## Society Position Statement

# Canadian Cardiovascular Society and Canadian Pediatric Cardiology Association Position Statement on the Approach to Syncope in the Pediatric Patient

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## ABSTRACT

Pediatric syncope is a common problem that peaks in adolescence, for which there are few data or evidence-based consensus on investigation and management. This document offers guidance for practical evaluation/management of pediatric patients (age < 19 years) with syncope encountered in the acute or primary care setting. The writing committee used the Grading of Recommendations, Assessment, Development, and Evaluation (GRADE) methodology. Most syncope is vasovagal, which is benign and does not require extensive investigation. This Position Statement presents recommendations to encourage an efficient and cost-effective disposition for the many patients with a benign cause of syncope, and highlights atypical or concerning clinical findings associated with other causes of transient loss of conscious-

## RÉSUMÉ

La syncope dans la population pédiatrique est un problème fréquent qui culmine à l'adolescence, et pour lequel il existe peu de données ou pas de consensus fondé sur des données probantes quant aux examens et à la prise en charge. Le présent document donne des conseils pratiques en matière d'évaluation et de prise en charge des patients d'âge pédiatrique (< 19 ans) avec syncope lors de la prise en charge en salle d'urgence ou de première ligne. Le comité de rédaction a utilisé la méthodologie GRADE (Grading of Recommendations, Assessment, Development, and Evaluation). La plupart des syncopes dites vasovagales sont bénignes et ne nécessitent pas d'examens approfondis. Cet énoncé de position présente des recommandations pour favoriser la prise en charge efficace et efficiente des nombreux

This document serves as a clinical guideline for the evaluation and management of pediatric patients, typically those younger than 19 years, with syncope encountered in the acute or primary care setting. It was developed after thorough consideration of the best available evidence and clinical experience, and represents the consensus of a Canadian panel

comprised of multidisciplinary experts with a mandate to formulate disease-specific recommendations. The purpose of this statement is to ensure that practitioners who encounter pediatric patients with syncope might readily recognize syncope due to an etiology other than transient autonomic nervous system dysfunction, and to encourage an efficient and

experts on this topic with a mandate to formulate disease-specific recommendations. These recommendations are aimed to provide a reasonable and practical approach to care for specialists and allied health professionals obliged with the duty of bestowing optimal care to patients and families, and can be subject to change as scientific knowledge and technology advance and as practice patterns evolve. The statement is not intended to be a substitute for physicians using their individual judgement in managing clinical care in consultation with the patient, with appropriate regard to all the individual circumstances of the patient, diagnostic and treatment options available and available resources. Adherence to these recommendations will not necessarily produce successful outcomes in every case.

ness. The prodrome and the circumstances around which the event occurred are the most important aspects of the history. Syncope occurring midexertion suggests a cardiac etiology. A family history, which includes sudden death in the young or from unknown causes or causes that might be suspected to be other than natural can be a red flag. The electrocardiogram is the most frequently ordered test, but the yield is low and the test is not cost-effective when applied broadly to a population of patients with syncope. We recommend an electrocardiogram when the history is not suggestive of vasovagal syncope and there are features suggestive of a cardiac cause like absence of a prodrome, midexertional event, family history of early-life sudden death or heart disease, abnormal physical examination, or new medication with potential cardiotoxicity. For most patients with syncope, medical testing is not required and lifestyle modifications without medications suffice to prevent recurrences.

patients dont la cause de la syncope est bénigne, et met l'emphase sur les résultats cliniques atypiques et inquiétants associés à d'autres causes de perte de connaissance. Le prodrome et les circonstances entourant la survenue de l'évènement sont les aspects les plus importants des antécédents médicaux. La syncope qui se présente pendant l'effort physique suggère une étiologie cardiaque. Les antécédents familiaux, dont la mort subite chez les jeunes ou de cause inconnue ou de cause que l'on soupçonne comme étant autre que naturelle, peuvent être des signes avant-coureurs. L'électrocardiogramme est l'examen le plus fréquemment prescrit, toutefois son rendement est faible, et l'examen n'est pas efficient lorsqu'il est appliqué à grande échelle à une population de patients atteints de syncope. Nous recommandons un électrocardiogramme lorsque les antécédents ne sont pas évocateurs de syncope vasovagale et s'il existe des caractéristiques qui évoquent une cause cardiaque comme l'absence de prodrome, l'évènement pendant l'effort physique, les antécédents familiaux de mort subite ou de maladie cardiaque précoces, les examens physiques anormaux ou les nouveaux médicaments potentiellement cardiotoxiques. Pour la plupart des patients atteints de syncope, les examens complémentaires ne sont pas nécessaires et les modifications du mode de vie sans médication suffisent pour prévenir les récurrences.

cost-effective disposition for those with evidence of a benign cause. A systematic and directed approach (Fig. 1) will prevent missing potentially dangerous diagnoses, the most common of which are cardiovascular and neurologic abnormalities (Table 1). With the exception of vasovagal syncope (VVS), the most common cause of syncope in children, this document does not discuss the management of the various conditions that can present with syncope.

