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Trends in Dermatopolymyositis Mortality, 1999–2022: A Nationwide Population-Based Study, United States

Elizabeth Matz and Ram R. Singh

Objective. We evaluated trends in dermatopolymyositis (DPM) mortality relative to all-cause mortality in the United States from 1999 to 2022.

Methods. We used the Centers for Disease Control and Prevention's databases (Multiple Causes of Death for 1999–2020 and Provisional Mortality Statistics for 2021 and 2022) to obtain death counts for DPM and non-DPM (all causes other than DPM). We calculated age-standardized mortality rates (ASMRs) for both groups and computed the ratio of DPM-ASMR to non–DPM-ASMR for each of the 24 years. We performed joinpoint regression analysis to estimate annual percent change (APC) in DPM-ASMRs and non–DPM-ASMRs and in the DPM-ASMR to non–DPM-ASMR ratios overall and by sex, age, and race and ethnicity.

Results. There were 12,882 DPM and 63,549,485 non-DPM deaths during 1999–2022. Mortality decreased at a higher APC (–3.8% [95% confidence interval (CI) –4.3% to –3.4%]) for DPM than for non-DPM (–1.2% [95% CI –1.5% to –0.9%]) until the start of the COVID-19 pandemic, when it increased for both at similar APCs. Consequently, the ratios of DPM-ASMRs to non–DPM-ASMRs decreased over these 24 years in all subgroups. The DPM-ASMR to non–DPM-ASMR ratios were higher in females than males and in younger individuals (≤64 years) than those aged ≥65 years. The odds of premature death were higher for DPM than for non-DPM. Individuals of non-Hispanic Black, Hispanic, and non-Hispanic other race and ethnicity had higher DPM-ASMR to non–DPM-ASMR ratios than White individuals.

Conclusion. DPM mortality decreased at a higher rate than all-cause mortality until the pandemic, when it proportionately increased for both DPM and all causes. Females, younger individuals, and individuals of Black, Hispanic, and non-Hispanic other race and ethnicity had higher DPM mortality relative to all-cause mortality. Findings highlight the need for improved screening, earlier intervention, and targeted efforts to address racial and ethnic disparities in DPM outcomes.

INTRODUCTION

Dermatopolymyositis (DPM) comprises chronic autoimmune inflammatory diseases that involve the muscles, skin, lungs, esophagus, heart, joints, and other organs.[1–4] DPM can lead to premature death due to active disease manifestations, including muscle weakness, lung and heart disease, adverse effects from treatment (including infections), and cancers.[5,6] The five-year mortality rates for DPM were as high as 23% to 73% before the introduction of glucocorticoids for its treatment.[6] Survival for DPM has improved since then, but initial treatment with high-dose glucocorticoids was also associated with lower survival[7] from 1997 to 2003. DPM outcomes have improved further with the fine-tuning of glucocorticoids and other immunosuppressive regimens, early recognition of high-risk disease features, and increasing use of intravenous Ig.[8,9] Assessing the impact of these advances on DPM mortality burden at the population level will further help improve DPM outcomes.

In a Michigan cohort of 160 patients, male sex and race and ethnicity other than White race were associated with lower survival.[7] However, mortality was not influenced by sex or race and ethnicity in a retrospective analysis of 831 patients with DPM from 2006 through 2014 at the Johns Hopkins Myositis Center.[10] Although the cohort-based studies provide useful information,

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SIGNIFICANCE & INNOVATIONS

- Using the largest dermatopolymyositis (DPM) death counts (12,882) to date, this study reports decreasing DPM mortality across the United States over the last quarter-century.
- This is the first study to assess DPM mortality in the context of all-cause mortality, reporting that DPM mortality decreased at a higher rate than all-cause mortality over a quarter-century.
- This is the first population-based study to compare DPM mortality before and during the COVID-19 pandemic, and it shows that the increase in DPM mortality during the pandemic was proportionate to the increase in all-cause mortality.
- Females, younger individuals, and individuals of non-Hispanic Black, Hispanic, and non-Hispanic other race and ethnicity had higher DPM mortality relative to all-cause mortality, and the odds of premature death were higher for DPM than for all other causes of death.

they may not represent the burden of disease at the population level.

Population-based data on DPM mortality are scarce in the United States. An early study of 1,986 DPM deaths from 1968 through 1978 revealed a higher age-adjusted mortality rate in race and ethnicity other than White females than in White females and both race and ethnicity other than White and White males.[11] Another study[12] retrieved DPM death counts from the US national mortality database from 1981 to 2020, but the association of DPM mortality with age and race and ethnicity was not explored. A recent population-based study of dermatomyositis in Olmsted County reported nine deaths from 1995 to 2019, giving the standardized mortality ratio of 2.0 (95% confidence interval [CI] 0.9–3.7).[13] However, a small sample size, particularly of individuals of Native American, Asian, Black, Hispanic, and Pacific Islander race and ethnicity or of male sex, may have limited its utility in assessing DPM burden in the US population.[11,13]

Here, we analyzed all recorded deaths in the United States from 1999 through 2022 to evaluate temporal trends in DPM mortality overall and by sex, age, and race and ethnicity. To evaluate DPM mortality in the context of changes in general population mortality, we calculated the ratios of age-standardized mortality rates (ASMRs) for DPM to ASMRs for all causes other than DPM (non-DPM).

