Mortality in idiopathic inflammatory myopathies

I.E. Lundberg[1], C.J. Forbess[2]

[1]*Rheumatology Unit, Department of Medicine at Karolinska University Hospital in Solna, Karolinska Institutet, Stockholm, Sweden;*
[2]*Division of Rheumatology, NYU Hospital for Joint Diseases, New York University School of Medicine, New York, NY, USA.*

Ingrid E. Lundberg, MD
Chelsey J. Forbess, BA

Please address correspondence to:
Ingrid E. Lundberg, Rheumatology Unit, Karolinska University Hospital, SE-17176 Stockholm, Sweden.
E-mail: Ingrid.Lundberg@ki.se

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The idiopathic inflammatory myopathies, collectively named "myositis," comprise a heterogeneous group of disorders with primary clinical features of muscle weakness and low muscle endurance. Based on some clinical and histopathological differences, the idiopathic inflammatory myopathies are classified into three major subgroups: polymyositis, dermatomyositis, and inclusion body myositis (1). Typical cases are characterized by inflammatory cell infiltrates in muscle tissue.

Organs other than muscle frequently are involved, particularly in polymyositis and dermatomyositis. Skin involvement is a characteristic feature of dermatomyositis. Other organs affected in many patients with polymyositis and dermatomyositis are the lungs, the heart, the gastrointestinal tract, and the joints. Lung involvement is common and may be seen in up to 70% of polymyositis and dermatomyositis patients if sensitive tools such as high resolution computerized tomography and pulmonary function tests are used as routine investigations (2). Lung involvement in myositis may be due to respiratory muscle involvement, interstitial lung disease, or less often pulmonary hypertension. The most frequent gastrointestinal symptoms are dysphagia, caused by weakness of the muscles in the tongue, pharynx, or esophagus, and dysmotility of the esophagus which may occur in up to 50% of myositis patients (3). Dysphagia is a common manifestation in patients with inclusion body myositis and may be an early feature of disease. Both the weakness of respiratory muscles and the dysphagia may lead to aspiration and pulmonary infections. Cardiac involvement, in particular subclinical electrocardiogram (ECG) changes, is frequently seen in polymyositis and dermatomyositis, although manifest heart problems are uncommon (4). Cardiomyopathy may occur due to inflammation of the heart muscle as a myocarditis, but rhythm disturbances and arteriosclerosis are more common.

The idiopathic inflammatory myopathies may occur together with other autoimmune diseases as a so-called overlap myositis. The most common auto-immune comorbidities are systemic sclerosis, Sjögren's syndrome, systemic lupus erythematosus (SLE), mixed connective tissue disease (MCTD), and rheumatoid arthritis. There is also an overrepresentation of malignancies in patients with myositis, foremost in adult dermatomyositis, whereas the association between malignancies and polymyositis is less established, and no association between malignancies and inclusion body myositis or juvenile dermatomyositis has been reported. As expected, involvement of extramuscular organs as well as the association between dermatomyositis and malignancies may affect morbidity and mortality.

Survival in myositis

Before the introduction of glucocorticoids in the treatment of polymyositis and dermatomyositis, the mortality was reported to be 50% in a study of untreated cases within a follow-up period of unspecified length (5). Studies from the first 2 decades with glucocorticoid treatments did not find any evidence that treatment had any positive effect on survival in polymyositis and dermatomyositis; 5-year survival rate was reported as 60% (6, 7). These early studies may be criticized for the heterogeneity of patients.

Later studies suggest improved prognosis and survival of patients with inflammatory myopathies. Although the proposed diagnostic criteria originally published by Bohan and Peter in 1975 have been used in most studies, patient selection continues to vary, rendering direct comparisons between different studies difficult (8, 9). Moreover, one obvious weakness of the Bohan and Peter criteria is that inclusion body myositis was not identified as a distinct subset when these criteria were proposed;

Table I. Mortality studies in myositis.[¶]

