SYNCOPE PATHWAY

EXECUTIVE SUMMARY

Physician Owner(s): Dr. Jeffrey Robinson

Primary Objective

The purpose of the Syncope Clinical Pathway intends to standardize the care of otherwise healthy children who experience syncope or near syncope events. Outpatient treatment standardization will address appropriate referrals to Pediatric Cardiology, Pediatric Neurology, and Adolescent Medicine, documentation of history and physicals and ECG's upon encounter, evaluate the need for more advanced follow-up and procedures and reduce healthcare utilization.

Inclusion and Exclusion Criteria

Inclusion Criteria

- Patient presenting with syncope or near syncope event

Exclusion Criteria

- Sepsis
- Shock
- Congenital heart disease
- Central nervous disease
- Previous cardiac surgery
- Epilepsy

Recommendations for Syncope Care

H&P and ECG for Syncope

The comprehensive evaluation of pediatric syncope necessitates a multifaceted approach, integrating both a thorough history and physical examination (H&P) and electrocardiography (ECG) screening. An estimated 2% of pediatric syncope patients presenting to the ED will have an underlying cardiac etiology[17].

Cardiac etiologies of syncope can generally be categorized as either structural or arrhythmogenic. Structural etiologies include cardiomyopathies, coronary artery anomalies, left ventricular outflow obstruction and primary pulmonary arterial hypertension. A variety of electrical disturbances can produce arrhythmogenic syncope. These include congenital long QT syndrome, bradyarrhythmias, high-degree AV block, Brugada syndrome, arrhythmogenic right ventricular dysplasia (ARVD), supraventricular tachycardia (SVT), Wolff-Parkinson-White syndrome (WPW), and catecholaminergic polymorphic ventricular tachycardia (CPVT).

ECG screening is a non-invasive and cost-effective tool and should be included in all initial evaluations of pediatric syncope[3,9]. The likelihood of cardiac syncope decreases when there is no history of heart disease and an ECG is normal[1]; an abnormal ECG is a primary indicator for the potential presence of cardiac syncope[15]. Table 1 outlines the Seattle Criteria and high-risk abnormal ECG findings in athletes[10].

A meticulous H&P provides essential context surrounding the syncopal episode and helps differentiate between benign vasomotor events and potentially more serious cardiac etiologies. The H&P aids in elucidating critical details such as the circumstances surrounding the syncopal episode, family history of cardiac conditions, and associated signs and symptoms. Also, a HEADSSS exam would be recommended. PHQ9A and GAD or similar anxiety screen would be recommended to help obtain further clues towards etiology. Review of growth chart to assess for weight loss. Lab assessment can be tailored based on information from the H&P but could

include CBC, CMP, TSH, ESR, urinalysis, ferritin, & Vit D[12]. Transthoracic echocardiography, exercise tests, and Holter/event monitoring have low diagnostic yield in the setting of pediatric syncope[3,18].

A screening protocol involving a detailed medical history, physical examination, and ECG facilitates the identification of a cardiac origin of syncope in the vast majority of pediatric patients[19].

