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RESEARCH ARTICLE

RECOGNIZING NON-COMPACTION CARDIOMYOPATHY IN ANESTHETIC PRACTICE: PERIOPERATIVE IMPLICATIONS AND MANAGEMENT STRATEGIES

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ABSTRACT

Background: Non-compaction cardiomyopathy is a rare and potentially serious form of primary cardiomyopathy characterized by prominent myocardial trabeculations and deep recesses resulting from a defect in embryonic myocardial compaction. Clinically, it can manifest with heart failure, potentially lethal arrhythmias, and thromboembolic events. It is frequently underdiagnosed, thus posing a high risk during anesthetic procedures. **Objective:** The objective of this mini narrative review is to highlight the importance of preoperative recognition of non-compaction cardiomyopathy by the anesthesiologist, highlighting its clinical implications in the perioperative period and the available anesthetic management strategies. **Conclusion:** Preoperative identification of non-compaction cardiomyopathy allows the anesthesiologist to adopt specific and individualized strategies, such as detailed cardiological evaluation, advanced hemodynamic monitoring, and careful choice of anesthetic agents that minimize changes in myocardial contractility. Cases with ventricular dysfunction or arrhythmias require additional care, including inotropic support, availability of defibrillation, and thromboembolic prophylaxis. The use of intraoperative transesophageal echocardiography has been useful in more complex procedures. Knowledge of non-compaction cardiomyopathy is essential for safe anesthetic practice, and consequently, appropriate management can significantly reduce the risk of intraoperative and postoperative complications, improving the prognosis for these patients.

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INTRODUCTION

Non-compaction cardiomyopathy is a rare primary cardiomyopathy characterized by anomalous myocardial morphology resulting from failure of embryonic compaction of the myocardial layers, resulting in prominent trabeculations and deep intertrabecular recesses.[1-3] This condition may be asymptomatic or manifest clinically with heart failure, potentially fatal arrhythmias, and thromboembolic events, posing a significant risk during surgical and anesthetic procedures. Although described decades ago, non-compaction cardiomyopathy remains underdiagnosed, in part due to the absence of specific symptoms and phenotypic variability.[1-3] Given the increased detection rates of this condition through advanced imaging methods such as echocardiography and cardiac magnetic resonance imaging, it is crucial that anesthesiologists be able to recognize this clinical entity and its perioperative implications. The anesthetic management of patients with non-compaction cardiomyopathy requires individualized planning, focusing on hemodynamic stability, arrhythmia prevention, and strict control of circulating volume, as well as special attention to thromboembolic risk.[4-6] Therefore, preoperative recognition of this cardiomyopathy is essential for defining safe and effective anesthetic procedures, with a direct impact on reducing complications and optimizing perioperative care.[4,6] In this mini narrative review, the importance of recognizing non-compacted cardiomyopathy in anesthetic practice was emphasized. Scientific databases, including PubMed, Web of Science, Medline Plus, Google Scholar, Scopus, and using the terms "non-compaction cardiomyopathy; ventricular non-compaction cardiomyopathy; left ventricular non-compaction; anesthesia management" were searched for this review. Within these search terms, every subtitle in the text was searched individually. Full-text papers

were reviewed after removing duplicates and screening titles and abstracts and the most relevant information found is presented in this mini narrative review.

Non-compaction Cardiomyopathy: Definition, Pathophysiology, and Clinical Implications: Left ventricular non-compaction cardiomyopathy is a rare form of primary cardiomyopathy, considered genetic in origin, characterized by anomalous myocardial morphology resulting from an incomplete process of endomyocardial compaction during embryonic development. This condition leads to the formation of excessive myocardial trabeculations, associated with deep recesses between the trabeculae, which communicate directly with the ventricular cavity, giving the myocardium a spongy appearance.[7,8] From a pathophysiological perspective, non-compaction cardiomyopathy occurs due to the interruption or delay in the normal process of myocardial compaction, which should occur between the 5th and 8th week of gestation. This failure results in a non-compaction of the myocardium, predominantly located in the apical, inferolateral, and midlateral segments of the left ventricle, although it can also affect the right ventricle in some cases.[8]

