SPA/AAP Pediatric Anesthesiology 2012 Winter Meeting

PBLD: Using the 2010 PALS guidelines to manage a pediatric patient with Long QT Syndrome

Learning Objectives:

1. Upon completion of this learning activity, participants should be able to describe the major changes to the new 2010 AHA PALS guidelines.

2. Upon completion of this learning activity, participants should be able to characterize the anesthetic implications for a pediatric patient with a prolonged QT interval.

3. Upon completion of this learning activity, participants should be able to create an effective anesthetic management strategy for the pediatric patient with Long QT Syndrome.

4. Upon completion of this learning activity, participants should be able to recognize the indications for infective endocarditis prophylaxis in the pediatric patient.
Case and Questions:

Your case is an 8 year old male scheduled for washout and debridement of suspected osteomyelitis in the right lower extremity.

The patient was seen in the Emergency Department (ED) 2 days ago with fever, right lower extremity pain, swelling, and inability to walk. Plain x-rays were unremarkable. The patient was started on cephalexin and acetaminophen for suspected cellulitis. The patient was subsequently discharged home.

Upon return to the ED tonight, the patient appears overall much worse than two days ago, according to the mother.

The patient had the following vital signs upon arrival to the ED:

Temperature (oral) 102.6 C, Respirations 42, Heart rate 164, Blood pressure 88/64, Room Air Oxygen saturation 95%. Patient weight: 30 kg.

The patient had a Magnetic Resonance Imaging (MRI) study tonight which was reported by the radiologist as a large abscess located in the right lower extremity.

In the ED over the last two hours, the patient has received acetaminophen and two fluid boluses of lactated ringers totaling 500 milliliters (ml). The patient is currently receiving intravenous fluids at a rate of 70 ml/hour. Clindamycin has already been administered. The patient has one 22 gauge peripheral intravenous line.

The surgeon has agreed, per the admitting physician’s request, to place a PICC (Peripherally Inserted Central Catheter) line during the surgical procedure due to suspected long-term intravenous antibiotic administration.

Additional information from the parent:

Drug allergies: Penicillin (rash)

Home medications: atenolol

Past Medical History: History of an abnormal electrocardiogram (EKG) due to Long QT Syndrome

Past Surgical History: tonsillectomy and adenoidectomy; Ventricular Septal Defect (VSD) repair several years ago

Last oral intake: 4 hours ago for solid food
Diagnostic tests:

Hematocrit 37%

The following EKG was obtained:

![EKG Image]

1. The QT interval is reported as 536; the QTc is reported as 569. What is a normal QT interval? Why is the QT interval corrected (QTc)?

2. What are some congenital and acquired causes for producing a prolonged QT interval.

3. What is Long QT Syndrome?

4. Does this patient require infective endocarditis prophylaxis? What additional information would you request in order to make that decision?

5. If the patient requires infective endocarditis prophylaxis, which medication(s) would you administer?

After consultation with a cardiologist and review of the electronic medical record, you have determined that the patient has Long QT Syndrome and is medically optimized to proceed with this surgical procedure.
6. What are the anesthetic implications for patients with prolonged QT intervals such as Long QT Syndrome?

7. Are there any medications that you would avoid in a patient with a prolonged QT interval?

Induction was uneventful. Vital signs are stable; the airway was secured with a tracheal tube. The surgical procedure has been underway for 20 minutes.

During PICC line placement, the patient suddenly develops a blood pressure of 58/36 with the following electrocardiogram (EKG) tracing:

![EKG Image]

The PICC line guidewire is promptly removed by the surgeon. The EKG rhythm and vital signs remain unchanged.

8. What is your interpretation of this EKG? What is your management plan? What is your plan if this therapy is ineffective?

The patient is stabilized using the appropriate Pediatric Advanced Life Support (PALS) guidelines. Surgery concludes after completing a large debridement and washout to the right lower extremity. The patient received 700 ml of lactated ringers. The estimated blood loss was 150 ml. The intraoperative hematocrit was measured at the end of the procedure and was reported to be 29%.

The surgical procedure has been completed. You planned to perform an awake extubation. During emergence, an alarm on the EKG monitor has been activated.

The following EKG is present:
9. What is your interpretation of this EKG?

10. Suppose the code cart was not present within the operating room. What are your initial steps as the code cart is being obtained?