Study	Year of study	Location	Source	No. of patients with myositis	Mean age	Mean duration of follow-up (years)	% women	Survival at 5 years	Survival > 5 years	Info on causes of death?	Data on risk factors?
Medsger *et al*. (6)	1971	TN, USA	Clinic	124 (PM)	--	7[§]	59.7	65%	53% (8 years)	No	Yes
Benbassat *et al*. (16)	1985	Israel	Population	92 (PM/DM)	--	20[§]	63.0	52%*	32% (total mortality)	Yes	Yes
Hochberg *et al*. (10)	1986	MD, USA	Rheum Unit	76 (PM/DM)	45.3	8	75.0	80.4%	72.8% (7 years)	Only for most common causes	Yes
Marie *et al*. (11)	2001	France	Clinic	77 (PM/DM)	52^	4^	57.1	77%	61% (15 years)	Yes	Yes
Sultan *et al*. (12)	2002	UK	Clinic (One rheum center)	46 (IIM)	38.9	20[§]	2.5:1 ratio	95%	83.8% (10 years)	Yes	No
Dankó *et al*. (13)	2004	Hungary	Clinic	162 (IIM)	39.2	8.46^	2.1:1 ratio	92%	89% (10 years)	Yes	Yes
Airio *et al*. (14)	2006	Finland	Population	176 (PM), 72 (DM)	56 (PM), 53 (DM)	11^	63 (PM), 53 (DM)	75% (PM), 63% (DM)	55% (PM), 53% (DM)	Yes	Yes
Torres *et al*. (15)	2006	Spain	Clinic	107 (IIM)	42.3	9[§]	70.1	80%	71% (10 years)	Yes	Yes

[¶] Some studies noted in the text that are not included in the tables do not include relevant quantitative information; [§] total number of follow-up years; ^ given as median, * as listed in review article.
IIM: idiopathic inflammatory myopathies; PM: polymyositis, DM: dermatomyositis.

therefore, these patients may have been included in older studies as polymyositis, which may affect the outcome and survival rate.

A limitation of most studies on mortality in myositis is that they are based on cohorts followed at a single or a few medical centers, and very few population-based studies have been presented. As the inflammatory myopathies are heterogeneous, these patients may be cared for by different specialists depending on the main clinical problem in the individual case. Thus cohort studies from one clinic or one medical center may infer a selection bias towards more or less severe cases. Another problem of some studies is that long-term survival, *e.g*., 5-year survival, cannot be determined if the follow-up period is less than 5 years.

With these limitations in mind, the 1-, 5- and 10-year survival rates have increased since the 1960s from a 5-year survival rate of 65% in 1971 (6) to 75-95% in later studies (10-15), with the exception of the 52% 5-year survival rate in an Israeli study from 1985 (16) (Table I). The wide variation in 5-year survival could be explained by patient selection. In general, a longer survival time is reported in cohorts in which patients with malignancies have been excluded or are few, and in studies with a bias towards inclusion of cases with overlap myositis, juvenile cases, and adults with lower mean age at onset (12, 13, 17). Moreover, in the study with the highest survival rate, there was a high dropout rate, and only half of the primary polymyositis and dermatomyositis patients had an observation time of a minimum of 5 years. However, these cases were still included in the data analyses, and the long-term survival data in this study are unreliable (13).

Survival in the few recent population-based studies was not as favorable (14, 18). In a recently published population-based study from Finland, including 248 patients with a follow-up period of 20 years, the 5-year survival was 75% for polymyositis and 63% for dermatomyositis, and the combined 10-year survival was 50% (14). The median survival for polymyositis was 11.0 years (95% CI: 9.5-13.3), and for dermatomyositis 12.3 years (95% CI: 5.5-20.7). A limitation of the Finnish study, which was based on the hospital discharge register, is the risk of a bias towards more severe cases, although the authors claim that most patients with a suspicion of polymyositis or dermatomyositis in Finland are still referred to a hospital for inpatient examinations. Furthermore, by definition, all patients presented many years ago, and the survival in Finland may be better today due in part to better general medical care. Information on survival rate on inclusion body myositis is limited, but it appears to be longer than for polymyositis or dermatomyositis, with 5-year survival reported as 95% (5).

