

LEFT VENTRICULAR NONCOMPACTION AND PRE-EXCITATION: AN UNUSUAL FINDING IN ADULTS

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ABSTRACT

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Left ventricular noncompaction (LVNC) is a rare form of cardiomyopathy characterized by prominent left ventricular (LV) trabeculae, deep intertrabecular recesses, and the thin compacted layer. The disease is potentially associated with sudden cardiac death due to LV dysfunction and ventricular arrhythmias. The presence of accessory pathway and Wolff-Parkinson-White syndrome is particularly rare in adults. Here we describe the rare association of LVNC and ventricular pre-excitation in an 18-year-old female with neonatal hypoxic brain injury.

Keywords: *Wolff-Parkinson-White syndrome; pre-excitation syndromes; isolated noncompaction of the ventricular myocardium*

Left ventricular noncompaction (LVNC) is a rare form of cardiomyopathy characterized by prominent left ventricular (LV) trabeculae, deep intertrabecular recesses, and the thin compacted (C) layer¹. The diagnosis is usually performed by echocardiography or magnetic resonance imaging (MRI). The disease is potentially associated with sudden cardiac death (SCD) due to LV dysfunction and ventricular arrhythmias. The presence of accessory pathway and Wolff-Parkinson-White (WPW) syndrome is particularly rare in adults^{2,3}. Here we describe the rare association of LVNC and ventricular pre-excitation in an 18-year-old female with neonatal hypoxic brain injury. There were no arrhythmias or WPW syndrome. Clinical follow-up was performed without complications.

CASE PRESENTATION

An 18-year-old female with severe neonatal hypoxic brain injury and chronic cognitive and motor disabilities was referred for outpatient cardiac consultation for preoperative evaluation. She presented severe scoliosis and motor disabilities requiring orthopedic surgery for spine stabilization. Her cardiac medical history was uneventful. Despite limited functional status, her parents denied any warning symptoms like chest pain, shortness of breath, palpitations, or syncope. Physical examination revealed good peripheral pulses, no peripheral edema, blood pressure 102/60 mmHg, and heart rate of 96 bpm. Cardiac auscultation showed regular rhythm with no murmurs or gallops. There were no signs of cardiomegaly.

A 12-lead electrocardiography (ECG) revealed ventricular pre-excitation with right-bundle branch block morphology compatible with a left wall accessory-pathway (Figure 1). A 24-hour Holter did not record arrhythmias. A transthoracic echocardiogram was also performed. Despite limited acoustic window, the left ventricular ejection fraction (LVEF) was normal. However, there were signs of prominent trabeculation and deep recesses that communicate with the left ventricular cavity (Figure 2). These signs were compatible with isolated LVNC.

An electrophysiological study (EPS) was considered, but deferred by patient's family. The patient underwent surgical procedure under cardiac monitoring. No arrhythmias or cardiovascular complications were reported. After 1 year of clinical follow-up, LVEF remained unaltered and the patient



Figure 1: The 12-lead electrocardiogram showing ventricular pre-excitation (short PR interval and delta wave). There is a right-bundle branch block pattern, negative delta wave in aVL and isoelectric delta waves in DI and positive delta waves in DII, DIII, aVF and V6.

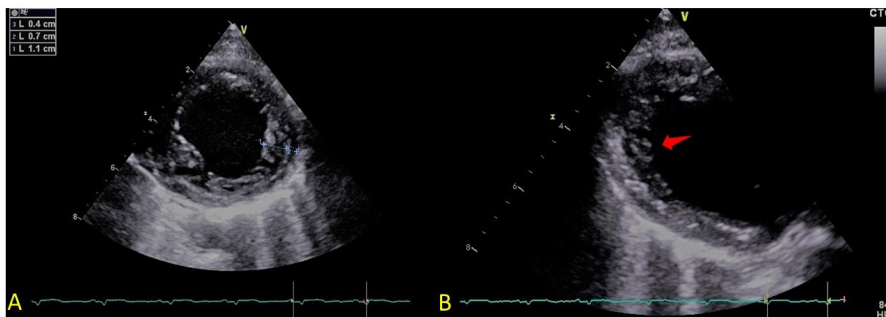


Figure 2: Transthoracic echocardiogram obtained in parasternal short axis view showing signs of left ventricular noncompaction, characterized by a relation of noncompacted endocardial to compacted epicardial layer above 2 during end systole (A). The hipertrabeculation is also observed in left ventricular apex (B, red arrow).

did not develop symptoms or clinical arrhythmias. Family screening (mother and sister) for cardiac abnormalities was negative.

