Short QT Syndrome in the Pediatric Age Group - Rare and Challenging Diagnosis

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Background: Short QT syndrome, is a fairly novel entity in the medical literature, but with very high morbidity and mortality. At this time, there is scarcity of information regarding all aspects of the disease, especially management in children.

Aim: We present a case of the new diagnosis of Short QT syndrome in a 13 months old baby that came to our attention prior to starting beta blockers for facial hemangeoma, due to a family history of aborted cardiac arrest.

Results: The child's father at age 31 years, prior to patient's birth, experienced sudden cardiac arrest and underwent resuscitation following which he had an intracardiac defibrillator implanted. At the time of presentation the child was asymptomatic and during the clinic visit had a normal cardiac physical exam and echocardiogram. On the EKG the patient was found to have a QTc interval of 0.294, no ST segment, and no peaked T-waves. It was decided to start Quinidine, to defer ICD implantation and refer the family for genetic testing.

Conclusions: The definition of the Short QT interval is not clear at this time, but appears to be QTc less than 320ms while QTc less than 360ms is highly suggestive. Most patients are male, with >50% of the cases having a significant family history of cardiac arrest. Present genetic studies have identified 3 genes; gain of function mutation of a potassium channel and 2 loss of function mutations in a calcium channel. Current treatment at this time is uncertain. It does appear that patients who carry the potassium channel mutation benefit from treatment with Quinidine. Among adults who experienced a sudden cardiac arrest an ICD is recommended, however in pediatrics it is a more complex decision, given the rapid growth of a child, device sizes and the overall higher complications rate. Further studies are needed and probably an establishment of international database could benefit gathering of information about this rare condition.