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## Original article

# Electrocardiographic changes in dermatomyositis and polymyositis



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### ABSTRACT

**Introduction:** Cardiac involvement is frequent in inflammatory myopathies. Electrocardiogram (ECG) may show evidence of this involvement and its changes should be well-known and described.

**Objectives:** Due to the lack of studies in the literature, we conducted an analysis of the ECG findings in patients with dermatomyositis (DM) and polymyositis (PM), comparing them with a control group.

**Methods:** This cross-sectional study compared the ECG of 86 individuals with no rheumatic disorders (controls) with 112 patients (78 DM and 34 PM), during 2010–2013. The ECG findings between DM and PM were also compared.

**Results:** Demographic characteristics, comorbidities and ECG abnormalities were similar between controls and patients ( $p > 0.05$ ), except for a higher frequency of left ventricular hypertrophy (LVH) in patients (10.7% vs. 1.2%,  $p = 0.008$ ). Demographic characteristics, comorbidities, clinical and laboratory manifestations, were also similar between the groups PM and DM, except for the presence of cutaneous lesions only in DM. One-third of the patients had ECG abnormalities, which were more prevalent in PM than DM (50% vs. 24.4%,  $p = 0.008$ ). LVH, left atrial enlargement, rhythm and conduction abnormalities were more frequent in PM than DM ( $p < 0.05$  for all), especially the left anterior fascicular block.

**Conclusions:** We showed distinct ECG changes between DM and PM and a higher frequency of LVH in patients compared to controls. Investigation of cardiac involvement should be considered even in asymptomatic patients, especially PM. Further studies are necessary in order to determine the correlation of ECG findings with other complementary tests, clinical manifestations, disease activity and progression to other cardiac diseases.

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## Alterações eletrocardiográficas em dermatomiosite e polimiosite

### R E S U M O

#### Palavras-chave:

Coração  
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**Introdução:** Acometimento cardíaco nas miopatias inflamatórias é frequente. Eletrocardiograma (ECG) pode mostrar indícios desse acometimento e suas alterações devem ser bem conhecidas e descritas.

**Objetivos:** Devido à escassez de trabalhos na literatura, analisamos as alterações de ECG em pacientes com dermatomiosite (DM) e polimiosite (PM) e as comparamos com um grupo controle.

**Métodos:** Este estudo transversal comparou ECGs de 86 indivíduos sem doenças reumatológicas (controles) com 112 pacientes (78 DM e 34 PM), de 2010 a 2013. Também comparamos os ECGs entre DM e PM.

**Resultados:** Características demográficas, comorbidades e alterações de ECG foram semelhantes entre controles e pacientes ( $p > 0,05$ ), exceto pela maior frequência de sobrecarga de ventrículo esquerdo (SVE) nos pacientes (10,7% vs. 1,2%;  $p = 0,008$ ). Características demográficas, comorbidades, manifestações clínicas e laboratoriais também foram semelhantes entre os grupos PM e DM, exceto por lesões cutâneas apenas em pacientes com DM. Um terço dos pacientes apresentou alterações de ECG, que foram mais prevalentes em PM do que em DM (50% vs. 24,4%,  $p = 0,008$ ). Sobrecarga de câmaras esquerdas (SCE), distúrbios do ritmo e da condução foram mais encontrados em PM do que em DM ( $p < 0,05$  para todos), sobretudo o bloqueio divisional do ramo anterossuperior.

**Conclusões:** Encontramos alterações distintas de ECG entre PM e DM e frequência aumentada de SVE em pacientes quando comparados com controles. Investigação do acometimento cardíaco nessas doenças deve ser considerada mesmo em pacientes assintomáticos, especialmente em se tratando de PM. Mais estudos são necessários para correlacionar os achados de ECG com outros exames complementares, manifestações clínicas, atividade das miopatias e evolução para outras doenças cardíacas.

