Isolated left ventricular noncompaction diagnosed by transthoracic three-dimensional echocardiography

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A 55-year-old man was admitted to our hospital because of chest distress, associated with activity. Two-dimensional echocardiography (2DE) demonstrated a suspected trabeculation versus false tendon of the left ventricular apex cordis but not meeting the diagnostic criteria of noncompaction of the ventricular myocardium (NVM). Three-dimensional echocardiography (3DE) revealed more prominent trabeculations and deeper intertrabecular recesses of the left ventricular apex, which were consistent with the diagnostic criteria of NVM. In contrast to 2DE, 3DE provides wide, pyramid-shaped datasets that encompass the entire left ventricle. (*Neth Heart J* 2009;17:208-10.)

Keywords: Three-dimensional echocardiography, ventricular noncompaction

Noncompaction of the ventricular myocardium (NVM) is a rare congenital cardiomyopathy, presumably resulting from an arrest in normal endomyocardial embryogenesis.¹ During the second month of embryonic life, two parallel processes normally occur: (a) gradual compaction of the ventricular myocardium with transformation of the intertrabecular spaces into capillaries, and (b) development of the coronary circulation. The process of compaction progresses from the epicardium to the endocardium and from the base of the heart toward the apex.² During this gradual process of compaction, different patterns of NVM are made by all kinds of known or unknown factors which could affect this gradual

process of compaction. The myocardium of the left ventricle is usually affected, but biventricular noncompaction is also possible. NVM is characterised by numerous excessively prominent trabeculations and deep intertrabecular recesses.² Although it has been known for more than a decade, NVM is still an unclassified cardiomyopathy according to the World Health Organisation classification of the cardiomyopathies.³

Numerous modalities have been used in the description, characterisation and diagnosis of NVM including, but not limited to, magnetic resonance imaging, two-dimensional echocardiography (2DE), contrast-enhanced 2DE, and angiography.⁴⁻⁶ 2DE is by far the most commonly used diagnostic modality. Recent studies have found that the diagnosis of this disorder is often delayed because of difficulty in elucidating the diagnostic findings.⁷ Frequently, multiple 2DE studies are required before the diagnosis is made. The recent availability of real-time transthoracic three-dimensional echocardiography (3DE) may provide an enhanced ability to diagnose and characterise the true morphology of complex myocardial disorders such as NVM.⁸ We present a case of an isolated left ventricular NVM which was confirmed by 3DE, rather than 2DE.

Case report

The patient was a 55-year-old man admitted to our hospital with chest distress, associated with activity. He had been diagnosed as having and was treated for cardiomyopathy of unknown cause and congestive heart failure during an extended period. He had no familial history of cardiovascular diseases or sudden cardiac death.

On examination, his blood pressure was 110/75 mmHg, pulse 89 beats/min and it was found that he had mild oedema of the lower limbs and rales at the bases of the left lung. Cardiovascular examination revealed a systolic murmur of grade 2/6 in the left 5th intercostal space in the apex region with an arrhythmic heart rate of 91 beats/min. Chest X-ray showed cardiomegaly and bilateral pulmonary oedema. The electrocardiogram revealed sinus rhythm, occasional

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polymorphic atrial beats, polymorphic ventricular beats and intraventricular bundle branch block.

2DE revealed hypokinesis of all the walls of the left ventricle. The left ventricular was markedly dilated and its function was severely impaired with a left ventricular ejection fraction of 36%. Pulsed Doppler echocardiography revealed restrictive patterns in the mitral inflow velocity. Furthermore, 2DE revealed a suspected trabeculation, versus false tendon, in the apex region of the cavity of the left ventricle, but did not meet the diagnostic criteria for NVM (figure 1). 3DE accurately characterised numerous large, central trabeculations in the apex region of the ventricular cavity that had been significantly underestimated by 2DE, which did met the diagnostic criteria of isolated NVM (figure 2).

