INCIDENCE OF DEVICE THERAPY AND COMPLICATIONS IN PATIENTS WITH ANDERSEN-TAWIL SYNDROME WITH ICDS

ACC Poster Contributions
Ernest N. Morial Convention Center, Hall F
Monday, April 04, 2011, 9:30 a.m.-10:45 a.m.

Session Title: Clinical and Financial Implications of Complications
Abstract Category: 48. Genetics and Clinical Outcomes
Session-Poster Board Number: 1101-137

Authors: Kelly Airey, Arthur Wilde, Nynke Hofman, Susan Etheridge, Louis Ptacek, Hussam Abuissa, Caroline Nubel, Martin Tristani-Firouzi, Creighton University Medical Center, Omaha, NE, Nora Eccles Harrison CVRTI, Salt Lake City, UT

Introduction: Andersen-Tawil Syndrome (ATS) is a rare genetic disorder characterized by ventricular arrhythmias, muscle weakness and dysmorphic features. There are no specific guidelines for implantation of ICD devices in this population. In this study, we determined the incidence of device therapy, burden of arrhythmias, indication for implant and device complications.

Methods: Our ATS registry consists of 147 patients including 52 patients with cardiac manifestations (ventricular arrhythmias, syncope and/or ICDs). The group ranged in age from 12-84 yrs (mean 38 yrs; 81 % female) with follow-up of 11 yrs ±12. The database was examined for ICD implants, therapy and complications. 16/52 ATS subjects with cardiac manifestations received an ICD.

Results: Three patients (19%) received ICD implantation for documented VF arrest. Four (25%) patients received an ICD for a history of non-sustained bidirectional or monomorphic VT and syncope. Three patients (19%) underwent implantation for QTc prolongation and syncope or family history of sudden cardiac death (SCD). The remaining 6/16 (38%) underwent implantation for ventricular ectopy without syncope.

The three patients with devices implanted for resuscitated VF arrest and one with bidirectional VT and syncope went on to have appropriate shock therapy. None of the remaining 12 patients had appropriate device intervention. Inappropriate shocks occurred in 8/16 (50%) patients, 5 due to lead fracture requiring extraction/revision and 3 due to ventricular high rates. Other device related complications included system extraction due to SVC syndrome and anxiety, pneumothorax at implant and atrial lead dislodgement.

Conclusions: We report the long-term follow up of ATS patients implanted with ICDs. Implant related complications, including lead fractures and inappropriate shocks occurred frequently in this cohort. Appropriate device therapy occurred only in subjects with a prior history of aborted SCD or bidirectional ectopy with syncope. Longitudinal, collaborative studies are needed to better define the subset of ATS patients who are at risk of SCD and require ICD implantation.