A rare case of left ventricular non-compaction in early infancy

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A 3-year old girl with multiple admissions for congestive heart failure associated with decompensation symptoms secondary to severe mitral insufficiency underwent valve repair (Fig. 1). Histopathological examination revealed endocardial fibroelastosis of dilated type (Fig. 2). Despite clinical recovery, the patient was listed for heart transplantation (Supplementary Videos 1 and 2).

Supplementary material (Video 1 and Video 2) is available at EJCTS online.

Figure 1: Preoperative echocardiographic image. Four-chamber view showing findings of atypical thickening and suspected mild non-compaction of the endomycocardium: scattered echogenic spots within the septum, echogenic papillary muscle. Severely dilated left atrium.

Figure 2: Histopathological testing. Left ventricular (LV) myocardial tissue with endocardial fibroelastosis, dilated type. (a) Hypocellular fibrous tissue with multiple lamellae of coarse elastic fibres arranged parallel to the luminal surface (van Gieson’s stain, ×200). (b) The endocardium is extremely thickened extending in the subendocardium (Masson’s trichrome stain, ×200). Note that this myocardial tissue represents the basis of a huge LV papillary muscle.