Discussion

Three major clinical manifestations of noncompaction have been described: Heart failure, arrhythmias, and embolic events.^{1-3,7} Findings vary among patients, ranging from asymptomatic left ventricular dysfunction to severe, disabling congestive heart failure. Over two-thirds of the patients in the largest series with NVM had symptomatic heart failure.⁹ The prompt recognition of the disease is extremely important because of its high mortality and morbidity due to progressive heart failure, thromboembolism, and malignant arrhythmias.^{1-3,7} NVM is commonly asymptomatic; thus, its incidence in the general population may be underestimated. Despite the widespread use of echocardiography, NVM is often misdiagnosed because of the lack of knowledge about this rare disorder. Differentiating NVM from other cardiomyopathies, especially hypertrophic and dilated cardiomyopathy, may also prove problematic.¹⁰ Many cases of dilated cardiomyopathy are revised to that of NVM at post-mortem examination.¹⁰

2DE provides excellent visualisation of left ventricular cavity, but is not necessarily adequate for thorough inspection of the myocardium and endocardium of the entire left ventricle. The prominent trabeculations that characterise NVM may curve into the left ventricular cavity in a manner that challenges the planar cuts that are inherent to 2DE. These limitations of 2DE may lead to misinterpretation of prominent trabeculations as 'false tendons', and may also lead to underestimation of the severity of NVM. As a result, the 2DE diagnosis of NVM is often not made until patients have had several 2DE. Ichida et al.⁷ found that the diagnosis was missed altogether in well over half of their study population of children. Similarly, the present patient underwent an initial 2DE that did not confirm NVM. This may be due to the nonuniform nature of NVM.⁸

In contrast to 2DE, 3DE provides pyramid-shaped datasets that encompass the entire left ventricle. Specifically, the left ventricle can be sectioned in user-selected planes and an unlimited number and angles of such planes can be used. Intracavitary echo densities that are suspicious for trabeculations can be tracked in

Figure 1. Left ventricular apex showing a suspected trabeculation, versus false tendon, in the apex region of the left ventricular cavity by two-dimensional echocardiogram.

Figure 2. Left ventricular apex showing the prominent trabeculations and deep intertrabecular recesses by three-dimensional echocardiogram.

multiple directions from base to apex. The resulting images provide the ability to visualise an entire trabeculation. The deep intertrabecular recesses and the distinction between the prominent noncompact left ventricular myocardium and the thin compact layer of left ventricular myocardium can also be well demonstrated.⁸

We have further provided preliminary data demonstrating the feasibility and utility of 3DE in the diagnosis and characterisation of NVM. In addition, in this case report, 3DE allowed improved characterisation of trabeculations using uniquely available cut planes and offline analysis, which are not possible with 2DE. As 3DE takes its place as part of the routine echocardiographic evaluation, it will likely enhance the sensitivity of echocardiography in this disorder.⁸

In addition, conventional echocardiography, including 2DE and 3DE, is the diagnostic modality of

choice for NVM. In classic cases, the diagnosis of NVM is not difficult. However, when noncompaction is subtle or incomplete, disorders such as prominent normal myocardial trabeculations, hypertrophic cardiomyopathy, dilated cardiomyopathy, and left ventricular-apical thrombus should be included in the differential diagnosis. Conventional echocardiography has some diagnostic limitations in obese patients or patients with lung disease who have poor acoustic windows. Currently, echocardiography with various contrast agents is being used to enhance endocardial border delineation.¹¹ Koo et al.¹² performed transpulmonary contrast echocardiography with intravenous injection of PESDA (a sonicated mixture of perflourocarbon [8 cc], 5% albumin [4 cc], and 5% dextrose water [8 cc]). Opacification of the ventricular cavity with a contrast agent clearly defined the endocardial border, and the authors obtained better images with this method than with conventional transthoracic 2DE. Therefore, when conventional echocardiographic images are poor or diagnosis is uncertain, contrast echocardiography can be helpful. This procedure can be performed without further patient discomfort, and may save time and money. Apical swirling or nonuniform distribution of contrast may simulate endocardial trabeculae, especially in patients with low ejection fraction. ■

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