Comparison of clinical features and drug therapies among European and Latin American patients with juvenile dermatomyositis

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Abstract

Objective

To compare the demographic features, presenting manifestations, diagnostic investigations, disease course, and drug therapies of children with juvenile dermatomyositis (JDM) followed in Europe and Latin America.

Methods

Patients were inception cohorts seen between 1980 and 2004 in 27 paediatric rheumatology centres. The following information was collected through the review of patient charts: sex; age at disease onset; date of disease onset and diagnosis; onset type; presenting clinical features; diagnostic investigations; course type; and medications received during disease course.

Results

Four hundred and ninety patients (65.5% females, mean onset age 7.0 years, mean disease duration 7.7 years) were included. Disease presentation was acute or insidious in 57.1% and 42.9% of the patients, respectively. The course type was monophasic in 41.3% of patients and chronic polycyclic or continuous in 58.6% of patients. The more common presenting manifestations were muscle weakness (84.9%), Gottron's papules (72.9%), heliotrope rash (62%), and malar rash (56.7%). Overall, the demographic and clinical features of the 2 continental cohorts were comparable. European patients received more frequently high-dose intravenous methylprednisolone, cyclosporine, cyclophosphamide, and azathioprine, while methotrexate and antimalarials medications were used more commonly by Latin American physicians.

Conclusion

The demographic and clinical characteristics of JDM are similar in European and Latin American patients. We found, however, several differences in the use of medications between European and Latin American paediatric rheumatologists.

Key words

juvenile dermatomyositis, onset manifestations, clinical features, disease course, drug therapy

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Introduction

Juvenile dermatomyositis (JDM) is a multisystem vasculopathic disease of presumed autoimmune etiology that involves primarily the skin and muscles. Other organs may be affected, including the gastrointestinal tract, heart and lungs, and, more rarely, the kidneys, eyes, and central nervous system (1-3). The onset of JDM is quite variable, with some patients experiencing the insidious development of progressive muscle weakness and skin rash, and others having a more acute onset with fever, profound muscle weakness, widespread cutaneous manifestations, and occasionally ulcerative lesions. The disease course is also heterogeneous. It may range from a monophasic course, with good response to treatment and full recovery within 2 years after diagnosis without relapse, to a chronic polycyclic or continuous course, with relapsing-remitting or persistently active disease for longer than 2 years after diagnosis and a significant risk of development of disease- or treatment-related complications (1).

Prior to the introduction of corticosteroids in the 1960s for treatment of the disease, almost one-third of patients with JDM died, one-third were left with permanent disabilities, and only one-third recovered without complications (4). Since then, the mortality has decreased to less than 2%, and there has been a considerable improvement in functional outcome. However, many patients are refractory or respond suboptimally to current treatments and are at risk of developing irreversible damage from the disease activity or its treatment (5-7). This morbidity may have a serious impact on the quality of life of patients and their family.

Recent improvement in patient outcomes is largely due to the refinement in protocols of corticosteroid administration, including the use of high-dose intravenous methylprednisolone pulses, and to the introduction of second-line medications, such as methotrexate, cyclosporine, and intravenous immunoglobulin (8-10). However, there have been no randomised controlled trials of any medications in children with JDM. Consequently, disease management remains largely empirical and based on observational studies and clinical experience.

A number of studies have described the clinical features and treatment modalities of series of patients with JDM (1, 3, 11, 12, 13). However, most studies come from single centres or are small. We recently collected a large sample of children with JDM in the context a multinational, multicenter study, whose primary aims were to investigate the long-term outcome of the disease and to search for prognostic factors. This study provided the opportunity to compare patient populations followed in paediatric rheumatology centres in different continents, namely Europe and Latin America. The results of outcome analysis have been reported previously (7). In this paper, we present the data regarding demographic features, presenting manifestations, diagnostic investigations, disease course, and use of drug therapies.

Patients and methods

Study design and patient selection.

The study protocol was described in detail elsewhere (7). Briefly, investigators in each participating centre were first asked to identify all patients seen between January 1980 and December 2004 who had a diagnosis of JDM by Bohan and Peter's criteria (14, 15), were age <18 years at disease onset, and had at least 24 months of disease (*i.e.* follow-up) duration between disease onset and the time of last follow-up observation. Next, each investigator was asked to collect retrospective data and to assess cumulative damage through the review of clinical data from disease onset to last follow-up visit or, if the patient had died, to the last visit before death. Investigators were also asked to make the cross-sectional assessment of all patients who were still followed or were no longer followed and were alive. Informed consent to participate in the study was provided by both the parent/guardian and the patient (when applicable). Ethics committee approval of the study was obtained in all participating countries. Outcome data were collected between 2003 and 2006.

