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Race, Income and Disease Outcomes in Juvenile Dermatomyositis

Kathryn Phillippi, DO[1], **Mark Hoeltzel, MD**[2], **Angela Byun Robinson, MD, MPH**[3], **Susan Kim, MD, MMSc**[4], and **for the CARRA Legacy Registry Investigators**[*]

[1]Fellow, Division of Pediatric Infectious Diseases and Rheumatology, Rainbow Babies and Children's Hospital / Case Medical Center, Cleveland, OH

[2]Assistant Professor, Division of Pediatric Rheumatology, C. S. Mott Children's Hospital, University of Michigan, Ann Arbor, MI

[3]Assistant Professor, Division of Pediatric Infectious Diseases and Rheumatology, Rainbow Babies and Children's Hospital / Case Medical Center, Cleveland, OH

[4]Associate Clinical Professor, Division of Rheumatology, University of California at San Francisco, Benioff Children's Hospital, San Francisco, CA

Abstract

Objective—To determine the relationship between race, income, and disease outcomes in children with juvenile dermatomyositis (JDM).

Study design—Data from 438 subjects with JDM enrolled in the Childhood Arthritis and Rheumatology Research Alliance (CARRA) Legacy Registry were analyzed. Demographic data included age, sex, race, income and insurance status. Clinical outcomes included muscle strength, presence of rash, calcinosis, weakness, physical function and quality of life measures. Disease outcomes were compared based on race and income.

Results—Minority subjects were significantly more likely to have low family income, and significantly worse scores on measures of physical function, disease activity and quality of life measures. Lower income subjects had worse scores on measures of physical function, disease activity and quality of life scores, as well as weakness. Black subjects were more likely to have calcinosis. Despite these differences in outcome measures, there were no significant differences in time to diagnosis or disease duration. Using calcinosis as a marker of disease morbidity, Black race, annual family income less than $50,000 per year, negative ANA, and delay in diagnosis greater than 12 months were associated with calcinosis.

Corresponding information: Kathryn Phillippi, 11000 Euclid Ave, Cleveland, Ohio 44106, Kathryn.phillippi@uhhospitals.org, (937) 524-4660 (phone), (216) 844-7587 (fax).

[*]List of members of the CARRA Legacy Registry is available at www.jpeds.com (Appendix)

The authors declare no conflicts of interest.

Conclusions—Minority race and lower income are associated with worse morbidity and outcomes in subjects with JDM. Calcinosis was more common in Black subjects. Future studies are needed to further understand these associations so that efforts may be developed to address health disparities in subjects with JDM and improve disease outcomes.

Keywords

JDM; Juvenile Dermatomyositis; Health Disparities; Calcinosis

Juvenile dermatomyositis (JDM), a rare, autoimmune, inflammatory myositis, with an annual incidence estimated between 1.9 and 4.1 per million children per year.[1,2,3] The diagnosis of JDM is confirmed in children with proximal muscle weakness, pathognomonic rash, and elevated muscle enzymes with typical electromyography (EMG) and/or muscle biopsy changes. In recent years, magnetic resonance imaging (MRI) has played an increasing important role in the diagnosis of inflammatory muscle disease in lieu of invasive testing.[4,5]

With improvements in treatment, survival among patients with JDM has improved over the past several decades, from 67% to 99%[6,7]. Although the survival in JDM has improved, many long-term morbidities remain, including persistent weakness, rash, and calcinosis. The presence of calcinosis is an accepted surrogate marker for JDM morbidity and is widely used in outcome measures to assess disease damage, including the Myositis Damage Index (MDI)[7–9].

The factors that contribute to the development of long term morbidity in JDM are unknown, but an earlier description of the Childhood Arthritis and Rheumatology Research Alliance (CARRA) Legacy Registry cohort reported an increased odds of developing calcinosis in Black subjects, suggesting a racial difference.[10] Other pediatric studies have shown racial and ethnic disparities may contribute to morbidity and mortality in chronic conditions such as systemic lupus erythematosus, arthritis, asthma, juvenile diabetes and kidney transplantation.[11–20]

The degree to which a patient's income or race impacts outcome has not been examined in depth in the JDM population. Determining if race and income are associated with increased morbidity is essential so that disparities can be identified and addressed. Such changes might include improved access to pediatric rheumatologists, improved insurance coverage, or increased education of primary care physicians, which may facilitate diagnosis of JDM.

