GENERAL ARTICLES

A CLINICAL APPROACH TO SYNCOPE

***Mani Ram Krishna**
****Mohammed Farooq Kunde**

Abstract: *Syncope is a common clinical complaint that a pediatrician encounters in the outpatient clinic or in the emergency room. The causes of syncope include autonomic disturbances, cardiovascular causes and neurological problems. Autonomic syncope is the commonest and is usually benign. Cardiovascular causes can potentially be life threatening and it is important to recognize them and refer these children to an appropriate specialist in a timely fashion. It is possible to identify the cause of syncope in most patients with a detailed history and physical examination. In this chapter, we list the causes of syncope and attempt to provide a clinical approach that will permit accurate triage of patients with syncope by a pediatrician.*

Keywords: *Syncope, Neurocardiogenic syncope, Cardiac arrhythmia, Fainting, Convulsive syncope.*

What is syncope and how common is it?

The word syncope is derived from the Greek work "synkoptein" meaning to cut short. Syncope is defined as an abrupt and transient loss of consciousness associated with loss of postural tone, typically followed by a rapid recovery.[1] The underlying event in all types of syncope is transient cerebral hypoperfusion. Syncope is a common clinical problem in the pediatric age group with most estimates quoting that 15 % of the population would have experienced at least one episode by the age of 18 years.[2] An analysis of pediatric emergency department (ED) visits from the Unites States identified that 0.9% (627,489 of 72,692,311 visits) of ED visits at pediatric teaching hospitals were due to syncope.[3] It is hence clear that syncope is an important clinical problem in the pediatric age group. The vast majority of syncope in this age group can be attributed to autonomic instability, which is usually benign. However, a small but significant group of children (5-10%) may experience symptoms due to a cardiac cause. The differentiation between the two is usually possible by a detailed history and physical examination. In this article, we will present a clinical approach to recognition and treatment of syncope.

* Clinical Assistant Professor

** Resident in Pediatric Cardiology,
Department of Pediatric Cardiology,
Amrita Institute of Medical Sciences, Kochi.
email: mann_comp@hotmail.com

What are the causes of syncope?

The causes of syncope can be broadly classified into three categories (Fig.1): 1) Autonomic Syncope 2) Cardiac Syncope and 3) Others including neurological causes.

Autonomic syncope

Autonomic Syncope accounts for close to 80% of pediatric cases with syncope. Improvement in our understanding of the pathophysiology has allowed us to further categorize this phenomenon into many types.[4]

The commonest and most well understood of these categories is the 'Neuro-cardiogenic syncope (NCS)' also referred to colloquially as "common faint". The typical NCS episode has three components - a prodrome which almost always precedes the loss of consciousness, which in turn is followed by a prompt and usually complete recovery. The pathophysiology of NCS is best explained by the Bezold-Jarisch reflex (Fig.2) - a paradoxical reflex where pooling of blood in the veins results in both a catecholaminergic surge as well as increased vagal tone.[5]

In addition to the 3 components, a careful history will also reveal the presence of precipitants, which tend to decrease the threshold for the event and triggers that bring about the event. The common precipitants in children include hunger, lack of sleep, dehydration, anemia and viral illnesses while typical triggers include sudden change of posture, prolonged upright posture and emotional stress.

The event itself as described earlier is preceded by a typical prodrome of dizziness or light-headedness and nausea. It almost always occurs when the child is standing upright. Syncope in the supine position is one of the pointers towards a non-autonomic cause of syncope and should be investigated further. Most children will be able to recollect the prodrome on questioning. Onlookers frequently mention noticing that the child looked pale just