Retrospective assessment

The following information was collected through the review of patient charts: sex; age at disease onset; date of disease onset and diagnosis (as recorded by the attending physician); onset type (acute: with high fever, prostration, prominent rash or profound muscle weakness, or insidious: progressive development of muscle weakness and rash); presenting clinical features (clinical manifestations observed in the first month of illness); diagnostic investigations (electromyography and muscle biopsy); course type, not including being off medications (monocyclic: full recovery within 2 years after diagnosis without relapse, chronic polycyclic: relapsing-remitting disease, or chronic continuous: persistently active disease for longer than 2 years after diagnosis) (16-18); and medications received during the disease course.

Damage assessment and cross-sectional evaluation

The methodology used for these assessments and the results obtained were reported elsewhere (7). Briefly, cumulative damage was assessed with the Myositis Damage Index (MDI) (19). This tool assesses the extent of damage in the muscle, skeletal, cutaneous, gastrointestinal, pulmonary, cardiac, peripheral vascular, endocrine, ocular, infectious, malignancy, and other organ/systems. The following clinical assessments were performed at cross-sectional visit: muscle strength and function/endurance through the 8-muscle Kendall Manual Muscle Testing (MMT) (20) and the Childhood Myositis Assessment Scale (CMAS) (21), respectively; overall disease activity through the Disease Activity Score (DAS) (22) and the Myositis Disease Activity Assessment VAS (MYOACT) (13); physical function through the Childhood Health Assessment Questionnaire (CHAQ) (23, 24); HRQL through the parent version of the Child Health Questionnaire (CHQ) (24, 25); satisfaction with illness outcome (very satisfied, moderately satisfied or not satisfied).

Statistics

Comparison of features between European and Latin American patients was made by means of the Mann-Whitney U-test in case of continuous variables and of chi-square or the Fisher exact test, as appropriate, in case of categorical data. Bonferroni adjustment was applied as a correction for multiple comparisons to explore post-hoc differences between pairs of patients groups.

The separate (univariate) and joint (multivariate) effects of predictor variables on long-term outcomes were examined. Predictor variables were sex, continent, ethnicity, and year of onset, onset age, onset type, onset manifestations, course type, disease duration and duration of active disease. Outcomes were muscle strength/endurance, continued disease activity, cumulative damage, muscle damage, cutaneous damage, calcinosis, lipodystrophy, physical function, and HRQL. Bivariate analyses were first made for each outcome. Then, multiple logistic regression analyses were carried out entering predictor variables as explanatory variables and each disease outcome as outcome variable. Cases with missing data were excluded. Variables that were significantly associated with the outcome in bivariate analyses were entered in multivariate procedures. Using a backward selection procedure, predictor variables that were significantly associated with the outcome were identified. The effect was expressed in terms of the odd ra-

Table I. Main demographic and clinical features and diagnostic procedures features in 490 children with juvenile dermatomyositis.