METHODS

The CARRA Legacy Registry is a multi-center registry developed within North America to capture information about multiple rheumatic diseases including juvenile idiopathic arthritis, mixed connective tissue disease, systemic lupus erythematosus, juvenile dermatomyositis, vasculitis, scleroderma, sarcoidosis, and primary juvenile fibromyalgia. Between May 2010 and July 2014, subjects with diagnosed rheumatic diseases were included from 55 pediatric CARRA centers in North America. It is an observational data capture registry, representing a convenience sample of eligible subjects. Institutional Review Board (IRB) approval was

obtained at each enrolling site. Subjects and/or parent/legal guardian were required to provide informed consent (in English or Spanish).

Subjects were eligible for inclusion if they were less than 21 years of age, with onset of JDM prior to 18 years of age, and met definite criteria for JDM as defined by modified Bohan and Peter criteria. Bohan and Peter criteria were modified by the CARRA Legacy Registry project investigators to include MRI as an acceptable diagnostic modality for JDM in order to reflect common practice and broaden subject inclusion. Subjects could be at any stage of disease and were not required to be recently or newly diagnosed. Subjects were excluded from analysis if age of onset could not be clearly identified as prior to 18 years old or if they did not meet criteria for a diagnosis of definite JDM by these modified standards[4,5]: Classic skin involvement for JDM, and at least 3 of the following (1) Muscle weakness; (2) Elevation of muscle enzyme(s); (3) Abnormal EMG suggestive of inflammatory myopathy; (4) Abnormal muscle biopsy suggestive of inflammatory myopathy; or (5) MRI evidence of myositis (modification introduced by CARRA Legacy Registry investigators [21]).

Data collection

Clinical data were collected from the subjects/guardians and enrolling physician using both general and JDM-specific case report forms at the time of enrollment. In addition, the subject's medical records were reviewed for previously obtained disease-specific clinical information. Subjects diagnosed prior to May 2010 were recruited retrospectively. Data was pooled and stored in a secure centralized electronic database and de-identified prior to analysis.

Demographic and Baseline Information

Baseline demographic information was obtained by the enrolling physician. Family income was determined by self-report from the parent or guardian of the enrolled subject. Medical insurance status was recorded by the enrolling physician as yes or no. No information was collected regarding public or private sources of insurance, or co-insurance.

Race and ethnic background were self-identified by the subject/guardian during enrollment. For the purposes of this study, a subject was defined as White if the subject self-identified as White and non-biracial. Subjects were defined as Black if they self-identified as Black or Black and biracial. All others were defined as minority, non-Black. Subjects were dropped from analysis if there was no race identified (n=3). Treatment history was recorded by the enrolling physician from chart abstraction or other records. Disease duration at enrollment was calculated by age at onset of symptoms subtracted from the age at enrollment. We defined a delay in diagnosis as being greater than 12 months after onset of symptoms to first appointment with a pediatric rheumatologist. Family income was reported by subjects in increments of $25,000. We compared patient characteristics according to reported annual family income, using a cutoff of greater or less than $50,000, which is closest to the low-income threshold for a family of four with two children in the United States[25].

Outcome Measures

Data were collected for muscle strength, physical functioning and quality of life of patient. The Childhood Myositis Assessment Scale (CMAS; maximum score 52), the Childhood Health Assessment Questionnaire (CHAQ; where 0=normal and 3=worst), health-related quality of life measures (HRQoL), the American College of Rheumatology functional class rating (ACR functional class), and global disease assessments. The CMAS has been validated to quantitatively assess muscle strength and endurance in children with idiopathic inflammatory myopathies [21–23]. The ACR functional class is described from Class I, able to perform usual activities of daily living including self-care, vocational, and avocational activities to Class IV, limited in ability to perform usual self-care, vocational, and avocational activities[24]. The ACR class was reported as worst ever during disease course by the enrolling physician either from subject report or chart abstraction. Proximal muscle weakness, rash (malar or facial erythema), Gottron rash and calcinosis were reported as present or absent by the enrolling physician. We used calcinosis as a surrogate marker of disease morbidity.

