Post-op Cardiac Arrest in Patient with Long QT syndrome

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Introduction:
Most cardiovascular events in pediatrics become apparent after an inciting respiratory event. Sudden death in pediatrics due to a cardiac cause such malignant dysrhythmia is less common. Dysrhythmia such as that of congenital long QT syndrome is relatively rare but an important consideration. Familial long QT syndrome is genetically transmitted due to defects in ion channels. It is can be first noticed by symptoms such as palpitations, syncope, and epilepsy. However, in this case present we had a patient with developmental delay who may not be able to report abnormal symptomatology.

Perioperatively, patients with prolonged QT may be at risk for life threatening dysrhythmias due to fluid shifts, electrolytes changes, and medications that may further prolong the QT interval. Nearly 75% of prolonged QTc are due to one of five gene mutations (KCNQ1, KCNH2, KCNE1, KCNE2, and SCN5A). Ion channel defects are inherited in an autosomal dominant manner and therefore genetic testing is warranted. Both the Heart Rhythm Society (HRS) and the European Heart Rhythm Association (EHRA) both recommend genetic testing to be offered for prepubescent individuals with QTc over 480ms and those with a prolonged QTc of greater than 500ms as adults. Perioperative management of patients with prolonged QT would necessitate optimization of electrolytes, optimization of hemodynamic parameters, and avoidance of drugs that prolong QT intervals. Preparation may also include the availability of defibrillation equipment if a malignant rhythm were to develop. An extensive updated list may be found on www.crediblemeds.org. Further, definitive management should include consultation with genetic counselors and a pediatric cardiologist for implantation of a pacemaker/ICD.

Discussion:
We present a case of a 4 year-old male with a history of spastic quadriaparesis, developmental delay, recurrent otitis media, chronic lung disease from chronic aspirations, and medically intractable epilepsy since the age of 6 months complicated by epileptic encephalopathy. He presented for G-tube placement, myringotomy, and Nissen fundoplication. No surgical complications were reported intra-op. Anesthetic was performed using mask induction with sevoflurane. During his anesthetic, he received ofloxacine ear drops, propofol, sevoflurane, fentanyl, and ondasentron. Recovery in post-op recovery unit was uneventful. However, post-operatively he decompensated into cardiac arrest, ventricular fibrillation with a prolonged QTc for which he was successfully resuscitated. Further, EKGs showed that his QTc remained prolonged at 567ms. No previous family history of prolonged QTc intervals were found in either of his parents. EKG analysis of his family members showed normal QTc intervals and did not reveal a genetic cause for his QT prolongation. Months later he underwent an uneventful placement of an pacemaker/ICD.

Case Report:
Identification of pediatric patients with prolonged QT syndrome starts with a thorough family history and review of patient’s symptoms. Malignant dysrhythmias can be prevented by avoidance of QT prolonging medications. Furthermore, patients at risk for dysrhythmias have life saving equipment available for emergent resuscitation.

Conclusion:
Identification of pediatric patients with prolonged QT syndrome starts with a thorough family history and review of patient’s symptoms. Malignant dysrhythmias can be prevented by avoidance of QT prolonging medications. Furthermore, patients at risk for dysrhythmias can have life saving equipment available for emergent resuscitation.

Areas of Improvement:
1) Avoid QT prolonging medications
2) Defibrillator pads (significant cost. Is it worth doing for every patient?)
3) If pacemaker, should we overpace patients if significant concerns.
4) Optimize electrolytes

References: