SAFETY THRESHOLD FOR CATECHOLAMINERGIC POLYMORPHIC VENTRICULAR TACHYCARDIA IN CHILDHOOD UNDER BETABLOCKER TREATMENT

Poster Contributions
Hall C
Sunday, March 30, 2014, 3:45 p.m.-4:30 p.m.

Session Title: Arrhythmias and Clinical EP: New Observations Affecting Clinical Management
Presentation Number: 1217-112

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Background: Catecholaminergic polymorphic ventricular tachycardia (CPVT) is a channelopathy characterized by high incidence of sudden cardiac death in the childhood and youth. However, very young children do not have events or symptoms in this disease. During 5 years, our group has performed a protocol for treatment and follow-up to the majority of the 190 CPVT-related mutation alive carriers (RyR2-G357S) found in Gran Canaria (Spain).

Methods: 17 carriers were identified since 2007 being less than 9 y. at the diagnosis. 3 were followed at another center and 3 were too young to perform any test. First ET was performed without treatment, and then all begun weight-adjusted BB. Data Pre Vs. Post 9 y. were compared using the McNemar test.

Results: 11 carriers (mean age at diagnosis ± SD: 5.96 ± 1.99 y. range 2.7-8.7) performed 88 ET (median ± SD: 10 ± 5.2, range 1-17) during follow-up. All ET under 9 y. (60) were negative for any VA (P<0.001, Figure). 5 children crossed the threshold of 9 y. Two of them (1 male) showed VA in ET after reach nine (9.25 y. & 9.58 y.), and since then showed VA in 2/6 and 7/15 ET, respectively. None of them presented symptoms or events. Weight or functional capacity did not seem to provide more information about safety threshold.

Conclusion: In our series, early childhood under age 9 is a safe period in carriers of a pathogenic mutation related to CPVT under BB. The identification of this safety age threshold may have consequences on the recommendations for the management of genetically positive CPVT children.