Cardiac dysfunction in juvenile dermatomyositis: a case-control study

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ABSTRACT

Objective To compare cardiac function in patients with juvenile dermatomyositis (JDM) with matched controls, and examine associations between pathological electrocardiography (ECG), echocardiographic abnormalities and disease variables in patients with JDM.

Methods A total of 59 patients with JDM, examined a median 16.8 years (range 2–38 years) after disease onset, were compared with 59 age-matched and sex-matched controls. Echocardiography, including early diastolic transmitral flow/early diastolic tissue velocity (E/E’) as a marker for diastolic dysfunction, and 12-channel ECG were performed and analysed blinded to patient information. Disease activity and damage were assessed by clinical examination at follow-up and chart review.

Results E/E’ was elevated (>9.5) in 13 (22%) patients versus 0 controls (p=0.001). In all, 10 patients presented with pathological ECG compared to 4 controls (p=0.054). Previous or current hypertension was found in 12 patients versus 0 controls (p=0.001). Among the patients, pathological ECG was found in 6/13 patients with versus 4/44 without elevated E/E’ (p=0.002); and systolic blood pressure was correspondingly 132±24 mm Hg versus 112±18 mm Hg in the groups (p=0.012). E/E’ correlated with cumulative organ damage assessed at follow-up (r=0.41, p=0.001) and disease activity at 1 year (r=0.56, p<0.001), which also predicted pathological E/E’ after controlling for age and gender. During disease course, 12% of patients with JDM developed pericarditis.

Conclusion Only patients with JDM and no controls had subclinical left ventricular diastolic dysfunction; the patients with elevated E/E’ also had high prevalence of pathological ECG and hypertension. High disease activity 1-year post diagnosis predicted high E/E’ at follow-up. The findings suggest that subclinical heart disease is related to the systemic nature of JDM.

INTRODUCTION

Cardiac affection is well documented in inflammatory autoimmune diseases such as rheumatoid arthritis and systemic lupus erythematosus.1–4 In adult polymyositis (PM) and dermatomyositis (DM), cardiac manifestations have been reported, but considered mainly subclinical.5 Studies on cardiac involvement in PM have shown pathological electrocardiography (ECG) in 33% to 72%.6–9 In 2 autopsy series from 1979 and 1982, myocarditis was found in 6 of 20 and 4 of 16 patients with PM.8,10 Two children were included in each autopsy study; none of these had cardiac disease.

Like adult PM/DM, juvenile dermatomyositis (JDM) is a disease of inflammation and vasculopathy.11 It is possible that the myocardium in patients with JDM can be affected by inflammation, as seen in the adult form. This may influence particularly left ventricular diastolic function, as observed in other rheumatic diseases.12–15 In a study on JDM from 1980, pathological ECG was present in 10 of 21 patients.16 Thus, it seems likely that the myocardium in patients with JDM can be affected by inflammation, just as the skeletal muscle. The knowledge on cardiac manifestations in adult myositis and JDM has been scarce, only based on few studies, case reports14,16 and clinical experience. Recently, two studies on JDM have suggested that cardiovascular complications are rare.17,18 However, no systematic, controlled study has been carried out.

In the present study we compared cardiac function in a JDM cohort with that in matched controls from the general population. The primary aims were to assess left ventricular diastolic function using early diastolic transmitral flow/early diastolic tissue velocity (E/E’),19 and to investigate for abnormalities in ECG. Secondary aims were to determine whether diastolic function and/or pathological ECG correlated with patient characteristics and disease variables.

PATIENTS AND METHODS

Patients and controls

The inclusion criteria were a probable or definitive diagnosis of DM according to the Bohan and Peter criteria for DM,20 disease onset before 18 years, minimum 24 months of disease duration and age ≥6 years at inclusion. By manual and electronic search in the in the medical records, we identified a retrospective inception cohort of patients with JDM diagnosed between January 1970 and June 2006 in Norway, as previously described in detail.21–23 A total of 66 patients fulfilled the inclusion criteria; 4 were deceased. The remaining 62 patients could all be tracked through the Norwegian population register and 59 (95%) participated in the study. Oslo University Hospital, Rikshospitalet (OUS) is the major referral centre for patients with JDM in Norway; hence our study population probably represents most of the patients with JDM in the country from this period.

One sex-matched and age-matched control per patient was randomly drawn from the National Population Register. Responders with lung or heart diseases (one with atrial fibrillation), was excluded, except for mild asthma and hypertension.