Mortality in polymyositis and dermatomyositis

The overall standard mortality ratio (SMR) in patients with polymyositis and dermatomyositis was 2.92 (95% CI 2.48-3.44) compared to the general population, in the Finnish study, which covered patients who were diagnosed between 1969 and 1985 and who had an adequate follow-up time of 10 years (in 1995) (14). In a smaller population-based study from New Zealand, the death rate was 33% after a median follow-up of 76 months (18).

The case fatality ratio (CFR) varies substantially between studies (2%-45%), which again can be explained by patient

Table II. Causes of death in patients with myositis (% total deaths).[¶]

	Henriksson *et al.* 1982 (21)	Benbassat *et al.* 1985 (16)	Marie *et al.* 2001 (11)	Sultan *et al.* 2002 (12)	Dankó *et al.* 2004 (13)	Airio *et al.* 2006 (14)	Torres *et al.* 2006 (15)	Bronner *et al.* 2006 (19)
Total no. deaths	25/107 (23.3%)	30/92 (32.6%)	17/77 (22.1%)	6/46 (13.0%)	20/162 (12.3%)	149/248 (60.1%)	28/102 (27.5%)	34/161 (21.1%)
Cardiovascular	64.0	16.7		50.0	55.0	36	21.4	14.7
(Myocardial Infarction)	20.0			(16.7)				
Cerebrovascular Disease	8.0	3.3				3		3.0
Cancer	12.0	16.7	47.1	#	10.0	16	35.7	26.5
Renal		10.0				1		
Respiratory	36.0					5		
Pulmonary		26.7	5.9		20.0		14.3	11.8
Pneumonia	20.0		29.4	33.3				
Infection						2.5	7.1	
Musculoskeletal Diseases			5.9*			30[§]		
Uncontrolled activity		10.0						
GI			11.8		5.0	3		
Accidents or intoxication	4.0			16.7		1		11.8
Other/unknown	16.0	16.7			10.0	2	21.4	32.3
Total percent	132.0¤	100.1	100.1	100.0	100.0	99.5	99.9	100.1

[¶] Some studies noted in the text that are not included in the tables do not include relevant quantitative information, ¤ more than one cause recorded in some patients, * classified as "generalized muscle weakness"; [#] cancer associated myositis excluded; [§] pneumonia was the cause of death in 40% of these.

selection and by variations in observation time. Furthermore, to be clinically relevant the CFR must be compared to the mortality ratio in the general control population (7, 10, 17). A mortality rate among myositis patients of 21% was reported in a recent multicenter study from the Netherlands that included 165 adult polymyositis and dermatomyositis patients with a median follow-up period of 5 years. This rate was substantially higher than the expected 1% to 2% in an age-matched healthy Dutch population (19).

Causes of death in myositis

The most common causes of death in patients with polymyositis and dermatomyositis are malignancies, infections, profound effects of muscle weakness, and cardiovascular disease (Table II) (11-16, 20, 21). The ranking order of causes of death in one more recent study was cardiovascular, infection (mainly pneumonia), and cancer (Table II) (14) . In other studies, cancer has been the most common cause of death, but the studies are relatively small and the number of fatalities in each study is low (11, 15, 18, 19). Respiratory failure is also among the more common causes of death (11, 13, 18, 19), considerably higher than in the general population. Opportunistic infections were recently found to be a frequent cause of death in a French study that included 156 patients from three medical centers (22). The most common sites of the opportunistic infections were the lungs and the digestive tract.

In a Hungarian study, pulmonary complications as a cause of death occurred within the first 12 months after diagnosis, while cardiovascular complications predominated among deaths after 5 years' disease duration (13). There may be a difference in causes of death between polymyositis and dermatomyositis, as, in the large Finnish study, patients with dermatomyositis had a greater risk of dying from cancer than polymyositis patients [hazard ratio (HR) 5.11, 95% CI: 2.31-11.3] (14).

Prognostic factors affecting survival in myositis patients

The most important predictor of mortality is age, with a worse prognosis with older age at onset (10, 13-16) (Table III). Male patients with myositis had a worse mortality prognosis compared to women in some studies (13, 15), but most studies report no sex difference in survival (10, 14, 16). In the study by Medsger *et al*., non-whites had worse prognosis than whites due to more severe muscle weakness and dysphagia among the non-whites (6). An ethnic difference could not be confirmed in the study by Hochberg *et al*. in 1986 (10). Most studies published thereafter have included primarily Caucasians. Smoking was a risk factor for mortality in the study by Torres *et al*. (15).

