APPROACH TO PEDIATRIC SYNCOPE

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November, 2020

Disclosures

- None

Objectives

- Develop a systematic approach to the management of pediatric patients presenting with syncope
- Be able to diagnose vasovagal syncope based on history and physical exam
- Identify red flags that will warrant further investigation or referral to specialist
- Become familiar with treatment of vasovagal syncope

Scope of the Problem

- 15-50% of normal children experience at least one syncopal event in their life
- 1% of all pediatric ER visits
- True incidence unknown as many do not seek care
- Peak in adolescence
- Lesser peak in 6-18 month-olds
- Mostly benign
- Need to exclude serious causes (cardiovascular, neurological)

Canadian Journal of Cardiology 33 (2017) 189–198

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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2016 CCS/CPS Position Statement

- Clinical guideline for evaluation of syncope
- Pediatric age (less than 19 years old)
- Acute or primary care setting
- Review of best available evidence and clinical experience
- Represents the consensus of a Canadian multidisciplinary expert panel

Goals

- Help practitioners recognize syncope due to an etiology other than transient autonomic system dysfunction
- Encourage a safe, efficient and cost-effective disposition in patients with evidence of a benign cause

Disclaimer

- Statement not a substitute for clinical judgment
- Adherence to recommendations may not always result in a successful outcome

Definition of Syncope

- Transient loss of consciousness (LOC) and inability to maintain postural tone
- Rapid and spontaneous recovery
- Absence of clinical features specific for another cause of LOC (ie epileptic seizure)
- Caused by global cerebral hypoperfusion

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 • 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 • Vascular events: subclavian steal phenomenon, vertebrobasilar insufficiency • Disrupted cerebrospinal fluid circulation: colloid cyst of the third ventricle, posterior fossa tumour • Vertiginous drop attack • Basilar migraine • 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	

Common Causes of Syncope in Children

Cause	Incidence
Vasovagal	64-73%
Breath-holding spells	6.4%
Cardiac	2.9-4.8%
Neurological	2.1-4.6%
Metabolic	0.8%
Psychiatric	2.2-2.3%
Unknown	8.2-18.9%

Cardiac Causes

- Primary electrical
  - Long QT / Short QT
  - Brugada syndrome
  - Catecholinergic polymorphic ventricular tachycardia
  - Wolff-Parkinson-White syndrome

- Structural heart disease
  - Hypertrophic / Dilated cardiomyopathy
  - Coronary artery anomalies
  - Arrythmogenic right ventricular cardiomyopathy
  - Severe valvular aortic stenosis
  - Pulmonary hypertension
  - Acute myocarditis

Neurological Causes

- Seizures
- Vascular events
  - Subclavian steal phenomenon
  - Vertebrobasilar insufficiency
- Basilar migraine
- Vertiginous drop attack
- Narcolepsia cataplexia

Other Causes

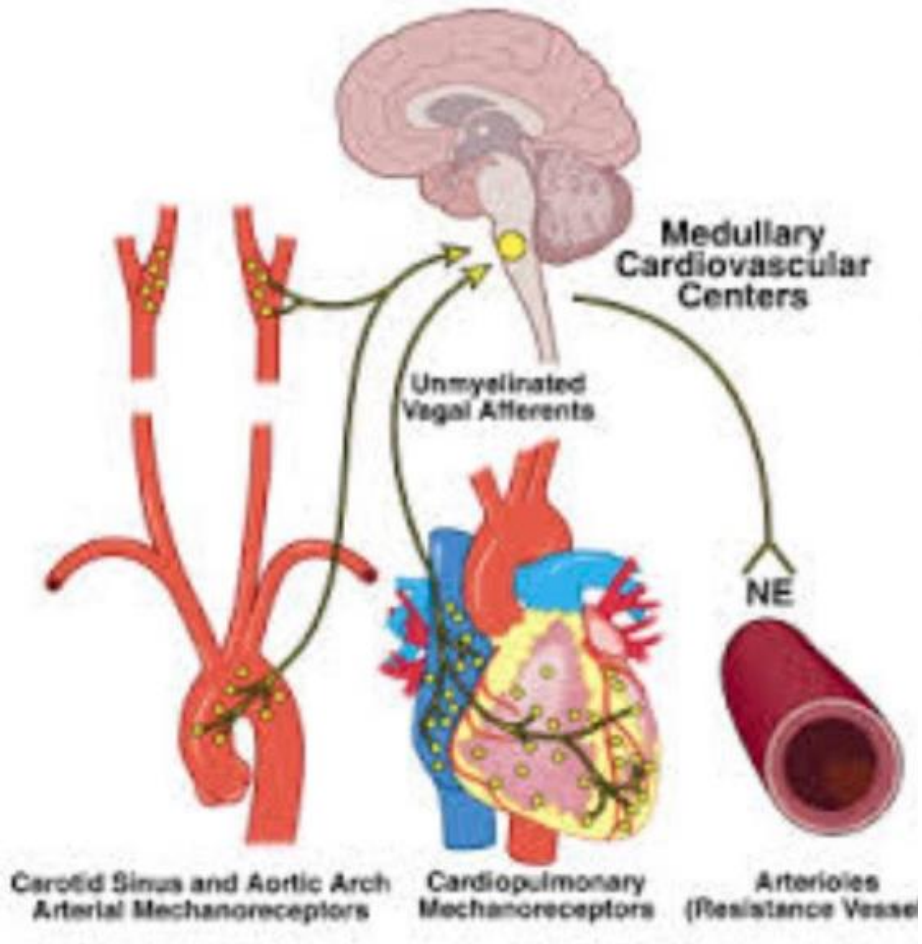
- Metabolic
  - Bleeding
  - Dehydration
  - Hypoglycemia
  - Electrolyte disturbances
- Psychiatric
  - Conversion disorder
  - Somatization
  - Munchausen syndrome
  - Anxiety-hyperventilation syndrome
- Unknown

