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Introduction

Noncompaction of the Ventricular Myocardium (NVM) is a rare condition with debatable clinical significance and no clear-cut morphological diagnostic criteria. The main feature is the excessive trabeculation of the myocardial walls with commitment of both ventricles. Non-systematic reviews show that some congenital cardiac defects can be associated with NVM.

Echocardiography is usually considered the gold standard for the diagnosis of NVM. There are 2 sets of echocardiographic criteria for NVM diagnosis: the Jenni criteria, which stress the presence of a 2-layered structure, and the Chin criteria, which focus on the depth of the recesses compared with the height of the trabecular layer.

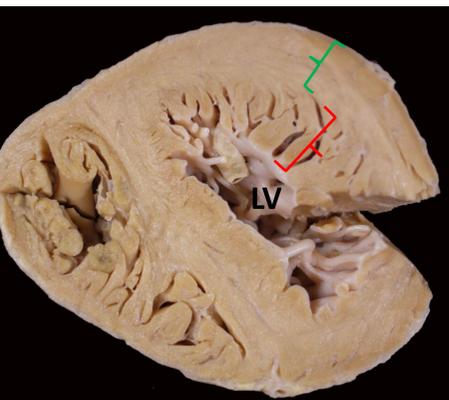
Objectives

The goal of this study was to evaluate the incidence of NVM in heart specimens with congenital cardiovascular defects.

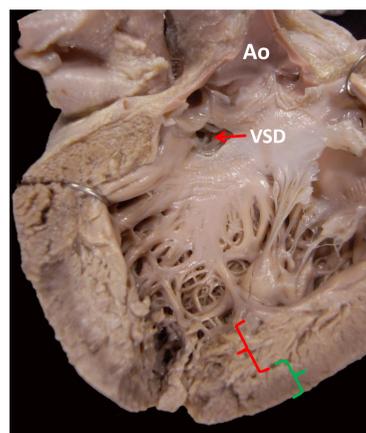
Methodology

The compacted and noncompacted myocardial layers were grossly measured in 206 heart specimens, at three different points of the left ventricular (LV) wall: inlet, apex and outlet. Coefficients of non-compaction were calculated according to the criteria described by Jenni and Chin. The Chin coefficient is calculated as the ratio between the distance from the epicardial surface to the trough of the trabecular recesses and distance from the epicardial surface to peak of trabeculation ≤ 0.5 . Jenni coefficient is defined by the ratio between the thicknesses of the noncompacted and the compacted layers, and considers as positive values those ≥ 2 for adults and 1.4 for children. This study includes 6 different congenital defects: isolated ventricular septal defect (VSD); isolated atrial septal defect (ASD); atrioventricular septal defect (AVSD); transposition of the great arteries (TGA); isomerism of the atrial appendages (ISO); Ebstein malformation of the tricuspid valve (EBS).

Gross evaluation



A short-axis section of the ventricles showing, at the apex of the left ventricle (LV), the prominent non-compacted layer (red bracket) as compared to the compacted one (green).

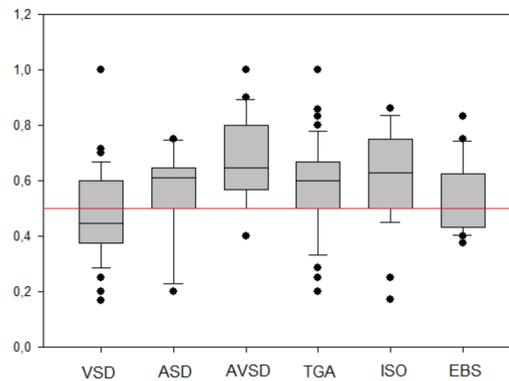


Left ventricle opened to show the perimembranous and sub-aortic (Ao) VSD and the hypertrabeculated myocardium at the apex. The non-compacted myocardial layer is marked by the red bracket while the compact layer by the green bracket.