Clinical factors associated with death are involvement of the cardiovascular system, with a cumulative survival rate after 8 years of 44.2% in patients with cardiac involvement, compared to 87% for those without clinical heart disease (10). More specific cardiovascular involvement, namely left ventricular dysfunction, conferred a risk factor for mortality in another study (15). Cardiac involvement was the major risk factor in some other recent studies (12, 13).

Involvement of the respiratory system may also confer a risk factor for mortality either from respiratory muscle involvement alone or together with dysphagia leading to pneumonia (11, 13, 15, 18). Interstitial lung disease is a frequent finding in polymyositis and dermatomyositis (2). In most cases, this is slowly progressive and improves with immunosuppressive treatment (23). However, in rare instances the interstitial lung disease may have a rapidly fatal course (23-26). In a report from one

Table III. Predictors of premature mortality in patients with myositis (hazard ratios with 95% confidence intervals).¶

	Medsger *et al.* 1971 (6)	Benbassat *et al.* 1985 (16)	Hochberg *et al.* 1986 (10)	Marie *et al.* 2001 (11)	Dankó *et al.* 2004 (13)	Airio *et al.* 2006 (14)	Torres *et al.* 2006 (15)	Bronner *et al.* 2006 (19)
Total no. deaths	43	30	13	17	20	149	28	34
Age (adults)	NS	p=0.0304* (for age>61)	p<0.005 (for age≥45)		p=0.0217 (for age≥45, in DM pts)	1.09 (1.06-1.11) (PM) p<0.001 1.07 (1.03-1.11) (DM) p<0.001	1.04 (1.02-1.07)	2.7 (1.0-7.5) (for age >60)
Age (between adults and children)	p<0.02							
Black (*vs.* White)	p<0.02		NS					
Male sex		p=0.8802*	NS		p=0.0382 (in DM pts)	1.29 (.83-2.02) (PM) p=0.26 1.35 (.54-3.70) (DM) p=0.051	0.1 (.01-.9) (in PM pts)	
Smoking habit							2.8 (1.2-6.6)	
Disease history		p=0.0082*						
Delay of diagnosis, months						1.01 (1.00-1.02) (PM) p=0.006		
Interval from clinical manifestation to treatment <6 months								7.7 (1.0- 58.0)
Pneumonitis	p<0.001							
Weakness (unable to move parts against gravity)	NS			0.18^ (.03-1.08) (p=0.03)				
Dysphagia (yes/no)	p<0.01	p=0.0066*	NS		p<0.01 (in PM pts)			
Decreased urine creatinine	p<0.05							
Low serum Albumin	NS							
SGOT elevation	NS							
Temperature (>38ºC)		p=0.0001*						
White blood cells (>10,000/μl)		p<0.0001*						
Antinuclear antibodies				1.05^ (.38-2.86)				
Anti-J01 antibody				1.29^ (0.11-9.85)				
ESR						NS		
Microangiography at nailfold capillaroscopy				0.70^ (0.23-8.16)				
Skin rash (yes/no)		p=0.1482*					NS	
Clinical remission achieved (yes/no)		p<0.0001*						
Cardiac involvement			p<0.05		3.182 (p<0.01) (for all IIM pts)		4.8 (2.2-10.5)	
Left ventricular dysfunction							5.2 (2.4-11.1)	
Respiratory Muscle Involvement					1.16 (p=0.045) (for all IIM pts)			
Initial myalgias				0.25^ (.04-1.32) p=0.056				
Raynaud's Phenomenon				1.17^ (.39-3.46) p=1.17			3.5 (1.2-10.2) (in IM group excluding cancer-associated IM)	
Esophageal dysfunction				1.50^ (.50-4.46)				
Dysphonia				1.37^ (.11-10.43)				
Joint involvement				0.74^ (.22-2.43)				
Pulmonary impairment				5.73^ (1.90-17.31) p=0.0011			2.6 (1.0-7.0) (in IM group excluding cancer-associated IM)	
Aspiration pneumonia				29.33^ (4.85-293.40) p<0.0001				
Ventilatory insufficiency				6.00^ (1.49-24.69)				
Interstitial lung disease				1.08^ (0.22-4.42)	p=0.0228 (in DM pts)			
Cancer				27.08^ (5.42-168.90) p<0.0011	1.99 (1.01-3.94) (for PM)	3.9 (1.8-8.5) p=0.047 2.16 (.95-4.50) (for DM) p=0.062	7.8 (1.5-40.4)	
Inflammatory infiltrates				1.14^ (0.28-5.59)				
Necrosis of muscle fibers				1.06^ (0.38-2.94)				
Vascular damage				3.39^ (1.15-10.02)				

