Pediatric Education and Active Resident Learning (PEARL) Conferences

Approach to Syncope

December 3, 2010

When working in the Emergency Department, you are called to see a 7-year-old girl who has “passed out” at school today. She has previously been healthy and has not had any recent illnesses. As you walk to her room, you think about what could have caused this and how you will go about evaluating her…

Core References

- Ng B, Maginot KR. Sudden cardiac death in young athletes. WMJ. 2007;106:335-42.

Review ECG interpretation, calculation of rate, rhythm, PR, QRS, QTc intervals. You do not have to go through the “cases” on this website.

What to bring to the conference:

- Calculator
- Calipers, if you have them
- Harriet Lane (for table on Normal Pediatric ECG Parameters)
CASE A.

10 year old previously healthy girl comes in for a pre-participation athletic screening.

Review of systems reveals 3 episodes of loss of consciousness in the past: the first occurred two years ago while standing in church. She reported feeling nauseous, appeared pale and then had a brief syncopal episode. The episode lasted about 10 seconds. When she awoke, she was diaphoretic and pale, but otherwise appeared normal. The second episode occurred 1 year ago: one morning, after a warm shower, she reported feeling nauseous and dizzy and appeared pale, she passed out for 20-30 seconds. She had extremity stiffening and “her eyes rolled back in her head.” The most recent episode was 6 months ago while she was attending a dinner theater and reported having mild abdominal discomfort. While walking into the theater she said she felt funny. She appeared pale and sat down. She was diaphoretic and had a syncopal episode that lasted about 10-20 seconds. She takes no medications. She has NKDA. PMH: negative. FH: negative except for her father who has HTN controlled with medications. Her examination is normal.

You should order which of the following tests initially:

1. ECG
2. Echocardiogram
3. EEG
4. 30 day cardiac event monitor

Her ECG is shown below.

Calculate the heart rate, PR interval, QRS duration, corrected QT interval, and final interpretation (including comments of abnormal rhythm, axis, and repolarization, if any).
Following the evaluation of her ECG, you recommend which of the following in terms of testing:

1. Further testing with echocardiogram.
2. Further testing with 30 day event monitor.
3. Further testing with EEG.
4. No further testing is necessary at this time.

True or False: Neurally-mediated syncope is also called neurocardiogenic syncope or vasovagal syncope.

True or False: Neurally-mediated syncope is the most common etiology for syncope and young healthy patients.

Neurally-mediated syncope may be initiated by multiple different stimuli. List several, from the most common to least common.

1. 
2. 
3. 
4. 
5. 
6.

Appropriate initial recommendations for this patient include which of the following:

1. Exercise restrictions.
2. Increasing fluid intake and avoidance of stimuli.
4. Initiation of beta-blockers or fludrocortisone.

True or False: Tilt table testing is highly specific and sensitive for diagnosis neutrally-mediated syncope.
CASE B.

A 16 year old previously healthy boy comes for an evaluation of a sore throat.

His mother mentions that last week the family was in Chicago and her son had brief syncopal episode after dinner when he was in the bathroom. He has had no fevers, rashes, vomiting, or diarrhea. He takes no medications and has NKDA. PMH: negative. FH: negative. Exam: Mild erythema of the posterior pharynx, no tonsillar hypertrophy.

You should order which of the following:

1. ECG
2. Echocardiogram
3. EEG
4. 30 day cardiac event monitor

His ECG is shown below.

Calculate the heart rate, PR interval, QRS duration, corrected QT interval, and final interpretation (including comments of abnormal rhythm, axis, and repolarization, if any).
Your recommendations include which of the following:

1. Further testing with echocardiogram.
2. Further testing with cardiac catheterization.
3. Further testing with electrolytes, Ca, Mg, TFTs
4. No further testing is necessary at this time.

The patient’s has obvious hypertrophic cardiomyopathy on echocardiogram.
Your recommendations are which of the following (may choose more than one):

1. Exercise restrictions.
2. Avoid medications that prolong repolarization.
3. Initiation of beta-blocker therapy.
4. Evaluation of family members with ECG and echocardiogram.

True or False: Patients with hypertrophic cardiomyopathy almost always have a heart murmur.

True or False: Patients with HCM have syncope only with exertional activities.