We obtained informed consent from all patients and controls (and their parents if aged <16 years), according to the Declaration of Helsinki. The study was approved by the Regional Ethics Committee for Medical Research.
**Data collection and clinical measures**

Clinical examination of all patients and matched controls was performed by a single doctor (HS) at OUS in the period September 2005 to May 2009. ECG and echocardiography were performed. In patients with JDM, blood samples included anti-nuclear antibodies (ANAs), myositis specific or associated antibodies, muscle enzymes and cardiac markers (pro-B-type natriuretic peptide (BNP) and troponin-T in 37 patients) were analysed. Disease activity was measured by disease activity score (DAS) for JDM,24 (range 0–20), 0 means no activity, which consists of DAS skin (0–9) and DAS muscle (0–11). Cumulative organ damage was measured by the Myositis Damage Index (MDI, range 0–35/40),25 as previously described;21 DAS and MDI were also scored retrospectively from the first-year post diagnosis based on chart review.21 The Health Assessment Questionnaire (HAQ)26 and the Child HAQ27 were used to measure physical function in patients aged ≥18 years (n=39) and <18 years (n=20), respectively. Physical health was measured by the Short Form 36 (SF-36) physical component summary (PCS).28 Disease onset was defined as time of the first muscle or skin symptom clearly related to JDM (found by chart review), and disease duration as the time from disease onset to the follow-up examination. Smoking habits (current and previous) and medication were obtained from the study cases and medical records.

**ECG and echocardiography**

A 12-channel ECG was performed on 58 patients and 57 controls. Rhythm and ST segment were assessed, and PR, corrected QT interval (QTc) and QRS duration were measured. Left ventricular hypertrophy was assessed by Cornell voltage × QRS duration product (ECG parameter indicating left ventricular (LV) hypertrophy)29; however, only validated in the adult population, it was not calculated in those <16 years. ECG was classified, blinded to clinical information and patient/control identity, as normal, borderline or pathological. Criteria for borderline ECG were: incomplete right bundle branch block, severe sinusarhythmia, STT changes or Sokalow criterion >35 mm. Standard criteria for pathological ECG were used.30

Two-dimensional, M-mode and Doppler echocardiography were performed for all patients and controls using a Vivid 7 ultrasound scanner (GE Vingmed Ultrasound, Horten, Norway).31 32 A minimum of three cycles were recorded, analysed and averaged. Data analysis was performed blinded to clinical information and to patient/control identity (TS). Colour coded Tissue Doppler was performed in 58 patients and 57 controls, with a frame rate of approximately 180. We used sample volumes from the mitral annulus, lateral and septal position in the four-chamber view and two corresponding positions in the two-chamber view. E and E’ were recorded, and E/E’ ratio, the most commonly used parameter reflecting left ventricular diastolic function, was calculated.19 Possible diastolic dysfunction was defined as E/E’ > mean+2SD (>9.5) of the matched control values, which corresponds to suggested cut-off values for possible diastolic dysfunction obtained by colour coded tissue Doppler.33 Valvular regurgitation less than grade 1 was considered insignificant.

**Statistical analysis**

Differences between patients and matched controls were tested by the paired sample t test (normally and continuous distributed variables), Wilcoxon’s rank sum test (continuous not normally distributed or ordinal variables) or McNemar’s test (binary data). Differences between patient groups were tested by the independent sample t test, Mann–Whitney U test or χ² test as appropriate. Correlations were determined by the Spearman correlation coefficient (rₛ). Two-tailed tests were used for all calculations except for ECG analyses between patients and controls, where a one-tailed significance test was used. It was considered unlikely that patients with JDM should have higher incidence of normal ECG than the controls since earlier studies have shown high incidences of abnormal ECG in JDM.6–9 The association between E/E’ at follow-up and DAS 1-year post diagnosis was controlled for age and gender in a linear regression model. SPSS V.16.0 (SPSS, Chicago, Illinois, USA) was used for statistical analyses.

**RESULTS**

**Characteristics and clinical cardiac assessment**

Baseline characteristics in patients and controls are shown in table 1. One JDM patient had coronary artery disease (woman, 54 years at follow-up). There were no known cases of diabetes, heart failure or cardiac arrhythmias in neither the patient nor the control group.

