Recognizing Life-Threatening Causes of Syncope

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KEYWORDS

• Syncope • Clinical history • Cardiac arrest • Structural heart disease • Risk • Repolarization • Family history • Ventricular arrhythmia

KEY POINTS

- The identification of potentially fatal causes of syncope is usually possible with a careful history and an electrocardiogram (ECG).
- The history should focus on the presence of structural heart disease, leading to the assumption that life-threatening ventricular arrhythmias may have caused syncope if present.
- Uncommon but concerning causes of syncope, including genetic causes, will often be evident based on an unusual context of syncope, a family history of suspicious events, and a careful evaluation of the ECG.
- Tailored investigations stemming from the initial clinical evaluation will confirm a suspected diagnosis and will rarely uncover an unexpected diagnosis.
- Involvement of specialist cardiology and cardiac electrophysiology services in such cases will help facilitate the selection of appropriate tests and subsequent therapeutic options.

INTRODUCTION

The diagnostic challenges posed by an episode of syncope are manifold. The identification of the cause is often far from straightforward, with a key concern for the small chance that the episode represents a potentially fatal process. Cardiovascular causes of syncope can be classified as either neurally mediated or attributable to cardiac abnormality. Cardiac causes of syncope include arrhythmic disturbances and obstructive cardiac lesions.[1] This distinction between potential attributable causes is of paramount importance because cardiac causes of syncope are associated with adverse outcomes, including an increased risk of death.[2,3] Approximately 35% of the population will experience syncope at some point in their lifetime.[4] As a significant proportion result from neurocardiogenic causes (21%–50%), identifying the small proportion with life-threatening causes is crucial in balancing reasonable reassurance in the vast majority of cases with thoughtful investigation in the remainder.[3,5] Accordingly, this article focuses on life-threatening causes of syncope and a diagnostic approach to facilitate their identification.

PROGNOSIS OF A PATIENT WITH SYNCOPE

Although the overall prognosis of an individual presenting with syncope is favorable, certain features reliably portend a poor prognosis. Chief among these is the identification of either a structural or electrical cardiac etiology for the syncopal episode. In early retrospective analyses of patients with syncope, cardiac syncope was associated with a 21% to 30% mortality rate at 1 year versus 4% to 12% in those with noncardiac syncope.[2,6] A more recent study has suggested a more conservative 15% mortality rate at 1 year in patients with cardiac syncope.[7] In the largest such study to

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date, cardiac syncope was associated with a 2-fold increase in the risk of death compared with those without a history of syncope, with an approximately 50% 5-year survival.[3] By comparison, patients with neurocardiogenic causes of syncope have a favorable prognosis that is comparable to asymptomatic controls.[3,8] However, this is confounded by the poor correlation of the cause of syncope with the cause of subsequent death. The SCD-HeFT trial identified syncope as a high-risk marker for subsequent mortality, but an implantable cardioverter-defibrillator (ICD) or the use of antiarrhythmic agents did not alter the outcome in a comparison with patients treated with placebo.[9]

Even in the absence of a firm diagnosis of cardiac syncope, the presence of structural cardiac abnormalities or evidence of a primary electrical disorder is associated with a poor prognosis and a hazard ratio (HR) for death of 5.57.[10] On the other hand, a structurally normal heart with a normal electrocardiogram (ECG) is usually associated with a benign etiology for syncope and a favorable prognosis.[10,11] Fundamental to the ability to identify high-risk cases of syncope, therefore, is the ability to effectively recognize, or at the very least infer, the presence of either structural or electrical cardiac anomalies. As these anomalies may be subtle, it is prudent to initially familiarize oneself with potentially lethal causes of syncope.

HIGH-RISK CAUSES OF SYNCOPE AND WHEN TO WORRY

Most cases for which syncope is life threatening are neither difficult to diagnose nor, for that matter, difficult to treat. Such cases represent clear evidence of manifest conduction system disease or ventricular arrhythmias, or a propensity to the same with left ventricular (LV) dysfunction or resting conduction disturbances. This article intentionally minimizes the discussion of these scenarios, because they represent established and familiar management scenarios to the reader, summarized in **Box 1** and **Fig. 1**. Acute ischemic syndromes may produce syncope through multiple mechanisms, and should be ruled out in the appropriate clinical setting. Having said this, they are a distinctly unusual cause of unexplained syncope and seldom warrant more than standard noninvasive clinical exclusion.

Arrhythmic Etiology

Arrhythmias are the most common cause of cardiac syncope, and can be further subdivided into (1) bradyarrhythmias and (2) tachyarrhythmias.[12]

Box 1
Life-threatening causes of syncope that typically present in the context of manifest cardiac disease

- Sinus node dysfunction
  - Syncope is a common presentation, but there is little concern regarding sudden death, typically not life threatening
- Atrioventricular node dysfunction
  - Infranodal atrioventricular block (Mobitz II second-degree block)
  - Complete heart block
  - Alternating bundle branch block
- Ventricular tachyarrhythmias
  - Polymorphic ventricular tachycardia secondary to ischemia
  - Scar-related monomorphic ventricular tachycardia
  - Ventricular fibrillation
- Reduction in stroke volume
  - Mechanical complication
    - Cardiac perforation ± pericardial tamponade
    - Ventricular septal rupture
    - Papillary muscle rupture

Bradyarrhythmias

Bradyarrhythmias can result from dysfunction of the conduction system at any level (see **Box 1**). Atrioventricular (AV) block can occur at the level of the AV node (ie, nodal) or below the AV node (ie, infranodal). This distinction is of importance, as infranodal block is more likely to be associated with the lack of an adequate escape rhythm and a small risk of sudden death.[13] Infranodal AV block is generally typified by the presence of Mobitz II second-degree AV block, or the presence of an escape rhythm with an intraventricular conduction delay or overt bundle branch block. Nodal AV block, on the other hand, is associated with Mobitz I (Wenckebach) second-degree AV block, or the presence of a narrow complex escape rhythm. The presence of alternating bundle branch block morphologies signals an individual at high risk of proceeding to complete heart block. Caution must be observed with apparent Wenckebach AV block in the presence of a wide QRS complex, as the level of block may in fact be infranodal (**Fig. 2**).

Tachyarrhythmias

Both supraventricular and ventricular tachyarrhythmias may result in syncope, with failed vascular