

Syncope Attacks Imitating Epileptic Seizures

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Syncope is among the most common non-epileptic attacks misdiagnosed as epileptic seizures, including GTCs.^{1,25-39}

A *syncope* is defined as a paroxysmal event of loss of consciousness and postural tone caused by cerebral hypoperfusion with spontaneous recovery.³⁸ There is an abrupt cutting off of the energy substrates to the cerebral cortex, usually through a sudden decrease in cerebral perfusion by oxygenated blood.³¹ If cerebral perfusion/oxygenation is cut off for a period of 8–10 s, then a clinical picture comprising loss of consciousness and postural tone, pallor and sweating, brief (lasting seconds) extensor stiffening or spasms, and a few irregular myoclonic jerks of the limbs may occur. The whole episode is brief, usually lasting less than 10 s. There is a great variety in the amplitude of the myoclonic jerks, the degree of stiffening and the recovery time after syncope.³¹

Cerebral syncope is defined as a syncope that results from derangement of cerebral autoregulation leading to cerebral vasoconstriction with resultant cerebral hypoxia in the absence of systemic hypotension.

Convulsive syncope is a term used for any type of syncope manifesting with convulsive movements.

Prodromal warning symptoms (presyncope) are commonly present but sometimes these are only recalled when syncope is reproduced, as in the head-up tilt test. They develop over 1–5 min and include light headedness, nausea, a feeling of warmth, sweating, palpitation, greying or blacking of vision, muffled hearing and feeling distant. At this stage a syncope may be averted by lying down in a horizontal position with the head down and legs up. Lack of prodromal symptoms, such as in a tussive syncope, are explained by the rapidly developing cerebral hypoperfusion.

The setting and stimulus are the most important identifiable factors/precipitants in allowing the presumptive diagnosis of syncope. Simple faints or vasovagal syncope most often occur upon getting up quickly or after prolonged standing, particularly if associated with peripheral vasodilatation (e.g. hot, unventilated, crowded places, or after drug or alcohol use) or increased vagal tone (e.g. bloody, terrifying, or obnoxious scenes, and painful stimuli).

Cerebral hypoperfusion most commonly results from conditions that decrease venous return (e.g. reflex-mediated vasomotor instability) or disorders that decrease cardiac output, such as primary cardiac disorders.

<p>Precipitating factors or triggers such as upright position, bathroom, crowded and humid places, lack of food, unpleasant circumstances, venipuncture</p> <p>Prodromal symptoms of cerebral ischaemia, such as dizziness, greying of vision and tinnitus</p> <p>Gradual onset over seconds to 1 min</p> <p>Pallor and sweating</p> <p>Lack or rare occurrence of convulsions (other than myoclonic jerks), urinary incontinence or tongue biting</p> <p>Brief duration (1–30 s)</p> <p>Rapid recovery with no post-ictal confusion</p>
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Table 2.1 Commonly emphasised clinical manifestations of syncope

<p>Convulsions occur in 70–90% of syncopes; symptoms include myoclonus, tonic flexion or extension, more complex movements and automatisms such as lip licking, chewing or fumbling</p> <p>Visual hallucinations (a perception of grey haze, coloured patches, glaring lights or more complex scenes involving landscapes, familiar situations or people) and, less often, auditory hallucinations (rushing and roaring sounds, traffic and machine noises, and talking and screaming human voices, but never intelligible speech) are frequent (60%) in both convulsive and non-convulsive syncope⁴⁰</p> <p>Syncope usually happens in an upright position but may also occur in the supine position (e.g. venipuncture)</p> <p>Sudden onset, urinary incontinence and trauma are not uncommon</p> <p>Abdominal pain that may be confused with epigastric aura may occur at onset. Auras comprising epigastric, vertiginous, visual or somatosensory experiences occur both in neurally mediated and cardiogenic syncope⁴¹</p> <p>Pallor and sweating are not invariable symptoms at onset and may be symptoms of autonomic epileptic seizures with or without secondarily GTCSs</p> <p>Complete recovery may not be rapid and post-ictal confusion may occur, although neither of them reaches even close to the severity of that after a GTCS</p> <p>Eyes, as in GTCSs, are always open during syncope and the most consistent oculomotor sign is an upward turning of the eyes early in its course, which may be followed by lateral eye deviation^{42,43}</p>
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Table 2.2 Clinical features of syncopes not emphasised in their differentiation from epileptic seizures

In general, syncopes are categorised into:

- Neurally mediated syncopes (neurocardiogenic or reflex-mediated syncope) that are usually benign
- Cardiogenic syncopes from either cardiac rhythm or structural cardiac disorders, which are potentially life threatening

Autonomic disturbances may also lead to cerebral hypoperfusion and fainting as with orthostatic syncope.

In differentiating syncopes from GTCSs, textbooks usually emphasise the characteristics of typical syncopes that are listed in Table 2.1.

