The prevalence, epidemiological characteristics and mortality trends of inflammatory myopathies patients in Oman: the Prevision study

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Abstract

Objective

This research aims to investigate the prevalence, epidemiological characteristics, mortality rates, survival rates and the rate of malignancy in patients diagnosed with inflammatory myopathies (IIM) in Oman.

Methods

This is a longitudinal study, that covered a span of 16 years at eight rheumatology centres in Oman. The study included all adults and paediatric patients diagnosed with different types of idiopathic inflammatory myopathies (IIM) and who fulfil either the Bohan classification criteria or the 2017 EULAR/ACR classification criteria.

Results

The study included a total of 116 patient with an average age of 38.78 (±17.61 SD) years. The most prevalent form of myositis was found to be dermatomyositis (DM) 48 (41.38%), followed by polymyositis (PM) 36 (31.03%) and juvenile myositis (JDM) 18(15.52%). However, inclusion body myositis and necrotising myopathy were relatively rare conditions. The prevalence rates for DM, PM and JDM were determined as 2.2, 2.2, and 1.14 per 100,000 population respectively. Cardiac complications were observed in 14.66% of cases. Among the individuals studied, a history of malignancy was present in around 1.72% of cases. ANA antibodies were present in 71.55% of the cases, anti-Jo 1 and anti-RNP/SM antibodies were detected in 8.62%, and Anti-Ro antibodies in 24.14%. The overall mortality rate was found to be 6.90% with a rate of 11.1% among JDM cases. The five-year survival rates for PM, DM and JDM were found to be 94.4%, 91.7% and 89.0% respectively. These rates decline over a 10-year period to 67%, 69% and 83.3% respectively.

Conclusion

The study highlights the prevalence, mortality, and survival rates of IIM in Oman. Patients with JDM had a higher mortality rate. This underscores the significance of using novel healthcare strategies to improve clinical outcomes and meet special requirements for this group of patients.

Key words

idiopathic inflammatory myopathies, dermatomyositis, polymyositis, immune-mediated necrotising myopathy, juvenile dermatomyositis, inclusion body myositis, prevalence, mortality, malignancy

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Introduction

Idiopathic inflammatory myopathies refer to a group of diseases characterised by progressive muscle weakness. The typical presentation of the disease is bilateral symmetrical weakness of several muscle groups. Muscle weakness can be experienced in different muscle groups according to the underlying conditions. Many patients with muscle disease will have elevated levels of the muscle enzyme creatine kinase (CK), which is one of the most sensitive and widely measured among all muscle enzymes (1, 33). Additionally, patients may often present with extra muscular manifestations, including musculoskeletal problems, respiratory problems, cardiovascular system, gastrointestinal system, etc. (2).

The idiopathic inflammatory myopathies (IIMs) group include dermatomyositis (DM), polymyositis (PM), immune mediated necrotising myopathy (IMNM), juvenile dermatomyositis (JDM), juvenile polymyositis (JPM) and inclusion body myositis (IBM).

The advancement of diagnostic techniques and the emergence of novel antibodies have led to the development of new classification criteria for inflammatory myopathies. In 1975, Bohan devised the initial set of classification criteria, which facilitated the categorisation of individuals with myopathies into distinct groups. The European League against rheumatism and the American college of Rheumatology (EULAR/ACR) released the updated classification criteria in 2017. These criteria had aided the identification of adult and juvenile idiopathic inflammatory myopathies, along with their primary subtypes (3-5).

The geographical distribution of IIM indicates that both environmental factors and genetics may contribute to the developmental of these diseases. Several epidemiological studies have examined these conditions and revealed diverse clinical outcomes. Prevalence studies on IIM in the Middle East, particularly in Oman, are limited. Using a comprehensive population database is crucial for conducting epidemiological studies on these diseases. This allows for the identification of disparities in terms of geography and demographics, as well as gaining a thorough understanding of their characteristics and different causes (6-11).

In terms of epidemiological clinical trials, a recent study conducted in Norway determined that the point prevalence of PM and DM to be 8.7 per 100,000. The study also revealed a calculated incidence rate of 6–10 incidents per 1,000 (9).

The rates of morbidity and mortality related to PM and DM are significant. Aside from the damage caused by the underlying disease, treatment side effects play an important role in the magnitude of damage that may occur. Survival rates of inflammatory arthritis had improved remarkably in the recent years. A recent report had estimated the five-year survival rate to range from 60% to 90% (13-17).

Therefore, this study aims to investigate the frequency and epidemiological features of IIM in Oman, together with patient characteristics and therapeutic interventions used for their management. In addition, we will analyse the mortality rate and survival rate of these patients, along with different variables that correlate with their increased risk of mortality. Furthermore, our research aims to examine the potential correlation between malignancy and IIM in our country, and to determine if this link has altered over time due to the introduction and advancement of new therapeutic approaches.

Methods

This study was conducted across eight rheumatology facilities in the Sultanate of Oman including government hospitals that provide rheumatology services. Ethical approval for the study was granted in 2022 under reference number MoH/CSR/22/25608. Data was collected from medical record systems. The study focused on both adult and paediatric patients diagnosed with myopathies who met either the criteria established by Bohan in 1975 for classification of PM and DM or the classification criteria for adult and juvenile idiopathic inflammatory myopathies published by EULAR/ACR in 2017. Data collection took place from 2006 to 2022 covering a period of 16 years.

To identify patients diagnosed with IIM; we conducted a search of records for both hospitalised and non-hospitalised patients. Various search phrases were used to facilitate this process, including myopathies, DM, PM, JDM, JDM, IMNM and IBD. Additionally, we used ICD codes (M33.0, M33.1, M33.2, M33.9, M36.0, M60.8 and M60.9) to assist in locating patient files.

The study's exclusion criteria included patients who did not meet the required eligibility criteria for classification and patients who lack sufficient clinical data. We used methods such as distribution charts, means, medians and interquartile ranges to analyse patient's characteristics. To examine variables, we employed the Chi statistic. For comparing means across groups, we used students t-tests. The analysis was carried out using STATA software v. 13 by StataCorp LP, in College Station, TX, USA.

Results

A total of 116 patients with IIM were included in this study which were representing a variety of diagnoses across the 8 centres. The most prevalent form of myositis was found to be DM (41.38%), followed by PM (31.03% and juvenile myositis (15.52%). However, IBM and necrotising myopathy are relatively rare conditions. In this study, females had higher prevalence (67.24%) compared to males (32.76%), with an average age of 38.78 (±17.61 SD). Heliotrope rash was the most frequent cutaneous symptom, occurring 49.14% of the patients. Gottron's papules and Gottron's sign closely followed the heliotrope rash as shown in Table I. Muscle weakness was observed in 93.10% of patients, highlighting its significance as a central pathology in myositis. A significant number of patients (60.34%) reported musculoskeletal symptoms, particularly arthralgia. In addition, vascular symptoms, such as Raynaud's phenomenon and digital ulcers were reported, although these are not as common (Table I).

