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TEMI CALDI IN CARDIOLOGIA DELLO SPORT

MIOCARDIO NON COMPATTO: QUANDO POSSIAMO FARE DIAGNOSI

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Un pò di storia…

Year	1st Author [Ref #]	Description
1926	Grant [1]	He was the first to describe a spongy myocardium as a congenital cardiac malformation in children.
1932	Bellet & Gouley [2]	Observed abnormally "spongy" myocardial walls linked to aortic atresia and coronary-ventricular fistula in an autopsy of a newborn with congenital heart disease
1975	Dusek et al. [3]	Described a persistent of spongy myocardium with embryonic blood supply in children.
1984	Engberding et al. [4]	Provided the first clinical description of spongy myocardium based on echocardiographic findings in an adult.
1990	Chin et al. [5]	Proposed the term LVNC following diagnosis in 8 cases in the absence of congenital cardiac abnormalities.

LVNC: Left Ventricular Non-Compaction

Albakri A (2018) Left ventricular non-compaction: A review of literature on clinical status and meta-analysis of diagnostic and clinical management methods

2008 position statement for ESC Working Group on Myocardial and Pericardial Diseases defines LVNC as a

(2006) The American Heart Association (AHA) defines LVNC as **"*a congenital cardiomyopathy characterized by distinctive (spongy) morphological appearance of the LV myocardium. Non-compaction involves predominantly the distal (apical) portion of the LV chamber with deep inter-trabecular recesses (sinusoids) in communication with the ventricular cavity, resulting from an arrest in the normal embryogenesis*" (2)**

(1) Elliott P et al. (2008). Classification of the cardiomyopathies: a position statement from the European Society of Cardiology Working Group on

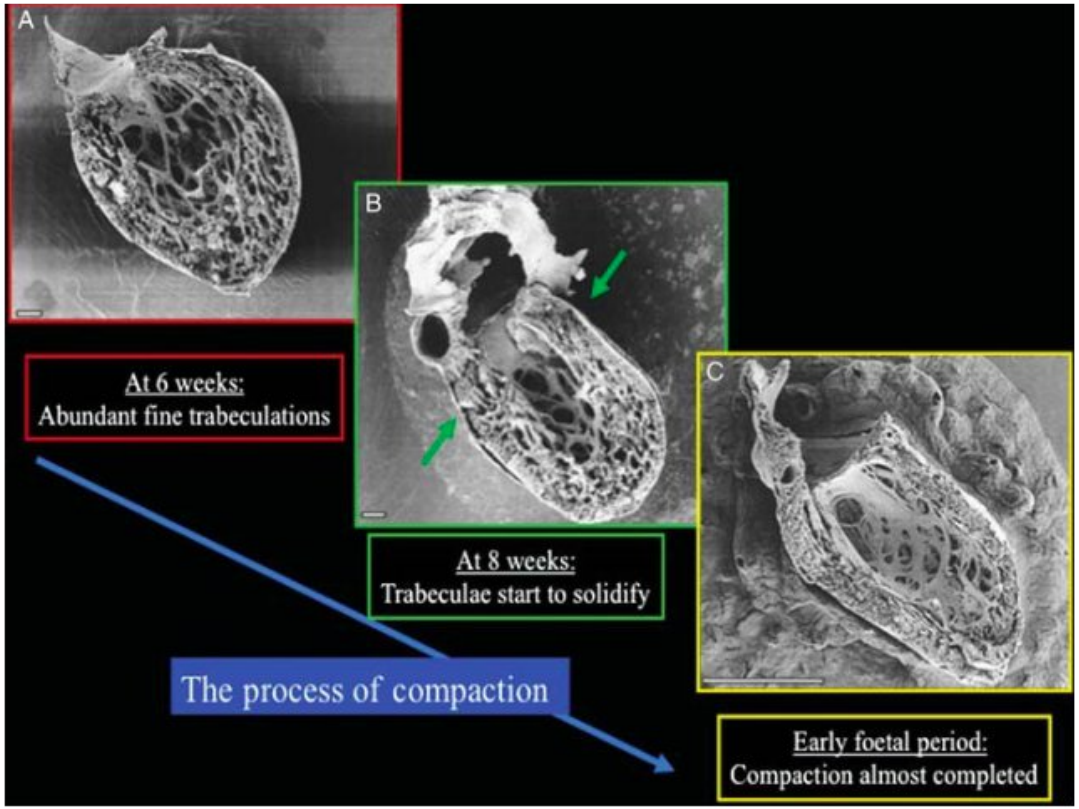
(2) Maron BJ et al.(2006). Contemporary definitions and classification of the cardiomyopathies: an American Heart Association Scientific Statemer