Prevalence studies indicate that non-compaction cardiomyopathy may be present in up to 0.01–0.3% of the general population undergoing echocardiographic evaluation. However, this is likely an underestimate, given that the image quality of this method has improved significantly in recent years. Studies indicate that the prevalence of non-compaction cardiomyopathy in adults was consistently higher among cohorts diagnosed by cardiovascular magnetic resonance imaging (14.79%) compared with echocardiograms (1.28%).[2,9,10] Due to its variable and often nonspecific clinical presentation, which can range from asymptomatic to more severe forms, this condition has been frequently underdiagnosed due to complications that include: progressive systolic heart failure, often associated with ventricular dilation; cardiac arrhythmias, especially ventricular tachyarrhythmias and conduction blocks; and systemic thromboembolism, resulting from blood stasis in the trabecular recesses and systolic dysfunction, justifying, in many cases, the prophylactic use of oral anticoagulants. The most frequently reported symptoms include dyspnea, fatigue, palpitations, syncope, and, in advanced cases, signs of low cardiac output or cardiogenic shock.[11,12]

The diagnosis of non-compaction cardiomyopathy is based on specific imaging criteria, with two-dimensional echocardiography being the most widely used initial method. However, cardiac magnetic resonance imaging has emerged as a complementary tool with high sensitivity and specificity, allowing for the accurate assessment of the ratio between the non-compaction and compaction layers of the myocardium, generally using a ratio greater than 2.3 in the diastolic phase as a diagnostic criterion. Furthermore, magnetic resonance imaging can identify areas of associated myocardial fibrosis, which correlate with a worse prognosis.[8,13,14] Genetically, non-compaction cardiomyopathy is inherited in many cases as an autosomal dominant trait, with mutations identified in genes related to sarcomeric, mitochondrial, and cytoskeletal proteins. Approximately 30% to 50% of patients with non-compaction cardiomyopathy have a family history of the disease or other associated cardiomyopathies, which reinforces the importance of genetic counseling and family screening.[15,16]

Genetic studies show that the *MYH7, MYBPC3*, and *TTN* genes account for approximately 70% of cases with detectable mutations, and variants in the transcription factors *TBX5* and *NKX25* are relevant in patients with altered electrocardiograms or conduction system dysfunction.[17-20] More than 40 associated genes have been identified, including ion channels (*SCN5A, HCN4*),[21,22] cytoskeletal proteins (*DTNA, LDB3, LMNA*),[23-25] and mitochondrial genes such as *TAZ*,[23] with diverse inheritance patterns (dominant, recessive, X-linked, or mitochondrial). Furthermore, mutations in the *DES* gene, such as p.A337P, have been associated with the arrhythmic form of this cardiomyopathy.[26] The prognosis for patients with non-compaction cardiomyopathy depends on the severity of ventricular dysfunction, the presence of complex arrhythmias, and previous episodes of thromboembolism. Patients with New York Heart Association functional class III or IV, reduced ejection fraction, and ventricular tachyarrhythmias are at increased risk of sudden death and may be candidates for advanced therapies, such as cardioverter-defibrillator implantation or heart transplantation.[27,28] Given its clinical heterogeneity and significant risk of serious complications, non-compaction cardiomyopathy requires a multidisciplinary approach, including specialized cardiology evaluation, risk stratification, and individualized treatment planning. Early detection and appropriate management are essential to improving the prognosis and quality of life of affected patients.[14]

Anesthetic Implications of Non-Compaction Cardiomyopathy: Perioperative Assessment, Planning, and Management: The presence of non-compaction cardiomyopathy poses a significant challenge in anesthetic practice due to the high risk of perioperative complications, such as hemodynamic instability, malignant arrhythmias, ventricular decompensation, and thromboembolism. Prior knowledge of the condition, combined with a careful and individualized anesthetic approach, is essential to ensure patient safety and the appropriate conduct of the surgical procedure.[12,29]