METHODS

Data source. Annual mortality data were obtained using the Centers for Disease Control and Prevention's (CDC's) Wide-ranging Online Data for Epidemiologic Research (WONDER) databases.[14] Death counts for DPM were retrieved from the Multiple Causes of Death (MCD) 1999–2020 database using *International Classification of Diseases, Tenth Revision* codes (Supplemental Table S1). Data for 2021 and 2022 were retrieved from MCD Provisional Mortality Statistics 2018–2022 and merged with the 1999–2020 data. Average population data were obtained from the US Census Bureau.

ASMR. Age-specific crude mortality rates for DPM and non-DPM were calculated as the number of deaths in each year divided by the number of persons in the US general population in that same year. The US population from the year 2000 was used to calculate ASMR per million, with 95% CIs,[15] overall as well as separately for males and females, for each of the four racial and ethnic groups, and for three age groups (≤44, 45–64, and ≥65 years) for each year from 1999 through 2022. Racial and ethnic groups included Hispanic, non-Hispanic Black, non-Hispanic White, and non-Hispanic other (Asian, Pacific Islander, American Indian, or Alaska Native). We then computed the ratios of DPM-ASMR to non–DPM-ASMR for each year from 1999 through 2022 overall and separately for each sex, age, and racial and ethnic subgroup. We also conducted a post hoc analysis comparing odds of premature death in DPM with two other autoimmune diseases: systemic lupus erythematosus (SLE) and systemic sclerosis (SSc).

Statistical analysis. The joinpoint regression model was used to evaluate mortality trends overall and by sex, age, and race and ethnicity. Joinpoint regression identifies changes in trend data by fitting a set of joinpoints—the calendar years at which the change in the slope (of ASMR or ratio) is statistically significant—over the entire period. The model then computes the slope (year-to-year percentage change in annual ASMR or ratio) and the 95% CI over each linear trend segment between adjacent joinpoints.[16,17]

The joinpoint regression model assumes that its regression mean function is piecewise linear and that the linear segments are continuously connected at the joinpoints. The program allows one of three options for handling heteroscedastic errors: constant variance (homoscedasticity), SE, or Poisson variance. The program uses one of two methods for model fitting: the grid search method or the Hudson method. The grid search method identifies a discrete number of locations for testing for changes in slope, and the Hudson method allows for continuous model fitting. For this study, we provided the SE and used the grid search method.

The Joinpoint Regression Program (version 4.2.0.2; National Cancer Institute) uses a permutation test to find the optimal number of joinpoints. For greater consistency in the permutation test *P* values, the Joinpoint Regression Program runs at least 4,499 permutations to select the optimal number of linear segments to fit the model. Because fitting all 4,499! (factorial) possible permutations would be computationally intensive, the program uses a

Monte Carlo simulation to conduct significance tests on a sample of the 4,499! permutations, which are adjusted using the Bonferroni correction to reduce the likelihood of false positive results.[18] The study is exempt from ethics committee approval requirement.

RESULTS

From 1999 through 2022, DPM was recorded as the underlying or a contributing cause of death in 12,882 decedents. Females (64.7%), non-Hispanic Black individuals (17.4%), and those in the 45 to 64 years age group (28.0%) were overrepresented among DPM decedents relative to their representation among non-DPM (all causes other than DPM) decedents (Table 1).

Trends in mortality: DPM versus non-DPM (all cause), 1999–2022. Mortality rates decreased for both DPM and non-DPM from 1999 through 2022 but at a higher pace for DPM (annual percent change [APC] –3.8 [95% CI –4.3 to –3.4]) than for non-DPM (APC –1.2 [95% CI –1.5 to –0.9]) until the start of the COVID-19 pandemic (Figure 1). The cumulative percent change over the entire 24-year period was –46.8% for DPM but only –4.8% for non-DPM (Supplemental Table S2). During the pandemic, mortality rates significantly increased for both DPM and non-DPM at a similar APC. Consequently, the ratio of DPM to non-DPM mortality rates continuously decreased throughout the 24-year period. This suggests that there was no disproportionate increase in DPM-specific mortality during the COVID-19 pandemic.

Trends in mortality by sex, age, and race and ethnicity: DPM versus non-DPM, 1999–2022. Females had higher DPM mortality rates than males; the converse was true for non-DPM mortality rates, which were higher for males (Figure 2). There were no significant differences in the cumulative percent changes for both DPM and non-DPM deaths by sex (Supplemental Table S2).

Similar mortality trends were observed at different age groups for DPM and non-DPM, with the highest ASMRs in the ≥65 years age group for both DPM and non-DPM (Figure 3). However, the ratio of DPM-ASMRs to non–DPM-ASMRs was higher in the 45 to 64 and ≤44 years age groups than in those aged ≥65 years, suggesting a disproportionately higher mortality rate at younger age groups for DPM than for non-DPM. The odds of premature death (<65 years) from DPM were significantly increased compared with non-DPM (odds ratio [OR] 1.6; 95% CI 1.5–1.6), which were less than in SSc (OR 1.9; 95% CI 1.9–2.0) and SLE (OR 4.3; 95% CI 4.2–4.4) (Table 2).

