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Exploring the clinical profiles and management of juvenile dermatomyositis in Africa: a survey of African rheumatology care providers

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Abstract

Background There are limited studies of juvenile dermatomyositis (JDM) in low and middle-income countries (LMIC). Many demonstrate delays to care, high prevalence of severe manifestations, and high mortality. Given the disease-associated damage with JDM, understanding JDM in Africa further is critical. Our objectives are to understand the burden of JDM in Africa and provider access to diagnostic tools and therapy through survey methodology.

Methods A survey (available in English and French) was distributed via WhatsApp to 363 total members of the African League of Associations for Rheumatology (AFLAR; $n = 233$) and Paediatric Society of the African League Against Rheumatism (PAFLAR; $n = 130$) from November 2022–January 2023. Topics included respondent specialty, number of JDM patients followed, severe manifestations, and available diagnostic tools and medications (with and without considering cost).

Results Forty-three (12%) of the 363 providers who received the survey started it. Among the 43 who started the survey, 37 (86%) provided consent and manage JDM patients; of these 37 providers, 4 (11%) partially and 16 (43%) fully completed the survey. Most were adult and/or pediatric rheumatologists ($n = 19$; 95%). Respondents represented all 5 African regions and described 216 children with JDM within the last 10 years. There was high prevalence of calcinosis (as high as 100%) and interstitial lung disease (ILD) (as high as 32%); mortality rates in Kenya (6/42; 14%) and Zambia (2/7; 29%) exceeded the 1–3% mortality reported in studies of high-income countries. Thirteen of 27 diagnostic tools and medications were accessible to $\leq 50\%$ of respondents after considering cost, mostly in Northern or Southern Africa (9/13; 69%). Despite being cost-free, disease assessment tools and physical exam to assess calcinosis were not reported as universally available or accessible.

Conclusions This is the first study to explore experiences of providers caring for children with JDM in Africa. Respondents identified 216 children with JDM seen within the last 10 years, exceeding the 196 children with JDM reported within the last 25 years but likely still underestimating prevalence. Our findings align with reports of severe

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manifestations and poor outcomes in African children with JDM. Access to many diagnostics and medications is limited, and differences in accessibility parallel regional healthcare disparities. The potential differences in JDM severity warrant systematic study and highlight the need to include patients and providers from LMIC in collaborative research efforts.

Keywords Juvenile dermatomyositis, Calcinosis, Interstitial lung disease, Low and middle-income countries, Africa

Background

There is a lack of data on pediatric rheumatic disease (PRD) in low and middle-income countries (LMIC) [1–3], and current reports are mostly limited to case studies or single center reports, with few national registries and no population-based studies [1]. While this may falsely create a perception of rarity of PRD in LMIC, the paucity of data is likely attributable to limited diagnostic capacities, scarcity of pediatric rheumatologists [4, 5], and diagnostic confusion due to limited awareness of PRD and clinical features mimicking infections and malignancy [5, 6]. Avoidable disability acquired in childhood due to PRD has lifelong medical, social, and economic burden on patients, families, and healthcare systems [1, 7].

Juvenile dermatomyositis (JDM), the most common idiopathic inflammatory myopathy affecting children, is a multisystem, autoimmune-mediated disease. JDM primarily causes proximal muscle weakness and pathognomonic cutaneous lesions [8, 9] but can also involve the joints, gastrointestinal tract, heart, lungs, and, more rarely, the kidneys, eyes, and central nervous system [10]. JDM is often a chronic, highly morbid disease with high rates of permanent damage ranging from 50 to 79% [11–13]. Due to its morbidity, understanding the burden of disease and long-term prognostic implications warrants particular attention in LMIC.

Several JDM studies have been reported in low middle-income countries [2, 3, 6, 10, 14–19], though many are descriptive small single-center reports [2, 3, 6, 14, 15, 19]. Several found higher mortality rates than those reported in North American and European studies [2, 17, 18]. There have been few African studies; the majority are single-patient case reports [19–23] or a few JDM patients among larger cohorts of other PRD [4, 5, 24]. To date, there are only three African case series from just 2 countries describing 46 South African [25, 26] and 134 Egyptian [27] children with JDM. These studies demonstrated high rates of severe disease manifestations both at diagnosis and throughout disease course, including cutaneous ulceration [27], global weakness, calcinosis, vasculitis, and pulmonary involvement [25, 26].

Calcinosis and interstitial lung disease (ILD) can be associated with severe disease in JDM, with calcinosis associated with increased morbidity [28] and ILD a risk factor for mortality [9]. The South African case series found higher rates of calcinosis and ILD compared not only to those reported in studies of the Childhood

Arthritis and Rheumatology Research Alliance (CARRA) JDM cohort [8] but also studies from other middle-income countries [3, 6, 14, 15, 17].