11. What is your management plan after the code cart has arrived? If these therapies are ineffective, what are your next steps in management of this rhythm? Would you administer vasopressin?

12. If you thought the rhythm is Torsade de pointes, are there any specific therapies in addition to the standard PALS guidelines?

The patient has been effectively resuscitated; you transferred the patient with a tracheal tube in place to the Pediatric Intensive Care Unit (PICU).

Vital signs: Temperature 101.2 C; Heart rate 124; Blood pressure 90/54; Oxygen saturation is 100% on FiO₂ of 1.0.

13. What is your recommendation to the PICU team regarding the amount of postoperative FiO₂ for this patient?

You have given report to the PICU physician and conclude transferring care to the PICU team.
Discussion:

**Prolonged QT interval/Long QT Syndrome (Questions 1-3):**

The QT interval represents the electrical depolarization and repolarization of the ventricles. The QT interval is a measure of the time interval between the start of the Q wave and the end of the T wave. The QT interval decreases with tachycardia and conversely increases with bradycardia. Therefore, the corrected QT interval (QTc) is used to compensate for the heart rate variation. The QT interval can be corrected by several formulas; the most popular method is by dividing the QT interval by the square root of the RR interval. A normal QTc interval is less than approximately 440 milliseconds (msec); a QTc interval of 450-470 msec is considered borderline prolonged. Patients with conditions such as Long QT Syndrome (LQTS) typically have a QTc interval greater than 480 msec.

Causes for producing a prolonged QT interval include congenital and acquired etiologies. Congenital forms of LQTS occur due to genetic mutations within the ion channels of cardiac muscle. These genetic mutations occur within the transmembrane protein mostly to the potassium channel; however, defects also occur in the sodium and calcium ion channel proteins. A prevalence as large as 1:5000 is estimated for all congenital forms of LQTS. It is proposed that there are many undiagnosed cases such as sudden death due to LQTS.

Acquired forms of LQTS commonly result from electrolyte disorders, predisposing medical conditions, and from the administration of drugs that increase the QT interval. Electrolyte disorders that increase the QT interval include hypomagnesemia, hypokalemia, and hypocalcemia. Numerous conditions predispose patients to these electrolyte disorders including diuretic administration, starvation, total parenteral nutrition, gastrointestinal losses, renal losses, transcellular shift, and endocrine conditions. Medical conditions that are associated with increasing the QT interval include hypothyroidism, hypothermia, subarachnoid hemorrhage, cocaine toxicity, organophosphate poisoning, autonomic neuropathy, and myocardial ischemia.

Long QT Syndrome (LQTS) is a disorder characterized by a prolonged QT interval on the electrocardiogram (EKG). LQTS is a condition becoming recognized more frequently with important implications to the anesthesiologist. The typical age for onset of symptoms due to LQTS places pediatric patients at significant risk. Patients with LQTS are at increased risk for cardiac events including Torsade de pointes and sudden death. Currently, 8 genes have been identified and implicated in approximately 70% of the congenital cases of LQTS.
Multiple congenital syndromes have been identified and associated with LQTS; a large amount of these cases have presented in the pediatric patient. Jervell Lange-Nielsen, was the first to report a case in 1957 of congenital deafness with an autosomal recessive pattern of inheritance. Romano and Ward have described a similar syndrome without deafness but inherited in an autosomal dominant pattern. Andersen syndrome and Timothy Syndrome are two newly recognized syndromes also thought to be associated with mutations to the cardiac muscle ion channels.

Patients with LQTS may report a history of ventricular arrhythmias, syncope, sudden death, and Torsade de pointes (TDP). However, many patients will be completely asymptomatic; some patients will report onset of symptoms i.e. syncope after physical exertion or sympathetic stimulation. Since documenting the occurrence of ventricular arrhythmias may prove to be difficult, patients with LQTS have frequently been misdiagnosed with epilepsy and vasovagal syndrome.

Current therapeutic options for patients with LQTS include beta blockade, pacemaker, and/or ICD placement. Left cardiac sympathetic denervation was previously utilized for patients who were refractory to medical management. Oral potassium supplementation and potassium sparing diuretics are currently being investigated for specific subtypes of LQTS.