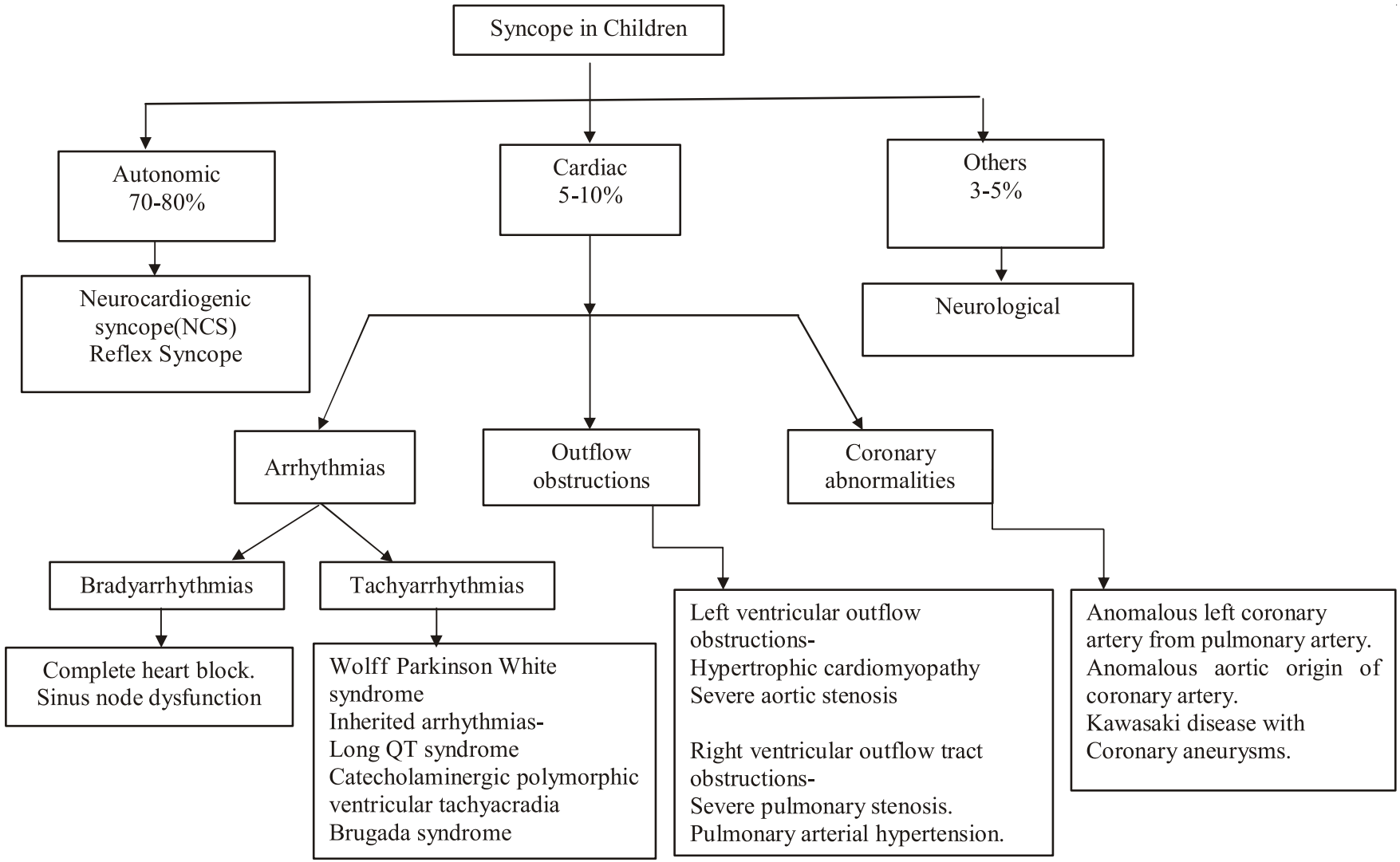

Fig.1. The causes of syncope in children

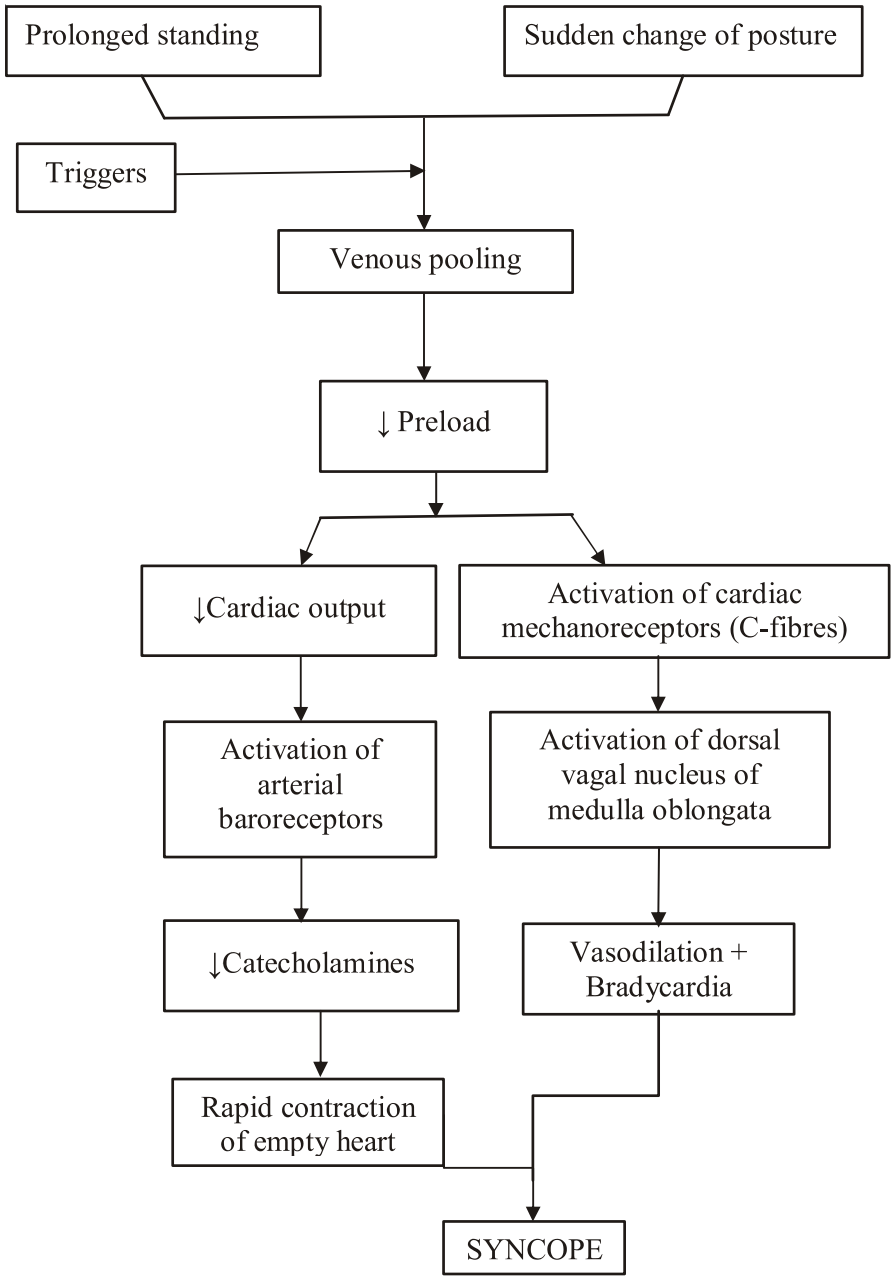

Fig.2. Pathophysiology of Neuro-cardiogenic syncope - The Bezold-Jarish reflex

before the fall. The recovery in NCS is usually instantaneous. There will also be no injury associated with the fall in the majority of cases.

Cardiac syncope

Syncope can be attributed to a cardiovascular cause in approximately 10% of cases.[4] The hallmark of cardiovascular syncope is syncope at the peak of exertion. The cardiovascular causes of syncope can be divided into arrhythmias, outflow tract obstructions and coronary problems.

Outflow tract obstructions (of both the left and right ventricle) are usually fixed mechanical causes. In the resting state, the cardiac output is adequate to match the metabolic demands. However, during exercise, the heart is unable to increase the cardiac output to meet the increased requirements and this results in cerebral hypoperfusion. Frequent co-existing symptoms in this subset include dyspnea on exertion and chest pain.