Table 1 Abnormal ECG findings in athletes

Abnormal ECG finding	Definition
T-wave inversion	>1 mm in depth in two or more leads V2–V6, II and aVF, or I and aVL (excludes III, aVR and V1)
ST segment depression	≥0.5 mm in depth in two or more leads
Pathologic Q waves	>3 mm in depth or >40 ms in duration in two or more leads (except for III and aVR)
Complete left bundle branch block	QRS ≥120 ms, predominantly negative QRS complex in lead V1 (QS or rS), and upright monophasic R wave in leads I and V6
Intraventricular conduction delay	Any QRS duration ≥140 ms
Left axis deviation	−30° to −90°
Left atrial enlargement	Prolonged P wave duration of >120 ms in leads I or II with negative portion of the P wave ≥1 mm in depth and ≥40 ms in duration in lead V1
Right ventricular hypertrophy pattern	R–V1+S–V5>10.5 mm *AND* right axis deviation >120°
Ventricular pre-excitation	PR interval <120 ms with a delta wave (slurred upstroke in the QRS complex) and wide QRS (>120 ms)
Long QT interval*	QTc≥470 ms (male) QTc≥480 ms (female) QTc≥500 ms (marked QT prolongation)
Short QT interval*	QTc≤320 ms
Brugada-like ECG pattern	High take-off and downsloping ST segment elevation followed by a negative T wave in ≥2 leads in V1–V3
Profound sinus bradycardia	<30 BPM or sinus pauses ≥ 3 s
Atrial tachyarrhythmias	Supraventricular tachycardia, atrial-fibrillation, atrial-flutter
Premature ventricular contractions	≥2 PVCs per 10 s tracing
Ventricular arrhythmias	Couplets, triplets and non-sustained ventricular tachycardia

Recommendations for Pediatric Cardiology Referral

While cardiac etiologies of pediatric syncope occur infrequently, their occurrence emphasizes the critical importance of referral to pediatric cardiology for a thorough investigation. A comprehensive history and physical examination, along with an electrocardiogram (ECG), can assist in identifying specific criteria or 'red flags' which may indicate a cardiac etiology and prompt a referral to pediatric cardiology for further evaluation.

Episodes of syncope occurring during or after exertion, as well as syncope accompanied by palpitations or exertional dyspnea, warrant evaluation by pediatric cardiology[1,3, 20, 26].

Additional indicators include instances where syncope is preceded by chest pain, occurs without prodrome, or leads to physical injury from a sudden fall or near drowning[1, 3, 20, 26]. The detection of a pathologic murmur during the physical examination is also an indication for pediatric cardiology referral[1,3, 20].

Special attention should be directed toward patients with syncope who have a known history of congenital/structural heart disease[1,3, 26]. An abnormal ECG during a pediatric syncope work-up also necessitates referral to pediatric cardiology[1, 3, 20]. Syncope involving a patient with a family history of sudden death under 50 years of age, cardiomyopathy, channelopathy, or the presence of a pacemaker/defibrillator should always raise concern and prompt a referral to pediatric cardiology[1, 3, 11, 20, 26].

Ferritin Levels in Syncope

The association between iron deficiency anemia and syncope is logical and well established. However, low total body iron stores can lead to neurological dysfunction and recurrent syncopal events without the presence of anemia in adolescents and children[16]. Low total body iron stores are most accurately detected by ferritin levels and levels should be interpreted as low if they at or below 25 microgram/L.

There is objective evidence in tilt table studies which show that children with low ferritin are more likely to have positive tilt table testing suggesting further that low total body iron stores plays a role in the pathogenesis of neurally mediated syncope[13]. For children with low ferritin levels and recurrent syncope, target treatment levels should be at or above 50 microgram/L[16]. Ferritin levels should be screened every 3 months in children who are symptomatic and/or on iron therapy.

Recommendations for Pediatric Neurology Referral

Neurally mediated syncope is the most common reason for syncope in the pediatric population; however, less common etiologies must be considered. Neurological etiologies of transient loss of consciousness that mimic syncope can grouped broadly into seizure, vascular events, disrupted cerebrospinal fluid (CSF) circulation, and others[3]. Children with concerning histories or risk factors for these symptoms should be referred accordingly.

Seizure is a common presentation to neurology clinics in the pediatric population. Several different types of seizures can be mistaken for syncope. Atonic seizures are seen in generalized epilepsy and characterized by a sudden fall or loss of tone[5]. Typical prodromal symptoms of syncope such as lightheadedness, nausea, and visual obscurations are not present. Myoclonic seizures are also brief seizures seen in generalized epilepsy which are associated with brief jerks of the extremities which may or may not be accompanied by loss of consciousness and collapse[5]. Myoclonic seizures generally do not have an extended post-ictal phase. Focal seizures and focal seizures with secondary generalization may share some semiological features similar to syncope; however, consciousness is typically altered prior to collapse and there is a prolonged post-ictal period. Interestingly, depending on the cause of syncope some patients will have convulsive movements (most typically myoclonic jerks) with the syncopal episode[6]. Careful history and, in some cases, confirmatory testing with electroencephalogram (EEG) are needed to help distinguish convulsive syncope from seizures.