The cumulative decreases in DPM mortality rates were also less in younger age groups than in older age groups (Supplemental Table S2). Notably, there was a cumulative increase of 21.1% in non–DPM-ASMRs for the ≤44 years age group, suggesting a higher premature all-cause mortality rate in 2022 than in 1999.

Mortality rates in non-Hispanic Black persons were higher than those in other racial and ethnic groups for both DPM and non-DPM causes (Figure 4). The ratio of DPM-ASMRs to non–DPM-ASMRs was higher in individuals of non-Hispanic Black, Hispanic, and non-Hispanic other race and ethnicity than in non-Hispanic White individuals. The 24-year cumulative decrease in DPM mortality rates was more than 50% for non-Hispanic Black and non-Hispanic White persons but only 8.7% for non-Hispanic Asian, Pacific Islander, American Indian, and Alaska Native individuals and 34.5% for Hispanic individuals (Supplemental Table S2).

Table 1. Demographic characteristics, 1999–2022*

Characteristics	DPM deaths[a]		Non-DPM deaths[a]		Average population[b]	
	No.	%	No.	%	No.	%
Total	12,882		63,549,485		7,410,144,137	
Sex						
Female	8,339	64.7	31,504,793	49.6	3,764,021,810	50.8
Male	4,543	35.3	32,042,977	50.4	3,646,122,327	49.2
Age						
≤44 y	1,024	8.0	4,837,629	7.6	4,508,987,499	60.8
45–64 y	3,610	28.0	12,049,566	19.0	1,860,984,067	25.1
≥65 y	8,248	64.0	46,657,324	73.4	1,040,172,571	14.0
Not stated	0	0	4,966	0.0	–	–
Race and ethnicity						
NH White	8,903	69.1	49,961,434	78.6	4,787,848,120	64.6
NH Black	2,240	17.4	7,585,215	11.9	947,648,882	12.8
Hispanic	1,128	8.8	4,028,734	6.3	1,139,927,382	15.4
NH A/PI/AN/AI	591	4.6	1,764,980	2.8	456,491,367	6.2
Not stated	20	0.2	209,122	0.3	78,228,386	1.1

* A, Asian; AI, American Indian; AN, Alaska Native; Black, Black or African American; DPM, dermatopolymyositis; Hispanic, Hispanic or Latino; NH, non-Hispanic; non-DPM, all-cause deaths other than DPM; PI, Pacific Islander.
[a] Number (percentage) of deaths from all 50 states and the District of Columbia as of September 14, 2023, in all categories except for total and sex in non-DPM (63,547,770 as of September 7, 2023).
[b] Mean annual population (percentage) derived from US Census Bureau files.

DISCUSSION

Analysis of all recorded deaths attributed to DPM in the United States from 1999 through 2022 revealed that mortality rates have decreased at a higher pace for DPM than for all-cause (non-DPM) deaths. There were significant differences between DPM and all-cause mortality by sex, age, and race and ethnicity. DPM was associated with higher odds of premature death compared to all-cause deaths. During the pandemic, mortality rates increased for both DPM and all causes at similar APCs; thus, the ratios of DPM to all-cause mortality rates continuously decreased throughout the 24-year period in all sex, age, and racial and ethnic groups, suggesting no disproportionate increase in DPM-associated mortality during the pandemic.

A few population-based studies have examined the changes in DPM mortality over time. An early study[11] reported increased DPM mortality rates in the United States from 1968 to 1978. A 1999–2014 study[19] of 303 DPM deaths from British Columbia, Canada, revealed a 2.4- to 5.5-fold increased risk of DPM death from 1997 to 2005 compared to a 2.0- to 4.1-fold increased risk of DPM death from 2006 to 2014. A 1999–2014 United Kingdom study of 114 DPM deaths did not report a significant improvement in excess mortality risk in DPM in recent years (2007–2014 vs 1999–2006).[20] However, we report that the mortality rates decreased at higher annual percent rates for DPM than for all-cause deaths from 1999 through 2018 in the US population.

The improving DPM mortality rate over the last two decades is likely attributable to a combination of factors (Figure 1, bottom panel),[2,3,8,21] including: (1) the 2003 European Neuromuscular Centre criteria, which updated the Bohan and Peter criteria by adding more muscle pathology details as well as incorporating amyopathic dermatomyositis[22]; (2) the increasing awareness

