly include (1) age-related changes that predispose the elderly to syncope; (2) varied clinical presentation that includes falls, gait disorders, dizzy spells, and amnesia; (3) drug interactions; and (4) multiple diseases.

Several age-related changes predispose the elderly to syncope. These alterations include reductions in thirst, ability to preserve sodium and water, baroreceptor response, and heart rate response to orthostatic stress, as well as autonomic dysfunction.(55,56) These physiological changes, in combination with the frequent use of multiple medications, are risk factors for orthostatic intolerance and syncope.

Aging is associated with a variety of diseases, including underlying heart disease, gait disorders, cardiovascular deconditioning, recurrent falls, and orthostatic hypotension. The elderly often are treated with multiple medications, including diuretics, β-blockers, calcium antagonists, ACE inhibitors, nitrates, antipsychotic agents, tricyclic antidepressants, antihistamines, dopamine agonists and antagonists, and narcotics, all of which may precipitate syncope. Alcohol also may be a contributing factor. The effects of these drugs and their interactions are exacerbated in the elderly because of the loss of peripheral autonomic tone that occurs with aging.(56)

Orthostatic hypotension is common in older patients and is the cause of syncope in 6% to 33% of patients.(57) A common clinical presentation of syncope in the elderly is postprandial hypotension that is frequently confused with transient ischemic attacks or seizures. Carotid sinus hypersensitivity is an underrecognized cause of syncope in the elderly.(58) A recent report suggests that 30% of unexplained syncope in the elderly is due to carotid sinus hypersensitivity, although pacing may not prevent syncope.(59) Neurally mediated causes remain a frequent mechanism of syncope in the elderly and may be underestimated because of an atypical presentation. Cardiovascular medications may be responsible for almost half of these episodes.

Elderly patients may present with recurrent falls resulting from syncope. Gait disorders secondary to central nervous system alterations may be associated with orthostatic hypotension and other chronic autonomic disorders. Unexplained syncope may be the first manifestation of degenerative disorders such as Parkinson's disease.(56,60) Classic clinical features of neurally mediated causes of syncope, including typical pre-episode and postepisode symptoms, often are absent in older patients.(11,56) Furthermore, complete amnesia is present in up to 40% of elderly patients with syncope.(61)

As with other patients with syncope, the goal of the diagnostic evaluation in the elderly is to exclude life-threatening illnesses and to prevent recurrent falls. Orthostatic blood pressure and heart rate measurements should be obtained.

The treatment of syncope is directed at its cause. In the elderly, multiple origins of syncope frequently coexist and need to be addressed. Particular emphasis should be given to the impact of polypharmacy, orthostatic intolerance, autonomic dysfunction, and carotid sinus hypersensitivity.

Neurological Evaluation

Neurological causes should be considered in patients with syncope, although syncope is an unusual manifestation of neurological processes. Neurological causes of syncope should be pursued only if suggested by the history or physical examination.

Cardiac causes of syncope can be accompanied by upward gaze deviation, asynchronous myoclonic jerks, and brief automatisms that result from global cerebral hypoperfusion and are not an indication for a neurological evaluation. Cerebrovascular disease can cause syncope in the presence of severe bilateral carotid or basilar artery disease, but syncope in these settings rarely occurs in the absence of other focal neurological signs or symptoms. Syncope in the supine position, preceded by an aura, or followed by confusion or amnesia is likely caused by a neurological disorder. Focal neurological signs such as diplopia, limb weakness, sensory deficits, or speech difficulties indicate an abnormality in a particular brain area, are usually caused by a seizure, and are an indication for a neurological evaluation.(62,63)

Neurological disorders that cause syncope typically result in orthostatic hypotension from dysautonomia, and they can be suspected by the history, screening neurological examination, and bedside testing of orthostatic vital signs. Orthostatic hypotension can be induced by tricyclic antidepressants, nitrates, and antiparkinsonian medications; neurodegenerative disorders of the central nervous system such as Parkinson's disease; or a peripheral autonomic neuropathy such as diabetes mellitus. Autonomic testing to confirm the presence of a dysautonomia, distinguish central from peripheral causes, and guide patient management includes tilt table testing, cardiac responses to deep breathing and the Valsalva maneuver, and sweat testing. Disorders that increase intracranial pressure, eg, subarachnoid hemorrhage or rarely brain tumors, can result in true syncope. The neurological basis of these disorders is usually obvious and associated with headache, meningismus, and/or other focal neurological findings.

Seizure disorders are the most common neurological cause of episodic unresponsiveness. Seizure disorders can be confirmed by an electroencephalogram, although repeated studies, prolonged electroencephalographic monitoring, or a sleep-deprived electroencephalogram may be necessary.

Cardiac involvement is frequent in neuromuscular diseases, eg, Duchenne dystrophy. As a result of cardiac involvement, these patients may develop cardiomyopathy, complete AV block, ventricular tachycardia, or ventricular fibrillation. Evaluation of syncope in the setting of a neuromuscular disease includes an ECG, an echocardiogram, and

an assessment for cardiac ischemia. Treatment may include pacemaker or implantable defibrillator therapy.(21)

When a neurological basis of syncope is suspected, imaging of the brain with either CT or MRI is indicated. If cerebrovascular disease is suspected, imaging of the extracranial and intracranial carotid arteries is appropriate.

Conclusions

Syncope can be a precursor to sudden death, particularly in patients with underlying heart disease. Most importantly, the evaluation of syncope includes an assessment for structural heart disease and ischemia. Less common causes of syncope that are associated with sudden death should be excluded and include Wolff-Parkinson-White syndrome and inherited cardiac ion channel abnormalities, eg, LQTS. When underlying heart disease is identified, the evaluation and treatment are generally 2 pronged. First, the underlying heart disease, with or without ischemia, and its possible contribution to the episode of syncope must be addressed. Second, a primary arrhythmia evaluation, with ventricular tachycardia or ventricular fibrillation in mind, should be pursued. A specific diagnosis should be obtained, particularly in high-risk patients, thus allowing appropriate and potentially life-saving therapy.

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