True or False: The ECG is usually normal in patients with HCM.

True or False: HCM is the most common cause of cardiac sudden death in young athletes in the US.

True or False: HCM is rarely inheritable and thus screening family members is reasonable.

Which of the following are high risk factors for patients with hypertrophic cardiomyopathy:

1. Drop in systolic blood pressure with exercise.
2. Left ventricular septal wall thickness > 3 cm.
3. Non-sustained ventricular tachycardia.
4. All of the above.
CASE C1.

An 18-month-old girl is sitting on the floor, playing with her stuffed cat. Her 3-year-old brother saunters over, takes the cat away, and runs out the room. The girl then cries, so hard that it appears that the girl stops breathing. She rapidly develops facial cyanosis then loses consciousness and falls over onto the floor, whereupon all limbs stiffen and she “trembles” for about 20 seconds. She then awakens and continues to cry. This scenario repeated itself two more times over the ensuing month, and the parents bring the girl into your office for evaluation.

What are the key history points? Is there any further history that you would like?

How would you work up this child?

What treatment is indicated?

How would you counsel the family?
CASE C2.

Now let’s change the scenario slightly:

An 18-month-old girl is sitting on the floor, playing with her stuffed cat. Her 3-year-old brother comes up behind her, and frightens her by whacking her on the head with his lightsaber. He then runs out the room. The girl suddenly becomes “as white as a ghost”, loses body tone and slumps onto the floor. Her mother, who witnessed the event, thinks the girl has “died” and begins mouth-to-mouth resuscitation. While performing mouth-to-mouth, the girl’s limbs stiffen followed by clonic jerks for about 20 seconds. She falls asleep for a few minutes and awakens back at her normal baseline. This scenario repeated itself two more times over the ensuing month, and the parents bring the girl into your office for evaluation.

What are the key history points? Is there any further history that you would like?

How would you work up this child?

What treatment is indicated?

How would you counsel the family?
CASE D.

A 10 year old boy presented with an episode of sudden loss of consciousness while riding his bike. After he lost consciousness, his jaw clenched, all limbs stiffened and jerked for at least 30 seconds. There was no loss of bowel or bladder control. Bystander CPR was initiated and he regained consciousness. PMH: one year ago, he lost consciousness while running the 50 yard dash and reportedly had bystander CPR. He is an only child, and there is no family history of early sudden death. His physical examination is normal. A sleep-deprived EEG showed frequent left frontotemporal spikes. Brain MRI scan showed thinning of the corpus callosum and cingulate gyrus, said to reflect cortical dysplasia.

His ECG is shown below.

Calculate the heart rate, PR interval, QRS duration, corrected QT interval, and final interpretation (including comments of abnormal rhythm, axis, and repolarization, if any).
You recommend which of the following:

1. Echocardiogram because his ECG is abnormal.
2. Echocardiogram, although his ECG is normal.
3. Exercise test because the ECG is normal.
4. Cardiac catheterization to check coronary artery anatomy due to findings of ECG.

In light of the abnormal EEG and seizure activity following the loss of consciousness, you recommend:

1. Starting ethosuximide.
2. Starting oxcarbazepine.
3. Starting valproic acid.
4. Starting levetiracetam.
5. Not starting any anticonvulsant.

The boy’s echocardiogram was normal, so your recommendations are:

1. Increase fluids and liberalize salt in the diet.
2. Exercise treadmill test.
3. Exercise restrictions and beta-blocker therapy.
4. Genetics testing to rule out an inherited arrhythmia.

True or false: Catecholaminergic polymorphic ventricular tachycardia (CPVT) is typically inherited in an autosomal recessive fashion.

True or false: Catecholaminergic polymorphic ventricular tachycardia (CPVT) is due to abnormal calcium regulation from a ryanodine receptor mutation.

True or false: Patients with CPVT typically present in teenager years.

True or false: Patients with CPVT should be treated with beta-blockers.

True or false: The patient will require lifelong anticonvulsant therapy in addition to treatment for his CPVT.
CASE E.