In high-income countries, particularly in North America and Europe, JDM outcomes have improved significantly due to early, aggressive treatment [29, 30], with mortality rates less than 1–3% [9, 31, 32], down substantially from 33% preceding corticosteroid use [9, 33]. In contrast, mortality rates in studies from LMIC range from 11% in India [2] to 8–50% in Africa [4, 25, 26]. These high mortality rates may be due to delays in diagnosis and treatment, late referrals to pediatric rheumatologists, financial constraints limiting medication adherence, and increased risk of infection [2].

While data is limited, existing reports seem to highlight severity of JDM in African patients. To our knowledge, there has not been a review of JDM across the African continent beyond reports from single centers. We suspect that existing literature does not fully capture the burden of disease. The objective of this survey is to better understand the provider experience of clinical burden of JDM in Africa and access to diagnostic tools and therapies by obtaining an overview of JDM and comparing this to data in previously published work.

Methods

Survey design and administration

An electronic, cross-sectional survey was designed by the authors using Qualtrics. It consisted of 29 questions in multiple choice, checkbox, and free-text format, with 5 additional questions to assess interest in future collaborative research efforts. Participants were excluded if they did not manage patients with JDM. To capture respondent demographic and geographic information, participants were queried about their role and practice setting. They could indicate the city and country of their clinical practice and the name of their institution; however, these questions were optional to protect anonymity. Participants were asked to indicate whether they were the only physician managing patients with JDM at their center or if there were multiple physicians to ensure that each child with JDM was included only once. Participants who were the sole physician managing children with JDM at their center answered the survey individually, accounting for all JDM patients at their center. At centers with two or more physicians managing children with JDM, participants were requested to either (a) coordinate with

other providers at their center and be the sole respondent for their center, with their responses reflective of all patients with JDM managed at their center; or (b) indicate that they answered the survey individually with their responses reflective of only patients they manage but not necessarily reflective of all patients at their center.

We aimed to understand the burden of JDM in Africa, thus participants were queried about the total number of JDM patients followed at their center currently or within the last 10 years but no longer actively followed. They were also queried about reasons for discontinuation of care. They were then asked to indicate how many of these patients achieved clinically inactive disease while on medication and remission off medications. Clinically inactive disease was defined as “lack of evidence of myositis disease activity as assessed by global and extra-muscular assessments, stable muscle strength and function, and normal muscle enzyme levels,” as per the International Myositis Assessment & Clinical Studies (IMACS) criteria for lack of evidence of active myositis [34]. Remission was defined as “clinically inactive disease while not receiving any drug therapy for a 6-month continuous period,” as per the IMACS 2005 definition [34]. In order to understand the spectrum of disease manifestations and outcomes, participants were queried about the number of patients who developed calcinosis or ILD and if any died. Respondents were queried about resources used to diagnose and/or monitor JDM generally, resources used specifically to diagnose and/or monitor calcinosis and ILD, and medications used to treat JDM. For each, respondents were first asked which resources were available at their center, regardless of access and/or cost-related issues. They were then asked to indicate which of these available resources were typically used after considering access and/or cost-related issues. Finally, respondents were queried about challenges encountered in managing children with JDM.

The survey was available in English and French given that these serve as the official and/or commonly spoken languages in 49 of the 54 African countries; in the other 5 countries (São Tomé and Príncipe, Mozambique, Guinea-Bissau, Cape Verde, and Angola), Portuguese and/or native African languages serve as the official and/or commonly spoken languages [35]. The survey was initially designed in English and translated into French using a certified medical translator. The survey was reviewed and revised by all authors prior to translation and distribution. A copy of the survey is available as Supplementary File 1.

The survey was distributed via WhatsApp groups to members of the African League of Associations for Rheumatology (AFLAR; $n=233$) and the Paediatric Society of the African League Against Rheumatism (PAFLAR; $n=130$) between November 2022 and January 2023.

WhatsApp was selected given that it is used for communication between AFLAR and PAFLAR members and allows for calculation of response rate given known number of participants in each group [36]; WhatsApp has been used for survey link distribution in several LMIC given that is a common method of communication [36–38]. AFLAR and PAFLAR members include physicians managing rheumatological diseases at the clinical and/or research level, paramedical professionals (including physiotherapists, nurses, biomedical engineers, and technicians), residents, and medical students.

Participant agreement to participate in the survey was obtained prior to initiating the survey. Survey responses remained anonymous unless participants chose to share their contact information for the purpose of future collaboration. Participation was voluntary, and no compensation was provided. Albert Einstein College of Medicine Institutional Review Board (IRB) approval was obtained (approval number 2022–13864) with waiver of informed consent.

