Race, Income, and Disease Outcomes in Juvenile Dermatomyositis

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Objective To determine the relationships among 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, annual family 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 annual family income and significantly worse scores on measures of physical function, disease activity, and quality of life measures. Subjects with lower annual family income 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 among the racial groups in time to diagnosis or duration of disease. Using calcinosis as a marker of disease morbidity, black race, annual family income <$50 000 per year, negative antinuclear antibody, and delay in diagnosis >12 months were associated with calcinosis.

Conclusion Minority race and lower family income are associated with worse morbidity and outcomes in subjects with JDM. Calcinosis was more common in black subjects. Further studies are needed to examine these associations in more detail, to support efforts to address health disparities in subjects with JDM and improve disease outcomes. *(J Pediatr 2017;184:38-44).*

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

With improvements in treatment, survival in patients with JDM has improved over the past several decades, rising from 67% to 99%.[6,7] However, although survival 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.[7-9]

The factors that contribute to the development of long-term morbidity in JDM are unknown, but an analysis 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 that 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 affects outcomes has not been examined in depth in the JDM population. Determining whether race and income are associated with increased morbidity is essential to identifying and addressing disparities through, for example, improved access to pediatric rheumatologists, improved insurance coverage, or increased education of primary care physicians, which may facilitate the diagnosis of JDM.

ACR	American College of Rheumatology
ANA	Antinuclear antibody
CARRA	Childhood Arthritis and Rheumatology Research Alliance
CHAQ	Childhood Health Assessment Questionnaire
CMAS	Childhood Myositis Assessment Scale
HRQoL	Health-related quality of life
EMG	Electromyography
JDM	Juvenile dermatomyositis
MRI	Magnetic resonance imaging

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Methods

The CARRA Legacy Registry is a multicenter registry developed in 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. It includes subjects with diagnosed rheumatic diseases from 55 pediatric CARRA centers in North America between May 2010 and July 2014. It is an observational data capture registry representing a convenience sample of eligible subjects. Institutional Review Board approval was obtained at each enrolling site. Each subject and/or a parent/legal guardian provided informed consent (in English or Spanish).

Subjects were eligible for inclusion who were aged <21 years with onset of JDM before age 18 years and met the 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, to reflect common practice and increase inclusion. Subjects could be at any disease stage and were not required to be recently or newly diagnosed. Subjects were excluded from our analysis if age of onset could not be clearly identified as before 18 years or if they did not meet the 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: muscle weakness, elevated muscle enzyme levels, abnormal EMG suggestive of inflammatory myopathy, abnormal muscle biopsy suggestive of inflammatory myopathy, or MRI evidence of myositis (modification introduced by CARRA Legacy Registry investigators[21]).

Data Collection

Clinical data were collected from the subjects/guardians and the enrolling physicians 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 before May 2010 were recruited retrospectively. Data were pooled and stored in a secure centralized electronic database and deidentified before analysis.

Demographic and Baseline Information

Baseline demographic information was obtained by the enrolling physician. Family income was based on 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 coinsurance.

Racial/ethnic background was self-identified by the subject or parent/guardian during enrollment. For the purpose of this study, subjects who self-identified as white and not biracial were classified as white, those who self-identified as black or black and biracial were classified as black, and all others were classified as minority, nonblack. Subjects without race identified were dropped from the analysis (n = 3). Treatment history was recorded by the enrolling physician from chart abstraction or other records. Disease duration at enrollment was calculated as age at onset of symptoms subtracted from the age at enrollment. A delay in diagnosis was as >12 months from the onset of symptoms to the first appointment with a pediatric rheumatologist. Annual 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 US family of 4 with 2 children.[22]

Outcome Measures

For each subject, data were collected on muscle strength, physical functioning, and quality of life. The Childhood Myositis Assessment Scale (CMAS) score (maximum score, 52), the Childhood Health Assessment Questionnaire (CHAQ) score (ranging from 0, normal to 3, worst), health-related quality of life (HRQoL) measures, the American College of Rheumatology (ACR) functional class rating, and global disease assessments were also recorded. The CMAS has been validated to quantitatively assess muscle strength and endurance in children with idiopathic inflammatory myopathies.[21,23,24] The ACR functional class rating ranges from class I (able to perform usual activities of daily, living including self-care, vocational, and avocational activities) to class IV (limited ability to perform usual self-care, vocational, and avocational activities).[25] The ACR class was reported by the enrolling physician as the highest (ie, worst) class occurring during the course of the disease based on either 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. Calcinosis served as a surrogate marker of disease morbidity.

Statistical Analyses

Statistical analysis was conducted using Stata version 10.0 (StataCorp, Austin, Texas). 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 mean ± SD or median (IQR) for continuous variables and frequency (percentage) for categorical variables. The χ^2 and Kruskal-Wallis tests were used to evaluate associations among demographic characteristics, income, treatments, and disease activity with disease duration, race, and income. All tests were 2-sided, with a P value <.05 considered to indicate statistical significance.

We performed multivariate logistic regression of the outcome calcinosis, which served as a surrogate marker of disease morbidity. Variables for inclusion in the multivariate model were selected based on results from bivariate testing following a stepwise approach, with previously associated variables, such as duration of disease, sex, and delay to treatment, selected automatically. Variables significantly ($P < .10$) associated with calcinosis (ie, race and income) and static variables associated with race and income (ie, antinuclear antibody [ANA] positivity) were included in the model. If the variable did not

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