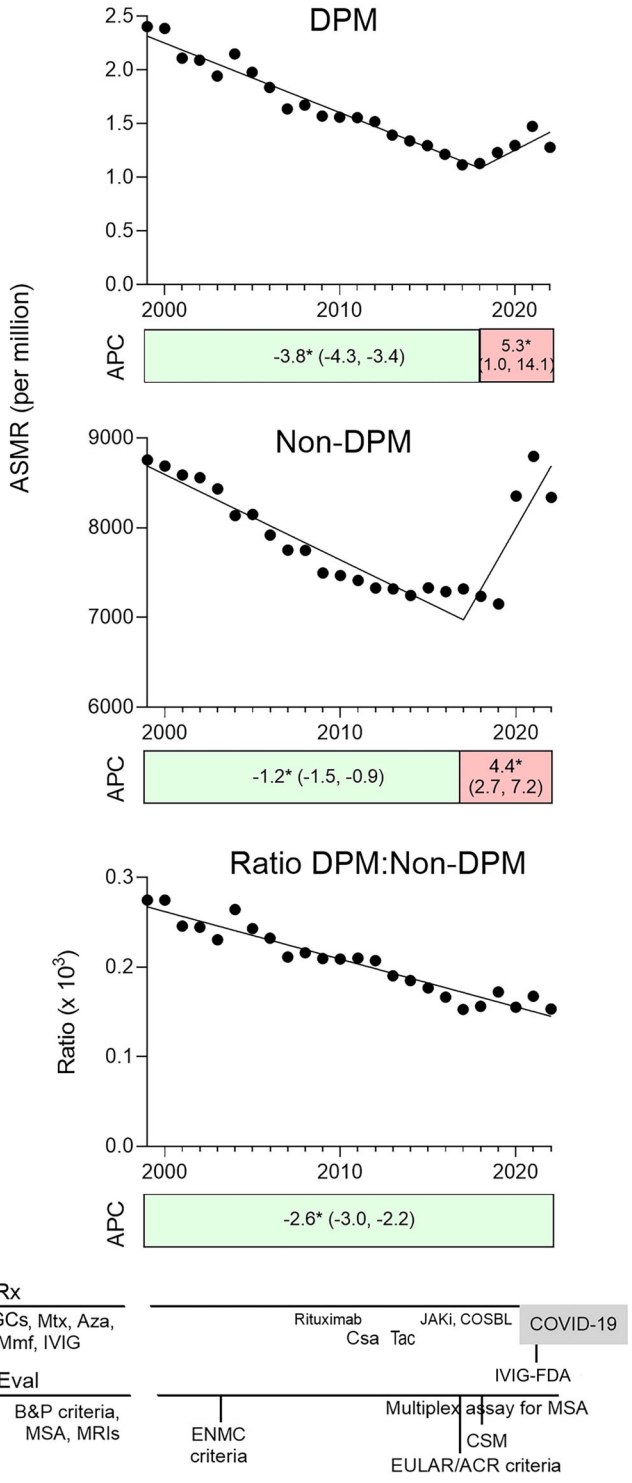

Figure 1. Legend on next page.

Figure 1. Trends in mortality rates for DPM and non-DPM causes and ratio of DPM to non-DPM mortality rates, 1999–2022. Results are expressed as the ASMR per million persons for DPM (top panel) and non-DPM (middle panel) and the ratio of DPM to non-DPM mortality rates (lower panel). Data are displayed per calendar year of death, with lines fitted on the basis of joinpoint trend analysis. The annual number of deaths ranged from 428 to 664 for DPM and from 2,391,374 to 3,464,216 for non-DPM. The APC for each trend is presented as stack bars below each panel. Each stack is segmented at the year in which the change in slope is statistically significant and is aligned with the trend line. Numbers in each stack denote the APC (95% confidence interval). The red-shaded stacks indicate an increasing trend, and the green-shaded stacks represent a decreasing trend. *$P < 0.05$ for slope change. (Bottom panel) DPM-ASMR is shown in relation to major therapeutic developments (Rx), advances in evaluation (Eval) of DPM, and the COVID-19 pandemic. ACR, American College of Rheumatology; APC, annual percent change; ASMR age-standardized mortality rate; Aza, azathioprine; B&P criteria, Bohan and Peter classification criteria; COSBL, T cell costimulation blockade; Csa, cyclosporine A; CSM, core set measures; DPM, dermatopolymyositis; ENMC, European Neuromuscular Centre; GCs, glucocorticoids; IVIG, intravenous Ig; IVIG-FDA, IVIG treatment approved by the Food and Drug Administration; JAKi, JAK inhibitors; Mmf, mycophenolate mofetil; MRI, magnetic resonance imaging; MSA, myositis-specific autoantibodies; Mtx, methotrexate; non-DPM, all-cause deaths other than DPM; Rx, therapeutic advances; Tac, tacrolimus.