A peculiar cause of cardiac syncope, which the general pediatrician may not be aware of, is inherited arrhythmias. These are caused by mutations in the genes encoding ion

channels (sodium, potassium and calcium channels). These ion channels control the depolarization and repolarization of cardiac cells and mutations in these channels result in ventricular arrhythmias. The inherited arrhythmias include Long QT syndrome (LQTS), Catecholaminergic polymorphic ventricular tachycardia (CPVT) and Brugada syndrome.[6] The clinical examination after recovery is often normal and the variations in the electrocardiogram (ECG) are often subtle and can be missed by an untrained eye. However, there are very significant clues in the history that raise the suspicion of these disorders. When there is a clinical suspicion, urgent referral to a pediatric cardiologist is indicated to ensure that the diagnosis is not missed.

Syncope due to coronary causes is rare. Kawasaki disease (KD) is fast replacing Rheumatic heart disease (RHD) as the commonest acquired heart disease in children. Children with KD and coronary aneurysm are at a risk for coronary insufficiency during exercise and this can manifest as chest pain or syncope depending on the severity. Anomalous origin of the coronary artery can occur from an unusual location in the aorta (AAOCA) or in rare cases from the pulmonary artery (ALCAPA). ALCAPA more frequently presents in infancy with heart failure. However, in the much rare, adult form of ALCAPA, chest pain and syncope on exertion may be a presenting feature.

Non-cardiac causes

Non-cardiac causes account for less than 10% of syncope. These include neurogenic causes like seizures. It is important to note that cerebral hypoxia may result in convulsive movements even during NCS – a phenomenon referred to loosely as convulsive syncope. A casual bystander may hence confuse this with epilepsy. While clinical differentiation from epilepsy may not always be possible, a few clues can be discerned from a careful history[7]:

1. Prodrome and presyncope – are typical of convulsive syncope and are very unusual in true seizures
2. Tongue biting - is very rare in convulsive syncope and if present is usually at the tip of the tongue unlike the sides during epilepsy
3. Incontinence – typically does not occur in convulsive syncope but is a frequent accompaniment in true seizures
4. Convulsive movement – are usually pleomorphic in convulsive syncope whereas they are rhythmic and uniform in seizures. In recurrent episodes, it may be useful to get one of the bystanders to record a video of the episode
5. Duration - The episode almost always lasts less than a minute in convulsive syncope while true seizures almost always last longer than this. The post-ictal phase is dominated by fatigue in convulsive syncope while confusion is typical of true seizures.

It is important to rule out malingering and other psychogenic causes of seizures especially in adolescents. This is usually difficult to establish clinically. Inconsistencies in the history offer the most important clue towards malingering as the cause of syncope.

Clinical evaluation of syncope

An oft quoted phrase among cardiologists is that "The only difference between syncope and sudden death is that the patient wakes up in one of them". A patient presenting with syncope hence presents a valuable opportunity to ensure that he is not at risk of sudden death. The importance of the clinical assessment in the evaluation of syncope cannot be stressed enough. Indeed, a detailed history is the single most important diagnostic factor in children with syncope.[8]

History

The 5 vital points in history are highlighted in Box 1 and it is imperative that each of these is recorded in detail for each patient. A detailed description of the event may not be available for every episode of syncope. Episodes that occur during school may not always happen under supervision of an adult and peer group children often do not recollect events accurately. In the authors' experience a trigger or precipitant is present in almost every case of NCS and can be obtained by a careful history interspersed with leading questions. Typically, the precipitants include a late-night movie, late night studying before an exam, skipping breakfast and not drinking adequate water in the summer. The triggers include assembly at school and midafternoon sports period.