Statistical analyses

Statistical analysis was conducted using STATA software, version 10.0 (StataCorp, Austin, TX, USA). All data analyses were preceded by extensive data checking and verification to identify and resolve the reasons for missing values, inconsistencies, and out-of-range values. Descriptive statistics were computed to summarize each variable, including the mean, median, standard deviation and interquartile range (IQR) for continuous variables; and frequencies (percentages) for categorical variables. Chi square and Kruskal-Wallis testing was used to evaluate associations between demographics, income, treatment and disease activity with disease duration, race and income. All tests were 2-sided and p-values less than 0.05 were considered statistically significant. We performed multivariate logistic regression of the outcome calcinosis, which was used as a surrogate marker of disease morbidity. Selection of variables in the multivariate model was performed using results from bivariate testing in a stepwise approach with previously associated variables such as duration of disease, sex, and delay to treatment selected automatically. Variables significantly ($p<0.1$) associated with calcinosis (race and income) and static variables associated with race and income (ANA positivity) were included in the model. If the variable did not add significantly to the model fit, and there was no evidence of interaction, the variable was dropped from the model. Tests for interactions were performed using interaction terms of binary outcomes for Black race, income less than $50,000 and ANA.

RESULTS

Between May 2010 and July 2014, 639 subjects meeting modified Bohan and Peter criteria for probable or definite JDM were enrolled from 55 CARRA sites. Confirmation of definite JDM by EMG, MRI, or biopsy was obtained in 441 cases. Of these, 438 subjects had full racial information available. Median disease duration from time of symptom onset to registry enrollment was 3.1 years (IQR 1.2–6.2 years), and the median time to diagnosis was 4 months (IQR 1.9–10.7).

Overall, 79% of subjects were identified as White (347/438), 13% (57/438) identified as Black, and 8% as minority, non-Black (34/438). Subjects were predominantly female (71%) with a median age of onset of 5.6 years. Over 90% of all subjects had received oral corticosteroids and disease antihermatic modifying drugs. Ten percent had clinical weakness, 32% had active rash (facial erythema) at the time of enrollment. The median CHAQ among all subjects at the time of enrollment was 0.12 and the CMAS was 50 (Table I).

Associations with Race and Income

Black children tended to be older at age of onset, but this was not statistically different (5.6, 6.4 and 5.0 years, for White, Black and minority non-Black, respectively, p=0.36). No differences between sex, time to diagnosis, and disease duration were found when comparing racial groups, and there were no differences in the treatments (intravenous corticosteroids, oral corticosteroids, disease modifying antirheumatic drugs, biologics, intravenous immunoglobulin, and cyclophosphamide) administered to subjects (Table I).

Family income was reported in 346 subjects (79%). Black subjects were more likely than other groups to report income below $50,000. The vast majority of patients had medical insurance (97.5%); 98.3%, 94.7% and 94.1%, for White, Black, and minority non-Black, respectively, p = 0.13. Subjects of Black or minority non-Black race were statistically more likely to have worse CHAQ scores, patient global, and HRQoL scores. Physician assessment of weakness, facial erythema and Gottron rash did not differ between races. Worst ACR functional class also did not differ between groups. Black and minority non-Black race were associated with a lower likelihood of positive ANA (41.9% and 45.8% vs. 68.1% in White subjects, p <0.01).

There were no measurable differences based on age of onset, sex and median length of time from symptom onset to first visit with a pediatric rheumatologist based on income cutoff of $50,000 per year (Table II). Subjects with a lower income compared with higher income, had statistically worse CHAQ, patient global, and, physician global scores and lower quality of life scores. In addition, moderate to severe weakness was more common in low income subjects at the time of enrollment (10.3% vs. 8.7%, p=0.03), though there was no difference based on CMAS. ACR functional class was worse in patients with lower incomes.

Associations with calcinosis

Of all subjects, 15% (63 out of 427) had calcinosis (Table I). A greater percentage of Black subjects (24%, 13/57) had calcinosis compared with White subjects (4%) and minority, non-Black subjects (9%), but this was not statistically significant (p=0.09). Subgroup analysis of Black subjects showed the univariate odds of calcinosis in Black subjects were 2 times higher than other subjects in this cohort, OR = 2.05 (95% CI 1.03, 4.09).