Table I. Patient diagnosis, demographics, and clinical manifestations of the Prevision study cohort.

(n=116)	Frequency (%)	Category	Frequency (%)
Patient diagnosis		**Cutaneous Manifestations**	
Polymyositis	36 (31.03%)	Heliotrop rash	57 (49.14%)
Dermatomyositis	48 (41.38%)	Gottron's papules	49 (42.24%)
Inclusion muscle myositis	7 (6.03%)	Gottron's sign	41 (35.34%)
Necrotising myopathy	1 (0.86%)	Shawal sign	12 (10.34%)
Amyopathic dermatomyositis	3 (2.59%)	V sign	13 (11.21%)
Juvenile dermatomyositis	18 (15.52%)	Nail abnormalities	14 (12.07%)
Juvenile polymyositis	3 (2.59%)	Photosensitive rash	24 (20.69%)
Gender distribution			
Male :	38 (32.76%)	Alopecia	13 (11.21%)
Female:	78 (67.24%)	Mucosal ulcers	11 (9.48%)
Mean age ± SD (years)	38.78 ±17.61 SD	**Musculoskeletal manifestations**	
Vascular manifestations		Arthritis/arthralgia	70 (60.34%)
		Muscle weakness	108 (93.10%)
Digital ulcers	7 (6.03%)	Mechanic's hands	12 (10.34%)
Calcinosis	5 (4.31%)		
Raynaud's phenomenon	13 (11.21%)	**Lymphadenopathy**	20 (17.24%)
Overlapping syndrome		**Neuropsychiatric involvement**	
Present	28 (24.14%)	Present	13 (11.21%)
Rheumatoid arthritis	8 (6.90%)	Seizure	1 (0.86%)
Scleroderma	11 (8.27)	Myelitis	1 (0.86%)
Lupus	11 (9.48%)	Neuropathy	5 (4.31%)
Sjogren's syndrome	2 (1.72%0)	Depression	3 (2.59%)
Mixed connective tissue disease	3 (2.59%)	**Pulmonary involvement**	
Cardiac involvement		Present	34 (29.31%)
Present	17 (14.66%)	Interstitial lung disease (ILD)	18 (15.52%)
Pericardial effusion/pericarditis	2 (1.72%)	Pulmonary embolism	2 (1.72%)
Myocarditis	3 (2.59%)	Pneumonitis	4 (3.45%)
Conduction defects/arrhythmias	3 (2.59%)	Pulmonary hypertension	4 (3.45%)
Valvular disease	3 (2.59%)	Pleural effusion	3 (2.59%)
Coronary artery disease	4 (3.45%)	**Gastrointestinal involvement**	
Peripheral vascular disease	2 (1.72%)	Present	9 (7.76%)
Malignancy		Ischaemic colitis	1 (0.86%)
History of benign tumour	5 (4.31%)	Spleen/liver infarction	5 (4.31%)
History of malignancy	2 (1.72%)	Enteritis	5 (4.31%)
History of ongoing malignancy	3 (2.59%)	Dysphagia/oesophageal dysmotility	29 (25.00%)

Internal organ involvement in inflammatory myopathies

According to the Prevision study, approximately 14.66% of patients experienced cardiac complications. A variety of cardiac manifestations were detected in our analysis, including pericardial effusion/pericarditis (1.72%), myocarditis (2.59%), conduction defects (2.59), valvular disease (2.59%), coronary artery disease (3.45%), and peripheral vascular disease (1.72%). An estimated 11.21% of the Prevision study cohort had neuropsychiatric symptoms. Among these were seizures (0.86%), neuropathy (4.31%), and depression (2.59%).

Approximately 29.31% of the patients in this cohort had pulmonary conditions. These include pleural effusion (2.59%), interstitial lung disease (ILD) (15.52%), pulmonary emboli (1.50%), pneumonitis (3.45%), and pulmonary hypertension (3.45%). In addition, an estimated 7.76% of patients identified in the Prevision study suffered from gastrointestinal symptoms. Approximately 0.86% of gastrointestinal symptoms were caused by ischaemic colitis, while about 4.31% were caused by spleen and liver infarctions, 4.31% by enteritis, and about 25.00% had history of dysphagia and dysmotility (Table I). The study also revealed that the prevalence of thyroid abnormalities and benign tumours in patients with dermatomyositis was significantly higher than other groups in the study (p=0.020, p=0.022 respectively). In the current study several other important findings stood out revealing disparities among the study different groups. It was particularly noteworthy that Gottron's papules were mainly observed in the JDM group (p=0.0001). Additionally, there was also significant difference in the occurrence of heliotrope rash and calcinosis in the JDM with a p-value

Table II. Laboratory findings, diagnostic procedures, and treatment metrics of the Prevision study cohort.

Laboratory/procedure	Frequency (%)	Serological marker	Frequency (%)	Imaging /medications	Frequency (%)
Laboratory finding		ANA	83 (71.55%)	**MRI Imaging**	
Anaemia presence	37 (31.90%)	Anti Jo-1	10 (8.62%)	Muscle MRI	78 (67.24%)
Iron deficiency anaemia	18 (15.52%)	Anti Sm	4 (3.45%)	MRI showed myositis	64 (81.01%)
Haemolytic anaemia	13 (11.21%)	Anti-Ro/SSA	28 (24.14%)	**EMG Test**	
Anaemia of chronic disease	8 (6.90%)	Anti La/SSB	4 (3.45%)	EMG performed	69 (60.00%)
Multifactorial anaemia	7 (6.03%)	Anti RNP/SM	10 (8.62%)	EMG with myopathic changes	62 (87.32%)
Elevated CRP	54 (46.55%)	Anti PM/SCL	12 (10.34%)	**Medications**	
Elevated ESR	67 (57.76%)	ACL IgG	1 (0.86%)	Patients on steroids	49 (36.84%)
Elevated creatinine kinase	92 (79.31%)	B2-glycoprotein I	0 (0.00%)	Average steroid dose (mg/day)	9.21
Elevated myoglobulin	42 (36.21%)	Lupus anticoagulant	4 (3.01%)	Antimalarials	41 (35.34%)
Elevated LDH	75 (64.66%)	Anti Mi-2	5 (4.31%)	IVIG	37 (31.90%)
Elevated TSH	18 (15.52%)	Anti PL-12	3 (2.59%)	Azathioprine	58 (50.00%)
Muscle biopsy		Anti TIF1-gamma	3 (2.59%)	Methotrexate	50 (43.10%)
Muscle biopsy performed	34 (29.31%)	Anti SAE	1 (0.86%)	Mycophenolate mofetil	49 (42.24%)
Positive report of DM	9 (25.00%)	Anti NXP-2	2 (1.72%)	Rituximab	22 (18.97%)
Positive report of PM	11 (30.56%)	Anti MDA5	5 (4.31%)	Cyclophosphamide	20 (17.24%)
Positive report of IBM	2 (5.56%)	Anti SRP	3 (2.59%)	TNF inhibitors	3 (2.59%)
Positive report of NM	14 (38.89%)	Anti OJ	1 (0.86%)	JAK inhibitors	4 (3.45%)
Tumour markers		Anti-HMGCR	0 (0.00%)	Sulfasalazine	1 (0.75%)
Tumour markers test present	22 (18.97%)	Anti PL-7 positive	0 (0.00%)	Tacrolimus	1 (0.75%)
Abnormal tumour markers	6 (5.17%)	Anti-EJ positive	0 (0.00%)	Abatacept	0 (0.00%)
Presence of comorbidities					
Present	41 (35.34%)	Hypertension	20 (17.24%)	Depression	5 (4.31%)
Ischaemic heart	7 (6.03%)	Hyperlipidaemia	13 (11.21%)	Obesity	8 (6.90%)
Diabetes	17 (14.66%)	Others	12 (10.34%)		