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## Introduction

Dermatomyositis (DM) and polymyositis (PM) are immune-mediated inflammatory disorders characterized by the presence of progressive proximal muscle weakness of limbs. In DM, typical skin changes, such as heliotrope and/or Gottron sign, still occur. Furthermore, extramuscular manifestations such as joint, cardiopulmonary and gastrointestinal tract involvement can also be present in these diseases.<sup>1,2</sup>

In particular, cardiac involvement in idiopathic inflammatory myopathies can occur in 9–72% of cases, depending on the diagnostic method used and selection of patients.<sup>3–6</sup>

Among patients with cardiac involvement, the clinical manifestations are infrequent and, when present, are mainly related to congestive heart failure.<sup>6,7</sup> In contrast, the vast majority of patients are asymptomatic and characterized especially by changes in the conduction system, hypertrophy and dilatation of the heart chambers and diastolic left ventricular dysfunction.<sup>4,5</sup>

To date, there are few studies in the literature specifically assessing changes in the electrocardiogram (ECG) in idiopathic inflammatory myopathies.<sup>3–5,7–13</sup> When they exist, are essentially limited to vague and brief descriptions of electrocardiographic findings in this population.<sup>7,9,10,12,13</sup> Furthermore, none of these studies<sup>7,9,10,12,13</sup> presented a control

group, nor examined systematically and comparatively possible electrocardiographic abnormalities found in patients with DM and PM, which motivated us to conduct this study.

## Materials and methods

This is a cross-sectional, single-center study. One hundred and twelve consecutive patients with DM and PM were evaluated according to the criteria of Bohan and Peter<sup>14,15</sup> in the period between 2010 and 2013, and followed at our department. No patients with amyopathic DM, inflammatory myopathies associated with other collagen diseases, malignancies or other causes of myopathy were included. As a control group, 86 outpatient subjects with no history of rheumatic disease were selected. All participants of this study were aged  $\geq 18$  years, did not use antiarrhythmic drugs nor had a history of angina, palpitations, myocardial infarction, heart failure, valvular disease and/or hyperthyroidism.

The study was approved by the local Ethics Committee [HC 0039/10].

Clinical and laboratory data of patients with DM and PM were obtained from a systematic review of the electronic medical record with previously parameterized and standardized data, including those of interest for the present study. The parameters analyzed were: disease duration and mean age at the time of performing ECG; gender; clinical

manifestations (constitutional symptoms, dysphagia, joint involvement [arthritis and/or arthralgia]); pulmonary involvement (confirmed by computed tomography: presence of interstitial lung disease, “ground-glass” or honeycombing lesions); skin changes (heliotrope, Gottron papules, ulcers, calcinosis, cutaneous vasculitis, V-sign, shawl sign); laboratory abnormalities (serum creatine phosphokinase [CPK], with a reference value of 24–173 U/L obtained by an automated kinetic method; antinuclear factor assessed by indirect immunofluorescence using Hep-2 cells as substrate, anti-Jo-1 antibody by immunoblotting method).

Demographic data and comorbidities (hypertension, diabetes mellitus and hypothyroidism) of all participants were also obtained.

A resting 12-Lead ECG was ordered to all patients for this study; these tests were performed and analyzed at the

Service of Electrocardiology in the same Institute. The control group consisted of sequential individuals who had an ECG performed in the same service, mostly in the preoperative workup of non-vascular/cardiac surgery. The following electrocardiographic changes were analyzed in the study: rhythm disturbances (ventricular extrasystole, sinus arrhythmia, atrial fibrillation, atrial ectopic rhythm, supraventricular tachycardia, atrioventricular block and supraventricular extrasystoles), chamber hypertrophy (left and right ventricles, left and right atria), conduction abnormalities (right bundle branch block [RBBB], left bundle branch block [LBBB], anterosuperior divisional branch block [ASDBB]) and diffuse ventricular repolarization changes (DVRC). The changes found were compared among control, PM and DM groups.