There are frequently no associated symptoms in children with NCS. However, associated symptoms especially dyspnea on exertion and history of palpitation

Box 1. Key elements in history

- Time of the event
- Activity leading to the event
- Associated symptoms
- Posture at the time of event
- Family history

carry very high specificity for a cardiac cause of syncope. In fact, syncope without prodrome and on exertion along with a history of palpitation has been shown to have a 100% sensitivity and specificity for a cardiac cause of syncope.[9]

The most important finding in history which suggests an inherited arrhythmia is a family history of Sudden Unexpected Death (SUD).[10] The symptoms in inherited arrhythmia disorders include syncope on exertion, syncope triggered by specific stimuli such as shrill sounds or during swimming, syncope while lying supine, family history of sudden unexplained deaths and family history of drowning deaths.[11] Recurrent episodes of syncope during febrile illnesses in a younger child should raise the suspicion of Brugada syndrome. The clinical pointers for a cardiac syncope are summarized in Box 2.

Box 2. The red flag signs which should raise suspicion about a cardiovascular cause of syncope

- Syncope on exertion
- Syncope without prodrome
- Syncope in supine position
- Known cardiac disease
- Known case of Kawasaki disease
- Sudden death in family members (<50years)
- Cardiomyopathy in family members
- Known history suggestive of arrhythmias - Death in other family members due to drowning, syncope after auditory stimulus

Physical Examination

The physical examination in syncope should include the cardiac vital signs i.e heart rate, pulse volume, blood pressure and oxygen saturation (SpO2). When the clinical suspicion is NCS, it is important to look for orthostatic changes in the heart rate and blood pressure. These should be recorded while sitting and after 2-3 minutes of standing. In most cases, these will be normal. However, a tachycardia or hypotension response can prove to be diagnostic. The heart rate is much lower than normal in complete heart block while a low volume, slow rising pulse (pulsus parvus et tardus) suggests significant left ventricular outflow tract obstruction.

The examination of the cardiovascular system should include looking for a parasternal heave (in right ventricular outflow obstruction) as well as cardiomegaly. A harsh ejection murmur at the base of the heart (left or right second inter-costal space) should raise the suspicion of outflow obstruction while a loud pulmonary component of the second heart sound (P2) suggests pulmonary hypertension. The clinical examination is usually normal in children with inherited arrhythmias.

A rapid neurological examination is essential and should focus on possible associations with epilepsy such as neuro-cutaneous markers, signs of raised intra-cranial tension and focal neurological deficits. Finally, it is important to rule out any external injuries that could potentially be related to the fall.

The Electrocardiogram (ECG)

An ECG should be performed in all children in whom a diagnosis of NCS cannot be reliably established at the end of history and physical examination. ECG is a readily available, relatively inexpensive investigation that probably has the highest diagnostic yield in syncope. Typical examples of abnormal ECG have been provided in Fig.3-5. Even if the pediatrician is not comfortable in interpreting the ECG, advancements in technology allows him to obtain the opinion of a pediatric cardiologist from the comfort of his clinic or ED.

In a very small proportion of patients in whom a suspicion of cardiac syncope persists at the end of history, physical examination and ECG, further evaluation by echocardiogram, ambulatory ECG monitoring (Holter) or exercise stress testing will be necessary and this should be done expeditiously by referring them to a pediatric cardiologist. Such tests if performed in those with a negative screen will result in additional economic burden with no diagnostic yield.

Sensitivity of clinical indicators for syncope

The clinical recommendations have been tested in both retrospective and prospective cohorts. Johnson and colleagues analyzed 617 patients who presented with syncope over an18-year period.[11] A screening test of abnormal family history, syncope during exercise, abnormal physical examination and ECG abnormalities had 100% sensitivity for diagnosing cardiac syncope. Tretter and colleagues applied these criteria as an indication for cardiac referral in children presenting with syncope.[12] Approximately 1/3rd of patients with NCS were referred for cardiac evaluation. More importantly, no child with a cardiac syncope was missed.