Pre-Anesthetic Evaluation

A complete pre-anesthetic evaluation should be performed, with emphasis on investigating the following aspects.[30-32]

- Detailed clinical history, including symptoms suggestive of heart failure (dyspnea on exertion, orthopnea, fatigue), syncope, palpitations, and a history of arrhythmias. Early recognition allows for risk stratification and the adoption of appropriate prophylactic measures.
- Additional tests, especially electrocardiogram, recent transthoracic echocardiogram, and, if necessary, cardiac magnetic resonance imaging. Identification of the left ventricular ejection fraction and the extent of myocardial trabeculations is essential.
- Assessment of thromboembolic risk and anticoagulation status. Many patients with non-compaction cardiomyopathy are on oral anticoagulants, requiring adjustment of the therapeutic regimen for the perioperative period.
- Presence of implantable devices, such as implantable cardioverter-defibrillators or pacemakers, which require functional assessment and, in some cases, perioperative reprogramming.[30-32]

Individualized Anesthetic Planning

The choice of anesthetic plan should consider:[33-35]

- Type and duration of the surgical procedure, as well as the patient's degree of cardiovascular complexity.
- Minimization of sudden hemodynamic changes by choosing anesthetic agents with less impact on myocardial contractility and systemic vascular resistance.
- Prevention of fluid overload and prolonged hypotension, which can precipitate cardiac decompensation.
- Prophylaxis against arrhythmias, with continuous cardiac rhythm monitoring and immediate availability of antiarrhythmic drugs and defibrillation equipment.
- Use of regional or locoregional anesthesia, when possible, may be beneficial, provided there are no contraindications. However, sympathetic blockade should be carefully titrated due to the risk of significant hypotension in patients with reduced cardiac reserve.
- Careful pharmacological approach: Certain anesthetic drugs can exacerbate myocardial depression or precipitate arrhythmias in patients with non-compaction cardiomyopathy. Agents such as volatile anesthetics, neuromuscular blockers, and opioids should be used with caution, and the use of drugs with more stable cardiovascular profiles is recommended. Individualization of anesthesia, combined with clinical experience, is essential to ensure safety.[33-35]

Prevention of Perioperative Complications

Prevention is based on knowledge of complications.[30,36]

- Acute heart failure: should be avoided with strict control of intravascular volume, maintenance of blood pressure within safe limits, and inotropic support when necessary.
- Ventricular arrhythmias: can be triggered by hypoxemia, acidosis, electrolyte disturbances, or hemodynamic stress. Correction of these factors and the use of antiarrhythmic agents should be promptly implemented.
- Thromboembolism: Discontinuation of oral anticoagulation should be carefully considered, considering the thromboembolic risk versus the hemorrhagic risk. In some cases, short-term parenteral anticoagulation may be indicated in the perioperative period.[30,36]

Advanced Intraoperative Hemodynamic Monitoring

Hemodynamic care addresses the following monitoring:[29,34,37]

- Basic monitoring: continuous electrocardiogram with specific leads for early detection of arrhythmias, pulse oximetry, capnography, and noninvasive blood pressure.
- Invasive monitoring: indicated in medium and major surgeries or in patients with significant ventricular dysfunction.