## Methods

The Canadian Cardiovascular Society (CCS) appointed cochairs, a primary panel, and a secondary panel to develop this statement. The primary panel developed the scope of the document, identified topics for review, performed the literature review, and critical appraisal of the identified literature using the Grading of Recommendations, Assessment, Development, and Evaluation (GRADE) methodology, drafted the recommendations, and voted on the recommendations. Peer review was provided by the secondary panel and the CCS Guidelines committee. The final draft was presented and approved by the CCS Executive Committee.

Using a validation set of relevant articles identified by the authors, a librarian drafted a search strategy for MedLine that was adapted and run in Embase, Cinahl, and PubMed. The strategy consisted of 2 blocks of search terms (subject heading and free text), combined using the AND operator. The first block contained the condition "syncope" and the terms in the second block were related to the diagnostic concept of syncope (eg, tilt table test, electrocardiogram [ECG], differential diagnosis). The set of articles retrieved using these 2 search blocks was further restricted to pediatric studies in English or French, and a filter to exclude review articles was applied. The search covered the time from the inception of each database through December 2015. A complete description of this strategy is available in Supplemental Appendix S2. The search retrieved 5997

references. After duplicates were removed, 4307 references were screened using Covidence. These were reviewed by the panel members to ensure they were pediatric, English or French, and original articles. Case reports were excluded, leaving 296 articles for full-text review and 231 that were included. The primary panel built the evidence for the recommendations on the basis of selected relevant articles (see Supplemental Appendix S3).

## Terminology

Syncope is a transient loss of consciousness, associated with an inability to maintain postural tone, followed by rapid and spontaneous recovery with the absence of clinical features specific for another form of transient loss of consciousness such as epileptic seizure.[3] It is caused by global cerebral hypoperfusion, secondary to a myriad of etiologies (Table 1). Unlike in the adult population, the distinction between syncope and transient loss of consciousness has not been applied systematically to children. In this document, we attempt to align with the international and adult communities in using the term "transient loss of consciousness" when the etiology might not be cerebral hypoperfusion, with the most notable example being seizures. There are many terms to describe the syncope due to a transient dysfunction of the autonomic nervous system, including "fainting," "vasovagal syncope," "reflex syncope," or "neurocardiogenic syncope." In this document, we will use the term, vasovagal syncope (VVS), which is defined as a syncope syndrome that typically (1) occurs with upright posture held for more than 30 seconds or with exposure to emotional stress, pain, or medical settings; (2) features diaphoresis, warmth, nausea, and pallor; (3) is associated with hypotension and relative bradycardia, when known; and (4) is followed by fatigue.[3]

## Epidemiology

Syncope is common, with a cumulative incidence of 30%-40% by age 60 years,[3] and accounts for approximately 1% of
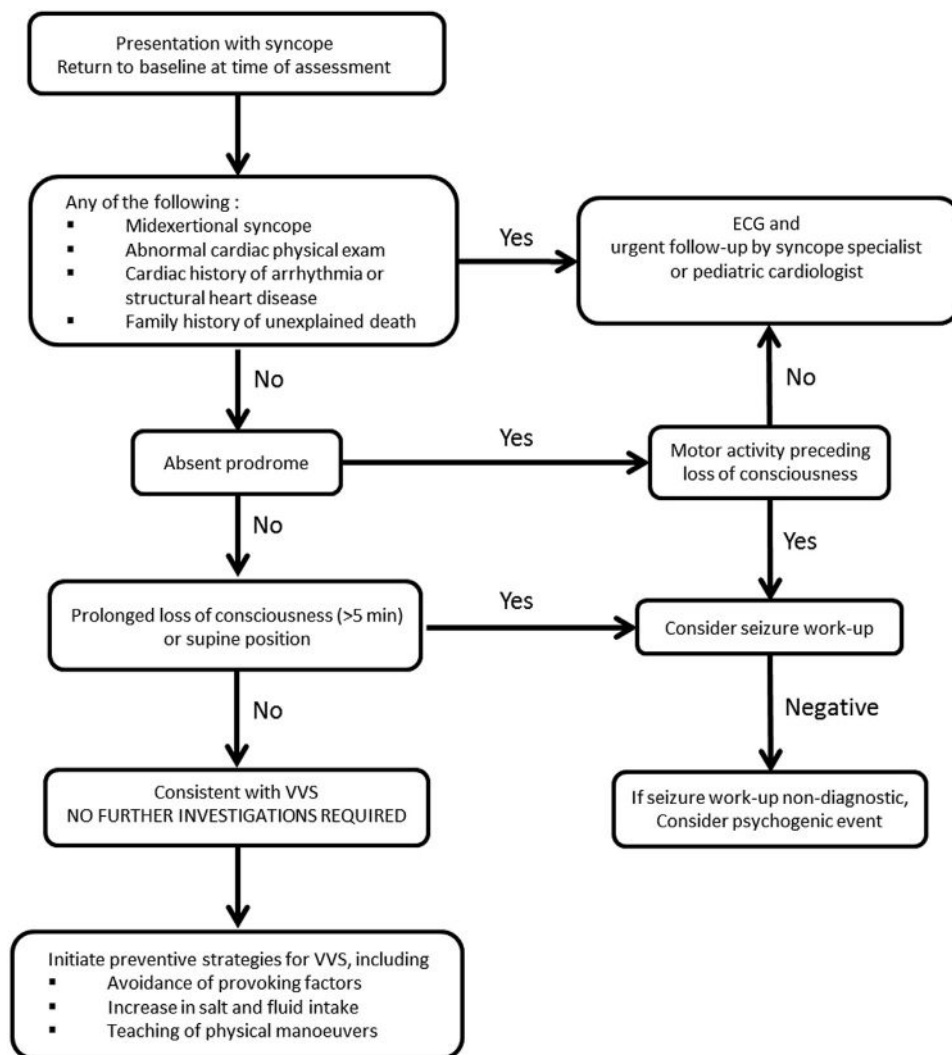
**Figure 1.** Clinical pathway for pediatric syncope patients. ECG, electrocardiogram; VVS, vasovagal syncope.