Annual family income less than $50,000 per year was associated with a calcinosis OR of 1.92 (95% CI 1.03, 3.59). Given missing data regarding family income status in 21% of the subjects, we conducted a sensitivity analysis assuming those who did not respond had income less than $50,000 annually. This assumption did not change the results. Delay to diagnosis and increased disease duration of disease were also independent predictors of

calcinosis; and 25% of subjects diagnosed after 12 months of symptoms were reported to have calcinosis.

In a multivariable analysis of calcinosis alone (Table III), Black race, time to diagnosis and duration of disease were found to be significant independent predictors of calcinosis. No significant interaction between Black race and positive ANA or Black race and family income less than $50,000 was found in this analysis.

DISCUSSION

Our analysis revealed independent health disparities in our JDM cohort based on race and income. Black and minority non-Black subjects had lower reported income. These subjects tended to have lower CHAQ scores, health related quality of life and patient global scores compared with White subjects. Black subjects had higher rates of calcinosis.

Similar results were found when comparing patient groups based on lower income. We found that more minority subjects reported an annual family income of less than $50,000; these subjects also had higher rates of calcinosis, worse CHAQ/functional scores, health-related quality of life, weakness, patient and physician global scores. Univariate analysis showed that lower income was associated with calcinosis, while multivariable analysis revealed that race, but not income, was an independent risk factor for the development of calcinosis; no effect of interaction was seen between race and income less than $50,000. This does not completely rule out the possibility of interaction, but helps inform our model.

The known correlation between minority race and lower income in the United States,[25] was also found in our JDM cohort. Black and minority non-Black race and low income were associated with worse outcome measures in our cohort, but it is not clear, from the available registry data and our analysis, if this correlation is due to genetic differences, environmental exposures, access to care, type of insurance, treatment adherence, or other treatment and non-treatment related factors.

Calcinosis occurs in up to 40% of subjects with JDM, although the current prevalence ranges from 10 to 70% [7,21,26–29]. We found that delay to diagnosis and duration of disease were associated with the development of calcinosis, as has been reported by others[30–32].

We found higher rates of calcinosis in Black subjects but overall rates of reported calcinosis in our cohort is similar to what is reported in previous populations[7,31–33]. Extremely high rates of calcinosis have been described in an African cohort of JDM subjects, with a 70% cumulative risk of calcinosis which is often associated with vasculitis in that patient population[34]. Rates of calcinosis in our Black North American cohort, though not as high as what has been reported in African JDM subjects, suggest a genetic component in the development or pathogenesis of calcinosis.

Similar to other inflammatory conditions such as familial Mediterranean fever, systemic lupus erythematosus, and psoriatic arthritis, genetic as well as environmental factors likely contribute to the expression of the disease. Some differences in autoantibodies and clinical expression of juvenile myositis have been described, such as anti-SRP antibodies, which is

associated with Black race, severe disease and juvenile polymyositis.[35] Differences in immune response genes between races have also been described in JDM [36]. Thus, our findings support the concept that JDM is a multifactorial, complex autoimmune disorder with a genetic contribution.

We were encouraged to find no significant differences in treatments administered based on race or income. However our findings of several worse outcome measures, based on race and income, suggest that there are additional factors that we are not measuring, recognizing or treating. In addition, we suspect, similar to what has been described in other pediatric chronic conditions such as asthma, inflammatory bowel disease and juvenile arthritis[37–41], there are differences in responses and efficacy of medications based on genetic variation. Therefore, this data suggests that improved pharmacogenetic assessment and approaches are necessary to improve future outcomes in JDM. We also note that detailed data regarding duration and total cumulative dose of treatments and adherence were not collected, which may have contributed to these differences in outcome measures. Overall, most subjects in this cohort have mild disease and a good outcome, including subjects of Black race and low income.