Results were expressed as mean  $\pm$  standard deviation (SD), median [25–75% interquartiles] or percentage (%). For statistical

**Table 1 – Demographic and clinical data and comorbidities (dermatomyositis and polymyositis) of patients and control group.**

	Control (n = 86)	DM/PM (n = 112)	<i>p</i>
Mean age (years)	49.0 $\pm$ 15.3	48.9 $\pm$ 15.4	0.944
Female gender	70 (69.0)	80 (71.4)	0.923
Disease duration (years)	–	5 [2–12]	–
<b>Comorbidities</b>			
Hypothyroidism	10 (11.9)	15 (13.4)	0.877
Hypertension	31 (36.9)	51 (45.5)	0.231
Diabetes mellitus	16 (19.0)	17 (15.2)	0.654
<b>Electrocardiographic changes</b>			
30 (35.8)	36 (32.1)	0.761	
1 item (A, B, C or D)	26 (31.0)	29 (25.9)	0.525
2 items (A, B, C or D)	4 (4.8)	5 (4.5)	1.000
3 items (A, B, C or D)	0	2 (1.8)	–
4 items (A, B, C and D)	0	0	1.000
<b>(A) Rhythm changes</b>			
7 (8.1)	12 (28.6)	0.631	
Ventricular extrasystole	2 (2.4)	3 (2.7)	1.000
Sinus arrhythmia	0	3 (2.7)	–
Atrial fibrillation	1 (1.2)	1 (0.9)	1.000
Atrial ectopic rhythm	0	2 (1.8)	–
First-degree A-V block	2 (2.4)	3 (2.7)	1.000
Supraventricular tachycardia	0	1 (0.9)	–
Supraventricular extrasystole	2 (2.4)	1 (0.9)	1.000
Change of 1 sub-item of A	7 (8.3)	10 (8.9)	1.000
Change of 2 sub-items of A	0	2 (1.8)	–
Change of 3 or more sub-items of A	0	0	1.000
<b>(B) Chamber hypertrophy</b>			
3 (3.6)	13 (11.6)	0.063	
Left ventricular	1 (1.2)	12 (10.7)	0.008
Right ventricular	0	0	1.000
Left atrial	2 (2.4)	5 (4.5)	0.607
Right atrial	0	0	1.000
Change of 1 sub-item of B	3 (3.6)	9 (8.0)	0.237
Change of 2 sub-items of B	0	4 (3.6)	–
Change of 3 or more sub-items of B	0	0	1.000
<b>(C) Conduction disturbance</b>			
6 (7.1)	6 (16.0)	0.766	
ASDBB	4 (4.8)	5 (4.5)	1.000
RBBB	1 (1.2)	1 (0.9)	1.000
LBBB	1 (1.2)	1 (0.9)	1.000
Change of 1 sub-item of C	6 (7.1)	5 (4.5)	0.766
Change of 2 sub-items of C	0	1 (0.9)	–
Change of 3 or more sub-items of C	0	0	1.000
<b>(D) DVRC</b>			
18 (21.4)	14 (12.5)	0.161	

DVRC, diffuse ventricular repolarization changes; A-V, atrioventricular; ASDBB, anterosuperior divisional branch block; RBBB, right bundle branch block; LBBB, left bundle branch block; DM, dermatomyositis; PM, polymyositis.

