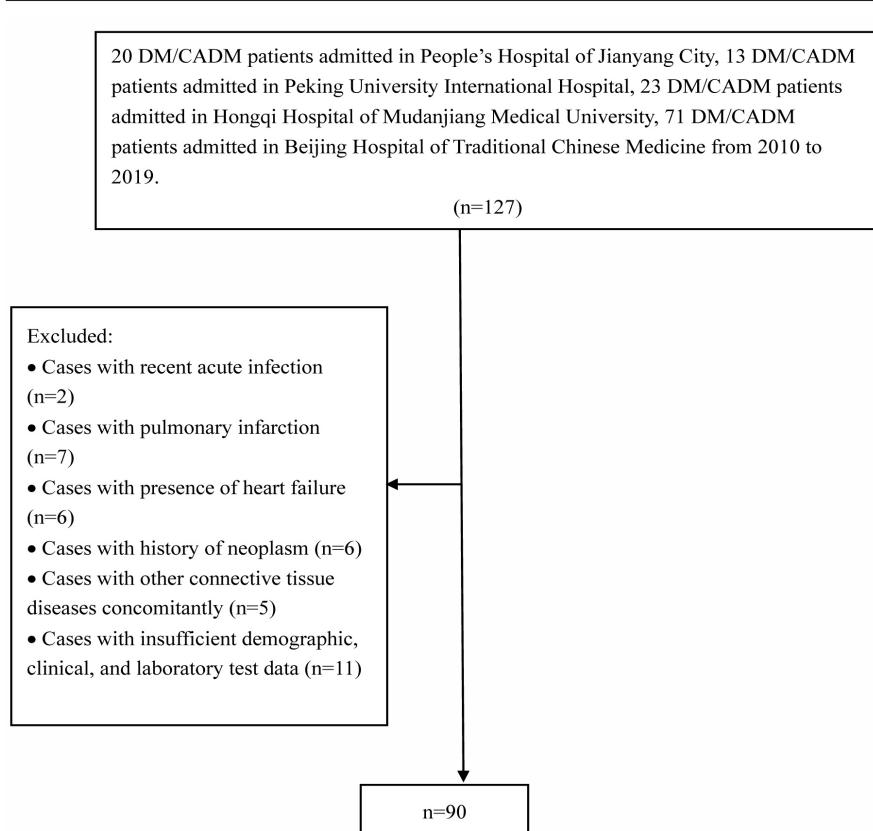


Supplementary Fig. S1. Flow chart of inclusion/exclusion criteria applied to Peking University People's hospital patients.



Supplementary Fig. S2. Flow chart of external validation set.

Supplementary Table S1. Comparison of clinical and laboratory manifestations between the development cohort and the validation cohort, the external validation cohort.