Vasovagal Syncope (VVS)

Also “fainting”, reflex syncope, neuro-cardiogenic syncope

- Transient autonomic system dysfunction
- Definition
  - Occurs with upright posture held >30 sec **OR** with exposure to emotion/ pain/ medical setting
  - Diaphoresis, warmth, nausea, pallor
  - Associated with hypotension and relative bradycardia
  - Followed by fatigue

Pathophysiology of VVS



Decrease in venous return (reduced preload)

↓

Reduced ventricular filling

↓

Increased sympathetic tone

↓

Hypercontractility of ventricles with underfilled chamber

↓

Ventricular mechanoreceptor activation

↓

Feedback to medulla (CNS) via afferent vagus nerve

↓

Sympathetic withdrawal, parasympathetic overdrive

↓

Bradycardia and hypotension

↓

SYNCOPE

Symptoms

- Prodrome (!!!)
  - Warm/ clammy, nausea, lightheadedness, visual changes
  - Irritability, confusion, auditory changes, dyspnea, abdominal symptoms
- Absence of prodrome suggests cardiac cause
- Palpitations / chest pain
  - Do not always indicate a cardiac etiology
  - May represent adrenergic surge preceding vagal response

I bet he wouldn’t faint if the needle had some ink in it!

Circumstances

- Mid-exertional vs post-exertional
- Change in position
- Poor hydration or nutrition
- Phlebotomy, blood, injury, hair grooming, micturition/defecation, emotion, pain, intercurrent illness (GI), hot or crowded environment

Collateral History
Interview witnesses!

- Duration of LOC
  - Less than 1 minute
  - Prolonged LOC suggests non-cardiac (neuro, psych)
- Degree of intervention required
- Change in colour
- Involuntary movements
  - Single twitch to violent jerks, not rhythmic, < 30 sec
  - LOC **precedes** movements

Medical & Family History

- Beta-blockers, Ca channel blockers, diuretics
- Other medications or drug use
- Previous syncope, heart disease, diabetes, seizures, psychiatric problems
- Family history of sudden death, unexplained accidents, arrhythmia, structural heart disease, seizures, migraine

Suggested Approach

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.

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.

RECOMMENDATION

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

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

History and Physical Findings	RED LIGHT Recommend Urgent Referral to Syncope Specialist	YELLOW LIGHT Consider Further Investigations or Semiurgent Referral to Syncope Specialist	GREEN LIGHT 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) Syncope while swimming (might be associated with LQTS)		Postexertional syncope 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 Palpitations just before syncope	
Position of child preceding event		Supine (consider seizure)	Prolonged or recent standing 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 Seizures	Diabetes Psychiatric disorder or medications Significant comorbidities	No relevant medical history Previous events consistent with vasovagal syncope or breath-holding spells
Family history (structural cardiac disease, arrhythmias, sudden death, migraines, or seizures)	Sudden death Arrhythmias	Seizures Structural heart disease	Vasovagal syncope Migraines
Focused cardiac and neurologic examinations	Pathologic murmur Sternotomy scar Persistent neurologic deficits (consider stroke, seizure, migraine)		Normal examination Typical flow murmur

Investigations

- Cost of diagnostic testing in syncope > $1000 per patient (1090s study)!
- Testing should be guided by history & physical
- No further testing needed in typical VVS
- ECG is very low yield and non-specific: routine use in acute setting not supported

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.