Seven patients had pericarditis during disease course; of these, five were within 1–2 years, one after 3 years and one 6 years after JDM diagnosis. A girl diagnosed as having JDM at age 3 years had high initial disease activity; after 3 years she became critically sick from pericarditis and later developed hypertension. Another girl with pericarditis at the age of 14 years presented with hypertension (185/80 mm Hg) at follow-up visit 27 years later. None of the controls had a history of pericarditis.

Seven patients had been diagnosed as having hypertension prior to follow-up, whereas none of the controls were (p=0.007). A total of 12 patients had a history of hypertension and/or systolic blood pressure (BP) >140 mm Hg (145–185 mm Hg) at follow-up; none of the controls did (p<0.001). Of these, only

| Table 1 Characteristics and cardiac symptoms in patients with juvenile dermatomyositis (JDM) and controls |
|--------------------------------------------------|---------------------|---------------------|
| Characteristics                                    | Patients with JDM | Controls* |
| Female sex                                        | 36 (61)            | 36 (61)            |
| Age at disease onset, years                        | 7.8 (1.4–17.3)     | NA                 |
| Age at diagnosis, years                            | 8.9 (2.1–19.2)     | NA                 |
| Variables assessed at 16.8-year follow-up          |                     |                     |
| Duration from disease onset, years                 | 16.8 (2.0–38.1)    | NA                 |
| Age, years                                        | 21.5 (6.7–55.4)    | 21.6 (6.2–55.4)    |
| Height, cm                                        | 165.5 (15.0)       | 167.5 (15.8)       |
| Weight, kg                                        | 62.8 (20.1)        | 64.9 (19.9)        |
| BP systolic, mm Hg                                | 117 (22.2)         | 111 (13.6)         |
| BP diastolic, mm Hg                               | 70 (13.3)          | 67 (9.3)           |
| HR, beats/min                                     | 69 (13.9)          | 63 (11.1)          |
| Body mass index, kg/m²                             | 22.3 (4.8)         | 22.5 (4.5)         |
| Smokers, daily                                    | 11 (23)            | 8 (17)             |
| Smokers, weekly                                   | 15 (31)            | 17 (35)            |
| Smokers, previously daily                         | 3 (6)              | 5 (10)             |
| Dyspnoea on exertion                              | 7 (12)             | 0\*                |
| Antihypertensive treatment                        |                     |                     |
| ACE inhibitor                                     | 2                   |                     |
| Calcium blocker                                   | 1                   |                     |
| β Blocker                                         | 1                   |                     |

Values are number (%), median (range) or mean (SD) unless otherwise stated; n=59.

*Sex-matched and age-matched controls from the general population.

†p=0.017.

‡Patients and controls ≥14 years at follow-up, n=48.

\*p<0.016.

ACE, angiotensin-converting enzyme; BP, blood pressure; HR, heart rate from electrocardiography (ECG); NA, not applicable.
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four patients received antihypertensive treatment at follow-up (table 1).

Pro-BNP, a marker of heart failure, was normal in the patients (4.45±3.99 pmol/litre; range 0–10 for men, 0–20 for women). Troponin T, a marker of myocardial disease, was not detectable by standard methods in a subset of 37 patients.

**ECG in patients with JDM and controls**

ECG was pathological in 10/58 (17%) patients (3 poor R-wave progression, 2 left ventricular hypertrophy signs, 2 right bundle branch block, 1 pathological Q-wave, 1 P pulmonale and 1 prolonged QTc) and borderline in 13 patients. Among the controls, 4/57 (7%) had a pathological ECG (2 ST-segment changes, 1 right bundle branch block and 1 left ventricular hypertrophy signs) and 10 borderline. Pathological/borderline ECG was more abundant in patients than controls (p=0.02, table 2), whereas pathological ECG was borderline significant (p=0.05). The Cornell product reflecting left ventricular hypertrophy was higher in patients than controls.

**Echocardiographic findings in patients with JDM and controls**

Possible diastolic dysfunction, defined as E/E’ >9.5, was found in 13/58 (22%) patients versus 0 controls (p=0.001). At the group level, E/E’ was 13.4% higher in patients with JDM than controls (p=0.007) (table 3 and figure 1A). Measured by conventional Doppler, patients had a reduced cardiac output and stroke volume compared to controls (table 3). No significant stenosis was found in any of the valves in neither patients nor controls, and the velocities did not differ significantly between the groups (data not shown). No patients or controls had evidence of elevated pulmonary pressure after assessing tricuspid and pulmonary velocities.