Statistical analysis

Statistical analysis was performed using STATA software, version 17.0. Individual responses were pooled together by country. All variables were examined to identify missing data and potential data entry errors. Descriptive statistics were applied to evaluate responses, which were summarized as frequencies for categorical variables; percentages were also reported when the total number of patients was reported by all participants of a country.

Results

Respondent characteristics

Forty-three individuals started the survey (12% of the 363 AFLAR and PAFLAR members who received the survey), of whom 37 (37/43; 86%) were eligible to complete the survey (participant provided consent and manages children with JDM). Among these 37 respondents eligible to complete the survey, 11 (11/37; 30%) did not proceed further with the survey and 6 (16%) only indicated their country of practice, therefore these 17 (46%) respondents were excluded, leaving 20 (54%) respondents for the remainder of the analyses. Of these 20 respondents, 4 (20%) partially and 16 (80%) fully completed the survey. The majority of respondents included in analysis (19/20; 95%) were adult and/or pediatric rheumatologists, with 1 general pediatrician also completing the survey (5%) (Fig. 1). Most respondents practice in an academic hospital setting ($n=17$; 85%), with a smaller number practicing in community hospitals ($n=4$; 20%) and/or private clinics ($n=8$; 40%); 6 respondents (30%) practice in multiple settings and therefore total percentages of responses to this question exceed 100. Respondents represented all 5 African regions, with most from Northern Africa ($n=12$;

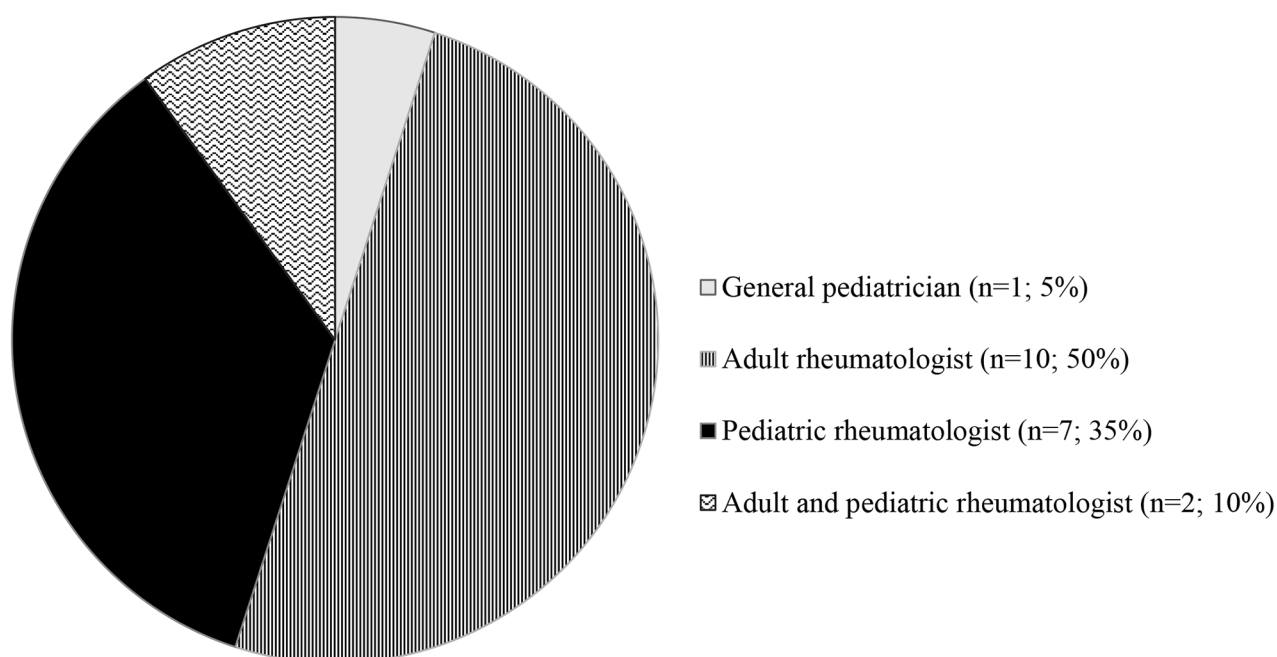


Fig. 1 Respondent specialty

60%) and smaller representation in Eastern Africa ($n=3$; 15%), Central Africa ($n=3$; 15%), Western Africa ($n=1$; 5%), and Southern Africa ($n=1$; 5%) (Fig. 2; Table 1). Six respondents (30%) were the sole providers at their center; of the remaining respondents who were not sole providers, 12 respondents (60%) completed the survey individually, answering only about their unique patients, and 2 respondents (10%) coordinated with other providers and completed the survey based on all patients at their center.