DISCUSSION

LVNC or “spongy myocardium” has been recognized as a distinct form of cardiomyopathy. It is thought to result from an arrest of the compaction process during the second month of embryological development, resulting in abnormal left ventricular myocardial architecture⁴. LVNC occurs in infants (0.81 cases/100,000 per year), children (0.12 cases/100,000 per year) and adults (prevalence 0.014%)¹. The disease is defined by three markers: prominent LV trabeculae, deep intertrabecular recesses, and a thin C layer¹. The myocardium will have an increased noncompacted (NC) endomyocardial layer depth compared to the C epicardial layer.

There is no consensus on diagnostic criteria. Two-dimensional echocardiography is the most common and useful tool for LVNC diagnosis. Jenni et al. proposed criteria based on an end systolic ratio of NC to C layers above 2⁵. More recently, cardiac magnetic resonance (CMR) has been considered a useful resource that may help to accurately describe and diagnose LVNC and distinguish true disease from the prominent hypertrabeculation that can be seen in normal hearts and individuals. A NC/C ratio > 2.3 measured at the end of diastole is considered the cutoff for proper diagnosis¹. Three-dimensional echocardiography may play a role in the future, although its role currently remains anecdotal⁶. Despite advances in imaging technologies, controversies persist and a “gold standard” for the diagnosis of LVNC continues to be lacking as no imaging or pathology signature has yet been agreed⁶.

The association of WPW with LVNC is well recognized. Previous reports include WPW associated with isolated LVNC^{3,7,8}; WPW associated with LVNC and Fabry disease⁹ or hypertrophic cardiomyopathy¹⁰ was also reported. During embryogenesis there is direct continuity of the atrial and ventricular myocardium, which is ultimately disrupted as the annulus fibrosus develops. Defects in the annulus

fibrosus are thought to account for the formation of accessory pathways. In ventricular noncompaction, the hypothesized arrest in development characterized by persistence of trabeculations might account for the persistence of myocardial channels between atrium and ventricle, allowing for the development of accessory pathways¹.

Electrocardiographic changes in LVNC most commonly reported in the literature are left bundle branch block, right bundle branch block, left anterior fascicular block, repolarization abnormalities, atrial fibrillation, and pre-excitation¹¹. Stöllberger and Finsterer² have analyzed the arrhythmias associated with LVNC and found a different prevalence between children and adults. In adults, the most frequent arrhythmias are ventricular tachycardia, atrial fibrillation, QT prolongation, and atrioventricular block. Ventricular pre-excitation was more frequently reported in children than in adults. In different series, the occurrence of ventricular pre-excitation is estimated in 15% in pediatric series but is rarely observed in adults. Most of ventricular pre-excitation cases associated with LVNC had signs and symptoms that corroborate a clinically relevant accessory pathway (WPW syndrome).

In the present case, the diagnosis of LVNC was confirmed by transthoracic echocardiography. Despite limited acoustic window, the NC/C ratio was measured at 2.75 in the inferolateral segment at mid short axis view (Figure 2A). Additional images also showed areas of hypertrabeculation at the apex (Figure 2B). Despite recent concerns about overdiagnosis, LVNC is the main diagnosis¹². The ECG also confirmed the presence of ventricular pre-excitation. The main algorithms for accessory pathway location supports a location around the mitral annulus. Despite indication for EPS for precise pathway location and risk stratification, patient’s clinical condition was determinant for a conservative approach.