Practical evaluation of syncope

There is ample scientific evidence that a detailed history, physical examination and an ECG can ensure that

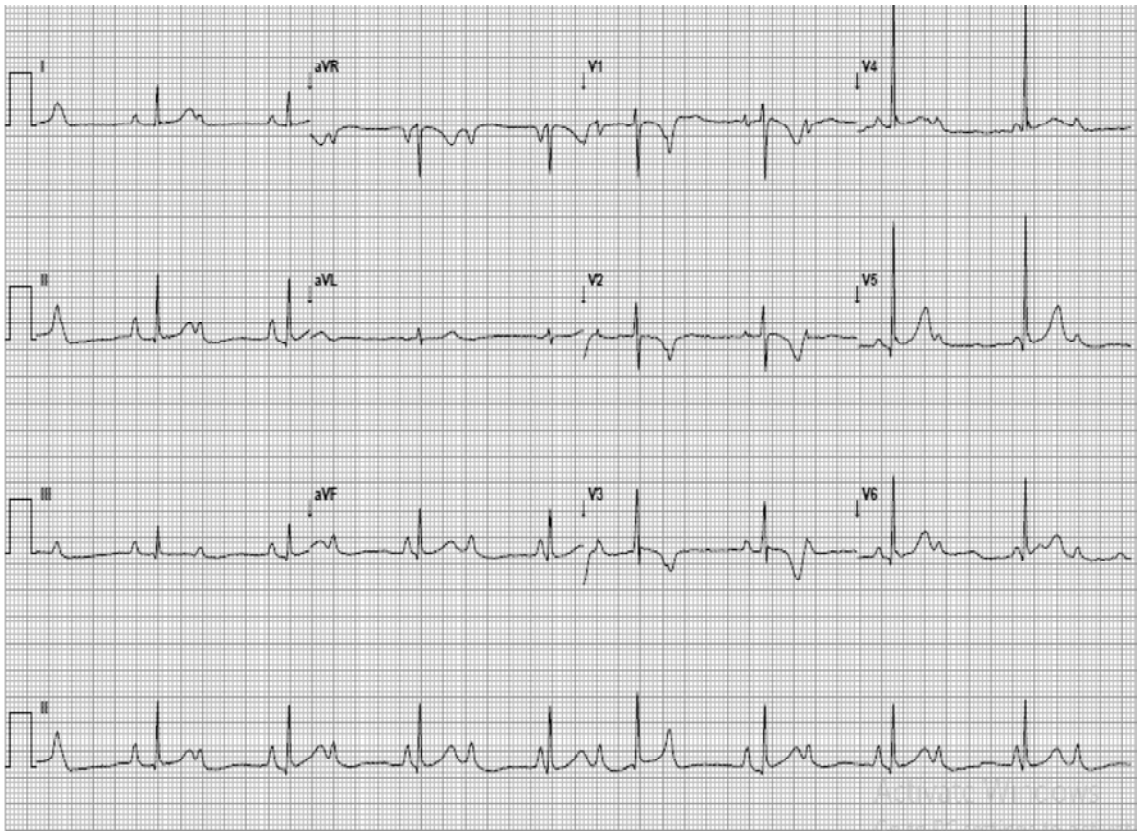

Fig.3. Standardised 12 lead ECG demonstrating bradycardia with no consistent relationship between the p wave and the QRS complex typical of Complete Heart Block (CHB)

