

**Supplementary Table S1.** Publications in adult DM that have used JAK inhibitors in refractory disease.

Publication	Type of study	N	JAK inhibitor (dose)	Refractory cutaneous disease, n	Refractory muscle disease, n	ILD, n
Hornung <i>et al.</i> (2014) (1)	Case report	1	Ruxolitinib (5 mg QD–15 mg BID)	1	1	0
Paik and Christopher-Stine (2017) (2)	Case report	1	Tofacitinib (5 mg BID)	1	1	0
Hornig <i>et al.</i> (2018) (3)	Case report	1	Tofacitinib (5–10 mg QD)	1	0	1
Kurasawa <i>et al.</i> (2018) (4)	Case series	5	Tofacitinib (5 mg BID)	5	0	5
Chen <i>et al.</i> (2019) (5)	Open label	18	Tofacitinib (5 mg BID)	NR	0	18
Kato <i>et al.</i> (2019) (6)	Case report	1	Tofacitinib (5 mg BID)	0	0	1
Landon-Cardinal <i>et al.</i> (2019) (7)	Case series	12	Baricitinib (NR), ruxolitinib (NR)	12	3	0
Allenbach <i>et al.</i> (2018) (8)*	Case series	3	Baricitinib (NR), ruxolitinib (NR)	3	1	0
Ladislau <i>et al.</i> (2018) (9)*	Case series	4	Ruxolitinib (30 mg QD)	4	2	0
Moghadam-Kia <i>et al.</i> (2019) (10)	Case series	4	Tofacitinib (5 mg BID)	4	1	0
Wendel <i>et al.</i> (2019) (11)	Case series	2	Tofacitinib (5 mg BID)	2	1	1
Conca <i>et al.</i> (2020) (12)	Case report	1	Tofacitinib (5 mg BID)	0	0	1
Delvino <i>et al.</i> (2020) (13)	Case report	1	Baricitinib (4 mg QD)	1	1	0
Fetter <i>et al.</i> (2020) (14)	Case report	1	Ruxolitinib (10 mg BID–30 mg QD)	1	1	0
Fischer <i>et al.</i> (2020) (15)	Case report	1	Baricitinib (4 mg QD)	1	1	0
Hosokawa and Oiwa (2020) (16)	Case report	1	Tofacitinib (5 mg BID)	0	0	1
Ishikawa <i>et al.</i> (2020) (17)	Case report	1	Tofacitinib (10 mg QD)	1	0	1
Jalles <i>et al.</i> (2020) (18)	Case report	1	Ruxolitinib (15 mg BID)	1	1	1
Riggle <i>et al.</i> (2020) (19)	Case series	4	Tofacitinib (11 mg QD)	4	0	0
Shinjo and de Souza (2020) (20)	Case report	1	Tofacitinib (5 mg BID)	1	0	0
Takatani <i>et al.</i> (2020) (21)	Case report	1	Tofacitinib (10 mg QD)	0	0	1
Williams and McKinney (2020) (22)	Case report	1	Tofacitinib (11 mg QD)	1	1	0
Crespo Cruz <i>et al.</i> (2021) (23)	Case report	1	Tofacitinib (5 mg BID)	1	0	0
Jasmine <i>et al.</i> (2021) (24)	Case report	1	Tofacitinib (11 mg QD)	1	0	0
Machiyama <i>et al.</i> (2021) (25)	Case report	1	Tofacitinib (5 mg BID)	1	0	1
Min <i>et al.</i> (2021) (26)	Retrospective	9	Tofacitinib (5–11 mg BID)	9	2	0
Kurtzman <i>et al.</i> (2016) (27)†	Case series	3	Tofacitinib (5–10 mg BID)	3	2	0
Alsarheed <i>et al.</i> (2018) (28)‡	Case series	7	Tofacitinib (5–10 mg BID)	7	5	0
Navarro-Navarro <i>et al.</i> (2021) (29)	Case series	2	Tofacitinib (5 mg BID)	2	1	0
Ohmura <i>et al.</i> (2021) (30)	Case report	1	Tofacitinib (5–10 mg BID)	1	0	1
Paik <i>et al.</i> (2021a) (31)	Open label	10	Tofacitinib (11 mg QD)	10	1	0
Paik <i>et al.</i> (2020) (32)‡	Open label	7	Tofacitinib (11 mg QD)	7	0	0
Paik <i>et al.</i> (2021b) (33)‡	Open label	7	Tofacitinib (11 mg QD)	7	0	0
Shneyderman <i>et al.</i> (2021) (34)‡	Open label	3	Tofacitinib (11 mg QD)	3	0	0

