Syncope in Children

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ABSTRACT

Syncope, or fainting, is a heterogeneous condition and hence difficult to diagnose because of its underlying physiology. Between 20 to 40% of the population would have had a syncopal attack at some point of their life time, and some affected individuals suffer from frequent episodes. Presyncope, orthostatic dizziness which occurs prior to fainting is commonly reported by pediatricians and physicians, but its prevalence is unknown. Both syncope and presyncope affect the quality of life of those who have suffered from frequent attacks, thus prompting investigations into their cause and prevention. The autonomic nervous system is mainly implicated in the development of (pre)syncope. This paper focuses on syncope in children, the etiology and the possible emergent treatment modalities.

Keywords: Syncope, Children, Dizziness, Arrhythmia, Reflex syncope.


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INTRODUCTION

Syncope is an abrupt and transient loss of consciousness associated with loss of postural tone that follows a sudden fall in cerebral perfusion; is a common, important medical problem caused by many conditions, ranging from benign and self-limiting to chronic, recurrent and even potentially fatal causes. Unfortunately, the differentiation between benign and malignant causes is difficult and quite challenging. Recurrent syncope is more common than epilepsy and accounts for 3% in emergency department and 1% of hospital admissions.1 There is a bimodal age distribution of episodes of both syncope and Presyncope; children and adolescents2 have a higher prevalence (first peak) and in the second peak which primarily occurs in the elderly age group.3-5 The main difference between the two age groups is the possible underlying mechanism which precipitates an attack. In children and adolescents, syncope generally occurs secondary to an inappropriately triggered cardiovascular reflex response; however in elderly, the etiology is more diverse. Females in all age groups are more prone to syncope than males, though the reasons are unclear.6

More often; syncope is often misdiagnosed and erroneously treated as an epileptic condition.7 Management of this condition can many a times be frustrating, confusing and often unrewarding; and the treatment will be impossible to achieve without understanding the cause behind it. The diagnosis relies mainly on the history, but more often, investigations may be required to support the diagnosis. It is imperative for a physician to distinguish between the benign, e.g. vasovagal syncope, and the potentially life-threatening, e.g. cardiac syncope (Fig. 1). With modern science and its advancements, fortunately, experienced, astute and highly-skilled physicians can deliver effective care when a careful attention is paid in full detail.

DEFINITION OF SYNCPE

Syncope (derived from the Greek words, ‘syn’ meaning ‘with’ and the verb ‘kopto’ meaning ‘I cut’ or more appropriately ‘I interrupt’) is a symptom, defined as a transient, self-limited loss of consciousness, usually leading to falling. The onset of syncope is relatively rapid, complete and usually prompt.7-11

Syncope can also be defined as transient loss of consciousness and postural tone due to transient global cerebral hypoperfusion (TGCH), fast start, short duration and complete and spontaneous recovery.12,13 According to this definition, those pathologies which does not involve a TGCH (epileptic attacks, psychogenic disorder) are excluded.

CLASSIFICATION, CATEGORIZATION AND CAUSES OF SYNCPE (REFER TABLE 1 to 3)14-16

Syncope must be differentiated from other nonsyncopal conditions associated with real or apparent transient loss of consciousness.

- Neurally-mediated syncope is a reflex response that, when triggered, gives rise to vasodilatation and/or bradycardia. But, the contribution of these factors to systemic hypotension and cerebral hyperfusion differs considerably.

- Presyncope describes the occurrence of warning symptoms without loss of consciousness; the symptoms include a faint or a dizzy feeling, nausea, feeling hot, cold or sweaty sometimes with a rushing noise in the ears.

- Mild syncope can be defined clinically by rapid recovery within seconds and neurophysiologically by an ictal/postictal EEG.

- Severe syncope can be defined clinically by a prolonged postictal cerebral impairment (confusion or sleep) that lasts minutes or hours and neurophysiologically by the ictal/postictal EEG demonstrating the ‘slow-fast-slow pattern’.
Syncope in Children

- The ‘classical vasovagal syncope’ is mediated by emotional/orthostatic stress and is often diagnosed during history taking.
- ‘Carotid sinus syncope’ occurs in close relationship to accidental mechanical manipulation of the carotid sinus massage.
- ‘Situational syncope’ is associated with specific scenarios such as coughing, micturition. Most often, neurally-mediated reflex syncopes have nonclassical presentations, which are diagnosed by minor clinical criteria or response to tilt test or cardiac massage.
- ‘Orthostatic hypotension’ refers to syncope in which the upright position causes arterial hypotension; this occurs when the autonomic nervous system is incapacitated and fails to respond to challenges imposed by the upright posture. The other important cause can be due to; volume depletion’ in which the autonomic nervous system is unable to maintain blood pressure due to decreased circulating volume.
- ‘Cardiac arrhythmias’ can cause a decrease in cardiac output, which usually occurs irrespectively of circulatory demands.
- ‘Structural heart disease’ can cause syncope when circulatory demands outweigh the impaired ability of the heart to increase its output.
- ‘Steal’ syndromes can cause syncope when a blood vessel has to supply both part of the brain and an arm.