Beta-adrenergic blockers are the mainstay of medical therapy for patients with LQTS. Beta-blockers have been shown to reduce the number of cardiac events, including TDP, in patients with LQTS. Therapy should be continued for life; this includes the perioperative period. In addition, many experts recommend beta-blocker therapy for asymptomatic patients with congenital forms of LQTS. Pacemakers and/or ICD’s should be considered for patients with LQTS who are symptomatic despite maximal medical therapy.

**Infective Endocarditis Prophylaxis (Questions 4-5):**

The current Infective Endocarditis (IE) prophylaxis guidelines, developed by the American Heart Association, were revised in 2007. These updated guidelines have advocated for a significantly lower amount of cases requiring prophylactic antibiotic administration. These new guidelines are based on the position that bacteremia is common; however, the development of Subacute Bacterial Endocarditis (SBE) is rare and requires abnormal cardiac endothelium for bacterial adherence. Abnormal cardiac endothelium is present in association with high risk cardiac conditions.

The experts that developed the current American Heart Association IE prophylaxis guidelines have concluded that SBE is rarely prevented with antibiotic prophylaxis. Furthermore, recent literature suggests that acquiring SBE
is much more likely from daily activities (e.g. oral hygiene) than in the perioperative period. The experts conclude that the risk from antibiotic administration may actually be higher than the potential benefit received from a risk reduction in IE.

In order for IE prophylaxis to be indicated, a patient must have the combination of an identified surgical procedure and a high risk cardiac condition. The current IE guidelines have eliminated antibiotic prophylaxis for gastrointestinal and genitourinary procedures.

Surgical procedures that may require IE prophylaxis include:

- Dental (such as gingival, periapical, and oral mucosa)
- Skin (such as infected skin and musculoskeletal tissues)
- Respiratory tract (such as incision or biopsy, e.g. tonsillectomy)

High risk cardiac conditions include:

- Use of prosthetic valve or materials
- Previous infective endocarditis
- Unrepaired cyanotic Congenital Heart Disease (CHD)
- Repaired CHD with prosthetic materials < 6 months
- Repaired CHD with residual defects
- Heart transplant with valvulopathy

Patients that require IE prophylaxis should receive antibiotic administration before surgical incision. The antibiotic should be administered as a single dose. The following are the suggested regimens and doses for patients not penicillin allergic:

- Amoxicillin (oral): 50 mg/kg; maximum 2g
- Ampicillin (Intravenous; IV): 50 mg/kg; maximum 2g
- Cefazolin (IV): 50 mg/kg; maximum 1g
- Ceftriaxone (IV): 50 mg/kg; maximum 1g

The following are the suggested regimens and doses for patients that are penicillin allergic:

- Clindamycin (IV): 20 mg/kg; maximum 600 mg
- Cefazolin* (IV): 50 mg/kg; maximum 1g
- Ceftriaxone* (IV): 50 mg/kg; maximum 1g
- Oral: cephalexin*, clindamycin, azithromycin, clarithromycin

* = contraindicated if anaphylaxis, angioedema, urticaria
**Intraoperative Management (Questions 6-7):**

Anesthetic implications for the patient with LQTS include an increased risk of ventricular arrhythmias, Torsade de pointes (TDP), and sudden death. The main goal is to avoid conditions that increase sympathetic stimulation, prevent the development of hypokalemia, hypocalcemia, hypomagnesemia, and substitute medications not associated with increasing the QT interval. However, TDP can also occur spontaneously. Conditions that increase sympathetic stimulation should be prevented by the use of sufficient preoperative anxiolysis, insuring adequate levels of anesthesia, and appropriate administration of postoperative analgesia. Conditions that increase sympathetic stimulation include hypothermia, hypertension, tachycardia, hypoxemia, and hypercapnia. In addition, conditions that cause bradycardia can also increase the QT interval and possibly increase the risk of TDP. Anesthetic techniques that should be considered may include deep extubation, parental presence during induction of anesthesia, and the use of regional anesthesia.

Anesthetic management for patients with LQTS requires appropriate preparation. A code cart with a defibrillator should be immediately available. Resuscitation medications including beta-blockers, calcium, and magnesium should also be readily accessible. Continuous EKG monitoring should occur in the intraoperative and postoperative periods. Increased vigilance for the development of PVC’s, ventricular arrhythmias, and TDP cannot be overemphasized. Preoperative beta blockade should be continued throughout the perioperative period.