The National Australian Childhood Cardiomyopathy study documented **9.2% of the primary cardiomyopathies in children younger**

In two observational studies patients with depressed ejection fraction (< 45%) and symptoms of heart failure referred for echocardiograp

With MR imaging the numbers were significantly higher, with 9.6% in cardiac patients

Gerecke B.J. Noncompaction Cardiomyopathy—History and Current Knowledge for Clinical Practice. J. Clin. Med. 2021, 10, 2457.
Albakri A (2018) Left ventricular non-compaction: A review of literature on clinical status and meta-analysis of diagnostic and clinical management methods

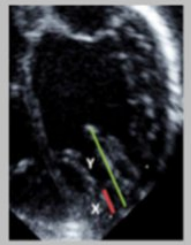
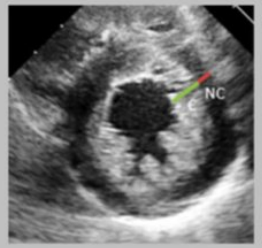
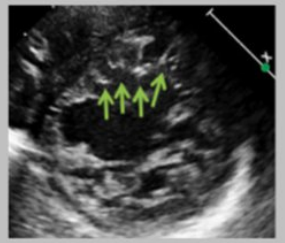


Albakri A (2018) Left ventricular non-compaction: A review of literature on clinical status and meta-analysis of diagnostic and clinical m

Diagnosi

At present, there is no consensus on practice guidelines or expert recommendations on diagnostic criteria for LVNC. The current criteria is based on findings from small single-center retrospective studies.

Albakri A (2018) Left ventricular non-compaction: A review of literature on clinical status and meta-analysis of diagnostic and clinical n

	Chin *et al.* [1]	Jenni *et al.* [3]	Stöllberger *et al.* [4,5]
Measurement Example			
Diagnostic Criteria	• Decreasing X/Y ratio* from base to apex • No specific cut-off reported	• Two-layer myocardium with NC/C* ratio >2 • Non-compaction predominantly located in mid-lateral, apical, and mid-inferior regions • Perfusion of intertrabecular spaces demonstrated by color Doppler • Absence of coexisting cardiac abnormalities	• >3 prominent trabeculations • Trabeculations move synchronously with myocardium • Trabeculations part of noncompacted layer of the two-layered myocardium • Perfusion of intertrabecular spaces demonstrated by color Doppler
Cardiac Phase	• End-diastole	• Systole	• Trabeculations: End-diastole • Two-layered myocardium: End-systole
Echocardiographic View/Segment	• Parasternal long-axis at level of mitral valve and papillary muscle; • Subcostal long-axis or apical four-chamber view at level of apex	• Parasternal short-axis	• Trabeculations: Parasternal short axis, apical level • Two-layered myocardium: "Atypical apical 2-chamber view"
LVNC Cases (n)	• 8	• 7	• 62[2], 115[3]
Control group (n)	• 8 (normal echocardiograms)	• 10 with DCM* • 9 with LVH*	• 0
Pathology data (n)	• 3 out of 8 LVNC cases	• 7 out of 7 LVNC cases • 19 out of 19 Control cases	• 0
Age range	• 11 months – 22 years	• 22 – 61 years	• 18 – 87 years

Joong A. et al, Comparison of Echocardiographic Diagnostic Criteria of Left Ventricular Noncompaction in a Pediatric Population, Pediatr Car

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Clinical Insights From Cardiac Imaging

Left Ventricular Non-Compaction

Insights From Cardiovascular Magnetic Resonance Imaging

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We analyzed magnetic resonance cine images, using the 17-segment model in 45 healthy volunteers, 25 athletes, 39 patients with hypertrophic cardiomyopathy and 14 with dilated cardiomyopathy, 17 with hypertensive heart disease, and 30 with aortic stenosis, as well as images from 7 patients previously diagnosed with LVNC whose diagnoses were supported by additional features.

