Diagnostic and classification criteria

JDM was diagnosed and categorized according to Bohan and Peter's criteria. The inclusion criteria were: 1) ≤18 years of age; 2) refractory JDM, defined as patients with no clinical improvement following treatment with combined therapy of glucocorticoids and over two kinds of immunosuppressant or biological agents; or recurrence during the tapering of glucocorticoids dosage; or relapse of JDM symptoms within 3 months after discontinuation of glucocorticoids [1]; and 3) treated with JAKi. The exclusion criteria were: 1) other chronic or inflammatory diseases; 2) ongoing infection; or 3) severe gastrointestinal tract involvement.

Supplementary Table 1. Characteristics of the patients

	N=25
Female	11 (44%)
Ethnic group (Han/Minority)	24/1
Age at disease onset (years)	4.6±3.3
Age at starting JAKi (years)	7.2±4.0
Age at the end of follow-up (years)	7.9±4.0
Diagnosis (JDM/CADM)	23/2
Refractory	
No response to GC+IA$^{>2}$/BA	8 (32%)
Relapse†	17 (68%)
Other diagnoses	
Cataract	3 (12%)
Compression fracture	1 (4%)
Hypertension	1 (4%)
Interstitial lung disease	4 (16%)
Joint contracture	1 (4%)
Symptoms at disease onset	
Rash‡	24 (96%)
Heliotrope rash	22 (88%)
Malar/facial erythema	24 (96%)
Gottron's sign	18 (72%)
Skin ulcer	10 (40%)
Muscle weakness	17 (68%)

Dysphagia/hoarseness/drinking cough/low voice	8 (32%)
Fever	6 (24%)
Maximum CK (U/L)	594 (276-1203)
Maximum LDH (U/L)	548 (389-660)
Myositis-specific antibodies (MSA)	
Negative¶	14 (56%)
Anti-NXP2 Positive	8 (32%)
Anti-MDA5 Positive	1 (4%)
Anti-JO-1 Positive	1 (4%)
Anti-PL-7 Positive	1 (4%)
Anti KU Positive	1 (4%)
ANA positive	6 (24%)
Anti-Ro-52 positive	5 (20%)
Therapy before JAKi started	
GC+IA$^{<=2}$	9 (36%)
GC+IA$^{>2}$	7 (28%)
GC+IA+IVIG	4 (16%)
GC+IA+IVIG+BA	3 (12%)
GC+IA+IVIG+plasma exchange/ASCT	2 (8%)
Therapy when JAKi started	
GC	3 (12%)
IA$^{<=2}$	2 (8%)
GC+IA$^{<=2}$	15 (60%)
GC+IA$^{>2}$	1 (4%)
GC+IA$^{<=2}$+IVIG	3 (12%)
GC+IA$^{<=2}$+BA	1 (4%)
Disease course of JDM before JAKi treatment (months)	21.0 (14.0-36.5)
JAKi, <25 kg, 2.5 mg bid	11 (44%)
JAKi, ≥25 kg, 5 mg bid	14 (56%)
Ruxolitinib	18 (72%)
Tofacitinib	7 (28%)
Follow up (months)	7 (4.0-12.0)

Continuous data are presented as mean ± SD or median (interquartile range). Category data are presented as number and percentage.

JAKi: Janus-kinase inhibitor; JDM: juvenile dermatomyositis; CADM: clinically amyopathic dermatomyositis; CK: creatinine kinase; LDH: lactate dehydrogenase; NXP2: nuclear matrix protein 2; MDA5: melanoma differentiation-associated gene 5; ANA: antinuclear antibodies; GC: glucocorticoids; IA: immunosuppressive agents; $IA^{<=2}$: two or fewer types of IA; $A^{>2}$: more than two types of IA; IVIG: intravenous immunoglobulins; BA: biological agents.

† Relapsing: manifestations recur during GC dosage reduction or within 3 months after GC withdrawal.

‡ Rash: Heliotrope rash; Malar/facial erythema; Gottron's sign.

¶ In six patients, the antibodies were tested after treatment in other hospitals.

Supplementary Table 2. Disease activity markers before and after JAK inhibitor treatment

	n	Before JAKi	After JAKi	P
CAT-BM activity score	25	7.0 (3.0-10.0)	0.0 (0.0-1.0)	<0.001*
CAT-BM damage score	25	0.0 (0.0-1.0)	0.0 (0.0-1.0)	--
CMAS	10	24.9±18.0	38.2±7.4	0.023*
CK (U/L)	12	107.5 (79.8-177.8)	124.0 (61.0-178.8)	0.814
LDH (U/L)	12	306.3 (361.5-463.3)	275.8 (291.0-394.8)	0.034*
SF (ng/mL)	25	81.2 (55.4-113.0)	64.3 (37.2-98.2)	0.010*
Hospitalizations in 6 months	12	2.5 (1.0-4.8)	0.0 (0.0-2.8)	0.028*

Continuous data are presented as mean ± SD or median (interquartile range). Category data are presented as number and percentage.

CAT-BM: Cutaneous Assessment Tool Binary Method; CMAS: Childhood Myositis Assessment Scale; CK: creatinine kinase; LDH: lactate dehydrogenase; SF: serum ferritin.

* Significantly statistic difference, P<0.05.