	All patients (n=490)		Europe (n=248)		Latin America (n=242)		*p*-value*
Females, no. (%)	490	321 (65.5)	248	169 (68.1)	242	152 (62.8)	0.21
Mean (SD) age at disease onset, years	486	7.0 (3.7)	247	6.9 (3.7)	239	7.0 (3.8)	0.68§
Age at disease onset, no. (%)	486		247		239		0.99
≤ 5 years		172 (35.4)		88 (35.6)		84 (35.1)	
5-10 years		211 (43.4)		107 (43.3)		104 (43.5)	
10-18 years		103 (21.2)		52 (21.1)		51 (21.3)	
Year of disease onset, no. (%)	487		248		239		0.80
1980-1990		98 (20.1)		47 (19.0)		51 (21.3)	
1991-2000		280 (57.5)		145 (58.5)		135 (56.5)	
2001-2004		109 (22.4)		56 (22.6)		53 (22.2)	
Mean (SD) time lag between disease onset and diagnosis, years	474	0.6 (1.0)	241	0.6 (1.1)	233	0.6 (0.9)	0.02§
Onset type, no. (%)	475		242		233		0.10
Acute		271 (57.1)		147 (60.7)		124 (53.3)	
Insidious		204 (42.9)		95 (39.3)		109 (46.8)	
Patients who underwent electromyography, no. (%)	479	287 (59.9)	244	153 (62.7)	235	134 (57.0)	0.20
Patients with abnormal findings		264 (55.1)		141 (57.8)		123 (52.3)	0.90
Patients with normal findings		23 (4.8)		12 (4.9)		11 (4.7)	
Patients who underwent muscle biopsy, no (%)	469	259 (55.2)	244	136 (55.7)	225	123 (54.7)	0.82
Patients who underwent needle biopsy		63 (13.4)		38 (15.6)		25 (11.1)	0.15
Patients who underwent surgical biopsy		196 (41.8)		98 (40.2)		98 (43.6)	
Course type, no. (%)	479		247		232		0.34
Monophasic		198 (41.3)		97 (39.3)		101 (43.5)	
Chronic polycyclic or continuous		281 (58.6)		150 (60.8)		131 (56.5)	

*Europe versus Latin America. SD: standard deviation.
P-values refer to the chi-square test unless otherwise specified. §Mann-Whitney U-test.

tio and 95% confidence intervals were calculated; statistical significance was tested by means of the likelihood-ratio test (LR test).

All statistical tests were two sided; a *p* value of less than 0.05 was considered as being statistically significant. The statistical packages used were the "Statistica" (StatSoft Corp., Tulsa, OK) and the "Stata release 7" (Stata Corporation, Texas, USA).

Results

Six hundred and six patients were identified at 27 paediatric rheumatology centres in 5 countries (Argentina, Brazil, Italy, Mexico, and the United Kingdom). Fifty-four patients (8.9%) were excluded because the clinical chart could not be retrieved and 62 patients (10.2%) were excluded because they had a disease onset before 1980 or a follow-up duration <2 years or undetermined. The remaining 490 patients, including 15 (3.1%) who had died, were included in the study. Of them, 248 (50.6%) were enrolled in Europe (168 in Italy and 80 in the United Kingdom), and 242 (49.4%) were enrolled in Latin America (117 in Brazil, 75 in Argentina and 50 in Mexico).

Table I shows the main demographic features of the study patients, considered as whole and by continent. Overall, two-thirds of patients were female and the mean age at disease onset was 7.0 years. The mean disease duration between disease onset and last follow-up visit or death was 7.7 years (range 2–25.2 years). Onset was most common between age 5–10 years and least common after age 10 years. Around one-third of patients were age <5 years at disease onset. There was a relatively greater prevalence of females in patients with onset between 5 and 10 years of age (71.1%) than in those with onset before 5 years (60.5%) or after 10 years (63.1%), but the difference was not significant. More than half patients had disease onset in the decade 1991-2000, whereas the percentage of patients who had disease onset between 1980 and 1990 and between 2001 and 2004 was comparable (around 20%). Gender ratio, age at disease onset and distribution of onset across decades were comparable between the 2 continental cohorts. European patients had a slightly longer time lag between disease onset and disease diagnosis than did Latin American patients. The age at onset for girls and boys is depicted in Figure 1. There was a peak age at 5 years for girls, whereas no definite peak age was seen for boys. Girls outnumbered boys at all ages, except at ages 12 and 15. The relative prevalence of females was more marked between ages 7 and 11 years. Overall, disease onset was less common after 11 years of age.

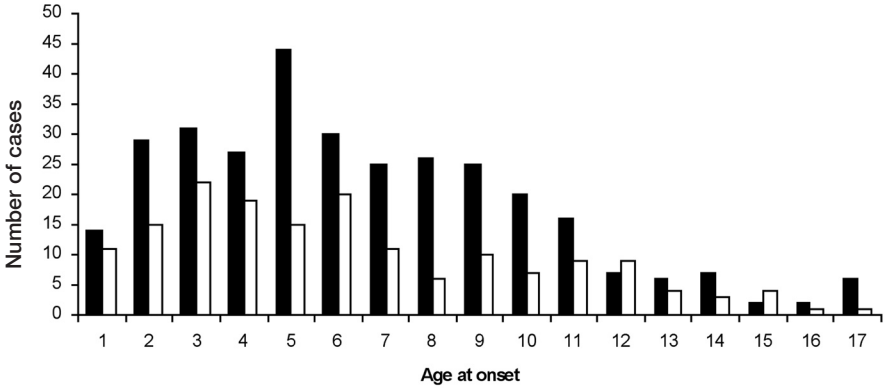

Fig. 1. Age at disease onset for girls (black bars) and boys (white bars).