Vascular events in the pediatric population are rare, but an important cause of transient loss of consciousness which has significant treatment and prognostic implications. Cerebral vascular disease such as stroke will typically present with persistent focal neurological deficit in older children, but most commonly present with seizure-like activity especially in younger children which may mimic a syncopal event[4]. There are typically post-event sequelae such as irritability, focal neurological signs, or altered mental status which are harbingers of more serious neurological disease. Emergent evaluation of such cases is imperative. There are several rare entities which may result in decreased cerebral perfusion and vertebrobasilar insufficiency which could present with similar syncopal prodromal symptoms of dizziness preceded by collapse such as subclavian steal syndrome, traumatic vertebral dissection, and Bowhunter syndrome[2]. Non-lethal strangulation by occlusion of the carotid arteries may also result in syncope[8].

Intracranial events leading to acute spike in CSF pressure may lead to episodic loss of consciousness. Most symptomatic hydrocephalus is due to a subacute or chronic process with distinct clinical findings to syncope including nausea, intractable vomiting, lethargy, irritability, and eye movement abnormalities. Certain intracranial pathologies, while rare, can cause hyperacute hydrocephalus that is positional in nature. If there is concern for acute hydrocephalus, emergent imaging is warranted.

Other neurological etiologies which mimic syncope include vertiginous drop attacks in Meniere disease and other vestibular disorders[14]. Basilar and vestibular migraine may also present with symptoms of dizziness, imbalance, and near syncope. Migraine is generally differentiated from syncope based on presence of headache and time course. Cataplexy is sudden loss of tone which may be confused with syncope. Cataplexy is typically triggered by emotional stimuli which can lead to collapse and is a sign of narcolepsy.

Recommendations for Adolescent Medicine

Adolescent aged patients present frequently for chief complaint symptoms such as light headedness, dizziness, or fatigue which can become recurrent and described as pre-syncope to syncope. Up to 15 percent of children experience a syncopal episode prior to the end of adolescence[11]. While these can be presenting signs of life-threatening cardiac conditions or neurological conditions such as seizure, the vast majority of these are common, benign occurrences but can still lead to significant functional impairment through missed school, inability to participate in activities, or fear of unfounded medical conditions resulting in anxiety and frequent medical visits looking for a specific answer[26]. Common adolescent conditions will have pre-syncopal sensations as a common symptom including but not limited to anxiety and depressive disorders, migraine or tension headaches, sleep deprivation, disordered eating, or iron deficiency anemia related to menstrual irregularities. Additionally, these symptoms are also associated with other forms of chronic discomfort[23,25].

Anxiety and depression are often associated with orthostatic intolerance or chronic fatigue which are often involved with recurrent pre-syncopal like events[22,23,25].

A common set of conditions relating to Orthostatic Intolerance are often among the differential diagnosis considerations but can also be misdiagnosed or misunderstood as the terms are often used interchangeably. These would include conditions such as POTS, orthostatic hypotension, postural vasovagal syncope, hypermobility syndromes, and MCAD. A careful approach to evaluating such conditions along with education to the family and patient regarding expectations and treatment for these is important to help with future functionality. An understanding of the impact of psychophysiology involved in these conditions is important to help with therapy and improvement of functionality[23].

