A Preliminary Study of Left Ventricular Rotational Mechanics in Children with Noncompaction Cardiomyopathy: Do They Influence Ventricular Function?

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Background: Current diagnostic criteria for noncompaction cardiomyopathy (NCC) lack specificity, and the disease lacks prognostic indicators. Reverse apical rotation (RAR) with abnormal rotation of the cardiac apex in the same clockwise direction as the base has been described in adults with NCC. The aim of this study was to test the hypothesis that RAR might differentiate between symptomatic NCC and benign hypertrabeculations and might be associated with ventricular dysfunction.

Methods: Echocardiograms from 28 children with NCC without cardiac malformations were prospectively compared with those from 29 age-matched normal control subjects. A chart review was performed to identify the patients' histories and clinical characteristics. Speckle-tracking was used to measure longitudinal strain, circumferential strain, and rotation.

Results: RAR occurred in 39% of patients with NCC. History of left ventricular (LV) dysfunction or arrhythmia was universal in, but not exclusive to, patients with RAR. Patients with RAR had lower LV longitudinal strain but similar ejection fractions compared with patients without RAR (median, -15.6% [interquartile range, -12.9% to -19.3%] vs -19% [interquartile range, -14.5% to -21.9%], $P < .01$; 53% [interquartile range, 43% to 68%] vs 61% [interquartile range, 58% to 67%], $P = .08$). Only a pattern of contraction with RAR, early arrest of twisting by mid-systole, and premature untwisting was associated with lower ejection fraction (46%; interquartile range, 43% to 52%; $P = .006$).

Conclusions: RAR is not a sensitive but is a specific indicator of complications in children with NCC. Therefore, RAR may have prognostic rather than diagnostic value. Premature untwisting of the left ventricle during ejection may be an even more worrisome indicator of LV dysfunction. (J Am Soc Echocardiogr 2018;■:■-■.)

Keywords: Cardiomyopathy, Mechanics, Noncompaction, Pediatrics, Torsion

Noncompaction cardiomyopathy (NCC) is a form of cardiomyopathy characterized by an expanded layer of trabeculations and deep intertrabecular recesses, in the inner ventricular muscle wall of the left ventricle.[1] This disease unfortunately lacks specific diagnostic criteria, leading to many normal individuals being diagnosed with NCC.[2] Therefore finding disease-specific characteristics that can differentiate patients with benign hypertrabeculations from those with true symptomatic NCC is crucial. The potential role of torsion as a diagnostic or prognostic parameter in this disease has been studied infrequently in children, although recent reports in adults with NCC describe replacement of the normal wringing motion of the heart by a roller pump–like motion.[3,4]

The normal wringing or twisting motion of the left ventricle during ejection consists of clockwise rotation of the base and counterclockwise rotation of the apex (when viewed from the apex). The roller pump–like motion found in some patients with NCC is characterized by rotation of the apex and base in the same clockwise direction. Hence, the wringing motion of the left ventricle is lost, and the left ventricle behaves like a roller pump. This motion has been labeled rigid body rotation or reverse apical rotation (RAR).[5,6] How this abnormal rotation pattern affects ventricular function and whether it alters prognosis is not known.

Therefore, in this pilot study, we sought to describe the rotational mechanics of the left ventricle in children with isolated NCC during systole and diastole. We sought to investigate the association of RAR with history of NCC complications and with ventricular dysfunction. We hypothesized that compared to children with NCC and normal rotation, children with NCC and RAR would be more symptomatic, have

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Conflicts of Interest: None.

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0894-7317/$36.00

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https://doi.org/10.1016/j.echo.2018.02.015

Abbreviations

EF = Ejection fraction

LV = Left ventricular

MRI = Magnetic resonance imaging

NCC = Noncompaction cardiomyopathy

RAR = Reverse apical rotation

more NCC-related complications, and manifest evidence of impaired left ventricular (LV) ejection and filling, secondary to loss of LV twist and untwist.

METHODS

Subject Enrollment

We prospectively enrolled children being evaluated in the echocardiography laboratory at the Children's Hospital of Philadelphia during diagnosis or routine follow-up of NCC if they were <18 years of age and had no associated congenital heart defects. Because of the low reproducibility[7] and specificity[2] of the current echocardiographic diagnostic criteria for NCC, we only included patients who met all three echocardiographic criteria proposed by Jenni *et al.*,[8] Stollberger and Finsterer,[9] and Chin *et al.*[10] (For measurement techniques used, see Figure 1 and its legend.) Available cardiac magnetic resonance imaging (MRI) data were also evaluated for additional confirmation of the diagnosis of NCC.