Patient characteristics

Respondents described at least 216 children with JDM within the last 10 years, of which 151 are followed currently and 65 are no longer actively followed. Several respondents from Egypt, Libya, and Tunisia were unable to provide the total number of patients followed: 3 respondents were unable to provide an estimate of the total number of children seen currently and 5 respondents were unable to provide an estimate of the total number of children seen previously within the last 10 years. In these cases, percentages were not able to be calculated. Respondents reported several reasons for discontinuation of care, including loss to follow-up (50% of respondents), death of patient (22%), patient older than the age cutoff for the center and/or was referred to an adult provider (22%), family could no longer afford medical care (17%), family moved (11%), and other reasons (22%) including remission off treatment and transfer to private clinic.

Table 1 depicts the prevalence of various clinical outcomes among respondents by country. Clinically inactive disease was achieved by 25–100% of children and remission was achieved by 21–46% of children. The prevalence of severe manifestations (defined as calcinosis, ILD, death) varied among countries. Prevalence of calcinosis ranged widely, from 0% in Nigeria to 100% in the Democratic Republic of the Congo. ILD was reported to be present in 0–32% of children. Respondents from most countries reported no patient deaths; however, 6 children (14%) in Kenya and 2 children (29%) in Zambia died. Respondents from Egypt and Libya also reported deaths, but percentages were not able to be calculated due to unknown number of total patients, and response was missing from the respondent in the Democratic Republic of the Congo (Table 1).

Table 2 shows the availability of and access to diagnostic tools and medications. Among resources used to diagnose JDM generally, irrespective of access and/or cost, all were available to at least 50% of respondents except for von Willebrand factor (vWF) antigen, which was available to only 4 respondents (22%). However, once access and/or cost were considered, all available resources were reported as typically used by fewer participants with the exception of muscle enzymes (creatinine kinase [CK], aldolase, lactate dehydrogenase [LDH], and/or aspartate aminotransferase [AST]), which were both available and accessible to all respondents. This discrepancy between availability and accessibility of diagnostic tools was also noted for all resources used specifically to diagnose and/