**Table 2 – Demographic and clinical data and comorbidities of patients with dermatomyositis and polymyositis.**

	Total (n = 112)	DM (n = 78)	PM (n = 34)	<sup>a</sup> p
Mean age (years)	48.9 ± 15.4	49.1 ± 15.9	48.3 ± 14.3	0.789
Female gender	80 (71.4)	58 (74.4)	22 (64.7)	0.298
Disease duration (years)	5 [2–12]	6 [3–15]	4 [1–10]	0.121
<b>Clinical manifestations</b>				
Constitutional symptoms	27 (24.1)	19 (24.4)	8 (23.5)	0.565
Dysphagia	27 (24.1)	20 (25.6)	7 (20.6)	0.329
Joint involvement	46 (41.1)	31 (39.7)	15 (44.1)	0.661
Pulmonary involvement	37 (33.0)	28 (35.9)	9 (26.5)	0.329
<b>Skin lesions</b>				
Heliotrope	65 (58.0)	65 (83.3)	0	–
Gottron's papules	73 (65.2)	73 (93.6)	0	–
Ulcers	32 (28.6)	8 (10.3)	0	–
Calcinosis	10 (8.9)	10 (12.8)	0	–
Cutaneous vasculitis	11 (9.8)	11 (14.1)	0	–
V sign	9 (8.0)	9 (11.5)	0	–
Shawl sign	3 (2.7)	3 (3.8)	0	–
<b>Laboratory abnormalities</b>				
Initial CPK (U/L)	723 [244–4012]	533 [170–3748]	2076 [728–4868]	0.221
Antinuclear factor	49 (43.8)	36 (46.2)	13 (38.2)	0.437
Anti-Jo-1 antibody	8 (7.1)	4 (5.1)	4 (11.8)	0.261
<b>Comorbidities</b>				
Hypothyroidism	15 (13.4)	10 (12.8)	5 (14.7)	0.770
Hypertension	51 (45.5)	37 (47.4)	14 (41.2)	0.541
Diabetes mellitus	17 (15.2)	10 (12.8)	7 (20.6)	0.390

CPK, creatine phosphokinase; DM, dermatomyositis; PM, polymyositis.

<sup>a</sup> Comparison between patients with polymyositis and dermatomyositis.

analysis, Student's t and Mann–Whitney tests for continuous variables and chi-squared or Fisher's exact test for categorical data were used. These calculations were performed with the computer program STATA version 7.0 (STATA, College Station, TX, USA). *p*-Values < 0.050 were considered statistically significant.

## Results

The general characteristics of the 112 patients (DM and PM) and of the control group (*n* = 86) are presented in Table 1. Demographic data and comorbidities were similar in both groups (*p* > 0.05). The ECG findings were also similar, except for a higher frequency of left ventricular hypertrophy (LVH) in patients when compared to the control group (10.7% vs. 1.2%, *p* = 0.008). Changes of rhythm were more frequent in patients versus controls, but without statistical significance (28.6% vs. 8.1%, *p* = 0.631).

An additional analysis comparing 78 DM and 34 PM patients will be presented in Table 2. Patients with DM and PM were comparable regarding demographic, clinical and laboratory data and also as to comorbidities, except for the presence of skin lesions only in patients with DM.

One-third of the patients had electrocardiographic abnormalities (Table 3), which included rhythm disturbances (ventricular extrasystole, sinus arrhythmia, atrial fibrillation, ectopic atrial rhythm, first-degree AV block, supraventricular tachycardia, supraventricular extrasystoles), chamber hypertrophy (atrium and left ventricles) and conduction disturbances (ASDBB, RBBB and LBBB) and DVRC. The presence

of changes was found more frequently in patients with PM when compared to patients with DM (50% vs. 24.4%, *p* = 0.008). More than a change of ECG per patient was observed only in PM group, and no cases have been reported in patients with DM. None of the study participants showed right-chamber changes.

The frequency of rhythm disturbances and chamber overload were higher in patients with PM compared to those with DM (*p* = 0.007 and *p* = 0.003, respectively), and both LVH and left atrium hypertrophy were predominant in patients with PM (*p* = 0.007 and *p* = 0.029, respectively) (Table 3). Atrial fibrillation, supraventricular extrasystoles and first-degree atrioventricular block were observed only in patients with PM, and no case have been reported in patients with DM. Supraventricular tachycardia was observed only in one case in the entire study, in a patient with DM.

Conduction disorders also were more frequent in patients with PM (*p* = 0.010), with emphasis for ASDBB (*p* = 0.029). No patient had RBBB or LBBB in the DM group compared to a case of each in PM group.