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)

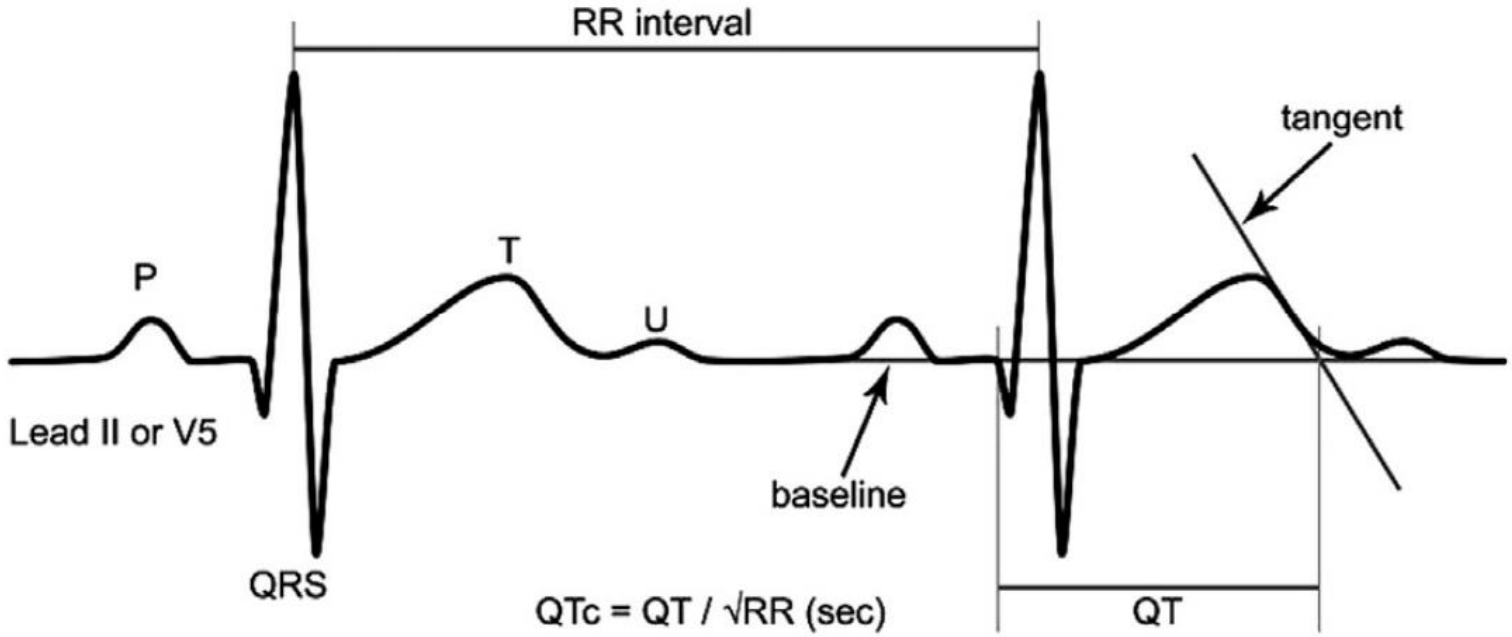


Figure 4. Measuring the QT interval. QTc is the heart rate corrected using the Bazett formula with use of the preceding RR interval. To determine QT, a tangent is drawn to the steepest slope of the last limb of the T wave in lead II or V_5. The end of the T wave is the intersection of the tangent with the baseline. Modified from Postema et al.[24] with permission from Elsevier.

	ECG Findings

RED LIGHT	• Abnormal QT interval*
	  ○ Long QT interval (QTc > 470 ms)
	  ○ Short QT interval (QTc ≤ 330 ms)
	• Type 1 Brugada pattern
	• Delta wave (ventricular pre-excitation or Wolff-Parkinson-White syndrome)
	• Signs of myocardial ischemia (ST-T wave changes, Q waves > 1 mm wide)
	• PVCs, polymorphic
	• 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) • PVCs, monomorphic • Second-degree AV block • Heart rate < 40 bpm in normally nourished, nonathletic individual
GREEN LIGHT	• Sinus arrhythmia • Wandering atrial pacemaker; atrial or junctional rhythm • First-degree AV block • Negative T waves in right precordial leads • Early repolarization • Incomplete right bundle branch block

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).

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).

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.