pediatric emergency room visits. Many patients with syncope do not seek medical attention.[4] The incidence peaks in adolescence, with a lesser peak between 6 and 18 months, related to breath-holding spells.[5] In breath-holding spells, the child experiences an emotional or minor physical trauma, resulting in a breath-hold and a brief loss of consciousness. The episodes are often associated with facial cyanosis or progressive pallor with an appearance of being disoriented, and can involve involuntary muscle twitching, all of which can be quite dramatic to the observer. However, these are benign self-limited episodes and do not require further investigation or treatment. Some infants with breath-holding spells will have VVS later in life.[6]

## History and Physical

### Symptoms

The prodrome is the most important aspect of the history. A warm or clammy sensation, nausea, light-headedness, or visual changes (eg, seeing spots, grey out, tunneling) are strongly suggestive of VVS. Other symptoms include irritability, confusion, auditory changes, dyspnea, or abdominal symptoms

(Fig. 2). The absence of a prodrome should raise suspicion of a possible cardiac cause.[4,7] In 154 children with syncope, 85% of children with VVS had a prodrome, whereas only 40% of those with cardiac conditions had prodromal symptoms.[8]

Palpitations and chest pain have been related to pediatric cardiac causes of syncope on the basis of very few data. Massin et al. reported that 9 of 25 patients with cardiac syncope had palpitations and/or chest pain; however, in this study the rates of such symptoms in patients with VVS were not reported.[7] Hurst et al. reported that the combination of the following features had 98% specificity for cardiac disease: absence of a prodrome, a midexertional event, and chest pain or palpitations preceding the event.[4] However, there were very few (3 of 3445; < 0.1%) cardiac syncope patients in the study. In the adult literature[9] and in the experience of the authors, palpitations are a relatively common finding in patients with VVS, often attributed to the adrenergic surge preceding the vagal response.

### Circumstances

The circumstances under which the syncope occurred often inform the diagnosis. Syncope occurring

**Table 1.** Common causes of transient loss of consciousness in children[1,2]

| Cause | Incidence, % | Example |
|---|---|---|
| Vasovagal syncope | 64-73 | |
| Breath-holding spell | 6.4 | |
| Cardiac | 2.9-4.8 | • Primary electrical disturbances: long QT syndrome, Brugada syndrome, catecholaminergic polymorphic ventricular tachycardia, short QT syndrome, Wolff-Parkinson-White syndrome<br>• Structural heart disease: Hypertrophic cardiomyopathy, coronary artery anomalies, arrhythmogenic right ventricular cardiomyopathy, valvular aortic stenosis, dilated cardiomyopathy, pulmonary hypertension, acute myocarditis |
| Neurologic | 2.1-4.6 | • Seizures: Panayiotopouolos syndrome<br>• Vascular events: subclavian steal phenomenon, vertebrobasilar insufficiency<br>• Disrupted cerebrospinal fluid circulation: colloid cyst of the third ventricle, posterior fossa tumour<br>• Vertiginous drop attack<br>• Basilar migraine<br>• Narcolepsia/cataplexia |
| Metabolic | 0.8 | • Bleeding, dehydration, hypoglycemia, electrolyte disturbances |
| Psychiatric | 2.2-2.3 | • Conversion disorder, somatization, Munchausen syndrome/malingering, anxiety and hyperventilation syndrome |
| Unknown | 8.2-18.9 | |