of 0.005. Furthermore, another crucial observation was the frequency of comorbidities among patients with PM compared to others with a significant *p*-value of 0.030. This emphasises the complexity and importance of management for PM patients. Moreover, cardiac involvement was more pronounced in the PM group as evidenced by a *p*-value of 0.007 underscoring the significance of assessment and monitoring for these individuals.

Analysis of laboratory parameters and diagnostic imaging findings in the Prevision study cohort

A variety of types of anaemia are present in this study (31.9%), with iron deficiency anaemia being the most prevalent at 15.52% and haemolytic anaemia at 11.21%. Additionally, multifactorial anaemia as well as chronic disease anaemia were observed as well. Creatine kinase and LDH were elevated in 79.31% and 64.66% of patients respectively. 67% of patients underwent MRIs of their muscles, and out of them, 81.01% had evidence of myositis. An electromyographic (EMG) was performed in 60% of cases, which revealed myopathic changes in 87.32% of those cases.

Muscle biopsies, which were performed on 29.31% of the cases, showed reports that included DM, PM, IBM, and necrotising myopathy. This can be explained by lack of such facility in many rural areas. Testing for tumour markers was conducted in 18.97% of the patients and abnormalities were revealed in 5.17% of the cases and 27.27% of the specimens sent. This underscores the importance of continuous malignancy surveillance in this patient population.

Furthermore, the study found that 36.84% of patients were prescribed steroids, with an average dosage of 9.21 mg per day. Among the most prescribed medications were antimalarials and immunosuppressants such as azathioprine, methotrexate, and mycophenolate mofetil. In this cohort, IVIG, cyclophosphamide, and biologic drugs such as Rituximab were among the treatments used (Table II).

Analysis of autoantibody profiles in the Prevision study cohort

The Prevision study had revealed a wide range of different autoantibody profiles which is commonly associated with these conditions. Around 71.55% of the patients tested positive for the ANA antibodies. The study also, reported the presence of anti Jo-1 antibodies in 8.62% and anti-RO antibodies in (24.14%) of the cases. Additionally, around 10.34% of patients tested positive for anti-PM/SCL antibodies. Furthermore, anti-RNP/SM antibodies were detected in around 8.62% of the patients.

Further analysis revealed that anti-TIF antibodies were present in 2.59% of the patients. From these cases, two patients had DM, and none of them had a history of malignancy. Additionally, we observed the presence of NXP 2 antibodies (1.72%) in individuals with both PM and DM.

Previous studies have reported an association between MDA5 antibodies and pulmonary complications. In our study cohort, 34.31% tested positive for these antibodies, and all (100%) patients had evidence of pulmonary involvement. From these patients, 80% had interstitial lung disease (ILD), suggesting that this antibody could be a useful marker for lung involvement. Interestingly there was no evidence of malignancy or mortality in this subgroup, which may indicate a divergence in clinical progression.

The study cohort exhibit a prevalence of 2.59% of SRP antibodies, with two

Table III. Statistics on disease outcome and mortality rate of the Prevision study cohort.

Outcome category	Frequency (%)	95% confidence interval
Surviving	79 (68.10%)	59.59% - 75.81%
Deceased	8 (6.90%)	3.17% - 12.34%
Disease outcome		
Remission	68 (58.62%)	49.45% - 67.34%
Low disease activity	17 (14.66%)	8.83% - 22.20%
Moderate disease activity	16 (13.79%)	8.09% - 21.19%
Very active disease activity	8 (6.90%)	3.17% - 12.34%
Causes of very active disease (out of 8 cases)		
New case	6 (75.00%)	44.96% - 94.29%
Refractory myositis	4 (50.00%)	23.94% - 76.06%
Poor compliance	4 (50.00%)	23.94% - 76.06%
Mortality causes (out of 8 cases)		
Sepsis	4 (50.00%)	23.94% - 76.06%
CV involvement	1 (12.50%)	0.32% - 52.77%
Lung involvement	2 (25.00%)	6.98% - 57.91%
Renal involvement	1 (12.50%)	0.32% - 52.77%
CNS involvement	1 (12.50%)	0.32% - 52.77%
Other causes of death	2 (25.00%)	6.98% - 57.91%

patients diagnosed with PM and one with DM. Remarkably, none of these patients had any history of malignancy or any other worse clinical outcome. It is important to mention that these patients did not exhibit necrotising myopathy typically associated with these antibodies and along with absence of HMGR antibodies. These findings point to a unique spectrum of these conditions within this cohort. This finding highlights the need for further research since the current study is of limited sample size.

Anti-Mi antibodies were detected in 4.31% of patients in our analysis with approximately 40% of these patients had pulmonary conditions. Furthermore, among those with positive anti-Mi antibodies 80% had DM while 20% had PM. None of these patients had any history of malignancy and all patients survived without complications, which suggests a less aggressive disease course. Lastly, it is worth mentioning that we did not detect any cases of positive HMGCR antibodies, anti-PL 7 antibodies or other less common myositis specific antibodies within this cohort (Table II).

Gender analysis

When comparing the gender distribution among the individuals included in the Prevision study cohort, we observe some trends. There was no significant gender disparity observed between PM and DM cases with p=0.310 and 0.106, respectively.

When examining the variations in DM manifestations between males and females within our cohort, we found that majority of the symptoms presented equally among both genders. These include arthritis, rashes, nail changes and other characteristic signs of the disease. However, one particular manifestation appeared to occur frequently in females: is the V-shaped rash (p= 0.010). Our analysis also indicated that gender did not significantly impact laboratory results, comorbidities, or serological markers in patients with DM. Although females exhibited a higher prevalence of ANA compared to males (p=0.063), this difference, however, did not achieve statistical significance.