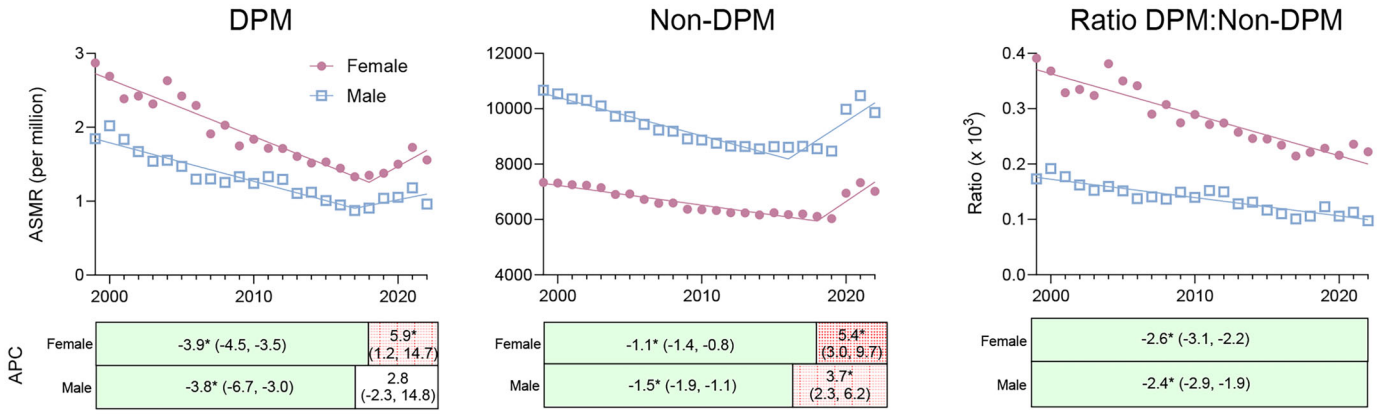

Figure 2. Trends in mortality rates for DPM and non-DPM and in the ratio of DPM mortality rates to non-DPM mortality rates by sex, 1999–2022. Results are expressed as the ASMR per million persons for DPM (left panel) and non-DPM (middle panel) and the ratio of DPM to non-DPM mortality rates (right panel). Data are displayed per calendar year of death, with lines fitted on the basis of joinpoint trend analysis. The annual number of DPM deaths ranged from 277 to 445 for females and from 151 to 240 for males. The APC for each trend for each subpopulation is presented as stack bars below each panel. Each stack is segmented at the year in which the change in slope is statistically significant and is aligned with the trend line. Numbers in each stack denote the APC (95% confidence interval). The red-shaded stacks indicate an increasing trend, unshaded stacks represent a nonsignificant trend, and the green-shaded stacks represent a decreasing trend. *$P < 0.05$ for slope change. APC, annual percent change; ASMR, age-standardized mortality rate; DPM, dermatopolymyositis; non-DPM, all-cause deaths other than DPM. Color figure can be viewed in the online issue, which is available at http://onlinelibrary.wiley.com/doi/10.1002/acr.25609/abstract.

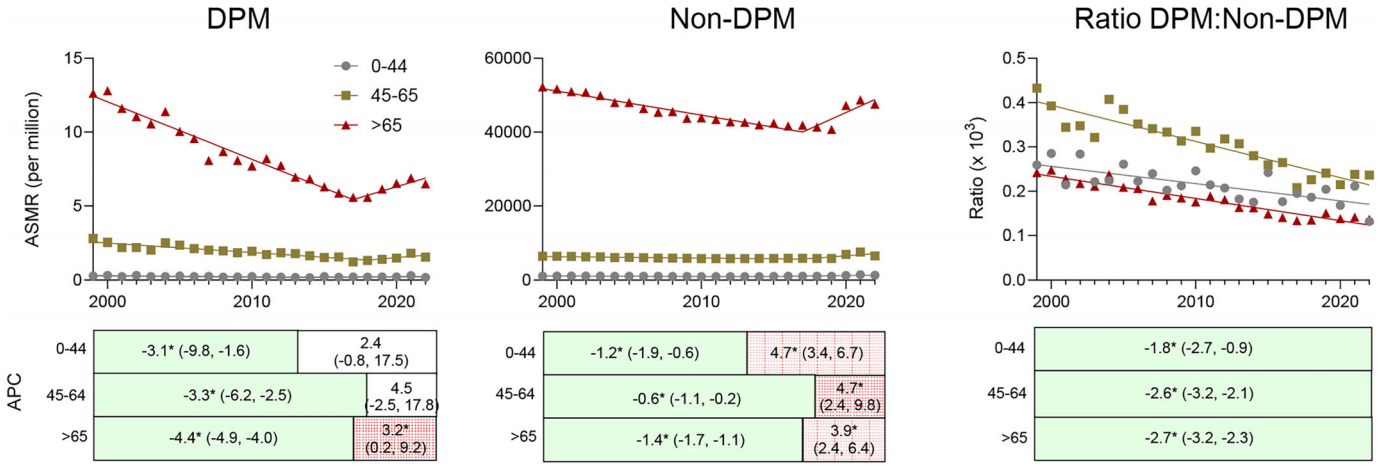

Figure 3. Trends in mortality rates for DPM and non-DPM and in the ratio of DPM mortality rates to non-DPM mortality rates by age, 1999–2022. Results are expressed as the ASMR per million persons for DPM (left panel) and non-DPM (middle panel) and the ratio of DPM to non-DPM mortality rates (right panel). Data are displayed per calendar year of death, with lines fitted on the basis of joinpoint trend analysis. The annual number of DPM deaths ranged from 33 to 57 for those aged ≤44 years, from 112 to 183 among those aged 45 to 64 years, and from 275 to 447 among those aged ≥65 years. The APC for each trend for each subpopulation is presented as stack bars below each panel. Each stack is segmented at the year in which the change in slope is statistically significant and is aligned with the trend line. Numbers in each stack denote the APC (95% confidence interval). The red-shaded stacks indicate an increasing trend, unshaded stacks represent a nonsignificant trend, and the green-shaded stacks represent a decreasing trend. *$P < 0.05$ for slope change. APC, annual percent change; ASMR, age-standardized mortality rate; DPM, dermatopolymyositis; non-DPM, all-cause deaths other than DPM. Color figure can be viewed in the online issue, which is available at http://onlinelibrary.wiley.com/doi/10.1002/acr.25609/abstract.

