

Left ventricular noncompaction in cardiac explants from paediatric patients and adults with congenital heart disease: prevalence and associated factors

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INTRODUCTION: Left ventricular noncompaction (LVNC) is a rare anomaly characterized by prominence of the ventricular trabeculations that may be associated with severe heart failure and need for a heart transplant or occur as an associated feature in other cardiomyopathies or congenital heart defects. There are no data concerning the prevalence of LVNC in patients who have undergone heart transplantation nor its associated factors. Our objectives are to assess the prevalence of LVNC phenotype (focal or diffuse) as the main or associated diagnosis in cardiac explants from pediatric patients or adults with congenital heart disease investigating the association between LVNC and sex/age, comparing the gross findings with the data obtained through pre-surgical imaging studies. **METHODS:** Cardiac explants from paediatric patients and adults with congenital heart defects were selected. Medical records were consulted for the demographic, anathomopathological, echocardiographic and MRI data. Measurement of the compacted and noncompacted portions of the left ventricular wall was performed with a pachymeter at 3 different points: the inflow, the apical, and the outflow walls. The echocardiographic-based criteria of Chin and the Jenni were used to determine whether LVNC was present.

RESULTS: 94 hearts were evaluated, being 51.06% from male individuals. The average age was 9.81 ± 6.87 years. Three (3.19%) had LVNC as the main pre-transplant diagnosis. However, 59.57% of the heart specimens fulfilled any of the criteria for LVNC in at least one ventricular segment. 32.9% presented focal LVNC at just one point, 17% presented at 2 different points, and 7.4% at all 3 points. The apex had the highest prevalence of LVNC (28.72% under Jenni and 41.48% under Chin). Dilated cardiomyopathy was the most prevalent anatomopathological diagnosis (67.02%) overall and among the hearts with LVNC at gross evaluation (69.09%). The prevalence of LVNC in hearts with structural and restrictive cardiopathies were 52,4% and 61,5%, respectively. The average ejection fraction was $34.16 \pm 15.30\%$, but no difference was found between hearts with or without LVNC. **CONCLUSION:** Focal LVNC is a prevalent feature in cardiac explants from the paediatric population, present mainly at the ventricular apex and frequently associated with the primary diagnosis of dilated cardiomyopathy.