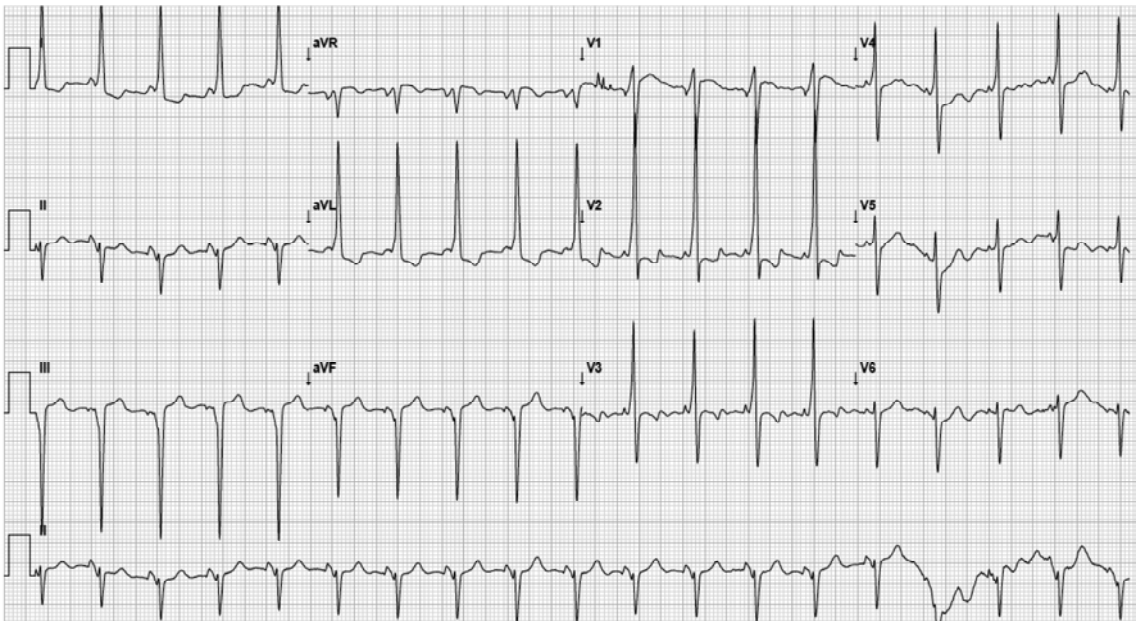

Fig.4. Standardised 12 lead ECG demonstrating sinus rhythm with a short P-R interval, broad QRS complex and a typical slurring of the initial part of the QRS complex (delta wave) in Wolff-Parkinson-White syndrome

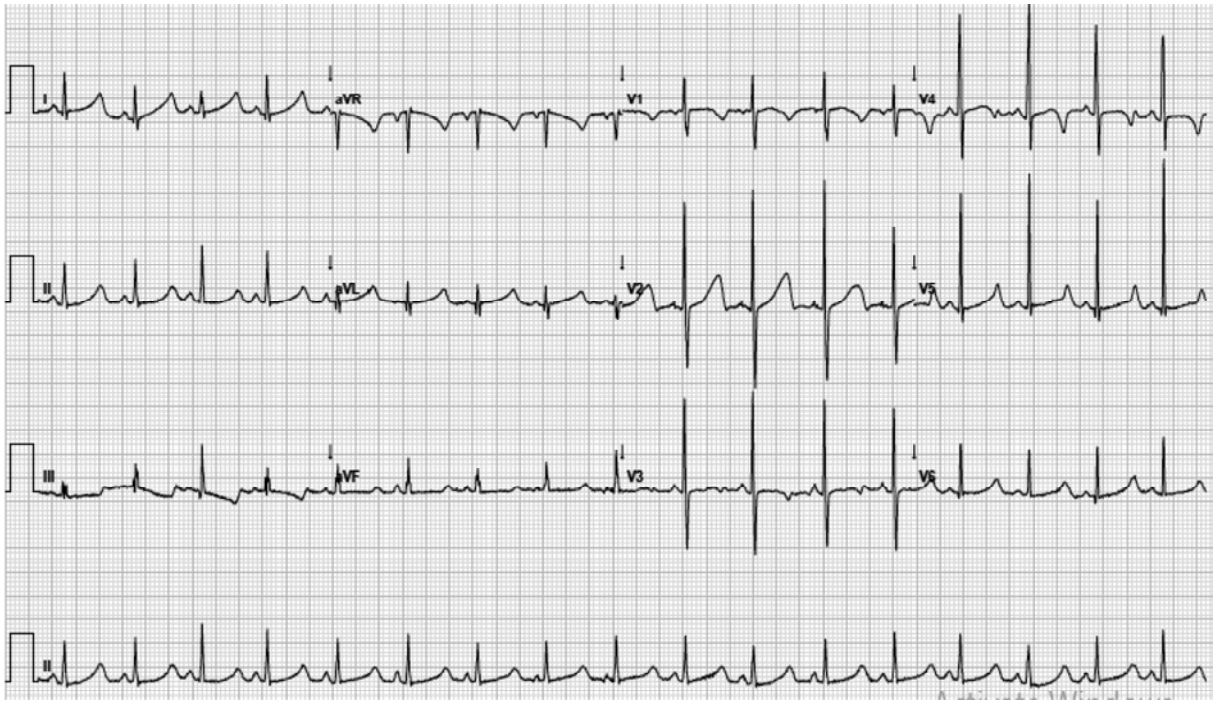

Fig.5. Standardised 12 lead ECG demonstrating sinus rhythm. The T wave is abnormally asymmetrical and the QT interval is prolonged. The QTc in this patient is 540 milliseconds.