and earlier recognition of interstitial lung disease as a manifestation of DPM[23,24]; (3) the recognition of myositis-specific antibodies as valuable tools in diagnosing and understanding DPM[25]; (4) the increased availability and reliability of myositis antibody testing, including the development of multiplex assays for testing[25]; (5) the development of DPM core set measures[26] and the 2017 EULAR/American College of Rheumatology criteria[42]; (6) earlier use of disease-modifying antirheumatic drugs, for example, methotrexate, azathioprine, and mycophenolate mofetil, with a more rapid tapering of glucocorticoids[21]; (7) increasing use of intravenous Ig[9,27]; (8) introduction of rituximab and the calcineurin inhibitors tacrolimus and cyclosporine A[21,28–31]; and (9) use of JAK inhibitors and costimulation blockers in patients refractory to current treatments.[21,32] Better management of comorbidities

Table 2. Odds of premature death, 1999–2022*

Age, y	DPM, n (%)	Non-DPM, n (%)	OR[a] (95% CI)	SSc, n (%)	Non-SSc, n (%)	OR[a] (95% CI)	SLE, n (%)	Non-SLE, n (%)	OR[a] (95% CI)
<65	4,634 (36.0)	16,887,195 (26.6)	1.6[b] (1.5–1.6)	17,654 (41.0)	16,885,593 (26.6)	1.9[b] (1.9–2.0)	32,009 (60.8)	16,884,595 (26.6)	4.3[b] (4.2–4.4)
≥65	8,248 (64.0)	46,657,324 (73.4)	1 (ref)	25,392 (59.0)	46,655,625 (73.4)	1 (ref)	20,659 (39.2)	46,656,511 (73.4)	1 (ref)

* CI, confidence interval; DPM, dermatopolymyositis; non-DPM, all causes other than DPM; OR, odds ratio; ref, reference; SLE, systemic lupus erythematosus; SSc, systemic sclerosis.
[a] Odds of premature death (≤65 y) from DPM, SSc, or SLE relative to all-cause (non-DPM, non-SSc, or non-SLE, respectively) deaths.
[b] $P < 0.0001$.