The disease characteristics, diagnostic investigations, and disease course are also presented in Table I. Disease presentation was recorded as being more frequently acute than insidious. Around 60% of patients underwent electromyography, which was abnormal in more than 90% of instances. Muscle biopsy was performed in 55.2% of patients, 75.7% of whom had the investigation performed surgically. Histological evaluation of muscle biopsy samples was pathologic in 88.6% of instances. The course type was monophasic in 41.3% of patients and chronic polycyclic or continuous in 58.6% of patients. There were no differences in any of these features between European and Latin American patients.

The main clinical manifestations observed at disease onset in the whole patient sample were, in order of frequency, muscle weakness (84.9%), Gottron's papules (72.9%), heliotrope rash (62%), malar rash (56.7%), arthritis (35.7%), fever (30.8%), dysphagia (17.8%), dysphonia (11.4%), skin ulcers (6.3%), Raynaud phenomenon (5.3%), calcinosis (3.7%), and intestinal vasculitis (0.2%). The frequency of presenting manifestations was comparable between the 2 continental cohorts, with the exception of malar rash and dysphonia, which were seen more commonly in European patients, and Gottron's papules, which were more frequent in Latin America patients (Fig. 2).

Table II shows the frequency of presenting clinical manifestations by age at disease onset. All clinical manifestations were comparable across onset age groups, with the exception of a greater frequency of dysphagia in older children. Analysis of predictors of long-term outcome showed that cutaneous manifestations at onset were protective for decreased muscle function on the CMAS, but predicted continued disease activity on the MYOACT. Children who presented with dysphagia or dysphonia were more likely to have decreased muscle function on the CMAS. Muscle weakness at onset was associated with long-term muscle damage (7).

The medications administered during the disease course were, in order of frequency, corticosteroids (98.5%), methotrexate (56.2%), high-dose (pulse) intravenous methylprednisolone (41.7%), antimalarials (32.4%), cyclosporine (25.5%), intravenous immunoglobulin (17.2%), cyclophosphamide (10.1%), bisphosphonates (10.0%), azathioprine (9.8%), and biologic medications (2.7%). The comparison of drug therapies between European and Latin American patients is illustrated in Figure 3. European patients had received more frequently high-dose intravenous

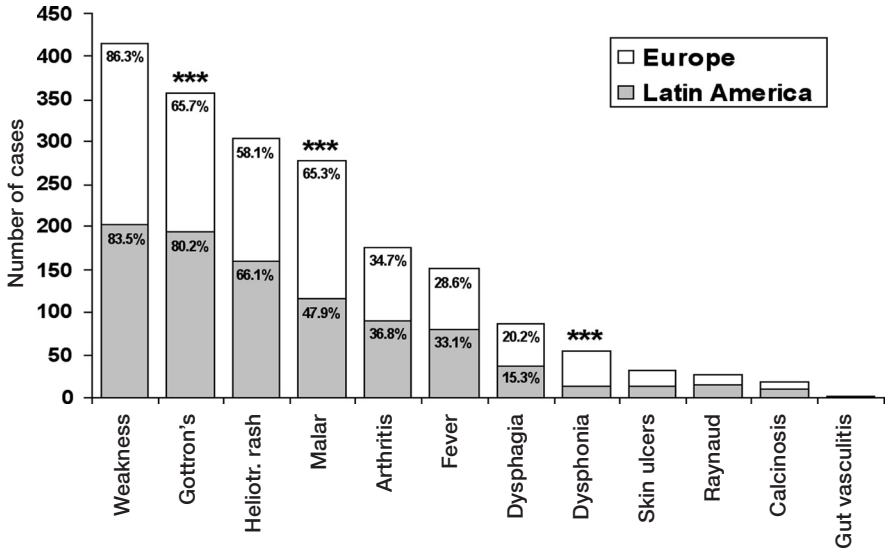

Fig. 2. Frequency of clinical manifestations at disease onset in European and Latin American patients. ***$p<0.001$.

