Diagnosis of biventricular non-compaction cardiomyopathy by real-time three-dimensional echocardiography

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Non-compaction of the ventricular myocardium is a recently recognized rare disorder of the endomyocardial morphogenesis. The disease can be characterized by systolic and diastolic heart failure, ventricular arrhythmias and systemic embolization. The present case suggests the clinical role of real-time three-dimensional echocardiography in the spatial evaluation of both ventricles in suspected biventricular non-compaction cardiomyopathy.

KEYWORDS

Biventricular; Non-compaction; Cardiomyopathy; Real-time; Three-dimensional; Echocardiography

Introduction

Non-compaction of the ventricular myocardium is a recently recognized rare disorder of the endomyocardial morphogenesis. In the early embryogenesis, the heart consists of loose mesh of muscle fibres that normally condense gradually, which process is more complete in the left side than in the right side. The reason for the stoppage for the compaction is unknown. The aim of the present study was to demonstrate the clinical role of non-invasive real-time three-dimensional echocardiography (RT3DE) in the evaluation of a patient with biventricular non-compaction cardiomyopathy.

Case study

A 26-year-old normotensive woman was referred to the Thoraxcentre, Erasmus MC, Rotterdam, with previously recognized and treated hypertrophic/non-compaction cardiomyopathy for further cardiological examination because of proposed pregnancy. In 2003, her first pregnancy discontinued in the 11th week. She complained shortness of breath, paroxysmal tachycardia and palpitation, but have never collapsed. Her father suffered from similar symptoms before he died. During a routine transthoracic echocardiographic examination, non-compaction of the left ventricle was demonstrated with 38% ejection fraction (Figure 1). For a better evaluation, RT3DE was performed, which confirmed the previous findings but completed with right ventricular non-compaction (Figure 2A and B; Movies I, II and III).

For RT3DE, a Philips Sonos 7500 ultrasound system (Philips Co., Eindhoven, the Netherlands) equipped with software for RT3DE with a 2–4 MHz matrix-array scanner was used. For the three-dimensional analysis, a TomTec 4D Echo 5.3

Figure 1 Two-dimensional transthoracic echocardiographic evaluation of the case. Myocardial trabecularizations and deep intertrabecular recesses are seen, but the structure of the right ventricle was not obvious.
workstation (TomTec, Inc., Unterschleissheim, Germany) was used.

Discussion

Ventricular non-compaction is a rare congenital cardiomyopathy characterized by prominent myocardial trabecularizations and deep intertrabecular recesses leading to the spongy appearance of the myocardium. The disease can be characterized by systolic and diastolic heart failure, ventricular arrhythmias and systemic embolization. Clear-cut echocardiographic diagnostic criteria for isolated ventricular non-compaction have even been established. Besides the classical two-dimensional transthoracic echocardiography, the role of angiography, contrast echocardiography, computed tomography and magnetic resonance imaging were also verified to assess non-compaction cardiomyopathy. Although right ventricular involvement is not uncommon, the correct diagnosis is often difficult by conventional echocardiography. In the present case the clinical role of RT3DE is suggested in the spatial evaluation of both ventricles in suspected non-compaction cardiomyopathy. It was confirmed that RT3DE shows the presence of non-compaction in both ventricles.

Conflict of interest: none declared.

References