A term newborn has a brief syncopal episode in his mother’s arms while feeding. He was born via NSVD to a G4, P1 to 2, SAb 2 28 yo female with good prenatal care and no problems during this pregnancy. Apgars were 8 at 1 min and 9 at 5 mins. The family history is notable for a maternal uncle who drowned at 16 years of age and a paternal cousin who is deaf but otherwise normal. The infant’s examination is normal except for bradycardia (heart rate is regular and 80 BPM).

An ECG was done.

Calculate the heart rate, PR interval, QRS duration, corrected QT interval, and final interpretation (including comments of abnormal rhythm, axis, and repolarization, if any).

![ECG Image]

What is the most common identifiable cause for neonatal complete AV block in patients with a structurally normal heart:

1. Long QT syndrome.
2. Viral myocarditis.
4. Lyme carditis.
List 3 symptoms that patients presenting with LQTS have:

1.
2.
3.

The life threatening arrhythmia associated with long QT syndrome is:
1. Rapidly conducting atrial fib-flutter down the accessory pathway.
2. Asystolic pauses due to a sodium ion channel defect.
3. Torsade de pointes.
4. Bradycardia due to complete AV block

True or false: acquired LQTS may occur due to electrolyte abnormalities or exposure to certain drugs that prolong repolarization.

Currently there are over 10 genes (and hundreds of mutations) that can cause LQTS, the known ion channel abnormalities include: Na, K, Ca, Mg, Phos (circle the ions channels that are correct).

True or false: patients with LQTS always have their events with exertional activities.

True or false: patients with the most typical form of LQTS respond well to beta-blockers.

True or false: Certain medications, such as erythromycin and azithromycin should be avoided in patients with the diagnosis of LQTS.

True or false: LQTS can be ruled out if a patient’s ECG shows a normal QTc.

What family history is important in the evaluation of inherited arrhythmia patients? List at least 5 questions:
1.
2.
3.
4.
5.

The patient’s mother asks if her other child should be evaluated. You should answer:
1. Biological family members should be screened.
2. Only symptomatic biological family members should be screened.
3. Genetic testing will be ordered first on the baby to determine the specific genetic mutation. If the testing is negative, other family members do not require any cardiac screening.
4. It is unlikely that the mother is affected since she has been completely asymptomatic; therefore, she does not require testing.
Learning Objectives

By the end of this session, each resident will be able to:

- Differentiate between benign and pathologic forms of syncope.
- Choose appropriate testing to make a diagnosis for a patient with syncope.
- Recognize the common presentations for breath holding spells and determine the appropriate therapy.
- Recognize the common presentations for neurally-mediated syncope (vasovagal syncope) and determine the appropriate therapy.
- Recognize typical ECG findings that suggest: Wolff-Parkinson-White (WPW) syndrome, long QT syndrome (LQTS), Brugada syndrome, AV block, and catecholacholaminergic polymorphic ventricular tachycardia (CPVT), and hypertrophic cardiomyopathy.
- Recognize the cardiac disorders that are inheritable and the importance of family screening.
- Determine the appropriate therapy for cardiac causes of syncope.

Top Pearls

1. Neurally mediated syncope (also known as vasovagal syncope or neurocardiogenic syncope) is the most common cause of syncope.
2. Neurally mediated syncope is diagnosed by history: it is typically non-exertional, brief, associated with a prodrome (lightheadness, nausea, tunnel vision). This form of syncope is considered benign.
3. Fainting due to breath holding spells and neurally mediated syncope may be associated with jerking of the extremities. These brief “seizures” are not considered epileptic.
4. Breath holding spells (BHS) come in 2 flavors: white (pallid) and blue (cyanotic). Pallid BHS are triggered by a bump or fright and result in vagally mediated decreased heart rate and syncope. Cyanotic BHS are triggered by crying that leads to expiratory apnea and involve syncope unrelated to in heart rate.
5. An electrocardiogram (ECG) should be performed on all patients with unexplained syncope (including all patients with first time seizures and syncope).
6. Patients with long QT syndrome, WPW, Brugada syndrome, AV block, and hypertrophic cardiomyopathy typically have abnormal ECGs.
7. The most common cause of sudden death in young athletes in the US is hypertrophic cardiomyopathy.
8. Exercise-induced seizures and exercise-induced syncope require a complete cardiac evaluation. Patients with catecholaminergic polymorphic ventricular tachycardia typically have a normal ECG and echocardiogram.
9. Long QT syndrome can be acquired or congenital and is due to abnormal repolarization that causes episodes of syncope, seizures, and sudden death.