Based on a proper synthesis of the quality and chronological order of the clustering of these symptoms, the differential diagnosis of syncopes from GTCSs is rarely a problem. However, there are important points to consider and these are listed in Table 2.2.

Neurally Mediated Syncope

Examples of neurally mediated or non-cardiogenic syncopes include:

- Vasovagal syncope (simple faint) – variants of which may include reflex anoxic seizures (reflex asystolic syncope) and cyanotic breath-holding attacks (of prolonged expiratory apnoea) in infants and children
- Situation-related syncope caused by increased intrathoracic pressure, as a result of, for example, cough (tussive syncope), Valsalva manoeuvres, or straining to void (micturition syncope) or defecate
- Carotid sinus syncope

Vasovagal Syncope

Vasovagal syncope is the most common and familiar form of neurally mediated syncope and results from a combination of excessive vagal tone, abnormal catecholamine response to stress, venous pooling during an upright stance and impaired cardiac filling. Episodes may begin in infancy, sometimes with reflex anoxic seizures, and thereafter are seen at all ages, although it predominates in otherwise normal children and adolescents. The frequency of vasovagal syncopes varies considerably from one to two during a lifetime to as common as more than once a day.

Reflex Anoxic Seizures

Reflex anoxic seizures (synonyms: *pallid breath-holding attacks*, *pallid infantile syncope*) are reflex asystolic syncopes in young children. An unexpected bump to the head, an occasional fright or seeing blood triggers a neurally mediated vagal discharge leading to severe bradycardia, asystole, syncope and anoxic seizure. The child falls unconscious, white as a sheet and looks dead.

Useful Clinical Note

Breath-holding attacks in children

In children, there are two main types of so-called 'breath-holding attacks':

- Cyanotic or blue breath-holding attacks (prolonged expiratory apnoea usually without syncope)
- Pallid or white breath-holding attacks (reflex anoxic seizures, which are reflex asystolic syncopes)

The term 'breath-holding' has been discouraged by Stephenson and Zuberi because it erroneously implies a voluntary action to obtain gains or a behavioural disorder; psychological disorders in those afflicted do not differ from those in control children.³⁷ In contrast to voluntary breath holding that occurs during inspiration, cyanotic breath-holding attacks occur during expiration.

The cyanotic or blue breath-holding attacks, which are more common than the pallid forms, are purely respiratory consisting of prolonged expiratory apnoea without any change in cardiac rate or rhythm. Attacks of prolonged expiratory apnoea have similar triggers to reflex anoxic seizures, but they usually occur when the child has reasons to suddenly become angry or frustrated or fearful. The child stops breathing and becomes cyanotic until spontaneous recovery with a deep breath.

Mixed breath-holding of expiratory apnoea and a degree of bradycardia or cardiac asystole occur.³⁷

Clarification on Terminology

- Reflex anoxic seizures are syncopes (and not epileptic seizures).
- Anoxic epileptic seizures are epileptic seizures caused by syncopes.
- Cyanotic or blue breath-holding attacks are not syncopes.

Orthostatic Syncope

Orthostatic syncope (autonomic failure) results from failure of normal mechanisms to compensate for the temporary decrease in venous return after standing.

Syncope occurs within seconds or minutes of becoming upright, especially when rising and after meals. Unlike with reflex vasovagal syncope, the skin stays warm, the pulse rate is unchanged despite the fall in blood pressure, and sweating is absent. Assuming a horizontal position results in complete recovery. Causes include autonomic dysfunction, cardiovascular disorders and drugs. Orthostatic syncope secondary to autonomic failure is rare in childhood. Dopamine β -decarboxylase deficiency is a possibility in such a clinical situation.

Syncopes Induced by Valsalva Manoeuvre

During a Valsalva manoeuvre (powerful effort to exhale against a closed glottis) increased intrathoracic pressure limits the venous return to the heart and increases vagal tone, resulting in decreased cardiac outflow and syncope.

Rarely, patients may have self-induced syncope by Valsalva manoeuvre. This particularly occurs in children with learning disabilities and Rett syndrome. The anoxic attacks may be severe.

Micturition Syncope

Micturition syncope is a reflex-mediated situational syncope usually occurring in men while standing for nighttime micturition.

Several mechanisms act in concert:

- Postural – standing on leaving a warm bed causing hypotension
- Straining – Valsalva manoeuvre increasing an already high nocturnal vagal tone, causing bradycardia
- Emptying bladder – abrupt decrease in stimulus to bladder stretch receptors causing reflex vasodilatation and hypotension

Carotid Sinus Syncope

Carotid sinus hypersensitivity is a common cause of unexplained falls in elderly people of over 50 years of age.^{44,45} The incidence steeply increases with age. Activation of one or both carotid sinuses causes peripheral vasodilation, hypotension and syncope in people with carotid sinus hypersensitivity. Clinical attacks of syncope or falls without definite loss of consciousness are attributed to carotid sinus pressure by head turning or tight collars. Some patients may suffer from orthostatic hypotension but usually there is no evidence of sympathetic or parasympathetic failure.