The following drugs that are commonly administered in the perioperative period have been shown to increase the QT interval: thiopental, volatile agents, succinylcholine, anticholinergics, neostigmine, and ondansetron. However, the clinical significance of this is unclear; there are many reports of patients receiving these drugs without cardiac complications or the development of TDP.

The following medications have not been shown to significantly increase the QT interval or increase the risk of TDP development: midazolam, propofol, nondepolarizing neuromuscular blocking agents, opioids, nitrous oxide, and local anesthetics.

Numerous drugs have been shown to increase the QT interval. Several drugs have been removed from clinical use or restricted due to reports of prolongation of the QT interval and the development of ventricular arrhythmias. A comprehensive list of medications and their risk of causing TDP can be found on the Arizona Center for Education and Research on Therapeutics website http://www.azcert.org/medical-pros/drug-lists/drug-lists.cfm (accessed October 2011).
Pediatric Advanced Life Support (Questions 8-13):

In late 2010, the American Heart Association (AHA) released updates to Pediatric Basic Life Support (BLS) and Pediatric Advanced Life Support (PALS). Most of the PALS algorithms have been left unchanged. A major change in the 2010 Pediatric BLS guidelines is regarding the order of cardiopulmonary resuscitation. The previous "ABC" order of airway, breathing, and then circulation has been replaced with "CAB"; this sequence requires chest compressions to be started before providing airway management.

The change from "ABC" to "CAB" was made by the AHA for the following reasons:
- Better outcome in adults if compressions started early
- Minimal delay in starting ventilations for children
- Less delay in ability to start compressions
  - Head positioning/opening airway
  - Unwillingness to provide rescue breaths

Other changes made to the 2010 Pediatric BLS guidelines include:
- Pulse check: Take no longer than 10 seconds
- If no pulse or unsure, start chest compressions
- "Look, listen, and feel" terminology eliminated
- Apply an Automatic External Defibrillator (AED) if available
  - Ideally with a pediatric attenuator if <8 years old
  - May use a standard AED in all patients if a pediatric attenuator is unavailable

Other significant changes made to the 2010 PALS guidelines include:
- Oxygen concentration
  - Use of continuous pulse oximetry is strongly recommended
  - Appropriate to use 100% oxygen concentration during resuscitation
  - Once stabilized, titrate the oxygen concentration to maintain an oxygen saturation of >94%
- No recommendation for routine cricoid pressure during intubation (for the prevention of aspiration)
- Use of the actual body weight for dose calculations
- Calcium nor sodium bicarbonate are routinely recommended
- Defibrillate first at 2 Joules/kg; the maximum has been increased to 10 Joules/kg
- New sections on Congenital Heart Disease
  - Single ventricle: consider the use of heparin, maintaining an oxygen saturation of approximately 80%, the use of medications to lower the systemic vascular resistance, and the use of extracorporeal membrane oxygenation (ECMO)
Pulmonary Hypertension: consider the use of inhaled nitric oxide, prostacyclin, and ECMO

- Etomidate is not recommended for patients with suspected septic shock
- Associated with increased mortality
- Wide-complex tachycardia is defined as QRS width >0.09 sec.
- No reliable predictors to guide termination of efforts
- Prefer IV or IO vs. ETT for drug delivery
- Vasopressin is still not part of the PALS guidelines

Initial steps during a cardiac arrest in the operating room should include calling for help and the code cart/defibrillator. Verification of a secure airway and ventilation should follow. Chest compressions should be started without delay. The FiO₂ should be 100% with the vaporizers turned off. Emergency drugs can be delivered by endotracheal or intraosseous routes if loss of intravenous access should occur. The smaller defibrillator paddles are used for infants less than 1 year old or less than 10 kg.

The most recent 2010 PALS Pulseless Arrest Algorithm incorporates 4 cardiac rhythms that are divided into shockable and not shockable categories. Pulseless electrical activity (PEA) and asystole comprise the not shockable rhythms. Ventricular fibrillation (VF) and pulseless ventricular tachycardia (VT) include the shockable rhythms. The first decision to be made is if the rhythm is shockable (VF/VT) or not shockable (Asystole/PEA).

