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Ventricular Noncompaction and the association with congenital heart defects

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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. Non-systematic reviews show that some congenital cardiac defects can be associated with NVM. The goal of this study was to evaluate the incidence of NVM in heart specimens with congenital defects.
Methods: The compacted and noncompacted myocardial layers were grossly measured in three different heart walls: inlet, apex and outlet of the left ventricle (LV). Coefficients of non-compaction were calculated according to the criteria described by Jenni and Chin.
Results: Five types of cardiac defects were studied: isolated ventricular septal defect (VSD, n=51), isolated atrial septal defect ASD, n=15), atrioventricular septal defect (AVSD, n=41), transposition of the great arteries (TGA, n=54), isomerism of the atrial appendages (n=29) and Ebstein malformation (n=24) (total=214 heart specimens; 46 % from male patients, 64% under 12 months of age). According to the Chin's criterium the percentages of non-compaction at the LV apex were respectively: 50.98% in VSD specimens; 25% in Ebstein malformation, 22.2% in TGA, 6.7% in ASD, 6.1% in Isomerism, and 2.4% in AVSD. According to Jenni's criterium the percentages were: 33.3% in VSD, 31.0% in Isomerisms, 16.7% in TGA, 8.33% in Ebstein, 6.7% in ASD and 2.4% in AVSD. Considering all hearts, the apex region showed the largest percentage of non-compaction (Chin=24.5%; Jenni=16.3%). Concomitance of non-compaction in different regions of the same heart was low (4 hearts according to Chin's criterium and 2 according to Jenni's).In the TGA group, there was no association of MVN and the presence of VSD (p=0.067). The Kappa test was used to compare the diagnosis of non-compaction according to both criteria. In ASD, AVSD, TGA and isomerism there was an almost perfect agreement ($p<0.001$; Kappa>0.8), in Ebstein there was a moderate agreement (p=0.023; Kappa=0.412), and in VSD there was a fair agreement (p=0.05; Kappa =0.289).
Conclusion: 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.