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

RECOMMENDATION

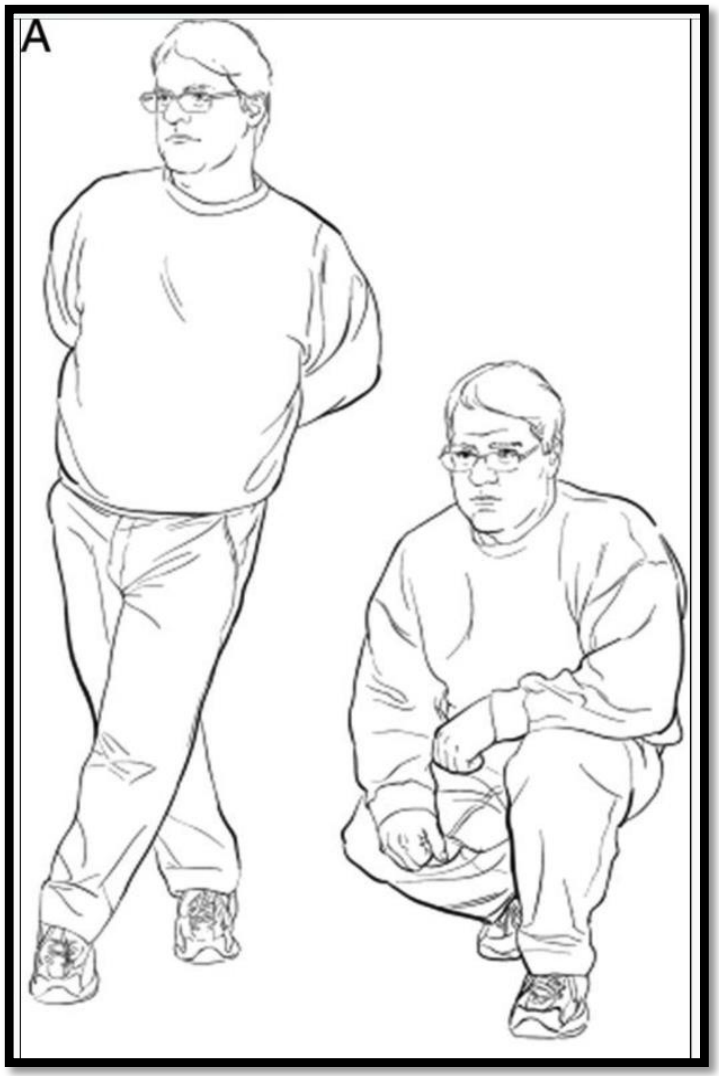
8. For children with syncope and a history atypical for VVS, a family history of arrhythmia or epilepsy, relevant abnormalities in physical examination, or an abnormal ECG, we recommend referral to a specialist with expertise in syncope (Strong Recommendation; Low-Quality Evidence).

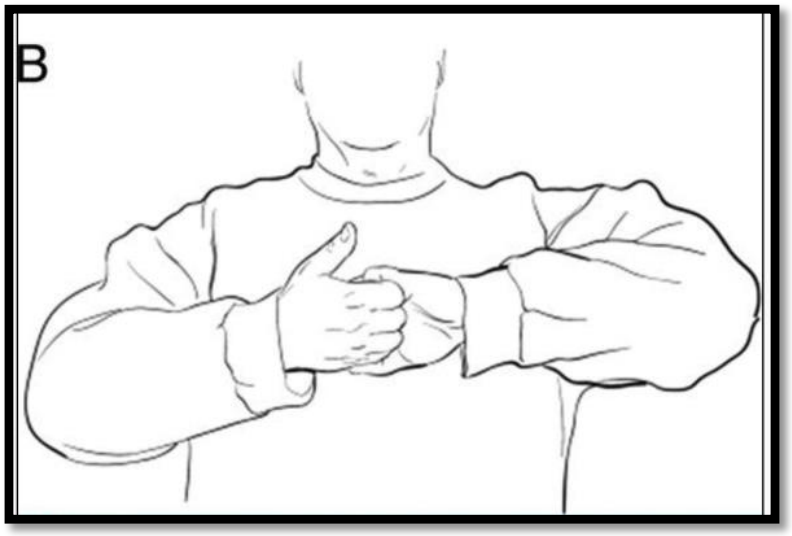
Management of VVS

- Children with VVS generally do well
- No need for hospitalization or intervention
- Provide explanations and reassurance
- Avoid provoking factors
- Educate patient to recognize prodrome
- Increase hydration and salt intake
- Teach preventative manoeuvers

Physical Manoeuvers to Counter VVS

Source: American College of Cardiology Foundation 2009





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).