Moreover, there was no disparity in mortality between genders (p=0.274). Furthermore, this research explored the occurrence of overlapping syndromes among individuals with myopathies. It was found that female patients had a prevalence (29.55%) compared to male patients (11.11%) with a p-value of 0.017.

Age-related prevalence of IIM in the Prevision cohort

Furthermore, a detailed analysis was conducted investigating the prevalence rates of various IIM among different age groups. According to this study the prevalence of JDM among children (up to 12 years old) was 0.777 per 100,000. In adolescents (13–18 years old) both DM and PM have a prevalence rate of 0.351 per 100,000, while JDM has a prevalence rate of 2.809 per 100,000. Among adults aged 19–24 and 25–29 years, the prevalence rates of DM gradually increase with values of 0.516 and 1.472 per 100,000, respectively. In the age group of 30 -39 years, the prevalence rate for DM is around 1.407 per 100,000, while for individuals aged between 40–49 years old, it increases further to 4.537 per 100,000 people. As

Table IV. The prevalence rate of IIM patients of different age group in Prevision cohort.

	DM patients/ total population	The prevalence of DM (per 100,000 inhabitants)	PM patients/ total population	The prevalence of PM (per 100,000 inhabitants)	JDM patients/ total population	The prevalence of JDM (per 100,000 inhabitants
Paediatric <12 years	---	---	0/771780	0	6/771780	0.777
Adolescents 13-18 years	1/284826	0.351	1/284826	0.351	8/284826	2.809
Adults 19-24 years	2/387448	0.516	0/387448	0	3/387448	0.774
Adults 25-29 years	7/475403	1.472	2/475403	0.421	1/475403	0.210
Adults 30-39 years	9/639636	1.407	6/639636	0.938	---	---
Adults 40-49 years	16/352632	4.537	10/352632	2.836	---	---
Adults >49 years	13/262192	4.958	17/262192	6.484	---	---
Prevalence mean for all patients	2.2		2.2		1.14	

Table V. Mortality rate of polymyositis/dermatomyositis in different cohorts.

Study	Total number of patients	Country	Overall mortality rate	Mortality rate of PM	Mortality rate of DM
Current study	**116**	**Oman**	**6.90%**	**5.56%**	**8.3%**
Essouma *et al*. 2022 (29)	683	Africa	7.8-45%	---	----
Hočevar *et al*. 2021 (30)	217	Slovenia	3.5%	---	---
D'Silva *et al*. 2020 (31)	817	UK	---	5.4%	7.5%
Kridin *et al*. 2020 (32)	166	Israel	7.4%	7.7%	7.2%
Li *et al*. 2020 (17)	801-1031	Canada	---	3.94 - 5.9%	5.13 - 8.06%
Yang *et al*. 2020 (34)	331	China	6.0%	---	---
Muhammed *et al*. 2019 (35)	372	India	10.2%	---	---
Dobloug *et al*. 2018 (36)	716	Sweden	9.0%	---	---
Wu *et al*. 2018 (37)	982	China	4.58%	---	---
Nuño-Nuño *et al*. 2017 (38)	467	Spain	24%	---	---
Ishizuka *et al*. 2016 (50)	124	Japan	15.3%	---	----
Galindo-Feria *et al*. 2016 (39)	333	Mexico	16.2%	8.7%	18%
Murray *et al*. 2015 (40)	15407	US	4.5%	----	---
Dobloug *et al*. 2015 (41)	326	Norway	27%	2.4%	2.6%
Schiopu *et al*. 2012 (42)	160	US	16.9%	---	---
Woo *et al*. 2012 (43)	162	South Korea	12.3%	4.8	12.8
Kuo *et a*l. 2011 (44)	1303	Taiwan	6.56%	5.29%	7.68%
Yu *et al*. 2011 (45)	192	China	28.6%	---	---
Limaye *et al*. 2010 (46)	370	Australia	1.75	1.56	2.40

age advances there is a rise in both DM and PM prevalence rates reaching up to 4.958 and 6.484 cases respectively per 100,000 people. In general, the average prevalence rates for DM, PM and JDM were 2.2, 2.2 and 1.14 per 100,000, respectively (Table IV).

Analysis of outcome, mortality, and survival rates in the Prevision study cohort

In this study the majority of patients (68.10%, 95% CI: 59.59–75.81%) adhered to their treatment. The outcome was favourable in 73.28%, with 58.62% (95% CI: 49.45–67.34%) achieving complete remission or had a low disease activity level (14.66%, CI: 8.83–22.20%). Despite this, (6.90%, CI: 3.17–12.34%) had very active disease level. Newly diagnosed cases constituted 75.00% (CI: 44.96–94.29%) in the subgroup with highly active illness, whereas poor compliance and treatment-resistant myositis each represented 50.00% (CI: 23.94–76.06%).

The mortality rates based on the patient diagnosis was to be as follow: for DM 8.33%, for PM 5.56%, and for JDM was found to be 11.1%. Interestingly, the JDM group had a higher mortality than other, and this finding warrant further research. The average JDM cases with mortality outcome age ranged between 11-15 years, and average disease duration of their illness was 3 years (Table V).

In terms of mortality, it was found that sepsis was responsible for 50.00% of deaths (CI: 23.94–76.06%). This was followed by lung involvement, which was responsible for 25.00% (CI: 6.98–57.91%). In addition, 12.50% (CI: 0.32–52.77%) of mortality cases were related to cardiovascular disease, renal disease, and CNS complications (Table III).

In addition, these patients lack autoantibodies that are known to be associated with a worst clinical outcome. In addition, none of the patient had history of associated malignancy. The commonest cause of mortality in this group was sepsis (50%) and there was one patient with a history of overlap syndrome with lupus. This finding suggests the need for targeted interventions and more aggressive treatment strategies in this subgroup of patients (Table V).

A total of 62.5% of patients with mortality outcome had pulmonary involvement, with 25% and 12.5% of cases showing ILD and pulmonary hypertension, respectively. Moreover, cardiac problems and overlap syndrome were associated with 25% of deaths, while dysphagia or oesophageal dysmotility were present in 50% of the patients. Autoantibodies such as anti SRP, anti MDA5 and Anti NXP were not found in patients with mortality outcome. In contrast, ANA antibodies were detected in 62.5% of the deceased individuals. Additionally, half of the deceased individuals had different comorbidities mainly DM, HTN and to lesser extent IHD. Furthermore, 50% of these patients were on steroid treatment, and had history sepsis as a contributing factor for mortality. The average steroid dose that was given is 6.08 mg. The incidence of malignancy was found in 12.5% of the patients. All these observations underscore the crucial need of effectively managing this group of patients especially those with pulmonary involvement.