PREVALENCE

It is not clear regarding the frequency of occurrence in children, partly due to the fact that most children with a single episode of syncope do not report for medical attention/help. One of the studies have reported that the incidence in children and adolescents to be 126/100,000.\textsuperscript{14}

It was also estimated that up to 15% of normal children would experience at least one syncope before the age of 18.\textsuperscript{14} The peak incidence occurs around the age of 15 to 19 years, most of them being neurocardiogenic (vasovagal) syncope with a predilection toward females.\textsuperscript{17}

NEUROCARDIOGENIC SYNOCOPE (VASOVAGAL SYNOCOPE OR THE COMMON FAINT)

This is caused by stimulation of the intramyocardial mechanoreceptor (stretch receptor or C fiber) as a result of

Table 1: Classification of syncopes by pathophysiology and causes\textsuperscript{14}

<table>
<thead>
<tr>
<th>Category</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Insufficient cerebral perfusion</td>
<td>Transient raised intracranial pressure (e.g. hydrocephalic attack due to papilloma of the third ventricle)</td>
</tr>
<tr>
<td></td>
<td>Transient isolated cerebral arterial hypotension (e.g. G-LOC, i.e. loss of consciousness associated with extreme gravitational force, vertebra-basilar insufficiency, adolescent stretch syncope)</td>
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<tr>
<td></td>
<td>Transient systemic arterial hypotension with</td>
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<tr>
<td></td>
<td>Reduced cardiac output</td>
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<td></td>
<td>Reduced stroke volume (e.g. from reduced venous return, tachycardia), e.g. hypovolemia, forced expiration (valsalva, expiratory apnea)</td>
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<tr>
<td></td>
<td>Bradycardia (e.g. heart attack)</td>
</tr>
<tr>
<td></td>
<td>Asystole (e.g. cardio inhibitory neurally mediated syncope/ reflex asystolic syncope)</td>
</tr>
<tr>
<td></td>
<td>Reduced vascular resistance (e.g. sepsis, anaphylaxis)</td>
</tr>
<tr>
<td></td>
<td>Reduced cardiac output and vascular resistance (e.g. vasodepressor and mixed neurally mediated syncope)</td>
</tr>
<tr>
<td>Hypoxemic cerebral perfusion</td>
<td>(e.g. acute left to right shunts, anesthetic, aviation, diving and mountaineering accidents)</td>
</tr>
<tr>
<td>Hypoxemic and insufficient cerebral perfusion</td>
<td>(e.g. expiratory apnea syncope or blue-breath holding)</td>
</tr>
<tr>
<td>Hypoxemic and/or insufficient cerebral perfusion</td>
<td>(not otherwise specified)</td>
</tr>
</tbody>
</table>
Table 2: Causes of syncope

(Source: ESC Guidelines; guidelines on management (diagnosis and treatment) of syncope update 2004 the task force on syncope, European Society of Cardiology, Europace 2004;6:467-537)

- Neurally-mediated (reflex)
  - Vasovagal syncope (common faint)
    - Classical
    - Nonclassical
  - Carotid sinus syncope
  - Situational syncope
    - Acute hemorrhage
    - Cough, sneeze
    - Gastrointestinal stimulation (swallow, defecation, visceral pain)
    - Micturition (post-micturition)
    - Postexercise
    - Postprandial
    - Others (e.g. brass instrument playing, weightlifting)
  - Glossopharyngeal neuralgia
- Orthostatic hypotension
  - Autonomic failure
    - Primary autonomic failure syndromes (e.g. pure autonomic failure, multiple system atrophy, Parkinson’s disease with autonomic failure)
    - Secondary autonomic failure syndromes (e.g. diabetic neuropathy, amyloid neuropathy)
  - Postexercise
  - Postprandial
  - Drug (and alcohol)-induced orthostatic syncope
  - Volume depletion
    - Hemorrhage, diarrhea, Addison’s disease
- Cardiac arrhythmias as primary cause
  - Sinus node dysfunction (including bradycardia/tachycardia syndrome)
  - Atrioventricular conduction system disease
  - Paroxysmal supraventricular and ventricular tachycardia
  - Inherited syndromes (e.g. long QT syndrome, Brugada syndrome)
  - Implanted device (pacemaker, ICD) malfunction
  - drug-induced proarrhythmias
- Structural cardiac or cardiopulmonary disease
  - Cardiac valvular disease
  - Acute myocardial infarction/ischemia
  - Obstructive cardiomyopathy
  - Atrial myxoma
  - Acute aortic dissection
  - Pericardial disease/tamponade
  - Pulmonary embolus/pulmonary hypertension
- Cerebrovascular
  - Vascular steal syndromes