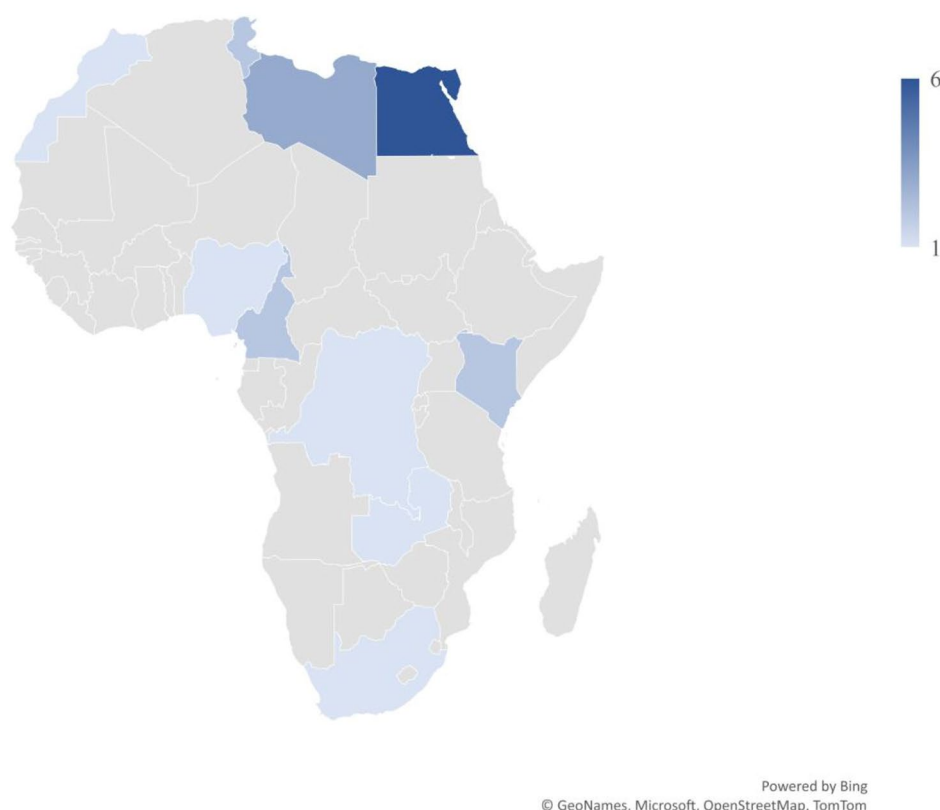


Fig. 2 Respondent country of origin

or monitor calcinosis and ILD. Oral corticosteroids and methotrexate were the only medications that were both equally available and accessible among all respondents. For all other medications, fewer respondents indicated that they were typically used after considering access and/or cost. Among the 27 diagnostic tools and medications included in the survey, 13 (48%) were accessible to $\leq 50\%$ of respondents; the majority (9/13; 69%) of these diagnostic tools or medications were only accessible in Northern (Egypt, Tunisia, Libya, Morocco) or Southern (South Africa) African countries. These are highlighted in Table 2 and include vWF antigen, myositis-specific antibodies (MSA) or myositis-associated antibodies (MAA), muscle biopsy, electromyography (EMG), magnetic resonance imaging (MRI), and ultrasound among the diagnostic resources; and hydroxychloroquine/chloroquine, cyclosporine, cyclophosphamide, rituximab, tumor necrosis factor (TNF) inhibitors, abatacept, and janus kinase (Jak) inhibitors among medications. Despite the fact that there is no cost associated with physical exam or disease assessment tools, these were not indicated as being available or accessible to all respondents.

Respondents identified several challenges in managing children with JDM. Specifically, respondents identified delayed presentation to care (12 of 16 respondents; 75%),

unfamiliarity of JDM among caregivers (11/16; 69%), lack of familiarity with JDM among other medical providers (7/16; 44%), caregivers and/or patients refused to initiate or continue treatment due to stigma surrounding JDM (4/16; 25%), and limited availability of resources – including medications (9/16; 56%), non-medical therapies such as physical and occupational therapy (5/16; 31%), and diagnostic tools (4/16; 25%). Additionally, general physicians caring for pediatric rheumatology patients expressed their own limited familiarity with JDM.

Discussion

Our survey of AFLAR and PAFLAR members identified at least 216 children with JDM cared for across the African continent within the last 10 years. Respondents were mostly rheumatologists practicing in academic hospitals and represented all 5 regions of Africa. While prevalence of severe disease outcomes varied among countries, prevalence was as high as 100% for calcinosis, 32% for ILD, and 29% for mortality. Respondents identified limitations in accessibility of medications, and almost half of all queried resources were accessible to $\leq 50\%$ of respondents, mostly in Northern and Southern Africa.

To date, only 196 children with JDM reviewed within the last 25 years have been reported in the literature,

Table 1 Clinical outcomes in children with juvenile dermatomyositis in Africa among survey respondents (n = 20)

Country (Number of Respondents)	Total Patients (n = 216) [†]	Clinically Inactive Disease [‡]	Remission [‡]	Calcinosis	Interstitial Lung Disease	Deaths
Northern Africa (12; 60%)						
Egypt [6]	68*	28	13	36	5	2
Libya [3]	55*	18 ⁺	39 ⁺	12	5 ⁺	3
Morocco [1]	19	6 (31.6%)	4 (21.1%)	7 (36.8%)	6 (31.6%)	0 (0%)
Tunisia [2]	3*	6	6	0	4	0 ⁺
Eastern Africa (3; 15%)						
Kenya [2]	42	15 (35.7%)	12 (28.6%)	13 (31.0%)	8 (19.0%)	6 (14.3%)
Zambia [1]	7	5 (71.4%)	0 (0%)	3 (42.9%)	1 (14.3%)	2 (28.6%)
Central Africa (3; 15%)						
Cameroon [2]**	11	6 (54.5%)	5 (45.5%)	3 (27.3%)	2 (18.2%)	0 (0%)
Democratic Republic of the Congo [1]**	3	3 (100%)	1 (33.3%)	3 (100%)	0 (0%)	+
Western Africa (1; 5%)						
Nigeria [1]	4	2 (50.0%)	1 (25.0%)	0 (0%)	1 (25.0%)	0 (0%)
Southern Africa (1; 5%)						
South Africa [1]	4	1 (25.0%)	1 (25.0%)	2 (50.0%)	0 (0%)	0 (0%)

Number of patients reported as n; (%) also reported when total patients known

*At least 1 respondent unsure of total number of patients, therefore number provided is an underestimation and percentages were unable to be calculated; Egypt = 3 of 6 respondents unsure about prior patients, Libya = 1 of 3 respondents unsure about total current and total prior patients, Tunisia = 2 of 2 respondents unsure about total current patients and 1 of 2 unsure about prior patients.

[†] Defined as lack of evidence of myositis disease activity as assessed by global and extra-muscular assessments, stable muscle strength and function, and normal muscle enzyme levels, as per the International Myositis Assessment & Clinical Studies (IMACS) criteria for lack of evidence of active myositis

[‡] Defined according to the IMACS 2005 definition: clinically inactive disease while not receiving any drug therapy for a 6-month continuous period

**Respondents answered questions only about current, not prior, patients (Cameroon = 1 respondent, Democratic Republic of Congo = 1 respondent)