Table 1. Characteristics of Patients Diagnosed With LVNC

LVNC Patients	Age (yrs)	Gender	Symptoms	FH	ECG Changes	Regional Wall Motion Abnormality	LVEF (%)	Neuromuscular Findings	NC/C Ratio	No. of Segments With NC
1	14	M	Heart failure as baby	+	+	—	48	—	2.4	12
2	15	F	—	+	+	—	64	—	1.1	9
3	38	M	—	+	—	—	61	—	2.3	8
4	41	M	—	+	—	+	53	—	3.3	12
5	46	M	Systemic embolus	—		+	17	—	6.1	15
6	26	F	—	—	—	+	68	—	2.9	9
7	25	M	Syncope	—	+	+	59	+	2.7	8

FH = family history; ECG = electrocardiographic; LVNC = left ventricular non-compaction; NC = non-compaction; NC/C = non-compacted to compacted myocardium.

Areas of non-compaction were common and occurred more frequently in all groups studied in apical and lateral, rather than in basal or septal, segments. A NC/C ratio of >2.3 in diastole distinguished pathological non-compaction, with values for sensitivity, specificity, and positive and negative predictions of 86%, 99%, 75%, and 99%, respectively.

	Jacquier
Year	2010
Total Patients; Patients with LVNC	64; 16
Selection Criteria	Patients fulling Jenni et al. criteria for LVNC; patients with hypertrophic or dilated cardiomyopathy and Controls
Age Range	25 to 74 years
Description of Criteria	1. Total left ventricular (LV) trabeculated mass ≥20% of the global LV mass
View	Short-axis stack
Phase	End-diastole
Outcomes	No

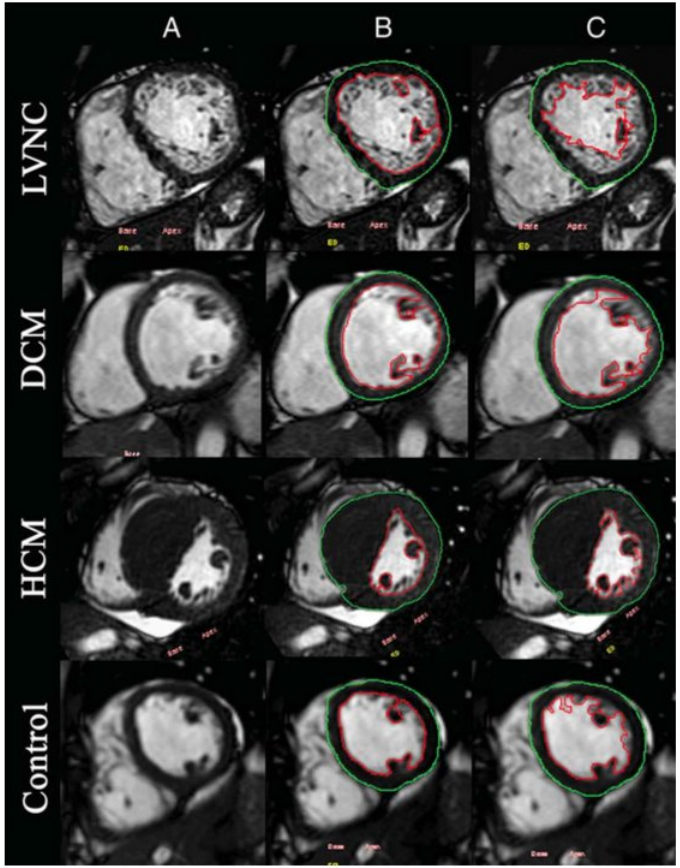

Figure 1 Illustration of the described method for measuring the global and trabeculated left ventricular masses in patients with left ventricular non-compaction, dilated cardiomyopathy, hypertrophic cardiomyopathy, and controls. Column A shows the short-axis end-diastolic cine images used for measurement without contouring. Column B shows the inclusion of papillary muscles and the exclusion of left ventricular trabeculation for the measurements of the compacted left ventricular mass. Column C shows inclusion of papillary muscles and trabeculation for the measurements of global left ventricular mass.

lf Jenni
et *al.*'s criteria alone are considered as the gold standard for LVNC diagnosis, a value of trabeculated LV mass above 20% of the total LV mass is predictive of LVNC with a sensitivity of 91.6% (CI, 64.6–98.5%) and a specificity of 86.5% (CI, 74.7–93.3%; $\kappa = 0.65$).

