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# Noncompacted myocardium and physical activity: a systematic review of case studies

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## ABSTRACT

### OBJECTIVE

To check whether physical activity is recommended for those who practice physical activity with left ventricular noncompaction (LVNC).

### METHODS

This is a systematic review of case reports published between 2010 and 2024 on the Pubmed and Scielo platforms related to the topic of the present study. To ensure greater reliability and quality of the study, the guidelines of PRISMA declaration were applied.

### RESULTS

From 684 encountered articles, ten belonged to the final sample only. Among 10 practitioners, 60% were male, amateur athletes and symptomatic. Symptoms presented during or after physical exercise were chest pain, palpitations, syncope and dyspnea. The average age was 24.3 years and the ejection fraction ranged from 32 to 65%. Anticoagulant therapies and defibrillator implantation were seen in four studies (40%). The evolution of the cases studied was positive, except for the single death; Of the 9 living patients, only 1 was suspended from physical activity due to the clinical condition and important family history.

### CONCLUSIONS

Among the nine patients, one died, one was suspended from sports practice and four were released to practice. They were asymptomatic and had preserved ejection fraction. Furthermore, it is not known whether or not they presented any event after the observation period of each study. Therefore, it is concluded that, for physical activity practitioners with MNC, regardless of the workload and intensity of the exercise, amateur or competitive sports practice should not be recommended due to the scarcity of studies proving long-term safety.

### DESCRIPTORS

Congenital heart diseases, Cardiomyopathies, Cardiology.

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## INTRODUCTION

Non-compaction myocardium (NCM) is a rare congenital cardiomyopathy whose etiology remains unknown. Its clinical triad consists of heart failure, atrial and/or ventricular arrhythmias, and thromboembolic events[1,2]. The age at diagnosis does not follow any specific pattern, as the disease can manifest in both children and adults, and, on the other hand, it can also be asymptomatic[2].

It was first described in 1926 by Grant, and in 1984 it was characterized following its detection on an echocardiogram[3]. In 1990, the disease was named "noncompaction of the myocardium" by Chin and others[4]. In the last few decades, NCM has been more widely studied by doctors around the world, but it is still a topic that lacks extensive information, making medical management challenging.

The current global prevalence is 0.5%, and it affects more males. In recent years, the incidence has increased due to advances in diagnostic tools, particularly echocardiography, which has led to a higher detection rate[4]. However, the prevalence and incidence among different populations remain uncertain due to the lack of larger studies. Brazil lacks epidemiological data on the disease.

Diagnostic methods have evolved significantly in recent years. Today, the primary choice for diagnosis is two-dimensional echocardiography with color Doppler due to its practicality and availability in healthcare settings. However, the gold standard is cardiac magnetic resonance imaging, which can diagnose NCM with high accuracy[1,2]. Additionally, cardiac biomarkers may show no abnormalities, such as troponin[2].

Its pathogenesis basically consists of genetic mutations that generate neuromuscular dysfunctions that result in a myocardium with two layers, one at the apex and another on the lateral wall of the left ventricle (LV), normally; they are a spongy endocardial layer containing prominent and numerous trabeculations and deep intertrabecular recesses that communicate with the cavity of the LV chamber (not compacted), but not with the coronary circulation; and a compacted and thin epicardial layer[1,5]. These characteristics result from a possible interruption of the myocardial compactation process during the first half of embryonic development, of still unknown origin. The earlier the non-compaction occurs, the larger the trabeculae and the greater the ventricular dysfunction[1,6].

Athletes with NCM are among the high-risk group for sudden death, regardless of age, especially those who are asymptomatic or have not yet been diagnosed. Therefore, every athlete should be closely monitored by healthcare professionals to facilitate early diagnosis. Analyzing the morphological and functional aspects of the affected heart, as well as the presence of conduction disturbances and arrhythmias, can assist the physician in recommending physical activity, distinguishing NCM from a benign or non-benign cardiomyopathy[7].

The American College of Cardiology (ACC)[8], in its guidelines on recommendations for eligibility and disqualification of competitive athletes with cardiovascular abnormalities, recommends the clearance of competitive sports for those patients with asymptomatic NCM, normal systolic function, no significant ventricular tachyarrhythmias on ambulatory monitoring or exercise testing, and specifically, no previous history of unexplained syncope (class IIb and level of evidence C). In 2005, the same international body recommended that any patient with NCM should refrain from participating in competitive sports[9]. However, despite the updated recommendations, there is still a lack of discussions and studies on the topic, mainly due to the wide variety of symptoms and the scarcity of studies on a disease that has only been more deeply studied in recent years.