midexertion, before the child has a chance to stop the activity, suggests a cardiac etiology.[10] Whereas postexertional syncope is typically benign, in the review by Miyake et al. of 60 patients with midexertional syncope, 32 patients had cardiac causes.[11] There were no other risk factors according to the history that differentiated the cardiac from noncardiac exertional syncope groups. The distinction between cardiac syncope and postural orthostatic tachycardia syndrome-associated syncope could be helped with the use of the modified Calgary score for the pediatric population.[12] The median of modified Calgary scores for cardiac syncope was −5.0, which significantly differs from that of postural orthostatic tachycardia syndrome. The sensitivity and specificity of a differentiation score of less than −2.5 are 96.3% and 72.7%, respectively (Table 2).[12]

Syncope associated with recent change of position, poor hydration or nutritional status, or a warm environment is usually VVS. The most common position in which VVS occurs is standing, although syncope when seated has also been observed. Syncope when supine suggests neurologic or psychologic causes. In 268 syncopal children, no VVS occurred in the supine position.[13] Other common precipitating factors for VVS include phlebotomy, the sight of blood or disfiguring injury (eg, fractures or soft tissue injuries), hair grooming, micturition/defecation, emotional upset, mild physical trauma or pain, intercurrent illness, especially those with gastrointestinal symptoms, and hot or crowded conditions.

## Collateral history

Interviewing persons who witnessed the syncope might be helpful. Details like duration of the loss of consciousness and required degree of intervention, should be solicited whenever possible. Pallor or loss of colour are frequently seen in VVS. Some involuntary movement is also common in VVS, leading to a suspicion of seizure (Table 3). Twelve percent of prospectively studied pediatric patients have been reported to be affected by the myoclonic variant of VVS.[14] Syncopal myoclonus might manifest as anything from a single muscle twitch to violent jerks affecting the entire body. Muscle jerks are often asynchronous in different parts of the body. Proximal and distal muscles are equally affected and facial involvement is common. In contrast to epileptic activity, syncopal myoclonus is not rhythmic and is only rarely sustained for more than half a minute.[15] VVS might also produce more complex movements, which raises a suspicion of epilepsy.[16] After evaluation with simultaneous tilt test and video electroencephalogram (EEG) recordings, most patients initially treated for epilepsy are subsequently diagnosed with VVS.[16]

In general, loss of consciousness occurs with the onset of movements in seizures, but loss of consciousness precedes movements in most cases of true syncope. Loss of consciousness from VVS frequently lasts < 1 minute. Prolonged events might be noncardiac (eg, seizures, stroke) or represent somatization. One study reported that almost 90% of those with eventual psychiatric diagnoses had loss of consciousness lasting over 5 minutes.[8] Inadequate history-taking and overemphasis on positive family history for seizures were important causes of misdiagnosis of epilepsy.[17]

## Medications, medical history, and family history

Medications that alter vascular tone or heart rate, like β-blockers, calcium channel blockers, and diuretics, might contribute to syncope. The medical history must include previous syncopal events, cardiac disease, diabetes, seizures, medication or drug use, and psychiatric or psychological problems. The family history is important. A family history of sudden death in young, apparently healthy individuals, or from unknown or incongruous causes, such as a "heart attack" in an apparently healthy person, drowning in a capable swimmer, or unexplained motor vehicle accidents, should prompt evaluation for cardiac causes. Although arrhythmias and inherited arrhythmia conditions are much less common than benign causes of syncope, many initially present with syncope. Any family history of structural cardiac disease, arrhythmias, migraine, or seizures is also relevant.

## Suggested approach

The history should focus on the symptoms and context in which syncope occurred (Fig. 2). The most useful information is obtained from the patient directly, even if they are young. The history should be taken before repetitive questions provide leading suggestions. As indicated previously, the prodrome is the most important aspect of the history. The historian must determine whether the patient had stopped their activity and had any prodromal symptoms, or whether the event occurred without any warning. Syncope during exertion can be secondary to peripheral vasodilatation

and hypotension: this should be verified with a stress test.[18] The presence of palpitations is of unclear significance and should be considered in the clinical context, rather than as an isolated finding indicating underlying cardiac disease. Mid-exertional syncope should prompt an urgent cardiac evaluation. New medications or recent dosage adjustments, particularly of antihypertensive or β-blocker agents, should prompt review by the prescriber.