Table II. Presenting manifestations by onset age. Data are number (percentage).

Manifestation	< 5 years (n=172)	5–10 years (n=211)	>10–18 years (n=103)	*p*-value	Comparisons significant on post-hoc test*
Muscle weakness	140 (81.4)	182 (86.3)	90 (87.4)	0.30	
Gottron's sign	122 (70.9)	157 (74.4)	75 (72.8)	0.75	
Heliotrope rash	103 (59.9)	135 (64.0)	62 (60.2)	0.67	
Malar rash	99 (57.6)	116 (55.0)	60 (58.3)	0.82	
Arthritis	57 (33.1)	79 (37.4)	35 (34.0)	0.65	
Fever	57 (33.1)	60 (28.4)	32 (31.1)	0.61	
Dysphagia	30 (17.4)	28 (13.3)	28 (27.2)	0.01	5-10 years *vs.* >10-18 years
Dysphonia	24 (14.0)	22 (10.4)	9 (8.7)	0.36	
Cutaneous ulcers	13 (7.6)	12 (5.7)	5 (4.9)	0.62	
Raynaud phenomenon	4 (2.3)	14 (6.6)	7 (6.8)	0.11	
Calcinosis	11 (6.4)	5 (2.4)	2 (1.9)	0.07#	
Intestinal vasculitis	0 (0.0)	1 (0.5)	0 (0.0)	0.52#	

p-values refer to the chi-square test unless otherwise specified. #Fisher's exact test. *Pairs of comparisons that are statistically significant after Bonferroni correction for multiple comparisons.

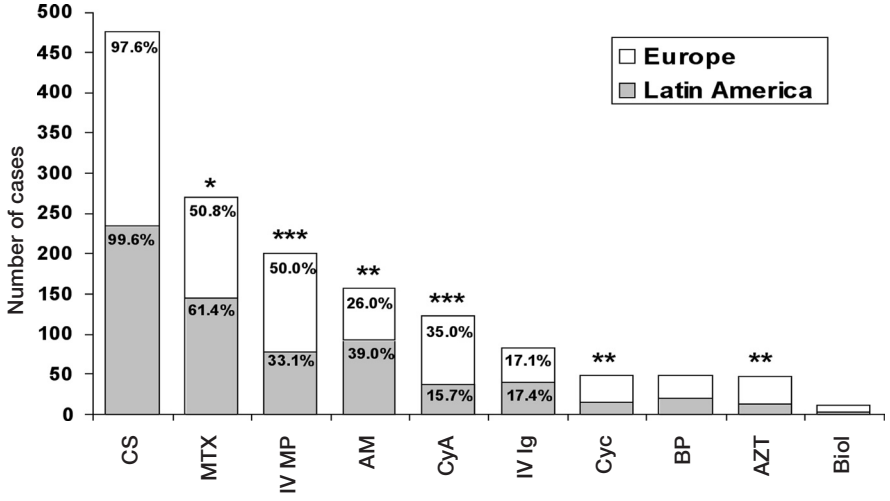

Fig. 3. Frequency of medications administered during the disease course in European and Latin American patients. CS: corticosteroids; MTX: methotrexate; IV MP: intravenous methylprednisolone; AM: antimalarials; CyA: cyclosporine A; IV Ig: intravenous immunoglobulin; Cyc: cyclophosphamide; BP: bisphosphonates; AZT: azathioprine; Biol: biologic medications. *$p<0.05$, **$p<0.01$, ***$p<0.001$.

methylprednisolone, cyclosporine, cyclophosphamide, and azathioprine, whereas methotrexate and antimalarials medications were used more frequently by Latin American paediatric rheumatologists. Intravenous immunoglobulins were administered with equal frequency in the 2 continental populations.

Table III reports the frequency of prescribed medications by decade of disease onset. Although corticosteroid therapy was given to nearly all patients seen throughout decades, use of high-dose intravenous methylprednisolone became more popular in the 90s and 2000s. Methotrexate administration increased sharply over the years, with patients seen in the 2000s being almost three times more likely to have received this drug than did those seen in the 80s. Cyclosporine, intravenous immunoglobulin, and antimalarials were given more frequently in the 90s than in the 80s, but their use remained stable thereafter. Prescription of azathioprine tended to decline over the years, whereas cyclophosphamide was used with similar frequency across decades. However, administration of cyclophosphamide was marked by a progressive decline in the use of the oral route and by a relative increase in the choice of the intravenous pulse regimen in the 2000s.