¶Some studies noted in the text that are not included in the tables do not include relevant quantitative information; *p-value from univariate Mantel-Cox test of factors affecting survival; ^listed as Odds Ratio (OR)
IIM: idiopathic inflammatory myopathies; PM: polymyositis; pts: patients; DM: dermatomyositis; NS: not significant.

hospital including 6 patients with dermatomyositis who required hospitalization in intensive care units (ICU), the primary reason for admission to ICU was acute respiratory failure (25). Two of these patients died from respiratory complications. In another study of 99 patients with newly diagnosed polymyositis or dermatomyositis, 11 patients had an acute form of interstitial lung disease and 8 of these patients died of respiratory failure within 1 to 2 months despite treatment with glucocorticoids (26). In the same study, patients with interstitial lung disease defined by chest radiograph had a significantly lower 3-year survival (61.6%) compared to patients who did not have interstitial lung disease (89.8%), and this increased risk of mortality was explained by the high death rate among the patients with the acute form of interstitial lung disease (26).

The presence of cancer was associated with increased mortality in several studies (11, 14, 15, 18, 19), but not in all (10). Cancer accounted for 50% of the deaths in the studies by Lynn et al (18) and Marie *et al*. (11), 36% in the study by Torres *et al*. (15), 20% in the study by Bronner *et al*. (19), and 16% in the study by Airio *et al*. (14).

Subgroup-specific prognostic factors

In the population-based Finnish study, the patients with dermatomyositis had a 1.47-fold (95% CI: 0.99 -2.12) age- and sex-adjusted mortality rate compared to those with polymyositis (p=0.08), mostly explained by the higher risk of dying from cancer among patients with dermatomyositis (14). After adjusting for age, patients with dermatomyositis had a 5-fold greater risk of dying from cancer than did polymyositis patients (14). A delay in diagnosis and a lower initial dose of glucocorticoids were prognostic factors for mortality in polymyositis but not dermatomyositis patients in the population-based study by Airio *et al*. (14).

Summary

Patients with polymyositis and dermatomyositis have increased mortality rates, almost 3-fold higher compared to age- and sex-matched individuals in one of the few published population-based studies (14). The 5-year survival rate has increased since the 1950s and 1960s but is still lower than expected in unselected population-based studies. Whether the 10-year survival rate has improved is uncertain. The primary causes of death are cardiovascular disease, aspiration pneumonia, cancer, and respiratory failure, with varying frequencies in different cohort studies most likely depending on patient selection and variation in observation times.

The most important variable affecting survival rate is age at disease onset. It remains uncertain whether gender or ethnicity is associated with reduced survival. The major disease-specific factors affecting survival are involvement of the cardiovascular and respiratory systems and presence of a malignancy. Patients with dermatomyositis have a 5-fold greater risk of dying from cancer compared to polymyositis patients, whereas delay in diagnosis is a risk factor for mortality in polymyositis. Mortality data in patients with inclusion body myositis are scarce, but survival seems to be longer than for polymyositis and dermatomyositis. In conclusion, survival prognosis has improved for polymyositis and dermatomyositis since the 1960s but the overall mortality is still increased compared to the general population. More population-based and clinic-based long-term studies are clearly needed to clarify risk factors for mortality and to evaluate how new interventions affect the long-term prognosis, including survival rates in myositis.

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