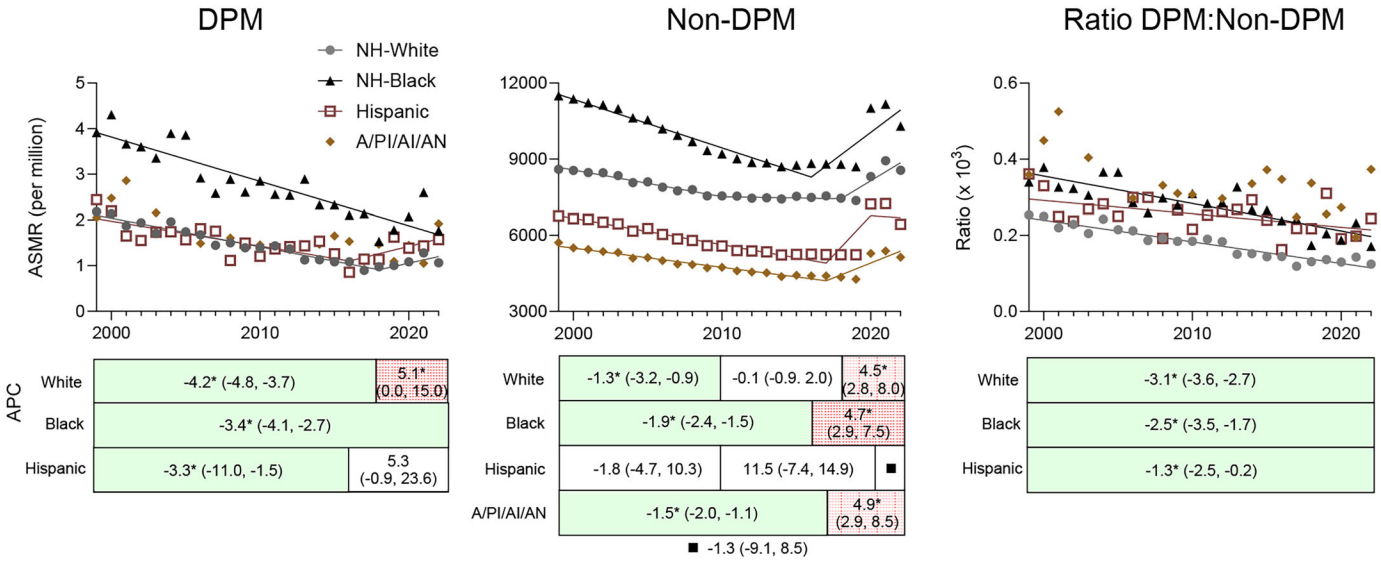

Figure 4. Trends in mortality rates for DPM and non-DPM and in the ratio of DPM mortality rates to non-DPM mortality rates by race and ethnicity, 1999–2022. Results are expressed as the ASMR per million persons for DPM (left panel) and non-DPM (middle panel) and the ratio of DPM to non-DPM mortality rates (right panel). Data are displayed per calendar year of death, with lines fitted on the basis of joinpoint trend analysis. The annual number of DPM deaths ranged from 270 to 491 for NH White individuals, from 61 to 116 for NH Black individuals, from 33 to 76 for Hispanic individuals, and from 14 to 46 for NH A/PI/AI/AN individuals. The APC for each trend for each subpopulation is presented as stack bars below each panel. Each stack is segmented at the year in which the change in slope is statistically significant and is aligned with the trend line. Numbers in each stack denote the APC (95% confidence interval). The red-shaded stacks indicate an increasing trend, unshaded stacks represent a nonsignificant trend, and the green-shaded stacks represent a decreasing trend. *$P < 0.05$ for slope change. Stack bars are not shown for NH A/PI/AI/AN under DPM and ratio trendline curves because the annual death counts were low in this group. A, Asian; AI, American Indian; AN, Alaska Native; APC, annual percent change; ASMR, age-standardized mortality rate; Black, Black or African American; DPM, dermatopolymyositis; Hispanic, Hispanic or Latino; NH, non-Hispanic; non-DPM, all-cause deaths other than DPM; PI, Pacific Islander. Color figure can be viewed in the online issue, which is available at http://onlinelibrary.wiley.com/doi/10.1002/acr.25609/abstract.

including ischemic heart disease and malignancy, which are common causes of death in DPM,[5,6] could also have contributed to the decreasing DPM mortality rate. The decreasing mortality rate is not likely due to decreasing prevalence, as a recent estimate from Olmsted County suggested a relatively higher prevalence of dermatomyositis (13 per 100,000) than previous prevalence estimates,[13] which ranged from 1.2 to 9.2 per 100,000.

Mortality was not influenced by sex in the Johns Hopkins Myositis cohort,[10] whereas an association between male sex and lower survival was reported in the Michigan cohort.[7] Consistent with the known higher prevalence of DPM in females,[33] DPM mortality rates were consistently 1.6-fold higher in females than males: 2.9 versus 1.8 (1999) and 1.6 versus 1.0 (2022), respectively (Supplemental Table S2). Because DPM is two to three times more prevalent in females than males, with an adjusted OR of 2.52 (95% CI 1.08–3.99),[33] and mortality rates were only 1.6-fold higher in females, it is likely that males would have a higher case fatality rate than females when adjusted for prevalence.

We also found that DPM mortality rates were higher in non-Hispanic Black individuals than in non-Hispanic White individuals throughout the 24-year period. In Hispanic individuals and those of non-Hispanic other race and ethnicity (Asian, Pacific Islander, American Indian, Alaska Native), DPM mortality rates were not

different relative to non-Hispanic White persons in 1999. However, Black and White individuals had a more than 50% cumulative decrease in DPM mortality rates over the 24-year period, whereas those of non-Hispanic other race and ethnicity had only an 8.7% cumulative decrease. Thus, DPM mortality rates were significantly higher in all individuals of non-Hispanic Black, Hispanic, and non-Hispanic other race and ethnicity relative to non-Hispanic White individuals in 2022. Previous studies have reported either no influence of race and ethnicity on DPM mortality[10] or an association of race and ethnicity other than White race with lower survival from DPM.[7] Poorer clinical outcomes have also been reported in Black and minority non-Black children with dermatomyositis compared with White children.[34] Multiple factors likely contribute to these disparities. For example, diagnostic delay of DPM, which has been linked to the lack of access to specialists, has extensive impacts on quality of life with this disease.[35] Significantly prolonged time to diagnosis and treatment of dermatomyositis occurs in all other races when compared with White patients, suggesting a lack of dermatology education of race and ethnicity other than White skin in DPM detection.[36] Another study reported that negative beliefs about biologic therapies were significantly more prevalent (60%) in patients of other ethnicities compared to the White cohort (35%),[37] which could have contributed to higher mortality in individuals of other races and ethnicities compared to non-Hispanic White individuals with DPM. Further studies are needed to understand reasons underlying racial and ethnic disparities in DPM mortality.

We found the highest mortality rates in those aged ≥65 years for both DPM and all-cause deaths, which is consistent with a previous report that increased age at diagnosis was associated with lower survival.[7] However, we found that the ratio of DPM to all-cause mortality rates was higher in the 45 to 64 years and ≤44 years age groups than in those aged ≥65 years, suggesting higher premature mortality for DPM than for all-cause deaths. Consistently, the OR for premature death (≤64 years) was significantly higher for DPM relative to all-cause deaths. Studies are underway to identify causes of premature death in DPM. Our preliminary analysis suggests that myositis and interstitial lung disease were more commonly recorded as the underlying causes of death in DPM at younger ages than ≥65 years.

