

**Clinical vignette**

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Isolated ventricular non-compaction with restrictive cardiomyopathy

Claudio Rapezzi1*, Ornella Leone2, Marinella Ferlito1, Elena Biagini1, Fabio Coccolo1, and Giorgio Arpesella3

1 Institute of Cardiology, University of Bologna and S. Orsola-Malpighi Hospital, Via Massarenti 9, 40138 Bologna, Italy; 2 Department of Pathology, University of Bologna and S. Orsola-Malpighi Hospital, Bologna, Italy and 3 Department of Cardiovascular Surgery, University of Bologna and S. Orsola-Malpighi Hospital, Bologna, Italy

* Corresponding author. Tel: +39 051349858; fax: +39 051344859. E-mail address: crapezzi@aosp.bo.it

A 42-year-old man with chronic heart failure and permanent atrial fibrillation was referred to our cardiology institute for diagnostic assessment and therapy. The patient denied any known family history of heart disease. No clinical/echocardiographic sign of heart disease was detected among living first-degree relatives. Atrial fibrillation and progressively worsening effort dyspnea had started at age 25. Physical examination showed moderate bilateral leg oedema, marked hepatomegaly, and raised jugular venous pressure and gallop rhythm. ECG showed atrial fibrillation with right bundle branch block and left anterior hemiblock. Cardiomegaly and chronic interstitial lung oedema were apparent on chest X-ray. Echocardiography revealed a normally sized, mildly hypokinetic left ventricle (ejection fraction = 45%) and an enlarged, mildly hypokinetic right ventricle, accompanied by massive right and left atrial enlargement and severe tricuspid regurgitation. The apical portions of both ventricles had a coarsely trabeculated, spongy appearance suggestive of non-compaction (Panel A). E-wave deceleration time was abnormally shortened in the echo-Doppler profile at transmitral level (Panel B). Coronary arteries were normal at angiography. At the right heart catheterization, restrictive physiology was evident (Panel C). Pulmonary artery pressures were 40/20/22 mmHg; mean pulmonary capillary wedge and right atrial pressures were 22 and 15 mmHg, respectively; cardiac index was 2.1 L/min/m². The patient was submitted to heart transplantation, which was successfully performed. The explanted heart (Panel D) provided definitive confirmation of biventricular non-compaction and restrictive cardiomyopathy (without signs of infiltrating myocardial diseases or desmin accumulation). Ventricular non-compaction has been shown to occur in the context of congenital anomalies, otherwise normal hearts or, most often, dilated cardiomyopathy. To our knowledge, the present case provides the first documentation of (b)iventricular non-compaction in the context of a restrictive cardiomyopathy. This observation is in line with the concept that isolated ventricular non-compaction is more likely to be a morphological trait that can be found within different types of cardiomyopathy rather than a distinct cardiomyopathy.

Panel A. Two-dimensional echocardiography in four-chamber view shows huge biastral dilation, a normally sized left ventricle, and a moderately enlarged right ventricle. The apical portions of both ventricles have a coarsely trabeculated, spongy appearance suggestive of ventricular non-compaction. E-wave deceleration time was abnormally shortened in the echo-Doppler profile at transmitral level (Panel B). Coronary arteries were normal at angiography. At the right heart catheterization, restrictive physiology was evident (Panel C). Pulmonary artery pressures were 40/20/22 mmHg; mean pulmonary capillary wedge and right atrial pressures were 22 and 15 mmHg, respectively; cardiac index was 2.1 L/min/m². The patient was submitted to heart transplantation, which was successfully performed. The explanted heart (Panel D) provided definitive confirmation of biventricular non-compaction and restrictive cardiomyopathy (without signs of infiltrating myocardial diseases or desmin accumulation). Ventricular non-compaction has been shown to occur in the context of congenital anomalies, otherwise normal hearts or, most often, dilated cardiomyopathy. To our knowledge, the present case provides the first documentation of (b)iventricular non-compaction in the context of a restrictive cardiomyopathy. This observation is in line with the concept that isolated ventricular non-compaction is more likely to be a morphological trait that can be found within different types of cardiomyopathy rather than a distinct cardiomyopathy.

Panel A. Two-dimensional echocardiography in four-chamber view shows huge biastral dilation, a normally sized left ventricle, and a moderately enlarged right ventricle. The apical portions of both ventricles have a coarsely trabeculated, spongy appearance suggestive of ventricular non-compaction. Panel B. Transmirtal echo-Doppler profile indicates very short deceleration time, suggesting restrictive physiology.

Panel C. Right ventricular catheterization trace (bottom) shows 'square-root' morphology in the diastolic portion of the curve accompanied by protodiastolic pressure above zero, confirming restrictive physiology.

Panel D. Macroscopic longitudinal section of the explanted heart (lacking the upper portion of the atrial cuff) clearly shows biventricular non-compaction with coarse apical trabeculation and deep inter-trabecular recesses. The non-compacted portions of both ventricles are predominant, drastically limiting cavity volumes. The histological detail (inset) after Mallory's trichrome staining additionally shows mild fibrotic endocardial thickening and moderate interstitial fibrosis.

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