The technique we describe was c
gold standard CMR technique defin
value of trabeculated LV mass above 2
and specificity for the diagnosis of LVN
criteria as the standard of reference
92.4%) and 72.2% (CI, 49.1–87.5%),

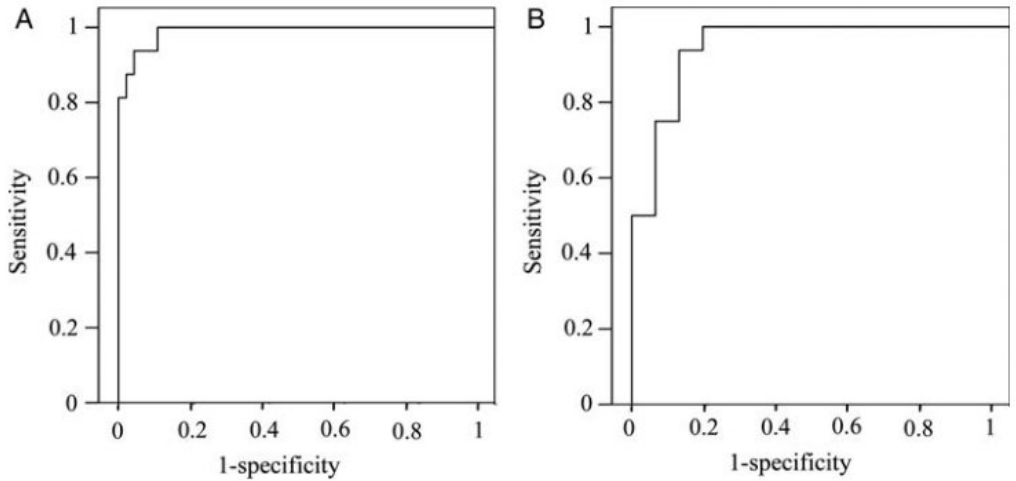

Figure 4 Receiver operating characteristic curve, using patient classification according to our echocardiographic diagnosis criteria for left ventricular non-compaction as a reference, describing the performance of the percentage of trabeculated myocardial mass (*A*) and Petersen *et al.*'s criteria (*B*) for left ventricular non-compaction diagnosis.

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DOI 10.1007/s

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Matthias Gr
Janine Hoffi
Sabine Klaa

	Grothoff
Year	2012
Total Patients; Patients with LVNC	57; 12
Selection Criteria	Patients with echo LVNC (Jenni criteria) plus one of the following: LVNC in first-degree relatives, neuromuscular disorder or complications such as systematic embolization or regional wall motion abnormalities or ventricular abnormalities; patients with hypertrophic or dilated cardiomyopathy and matched controls
Age Range	11 to 71 years
Description of Criteria	1. Percentage of non-compacted mass > 25% 2. Total indexed myocardial mass > 15 g/m^2 3. A non-compacted to compacted myocardial ratio ≥ 3:1 in segments 1–3 or 7–16 excluding the apex 4. A non-compacted to compacted myocardial ration ≥ 2:1 in segments 4–6
View	Short-axis stack
Phase	End-diastole
Outcomes	No