+ Response missing from 1 respondent

with the vast majority described in case series from just 2 countries, Egypt (134 children) and South Africa (46 children) [3, 25, 26]; the remainder have been described in single-patient case reports [20–23] or a few JDM patients among larger cohorts of other pediatric rheumatic diseases or idiopathic inflammatory myopathies [4, 5, 24]. The 216 children with JDM seen within only the last 10 years described in our survey exceeds what has been reported in the past 25 years. Given that as of 2021, there were known rheumatology centers in only 10 of the 54 African countries [39], this number still likely vastly underestimates true prevalence as our survey did not capture all known patients with JDM and does not account for the likely many children who remain undiagnosed. While epidemiologic studies of JDM have focused mostly on incidence, with less known about prevalence, one study in France determined the prevalence of JDM to be 3.78 per 100,000 children age 0–16 years, with similar incidence to what is reported in the literature [40]. There are an estimated approximately 650 million children age 0–17 years in Africa as of 2021 [41]; assuming similar prevalence, there may be 24,000–25,000 children with JDM on the continent. While our understanding of the prevalence of JDM in Africa is limited, the findings of our survey highlight that existing literature likely greatly underestimates the true burden of JDM on the continent.

Overall, the prevalence of severe disease manifestations and outcomes in several countries is consistent with what is reported in the literature. However, in other countries represented in our survey, observed prevalence was higher than what has been reported. Prevalence of calcinosis ranges widely and is thought to depend on various factors, including MSA and/or MAA [29], diagnostic delays, disease duration, and Black race [28, 42]; studies of national and/or international registries estimate prevalence between 12 and 47% [8, 11, 12, 16, 17, 43], though a single-center case series in South Africa found a prevalence of 71% [26]. Respondents to our survey generally identified similar prevalence of calcinosis, though the prevalence reported in South Africa (50%) and Democratic Republic of the Congo (100%) was higher than estimates from national and/or international registries [8, 11, 12, 16, 17, 43]. Prevalence estimates of ILD in JDM range from 7 to 19% [44]; while prevalence estimates in Kenya, Zambia, and Cameroon were consistent, ILD was reported in 25% of Nigerian and 32% of Moroccan children. Finally, mortality estimates are 1–3%, though these are mostly based on studies from high-income countries [9, 31, 32]. While most respondents reported similar estimates, higher mortality was reported in Kenya (14%) and Zambia (29%).

Table 2 Resources available and accessible for diagnosing and managing JDM in Africa ($n = 17^*$)

	Resources available, not considering access and/or cost	Resources typically used after considering access and/or cost	Countries [†]
General Diagnostic Tools	$n = 18$	$n = 17$	
None of the above	n/a	0 (0%)	
Inflammatory markers	18 (100%)	15 (88.2%)	
Muscle enzymes	18 (100%)	17 (100%)	
vWF antigen	4 (22.2%)	1 (5.9%)	Egypt
ANA	15 (83.3%)	9 (52.9%)	
MSA and/or MAA**	10 (55.6%)	5 (29.4%)	Egypt, Tunisia, South Africa
Muscle biopsy	11 (61.1%)	1 (5.9%)	Tunisia
EMG	13 (72.2%)	7 (41.1%)	Egypt, Libya, Tunisia
Disease assessment tools***	13 (72.2%)	9 (52.9%)	
Other [†]	2 (11.1%)	2 (11.8%)	
Diagnostic Tools for Calcinosis	$n = 17$	$n = 17$	
None of the above	n/a	1 (5.9%)	
Physical exam	14 (82.4%)	13 (76.5%)	
X-rays	15 (88.2%)	12 (70.6%)	
MRI	9 (52.9%)	4 (23.5%)	Egypt, Libya
Ultrasound	12 (70.6%)	7 (41.2%)	Egypt, Tunisia, Zambia, Cameroon
Other [†]	1 (5.9%)	0 (0%)	
Diagnostic Tools for ILD	$n = 17$	$n = 17$	
None of the above	n/a	0 (0%)	
X-rays	10 (58.8%)	9 (52.9%)	
CT	15 (88.2%)	12 (70.6%)	
PFTs	14 (82.4%)	13 (76.5%)	
Other	0 (0%)	0 (0%)	
Medications	$n = 17$	$n = 16$	
None of the above	n/a	1 (6.3%)	
Steroids			
Prednisone or prednisolone	17 (100%)	16 (100%)	
Intravenous Methylprednisolone Pulse Dosing (15-30 mg/kg)	16 (94.1%)	14 (87.5%)	
Conventional DMARDs			
Hydroxychloroquine or chloroquine	14 (82.4%)	8 (50%)	Egypt, Tunisia, Morocco, Zambia
Methotrexate	17 (100%)	16 (100%)	
Mycophenolate mofetil or mycophenolic acid	14 (82.4%)	11 (68.8%)	
Azathioprine	14 (82.4%)	10 (62.5%)	
Tacrolimus	7 (41.2%)	0 (0%)	
Cyclosporine or ciclosporin	10 (58.8%)	3 (18.8%)	Egypt, Morocco
Cyclophosphamide	12 (70.6%)	8 (50%)	Egypt, Tunisia, Morocco, South Africa, Kenya
Biologic DMARDs			
Rituximab	13 (76.5%)	8 (50%)	Egypt, Tunisia, Morocco, Libya, South Africa, Kenya
TNF inhibitors	9 (52.9%)	4 (25%)	Egypt, South Africa
Abatacept	2 (11.8%)	1 (6.3%)	Egypt
Other			
IVIg	14 (82.4%)	9 (56.3%)	