It includes invasive blood pressure and, if necessary, central venous catheter and cardiac output monitoring. In selected cases, the use of intraoperative transesophageal echocardiography can provide dynamic, real-time information on ventricular function, cardiac filling, and the presence of intracavitary thrombi.[29,34,37]

Postoperative Procedures

Special attention to postoperative care:[32,37]

- Monitoring in an intensive care unit or advanced post-anesthesia care unit may be necessary in patients with compromised ventricular function or who presented intraoperative instability.
- Restarting anticoagulation should be assessed individually, based on the type of surgery and bleeding risk.
- Close monitoring for signs of cardiac decompensation, arrhythmias, and thromboembolic events.[32,37]

CONCLUSION

The anesthetic management of patients with non-compaction cardiomyopathy requires in-depth knowledge of the pathophysiology and potential complications associated with the disease. Early identification, thorough preoperative evaluation, individualized anesthetic planning, and intensive monitoring are fundamental strategies to ensure safe and effective anesthetic care and minimize the risk of serious complications. Furthermore, an interdisciplinary approach involving cardiology, anesthesiology, and intensive care is recommended, especially in complex or high-risk cases.

Given the potential severity of the clinical picture and the frequency of underdiagnosis, it is recommended that anesthesiologists be up to date on the clinical manifestations of non-compaction cardiomyopathy and be trained to intervene immediately in its possible occurrences and complications, thus contributing to better surgical and anesthetic outcomes.

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Glossary of Abbreviations

HCN4 - Hyperpolarization-activated Cyclic Nucleotide-gated Potassium Channel 4
LDB3 - LIM Domain-Binding Protein 3
LMNA - Lamin A/C
MYBPC3 - Myosin-Binding Protein C, Cardiac
MYH7 - Myosin Heavy Chain 7
NKX2-5 - NK2 Homeobox 5
SCN5A - Sodium Voltage-Gated Channel, Alpha Subunit 5
TAZ - Tafazzin
TBX5 - T-Box Transcription Factor 5
TTN - Titin