The tachycardia algorithm of the 2010 PALS guidelines assumes a pulse is present along with evidence of poor perfusion. First, determine if the QRS duration is narrow or wide. If the QRS duration is narrow, assume supraventricular tachycardia (SVT) is present. SVT is a rapid, regular rhythm with nonidentifiable P waves. SVT may be distinguished from sinus tachycardia by a rate that is usually greater than 220 in infants and greater than 180 in children. SVT with a pulse can initially be treated with adenosine if intravenous access is present or synchronized cardioversion. Vagal maneuvers may be attempted first as long as no delay occurs in providing drug or electrical therapy. Adenosine is administered at a dose of 0.1 mg/kg followed by 0.2 mg/kg if the first dose is ineffective. Adenosine must be flushed in rapidly during its administration. If adenosine is ineffective or if intravenous access is not present, synchronized cardioversion is used with an initial dose of 0.5-1 J/kg followed by 2 J/kg for further attempts. If cardioversion remains ineffective, consultation with a pediatric cardiologist is strongly encouraged. In addition, amiodarone 5 mg/kg given over 20-60 minutes or procainamide 15 mg/kg given over 30-60 minutes should be administered.

If a wide QRS duration is present, treat as presumptive ventricular tachycardia using synchronized cardioversion at 0.5-1.0 J/kg followed by 2 J/kg if ineffective. If cardioversion is ineffective, consultation with a pediatric cardiologist is strongly encouraged. In addition, amiodarone 5 mg/kg IV given over 20-60 minutes or procainamide 15 mg/kg IV given over 30-60 minutes should be administered.
A summary for the management of a pediatric patient with a rapid pulse and with poor perfusion includes:

- Determine the QRS interval
- Normal QRS interval (Sinus Tachycardia): no treatment vs. underlying cause
- Narrow QRS interval (Supraventricular Tachycardia): vagal maneuvers, adenosine (0.1 then 0.2 mg/kg), cardioversion (0.5-1 J/kg then 2 J/kg)
- Wide QRS (Ventricular Tachycardia): cardioversion, then amiodarone 5 mg/kg over 20-60 min or procainamide 15 mg/kg over 30-60 min
  - Expert consultation recommended; don’t give both medications
  - Avoid adenosine if known Wolff-Parkinson-White syndrome

For shockable rhythms such as VF or VT, defibrillate one time at 2 J/kg and resume cardiopulmonary resuscitation (CPR) immediately. Five cycles of CPR follow. If the rhythm is still shockable, defibrillate once at 4 J/kg and resume CPR. After defibrillation, epinephrine is given every 3-5 minutes. The dose for epinephrine is 10 mcg/kg IV or IO and 100 mcg/kg if via the endotracheal tube. Five cycles of CPR occur followed by rhythm evaluation. If the rhythm is shockable, defibrillate at 4 J/kg and resume CPR. One then will administer a bolus of amiodarone 5 mg/kg IV. Cycles of 1 shock followed by 5 rounds of CPR is repeated until the rhythm is not shockable or efforts are terminated.

A summary for the management of a pediatric patient without a pulse and with a shockable rhythm includes:

- Defibrillate 2 J/kg, 5 cycles CPR, Reassess
- If still shockable:
  - Defibrillate 4 J/kg
  - Administer epinephrine bolus 10 mcg/kg every 3-5 minutes
  - Provide 5 cycles of CPR
  - Reassess and repeat if applicable
- If still shockable:
  - Defibrillate 4 J/kg then bolus amiodarone 5 mg/kg
  - Magnesium only if suspected Torsades (25-50 mg/kg)
  - Provide 5 cycles of CPR then Reassess

Torsade de pointes (TDP), a French term meaning "twisting of the points", can be classified as a less common form of polymorphic, ventricular tachycardia. TDP commonly spontaneously resolves or can progress into other arrhythmias such as ventricular fibrillation and ultimately asystole. The ventricular rate in TDP can vary from 150-250 beats per minute. In TDP, the QRS complexes will be seen rotating around the horizontal axis. Patients with TDP may present pulseless and in cardiac arrest. However, many patients may present with ventricular tachycardia with a pulse. Management of TDP centers on magnesium sulfate administration and the implementation of emergency cardiac care guidelines such as PALS.
References:

**General Textbook:**


**Journal Articles; PALS:**


**Journal Articles; Prolonged QT/Long QT Syndrome:**


**Journal Articles; Infective Endocarditis Prophylaxis:**