Table 2 (continued)

	Resources available, not considering access and/or cost	Resources typically used after considering access and/or cost	Countries ⁺
Janus kinase inhibitors	6 (35.2%)	2 (12.5%)	Egypt
Other	0 (0%)	0 (0%)	

JDM: juvenile dermatomyositis, vWF: von Willebrand factor, ANA: antinuclear antibody, MSA: myositis-specific antibody, MAA: myositis-associated antibody, EMG: electromyography, MRI: magnetic resonance imaging, ILD: interstitial lung disease, CT: computed tomography, PFTs: pulmonary function tests, DMARD: disease-modifying antirheumatic drugs, TNF: tumor necrosis factor, IVIG: intravenous immune globulin

* 17 respondents except where otherwise noted

+ Country listed when a resource was available and/or accessible to ≤ 50% of respondents; countries in bold represent Northern or Southern African countries

** MSA include: Mi-2, MDA5 (CADM140), NXP-2 (MJ), TIF1 (p155/140), SRP, anti-synthetase (Jo-1, PL-7, PL-12, EJ, OJ, KS, Zo, Ha, YRS); MAA include: Pm-Scl, U1RNP, U1/2RNP, U3RNP, Ro, La, Ku

*** Including any of the following: Childhood Myositis Assessment Scale (CMAS), Childhood Health Assessment Questionnaire (CHAQ), Manual Muscle Testing (MMT), Physician Global Activity Visual Analogue Scale (VAS), Patient/Parent Global Activity VAS, Myositis Disease Activity Assessment Tool (MDAAT), Myositis Disease Damage Index (MDI), Physician Global Assessment of Disease Damage, Patient/Parent Global Assessment of Disease Damage

† Other = MRI [1], ultrasound [1]

‡ Other = diagnosis made clinically but all tools available [1]

When exploring availability and accessibility of diagnostic tools and medications commonly used in the management of JDM, regional differences were noted. 48% of the queried resources were accessible to 50% or fewer of respondents, mostly in Northern or South Africa. This reflects regional healthcare disparities on the continent, with Northern and Southern African countries having the highest Healthcare Access and Quality (HAQ) Index in the continent [45]. Interestingly, diagnostic tools without any associated cost, namely disease assessment tools (including the Childhood Myositis Assessment Scale [CMAS], Manual Muscle Testing (MMT), and Physician and Patient/Parent Global Activity Visual Analogue Scale [VAS], among others) and physical exam to assess calcinosis, were not indicated as being universally available or accessible, despite their important role in the management of JDM [46]. This may reflect limitations in time required to utilize these tools and/or lack of familiarity with their use; increasing their use may reflect a cost-effective way to improve the diagnosis and management of JDM.

Our study had several notable strengths. To our knowledge, this is the first study to explore the scope of JDM across the African continent as a whole, including an overview of availability and accessibility of diagnostic tools and medications. Furthermore, we had representation from all 5 regions of the continent, ensuring our findings were broadly representative of the continent. Our respondents identified more children with JDM seen in the last 10 years than have been represented in all the literature to date in the last 25 years, suggesting that the prevalence of JDM is not fully captured by existing publications. Furthermore, as of 2021, there were known rheumatology centers in only 10 of the 54 African countries [39]; however, rheumatologists from 5 countries not included in this report (Cameroon, Democratic Republic of the Congo, Morocco, Tunisia, and Zambia) responded

to our survey. This suggests that there may have been interim growth in the rheumatology workforce and/or these practitioners were not captured by the 2021 report. Given that our survey was distributed to all members of AFLAR and PAFLAR via WhatsApp groups, we were able to minimize sampling bias. Finally, our survey was distributed in both English and French, the official and/or commonly spoken language in 90% of African countries, thereby ensuring equity in participation.