## Discussion

In the present study, we analyzed the electrocardiographic changes (rhythm, chamber hypertrophy and conduction) in a large case series of patients with DM and PM, compared to a control group. Our results showed that the frequency of ECG changes in patients with DM and PM was similar to that observed in the control group, except for a higher prevalence of LVH in our patients. Further analysis showed that changes

**Table 3 – Demographic, clinical and laboratory data and comorbidities of patients with dermatomyositis and polymyositis.**

	Total (n = 112)	DM (n = 78)	PM (n = 34)	p
<i>Electrocardiographic changes</i>	36 (32.1)	19 (24.4)	17 (50.0)	0.008
1 item (A, B, C or D)	29 (25.9)	19 (24.4)	10 (29.4)	0.367
2 items (A, B, C or D)	5 (4.5)	0	5 (14.7)	–
3 items (A, B, C or D)	2 (1.8)	0	2 (5.9)	–
4 items (A, B, C and D)	0	0	0	1.000
(A) <i>Rhythm changes</i>	12 (28.6)	4 (5.1)	8 (23.5)	0.007
Ventricular extrasystole	3 (2.7)	1 (1.3)	2 (5.9)	0.166
Sinus arrhythmia	3 (2.7)	2 (2.6)	1 (2.9)	1.000
Atrial fibrillation	1 (0.9)	0	1 (2.9)	0.304
Atrial ectopic rhythm	2 (1.8)	1 (1.3)	1 (2.9)	0.517
First-degree A-V block	3 (2.7)	0	3 (8.8)	–
Supraventricular tachycardia	1 (0.9)	1 (1.3)	0	–
Supraventricular extrasystole	1 (0.9)	0	1 (2.9)	–
Change of 1 sub-item of A	10 (8.9)	3 (3.8)	7 (20.6)	0.008
Change of 2 sub-items of A	2 (1.8)	1 (1.3)	1 (2.9)	0.517
Change of 3 or more sub-items of A	0	0	0	1.000
(B) <i>Chamber hypertrophy</i>	13 (11.6)	4 (5.1)	9 (26.5)	0.003
Left ventricular	12 (10.7)	4 (5.1)	8 (23.5)	0.007
Right ventricular	0	0	0	1.000
Left atrial	5 (4.5)	1 (1.3)	4 (11.8)	0.029
Right atrial	0	0	0	1.000
Change of 1 sub-item of B	9 (8.0)	3 (3.8)	6 (17.6)	0.022
Change of 2 sub-items of B	4 (3.6)	1 (1.3)	3 (8.8)	0.083
Change of 3 or more sub-items of B	0	0	0	1.000
(C) <i>Conduction disturbance</i>	6 (16.0)	1 (1.3)	5 (14.7)	0.010
ASDBB	5 (4.5)	1 (1.3)	4 (11.8)	0.029
RBBB	1 (0.9)	0	1 (2.9)	–
LBBB	1 (0.9)	0	1 (2.9)	–
Change of 1 sub-item of C	5 (4.5)	1 (1.3)	4 (11.8)	0.029
Change of 2 sub-items of C	1 (0.9)	0	1 (2.9)	1.000
Change of 3 or more sub-items of C	0	0	0	1.000
(D) <i>DVRC</i>	14 (12.5)	10 (12.8)	4 (11.8)	1.000

DVRC, diffuse ventricular repolarization changes; A-V, atrioventricular; ASDBB, anterosuperior divisional branch block; RBBB, right bundle branch block; LBBB, left bundle branch block; DM, dermatomyositis; PM, polymyositis.

in ECG were found in most patients with PM, when compared to patients with DM, especially for the higher frequency of left chamber hypertrophy, changes of rhythm and the presence of ASDBB in the first group.

