

Non-compacted cardiomyopathy: clinical characteristics, evolution and prognostic data in childhood. Results of a multicentre study

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INTRODUCTION: Noncompaction of the ventricular myocardium (NCVM) is a rare congenital heart disease (CHD). Heightened awareness has resulted in an increased detection of the morphological features of NCVM in routine clinical practice.

METHODS: Multicentre study (6 Catalan hospitals) including paediatric patients affected by NCVM according to echocardiographic criteria of Chin and Jenni. Clinical features, echocardiographic, MRI, complications and treatments are reviewed. Descriptive data and statistical analysis is provided.

RESULTS: A total of 29 patients were included, 15 female and 14 male, the median age at diagnosis was 5 years and 7 months (range from birth to 17 years). In four cases (14%) cardiomyopathy was detected in utero. Sixteen patients (55%) presented as an isolated lesion, 8 (27,5%) had a ventricular septal defect associated one of them with aortic coarctation, 3 (10%) had an inborn error of metabolism, 1 (3,5%) had Juvenile Idiopathic Arthritis and 1 (3,5%) has a syndrome being studied. The location of the trabeculae has been predominantly at the apex, but also affected the left ventricle free wall in 11 patients (40%) and right ventricle in 2 (7%). MRI findings provided no further information. No complications have occurred in 12 patients (41%), cardiac failure 12 patients (41%), implantable cardioverter defibrillator was placed for ventricular arrhythmias in 2 patients (7%), stroke 1 patient (3,5%) and death 2 patients (7%), both of them less than 6 month of age ($p < 0,05$). Median follow up has been 12 month (range from 2 months to 8 years). Current treatment includes carvedilol, ACEI's and ASA and one patient is waiting for cardiac transplantation.

CONCLUSIONS: Early onset of symptoms is related with poor prognosis. The clinical and prognostic heterogeneity described supports the theory that NCVM is not a disease but a morphologic feature.