Congenital long QT syndrome type 1

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Introduction: Congenital long QT syndrome (LQTS) is a potentially lethal cardiac channelopathy and one of the leading causes of sudden cardiac death in the young. Disease prevalence is estimated nearly 1 in 3000 people. There have been impressive advancements in pathogenic mechanisms for LQTS, which enable us to classify the disease into different types. Long QT syndrome type 1 is triggered by exercise, associated with mutations in KCNQ1 gene which is responsible for slow component of the delayed rectifier repolarizing current (IKs). We report the case of type 1 LQTS based on the result of epinephrine QT stress testing.

Case: A 20-year old man has been presented with syncope and seizure like episodes since he was 3 years old. These episodes usually lasted for 5-10 minutes and spontaneously subsided. His EKG showed a sinus rhythm with prolong QTc interval of 476ms. Diagnosed with congenital LQTS, he took propranolol and atenolol, which were effective until 19 years of age. However, after that, he felt fluttering and dizziness again. In most cases, this symptom developed after exercise, especially running. His family history was not significant for syncope or premature sudden death. His lab findings, 2-D echo and holter EKG test results were also unremarkable. Thus we conducted an epinephrine infusion test according to the protocol of Shimizu and Antzelevitch. Immediately after the bolus injection of 0.1 μg/kg of epinephrine, the EKG showed marked prolongation of QTc from 512ms to 617ms. When he was challenged with isoproterenol after head up tilt test, blood pressure decreased accompanied by dizziness and nausea and T-alterans EKG pattern. Putting together his clinical history and provocation results, we made the diagnosis of a long QT syndrome, probably type 1. He was advised to avoid strenuous exercise and ensure regular full doses of beta-blocker.

Conclusion: In this case, the most likely cause of the QT prolongation is exercise, but his several holter EKG indicated no ventricular tachycardia. A provocative test using epinephrine showed the marked increase in QTc interval. Therefore, the diagnosis is LQTs type 1, but further genetic tests are required to confirm that.

Effect of triple antiplatelet therapy on small coronary artery treated with Zotarolimus Eluting Stent

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Background: In previous study, target lesion revascularization was greater in small caliber vessel treated with Zotarolimus Eluting Stent (ZES). Cilostazol has been known that it reduced restenosis and repeat intervention after drug-eluting stent (DES) implantation. Objectives: We evaluate clinical outcome and angiographic finding of patients who had small coronary artery with treated ZES. SUBJECTS AND Methods: We searched our database (from January 2006 to December 2008) and found 125 patients who were undergoing percutaneous revascularization with 2.25mm or 2.5mm ZES in de novo native coronary lesion. 32 patients were treated with triple antiplatelet therapy. The primary end points were follow-up angiographic binary restenosis rate and Major adverse cardiac event (cardiac death, myocardial infarction, target lesion revascularization percutaneous coronary intervention(TLR-PCI), Target vessel revascularization -PCI, stent thrombosis) Results: 125 patients were eligible for analysis. The patients who were treated with triple antiplatelet therapy were 32. For baseline characteristics, there was no significant difference between both groups. In angiographic finding, Patients with triple antiplatelet therapy had more stent number, longer lesion length, stent length, smaller minimal lumen diameter, more severe diameter stenosis. But about lesion type, location were not difference statistically between both groups. There were no significant reduction of TLR-PCI, MACE in triple versus dual antiplatelet therapy. Also restenosis rate was not different in stenotically. (triple 21.9% vs. dual 16.5%). Conclusion: In this study, Patients with triple antiplatelet therapy had more tough coronary artery lesion angiographically. But clinical outcome was similar between two group. For small coronary artery lesion treated with ZES, triple antiplatelet therapy with cilostazol may be effective. A larger scale study is needed to clarify efficacy of the ZES in this setting.