Standard two-dimensional and M-mode echocardiography showed no significant differences between patients and controls in left ventricular ejection fraction (LVEF), wall thickness and internal diameter of LV, or size of the atrias (table 3). All patients (55) and controls (52) with recordings allowing measurement of LVEF, had normal values (>50%).

**Associations between cardiac parameters, patient characteristics and disease variables assessed at follow-up**

Pathological ECG was found in 6/13 (46%) of patients with possible diastolic dysfunction (E/E’ >9.5) versus 4/44 (9%) without diastolic dysfunction (p=0.002). No correlation was present between E/E’ and Cornell voltage × QRS duration product, QTc interval, QRS or PR duration (data not shown). However, systolic BP was higher in patients than those with normal E/E’; 132±24 mm Hg versus 112±18 mm Hg (p=0.012).

**Table 1 12-channel electrocardiography (ECG) in patients with juvenile dermatomyositis (JDM) (median 16.8 years after onset) and controls**

| Parameters                  | Patients with JDM | Controls | p Value |
|-----------------------------|-------------------|----------|---------|
| Pathological ECG            | 10 (17)           | 4 (7)    | 0.054*  |
| Pathological/borderline ECG | 23 (39)           | 14 (25)  | 0.020*  |
| PR, ms                      | 139 (19)          | 146 (22) | 0.082   |
| QRS, ms                     | 91 (11)           | 90 (12)  | 0.395   |
| QTc, ms                     | 405 (24)          | 407 (23) | 0.631   |
| Cornell, mm × ms1           | 1451 (550)        | 1157 (478) | 0.002  |

Values are number (%) or mean (SD); n=56 pairs (three missing due to technical problems).

Early predictors for cardiac outcome

E/E’ at follow-up correlated with DAS skin, DAS muscle and DAS total 1 year after diagnosis (all ps<0.001) (table 5 and figure 1B). E/E’ also correlated mildly with DAS total 6 months (data not shown), although weaker than with DAS total 1 year. Pathological ECG correlated with DAS skin 1 year. Neither E/E’ or pathological ECG correlated with MDI 1-year post diagnosis.

**DISCUSSION**

In our study, we found E/E’ elevated in patients with JDM compared to controls; E/E’ values suggestive of diastolic dysfunction was only present in patients with JDM. Furthermore, high E/E’ median 16.8 years after disease onset, was predicted by DAS 1-year post diagnosis. Pathological ECG was prevalent among patients with high E/E’. Patients also had increased risk of

**Table 3 Echocardiography at follow-up in patients with juvenile dermatomyositis (JDM) and controls**

| Two-dimensional echocardiography | Patients with JDM | Controls | p Value |
|----------------------------------|-------------------|----------|---------|
| LA area, cm²                     | 59                | 15.5 (3.4) | 16.5 (3.7) | 0.095 |
| RA area, cm²                     | 59                | 13.3 (3.1) | 14.2 (3.5) | 0.086 |
| LV ejection fraction, %          | 47                | 63.2 (5.1) | 62.5 (5.5) | 0.487 |
| LV diastolic volume, cm³         | 59                | 49.0 (25.4) | 97.5 (25.7) | 0.029 |
| LV systolic volume, cm³          | 47                | 33.8 (11.6) | 37.1 (12.5) | 0.051 |

**M-mode echocardiography**

| Pathological Doppler | Patients with JDM | Controls | p Value |
|----------------------|-------------------|----------|---------|
| E velocity, m/s      | 59                | 0.86 (0.17) | 0.86 (0.15) | 0.858 |
| A velocity, m/s      | 59                | 0.49 (0.11) | 0.47 (0.10) | 0.132 |
| A/E ratio            | 59                | 1.81 (0.50) | 1.95 (0.54) | 0.116 |
| MV deceleration time, ms | 59             | 193 (42) | 200 (42) | 0.278 |
| LVOT stroke volume, ml | 56               | 21.4 (16.8) | 17.9 (18.9) | 0.008 |
| LVOT cardiac output, litres/min | 55        | 4.1 (1.22) | 4.5 (1.20) | 0.021 |

**Valvular heart disease**

Valvar heart disease

Mitral regurgitation

Aortic regurgitation

**Tissue Doppler**

E’ velocity, cm/s

E’/E ratio

Values are mean (SD); n=47 is due to difficulties with data acquisition when measuring ejection fraction. n=55 and 56 is due to missing Doppler data in 3–4 of the pairs.