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**RECOMMENDATION**

1. We recommend a detailed history in all cases (Fig. 2) (Strong Recommendation; Moderate-Quality Evidence).

   **Values and preferences.** Because of the unique diagnostic information obtained from the history, the committee placed emphasis on an accurate and detailed history, as supported by all of the available data. The history is the diagnostic test of most utility in managing pediatric syncope.

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**Practical tip.** The history should focus on accompanying symptoms and the context in which syncope occurred. The prodrome and timing of syncope in relation to exercise are particularly important. The most informative aspects are obtained directly from the patient.

### Physical examination

The physical examination is rarely helpful. Evidence of trauma related to the syncopal event, as well as potential diagnostic clues for the cause, should be sought. Complete cardiac and neurologic examinations should be performed to identify the rare occurrence of occult structural cardiac or neurological disease.[19] Assessing the heart rate and blood pressure responses to standing—postural vital signs—are helpful in assessing hydration and showing dysautonomic responses to orthostatic stress. Pulse rate and regularity at the time of syncope are useful. An irregular pulse is usually due to sinus arrhythmia, which is common in children and of no significance. Innocent murmurs are common and should not cause alarm. In contrast, cyanosis, a pathologic murmur, diminished pulse volume, or a sternotomy scar point to underlying structural cardiac disease.[7,8,20] Persistent neurologic deficits should raise concern for underlying neurologic conditions like stroke, seizure, or complex migraine.

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**RECOMMENDATION**

2. A focused physical examination should always be performed (Strong Recommendation; Low-Quality Evidence).

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**Practical tip.** Postural vital signs are helpful in assessing hydration. An abnormal cardiac or neurologic examination warrants further investigation.

### Investigations

In a review of costs associated with diagnostic testing in a tertiary centre, not including hospital costs and physician fees, the mean cost per patient was > \$1000 in the 1990s.[21] Considering that between 15% and 50% of children have at least 1 syncopal event, the consequences and costs of overinvestigation are significant.[21] Any further testing should be guided by the history and physical examination. In typical VVS, no further investigations are needed. Otherwise, investigations are guided by the history and physical examination to detect life-threatening arrhythmias and causes of syncope. Bloodwork is not required in cases of typical VVS. Hypoglycemia is a rare cause of syncope and is typically associated with autonomic symptoms like sweating, weakness, tachycardia, tremor, nervousness, and hunger.

The ECG is the most frequently ordered test, accounting for more than one-third of all tests in a study of 169 pediatric patients with new-onset syncope.[21] Of these, only 1 ECG was of diagnostic utility, rendering it the least cost-effective test of those performed. A combination of history, physical examination, and ECG had 96% sensitivity for cardiac syncope, with exertional syncope being the most consistent factor.[22] The ECG was the only indicator of cardiac disease in 5 of the 480 patients (1%), and causality could not be determined.

Despite the fear of missing a cardiac cause of syncope, the low yield and specificity do not support routine use of the ECG in the acute setting. For cases in which an ECG is indicated (Table 4), the features that should raise concern are listed in Figure 3. Long QT syndrome (LQTS), due to abnormalities in the function of ion channels controlling repolarization ("ion channelopathy") can cause syncope and sudden death in otherwise healthy individuals. LQTS is the most common ion channelopathy, occurring in 1 in 2500 individuals.[23] The QT interval should be verified and corrected for heart rate according to the Bazett formula (Fig. 4). An ECG done acutely might not be optimal. In the emergency department, approximately one-third of pediatric patients had a QTc interval ≥ 440 ms and normalization of QTc values on follow-up.[23] Thus, first-time ECGs obtained after a syncopal episode must be interpreted with caution to avoid overdiagnosis of a LQTS. Wolff-Parkinson-White syndrome is due to an accessory pathway connecting the atria and the ventricles. It never causes syncope in the absence of arrhythmia.

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**RECOMMENDATION**

3. For all children with atypical syncope or who have additional risk factors (Table 4), we recommend a 12-lead ECG (Strong Recommendation; Low-Quality Evidence).

   **Values and preferences.** Whereas the ECG is the most often ordered test in children with syncope, the data do not support its routine use. The yield is very low (1%), the cost is significant, and abnormal ECGs at the time of the acute event are often false-positives subject to misinterpretation. Therefore, the committee deliberately emphasizes that ECGs are not required in typical syncope and should be obtained only when there is a particular indication, such as those provided in Table 4.