So far, there are few studies evaluating electrocardiographic changes in patients with DM and PM; and, when present, these are limited to a small number of cases and/or in the absence of a control group,<sup>7,9,10,12,13</sup> differently of the present study. ECG changes in idiopathic inflammatory myopathies occur in 33–72%.<sup>3–5,7–13</sup>

Our data show that LVH was significantly present in patients with DM and PM compared to the control group. Furthermore, an additional analysis showed the persistence of left-chamber hypertrophy in patients with PM, when compared to patients with DM. This finding could be explained by the high prevalence of systemic hypertension, commonly found in patients with DM and PM,<sup>16,17</sup> although this finding was similar to the prevalence in the control group. It is necessary, however, to evaluate the duration and degree of hypertension in these individuals, as well as their blood pressure control. And the pathogenesis of DM and PM itself also may have contributed to left chamber hypertrophy. There are indications of presence of infiltration of mononuclear inflammatory cells in the endomysium and in perivascular areas

of infarction, as well as degeneration of cardiac myocytes and areas of myocardial fibrosis.<sup>12,13,18,19</sup> This inflammatory process in the myocardium can lead to remodeling, with anatomical and functional changes of the myocardium and, ultimately, possible left ventricular dysfunction and restrictive cardiomyopathy and heart failure. Thus, ECG changes may be an early finding of these complications.

Corroborating our data, Sharratt et al.<sup>10</sup> showed that 5 of 13 patients with PM had cardiac abnormalities seen by clinical examination, ECG and/or chest radiography. Of these five, four patients had abnormalities of left ventricular dysfunction, diagnosed by ECG. On the other hand, Gonzalez-Lopez et al.<sup>4</sup> observed in a case series of 32 patients the presence of diastolic dysfunction of left ventricle in 42% of cases. Schwartz et al.<sup>11</sup> compared 59 patients with juvenile DM with 59 healthy subjects and observed that only their patients had subclinical diastolic (left ventricular) dysfunction, showing that such change is inherent to the disease (juvenile DM).

Regarding the changes of rhythm and conduction disorders, there was no significant difference between patients and the control group analyzed in this study. However, when comparing patients with DM and patients with PM, there was a higher frequency of disturbances of rhythm and conduction, especially ASDBB, in patients with PM. Previous studies



with ECG and Holter monitoring in patients with DM and PM showed several abnormalities in this population: atrial and ventricular extrasystoles, atrial tachycardia, ventricular tachycardia, atrial fibrillation, atrioventricular conduction block, bundle branch blocks, abnormal Q waves and nonspecific ST-T segment changes.<sup>3,4,6,7</sup>

In studies with autopsy, histological changes were found in the conduction system, with the presence of lymphocyte infiltration and sinoatrial node fibrosis,<sup>18,19</sup> which may explain the presence of conduction findings in patients with idiopathic inflammatory myopathies.

Our study has limitations, by not presenting a structural cardiac evaluation nor an assessment for myocardium inflammation. However, as the cardiac involvement is a factor of worse prognosis for patients with myopathies,<sup>20</sup> the ECG may serve as a noninvasive, cheap and practical tool for an initial assessment and in tracking this problem. Another limiting factor is the failure to establish a correlation of ECG findings, particularly with disease activity (DM/PM). Furthermore, we must consider that the lack of electrocardiographic differences between patients and controls may also be due to a selection bias, since the control group came from the preoperative (and clinical) sector, with a frequency of comorbidities similar to that of our patients and perhaps with a higher frequency of electrocardiographic changes versus healthy subjects. However, we believe that this bias is not significant, due to the fact that most ECGs in control group were performed on patients involved in preoperative non-cardiac/vascular surgery.

In summary, we observed a higher frequency of LVH in patients with inflammatory myopathies. Furthermore, there was a distinction of the ECG findings between patients with DM and PM, with higher prevalence of left-chamber hypertrophy and rhythm and conduction disturbances in patients with PM compared to DM.

More studies are needed to determine the correlation of ECG findings with other complementary tests, clinical manifestations, myopathy activity and evolution to other cardiac diseases.

### Conflicts of interest

The authors declare no conflicts of interest.

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