Table 3: Causes of nonsyncopal attacks (commonly misdiagnosed as syncope)

(Source: Guidelines for the diagnosis and management of syncope (ver 2009); The task force for the diagnosis and management of syncope of the European Society of Cardiology (ESC) Developed in collaboration with, European Heart Rhythm Association (EHRA), Heart Failure Association (HFA), and Heart Rhythm Society (HRS): European Heart Journal 2009;30:2631-2671)

- Disorders without any impairment of consciousness
  - Falls
  - Cataplexy
  - Drop attacks
  - Psychogenic pseudosyncope
  - Transient ischemic attacks (TIA) of carotid origin
- Disorders with partial or complete loss of consciousness
  - Metabolic disorders, including hypoglycemia, hypoxia, hyperventilation with hypocapnia
  - Epilepsy
  - Intoxications
  - Vertebro-basilar transient ischemic attack

Vasovagal syncope is more likely to occur as a result of being tired, hungry, stressed, feeling crowded, with pain, forceful myocardial contraction from various etiologies, such as decreased cardiac preload from prolonged standing. The relative contributions of ‘vaso’ and ‘vagal’ components varies considerably from being predominantly ‘vaso’ (as in vasodepressor syncope) to exclusively ‘vagal’ (as in reflex asystolic syncope).
sight of blood or injury, unwell, fright, anemic, dehydrated, immediately after exercises with combinations of such provoking or risk factors. These risk factors comprises emotional or psychological stress, hypotensive effects of hypovolemia or vasodilation and orthostatic challenge. Because there is a clear provocation, some authors prefer to call this as ‘reflex syncope.’

- **Bezold-Jarisch reflex:** A neural impulse from the myocardium is sent to the brainstem, causing a reflex that leads to a withdrawal of sympathetic stimulation and/or an increase in vagal tone, resulting in paradoxical bradycardia and/or vasodilation. The resultant mechanism is called Bezold-Jarisch reflex and this is commonly seen in children standing in school during hot weather or while standing at a church.

**CARDIAC SYNCOPE**

Heart diseases causing syncope in children are generally severe structural defects, coronary anomalies or aneurysm, or are arrhythmia syndromes that causes ventricular tachyarrhythmia. Any of these etiologies can be lead to sudden death. Some of the examples are severe pulmonary hypertension, cyanotic spells, sick sinus syndrome, heart block, long QT syndrome, Wolff-Parkinson-White syndrome, coronary obstruction post Kawasaki disease, etc.

These patients usually have a left or right heart outflow obstruction with a loud heart murmur (E.g. severe aortic stenosis and severe pulmonary stenosis); severe pulmonary hypertension or any coronary anomaly such as the presence of a left main coronary artery (LMCA) arising from right sinus of Valsalva. The LMCA in this condition may take a course in-between the aortic root and the main pulmonary artery. Expansion of both great arteries with exercise can compress the LMCA and can cause cardiac ischemia.

Very rarely cardiac syncope in myocardial bridging, coronary spasms, coronary-cameral fistula or in coronary atherosclerotic disease. However, for this to happen there should be a familial history of hypercholesteremia or progeria.

**CHRONIC ORTHOSTATIC INTOLERANCE SYNCOPE**

Orthostatic intolerance is seen in some patients with recurrent vasovagal syncope. Chronic orthostatic intolerance is defined as the occurrence of various symptoms including syncope on standing for a duration of at least 3 months. The other features include symptoms of Presyncope: lightheadedness, ‘dizziness’, blurred vision and chronic fatigue. Common features include migranous headache, nausea, chest and abdominal discomfort, palpitations, shortness of breath, hyperventilation, peripheral cyanosis, etc. Chronic orthostatic intolerance is often part of the clinical picture of chronic fatigue syndrome and it may be helpful to consider this disorder as a differential of idiopathic chronic fatigue syndrome.

**REFLEX ASYSTOLIC SYNCOPE OR REFLEX ANOXIC SEIZURES**

Reflex anoxic seizures is the preferred term for paroxysmal episodes of collapse (usually with stiffening or jerking) provoked by pain or surprise in children and some adults. The term ‘infantile vasovagal syncope’ has also been suggested, although there is no evidence of a ‘vaso’ component. Older children and adults with the same disorder have ‘convulsive cardioinhibitory NMS’.

In a typical attack, a toddler will have had a bump to his head or any other unexpected, sudden pain or shock. He will cry out and fall limply to the floor within a few seconds. Because there is no warning, injury is common, although usually mild. The child will appear deathly pale and ‘lifeless’ for around 5 to 30 seconds (asystole lasting 5-30 seconds). Frequently, there will be, an upward eye gaze and stiffening of the trunk in extension, typically with the upper limbs extended at the elbows, wrists and hands clawed in a dystonic posture for several seconds, sometimes with asymmetric limb jerking. During the extensor spasm however, there is apnea. After a minute or so, the child comes in a rather confused or distressed state before finally going off to sleep for an hour or so.