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| History and Physical Findings | RED LIGHT<br><br>Recommend Urgent Referral to Syncope Specialist | YELLOW LIGHT<br><br>Consider Further Investigations or Semiurgent Referral to Syncope Specialist | GREEN LIGHT<br><br>Reassuring for Nonserious Cause; No Further Investigation Required |
|---|---|---|---|
| Hydration status and timing of most recent meal | | | Missed meals, poor fluid intake |
| Environmental conditions | Syncope triggered by loud noise-look for long QT and refer if ECG positive | | Painful stimulus, sight of blood, very warm environment |
| Activity preceding the syncopal event | Midexertional syncope (consider cardiac causes)<br><br>Syncope while swimming (might be associated with LQTS) | | Postexertional syncope<br><br>Prolonged standing |
| Use of drugs and medications | | Medications that may prolong QT (refer if ECG abnormal) | No medications |
| Prodrome | No prodrome (concerning for arrhythmia) | Short or atypical prodrome | Warmth, nausea, light-headedness, a visual grey-out or tunneling of vision |
| Other symptoms | | Acute chest pain followed by syncope<br><br>Palpitations just before syncope | |
| Position of child preceding event | | Supine (consider seizure) | Prolonged or recent standing<br><br>Position change from seated or lying to standing |
| Duration of loss of consciousness | | Prolonged > 5 minutes (consider seizure or somatization) | Short; < 1-2 minutes |
| Movement during event | Tonic-clonic movements or motor activity preceding LOC (consider seizure) | Exaggerated or flailing movements (consider somatization) | Myoclonic jerks after loss of consciousness |
| History (previous syncopal events, cardiac disease, diabetes, seizures, and psychiatric or psychological problems) | Arrhythmia, structural heart disease<br><br>Seizures | Diabetes<br><br>Psychiatric disorder or medications<br><br>Significant comorbidities | No relevant medical history<br><br>Previous events consistent with vasovagal syncope or breath-holding spells |
| Family history (structural cardiac disease, arrhythmias, sudden death, migraines, or seizures) | Sudden death<br><br>Arrhythmias | Seizures<br><br>Structural heart disease | Vasovagal syncope<br><br>Migraines |
| Focused cardiac and neurologic examinations | Pathologic murmur<br><br>Sternotomy scar<br><br>Persistent neurologic deficits (consider stroke, seizure, migraine) | | Normal examination<br><br>Typical flow murmur |

Figure 2. History and physical examination. ECG, electrocardiogram; LOC, loss of consciousness; LQTS, long QT syndrome.

When the history, physical examination, or ECG suggests the possibility of cardiac disease, further testing is warranted. This should be directed by a pediatric cardiologist with expertise in syncope, and will often include an echocardiogram. In one study, echocardiograms were obtained in 67% of patients and none revealed a pertinent abnormality not already suspected on the basis of the history, physical findings, or ECG.[22] A treadmill test should be done when patients have exertional symptoms. In those too young for structured exercise testing, a Holter monitor during active play can be useful. Longer-term monitoring strategies and event monitors are reserved for patients with associated palpitations or recurrent unexplained syncope. Recent attention has focused on implantable cardiac monitors, because of their small size and ease of deployment. These can be useful for complex cases in which extensive investigations have not confirmed a cause of syncope. Their greatest utility might be in patients with established heart disease and new paroxysmal symptoms.

Reports on the specificity of tilt tests in children vary tremendously and results are difficult to interpret.[25] A positive

tilt test might not reliably exclude other conditions and this test is of limited utility.[26] Tilt testing should not be used to establish a diagnosis of VVS. It might be useful to explore episodes of psychogenic syncope.

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**RECOMMENDATION**

4. For children with a history typical of VVS, no family history of arrhythmia, and normal physical examination, we suggest that further cardiac investigations not be performed (Strong Recommendation; Low-Quality Evidence).

   **Values and preferences.** The echocardiogram, treadmill test, Holter monitor, long-term monitoring strategies, and tilt test do not help to establish a diagnosis of VVS. They should generally be prescribed only by specialists with expertise in pediatric syncope in specific situations (eg, treadmill test for exertional syncope).

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Atypical presentations like onset in the supine position, a preceding aura, or subsequent significant confusion or amnesia suggest an epileptic cause. In this case, a sleep-deprived EEG or ambulatory EEG (if the episodes of syncope are frequent enough) might be useful. Standard EEG should include concomitant ECG. Focal slowing or interictal discharges on the EEG, or neurological deficits such as diplopia, dysarthria, and paresis, should be investigated further with brain imaging. In the absence of such focal signs or deficits, neuroimaging is of very low yield.

It is important to remember that other causes of syncope and epilepsy can coexist. If doubts persist after initial investigations, simultaneous tilt testing and video EEG monitoring can help distinguish other forms of syncope from epilepsy in patients with recurrent loss of consciousness.[16]

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**RECOMMENDATION**

5. For children who present with a history typical of VVS, no family history of epilepsy, and normal physical examination, we suggest that an EEG or neuroimaging not be performed (Strong Recommendation; Low-Quality Evidence).