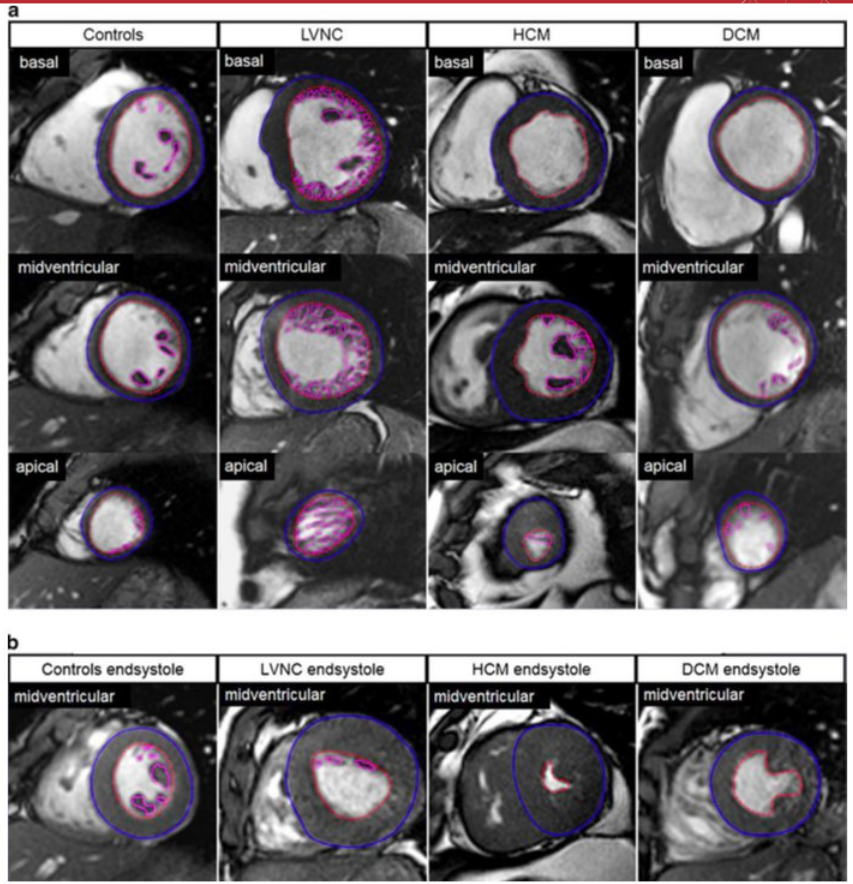

Fig. 1 **a** Assessment of non-compacted (*pink contours*) and compacted (*red and blue contours*) left ventricular myocardium using CAAS MRV software in controls, patients with left ventricular non-compaction cardiomyopathy (LVNC), dilated cardiomyopathy (DCM) and hypertrophic cardiomyopathy (HCM). Basal, midventricular and apical cine steady-state free precession images in short-axis orientation during end-diastole (echo time 1.8 ms, repetition time 3.6 ms, flip angle 50°). All masses were assessed in end-diastole. **b** Midventricular steady-state free precession (SSFP) images in short-axis orientation in end-systole. In this phase trabeculation is compressed which makes it almost impossible to differentiate non-compacted from compacted myocardium. Therefore, the end-systolic phase is not suitable either for measuring the ratio of non-compacted/compacted myocardium or for quantifying the amount of non-compacted myocardium. Furthermore, this may also lead to an overestimation of LV ejection fractions (EF) in LVNC and HCM patients

- No trabeculation
- Non-compacted/compacted myocardium ratio of <2:1
- Non-compacted/compacted myocardium ratio of ≥2:1 to <3:1
- Non-compacted/compacted myocardium ratio of ≥3:1

Table 4 Impact of criteria

Criteria met	Sensitivity	95 % CI	Specificity	95 % CI	PPV	NPV
Single criteria						
1. Percentage LV- $MM_{non\text{-}compacted}$>25 %	91 (10/11)	62.3–98.4	100 (45/45)	92.1–100	100 (10/10)	98 (45/46)
2. Total LV-$MMI_{non\text{-}compacted}$>15 g/m^2	91 (10/11)	62.3–98.4	91 (41/45)	79.3–96.5	71 (10/14)	98 (41/42)
3. Cut-off≥3:1 (segment 17 excluded)	100 (12/12)	75.8–100	93 (42/45)	82.1–97.7	80 (12/15)	100 (42/42)
4. Trabeculation in segments 4–6≥2:1	67 (8/12)	39.1–86.2	91 (41/45)	79.3–96.5	67 (8/12)	91 (41/45)
Cut-off≥3:1 (segment 17 included)	100 (12/12)	75.8–100	73 (33/45)	59.0–84.1	50 (12/24)	100 (33/33)
Cut-off>2.3:1 (segment 17 excluded)	100 (12/12)	75.8–100	80 (36/45)	66.2–89.1	57 (12/21)	100 (36/36)
Cut-off>2.3:1 (segment 17 included)	100 (12/12)	75.8–100	58 (26/45)	43.3–71.0	39 (12/31)	100 (19/19)
Combined criteria (1–4)						
Two of four criteria (1–4)	100 (12/12)	75.8–100	95 (43/45)	85.2–98.8	86 (12/14)	100 (43/43)
Three of four criteria (1–4)	92 (11/12)	64.6–98.5	100 (45/45)	92.1–100	100 (9/9)	96 (45/48)
Four of four criteria (1–4)	75 (9/12)	46.8–91.1	100 (45/45)	92.1–100	100 (7/7)	94 (45/49)

