A 28-year-old male was admitted to our hospital due to heart failure secondary to dilated cardiomyopathy. His first heart failure episode occurred when he was 8 months old, triggered by a respiratory infection, with good response to medical treatment. He was diagnosed dilated cardiomyopathy. Afterwards, he remained asymptomatic until he reached the adult age.

The patient referred severe functional capacity impairment in the last 3 months. A cardiac MRI was performed, which showed marked severe dilatation and systolic dysfunction and heavy apical trabeculation of both ventricles (Panel A, short-axis view, Panel B, four-chamber view). After contrast administration, few small patchy endocardial areas of hyperenhancement could be seen in the left ventricle (Panel C, arrows).

Owing to worsening heart failure, the patient was transplanted 9 days later, with uneventful postoperative course. He remained in NYHA functional class I for 3 years.

The gross specimen of the explanted heart showed severe dilatation of both ventricles and heavy apical hypertrabeculation (Panel D). A fibrous layer with some thickened areas existed in the endocardium, including papillary muscles and chordae tendinae. There were neither valvular abnormalities nor other malformations. Histological examination showed thickened endocardium with fibrosis and elastic fibres in the left ventricle, compatible with the diagnosis of endocardial fibroelastosis (Panel E, Masson’s trichrome, collagen fibres stained in blue, Panel F, orcein staining for elastic fibres).

Endocardial fibroelastosis consists of a diffuse thickening of the endocardium resulting from proliferation of collagen and elastic fibres. It may be secondary to congenital heart disease, mainly left ventricle obstructive malformations and hypoplastic left ventricle. As for primary endocardial fibroelastosis, different pathogenetic mechanisms have been suggested, such as genetic factors, viral infections, carnitine deficiency, or transplacental crossing of maternal antibodies (anti-Ro, anti-La); it has also been suggested to be a nonspecific response to chronic myocardial dysfunction.

Endocardial fibroelastosis is an adverse prognostic factor for children and adolescents with dilated cardiomyopathy. Interestingly, our patient showed an unusually benign course despite extensive fibroelastosis and remained in a good functional class until his 20s.