Methylprednisolone pulses were found to be protective toward the development of global and muscle damage. Therapy with intravenous immunoglobulin methotrexate, and cyclophosphamide was associated with continued muscle weakness, ongoing disease activity, long-term damage, and worse functional outcome (results not shown). However, this association is likely explained by patients with more severe disease being more likely to have received such treatments.

Discussion

We evaluated the demographic and clinical features, diagnostic procedures, and frequency of medication use in 490 children with JDM seen over a 25-year period in 27 paediatric rheumatology centres in 2 continents, Europe and Latin America. The female-to-male ratio (2:1) and the average age at disease onset (7.0 years) seen in our patients are similar to those reported

Table III. Medication choices by decade. Data are number (percentage).

Medication	1980 -1990 (n=94)	1991-2000 (n=277)	2001-2004 (n=108)	*p*-value	Comparisons significant on post-hoc test*
Corticosteroids	93 (98.9)	274 (98.9)	105 (97.2)	0.51#	
Intravenous methylprednisolone pulses	26 (27.7)	120 (43.3)	54 (50.0)	0.004	1980-1990 *vs*. 1991-2000 1991-2000 *vs*. 2001-2004
Methotrexate	27 (28.7)	153 (55.2)	89 (82.4)	<0.0001	1980-1990 *vs*. 2001-2004 1980-1990 *vs*. 1991-2000 1991-2000 *vs*. 2001-2004
Cyclosporine	10 (10.6)	85 (30.7)	28 (25.9)	0.0006	1980-1990 *vs*. 1991-2000 1980-1990 *vs*. 2001-2004
Intravenous immunoglobulin	6 (6.4)	55 (19.9)	22 (20.4)	0.007	1980-1990 *vs*. 1991-2000 1980-1990 *vs*. 2001-2004
Antimalarials	22 (23.4)	92 (33.2)	42 (38.9)	0.06	
Azathioprine	14 (14.9)	25 (9.0)	8 (7.4)	0.16	
Cyclophosphamide	12 (12.8)	21 (7.6)	15 (13.9)	0.11	
Oral cyclophosphamide	10 (10.6)	7 (2.5)	1 (0.9)	0.001#	1980-1990 *vs*. 1991-2000 1980-1990 *vs*. 2001-2004
Intravenous cyclophosphamide pulses	3 (3.2)	14 (5.1)	14 (13.0)	0.006	1980-1990 *vs*. 2001-2004 1991-2000 *vs*. 2001-2004

p-values refer to the chi-square test unless otherwise specified. #Fisher's exact test. *Pairs of comparisons that are statistically significant after Bonferroni correction for multiple comparisons.

in previous studies (1, 2). The greater frequency of disease onset in the 5-to 10-year old range is consistent with the peak onset age reported in previous studies (26). The proportion of children with disease onset before 4 years of age in our cohort (25.7%) is comparable to that found by Pachman and co-workers (25%) (27). Demographic features were remarkably similar in the 2 continental cohorts, indicating that the paediatric age group targeted by the disease is the same in different ethnic groups or geographic areas.

As expected, proximal muscle weakness and the characteristic skin rashes were the most common presenting features in the large majority of our children with JDM. The frequency of onset manifestations observed in the study sample is in the range of that reported in other series of JDM patients (1, 12, 28-30). The frequency of presenting symptoms was comparable between European and Latin American patients, with the sole exception a greater frequency of malar rash and dysphonia in European patients and of Gottron's papules in the Latin American population. Presenting features were also similarly distributed across different onset age groups, although dysphagia was reported more frequently in patients with onset after 10 years of age (perhaps owing to the greater capacity of older children to self report this complaint). These findings suggest that the clinical spectrum of JDM is similar in different geographic or ethnic groups and is not influenced by age at disease onset.

Although the presentation of JDM is variable, it is seen that onset is usually insidious, with development of progressive muscle weakness and pain; a more acute onset, with fever, prostration and profound muscle weakness occurs in approximately one third of children (1). In our population, acute onset was recorded more frequently than insidious onset, with both continental samples revealing the same trend. Our findings should be regarded with caution, however, due to the difficulty in ascertaining the severity and acuteness of disease presentation in a retrospective analysis.