LV myocardial mass index (LV- MMI)

LV-MMI compacted

LV- MMI non-compacted

Percentage LV-MM non-compacted

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Evaluation of isolated left ventricular noncompaction using cardiac magnetic resonance tissue tracking in global, regional and layer-specific strains

Scientific Reports | (2021) 11:7183 | https://doi.org/10.1038/s41598-021-86695-0

52 patients without ILVNC (control group)

63 patients with ILVNC

29 patients LGE+

34 patients LGE-

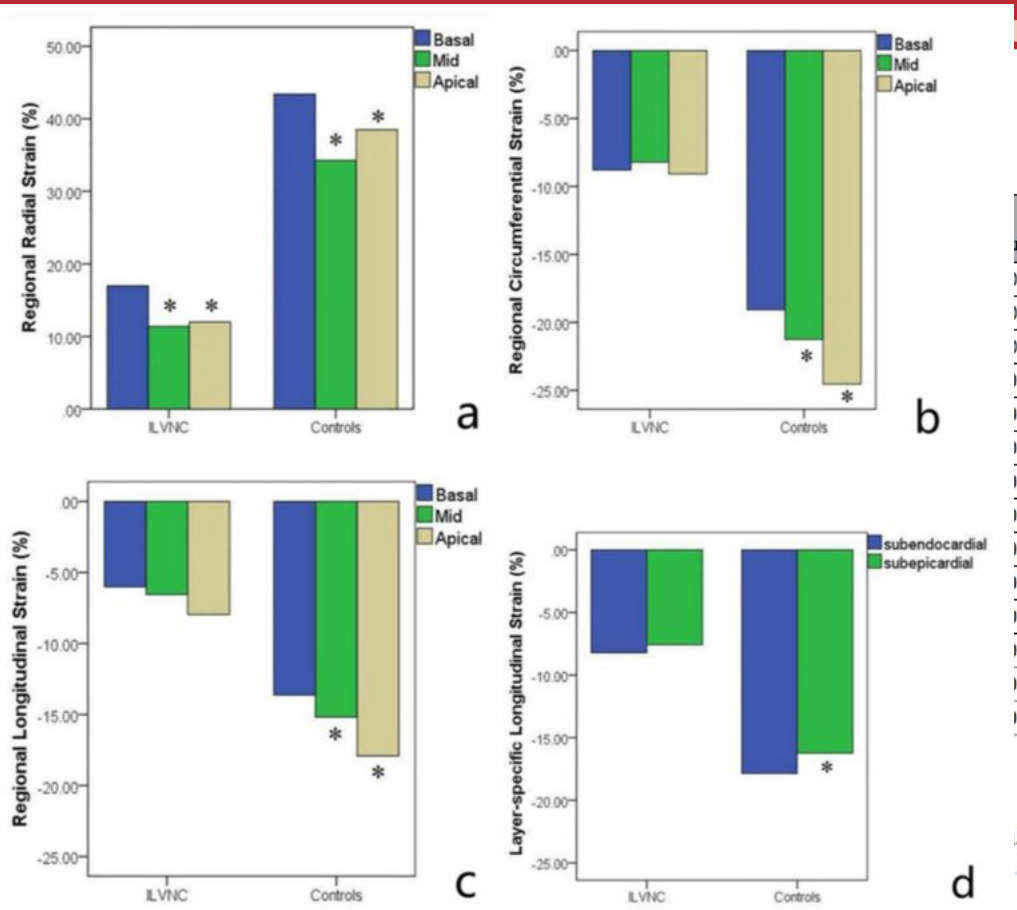

Figure 4. Comparison of basal to apical radial, circumferential, and longitudinal strain values in the isolated left ventricular noncompaction (ILVNC) group and the control group (**a**–**c**), *means compared with the basal strain, $P<0.05$. Comparison of subepicardial to subendocardial longitudinal strain values in the ILVNC group and the control group (**d**), *means compared with the subendocardial longitudinal strain, $P<0.05$.

NCCM: strictly congenital or also postnatal?