The results from this analysis are not without limitations. The CARRA Legacy Registry may not represent the full spectrum of patients with JDM. Subjects were only enrolled from academic centers, where CARRA members, who are pediatric rheumatologists, care for patients and may not represent a subset of patients who do not have access to care at these centers. In addition, we may be missing a subset of patients followed by other specialists, such as dermatologists and neurologists. Furthermore, the method of accrual into the registry may have influenced subject enrollment to the registry: subjects were enrolled by researchers at individual centers and though the intent was to enroll all subjects with JDM, we cannot rule out the possibility that there was an ascertainment bias in the subject enrolled in the CARRA Legacy Registry. This cohort is not an inception cohort and therefore can lead to selection bias as those patients with severe disease or mild disease might not be included. Because subjects were enrolled at any stage of disease, this registry represents a cross-sectional, heterogeneous population. Families with limitations with the English language may have been excluded. In addition, many items were self-reported from subjects and families, including income, race and insurance status, and data collection was often retrospective. Information about when the subject met diagnostic criteria for JDM is not known; only information on when the first symptom was noted by the parent and when the subject was first seen by a pediatric rheumatologist was included in the data collection. More details related to parent education level, subtype of insurance (i.e. private vs. Medicaid/government supported) would have been helpful to decipher the implications of income and outcomes in JDM. This analysis was not adjusted for multiple comparisons; the results of this study should be viewed as hypothesis generating rather than hypothesis testing.

In conclusion, this study of North American children with JDM enrolled in the CARRA Legacy Registry suggest that minority race and lower income are associated with worse outcomes, increased morbidity, and decreased physical function. Registries such as the CARRA Legacy Registry, aimed to study rare conditions like JDM are powerful, because

they allow us to ask and answer questions beyond the reach of any single rheumatology practice. Prospective, multicenter, longitudinal research is required to clarify the observations made in this study between different racial and socioeconomic groups.

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Appendix

Additional CARRA Registry site principal investigators and research coordinators are:

Leslie S. Abramson, MD, University of Vermont College of Medicine, Burlington, VT

Eleanor S. Anderson, MD, Permanente Medical Group, Union City, CA

Mara L. Becker, MD, Children’s Mercy Hospitals and Clinics, Kansas City, MO

Heather Benham, CPNP, RN, Texas Scottish Rite Hospital, Dallas, TX

Timothy. Beukelman, MD, MSCE, University of Alabama-Birmingham, AL

Peter R. Blier, MD, PhD, Baystate Children’s Hospital, Springfield, MA

Hermine I. Brunner, MD, MSc, MBA, Cincinnati Children’s Hopsital Medical Center, Cincinnati, OH

Joni Dean, MSN, American Family Children’s Hospital, Madison, WI

Fatma Dedeoglu, MD, Boston Children’s Hospital, Boston, MA

Brian M. Feldman, MD, MSc, The Hospital for Sick Children, Toronto, ON

Polly I. Ferguson, MD, University of Iowa Carver College of Medicine, Iowa City, IA

Donald P. Goldsmith, MD, St. Christopher’s Hospital for Children, Philadelphia, PA

Beth S. Gottlieb,MD, MS, Cohen Children’s Medical Center, New Hyde Park, NY

Thomas B. Graham, MD, MS, Vanderbilt Children’s Hospital, Nashville, TN

Thomas A. Griffin, MD, PhD, Levine Children’s Hospital, Carolinas Medical Center, Charlotte, NC

Hilary M. Haftel, MD, MHPE, University of Michigan Medical Center, Ann Arber, MI

Gloria C. Higgins, MD, PhD, Nationwide Children’s Hospital, Columbus, OH

J.R. Hollister,MD, MD, University of Colorado, Colorado Springs, CO

Joyce J. Hsu, MD, Stanford University, Palo Alto, CA

Anna Huttenlocher, MD, University of Wisconsin School of Medicine, Madison, WI

Norman T. Ilowite, MD, Children's Hospital of Montefiore, Bronx, NY

Lisa F. Imundo, MD, Columbia University Medical Center, New York, NY

Rita S. Jerath, MBChB, Children's Hospital of Georgia, Augusta, GA

Lawrence K. Jung, MD, Children's National Medical Center, Washington, DC

Philip J. Kahn, MD, New York University, New York, NY

Daniel J. Kingsbury, MD, Randall Children's Hospital at Legacy Emanuel, Portland, OR

Kristin E. Klein, RN, BSN, University of Louisville, Louisville, KY

Marisa S. Klein-Gitelman, MD, MPH, Lurie Children's Hospital of Chicago, Northwestern University, Chicago, IL

