Arrhythmias and Clinical EP

IMPLANTABLE CARDIOVERTER DEFIBRILLATORS IN PRIMARY ARRHYTHMIA SYNDROMES: OUTCOME RELATIVE TO MEDICATION

Poster Contributions
Hall C
Monday, March 31, 2014, 9:45 a.m.-10:30 a.m.

Session Title: Device Therapy in Patients with Heart Failure and Cardiomyopathy
Abstract Category: 8. Arrhythmias and Clinical EP: Devices
Presentation Number: 1255-107

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Background: The initial presentation of catecholaminergic polymorphic ventricular tachycardia (CPVT) and long QT syndrome (LQTS) may be aborted sudden cardiac death. Management commonly includes secondary prevention ICD implantation, along with medication. We hypothesize that appropriate medical therapy is adequate first line treatment and ICD implantation may be avoided in these patients (pts). In this study we aim to describe the outcome of CPVT and LQTS pts receiving an ICD at a single center with attention to adequacy of medical therapy.

Methods: Retrospective cohort study of all patients at a single center with an ICD for CPVT or LQTS followed from 2005-2013. Adequate drug therapy (ADT) defined as β-blocker dose of at least 1mg/kg/day for LQTS and CPVT or mexiletine for LQTS type 3 and documented compliance at time of appropriate shock.

Results: There were 24 LQTS and 13 CPVT pts. Median age at implant 13.3 (IQR 10.3-16) yrs. Secondary prevention (2P) in 31 (72%). Five CPVT and 19 LQTS pts were on ADT. Total follow-up was 285 pt-yrs. Median 5.8 yr (IQR 3.6 - 9.6)/pt. Ten pts received at least 1 appropriate therapy; 8 CPVT (1 on ADT) and 2 LQTS pts (non genotyped siblings both on ADT). No genotype positive LQTS pt received an appropriate therapy. LQTS and CVPT pts on ADT were more likely to not receive appropriate ICD therapy vs. pts not on ADT, OR 6.6 (CI 1.3 - 32.5; p=0.02); in 2P only, OR 5.5 (95% CI: 0.9 - 33.2, p=0.09). Kaplan-Meier showed 75% of pts on ADT were shock-free at max follow-up 12.6 yr vs median shock-free survival in non-ADT pts 8.1 yr (log-rank p= 0.02). Median shock-free survival after 2P was 5.2 yr for pts on ADT and 3.3 yr for non-ADT (log-rank p=0.2, Wilcoxon p=0.09). Cohort complications included 141 inappropriate shocks in 16 pts, 24 non-routine procedures (including 11 lead extractions) and 2 lead perforations.

Conclusions: Shock free survival was 75% at max follow-up of 12.6 yrs, if pts were on ADT. The risk of shock was significantly reduced by ADT. However an acceptable incidence of appropriate shocks was observed in these pts. The study was underpowered to detect a difference after 2P ICD. Larger multi-center studies are needed to evaluate the outcome of ICD prior to failure of ADT in this population.