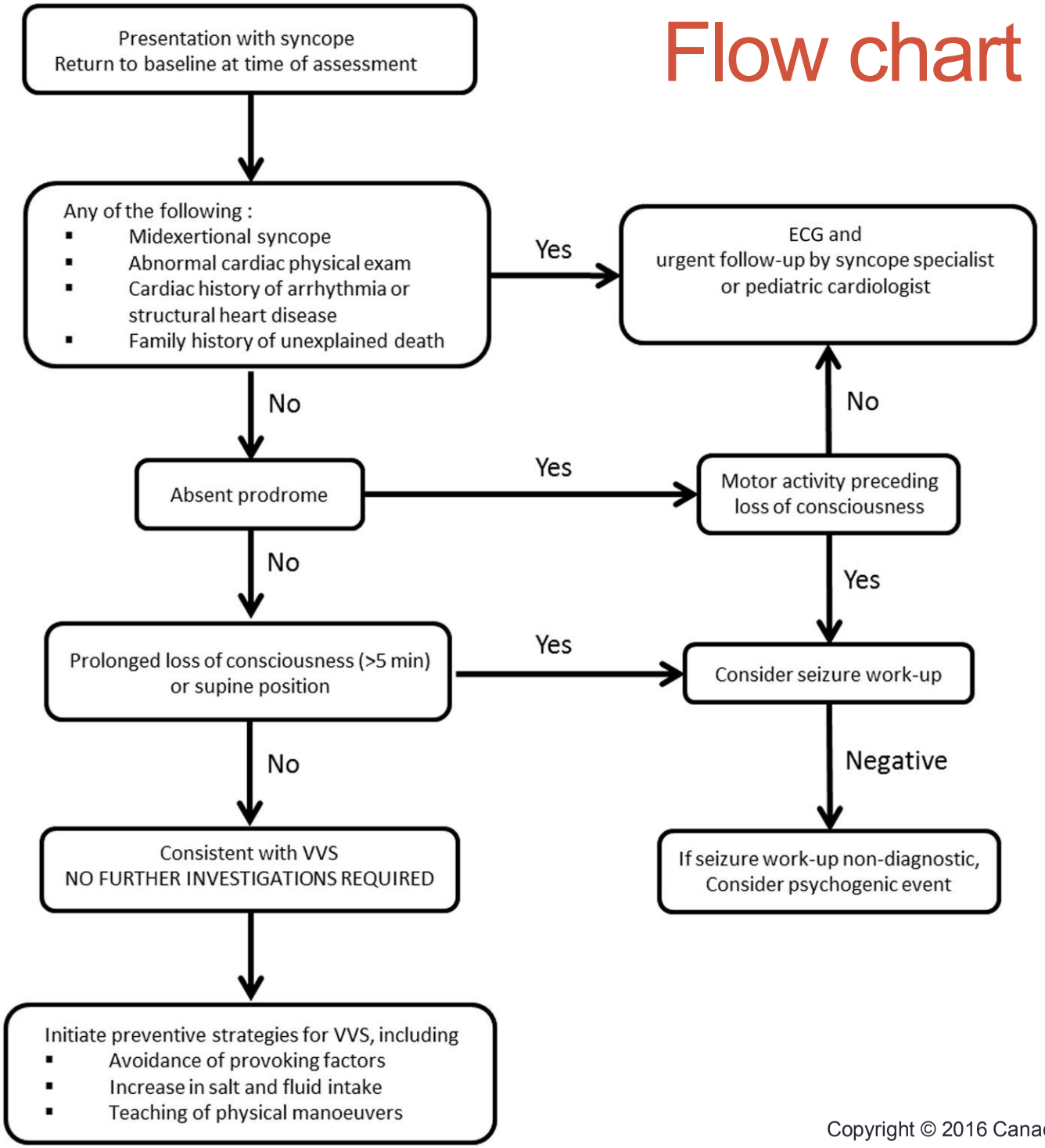
RECOMMENDATION

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

Pharmacological Treatment

- Medication rarely required
- Weak evidence to support the use of beta-blockers or fludrocortisone
- Small prospective study shows that midodrine prevents recurrent VVS in children refractory to conservative management for >6 months
  - Alpha1-agonist
  - Should be used ONLY if ADL significantly impaired
  - An increase in systolic blood pressure measured one minute after standing should be documented
  - Can cause marked elevation of supine blood pressure
  - Blood pressure should be carefully monitored during therapy
  - Should be stopped if no clinical effect

Flow chart



Summary

- Vasovagal syncope is a clinical diagnosis
- A detailed history is the most important part of the assessment
- Further investigations are not required as they are low yield and cost-inefficient
- Treatment of vasovagal syncope comprises education, salt and water intake, physical manoeuvres, and very rarely medication
- Red flags in history, physical, and ECG findings should trigger referral to the appropriate specialist

Thank you!