REFERENCES

1. Gerecke BJ, Engberding R. Noncompaction Cardiomyopathy-History and Current Knowledge for Clinical Practice. J Clin Med. 2021;10(11):2457. doi: 10.3390/jcm10112457.
2. Paluszkiewicz J, Milting H, Kałużna-Oleksy M, *et al*. Left Ventricular Non-Compaction Cardiomyopathy-Still More Questions than Answers. J Clin Med. 2022;11(14):4135. doi: 10.3390/jcm11144135.
3. Przytuła N, Dziewięcka E, Winiarczyk M, *et al*. Hypertrophic cardiomyopathy and left ventricular non-compaction: Distinct diseases or variant phenotypes of a single condition? World J Cardiol. 2024;16(9):496-501. doi: 10.4330/wjc.v16.i9.496.
4. Huda AU, Ahmed T. Anaesthetic Management of a Patient with Left Ventricular Non-Compaction Undergoing Laparotomy. J Coll Physicians Surg Pak. 2018;28(9):S152-S153. doi: 10.29271/jcpsp.2018.09.S152.
5. Rodríguez-de-Vera JM, Bernabé G, García JM, *et al*. Left ventricular non-compaction cardiomyopathy automatic diagnosis using a deep learning approach. Comput Methods Programs Biomed. 2022;214:106548. doi: 10.1016/j.cmpb.2021.106548.
6. Gallucci AE, Grewal MR, Alexander BT, *et al*. Left Ventricular Non-compaction Cardiomyopathy: The Key to Its Diagnosis and Implications for Management. Cureus. 2023;15(10):e47121. doi: 10.7759/cureus.47121.
7. Akhtar T, Bandyopadhyay D, Ghosh RK, *et al*. Left ventricular noncompaction cardiomyopathy: Recent update on genetics, usefulness of biomarker, and speckle imaging. J Cardiol. 2019;73(1):94-5. doi: 10.1016/j.jjcc.2018.05.016.
8. 8.Pittorru R, De Lazzari M, Migliore F, *et al*. Left Ventricular Non-Compaction: Evolving Concepts. J Clin Med. 2024;13(19):5674. doi: 10.3390/jcm13195674.
9. Ivanov A, Dabiesingh DS, Bhumireddy GP, *et al*. Prevalence and Prognostic Significance of Left Ventricular Noncompaction in Patients Referred for Cardiac Magnetic Resonance Imaging. Circ Cardiovasc Imaging. 2017;10(9):e006174. doi: 10.1161/CIRCIMAGING.117.006174.
10. Ross SB, Jones K, Blanch B, *et al*. A systematic review and meta-analysis of the prevalence of left ventricular non-compaction in adults. Eur Heart J. 2020;41(14):1428-36. doi: 10.1093/eurheartj/ehz317. PMID: 31143950.
11. Novo G, Vernuccio F, Fazio G, *et al*. Left ventricular non compaction: epidemiology, clinical findings, diagnosis and therapy. Minerva Cardioangiol. 2015;63(3):239-51.
12. Casas G, Rodríguez-Palomares JF, Ferreira-González I. Left ventricular noncompaction: a disease or a phenotypic trait? Rev Esp Cardiol (Engl Ed). 2022;75(12):1059-69. doi: 10.1016/j.rec.2022.07.002.
13. Jacquier A, Thuny F, Jop B, *et al*. Measurement of trabeculated left ventricular mass using cardiac magnetic resonance imaging in the diagnosis of left ventricular non-compaction. Eur Heart J. 2010;31(9):1098-104. doi: 10.1093/eurheartj/ehp595.
14. Mirchuk M. Risk Stratification of Left Ventricle Hypertrabeculation Versus Non-Compaction Cardiomyopathy Using Echocardiography, Magnetic Resonance Imaging, and Cardiac Computed Tomography. Echocardiography. 2025;42(5):e70165. doi: 10.1111/echo.70165.
15. van Waning JI, Moesker J, Heijsman D, *et al*. Systematic Review of Genotype-Phenotype Correlations in Noncompaction Cardiomyopathy. J Am Heart Assoc. 2019;8(23):e012993. doi: 10.1161/JAHA.119.012993.
16. Ichida F. Left ventricular noncompaction—risk stratification and genetic consideration. J Cardiol. 2020;75(1):1-9. doi:10.1016/j.jjcc.2019.08.002.
17. Zhang YH, Li XY, Song BR, *et al*. [Clinical phenotype and genetic analysis of patients with left ventricular noncompaction caused by the biallelic mutation of MYBPC3 and MYH7]. Zhonghua Xin Xue Guan Bing Za Zhi. 2023;51(11):1160-5. doi: 10.3760/cma.j.cn112148-20230929-00200.
18. Tian S, Liang H, Li X, *et al*. A novel mutation in the TTN gene resulted in left ventricular noncompaction: a case report and literature review. BMC Cardiovasc Disord. 2023;23(1):352. doi: 10.1186/s12872-023-03382-w.
19. Ross SB, Singer ES, Driscoll E, *et al*. Genetic architecture of left ventricular noncompaction in adults. Hum Genome Var. 2020;7:33. doi: 10.1038/s41439-020-00120-y. PMID: 33082984; PMCID: PMC7566488.
20. Yamada Y, Yasuda K, Hata Y, *et al*. A Novel *NKX2-5* Variant in a Child with Left Ventricular Noncompaction, Atrial Septal Defect, Atrioventricular Conduction Disorder, and Syncope. J Clin Med. 2022;11(11):3171. doi: 10.3390/jcm11113171.
21. Shan L, Makita N, Xing Y, *et al*. SCN5A variants in Japanese patients with left ventricular noncompaction and arrhythmia. Mol Genet Metab. 2008;93(4):468-74. doi: 10.1016/j.ymgme.2007.10.009.
22. Yonehara K. Fetal bradycardia associated with left ventricle noncompaction diagnosed as HCN4 mutations. Ann Pediatr Cardiol. 2024;17(4):295-7. doi: 10.4103/apc.apc_138_24.
23. Xing Y, Ichida F, Matsuoka T, *et al*. Genetic analysis in patients with left ventricular noncompaction and evidence for genetic heterogeneity. Mol Genet Metab. 2006;88(1):71-7. doi: 10.1016/j.ymgme.2005.11.009.
24. Finsterer J. Cardiogenetics, neurogenetics, and pathogenetics of left ventricular hypertrabeculation/noncompaction. Pediatr Cardiol. 2009;30(5):659-81. doi: 10.1007/s00246-008-9359-0.
25. O'Connor W, Arshia A, Prabakar D, *et al*. Nuclear envelope lamin-related dilated cardiomyopathy: a case series including histopathology. Eur Heart J Case Rep. 2024;8(8):ytae412. doi: 10.1093/ehjcr/ytae412.
26. Kulikova O, Brodehl A, Kiseleva A, *et al*. The Desmin (*DES*) Mutation p.A337P Is Associated with Left-Ventricular Non-Compaction Cardiomyopathy. Genes (Basel). 2021;12(1):121. doi: 10.3390/genes12010121.
27. Macaione F, Meloni A, Positano V, *et al*. The prognostic role of CMR using global planimetric criteria in patients with excessive left ventricular trabeculation. Eur Radiol. 2021;31(10):7553-65. doi: 10.1007/s00330-021-07875-0.