Sivia K. Lapidus, MD, Goryeb Children's Hospital, Morristown, NJ

Thomas JA. Lehman, MD, Hospital for Special Surgery, New York, NY

Carol B. Lindsley,MD, University of Kansas City Medical Center, Kansas City, KS

Michael A. Malloy, MD, Warren Clinic, Tulsa, OK

Deborah K. McCurdy, MD, Mattel Children's UCLA, Los Angeles, CA

Eyal Muscal, MD, Baylor College of Medicine, Houston, TX

Judyann C. Olson, MD, Medical College of Wisconsin, Milwaukee, WI

Kathleen M. O'Neil, MD, Riley Hospital for Children, Indianapolis, IN

Karen Onel, MD, University of Chicago, Chicago, IL

Sampath Prahalad, MD, MSc, Emory Children's Center, Atlanta, GA

Marilynn G. Punaro, MD, Texas Scottish Rite Hospital, Dallas, TX, C. Egla Rabinovich, MD, MPH, Duke University Medical Center, Durham, NC

Ann M. Reed, MD, Duke University, Durham, NC

Sarah Ringold, MD, MS, Seattle Children's Hospital, Bellevue, WA

Mary Ellen Riordan, BSN, Hackensack University Medical Center, Hackensack, NJ

Angela B. Robinson, MD, MPH, Rainbow Babies and Children's Hospital, Cleveland, OH

Deborah Rothman, MD, PhD, Albany Medical Center, Albany NY

Natasha M. Ruth,MD, MS, The Medical University of South Carolinam, Charleston, SC

Kenneth N. Schikler, MD, University of Louisville School of Medicine, Louisville, KY

Nora G. Singer, MD, MetroHealth Center, Cleveland, OH

Steven Spalding, MD, Phoenix Children's Hospital, Phoenix, AZ

Reema H. Syed, MD, MBBS, St. Louis University, St. Louis, MO

Kathryn S. Torok, MD, University of Pittsburgh Medical Center, Pittsburgh, PA

Jenna Tress, BA, The Children's Hospital of Philadelphia, Philadelphia, PA

Richard K. Vehe, MD, University of Minnesota, Minneapolis, MN

Emily Von Scheven, MD, University of California-San Francisco, San Francisco, CA

Lydia M. Walters, MSN, University of Miss Medical Center, Jackson, MS

Jennifer E. Weiss, MD, Hackensack University Medical Center, Hackensack, NJ

Pamela Weiss, MD, MSCE, The Children's Hospital of Philadelphia, Philadelphia, PA

Andrew J. White, MD, St. Louis Children's Hospital, St. Louis, MO

Jennifer M. Woo, MPH, Milwaukee, WI

Ali Yalcindag, MD, Hasbro Children's Hospital, Providence, RI

Lawrence S. Zemel, MD, Connecticut Children's Medical Center, Harford, CT

Glossary

ACR	American College of Rheumatology
CARRA	Childhood Arthritis and Rheumatology Research Alliance
CHAQ	Childhood Health Assessment Questionnaire
CMAS	Childhood Myositis Assessment Scale
EMG	Electromyography
JDM	Juvenile Dermatomyositis
MRI	Magnetic resonance imaging

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Table I

Patient demographics and disease characteristics by race.