Diagnostic carotid sinus massage may be positive in asymptomatic elderly patients⁴⁵ and carries a risk of prolonged asystole, transient or permanent neurological deficit, stroke and sudden death.

Cardiogenic Syncope

Cardiogenic syncope result from either rhythm (e.g. tachyarrhythmias or bradyarrhythmias) or structural (e.g. aortic or mitral stenosis, intracardiac tumours, cardiomyopathy, ischaemic heart disease) cardiac disorders. Cardiogenic syncope are potentially life threatening and may be treatable. Morbidity and mortality is up to 50% within the first 3 years following the initial attack, and it is cause-dependent.

Palpitations, chest pain, shortness of breath, extreme fatigue or other features of cardiovascular insufficiency occur with other presyncopal symptoms of simple faints. Cardiac syncope occur from any posture (e.g. arrhythmogenic syncope is common in bed), and during periods of high exertion or emotion. Anoxic seizures precipitated by exercise require the urgent exclusion of cardiac causes, although most turn out to be neurally mediated syncope.

In tachyarrhythmias, a heart rate of more than 150–180 beats/min prevents adequate ventricular filling. In bradyarrhythmias, a heart rate of less than 30–35 beats/min prevents adequate cardiac output.

Attacks due to transient complete heart block are abrupt and short with rapid loss of consciousness. Lack of cardiac output may be due to short episodes of ventricular tachycardia or fibrillation. Prolongation of the QT interval may lead to such events. Attacks may be preceded by palpitations, extreme fatigue or presyncopal features. Mitral valve prolapse and aortic stenosis may present with episodic loss of awareness due to fluctuating cardiac output or associated arrhythmias. Aortic stenosis and hypertrophic cardiomyopathy is especially prone to episodes of sudden collapse with loss of awareness during exercise. Sometimes ventricular tachyarrhythmias occur with normal QT intervals.

The Long QT Syndrome

Of various cardiogenic syncope, the long QT syndrome is of particular significance because it may be associated with convulsive syncope that cause unexplained sudden death in a young person and closely imitate GTCSs. It is often of autosomal dominant inheritance with mutations in genes encoding

potassium or sodium channels, depending on lineage. The mechanism of the syncope is a ventricular tachyarrhythmia, normally torsades de pointes triggered by fear or fright, particularly during exercise (especially when that exercise is emotionally charged) and during sleep.

The ECG is diagnostic. Genetic testing is available.

A 33-year-old apparently normal man had, over a period of 6 months, three unwitnessed rather severe falls due to loss of consciousness that resulted in head trauma. All of them occurred while cycling or jogging in days that he recalls were particularly intense. EEG was normal but a synchronous ECG showed long QT syndrome, verified with appropriate cardiological evaluation.

Brugada syndrome, characterised by 3 types of ST-segment abnormalities, is another genetic ion channelopathy (usually SCN5A mutation) that may imitate epileptic attacks and other causes of sudden death. Genetic testing is available.

Useful Note

A proper ECG should be obtained in any patients suspected or newly diagnosed with epilepsy. Misdiagnosis of cardiac diseases as epilepsy is common and often difficult to recognise on clinical information alone. Therefore a proper ECG is mandatory for this crucial differentiation. Also consider that the adverse cardiac effects of some AEDs may have fatal consequences for patients with conditions such as long QT or Brugada syndrome and other disorders of cardiac conductivity.

Syncopes Induced by Drugs and Electrolyte Abnormalities

Drugs and electrolyte abnormalities are common causes of syncope secondarily to cardiac rhythm changes or autonomic disturbances. Some examples include beta-blockers, quinidine, calcium channel blockers, digoxin, hypomagnesaemia and hypokalaemia.

Syncopal Attacks Provoking Epileptic Seizures: Anoxic Epileptic Seizures

Occasionally, but likely more often than is reported, true epileptic seizures are triggered by non-epileptic syncopes in children and adults.^{39,46} This combination of syncope and epileptic seizure has been called an *anoxic epileptic seizure* (do not confuse with reflex anoxic seizure, which is a syncope). Of anoxic epileptic seizures documented with home video recordings, examples include a neurally mediated syncope inducing a long, clonic epileptic seizure with some features of myoclonic absence, and a compulsive Valsalva manoeuvre in an older autistic child provoking a vibratory tonic epileptic seizure.⁴⁶

Epileptic Seizures Imitating Syncope

Epileptic seizures may manifest with syncopal-like attacks which are common in Panayiotopoulos syndrome.^{46a}



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Imitators of epileptic seizures

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