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**Practical tip.** Sleep-deprived EEG, ambulatory EEG, and neuroimaging should be reserved for specific situations like syncope in a supine position, with a preceding aura, or with subsequent significant confusion or amnesia.

## Treatment of VVS in Children

Children with VVS generally do well, with few recurrences, and do not require hospitalization or intervention. The evidence that specific treatments help is sparse, and most recommendations are adopted from the adult literature. Management should begin with explanation and reassurance. Patients can look alarmingly ill when unconscious, so family members and witnesses might benefit from this education. It

**Table 2.** Individual items of the Modified Calgary score

| Question | Point (if yes) |
| --- | --- |
| 1. Is there a history of bifascicular block, asystole, or supraventricular tachycardia? | −5 |
| 2. At times have bystanders noted that you turn blue during your faint? | −4 |
| 3. Did your syncope start when you were 5 years of age or younger? | −3 |
| 4. Do you remember anything about being unconscious? | −2 |
| 5. Do you feel faint with prolonged sitting or standing? | 1 |
| 6. Do you sweat before a faint? | 2 |
| 7. Do you feel faint with pain or in medical settings? | 3 |

The Modified Calgary score is derived from the Calgary score, originally developed to make differential diagnoses between adult vasovagal syncope and other types of syncope. On the basis of a cohort of 213 children, this modified score consists of 7 diagnostic questions related to medical history, triggers, circumstances as well as signs and symptoms of transient loss of consciousness. All questions were answered with "yes" or "no," and then summarized to obtain a total score, ranging from −14 to 6 points. A score of < −3.0 suggests a diagnosis of cardiac syncope, as opposed to postural orthostatic tachycardia syndrome-associated syncope.

Adapted from Yang et al.[12] with permission from Cambridge University Press.

is crucial to educate the patient about the importance of recognizing and reacting to presyncopal prodromes. Avoidance of precipitating factors like prolonged standing, dehydration, and hot crowded environments is beneficial in preventing recurrences.

An increase in dietary salt and fluid intake should be encouraged. In a randomized trial of 166 children and adolescents oral rehydration salts (a mixture of NaCl, KCl, and $NaHCO_3$) decreased syncope recurrences for up to 6 months (56% vs 39%; $P = 0.029$).[27] Both groups also received education and tilt training consisting of periods of standing upright. In adults, physical techniques like squatting, crossing legs, or buttocks-clenching while upright prevented syncope in randomized open-label trials.[3] These manoeuvres are safe and, even in the absence of specific pediatric data, should be taught to all children with VVS.

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**RECOMMENDATION**

6. For children with typical VVS, we recommend a conservative strategy including education, avoidance of provoking factors, increase in salt and fluid intake, and teaching physical manoeuvres as a preventative and rescue strategy. For most patients with VVS, education and hydration strategies suffice (Strong Recommendation; Low-Quality Evidence).

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Medication is rarely required to treat VVS. A small prospective randomized study showed that midodrine prevents recurrent VVS in children who are refractory to conservative management for up to 6 months (recurrence rates in placebo and midodrine groups 80% and 22%, respectively).[28] Side effects from midodrine (principally supine hypertension) are rare. The evidence for benefit from fludrocortisone or β-blockers is weak. A small randomized clinical trial showed that outcomes with placebo were better than with fludrocortisone,

**Table 3. Events easily mistaken for cardiovascular syncope**

| Condition | Distinguishing characteristics |
|---|---|
| Basilar migraine | Headache, rarely loss of consciousness, other neurologic symptoms such as dysarthria, ataxia, and paresthesia |
| Seizure | Loss of consciousness simultaneous with motor event, prolonged postictal phase (eg, bladder and bowel incontinence, tongue-biting) |
| Vertigo | Rotation or spinning sensation, no loss of consciousness |
| Hyperventilation | Inciting event, paresthesias or carpopedal spasm, tachypnea |
| Psychosocial causes | No loss of consciousness, indifference to event |
| Hypoglycemia | Confusion progressing to loss of consciousness, with adrenergic manifestations (sweating, pallor, shivering); requires glucose administration to terminate |

and that most of the outcomes in the placebo arm occurred after the placebo was stopped.[29] Similarly in a prospective, randomized trial, treatment with metoprolol was associated with a higher recurrence rate (43%) compared with conventional treatment (29%).[30]

In patients who undergo tilt testing and other cardiac monitoring strategies, sinus arrest, transient heart block, and asystole can accompany VVS. These findings raise the possibility of treatment with a permanent pacemaker. Although a small double-blind study reported improvement with pacing in very young patients with frequent VVS,[31] this is a highly invasive treatment for a generally benign condition, and should be undertaken only in exceptional cases, and only in consultation with a pediatric arrhythmia specialist.