*Number (%) with ≥ grade 1 regurgitation.

A, late diastolic transmitral flow; E, early diastolic transmitral flow; E’, early diastolic tissue velocity; LA, left atrium; LV, left ventricle; LVOT, left ventricular outflow tract; MV, mitral valve; n, number of pairs; PWV, posterior wall; RA, right atrium.
Pathological ECG was more prevalent in patients with possible diastolic dysfunction than in those without (46% vs 9%). The two cardiac parameters might address the same aspects of cardiac remodelling; E/E’ reflecting inflammation and fibrosis in the developing pericarditis during disease course. To our knowledge, this is the first controlled study to examine cardiac function in an unselected JDM cohort.

The representativeness of our inception cohort has previously been described in detail.21–23 The frequency of dyspnoea in the present study is similar to previous reports.11 Apart from the patient with coronary artery disease, none had clinically cardiac disease which is in accordance with previous reports.17 A strength of our study is the random selection of age and sex matched controls from the National Population Register. Height, weight and smoking habits did not differ between the groups.

A primary aim of the study was to assess diastolic function of patients with JDM versus controls; we found E/E’ to be higher in patients. We used a cut-off value derived from the controls (> mean+2SD, E/E’ >9.5); 13 patients were above this limit and could be classified with possible diastolic dysfunction, whereas none of the controls. This suggests that JDM induces myocardial remodelling; resulting in slower left ventricular relaxation. Although cardiac biopsy studies are unavailable for patients with JDM, autopsy series of adult myositis have shown inflammatory changes in the myocardium; perivascular mononuclear cell infiltrates with or without small vessel disease and replacement fibrosis. We suggest that a similar pathophysiological process in JDM can explain the observed increase in left ventricular stiffness and the impaired relaxation with raised E/E’.34–36

Pathological ECG was more abundant in patients than in controls, albeit only borderline significant values. Our study may have been underpowered due to too few patients to obtain significant differences. As for the combined endpoint pathological/borderline ECG, this was present in more patients than controls. In the literature, a high prevalence of abnormal ECG is often reported (35% to 72%) in uncontrolled studies of PM/DM,6–8 and this is in accordance with our combined endpoint. However, we only found pathological ECG in 17% of our patients. This discrepancy might be explained by our strict criteria for pathological ECG, but our cohort might also be more representative for the JDM population than earlier studies.

Table 4 Correlations between cardiac outcome and disease variables at follow-up

| E/E’ | Pathological ECG |
|------|-----------------|
|      | r<sub>sp</sub> | p Value | r<sub>sp</sub> | p Value |
| BP systolic | 0.40 | 0.002 | −0.03 | 0.819 |
| Male sex | 0.18 | 0.176 | NA | NA |
| Age | 0.23 | 0.080 | 0.22 | 0.092 |
| Disease duration | 0.29 | 0.025 | 0.27 | 0.039 |
| DAS muscle | 0.42 | 0.001 | 0.00 | 1.00 |
| DAS skin | 0.01 | 0.570 | 0.22 | 0.099 |
| DAS total | 0.27 | 0.042 | 0.12 | 0.352 |
| MDI | 0.41 | 0.001 | 0.19 | 0.160 |
| SF-36 PCS | −0.34 | 0.017 | 0.17 | 0.243 |
| CHAQ/HAQ | 0.26 | 0.047 | 0.14 | 0.292 |
| Pericarditis | −0.04 | 0.786 | NA | NA |
| Prednisolone | 0.29 | 0.030 | 0.00 | 0.904 |

Table 5 Correlations between cardiac outcome at follow-up and disease variables at 1 year

| E/E’ | Pathological ECG |
|------|-----------------|
|      | r<sub>sp</sub> | p Value | r<sub>sp</sub> | p Value |
| DAS muscle | 0.41 | 0.001 | 0.12 | 0.371 |
| DAS skin | 0.55 | <0.001 | 0.26 | 0.047 |
| DAS total | 0.56 | <0.001 | 0.24 | 0.066 |
| MDI | 0.22 | 0.09 | 0.18 | 0.169 |

BP, blood pressure; CHAQ, Child Health Assessment Questionnaire; DAS, disease activity score; E, early diastolic transmitral flow; E’, early diastolic tissue velocity; ECG, electrocardiography; HAQ, Health Assessment Questionnaire; MDI, Myositis Damage Index; Prednisolone, cumulative prednisolone dose during disease course; r<sub>sp</sub>, Spearman correlation coefficient; SF-36 PCS, Short Form 36 physical component summary.