The resemblance to epileptic seizures is clear enough and traditionally, this has been viewed only as an important exclusion in the diagnosis of pediatric epilepsy. The fact that it falls between pediatric neurology and cardiology has added to this reluctance.

**CYANOTIC SPELL SYNCOPE**

Severe cyanosis from congenital heart disease can result in the loss of consciousness and postural tone. The child usually has visible cyanosis and examination reveals abnormal heart sounds.

**BLOOD INJURY PHOBIA SYNCOPE**

Blood injury phobia (BIP) demonstrates transient tachycardia followed by bradycardia and even Asystole after exposure to the feared stimuli. Here the child will link blood or injury to the previous unpleasant reaction and starts to display phobic avoidance behaviors. The clinical histories when corroborated are relative with symptoms of nausea, flushing, sweatiness and perceptual disturbance. The
investigation of choice is head-up tilting, which typically causes bradycardia and hypotension, with a preceding tachycardia.

**OTHER FORMS OF RARE SYNCOPE IN CHILDREN**

- Vagovagal syncope (here there is direct vagal stimulation that occurs with swallowing, coughing, vomiting or gastrointestinal reflux or even during intubation).
- Apnoeic syndrome (also known as reflex prolonged expiratory apnoea syndrome or reflex apnoeic syncope or blue-breath-holding spells).
- Vascular syncope which is caused by a sudden drop in blood pressure (e.g. hypovolemic shock from a sudden hemorrhage or extensive burns or in septicemic shock).
- Cerebral syncope occurs due to impaired cerebral blood flow in the presence of a normal systemic blood pressure and cardiac output.

**GUIDELINES FOR ASSESSMENT OF SYNCOPE IN CHILDREN**

- History
  - Presyncopal activity and position
  - Presyncopal associated symptoms
  - Precipitating factors for vasovagal syncope
  - Estimated ‘Duration of Warning’-this is one of the most important source of information and are characteristic of cardiac, orthostatic or micturition syncope.
  - Estimated loss of consciousness
  - Reports from witness (including convulsions, loss of urine)
  - Post syncopal residua
  - Family history (unexplained death, deafness, congenital heart diseases, metabolic disorders, MI at young age)
  - Medications (certain drugs causes prolongation of QT interval and predisposes to Toursades de Points)
- Physical
  - Orthostatic blood pressure/pulse
  - CVS examination (listen carefully to the murmur of aortic stenosis)
  - CNS examination
  - Stool for OB
- Investigations
  - ECG monitoring (15-20% yield). ECG yields the following:
    i. Arrhythmias (e.g. bradycardia, atrial fibrillation)
    ii. Heart blocks
    iii. Interval abnormalities: short PR segment (sign of WPW or other bypass diseases, prolonged QT interval which is a risk factor for Toursades de Points)
    iv. Sinus tachycardia
    v. Signs of ischemia

The choice of further investigations depend on the differential diagnosis. EEG and neurologic referral is indicated in a patient who is suspected of having a seizure. Holter monitoring and event recorder (loop recorder) is useful to diagnose arrhythmias and cardiac MRI is useful to diagnose arrhythmogenic right ventricular dysplasia or other types of silent cardiomyopathy. Finally, an invasive cardiac catheterization, angiography and electrophysiological study may be needed in rare cases.

**TILT TABLE TESTING**

Head up passive tilt testing (HUTT) has assisted the diagnosis of vasovagal syncope and the test protocol typically involves testing the patient in the morning having fasted. After lying supine for 30 minutes, the patient is tilted to 60 to 80° for <45 minutes and asked to report any symptoms. A test is positive only if the patient’s original presyncopal symptoms are reproduced entirely, and accompanied by arterial hypotension, bradycardia or both.

HUTT is contraindicated in proximal coronary artery disease, critical mitral stenosis, clinically severe left ventricular outflow obstruction and severe cerebrovascular disease.

**OTHER INVESTIGATIONS**

- Carotid sinus massage
- Cardiac electrophysical (EP) study
- Brain imaging
- Carotid imaging
- Autonomic function tests
- Hyperventilation tests

**CONCLUSION**

Recurrent syncope can be distressing and disabling for a child and his family. Although many forms of syncope have an underlying benign cause, there are potentially life-threatening differentials that must be considered when assessing a child with syncope. Timely and selected investigations helps to elucidate the underlying mechanism and improves the treatment options. A detailed history and most importantly an ‘eye-witness account’ are critical to save inappropriate and potentially confusing syncopal conditions.
REFERENCES

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