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**RECOMMENDATION**

7. For children with highly symptomatic VVS resistant to conservative measures, we suggest treatment with midodrine during active hours (Strong Recommendation; Low-Quality Evidence).

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### Prognosis and Disposition

In children who present with syncope, it is critical to identify the rare patients at risk of sudden death. A careful history as outlined previously, as well as a detailed family history, will identify most at-risk patients.[4,32] Atypical features, such as syncope without a prodrome, syncope while supine or during exercise, a family history of early or sudden death, or an ECG suggestive of electrical or structural heart disease, should prompt an urgent referral to a specialist. The prognosis for these patients depends on recognition and treatment of the underlying cause.

**Table 4. When to do an electrocardiogram in syncope**

History is not diagnostic of vasovagal syncope
No prodrome before syncope
Midexertional event (eg, swimming)
Syncope triggered by loud noise or startle
Family history of sudden death or heart disease in young individuals
Abnormal cardiac examination
New medication with potential cardiac side effects (eg, https://crediblemeds.org)



| | ECG Findings |
|---|---|
| **RED LIGHT** | • Abnormal QT interval'<br>   ○ Long QT interval (QTc > 470 ms)<br>   ○ Short QT interval (QTc ≤ 330 ms)<br>• Type 1 Brugada pattern<br>• Delta wave (ventricular pre-excitation or Wolff-Parkinson-White syndrome)<br>• Signs of myocardial ischemia (ST-T wave changes, Q waves > 1 mm wide)<br>• PVCs, polymorphic<br>• Third-degree AV block |
| **YELLOW LIGHT** | • Left ventricular hypertrophy (including left axis deviation, tall R wave in V$_6$, tall S wave in V$_1$, deep Q waves in II, III, and aVF and ST-T wave changes)<br>• PVCs, monomorphic<br>• Second-degree AV block<br>• Heart rate < 40 bpm in normally nourished, nonathletic individual |
| **GREEN LIGHT** | • Sinus arrhythmia<br>• Wandering atrial pacemaker; atrial or junctional rhythm<br>• First-degree AV block<br>• Negative T waves in right precordial leads<br>• Early repolarization<br>• Incomplete right bundle branch block |

**Figure 3.** ECG findings in syncope. "Red light" might suggest malignant arrhythmia in certain contexts; "yellow light" might require a nonurgent evaluation in cardiology; "green light" indicates normal variants with no further management needed. AV, atrioventricular; bpm, beats per minute; ECG, electrocardiogram; PVC, premature ventricular complexes. *Manual measurement using tangent; see Figure 4.

There are few data about the prognosis of young patients with VVS. Although there are no prospective long-term studies, there is a general sense that pediatric VVS resolves, but that before this, it can be recurrent and troublesome. The only predictor of recurrence is the recent syncope frequency.[33] The likelihood of recurrence decreases markedly after a proper assessment and reassurance, and the recurrence rate is proportional to syncope frequency in the preceding year. Generally, patients have a 20%-50% likelihood of having at least 1 more syncopal spell in the year after assessment. Routine follow-up in 3-6 months is reasonable.
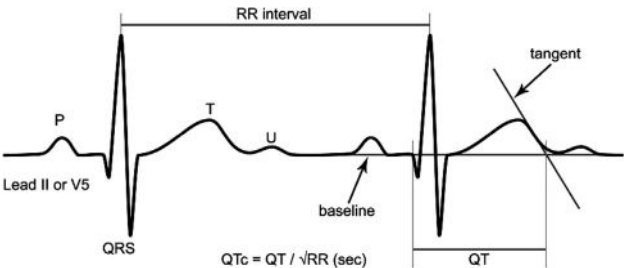


**Figure 4.** Measuring the QT interval. QTc is the heart rate corrected using the Bazett formula with use of the preceding RR interval. To determine QT, a tangent is drawn to the steepest slope of the last limb of the T wave in lead II or V$_5$. The end of the T wave is the intersection of the tangent with the baseline. Modified from Postema et al.[24] with permission from Elsevier.

Young people with a classic history of VVS should best be managed by their primary care physician. For patients with significant issues around quality of life, return to activity, or anxiety, referral to a clinic with expertise in syncope is warranted, with concurrent initiation of education and preventative measures.

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**RECOMMENDATION**

8. For children with syncope and a history atypical for VVS, a family history of arrhythmia or epilepsy, relevant abnormalities in physical examination, or an abnormal ECG, we recommend referral to a specialist with expertise in syncope (Strong Recommendation; Low-Quality Evidence).

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## Acknowledgements

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## Supplementary Material

To access the supplementary material accompanying this article, visit the online version of the *Canadian Journal of Cardiology* at www.onlinecjc.ca and at http://dx.doi.org/10.1016/j.cjca.2016.09.006.