Figure 1 Early diastolic transmitral flow/early diastolic tissue velocity (E/E’) in patients with juvenile dermatomyositis (JDM) compared to controls and correlation with disease activity score (DAS) total 1 year. A. E/E’ in patients compared to controls, values are median with CIs. B. E/E’ in patients correlates with DAS total 1 year, r<sub>sp</sub> = 0.56, p<0.001.
myocardium, and pathological ECG the damage this process does to the conduction system. We also compared the composite pathological/borderline ECG outcome with E/E', but found no association. This might suggest that ECG only gives clinically relevant information in patients with JDM if strict criteria for abnormal ECG are used.

Although as many as 22% of our patients had possible diastolic dysfunction, only one had known cardiac disease. Even though 12% of our patients reported pathological dyspnoea on exertion at follow-up, this was not associated with E/E' >9.5 or pathological ECG. Also, we have previously reported that dyspnoea was associated with restricted pulmonary function, which therefore is a more likely explanation for the dyspnoea.

Therefore, the clinical implications of increased E/E' and ECG abnormalities in patients with JDM remains uncertain. However, the correlation between E/E' and cumulative organ damage and patient-reported health status at follow-up, indicate a clinical relevance of our findings. The correlations between disease duration and E/E' and pathological ECG suggest a gradually developing process of cardiac remodelling. Most of our patients are young and might therefore face an increased risk of cardiac disease later in life. To assess this, follow-up studies are needed.

The patients had higher Cornell voltage x QRS duration product compared to controls, suggesting cardiac hypertrophy. On M-mode echocardiography, however, no difference in left ventricular wall thickness was found between the groups. This could suggest subtle differences in left ventricular mass between patients and controls, although this was not detected by echocardiography.

Hypertension is recognised as an important long-term cardiovascular complication in myositis, and it is included as an item in MDI. We found that 20% of the patients had systolic BP >140 mm Hg and/or a history of hypertension, whereas none of the controls. Also, we found a correlation between E/E' and systolic BP, and systolic BP was higher in patients with E/E' >9.5 than those <9.5. Other studies with shorter follow-up time have found a lower prevalence of hypertension, but we do not know when in disease course hypertension develops. There has been no report of renal impairment or activation of the renin–angiotensin system in JDM. The mechanism for hypertension might therefore well be the generalised microvasculopathy seen in the disease, which also is closely related to high early disease activity.

The strong correlation between early disease activity (1-year DAS total, muscle and skin) and E/E' is a main finding in our study. This correlation supports that myocarditis is the primary process inducing diastolic dysfunction. Previously we have also identified early disease activity as predictor of cumulative organ damage, muscle weakness and muscle damage (by MRI) at follow-up in the same cohort. This corresponds to the finding in the present study, where high E/E' was predicted by 1 year DAS; thus the disease process in skeletal muscle might be associated with the disease process in the myocardium. However, unfavourable pulmonary outcome was not predicted by early disease activity, and thus the pathological mechanisms in JDM might be organ specific. However, we are aware of the limitations in our study due to retrospective assessment of early disease activity.

From a clinical perspective, the prevalence of pericarditis in our patients (12%) is high. Recently Rider et al. found that pericarditis was a predictor for muscle damage in 96 patients with adult myositis; however, they did not report any cases of pericarditis among the 154 patients with JDM. Sallum et al. however, found eight cases of pericarditis and/or myocarditis in a cohort of 54 patients. In our material, we found no association between pericarditis and pathological ECG or E/E'. This is not surprising, since it is well known that the myocardium often remains unaffected even in acute pericarditis. In the present study, the retrospective assessment of pericarditis is a limitation.

In conclusion, patients with JDM had higher E/E' and higher prevalence of pathological/borderline ECG than controls. Although the patients did not report symptoms of cardiac disease, E/E' correlated with early disease activity, and was predicted by 1 year DAS. Patients with high E/E' also had increased prevalence of pathological ECG. Although the cardiac dysfunction was subclinical, these patients might have increased risk of developing clinical cardiac disease later in life; and thus might benefit from treatment aimed at reducing myocardial remodelling.

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Competing interests None.

Ethics approval This study was carried out in compliance with the declaration of Helsinki, and was approved by the Regional Ethics Committee (Helse Sør-Ost).

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