Despite these strengths, our study also had several limitations inherently seen with surveys. While the potential reach of our survey was broad, only a small proportion of recipients initiated the survey; while over half of those who started the survey partially or fully completed it, overall the number of respondents was small. Furthermore, not all countries were represented by respondents, therefore our understanding of the scope of JDM across the entire African continent, while an improvement on existing literature, remains limited. As a result, while our survey provides improved insight into the burden of JDM in Africa, it still likely vastly underestimates the true prevalence of disease since we (1) did not capture all known patients with JDM and (2) did not account for the, likely many, children with JDM who remain undiagnosed. While we strived to count unique JDM patients by asking respondents who share patients at a single center to coordinate their responses, it is possible that the same patient may have been included in multiple survey responses. Though most African countries officially and/or commonly use English and/or French, the remaining 5 countries speak Portuguese and/or native African languages, therefore providers from these countries may not have been able to participate in the survey. Given that this was a survey, we did not analyze outcomes based on MSA or MAA, the distribution of which may have influenced the prevalence of outcomes such as calcinosis and ILD. Finally, given that we inquired about children seen within

the last 10 years but who are not currently followed, there may have been recall bias; respondents may have been more likely to remember severe outcomes (leading to overestimation), and their estimation of the number of patients with JDM may not have been fully accurate since we did not ask them to confirm with medical record review as this would have been cumbersome and reduced response rate.

Importantly, this survey identifies not only existing limitations in care of children with JDM but also highlights opportunities to improve their care and outcomes. Our survey demonstrates that existing literature underestimates disease burden; furthermore, the number of children identified in our survey is still likely a vast underestimation of disease prevalence. Understanding epidemiology of disease is critical and can be achieved through the establishment of international patient registries [46], as has recently been successfully achieved by PAFLAR for juvenile idiopathic arthritis (JIA) [47]. Limited familiarity of JDM among healthcare providers leads to missed diagnoses and inadequate management, challenges that have been identified not only in our survey but in prior work [1, 48]. While increasing the pediatric rheumatology workforce is a crucial long-term goal, educating general practitioners, pediatricians, allied healthcare professionals, and community health workers is perhaps a more achievable goal that can have widespread impact on recognition and initial management of disease. A variety of strategies have been proposed, including local education campaigns, use of free and accessible online educational materials such as Pediatric Musculoskeletal Matters, and use of telemedicine to improve access to care [1, 48]. Our survey also demonstrates that caregivers are unfamiliar with JDM and that there is stigma surrounding disease; educating caregivers and patients and providing access to support networks and resources are critical steps in improving the patient and caregiver experience. Finally, our survey identifies limited availability of and access to a variety of diagnostic tools and medications. There are efforts underway to revise and expand the World Health Organisation (WHO) Essential Medicines List for pediatric rheumatology. This list informs policy makers about which medications are most important for the management of PRD, particularly in LMIC; however, the list currently lacks many critical medicines needed to appropriately manage PRD [49, 50]. These efforts require funding and advocacy but can have huge impacts on improving the equity of care for children globally.

Conclusions

In conclusion, our survey of African providers identified a greater number of children with JDM than what is reflected in existing literature, supporting the notion that limited existing reports do not reflect disease rarity on

the continent but rather underreporting and undercounting true disease burden [1, 4]. The potential differences in JDM manifestations associated with increased morbidity and mortality warrant further systematic study; if there are indeed differences in African patients, it is critical to understand why these differences occur and to develop tailored management guidelines that account for these differences. The inclusion of children and providers from African countries and other LMIC in global collaborative research is critical to ensure equity and generalizability of research and improve outcomes.

Abbreviations

PRD	Pediatric rheumatic diseases
LMIC	Low and middle-income countries
JDM	Juvenile dermatomyositis
ILD	Interstitial lung disease
CARRA	Childhood Arthritis and Rheumatology Research Alliance
IMACS	International Myositis Assessment & Clinical Studies
AFLAR	African League of Associations for Rheumatology
PAFLAR	Paediatric Society of the African League Against Rheumatism
IRB	Institutional Review Board
vWF	von Willebrand factor
CK	Creatinine kinase
LDH	Lactate dehydrogenase
AST	Aspartate aminotransferase
MSA	Myositis-specific antibodies
MAA	Myositis-associated antibodies
EMG	Electromyography
MRI	Magnetic resonance imaging
TNF	Tumor necrosis factor
Jak	Januse kinase
JIA	Juvenile idiopathic arthritis
WHO	World Health Organisation

Supplementary Information

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Supplementary Material 1

Supplementary Material 2

Supplementary Material 3

Supplementary Material 4

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Author contributions

JP, LBL, DMW, KW, CS, and AM conceptualized the study; JP drafted the survey; CS, KW, and AM distributed the survey; JP performed data analysis; JP drafted the manuscript; all authors read and approved the final manuscript.

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Data availability

The datasets used and/or analyzed during the current study are available from the corresponding author on reasonable request.

Declarations

Ethics approval and consent to participate

Albert Einstein College of Medicine Institutional Review Board (IRB) approval was obtained (approval number 2022–13864) with waiver of informed consent.

Consent for publication

Not applicable.

Competing interests

The authors declare that they have non-competing interests.

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