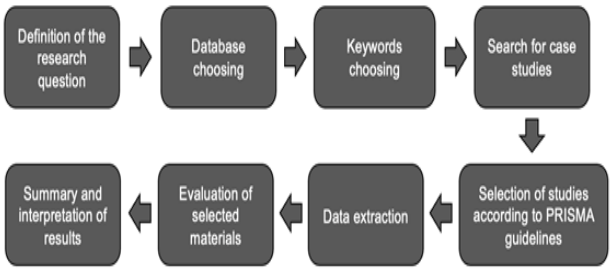
The objective of this study is to determine whether physical activity is recommended for individuals who engage in physical activity and competitive athletes with NCM, and to compare the different medical approaches regarding the release of studied patients from sports practice.

## METHODS

This is a systematic review of case reports published between 2010 and 2024 on the PubMed and SciELO platforms related to the topic of the present study. The search and selection terms used were: "non-compaction cardiomyopathy," "spongy myocardium," "non-compaction of the myocardium," and "left ventricular noncompaction cardiomyopathy," which were then combined with the terms "physical activity," "athlete," and "exercise". Among the selected articles, those that were not case reports or were unrelated to the topic or the specified time period were excluded from the sample. After selecting the articles for the final sample, a full reading of each one was conducted to include the most important findings in the discussion of the study.

To ensure greater reliability and quality of the study, PRISMA guidelines (Preferred Reporting Items for Systematic Reviews and Meta-Analyses)[10] were used to select the articles for the final sample. Figure 1 summarizes the steps required in the development of a systematic review, steps that were followed in this study. The PICOS acronym[11] was applied to define the research question, corresponding to the first step in Figure 1.

**Figure 1** - Flowchart of the steps for conducting a systematic review.




**Source:** authors.

## RESULTS AND DISCUSSION

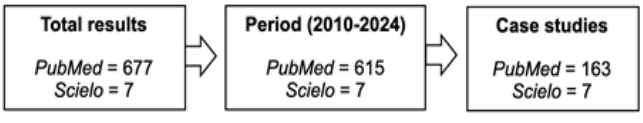
Table 1 refers to the PICOS acronym applied in the study methodology to determine the main research question. Figure 2 summarizes the search for case studies in the Scielo and PubMed databases.

**Table 1** - Research Question Components According to the PICOS Acronym.

| Description | Abbreviation | Question Components |
|---|---|---|
| Population | P | Amateur or professional physical activity practitioners with NCM |
| Intervention | I | Physical activity |
| Comparison | C | Different approaches regarding physical activity among the studied cases |
| Outcome | O | Improvement or worsening of symptoms, sudden death, suspension of physical activity |
| Study Type | S | Systematic review of case reports |

Source: authors.

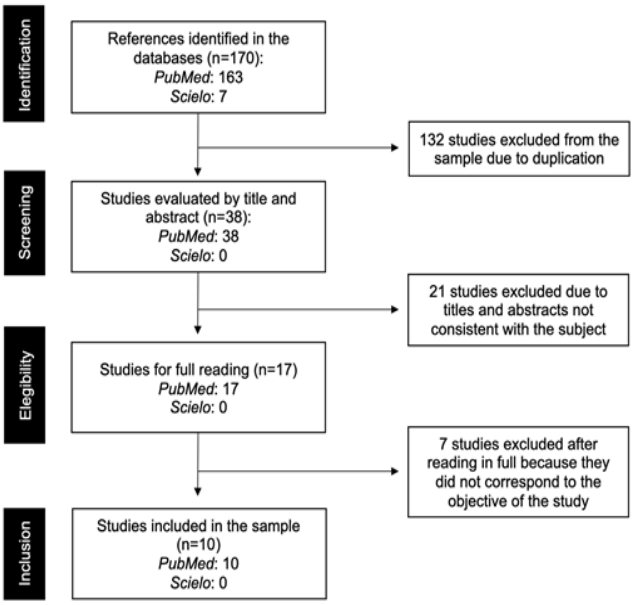
**Figure 2** - Flowchart of bibliographic findings according to the filters applied in the database, based on the keywords used.