no serious cause of syncope will be missed.[8,11,12] This has been made part of recommendations for many years. However, this has not been reflected in clinical practice. Goble and colleagues reviewed workup of syncope in a pediatric ED over a 1-year period.[13] Of the 113 patients who presented with syncope, a minimum of 3 of the 5 key elements of history were recorded in only 60% of patients. Investigations other than ECG were ordered in 89% of patients but a vast majority of tests were non-diagnostic. More than half the patients (58%) underwent a computerized tomogram (CT) of the brain and none of these were diagnostic. Only 10% of patients were admitted but half of these were due to an inappropriate interpretation of the ECG. It is hence clear that the established guidelines are seldom followed in the real world. Unnecessary investigations are frequently ordered. This results in increased anxiety for the patients and also increases the cost of healthcare significantly, an important consideration in resource limited settings in our country. In contrast, the yield of these investigations is virtually nil.

Illustrative case scenarios

Case Scenario 1

A 13-year-old boy was brought to our ED by his physical education teacher on a summer afternoon after he had collapsed at the end of his playtime at school. The boy had played football at school and at the end of the session had gone to drink water with his friends. While awaiting his turn, he had slouched on to the bench behind and fell unconscious. He had recovered within a minute. The boy recollected feeling dizzy but then could only remember waking up surrounded by his teachers. His physical examination was completely normal. His parents arrived half an hour later after being alerted by the school authorities. They confirmed that the boy had no significant past medical history and that there was no family history of SUD. The family had gone to a late-night movie the previous day and the boy had slept for less than 5 hours. A clinical diagnosis of NCS was made. The family was reassured and lifestyle modifications were advised. He was then discharged without further investigations.

Case Scenario 2

A 9-year-old girl was referred to our outpatient clinic from pediatric neurology for evaluation of recurrent syncope. She had been diagnosed as complex partial seizures. However, as the episodes did not improve with treatment, cardiology evaluation was suggested. The parents gave a detailed description of the previous 2 episodes. Once she was carrying a bucket full of clothes up the stairs and collapsed on the top stair. During the other episode, she was dancing along with her sister in front of the television when she collapsed suddenly. Both episodes had clearly occurred at peak exertion. On examination, her vital signs were normal. She however had a grade II para-sternal heave and the pulmonary component of her

second heart sound was very loud, An ECG showed evidence of right ventricular hypertrophy with a strain pattern. An echocardiogram confirmed the diagnosis of severe pulmonary arterial hypertension. There were multiple red flag signs on history and physical examination to merit further evaluation for a cardiac cause of syncope.

Conclusion

Syncope is a common clinical presentation in the pediatric age group. While benign neuro-cardiogenic syncope is the commonest cause, cardiac causes contribute to a small but important subset of these patients. A detailed history and physical examination along with an ECG is adequate to identify all cases of cardiac syncope which require further evaluation.

Points to Remember

- ***Syncope is an important clinical problem in the pediatric age group attending ED.***
- ***There are three groups of causes, autonomic instability which is usually benign and more common, cardiac cause which are serious but less common and others including neurological causes (e.g. convulsive syncope) which are rare.***
- ***The differentiation is usually possible by a detailed history, physical examination and basic investigation like ECG.***
- ***ECG is an inexpensive investigation that probably has the highest diagnostic yield in the evaluation of syncope.***
- ***CT has the lowest diagnostic yield and can be replaced by a good focused neurological examination.***
- ***Unnecessary investigations can be avoided and diagnostic yield can be increased if the pediatrician meticulously takes the history and performs the clinical examination.***

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