Result of implantatable cardioverter-defibrillator in children with hypertrophic cardiomyopathy: an european experience

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Background: Hypertrophic cardiomyopathy (HCM) is an important cause of life-threatening arrhythmias. Demonstrated risk factors (RF) guide primary prevention indication of cardiac defibrillator implantation in adult. Attitude toward pediatric patient is less consensual.

Objective: Describe result of primary and secondary prevention cardiac defibrillator implantation in pediatric patients.

Material and method: Multicenter retrospective study from 5 centers in Europe. 45 patients under 20 years old were gathered, 16 from Paris, 9 from Zurich, 7 from Madrid, 7 from Roma and 6 from Ljubljana.

Results: Among the 45 patient 31 are male. Median age at implantation is 12.4 years old (3-20). Family history of HCM was described for 20 patients (44%). Most of the patient, 35 (78%), were implanted for primary prevention of sudden death, 10 (22%) for secondary prevention. The average number of risk factor per patient is 2: 11 patients were implanted for only 1 RF, 14 patients had 2 RF and 10 patients had 3 or more RF. A great majority of them (26/45) were implanted transvenously. Appropriated shocks were experimented by 12 patients (27%): 5 in the primary prevention group (14%) and 7 in the secondary prevention group (70%). Thirteen patients had an ICD-related complication: 8 inappropriate therapies, 4 device or lead failures and 1 delayed therapy. Two patients died and one had major neurological complication after electric storm.

Conclusion: The rate of appropriated shock in the secondary prevention group is significantly higher than in the primary prevention group. The annual rate of appropriated shock in the primary prevention group is very low but the complications are frequent. This result suggests that the demonstrated risk factor for adult may not be accurate at the pediatric age.