**Source:** authors.

The 170 case studies found in the databases underwent PRISMA selection to filter the studies that would be included in the final sample. Figure 3 refers to the PRISMA flowchart of the steps applied in the study.

**Figure 3** - Flowchart of the steps of the PRISMA guidelines for selecting case studies.




**Source:** authors.

In summary, 10 case studies involving individuals who engage in physical activity and have non-compaction cardiomyopathy were included in the sample. Table 2 presents the case reports studied and their key information.

**Table 2** - Main data obtained from the case reports studied.

| Sex and age | Comorbidities | Physical exercise | Symptoms | EF | Management |
|---|---|---|---|---|---|
| Woman, 14y. [12] | Biventricular systolic dysfunction | Goalball parathlete (6h weekly) | - | N/A | Aspirin 100mg/day. Patient cleared for sports with annual clinical follow-up |
| Man, 18y. [20] | - | Soccer | - | 65% | Patient cleared for sports. Annual clinical follow-up |
| Man, 20y. [13] | Asthma and systolic murmur | Basketball | - | 45% | Patient cleared for sports. Semi-annual clinical follow-up |
| Man, 18y. [14] | Asthma and systolic murmur | Soccer | - | 60% | Referral to two specialists: (1) MNC diagnosis and 81mg aspirin (2) Physiological hypertrabeculation without pharmacotherapy. Patient cleared for sports with quarterly clinical follow-up |
| Woman, 52y. [21] | Hypertension and benign ventricular tachycardia | N/A | Chest pain and palpitations during exercise | 45% | Bisoprolol and repeat stress ECG. No specific guidance provided for physical activity |
| Man, 21y. (deceased) [15] | Arrhythmogenic right ventricular cardiomyopathy | Futsal and soccer (2x/week) | Sudden death | N/A | N/A |
| Man, 17y. [22] | Asthma | Wrestler | Recurrent syncope and symptomatic bradycardia | 60% | Dual-chamber pacemaker implantation and ICD. **Permanent suspension from any physical activity** |
| Woman, 31y. [23] | Neuromuscular disorder and mild mitral insufficiency | N/A | Muscle weakness in limbs, dyspnea, palpitations, and chest pain after exercise | 39% | Coenzyme Q10, B1, B2, B6 vitamins, and ATP for 2 months. Subsequent therapy: perindopril 4mg/day + spironolactone 20mg/day. No specific guidance for physical activity |
| Woman, 20y. [24] | Catecholaminergic polymorphic ventricular tachycardia (CPVT) | Running, basketball, dance | Syncope during exercise | 59% | Diltiazem at age 16. At 17: ICD, but symptoms persisted, and sotalol therapy began. At 20, sudden death during walking. After CPVT diagnosis, therapy with flecainide and nadolol started, and bilateral sympathectomy performed. No specific guidance for physical activity |
| Man, 32y. [25] | N/A | N/A | Dyspnea and persistent AF | 32% | ICD implantation, pulmonary vein isolation + amiodarone 200mg, eplerenone 25mg, rivaroxaban 20mg, and perindopril 4mg. No specific guidance for physical activity |

**Legend:** ICD: Implantable Cardioverter Defibrillator. AF: Atrial Fibrillation. NYHA: New York Heart Association functional classification. N/A: "Not available" or "Not applicable."

**Source:** authors.

From the 10 individuals studied, the majority (60%) were male and presented symptoms during or after physical activity, including chest pain, muscle weakness, dyspnea, palpitations, and syncope. The average age was 24.3 years (± 14-52 years).

In the case reports studied, only one patient identified as an athlete; six others were amateur physical activity practitioners, and three did not report which sport they practiced. The only athlete presented[12] was a 14-year-old para-athlete who, in addition to NCM, was diagnosed with mild biventricular systolic dysfunction. She was cleared to practice sports with prophylactic aspirin (100mg/day) and clinical follow-up every six months after a thorough investigation of her clinical condition. The use of anticoagulants in patients with NCM is still controversial; while some authors suggest that all patients with NCM should use the medication, others believe that only those with NCM and high thromboembolic risk should do so. This includes individuals with a history of atrial fibrillation, previous systemic embolism, severely impaired systolic function, and left ventricular thrombus[13].

It is common for patients with NCM to be asymptomatic, as well as to have a normal heart auscultation. All patients, except for those in two studies where patients had a systolic murmur[13,14], had a physical heart exam without any abnormalities.

