Evolutionary Persistence of Spongy Myocardium in Humans

Annalisa Angelini, MD; Paola Melacini, MD; Fabio Barbero, MD; Gaetano Thiene, MD

A 15-year-old boy underwent successful cardiac transplantation for severe congestive heart failure. His sister had died at the age of 9 years of undefined congenital heart disease. At the age of 10 years, during a school medical screening, the patient was diagnosed with a cardiomyopathy. At cardiac catheterization, the right ventricle disclosed an increased trabecular pattern, mainly in the region of the apex, and the left ventricle showed an increased wall thickness, with mild hypokinesia. Subsequently, the patient complained of dyspnea on effort and presented signs of congestive heart failure. Five years later, the boy was admitted to an intensive care unit requiring a continuous inotropic drug infusion. Two-dimensional echocardiography disclosed biventricular dilatation with diffuse hypokinesia and a hyperechogenic coarse endocardial shape (a and b) interpreted as apical mural thrombi (arrow). The heart removed at transplantation was markedly dilated (weight, 720 g) and revealed the gross morphological features of noncompaction (spongy myocardium) with numerous, excessively prominent trabeculations and deep intertrabecular recesses penetrating into the endocardium (c). Histological examination confirmed that the spongy appearance was due to the deep intertrabecular recesses, lined by endothelium, which spread close to the epicardial surface (d). This feature strongly resembles the spongy myocardium pattern of nonmammalian vertebrates such as fish, amphibians, and reptiles.
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_Circulation_. 1999;99:2475
doi: 10.1161/01.CIR.99.18.2475

_Circulation_ is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/99/18/2475

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