In the Prevision study, the overall survival rate, for the group showed a decline as time passed. At the 5-year mark, the overall survival rate 93.1%. However, by the 10 years mark this rate dropped to 75% indicating that these conditions are progressive in nature. Specifically, PM patients had a survival rate of 94.4% at 5 years and 67% at 10 years. For DM patients the survival rates were 91.7% at 5 years and 69% at 10 years. On the other hand, JDM patients had lower long-term outcomes with a survival rate of 89% at 5 years and rate of 83.3% at 10 years. These findings underscore the discrepancies in the long-term outcomes based on the underlying conditions (Table VI).

Table VI. Survival rate of polymyositis/dermatomyositis in different cohorts.

Study	Total number of patients	Country	Survival rate of PM 5-year	Survival rate of DM 5-year	Survival rate of PM 10-year	Survival rate of DM 10-year
Current study	**116**	**Oman**	**94.4%**	**91.7%**	**67%**	**69%**
Hochberg *et al*. 1986 (16)	76	USA	73%		---	
Hočevar *et al*. 2021 (30)	217	Slovenia	75.5%		---	---
Yang *et al*. 2020 (34)	331	China	87.5%	76.6%	84.8%	71.7%
Kridin *et al*. 2020 (32)	166	Israel	82.8%	59.6%	51.5%	40.0%
Nuño-Nuño *et al*. 2017 (38)	467	Spain	86.7%		77%	
Ishizuka *et al*. 2016 (50)	124	Japan	95%	90%	90%	79%
Danieli *et al*. 2014 (51)	91	Italy	96.2%	93.9%	88.8%	90%
Schiopu *et al*. 2012 (42)	160	USA	87%	70%	69%	57%
Yu *et al*. 2011 (45)	192	China	69.9%		66.2%	
Airio *et al*. 2006 (52)	248	Finland	75%	63%	55%	53%
Torres *et al*. 2006 (53)	107	Spain	80%		71%	
Dankó *et al*. 2004 (54)	162	Hungary	94.2%	90.1%	89.4%	89.4%
Sultan *et al*. 2002 (55)	46	UK	95%		83.8%	
Maugars *et al*. 1996 (56)	69	France	73.9%		55.4%	

Malignancy

In the Prevision group 1.72% of patients had a history of malignant neoplasms. The rate of patients with benign tumour growths was slightly higher at 4.31%. Moreover around 2.59% of the group were still undergoing treatment for malignancies when the investigation took place. Among individuals with IIM approximately 18.97% underwent tumour marker testing. Approximately, around 27.27% showed positive results, accounting for 5.17% of the entire cohort. Breast cancer was found to be the most common diagnosed malignancy in this cohort.

Hence, it is crucial to conduct screenings and maintain close monitoring during IIM management since specific myopathies are associated with an elevated risk of developing malignancies (Table I).

Discussion

The incidence and prevalence rates of IIMs demonstrate significant variation, across different geographic regions worldwide. According to a study conducted by Khoo *et al*. in 2023, the estimated incidence rate of this condition is, between 0.2 to 2 cases per 100,000 person per year, while the prevalence ranges from 2 to 25 cases per 100,000 people per year. These variations can be attributed to factors such as diagnostic criteria, population genetics and environmental influences (7). A recent report by Cho *et al*. showed that the prevalence of IIM in Korea ranges from 2.3 to 4.0 per 100,000 person-years. In Turkey the reported prevalence rate was 3.22 per 100,000 persons. In contrast, studies from Minnesota (USA) reported a higher prevalence rate of 13 per 100,000 people (2, 6, 12).

The Prevision cohort study provides a nuanced view, showing an age-related increase in DM and PM prevalence, with a peak in adults over 49 years of age. According to the results of this study, there was an average prevalence rate of 2.2 for DM, 2.2 for PM, and 1.14 for JDM per 100,000. (Table IV).

Moreover, regarding patient characteristics in the Prevision study, we found pulmonary abnormalities in 29.31% of patients. Specifically, 15.52% of participants in this study had ILD, which falls within the lower range of 17–36% described in other literature (20). The incidence of oesophageal disorders, such as dysphagia and dysmotility, was 25.00%, considerably lower than the estimated 60 percent rate previously reported (21). Cardiovascular complications are prevalent among patients with idiopathic inflammatory myopathies, ranging from 67 to 75 percent. Among the patients in this study, 14.66% had cardiac complications, falling under the broad category of cardiac manifestations (22). Furthermore, the prevalence of auto-antibodies in the Prevision cohort offers contrasts when compared to the literature. The prevalence of ANA antibodies in this group was found to be 71.55%, while the reported positive ANA in literature was found to be around 50% (23). The prevalence of anti-Jo-1 in the current cohort was found to be 8.62%, which is lower than the literature-reported range of 11.3–16.7%. The prevalence of anti-Ro/SSA in the current cohort was 24.14%, which is higher than the literature-reported range of 4.9–13.5%. The elevated prevalence of anti-Ro antibodies, points to an increased incidence of conditions like Sjogren's syndrome within this population. The prevalence of overlap syndromes in this cohort was determined to be 24.14%. In addition, the prevalence of anti-PM/SCL in the current cohort was 10.34%, compared to the literature range of 4.8–6.9% (24).

In addition, the prevalence of anti-Mi-2 in the cohort was 4.31%, which was in line with what reported (1.5–16.5%). Similarly, the prevalence of anti-TIF1-gamma was found to be 2.59%, which is within the reported range of (0.7–36.4%). Furthermore, the occurrence of anti-NXP-2 was 1.75%, which is again falls within the reported range of 0.5–13.8%. The prevalence of anti-MDA5 in the current cohort was (4.31%) lower than the reported range of 13.6-48.5%. In contrast, the prevalence of anti-SRP was (2.59%), which aligns with the documented range of (2.1–8.4%). To sum up, the prevalence of different antibodies within the Prevision cohort revealed both similarities

and differences in comparison to what is reported in literatures, highlighting diversity according to the genetic background of the studied population (18, 19, 24, 25, 33).

Several studies have demonstrated a significant correlation between adult-onset idiopathic inflammatory myopathies (IIM) and the development of malignancy. It was found that approximately one-fourth of patients with IIM develop malignancy within three years of the onset of their illness. Various studies have identified several risk factors for IIM-associated malignancy, include: the diagnosis of DM, advanced age, male gender, dysphagia, cutaneous ulceration, and the presence of anti-TIF1c antibodies (26). Compared to their adult counterparts, juvenile dermatomyositis patients appear not to have a higher risk of malignancy (25). Studies had shown a correlation between patients with positive anti-TIF1-γ and anti-NXP2 antibodies and malignancy. They found 9.37-fold and 3.68-fold higher risks of getting cancer in association with anti-TIF1-γ and anti-NXP2 antibodies respectively. Furthermore, patients with positive anti-SAE antibodies had 15–57% higher risks of getting cancer. Among reported malignancies, breast cancer, haematological malignancies, lung cancer, ovarian cancer, colorectal cancer, and prostate cancer are the most common one. A recent study from Asian populations found that the most prevalent site of malignancy was the nasopharynx (18, 25, 33).