Orthostatic assessments or stress tests are standard for assessing orthostatic intolerance conditions however the logistics and reliability of these tests are challenging. Tilt table testing remains the standard for diagnosing POTS in pediatric populations, but this is not feasible in most clinic locations, whereas a standing test is more feasible but more difficult to standardize. Also, a tilt table test is arguably less physiologic than a standing test as there is no muscle venous pumping along with other physiologic changes during a tilt table exam. In adults, there a high false positive rate for POTS has been shown during tilt table testing. Orthostatic hypotension can be reliably diagnosed by tilt or standing for 3 minutes. POTS requires a 10 minute evaluation in the upright position and a standing test could be performed in lieu of lack of tilt table availability[7,23].

Recommendations for Primary Care Providers and Follow-Up

Primary care providers should consider rehydration in this population, aiming for a goal of 80-100 oz of water per day, or in smaller children setting a goal of rehydration with number of ounces of water per day equal to their weight in kilograms. Increased salt intake can help with rehydration, utilizing “healthy, salty snacks” such as nuts, string cheese, veggies and dip, or popcorn. These children should be taught to avoid caffeine, start a daily exercise routine, get consistent sleep, and set up a daily routine. It is very important for these children to eat breakfast, lunch, dinner, and at least 2 snacks. Appropriate stress management techniques should also be provided to these children.

Iron replacement can become difficult in children and adolescents due to the poor palatability and stomach upset associated with oral iron products. Traditionally, this was accomplished through multiple daily doses. However, multiple well-structured studies have demonstrated that daily and multiple times daily dosing decreases cumulative iron absorption due to concurrent upregulation of hepcidin[24]. This committee recommends iron replacement through ferrous sulfate or ferrous gluconate, 3-6 mg/kg/dose, administered daily or every other day.

Iron is best absorbed with increased gastric acidity, so it should ideally be administered apart from antiacids, meals and separate from dairy products which inhibit absorption. Administration with Vitamin C containing foods (orange juice, citrus) can improve absorption[21].

For children requiring iron replacement, follow-up with a repeat Hemoglobin lab draw should performed in 2 weeks. Ferritin levels should be rechecked in 3 months. Continued rehydration should be reinforced in this population.

For children that do not require iron replacement, they should perform blood pressure checks at home 1-2 times per week for 2 weeks and report back readings to the provider. If blood pressures remain appropriate, they should be seen for a follow-up in 2 months. For blood pressures that are not appropriate, follow-up should be provided at the provider's discretion.

Rationale

Safety: will be improved by utilizing consistent practices when referring to specialties, providing guidance on how to treat syncope in the outpatient setting, and consistent follow-up recommendations.
Quality: will be improved by providing consistent care and follow-up recommendations across outpatient providers.
Cost: will be reduced by improving the identification of children who necessitate an ECG and those children who need additional specialty referrals.
Delivery: will be improved by standardizing the way providers engage with children with syncope and pre-syncope.
Engagement: is created by the engagement of a multidisciplinary team in the development and maintenance of the pathway.
Patient/Family Satisfaction: will be improved by providing the highest quality of care based on the latest evidence in the literature.

Metrics

Outcome

- Proportion of patients with an H&P or Office Visit Progress Note documented within 30 days before or after the syncope visit
- Proportion of patients with a referral to Pediatric Cardiology, Pediatric Neurology, or Adolescent Medicine and an ECG documented within 30 days before or after the referral date

Process

- Proportion of referrals related to ICD 10 codes associated with Pediatric Cardiology from the total number of referrals for Cardiology
- Proportion of referrals related to ICD 10 codes associated with Pediatric Neurology from the total number of referrals for Neurology
- Proportion of referrals related to ICD 10 codes associated with Adolescent Medicine from the total number of referrals for Adolescent Medicine
- Proportion of patient visits for syncope that utilize the SmartSet "Syncope/dizziness"

Balancing

- Proportion of patients who have 3 or more syncopal encounters without a referral to Pediatric Cardiology, Pediatric Neurology, or Adolescent Medicine
- Proportion of patients who are seen at Children's Physicians or Urgent Care for syncope and are then seen in the Emergency Department for syncope within 3 months

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Physician Owner(s): Dr. Jeffrey Robinson

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