The present case highlights an unusual finding of LVNC and ventricular pre-excitation in an adult patient. Despite the theoretical risks of arrhythmias and even sudden cardiac death, there is no clear warning signs in the present case. Clinical follow-up is important in such cases.

REFERENCES

1. Arbustini E, Weidemann F, Hall JL. Left ventricular noncompaction: a distinct cardiomyopathy or a trait shared by different cardiac diseases? *J Am Coll Cardiol*. 2014;64(17):1840-50. PMID:25443708. <http://dx.doi.org/10.1016/j.jacc.2014.08.030>.
2. Stöllberger C, Finsterer J. Arrhythmias and left ventricular hypertrabeculation /noncompaction. *Curr Pharm Des*. 2010;16(26):2880-94. PMID:20632950. <http://dx.doi.org/10.2174/138161210793176518>.
3. Fichet J, Legras A, Bernard A, Babuty D. Aborted sudden cardiac death revealing isolated noncompaction of the left ventricle in a patient with wolff-Parkinson-white syndrome. *Pacing Clin Electrophysiol*.

- 2007;30(3):444-7. PMID:17367369. <http://dx.doi.org/10.1111/j.1540-8159.2007.00690.x>.
4. Chin TK, Perloff JK, Williams RG, Jue K, Mohrmann R. Isolated noncompaction of left ventricular myocardium: a study of eight cases. *Circulation*. 1990;82(2):507-13. PMID:2372897. <http://dx.doi.org/10.1161/01.CIR.82.2.507>.
 5. Gopalumurugan AB, Kapetanakis S, Monaghan MJ, Murgatroyd F. Left ventricular non-compaction diagnosed by real time three dimensional echocardiography. *Heart*. 2005;91(10):1274. PMID:16162615. <http://dx.doi.org/10.1136/hrt.2005.063602>.
 6. Dawson DK, McLernon DJ, Raj VJ, Maceira AM, Prasad S, Frenneaux MP, et al. Cardiovascular magnetic resonance determinants of left ventricular noncompaction. *Am J Cardiol*. 2014;114(3):456-62. PMID:24934759. <http://dx.doi.org/10.1016/j.amjcard.2014.05.017>.
 7. Nihei K, Shinomiya N, Kabayama H, Ikeda C, Hosono T, Aoki T, et al. Wolff-Parkinson-White (WPW) syndrome in isolated noncompaction of the ventricular myocardium (INVM). *Circ J*. 2004;68(1):82-4. PMID:14695471. <http://dx.doi.org/10.1253/circj.68.82>.
 8. Brembilla-Perrot B, Codreanu A, Marie PY, Beurrier D, Husson JL, Hutin O, et al. Association of Wolff-Parkinson-White syndrome with isolated non-compaction of the left ventricle: a case report. *Arch Mal Coeur Vaiss*. 2006;99(6):626-8. PMID:16878725.
 9. Alper AT, Kaya A, Tekkesin AI, Öz A. Wolff-Parkinson-White and left ventricular noncompaction in a Fabry patient: a case report. *Turk Kardiyol Dern Ars*. 2016;44(3):248-50. PMID:27138316.
 10. Alday L, Moreyra E, Bruno E, Rossi N, Maisuls H. Left ventricular noncompaction associated with hypertrophic cardiomyopathy and Wolff-Parkinson-White syndrome. *Health*. 2010;2(3):200-3. <http://dx.doi.org/10.4236/health.2010.23029>.
 11. Jenni R, Oechslin E, Schneider J, Attenhofer Jost C, Kaufmann PA. Echocardiographic and pathoanatomical characteristics of isolated left ventricular non-compaction: a step towards classification as a distinct cardiomyopathy. *Heart*. 2001;86(6):666-71. PMID:11711464. <http://dx.doi.org/10.1136/heart.86.6.666>.
 12. Niemann M, Störk S, Weidemann F. Left ventricular noncompaction cardiomyopathy: an overdiagnosed disease. *Circulation*. 2012;126(16):e240-3. PMID:23071178. <http://dx.doi.org/10.1161/CIRCULATIONAHA.112.095059>.

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