Earlier studies showed that 15–25% of JDM patients had a monophasic course, with the remaining patients demonstrating a chronic course with either flares or unremitting disease activity (16, 17). The more recent analyses have reported a greater proportion of patients with monophasic course (37–40%) (5, 18), which is comparable to that found in our study (41.3%). The increase over time in the proportion of JDM patients with milder disease course may reflect the recognition of the importance of prompt aggressive therapy with high-dose corticosteroids after the 1980s.

The detection of the typical myopathic abnormalities on electromyography and the demonstration of the characteristic pathological changes on muscle biopsy are still the mainstay diagnostic procedure for JDM. They are part of the Bohan and Peter criteria used to diagnose JDM (14, 15). However, it has become clear that few clinicians subject children to the full work-up that would be required to fulfil the Bohan and Peter criteria. A recent international survey has shown that only 56% of paediatric rheumatologists used electromyography, and only 61% used muscle biopsy to diagnose JDM (31). Likewise, only 59.9% and 55.2% of our patients underwent electromyography or muscle biopsy, respectively.

Our analysis provided the opportunity to compare the medications used in the treatment of children with JDM by paediatric rheumatologists working in different areas of the world and to examine the trend in therapeutic choices throughout decades. As expected, nearly all patients in both continental cohorts were given corticosteroids during

their disease course. However, European physicians used more frequently high-dose intravenous methylprednisolone, cyclosporine, cyclophosphamide, and azathioprine, whereas Latin American physicians were more likely to prescribe methotrexate and antimalarial drugs. These differences are unlikely to depend on diversities in disease manifestations as the frequency of presenting features of the 2 continental populations was comparable. A recent survey of treatment in JDM among North American paediatric rheumatologists showed considerable variation in the choice of medications and the doses administered (32). These observations reflect the lack of data on which to base treatment decisions.

We found a remarkable change over time in the frequency of use of medications or therapeutic protocols. As compared to the 80s, there was a significant increase after the 90s in the frequency of administration of high-dose (pulse) intravenous methylprednisolone and second-line drugs, namely methotrexate, cyclosporine, and intravenous immunoglobulin. This reflects the recent shift toward early aggressive treatment of JDM, aimed to achieve rapid and sustained remission and to prevent disease- and treatment-related complications (2, 8, 33, 34).

Although the increase in the frequency of use of antimalarial agents across decades was not significant, these medications were prescribed to as many as 39% of patients managed in the 2000s. This suggests that antimalarial drugs, particularly hydroxychloroquine, remain popular for the treatment of skin manifestations of JDM, which is the indication for which they were initially proposed (35). Use of azathioprine was found to decline over time, whereas cyclophosphamide was still administered to a sizable proportion of patients seen in the current decade, particularly in Europe. This drug was likely used in patients with severe, refractory disease, particularly those with the most serious complications (36). Administration of cyclophosphamide through the oral route was almost abandoned in the 2000s, whereas the regimen based on intravenous pulse infusions became increasingly more popular over time. This may reflect the notion that intravenous pulse regimens lead to a lesser toxicity than does oral daily administration (37). Only a few patients received the novel biologic medications, reflecting the still limited experience with these agents in JDM (38, 39). Bisphosphonates were prescribed to approximately 10% of patients seen in each decade. Indications of these drugs in JDM include management of steroid-related osteoporosis and calcinosis (40).

Our findings should be interpreted in the light of some potential limitations. All study data were recorded through the retrospective review of clinical charts. A retrospective analysis is subject to missing and possibly erroneous data. We could not include information regarding features of muscle biopsy, nailfold capillary studies, magnetic resonance imaging, or the presence of myositis-specific autoantibodies or HLA alleles.

In summary, we have described the demographic features, disease characteristics, diagnostic investigations, and treatment modalities in the largest series of children with JDM reported to date. Our results highlight the current tendency toward early aggressive treatment, namely with high-dose intravenous methylprednisolone and methotrexate. There were, however, several differences in the frequency of therapeutic choices between European and Latin American paediatric rheumatologists, which reflect the scarcity of evidence-based data on which to base treatment decisions. This underscores the need of developing uniform therapeutic protocols for JDM, possibly based on the investigation of currently available therapeutic regimens and novel medications in randomised controlled trials.

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