The incidence of cancer in Oman varies from 0.07% to 0.11% annually as per a recent report by Al Mahdi *et al*. group (28) in the Prevision study, a remarkable number of patients (1.72%) had history of malignant tumours, which is higher than the reported national average in the general population. The commonest malignancy in this cohort was found to be breast cancer. Furthermore, benign tumours were found to be 4.31% of the cohort.

It is important to note that malignancies and infections are among the leading causes of mortality and morbidities in these patients. DM, in particular, is associated with pronounced higher risk of malignancy than PM. As demonstrated in a report by Moghadam-Kia *et al*., which included 1078 patients with myositis, the standardised incidence ratios (SIRs) for DM were found to be nearly double those for PM. According to this study, cancer associations were 4.4 for DM and 2.1 for PM indicating an elevated risk of malignancy associated with these conditions (27). Despite the fact that the prevalence of malignancy among the Prevision cohort exceeds the general population, this finding was in line with the reports from other international reports of inflammatory myopathies (27).

It was found that the overall mortality rate of current cohort is 6.90%, with PM at 5.56% and DM at 8.3%. A study from UK study reported slightly a lower mortality rates for PM (5.4%) and DM (7.5%). On the other hand, a report from a Chinese study found a uniform mortality rate of 6.0% for all IIMs (13-17, 31-34).

There are also substantial regional differences in mortality rates in IIM. Studies from Africa reported mortality rates that range from 7.8 to 45%, these rates were significantly higher than the results from Oman. Another study from India reported an overall mortality rate of 10.2%. The variability of mortality rates across different geographical regions illustrates the importance of healthcare infrastructure, access to medical treatment, and socioeconomic conditions in determining these diseases outcomes (Table V).

In contrast to the adult patients, the mortality rate for JDM (11.1%) was found to be higher than what is reported in the literature for this condition (2-5.1%) (47-49). This higher rate of mortality in this group, highlights the importance of further investigation of various contributing factors to improve future patient outcome.

In terms of survival rate analysis, international studies have reported various survival rates. For instance, the survival rate observed in the Prevision study cohort was higher than the rates reported by Yang *et al*. from China. In addition, Kridin *et al*. reported a 5-year survival rate of 82.8% for PM, and 59.6% for DM in their study but these rates declined significantly to 51.5% and 40.0%, respectively at the end of ten years. In contrast, another study conducted in Japan reported a higher survival rate in these conditions with 95% for PM and 90% for DM at 5 years (Table VI).

Finally, it is important to note that this current study is retrospective in nature, which indicates that it has a limitation that could affect its findings. The recruitment of participants from clinics and hospitals may limit the representativeness of our sample in relation to the population affected by inflammatory myopathies. Additionally, collecting data based on health records could introduce inconsistencies in data quality. Retrospective analyses encounter challenges when it comes to controlling variables and establishing causal relationships.

However, the strength of this study relies on its comprehensive analysis of data which was collected over a period of time. This allows for more in depth understanding of the trends surrounding myopathy over an extended duration. Moreover, it offers insights, into the care provided to patients and the real-world consequences that arise in clinical settings. Additionally, it sheds light on the unique characteristics exhibited by patients with different myopathies in this region.

In conclusion, the Prevision study significantly adds to our knowledge the epidemiology, mortality rates and unique autoantibody profiles associated with IIM within the cohort. It reveals significant disparities in morality and survival rates, various organ involvement and the risk of malignancy in this cohort. Specifically, this research shed light on possible higher mortality rates in patients with JDM in this cohort, which in turn signifies the importance of investigating this observation in a larger-scale clinical trial.

Furthermore, there is a great need to adapt new healthcare strategies to improve clinical outcomes and meet special requirements for this group of patients. Moreover, we should encourage more clinical trials to introduce new therapies to manage patients with these specific disorders.