	n	All (n = 438)	White (n = 347)	Black (n = 57)	Minority, non-Black (n = 34)	p-value
Demographics						
Age at onset, years	427	5.6 (3.7, 9.5)	5.6 (3.6, 9.4) N=337	6.4 (4.8, 10.4) N = 56	5.0 (3.0, 10.7) N = 34	0.36
Sex(female)	438	309 (70.6)	245 (70.6%)	41 (71.9%)	23 (67.7%)	0.91
Time to diagnosis, months	423	4.0 (1.9, 10.7)	4.3 (2.0, 10.7) N = 335	3.1 (1.9, 11.9) N = 55	3.1 (1.0, 7.8) N = 33	0.25
Duration of disease, years	427	3.1 (1.2, 6.2)	3.3 (1.2, 6.3) N = 337	2.8 (1.2, 4.5) N = 56	3.6 (1.2, 6.8) N = 34	0.75
Socioeconomic status						
Insurance (yes)	433	422 (97.5%)	336 (98.3%)	54 (94.7%)	32 (94.1%)	0.13
Income						
<$25,000/yr	438	60 (13.7%)	38 (11.0%)	16 (28.1%)	6 (17.7%)	<0.01
$25–49,999/yr		68 (15.5%)	46 (13.3%)	16 (28.1%)	6 (17.7%)	
$50–74,999/yr		66 (15.1%)	59 (17.0%)	5 (8.8%)	2 (5.9%)	
$75–99,999/yr		48 (11.0%)	42 (12.1%)	1 (1.8%)	5 (14.7%)	
$100–150,000/yr		63 (14.4%)	61 (17.6%)	0 (0.0%)	2 (5.9%)	
>$150,000/yr		41 (9.4%)	31 (8.9%)	5 (8.8%)	5 (14.7%)	
Unknown		92 (21.0%)	70 (20.2%)	14 (24.6%)	8 (23.5%)	
Patient characteristics						
Lipodystrophy	434	20 (4.6%)	14 (4.1%)	3 (5.4%)	3 (8.8%)	0.43
Calcinosis	427	63 (14.8%)	47 (13.8%)	13 (24.1%)	3 (9.1%)	0.09
ANA positive	349	221 (63.3%)	192 (68.1%)	18 (41.9%)	11 (45.8%)	<0.01
Treatments						

	n	All (n = 438)	White (n = 347)	Black (n = 57)	Minority, non-Black (n = 34)	p-value
IV/Pulse corticosteroids	406	248 (61.1%)	195 (60.0%)	33 (61.1%)	20 (74.1%)	0.69
Oral corticosteroids	410	398 (97.1%)	319 (97.3%)	53 (98.2%)	26 (92.9%)	0.20
DMARDS/non-biologic DMARDS	438	401 (91.6%)	319 (91.9%)	51 (89.5%)	31 (91.2%)	0.82
Cytotoxic therapy [1]	398	6 (1.5%)	4 (1.3%)	2 (3.9%)	0 (0.0%)	0.27
Biologics	438	182 (41.6%)	141 (40.6%)	22 (38.6%)	19 (55.9%)	0.20
IVIG	437	172 (39.4%)	134 (38.7%)	20 (35.1%)	18 (52.9%)	0.47
Disease activity						
CHAQ [2]	427	0.12 (0, 0.63)	0 (0, 0.6) N = 336	0.25 (0, 1) N = 57	0.25 (0, 1) N = 34	<0.01
Patient global	428	1 (0, 4)	1 (0, 3) N = 337	3 (1, 4) N = 57	3 (0, 5) N = 34	0.02
Physician global	415	1 (0, 3)	1 (0, 2) N = 327	1 (0, 3) N = 54	1.5 (0, 3) N = 34	0.25
Weakness [3]	434	45 (10.4%)	30 (8.8%)	9 (15.8%)	6 (17.6%)	0.10
Rash (facial erythema)	433	137 (31.6%)	114 (33.2%)	14 (25%)	9 (26.5%)	0.37
Gottron	434	196 (45.2%)	154 (44.8%)	27 (48.2%)	15 (44.1%)	0.88
CMAS [4]	271	50 (45, 52)	50 (45, 52) N = 215	49 (43, 52) N = 33	48 (43, 52) N = 23	0.49
HRQOL [5]	424	2 (2, 3)	2 (2, 3) N = 333	3 (2, 3) N = 57	3 (2, 3) N = 34	0.02
ACR [6] class (worst)	384					0.90
I		80 (20.8%)	65 (21.5%)	9 (18.4%)	6 (18.8%)	
II		99 (25.8%)	81 (26.7%)	12 (24.5%)	6 (18.8%)	
III		90 (23.4%)	68 (22.4%)	12 (24.5%)	10 (31.3%)	

	n	All (n = 438)	White (n = 347)	Black (n = 57)	Minority, non-Black (n = 34)	p-value
IV		115 (30.0%)	89 (29.4%)	16 (32.7%)	10 (31.3%)	

Parenthesis indicates percentage or median interquartile 25–75% range (IQR). Chi square testing was used for comparison of counts; Kruskal-Wallis nonparametric testing was used for comparison of ordinal results. The p-values are for the comparisons between the White group and the other two groups.