Comorbidities associated with NCM can impact the prognosis of the disease and influence whether physical activity practices are allowed or suspended. The individual studied by Razuin and colleagues[15] died of sudden cardiac death during an amateur futsal match. However, after an autopsy, doctors concluded that, despite having NCM, his arrhythmogenic cardiomyopathy may have further contributed to his sudden death. It is known that young and athletic individuals have a lower risk of cardiovascular diseases, especially sudden cardiac death. However, individuals—whether young or not—with congenital cardiovascular disorders may face a higher risk of sudden death, up to 2.8 times greater compared to non-athletes[15]. However, among the 10 active patients studied, one passed away only. Furthermore, five individuals studied had another cardiac comorbidity in addition to NCM, but none of the studies examined whether the symptoms presented were due to NCM, the other clinical condition, or both of them.

The ejection fraction (EF), an index that measures the left ventricle's contraction capacity, ranged from 32% to 65% among the individuals. Two studies did not specify the ejection fraction. According to the ACC guidelines[8], athletes with impaired systolic function should not engage in competitive sports, except for those with low-intensity activities. However, among the nine living individuals studied, four had impaired systolic function, and in three of these cases, the approach regarding physical activity was not specified.

NCM remains an underdiagnosed disease. Most individuals with NCM are diagnosed when the disease is already advanced or through exams required for competitions (as is the case with the individuals studied). The lack of awareness of NCM further contributes to underdiagnosis, medical errors, or late diagnosis[16]. In the study by Peritz and colleagues[14], included in the sample studied, to participate in his college competitions, the patient was referred to two specialists who disagreed in their diagnoses: one concluded the diagnosis of NCM, while the other stated it was simply a case of physiological hypertrabeculations due to increased myocardial demand. This diagnostic disagreement could interfere with both his routine as a college football player and the management of the disease itself.

NCM is often confused with athlete's heart syndrome, which involves myocardial hypertrabeculation as an adaptation to the excessive physical exertion by athletes. Many athletes end up being suspended from their activities due to echocardiographic changes that confuse doctors who suspect NCM. These athletes are asymptomatic, but the absence of symptoms combined with hypertrabeculations of the left ventricle does not rule out the diagnosis of NCM. There is still a lack of data supporting the theory that athlete's heart is exclusively a benign condition[17]. In the present study, some case reports were removed from the final sample because they concluded that the athletes studied, after thorough investigations over several months, actually had athlete's heart and not NCM[18-20].

The present study had some limitations. The first is that it

was not possible to classify the individuals according to the duration and intensity of the physical activity they practiced. Although only one individual identified as an athlete, their weekly training time was six hours—this was the only study that specified training hours. Additionally, three studies did not specify the type of sport practiced. The sample size and the short follow-up period (ranging from six months to two years) limited the ability to conduct a more complex evaluation. Another limitation relates to the approach regarding suspension or clearance for sports participation: only four of the ten studies cleared the individuals for physical activity, while one was suspended and another four did not specify the course of action. It is hoped that the present study will contribute to encouraging other researchers to continue in this line of inquiry so that the issues related to NCM and physical activity can be addressed.

## CONCLUSION

Among the nine patients, one died and one was suspended from sports participation; four were liberated to continue practicing, and the others have had not a specific course of action. The patients who were released for physical practice were asymptomatic and had preserved ejection fraction, although one of them had biventricular systolic dysfunction. However, it is not possible to determine whether they experienced any events after the observation period of each study, which ranged from six months to two years—a relatively short period. It is also not possible to determine the maximum duration and intensity of exercise that can be safely practiced. Therefore, it is concluded that, for individuals with NCM who engage in physical activity, regardless of exercise duration and intensity, amateur or competitive sports should not be recommended due to the lack of studies proving the long-term safety of physical exercise for cardiac health, ensuring that irreversible consequences of the disease do not occur.

Attention should be paid to those patients who are symptomatic, with reduced ejection fraction and/or a personal or family history of cardiovascular events due to the higher risk of future events and clinical deterioration. Half of the patients studied had another cardiopathy, making it unclear whether the symptoms were truly due to NCM or another heart condition. This highlights the importance of follow-up with a cardiologist who has advanced knowledge of the disease, as it should be mandatory to prevent misdiagnoses.

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