Cardiac Defect	number of specimens	Number of cases with NVM - left ventricular apex, Chin coefficient	Number of cases with NVM - left ventricular apex, Jenni coefficient	Mean age (months)
VSD	48	25 (52.08%)	15 (31.25%)	4.5
ASD	13	1 (7.69%)	1 (7.69%)	13
AVSD	41	1 (2.44%)	1 (2.44%)	17
TGA	54	12 (22.22%)	9 (16.66%)	2
ISO	28	2 (6.9%)	2 (7.14%)	1
EBS	22	6 (27.27%)	2 (9.09%)	0
Total	206	47 (22.8%)	30 (14.6%)	4

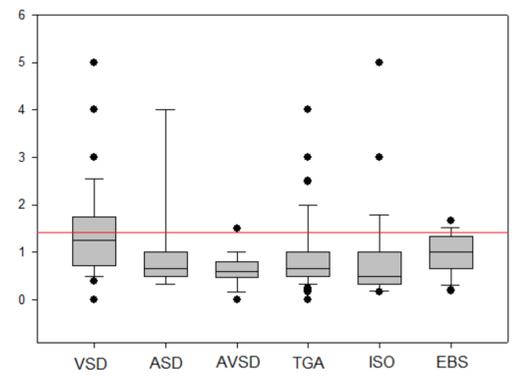
VSD: ventricular septal defect; ASD: atrial septal defect; AVSD: atrioventricular septal defect; TGA: transposition of the great arteries; ISO: isomerism of the atrial appendages; EBS: Ebstein malformation

Left ventricle apex, Chin coefficient



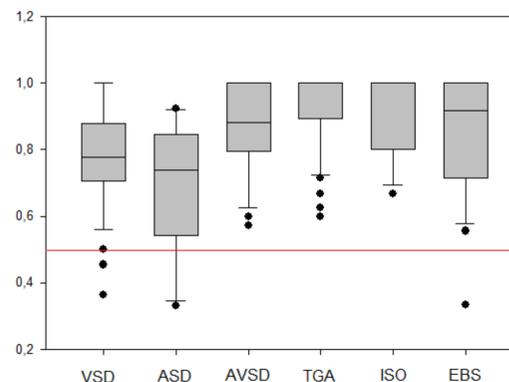
Abbreviations as the table above. Positive results for NVM are values below 0,5 (red line)

Left ventricle apex, Jenni coefficient. Children



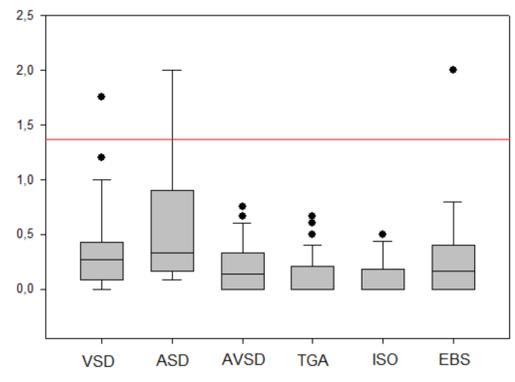
Abbreviations as the table above. Positive results for NVM are values greater than 1,4 (red line)

Left ventricle inlet, Chin coefficient



Abbreviations as the table above. Positive results for NVM are values below 0,5 (red line)

Left ventricle inlet, Jenni coefficient. Children



Abbreviations as the table above. Positive results for NVM are values greater than 1,4 (red line)

The Kappa test was used to compare the Chin and Jenni results.

In ASD, AVSD, TGA and isomerism there was an almost perfect agreement ($p < 0.001$); in Ebstein malformation there was a moderate agreement ($p = 0.023$), and in VSD a fair agreement ($p = 0.05$).

Once there was higher incidence in isolated VSD as compared to the other defects, the TGA cases were split into two groups, one with and other without VSD. The comparison failed to demonstrate any significant difference between them ($p > 0.05$).

CONCLUSIONS

Predominance of non-compaction at the LV apex of VSD heart specimens has not been previously described. The true significance of regional myocardial non-compaction in congenital heart defects is unknown, and needs correlation with clinical and outcome data.

RESULTS

Overall, NVM was more prevalent at the LV apex (Chin 22.8% and Jenni 14.6%) than at the inlet (1.46%) or the outlet (1.46%).