RESULTS OF IMPLANTABLE CARDIOVERTER-DEFIBRILLATOR (ICD) IN CHILDREN WITH HYPERTROPHIC CARDIOMYOPATHY: AN EUROPEAN EXPERIENCE


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INTRODUCTION
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.

PATIENTS & METHODS
Retrospective, multi-centric study including hypertrophic cardiomyopathy patients aged under 18 from 5 tertiary paediatric cardiology centers in Europe

Risk markers of sudden cardiac death were defined as:
1) Family history of sudden death (SFam)
2) Unexplained syncope (syncope)
3) Abnormal blood pressure response during treadmill test (exBPres)
4) Non-sustained ventricular tachycardia (NSVT)
5) Massive left ventricular hypertrophy (mass/LVH)

Survival free of appropriate therapy was estimated by Kaplan-Meier method. Rates were compared by chi^2 test. A p value < 0.05 was considered statistically significant.

Patients 44
Male/Female 30/14
Age at implantation 11.6 years (3-18)
Weight at implantation 43 Kg (13.5-92)
ICD indication
- Primary prevention (PP) 35 (80%)
- Secondary prevention (SP) 9 (20%)
Risks Factors (SP/PP)
- 0-1 17 (8/8)
- 2 16 (0/16)
- 3 or more 11 (0/11)
Beta-blockers medication 39 (89%)
Surgical Septal Myectomy 11 (25%)
Heart transplant 3
Survival 42

RESULTS (I)
Appropriate therapy was experimented by 13/44 (29%) patients
7/35 (20%) in the PP group
6/9 (67%) in the SP group

RESULTS (II)
The rate of appropriate therapy is not influenced by the number of risk factors. As shown in Figure 3 there is no significant difference whether the patient had less than 2, 2 or more than 2 risk factors (Figure 3).

Considering each risk factor one by one, the rate of appropriated therapy was significantly more important when the patient had either history of unexplained syncope or massive left ventricle hypertrophy among its risk factors. (figure 4)

Thirteen patients had ICD-related complication:
8 inappropriate therapies
4 device or lead failures
1 delayed therapy

At 5 years the survival free of ICD related complication is 44%.

Two patients died and one had major neurological complication after electric storm.

DISCUSSION
Recently, a debate has arisen as to whether or not the adult’s established risk factors were accurate to identify, in the hypertrophic cardiomyopathy paediatric patient, the one at risk of cardiac sudden death.

Consistent with previous report, this study suggested that the type of RF matters more than the number.

Once implanted the likelihood of having a first cardiac event, for the Primary prevention group, or of having an other cardiac event, for the Secondary prevention group, is significantly higher for patient with massive left ventricle hypertrophy or prior unexplained syncope or pre-syncope.

CONCLUSION
Sudden death is a devastating consequence of hypertrophic cardiomyopathy. Implantable cardioverter-defibrillator has been shown to successfully prevent such event in selected population with hypertrophic cardiomyopathy. Since the complication rate of ICD is high in paediatric population, the indication of implantation in primary prevention is challenging. This study highlights the importance of syncope and massive left ventricle hypertrophy as makers of cardiac event.