	DC (n =239)	IVC (n=128)	EVC(n=90)	P DC vs. IVC	P DC vs. EVC	P IVC vs. EVC
Demographics						
Age at onset, years	48.0 ± 14.3	48.7 ± 13.5	57.4 ± 12.1	0.617	0.000	0.000
Female (n, %)	188 (78.7)	97 (75.8)	60 (66.7)	0.528	0.024	0.140
Subtype of IIM				0.619	0.000	0.000
DM (n, %)	113 (47.3)	64 (50.0)	77 (85.6)			
CADM (n, %)	126 (52.7)	64 (50.0)	13 (14.4)			
Clinical characteristics						
RP-ILD (n, %)	70 (29.3)	40 (31.3)	18 (20.0)	0.696	0.090	0.064
Gottron's sign/papules (n, %)	173 (72.4)	90 (70.3)	37 (41.1)	0.675	0.000	0.000
Mechanic's hands (n, %)	76 (31.8)	46 (35.9)	38 (42.2)	0.423	0.077	0.348
Heliotrope rash (n, %)	120 (50.2)	56 (43.8)	28 (31.1)	0.238	0.002	0.059
V sign (n, %)	113 (47.3)	52 (40.6)	22 (24.4)	0.222	0.000	0.013
Shawl sign (n, %)	64 (26.8)	28 (21.9)	19 (21.1)	0.302	0.291	0.893
Skin ulceration (n, %)	11 (4.6)	10 (7.8)	4 (4.4)	0.207	1.000	0.473
Periungual erythema (n, %)	48 (20.1)	17 (13.3)	7 (7.8)	0.104	0.008	0.201
Myalgia (n, %)	26 (10.9)	16 (12.5)	28 (31.1)	0.642	0.000	0.001
Myasthenia (n, %)	117 (49.0)	62 (48.4)	43 (47.8)	0.925	0.849	0.924
Velcro rales (n, %)	38 (15.9)	23 (18.0)	77 (85.6)	0.612	0.000	0.000
Fever (n, %)	100 (41.8)	53 (41.4)	15 (16.7)	0.936	0.000	0.000
Dysphagia (n, %)	4 (1.7)	1 (0.8)	8 (8.9)	0.818	0.005	0.009
Arthralgia (n, %)	111 (46.4)	65 (50.8)	13 (14.4)	0.428	0.000	0.000
Raynaud's phenomenon	10 (4.2)	7 (5.5)	4 (4.4)	0.577	1.000	0.979
Myositis-specific antibodies						
Anti-ARS positivity	53 (22.2)	29 (22.7)	57 (63.3)	0.916	0.000	0.000
Anti-Jo-1 positivity	19 (7.9)	6 (4.7)	14 (15.6)	0.237	0.041	0.006
Anti-MDA5 positivity	32 (13.4)	17 (13.3)	19 (21.1)	0.977	0.084	0.125
Myositis-associated antibodies						
Anti-Ro-52 positivity	68 (28.5)	42 (32.8)	30 (33.3)	0.385	0.388	0.936
Anti-PM/Scl-75/100 positivity	1 (0.4)	1 (0.8)	6 (6.7)	1.000	0.002	0.042
ANA	159 (66.5)	88 (68.8)	35 (38.9)	0.665	0.000	0.000
Laboratory features						
Arterial blood gas						
PaO ₂ , mean ± SD mmHg	85.0 ± 14.4	71.3 ± 14.8	80.8 ± 26.8	0.000	0.401	0.050
HRCT						
NSIP	5 (2.1)	9 (7.0)	6 (6.7)			
OP	215 (90)	69 (53.9)	47 (52.2)			
NSIP+OP	19 (7.9)	46 (35.9)	24 (26.7)			
UIP	0 (0)	3 (2.3)	3 (3.3)			
DAD	0 (0)	0 (0)	0 (0)			
Pulmonary function tests ^a						
FVC % predicted	83.9 ± 17.5	67.0 ± 18.1	85.2 ± 17.8	0.000	0.776	0.000
DLco % predicted	67.4 ± 19.6	53.5 ± 18.2	78.3 ± 22.1	0.000	0.032	0.000
TLC % predicted	85.7 ± 16.9	73.3 ± 17.3	102.3 ± 45.7	0.000	0.003	0.000
Bronchoalveolar lavage ^b						
Total cell number, 10 ⁵ /ml	0.40 ± 0.37	0.30 ± 0.28	0.17 ± 0.03	0.068	0.234	0.502
Macrophage, %	63.7 ± 21.6	48.0 ± 22.2	57.0 ± 13.7	0.000	0.606	0.484
Lymphocyte, %	27.4 ± 20.0	37.8 ± 23.1	36.0 ± 17.0	0.005	0.503	0.886
Neutrophil, %	7.4 ± 10.1	12.9 ± 18.3	5.7 ± 4.7	0.033	0.848	0.418
Eosinophil, %	1.5 ± 2.7	1.2 ± 2.8	1.3 ± 1.5	0.429	0.902	0.915
Elevated ALT (n, %)	95 (39.7)	54 (42.2)	34 (37.8)	0.650	0.744	0.514
Elevated AST (n, %)	106 (44.4)	66 (51.6)	38 (42.2)	0.187	0.729	0.174
Elevated LDH (n, %)	159 (66.5)	88 (68.8)	60 (66.7)	0.665	0.981	0.746
ESR	29.7 ± 25.4	29.2 ± 22.9	24.7 ± 21.5	0.834	0.094	0.180
Death	70 (29.3)	40 (31.3)	18 (20.0)	0.907	0.012	0.023
OS, years	5.2 ± 5.7	5.0 ± 5.5	2.3 ± 1.1	0.647	0.000	0.000

^aNumber of subjects with DC, n=106, Number of subjects with IVC, n=58, Number of subjects with EVC, n=17; ^bNumber of subjects with DC, n=62, Number of subjects with IVC, n=80, Number of subjects with EVC, n=3. The LSD of analysis of variance and chi square test were used in a pair-wise post-hoc analysis. P values were two-sided, and values of < 0.05 were considered statistically significant. DC: development cohort; IVC: internal validation cohort; EVC: external validation cohort; RP-ILD: rapidly progressive interstitial lung disease; DM: dermatomyositis; CADM: clinically amyopathic dermatomyositis; ARS include Jo-1, EJ, OJ, PL-7, PL-12, KS. ARS: aminoacyl-tRNA synthetase; MDA5: melanoma differentiation-associated 5; PM/Scl: polymyositis/scleroderma; ANA: antinuclear antibodies; RF: rheumatoid factor; ALT: alanine transaminase; AST: aspartate aminotransferase; ESR: erythrocyte sedimentation rate; LDH: lactate dehydrogenase.