A control group consisting of children with noncardiac chest pain, vasodepressor syncope, or functional heart murmur was also enrolled. Control subjects were approached for inclusion in the study if they had no evidence of structural heart disease, ejection fractions (EFs) > 55%, and no evidence of diastolic dysfunction on mitral inflow spectral Doppler or tissue velocities of the lateral mitral valve annulus. Subjects were excluded from the study if they had evidence of elevated pulmonary arterial pressure, on the basis of ventricular septal position or a tricuspid regurgitant jet >3 m/sec, elevated systemic blood pressure, or any arrhythmia at the time of the study.

Echocardiography

Complete transthoracic echocardiography was performed on all subjects to assess their eligibility for inclusion in the study on the basis of the criteria discussed earlier. To assess LV torsion, we used our published protocol for image acquisition.[11] Parasternal short-axis cine clips of the base of the heart at the level of the mitral valve leaflets and the apex of the heart were obtained. The cardiac apex was defined as the furthest apical extent of the LV cavity, distal to the base of the papillary muscles and just proximal to the level of cavity obliteration. A target frame rate of >60 Hz was achieved in all subjects. The timing of aortic valve closure and mitral valve opening was determined from

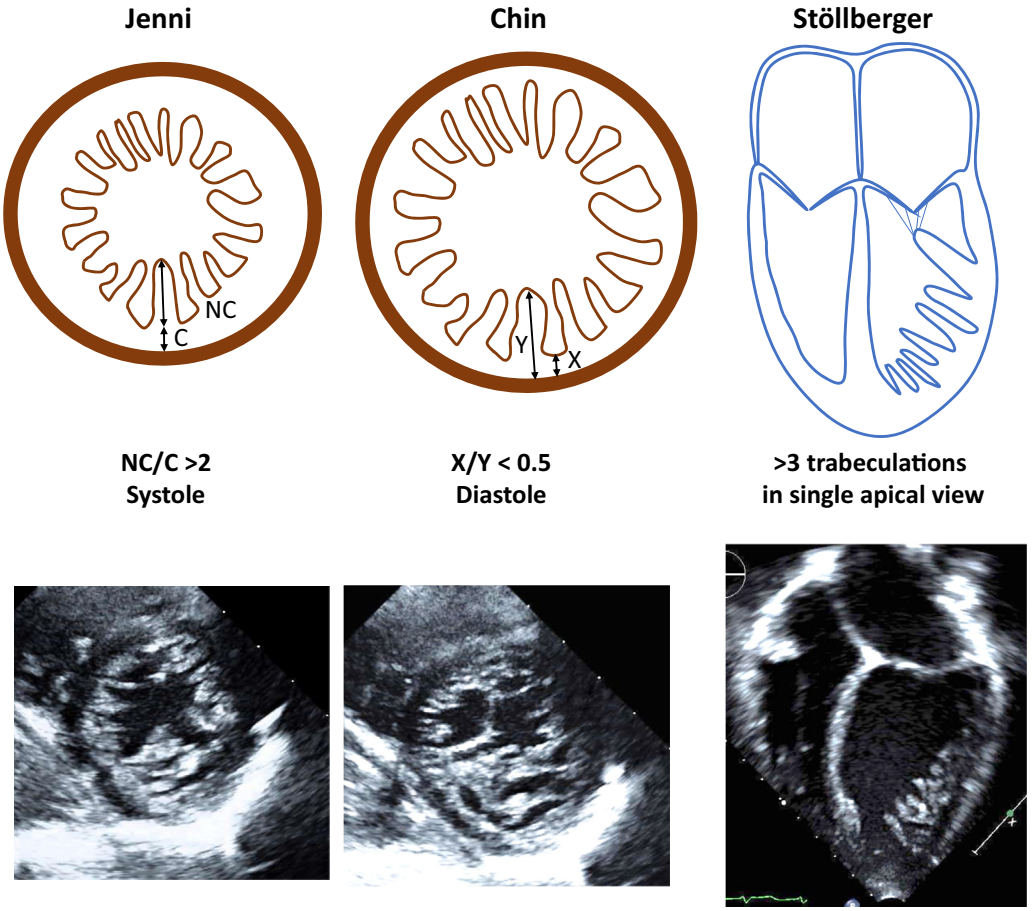

Figure 1 Schematic and two-dimensional echocardiographic images depicting the three methods used in the diagnosis of NCC (Jenni, Chin, and Stöllberger methods) from apical short-axis and four-chamber views. In the Jenni and Chin methods, the noncompacted segments are measured differently. In the Jenni method, the noncompacted segment is measured up to the bottom of the recesses (NC), whereas in the Chin method, it is measured up to the epicardium (Y). C and X are compacted segments for respective methods. The criteria for the diagnosis of NCC by the three methods are as follows: Jenni, NC/C ratio > 2; Chin, X/Y ratio < 0.5; and Stöllberger, more than three trabeculations in one imaging plane below the level the papillary muscles.

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