Some echocardiographic studies reported cases where LVNC was absent in initial studies, but became evident in subsequent ex

Gati *et al.* observed in highly trained athletes increased LV trabeculation might represent cardiac remodeling, where trabeculati

In a related study recruiting pregnant women, Gati *et al*. observed de-novo LV trabeculations in > 25% of the patients, suggest

(1) Stöllberger C, Finsterer J (2004). Left ventricular hypertrabeculation/noncompaction. J Am Soc Echocardiogr., 17: 91-100.

(2) Gati S, et al. (2013). Increased left ventricular trabeculation in highly trained athletes: do we need more stringent criteria for the diagnosis o

(3) Gati S, et al. (2014). Adult left ventricular noncompaction: Reappraisal of current diagnostic imaging modalities. JACC Cardiovasc Imaging,

Left Ventric

Anatomical Phen

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Kerrie Fitzgerald, MBChB,[a] Stephen J. Gandy, PhD,[b] Matthew Lambert, MBChB,[a] Jill J.F. Belch, MD,[a]
Ian Cavin, PhD,[b] Roberta Littleford, PhD,[a] Jennifer A. Macfarlane, PhD,[b] Shona Z. Matthew, PhD,[a]
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Richard D. White, MBChB,[d] J. Graeme Houston, MD[a]

ABSTRACT

BACKGROUND There is considerable overlap between left ventricul
cardiomyopathies. LVNC has been reported in up to 40% of the genera
a distinct pathological entity, a remodeling epiphenomenon, or merel

OBJECTIVES The authors determined the prevalence and predictors
magnetic resonance imaging diagnostic criteria.

METHODS Volunteers >40 years of age (N = 1,651) with no history
CVD < 20%, and a B-type natriuretic peptide level greater than their
resonance imaging scan as part of the TASCFORCE (Tayside Screening
measured on the horizontal and vertical long axis cine sequences. All
underwent short axis systolic and diastolic LVNC ratio measurements,
pacted myocardial mass ratios. Those who met all 4 criteria were considered to have LVNC.

RESULTS Of 1,480 participants analyzed, 219 (14.8%) met ≥1 diagnostic criterion for LVNC, 117 (7.9%) met 2 criteria, 63 (4.3%) met 3 criteria, and 19 (1.3%) met all 4 diagnostic criteria. There was no difference in demographic or allometric measures between those with and without LVNC. Long axis noncompaction ratios were the least specific, with current diagnostic criteria positive in 219 (14.8%), whereas the noncompacted to compacted myocardial mass ratio was the most specific, only being met in 61 (4.4%).

CONCLUSIONS A significant proportion of an asymptomatic population free from CVD satisfy all currently used cardiac magnetic resonance imaging diagnostic criteria for LVNC, suggesting that those criteria have poor specificity for LVNC, or that LVNC is an anatomical phenotype rather than a distinct cardiomyopathy. (J Am Coll Cardiol 2016;68:2157–65) © 2016 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation.

RESULTS Of 1,480 participants analyzed, 219 (14.8%) met ≥1 diagnostic criterion for LVNC, 117 (7.9%) met 2 criteria, 63 (4.3%) met 3 criteria, and 19 (1.3%) met all 4 diagnostic criteria. There was no difference in demographic or allometric measures between those with and without LVNC.

CONCLUSIONS

A significant proportion of an asymptomatic population free from CVD satisfy all currently used CMR diagnostic criteria for LVNC, suggesting that either these all have poor specificity for LVNC, or that LVNC is an anatomical phenotype rather than a distinct cardiomyopathy.

Conclusioni

Meta-Analysis > Eur Heart J. 2020 Apr 7;41(14):1428-1436. doi: 10.1093/eurheartj/ehz317.

A systematic review and meta-analysis of the prevalence of left ventricular non-compaction in adults

Samantha B Ross [1 2], Katherine Jones [1], Bianca Blanch [1], Rajesh Puranik [2 3], Kevin McGeechan [4 5], Alexandra Barratt [4 5], Christopher Semsarian [1 2 3 5]

Conclusion: Left ventricular non-compaction in adult populations is a poorly defined entity which likely encompasses both physiological adaptation and pathological disease. There is a higher prevalence with the introduction of newer imaging technologies, specifically CMR imaging, which identify LVNC changes more readily. The clinical significance of these findings remains unclear; however, there is significant potential for overdiagnosis, overtreatment, and unnecessary follow-up.

Grazie per l'attenzione

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