28. Heidenreich PA, Bozkurt B, Aguilar D, *et al*. . 2022 AHA/ACC/HFSA guideline for the management of heart failure: a report of the American College of Cardiology/American Heart Association Joint Committee on Clinical Practice Guidelines. Circulation. 2022;145(18):e895-e1032. doi:10.1161/CIR.0000000000001063.
29. Nishikawa K, Kariya N, Shin W, *et al*. Pronounced QT prolongation during general anesthesia in a child with left ventricular noncompaction cardiomyopathy: a case report. JA Clin Rep. 2023;9(1):70. doi:10.1186/s40981-023-00631-0.
30. Sviggum HP, Kopp SL, Rettke SR, *et al*. Perioperative complications in patients with left ventricular non-compaction. Eur J Anaesthesiol. 2011;28(3):207-12. doi: 10.1097/EJA.0b013e328341014c.
31. Ramachandran P, Woo JG, Ryan TD, *et al*. The Impact of Concomitant Left Ventricular Non-compaction with Congenital Heart Disease on Perioperative Outcomes. Pediatr Cardiol. 2016;37(7):1307-12. doi: 10.1007/s00246-016-1435-2.
32. Jackson AR, Kuller JA, Rhee KY, *et al*. Anesthetic challenges of labor and delivery in a patient with catecholaminergic polymorphic ventricular tachycardia and left ventricular non-compaction cardiomyopathy. J Clin Anesth. 2025;84:111059. doi:10.1016/j.jclinane.2024.111059.
33. De Hert SG, Turani F, Mathur S, *et al*. Cardioprotection with volatile anesthetics: mechanisms and clinical implications. Anesth Analg. 2005;100(6):1584-93. doi: 10.1213/01.ANE.0000153483.61170.0C.
34. Bazoukis G, Tyrovolas K, Letsas KP, *et al*. Predictors of fatal arrhythmic events in patients with non-compaction cardiomyopathy: a systematic review. Heart Fail Rev. 2022;27(6):2067-76. doi: 10.1007/s10741-022-10257-3.
35. Hochfelder MA, White RD, McGregor DG, *et al*. Peripartum anesthetic management in patients with left ventricular noncompaction: a case series and review of the literature. J Cardiothorac Vasc Anesth. 2022;36(9):3003-9. doi:10.1053/j.jvca.2022.06.010.
36. Shapiro RS, Bohanon TM, Rupright J, *et al*. Transient cardiac arrest during general anesthesia in a child with left ventricular noncompaction (spongiform cardiomyopathy). Anesth Prog. 2011;58(1):22-4. doi:10.2344/0003-3006-58.1.22.
37. Kim D, Kim E, Lee JH, *et al*. Anesthetic experience of patient with isolated left ventricular noncompaction: a case report. Korean J Anesthesiol. 2016;69(3):275-8. doi: 10.4097/ kjae.2016.69.3.275.

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