[1]Cytotoxic therapy=cyclophosphamide (pulse or oral)

[2]CHAQ= Childhood Health Assessment Questionnaire, visit (higher is worse

[3]Weakness = moderate or severe proximal weakness by evaluating physician, visit

[4]CMAS= Childhood Myositis Assessment Scale, visit (higher is better)

[5]HRQOL= Health Related Quality of Life, visit

[6]ACR= American College of Rheumatology

Table II

Patient characteristics according to reported annual family income

Subject characteristics	**<$50,000/yr (n = 128)**	**>$50,000/yr (n = 218)**	**p-value**
Demographics			
Age at onset	6.5 (3.9, 10.1) N = 124	5.4 (3.4, 8.7) N = 216	0.13
Female	90/128 (70.3%)	153/218 (70.2%)	0.98
Time to Diagnosis (months)	4.9 (2.0, 12.1) N = 122	4.3 (2.0, 10.1) N = 216	0.28
Race			
White	84 (65.6%)	193 (88.5%)	<0.01
Black	32 (25.0%)	11 (5.1%)	
Minority, non-Black	12 (9.4%)	14 (6.4%)	
Patient characteristics			
Lipodystrophy	6/127 (4.7%)	9/217 (4.2%)	0.80
Calcinosis	23/123 (18.7%)	23/215 (10.7%)	0.04
ANA	61/97 (62.9%)	122/181 (67.4%)	0.45
Treatments			
IV/Pulse corticosteroids	73/121 (60.3%)	130/199 (65.3%)	0.10
Oral corticosteroids	118/122 (96.7%)	197/202 (97.5%)	0.12
DMARDS/non-biologic DMARDS	120/128 (93.8%)	199/218 (91.3%)	0.41
Cytotoxic therapy[1]	3/119 (2.5%)	1/196 (0.5%)	0.12
Biologics	61/128 (47.7%)	86/218 (39.5%)	0.14
IVIG	55/128 (43.0%)	83/217 (38.5%)	0.20
Disease activity			
CHAQ[2]	0.38 (0, 1.00) N = 125	0 (0, 0.38) N = 215	<0.01
Patient global	3 (1, 5) N = 127	1 (0, 3) N = 214	<0.01
Physician global	1 (0, 3) N = 125	1 (0, 2) N = 207	0.02
Weakness[3] (moderate/severe)	13/126 (10.3%)	19/218 (8.7%)	0.03

Subject characteristics	**<$50,000/yr (n = 128)**	**>$50,000/yr (n = 218)**	**p-value**
Rash (facial erythema)	38/126 (30.2%)	65/217 (30.0%)	0.97
Gottron	59/127 (46.5%)	96/217 (44.2%)	0.69
CMAS[4]	49 (43, 52) N = 76	50 (45, 52) N = 141	0.44
HRQOL[5]	3 (2, 3) N = 127	2 (1, 3) N = 213	<0.01
ACR[6] class (worst)			0.05
I	17 (15.2%)	44 (22.8%)	
II	26 (23.2%)	52 (26.9%)	
III	30 (26.8%)	43 (22.3%)	
IV	39 (34.8%)	54 (28.0%)	

Parenthesis indicates percentage or interquartile 25–75% range (IQR). Chi square testing was used for comparison of counts; Kruskal-Wallis nonparametric testing was used for comparison of ordinal results. The p-values are for the comparisons between the two groups.

[1]Cytotoxic drugs = cyclophosphamide (pulse or oral)

[2]CHAQ = Childhood Health Assessment Questionnaire (higher is worse)

[3]Weakness = moderate or severe proximal weakness by evaluating physician

[4]CMAS = Childhood Myositis Assessment Scale (higher is better)

[5]HRQOL = Health Related Quality of Life

[6]ACR= American College of Rheumatology

Table III

Associations of risk factors and identification of calcinosis, multivariable analysis

Variable	Odds ratio	95% CI	p-value
Black race	2.42	1.11, 5.27	0.026
Time to diagnosis (months)	1.02	1.00, 1.04	0.017
Sex (female)	0.55	0.30, 1.02	0.057
Length of illness (years)	1.22	1.12, 1.33	<0.001

* Overall model shows a p-value<0.01 with R^2 =12.5%. R^2 is a statistical measure of how close the data are to the fitted regression line.