Notably, the all-cause (non-DPM) mortality rate was not substantially lower in 2022 than in 1999. In fact, the all-cause mortality rate at younger ages (≤44 years) has steeply increased by 4.7% each year since 2014, with a cumulative increase of 21.1% over 24 years. This worrisome increase in premature mortality in the general population has been attributed to increases in firearm-related deaths and drug overdose and poisoning among children and adolescents.[38]

About half of US rheumatologists in 2020 believed that patients with autoimmune and rheumatic diseases were at a higher risk for COVID-19, and as a result, there was a decreased use of biologics to treat these conditions during the pandemic,[39] which could have increased DPM mortality. However, we found that the overall DPM mortality increase during the pandemic was proportionate to the increase in all-cause mortality rates. We further calculated mortality rates during pandemic years as compared to three preceding prepandemic years for DPM, other autoimmune diseases with the risk of immune suppression and infection including myasthenia gravis and multiple sclerosis, and diseases with little or no concern for immune suppression (including muscular dystrophy and thyroid gland disorders). As shown in Supplemental Table S3, mortality rates increased by 15% during the pandemic for DPM as compared to 28% for myasthenia gravis, 20% for multiple sclerosis, 7% for muscular dystrophy, and 29% for thyroid gland disorders. Consistently, a retrospective analysis of the TriNetX database between January 2020 and August 2021 showed that individuals with dermatomyositis with COVID-19 had a lower risk of death in comparison to the general population with COVID-19.[40] Finally, the proportions of deaths attributed to COVID-19 as the underlying cause of death during the pandemic were different in three autoimmune disorders, ranging from 8% in multiple sclerosis to 10% in DPM and 14% in myasthenia gravis (Supplemental Table S4).

Limitations of the study using mortality databases include the lack of individualized data on clinical, laboratory, and treatment variables, as well as difficulty in ascertaining the accuracy of the physicians' coding on death certificates.[14,15,41] However, misclassification of an autoimmune disease (ie, recording an autoimmune disease as the cause of death on death certificates for decedents who did not have that autoimmune disease) is unlikely to have influenced our inferences in a substantial way.[15] Unlike administrative health care databases, where possible and probable diagnoses are often recorded, death certificates usually have the most proximate illness recorded as the cause of death. Consequently, DPM is unlikely to be recorded as a cause of death for decedents who did not have DPM. However, DPM may be underrecorded as the cause of death in some proportions of patients whose proximate causes of death were cancer, infection, and ischemic heart disease,[5,6] as suggested by a previous study that showed that DPM was recorded as the underlying (primary) cause of death in only about half of all DPM deaths and as a contributing cause of death in the other half.[12] Therefore, we used the MCD database to capture all DPM deaths across the United States in our study. A previous study showed that the survival rates were similar for dermatomyositis and polymyositis.[7] Hence, we analyzed all DPM deaths together instead of separately analyzing mortality for individual DPM subsets, which might differ. Finally, inclusion of data from 2020 to 2022 allowed us to assess the impact of the COVID-19 pandemic on DPM mortality. However, data for the years 2021 and 2022 were retrieved from the Provisional Mortality Statistics database, which may undergo minor updates when finalized.

Nevertheless, our study provides an unbiased assessment of population-based burden of annual DPM mortality using a large

sample size. Such mortality statistics from national datasets serve as important indicators of the disease-specific burden at the population level, with implications for health policy planning and resource allocation. Use of direct age standardization to calculate annual mortality rates and use of joinpoint regression as a computational approach to identify long-term mortality trends are other strengths of our approach. Finally, computation of the ratio of DPM to all-cause (non-DPM) mortality to assess DPM mortality in the context of overall changes in mortality in the general population is another unique strength of this study.

In conclusion, we report a steady decrease in DPM mortality relative to all-cause mortality over the last two decades. It is also encouraging that there was no disproportionate increase in DPM-specific mortality during the COVID-19 pandemic. However, significantly higher odds of premature death from DPM are concerning, highlighting the need to identify its causes, improve screening to facilitate early diagnosis, and earlier intervention. Studies are needed to determine if more aggressive screening and treatments at early stages of active myositis and interstitial lung disease will reduce the risk of premature death in DPM. A persistently higher DPM mortality rate in non-Hispanic Black individuals also warrants targeted efforts to identify high-risk subpopulations and evaluate socioenvironmental and biologic determinants of adverse DPM outcomes in these populations. Finally, large-scale, population-based, prospective studies are needed to verify differences in DPM mortality by race and ethnicity and age and understand causes underlying such differences, which, along with targeted public health resource allocations, may reduce disparities in DPM outcomes.

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AUTHOR CONTRIBUTIONS

Both authors contributed to at least one of the following manuscript preparation roles: conceptualization AND/OR methodology, software, investigation, formal analysis, data curation, visualization, and validation AND drafting or reviewing/editing the final draft. As corresponding author, Dr Singh confirms that all authors have provided the final approval of the version to be published and takes responsibility for the affirmations regarding article submission (eg, not under consideration by another journal), the integrity of the data presented, and the statements regarding compliance with institutional review board/Declaration of Helsinki requirements.

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