References

1. LUNDBERG I: Expert perspective: management of refractory inflammatory myopathy. *Arthritis Rheumatol* 2021; 73(8): 1394-407. https://doi.org/10.1002/art.41762
2. CHO S, KIM H, MYUNG J *et al*.: Incidence and prevalence of idiopathic inflammatory myopathies in Korea: a nationwide population-based study. *J Korean Med Sci* 2019; 34(8): e55. https://doi.org/10.3346/jkms.2019.34.e55
3. LUNDBERG IE, TJÄRNLUND A, BOTTAI M *et al*.: 2017 European League Against Rheumatism/American College of Rheumatology classification criteria for adult and juvenile idiopathic inflammatory myopathies and their major subgroups. *Ann Rheum Dis* 2017; 76(12): 1955-64. https://doi.org/10.1136/annrheumdis-2017-211468
4. BOHAN A, PETER JB: Polymyositis, and dermatomyositis (first of two parts). *N Engl J Med* 1975; 292: 344-47. https://doi.org/10.1056/nejm197502132920706
5. BOHAN A, PETER JB: Polymyositis, and dermatomyositis (second of two parts). *N Engl J Med* 1975; 292: 403-7. https://doi.org/10.1056/nejm197502202920807
6. KRONZER V, KIMBROUGH B, CROWSON C, DAVIS J, HOLMQVIST M, ERNSTE F: Incidence, prevalence, and mortality of dermatomyositis: a population-based cohort study. *Arthritis Care Res* 2023; 75(2): 348-55. https://doi.org/10.1002/acr.24786
7. KHOO T, LILLEKER JB, THONG BYH, LECLAIR V, LAMB JA, CHINOY H: Epidemiology of the idiopathic inflammatory myopathies. *Nat Rev Rheumatol* 2023; 19(11): 695-712. https://doi.org/10.1038/s41584-023-01033-0
8. BERNATSKY S, JOSEPH L, PINEAU C *et al*.: Estimating the prevalence of polymyositis and dermatomyositis from administrative data: age, sex and regional differences. *Ann Rheum Dis* 2008; 68(7): 1192-96. https://doi.org/10.1136/ard.2008.093161
9. DOBLOUG C, GAREN T, BITTER H *et al*.: Prevalence and clinical characteristics of adult polymyositis and dermatomyositis; data from a large and unselected Norwegian cohort. *Ann Rheum Dis* 2014; 74(8): 1551-56. https://doi.org/10.1136/annrheumdis-2013-205127
10. SMOYER-TOMIC K, AMATO A, FERNANDES A: Incidence and prevalence of idiopathic inflammatory myopathies among commercially insured, Medicare supplemental insured, and Medicaid enrolled populations: an administrative claims analysis. *BMC Musculoskelet Disord* 2012; 13: 103. https://doi.org/10.1186/1471-2474-13-103
11. MEYER A, MEYER N, SCHAEFFER M, GOTTENBERG J, GENY B, SIBILIA J: Incidence and prevalence of inflammatory myopathies: a systematic review. *Rheumatology* 2015; 54(1): 50-63. https://doi.org/10.1093/rheumatology/keu289
12. BALCI M, DONMEZ S, SARITAS F, BAS V, PAMUK O: The epidemiology of dermatomyositis in northwestern Thrace region in Turkey: epidemiology of dermatomyositis in Turkey. *Rheumatol Int* 2017; 37(9): 1519-25. https://doi.org/10.1007/s00296-017-3710-9
13. PAVON MR, SANCHEZ JE, PESCATORE J, EDIGIN E, RICHARDSON C, MANADAN A: Reasons for hospitalization and in-hospital mortality in adults with dermatomyositis and polymyositis. *J Clin Rheumatol* 2022; 28(2): e433-e439. https://doi.org/10.1097/rhu.0000000000001754
14. VATTEMI G, MIRABELLA M, GUGLIELMI V *et al*.: Muscle biopsy features of idiopathic inflammatory myopathies and differential diagnosis. *Auto Immun Highlights* 2014; 5(3): 77-85. https://doi.org/10.1007/s13317-014-0062-2
15. LECLAIR V, LUNDBERG I: New myositis classification criteria - what we have learned since Bohan and Peter. *Curr Rheumatol Rep* 2018; 20(4): 18. https://doi.org/10.1007/s11926-018-0726-4
16. HOCHBERG MC, FELDMAN D, STEVENS MB: Adult-onset polymyositis/dermatomyositis: an analysis of clinical and laboratory features and survival in 76 patients with a review of the literature. *Semin Arthritis Rheum* 1986, 15:168-78. https://doi.org/10.1016/0049-0172(86)90014-4
17. LI L, D'SILVA K, LU N *et al*.: Mortality trends in polymyositis and dermatomyositis: a general population-based study. *Semin Arthritis Rheum* 2020; 50(5): 834-9. https://doi.org/10.1016/j.semarthrit.2020.08.009
18. OKIYAMA N: Clinical features and cutaneous manifestations of juvenile and adult patients of dermatomyositis associated with myositis-specific autoantibodies. *J Clin Med* 2021; 10(8): 1725. https://doi.org/10.3390/jcm10081725
19. HODGKINSON LM, WU TT, FIORENTINO DF: Dermatomyositis autoantibodies: how can we maximize utility? *Ann Transl Med* 2021; 9(5): 433. https://doi.org/10.21037/atm-20-5175
20. LEGA JC, REYNAUD Q, BELOT A, FABIEN N, DURIEU I, COTTIN V: Idiopathic inflammatory myopathies and the lung. [published correction appears in *Eur Respir Rev* 2015; 24(137): 545. Dosage error in article text]. *Eur Respir Rev* 2015; 24(136): 216-38. https://doi.org/10.1183/16000617.00002015
21. LUNDBERG IE, FUJIMOTO M, VENCOVSKY J *et al*.: Idiopathic inflammatory myopathies. *Nat Rev Dis Primers* 2021; 7(1): 87. https://doi.org/10.1038/s41572-021-00325-7
22. SHAH M, SHINJO SK, DAY J, GUPTA L: Cardiovascular manifestations in idiopathic inflammatory myopathies. *Clin Rheumatol* 2023; 42(10): 2557-75. https://doi.org/10.1007/s10067-023-06599-4
23. AGGARWAL R, DHILLON N, FERTIG N, KOONTZ D, QI Z, ODDIS CV: A negative antinuclear antibody does not indicate autoantibody negativity in myositis: role of anticytoplasmic antibody as a screening test for antisynthetase syndrome. *J Rheumatol* 2017; 44(2): 223-9. https://doi.org/10.3899/jrheum.160618
24. AGUILAR-VAZQUEZ A, CHAVARRIA-AVILA E, PIZANO-MARTINEZ O *et al*.: Geographical latitude remains as an important factor for the prevalence of some myositis autoantibodies: a systematic review. *Front Immunol* 2021; 12: 672008. https://doi.org/10.3389/fimmu.2021.672008
25. MARZĘCKA M, NIEMCZYK A, RUDNICKA L: Autoantibody markers of increased risk of malignancy in patients with dermatomyositis. *Clin Rev Allergy Immunol* 2022; 63(2): 289-96. https://doi.org/10.1007/s12016-022-08922-4
26. OLDROYD AGS, ALLARD AB, CALLEN JP *et al*.: A systematic review and meta-analysis to inform cancer screening guidelines in idiopathic inflammatory myopathies. [published correction appears in *Rheumatology* (Oxford) 2021; 60(11): 5483]. *Rheumatology* (Oxford) 2021; 60(6): 2615-28. https://doi.org/10.1093/rheumatology/keab166
27. MOGHADAM-KIA S, ODDIS CV, ASCHERMAN DP, AGGARWAL R: Risk factors and cancer screening in myositis. *Rheum Dis Clin North Am* 2020; 46(3): 565-76. https://doi.org/10.1016/j.rdc.2020.05.006
28. MEHDI I, AL FARSI AA, AL BAHRANI B, AL-RAISI SS: General Oncology Care in Oman. *In*: AL-SHAMSI HO, ABU-GHEIDA IH, IQBAL F, AL-AWADHI A (Eds.): Cancer in the Arab World. Springer, Singapore, 2022. https://doi.org/10.1007/978-981-16-7945-2_12
29. ESSOUMA M, NOUBIAP JJ, SINGWE-NGANDEU M, HACHULLA E: Epidemiology of idiopathic inflammatory myopathies in Africa: a contemporary systematic review. *J Clin Rheumatol* 2022; 28(2): e552-e562. https://doi.org/10.1097/rhu.0000000000001736
30. HOČEVAR A, VIRŠČEK A, KROŠEL M *et al*.: Survival of patients with idiopathic inflammatory myopathies in Slovenia. *Front Med* (Lausanne) 2021; 8: 801078. https://doi.org/10.3389/fmed.2021.801078
31. D'SILVA KM, LI L, LU N, OGDIE A, AVINA-ZUBIETA JA, CHOI HK: Persistent premature mortality gap in dermatomyositis and polymyositis: a United Kingdom general population-based cohort study. *Rheumatology* (Oxford) 2020; 60(6): 2653-60. https://doi.org/10.1093/rheumatology/keaa661
32. KRIDIN K, KRIDIN M, AMITAL H, WATAD A, KHAMAISI M: Mortality in patients with polymyositis and dermatomyositis in an Israeli population. *Isr Med Assoc J* 2020; 22(10): 623-7.
33. CONTICINI E, DOURADO E, BOTTAZZI F *et al*.: Idiopathic inflammatory myopathies: One year in Review 2023. *Clin Exp Rheumatol* 2024; 42(2): 213-24. https://doi.org/10.55563/clinexprheumatol/dh5o6c
34. YANG X, HAO Y, ZHANG X *et al*.: Mortality of Chinese patients with polymyositis and dermatomyositis. *Clin Rheumatol* 2020; 39(5): 1569-79. https://doi.org/10.1007/s10067-019-04910-w
35. MUHAMMED H, GUPTA L, ZANWAR AA *et al*.: Infections are leading cause of in-hospital mortality in Indian patients with inflammatory myopathy. *J Clin Rheumatol* 2021; 27(3): 114-9. https://doi.org/10.1097/Rhu.0000000000001214
36. DOBLOUG GC, SVENSSON J, LUNDBERG IE, HOLMQVIST M: Mortality in idiopathic inflammatory myopathy: results from a Swedish nationwide population-based cohort study [published correction appears in *Ann Rheum Dis* 2018; 77(5): 786]. *Ann Rheum Dis* 2018; 77(1): 40-47. https://doi.org/10.1136/annrheumdis-2017-211402