Additional References


Ng B, Maginot KR. Sudden cardiac death in young athletes. WMJ. 2007;106:335-42.

Question 1
The most common cause of syncope in young children is:
1. Autonomic dysfunction associated with prolonged standing.
2. Breath-holding spells.
3. Cataplexy.
4. Panic attack associated with hyperventilation.
5. Prolonged QT syndrome.

Question 2
A 9 year old girl had a history of syncope at recess. It was unwitnessed by an adult, but she reportedly was fine by the time the paramedics arrived. She had one previous episode in kindergarten while running at recess. Her PMH is otherwise negative. She has mild ADHD and takes guanfacine on a regular basis (this medication is not expected to have cardiac side effects). The family history is notable for her father who had arrhythmias as a child and had a pacemaker implanted at 20 years of age. He takes metoprolol and mexiletine. The girl’s examination is normal. If her echocardiogram and ECG are normal, your recommendations are:
1. Parental reassurance and have the patient increase her fluid intake.
2. Arrange a cardiac catheterization to rule out coronary abnormality.
3. Order an exercise test.
4. No further testing is necessary.

Question 3
Of the following disorders that may result in syncope, which is correctly paired with its etiologic mechanism?
1. Breath-holding spells and prolongation of the QT interval.
2. Common fainting and autonomic dysfunction.
3. Cough syncope and increased venous return to the right heart.
4. Exercise-induced syncope and an abnormal EEG reading.
5. Panic attacks and a metabolic alkalosis.

Question 4
One of the most important aspects in the management of a child who has cyanotic breath-holding spells is:
1. Iron therapy.
2. Oral anticholinergic therapy.
3. Parental reassurance.
4. Parental training in cardiopulmonary resuscitation.
5. Strategies to prevent crying.
Question 5
Which of the following statements about syncope is true?
1. Cardiac disease is the most common cause.
2. Electrocardiography is an essential part of the evaluation for unexplained syncope.
3. Fainting usually results in injury.
4. It is more common in children younger than 10 years of age.
5. Most children who have syncope have an underlying pathologic cause.

Question 6
You are evaluating a 13-year-old girl who fainted at school. Which of the following histories would raise your suspicion most for a vasovagal cause for the syncope?
1. She fainted while playing soccer.
2. She felt a bitemporal headache shortly before the episode.
3. She was unconscious for approximately 5 minutes.
4. The episode was followed by a prolonged period of disorientation and sleepiness.
5. The episode was precipitated by a frightening event.

Question 7
A 14 yo previously healthy girl has had 5 episodes of syncope. The first occurred in science class at school during a movie on dissection of a mouse. The second occurred while her mother was combing her hair in the morning. One episode occurred when she cut her finger while carving a pumpkin. Two recent episodes occurred with a flu shot and a blood draw. She was noted to have pallor for all the episodes and reported tunnel vision just prior to the episodes. The episodes lasted 10-30 seconds. She had jerking of her extremities noted with the syncope associated with her most recent episodes. Her PMH and FH is negative except for coronary artery disease and CABG in the maternal grandparents. Her examination and ECG are normal. Your recommendations are:
1. EEG to rule out seizures.
2. Echocardiogram, consider exercise test.
3. Event monitor.
4. No further testing is necessary.

Question 8
About 9% of patient with congenital Long QT syndrome present with seizures.
1. True
2. False

Question 9
You have a patient with neurally-mediated syncope. Which of the following statements is INCORRECT?
1. This is the most common form of syncope in young patients.
2. Patients typically have a prodrome associated with this form of syncope.
3. Seizure-like activity (jerking of the extremities) is uncommon with this type of syncope.
4. First line therapy is fluid therapy and avoidance of precipitating stimuli.
Question 10
The rhythm strip below was noted on an exercise test of a patient with exertional seizures.

Which of the following disorders would you be concerned about?
1. Wolff-Parkinson-White syndrome with rapidly conducting atrial fib-flutter.
2. Complete AV block with a rapid ventricular escape rhythm
3. Catecholaminergic polymorphic ventricular tachycardia
4. All of the above.