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Risk stratification for AICD implantation for hypertrophic cardiomyopathy in the young

Introduction: Ninety-five percent of hypertrophic cardiomyopathy are related to defects present in four genes MYH7, MYBPC3, TNNT2 and TNN13. The cause of sudden cardiac arrest (SCA) in hypertrophic cardiomyopathy has not been determined. The guidelines for risk stratification for SCA and AICD implant from the ACC and AHA are not based on any large studies in children. Anecdotal cases do not support their usefulness. We report our data base analysis of a cohort of patients with HCM followed at a single center from 1995-2017 and found no consistent risk factors for SCA.

Methods: We reviewed our database for all hypertrophic cardiomyopathy patients followed by pediatric cardiac electrophysiology. Data was tabulated for presenting symptoms, EKG finds, ambulatory monitoring, and echocardiogram measurements. In decreased patients, autopsy results were also compiled and tabulated.

Results: Total of 57 candidates (37 male and 20 female) of age 1-29 (mean 19) years with 3 syncope 6 SCA as first symptom (3 resuscitated) and palpitations 20/57 were included in the study. 7/57 had family history of HCM 3/57 (SCA). 16/57 had no sustained VT (ambulatory monitor). 5/47 AICD received appropriate therapy (only 1/5 met guidelines for AICD) 2/47 patients inappropriate therapy. 1/47 with AICD died from ventricular fibrillation (patient within guidelines for AICD).

Conclusions: We could not identify specific criteria for risk stratification of SCA/AICD implantation in our population of patients with hypertrophic cardiomyopathy. The current guidelines are not sensitive or specific enough in children to guide AICD implant. The risk for SCA likely resides in the cellular dysfunction and may be related to the genetics. Until larger studies could better risk stratify SCA the decision for AICD should be discussed with the patient and a decision made even if the guidelines are not met.

Biography

Charles H Gaymes was has completed his graduation from High School with top honors and attended the University of the West Indies Medical School, graduating class of 1978. He completed Residency in Pediatrics at the University of Mississippi Medical Center in 1989 and was "Outstanding Resident" in 1989. He completed his Pediatric Cardiology Residency at MUSC, South Carolina in 1992. He has been on faculty at UMMC 1992 – Present. He was "Outstanding Pediatric Faculty Member" in 1995. Currently, he practices as a Professor of Pediatrics/Cardiology and Director of Children Arrhythmia Services.

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