37. WU C, WANG Q, HE L, YANG E, ZENG X: Hospitalization mortality and associated risk factors in patients with polymyositis and dermatomyositis: A retrospective case-control study. *PLoS One* 2018; 13(2): e0192491. https://doi.org/10.1371/journal.pone.0192491
38. NUÑO-NUÑO L, JOVEN BE, CARREIRA PE *et al*.: Mortality and prognostic factors in idiopathic inflammatory myositis: a retrospective analysis of a large multicenter cohort of Spain. *Rheumatol Int* 2017; 37(11): 1853-61. https://doi.org/10.1007/s00296-017-3799-x
39. GALINDO-FERIA AS, ROJAS-SERRANO J, HINOJOSA-AZAOLA A: Clinical and prognostic factors associated with survival in mexican patients with idiopathic inflammatory myopathies. *J Clin Rheumatol* 2016; 22(2): 51-56. https://doi.org/10.1097/rhu.0000000000000365
40. MURRAY SG, SCHMAJUK G, TRUPIN L *et al*.: A population-based study of infection-related hospital mortality in patients with dermatomyositis/polymyositis. *Arthritis Care Res* (Hoboken) 2015; 67(5): 673-80. https://doi.org/10.1002/acr.22501
41. DOBLOUG GC, GAREN T, BRUNBORG C, GRAN JT, MOLBERG Ø: Survival and cancer risk in an unselected and complete Norwegian idiopathic inflammatory myopathy cohort. *Semin Arthritis Rheum* 2015; 45(3): 301-8. https://doi.org/10.1016/j.semarthrit.2015.06.005
42. SCHIOPU E, PHILLIPS K, MACDONALD PM, CROFFORD LJ, SOMERS EC: Predictors of survival in a cohort of patients with polymyositis and dermatomyositis: effect of corticosteroids, methotrexate and azathioprine. *Arthritis Res Ther* 2012; 14(1): R22. https://doi.org/10.1186/ar3704
43. WOO JH, KIM YJ, KIM JJ *et al*.: Mortality factors in idiopathic inflammatory myopathy: focusing on malignancy and interstitial lung disease. *Mod Rheumatol* 2013; 23(3): 503-8. https://doi.org/10.1007/s10165-012-0673-2
44. KUO CF, SEE LC, YU KH *et al*.: Incidence, cancer risk and mortality of dermatomyositis and polymyositis in Taiwan: a nationwide population study. *Br J Dermatol* 2011; 165(6): 1273-79. https://doi.org/10.1111/j.1365-2133.2011.10595.x
45. YU KH, WU YJ, KUO CF *et al*.: Survival analysis of patients with dermatomyositis and polymyositis: analysis of 192 Chinese cases. *Clin Rheumatol* 2011; 30(12): 1595-601. https://doi.org/10.1007/s10067-011-1840-0
46. LIMAYE V, HAKENDORF P, WOODMAN RJ, BLUMBERGS P, ROBERTS-THOMSON P: Mortality and its predominant causes in a large cohort of patients with biopsy-determined inflammatory myositis. *Intern Med J* 2012; 42(2): 191-98. https://doi.org/10.1111/j.1445-5994.2010.02406.x
47. KIM H, HUBER AM, KIM S: Updates on juvenile dermatomyositis from the last decade: classification to outcomes. *Rheum Dis Clin North Am* 2021; 47(4): 669-90. https://doi.org/10.1016/j.rdc.2021.07.003
48. AL-MAYOUF SM, ALMUTIARI N, MUZAFFER M *et al*.: Phenotypic characteristics and outcome of juvenile dermatomyositis of Arab children. *Rheumatol Int* 2017; 37(9): 1513-7. https://doi.org/10.1007/s00296-017-3770-x
49. RHIM JW: Juvenile dermatomyositis. *J Rheum Dis* 2022; 29(1): 14-21. https://doi.org/10.4078/jrd.2022.29.1.14
50. ISHIZUKA M, WATANABE R, ISHII T *et al*.: Long-term follow-up of 124 patients with polymyositis and dermatomyositis: statistical analysis of prognostic factors. *Mod Rheumatol* 2016; 26(1): 115-20. https://doi.org/10.3109/14397595.2015.1054081
51. DANIELI MG, GAMBINI S, PETTINARI L, LOGULLO F, VERONESI G, GABRIELLI A: Impact of treatment on survival in polymyositis and dermatomyositis. A single-centre long-term follow-up study. *Autoimmun Rev* 2014; 13(10): 1048-54. https://doi.org/10.1016/j.autrev.2014.08.023
52. AIRIO A, KAUTIAINEN H, HAKALA M: Prognosis and mortality of polymyositis and dermatomyositis patients. *Clin Rheumatol* 2006; 25(2): 234-39. https://doi.org/10.1007/s10067-005-1164-z
53. TORRES C, BELMONTE R, CARMONA L *et al*.: Survival, mortality and causes of death in inflammatory myopathies. *Autoimmunity* 2006; 39(3): 205-15. https://doi.org/10.1080/08916930600622603
54. DANKÓ K, PONYI A, CONSTANTIN T, BORGULYA G, SZEGEDI G: Long-term survival of patients with idiopathic inflammatory myopathies according to clinical features: a longitudinal study of 162 cases. *Medicine* (Baltimore) 2004; 83(1): 35-42. https://doi.org/10.1097/01.md.0000109755.65914.5e
55. SULTAN SM, IOANNOU Y, MOSS K, ISENBERG DA: Outcome in patients with idiopathic inflammatory myositis: morbidity and mortality. *Rheumatology* (Oxford) 2002; 41(1): 22-26. https://doi.org/10.1093/rheumatology/41.1.22
56. MAUGARS YM, BERTHELOT JM, ABBAS AA, MUSSINI JM, NGUYEN JM, PROST AM: Long-term prognosis of 69 patients with dermatomyositis or polymyositis. *Clin Exp Rheumatol* 1996; 14(3): 263-74.