\*Some or all patients likely to have been reported in Landon-Cardinal *et al.* (2019) and were not included in the count of unique patients (7).

†Some or all patients likely to have been reported in Min *et al.* (2021) and were not included in the count of unique patients (26).

‡Patients reported in Paik *et al.* (2021a) and were not included in the count of unique patients (31).

BID: twice daily; DM: dermatomyositis; ILD: interstitial lung disease; JAK: Janus kinase; NR: not reported; QD: once daily.

**Supplementary Table S2.** Publications in JDM that have used JAK inhibitors in refractory disease.

Publication	Type of study	N	JAK inhibitor (dose)	Refractory cutaneous disease, n	Refractory muscle disease, n	ILD, n
Sabbagh <i>et al.</i> (2019) (35)	Case series	2	Tofacitinib (5–10 mg BID)	2	2	2
Papadopoulou <i>et al.</i> (2019) (36)	Case report	1	Tofacitinib (6 mg BID)	1	1	0
Ding <i>et al.</i> (2020) (37)	Retrospective	25	Ruxolitinib (2.5 mg BID), tofacitinib (5 mg BID)	24	10	4
El-Lateef (2020) (38)	Case report	1	Ruxolitinib (5 mg BID)	1	0	0
Kim <i>et al.</i> (2020a) (39)	Open label	4	Baricitinib (2–4 mg BID)	4	2	0
Kim <i>et al.</i> (2020b) (40)*	Open label	4	Baricitinib (2–4 mg BID)	4	2	0
Sozeri and Demir (2020) (41)	Case series	2	Tofacitinib (5 mg BID)	2	2	0
Heinen <i>et al.</i> (2021) (42)	Case report	1	Ruxolitinib (20–30 mg QD)	1	1	0
Kostik <i>et al.</i> (2021) (43)	Case series	2	Tofacitinib (0.5–0.7 mg/kg QD)	2	0	0
Min <i>et al.</i> (2021) (26)	Retrospective	2	Tofacitinib (5–11 mg BID)	2	0	0
Le Voyer <i>et al.</i> (2021)(44)	Retrospective	10	Ruxolitinib (7.5–20 mg BID), Baricitinib (2–4 mg BID)	10	9	0
Aeschlimann <i>et al.</i> (2018) (45)†	Case study	1	Ruxolitinib (10 mg BID)	1	1	0
Wang <i>et al.</i> (2021) (46)	Retrospective	7	Ruxolitinib (NR), Tofacitinib (NR)	7	5	2
Yu <i>et al.</i> (2021) (47)	Open label	3	Tofacitinib (5 mg BID)	3	3	2
Zhou <i>et al.</i> (2021) (48)	Case report	1	Tofacitinib (5 mg QD)	1	1	0

\*Patients were included in the analysis in Kim *et al.* (2021a) and were not included in the count of unique patients (39).

†Patient likely to have been reported in Le Voyer *et al.* (2021) and were not included in the count of unique patients (44).

BID: twice daily; ILD: interstitial lung disease; JAK: Janus kinase; JDM: juvenile dermatomyositis; NR: not reported; QD: once daily.

**Supplementary Table S3.** Prior therapies in patients refractory to SOC therapy among publications reporting therapy for individual patients.

Individual therapy, n (%) <sup>*†</sup>	DM without ILD N=43	DM-ILD N=15	All DM N=58	JDM without ILD N=25	JDM-ILD N=4	All JDM N=29
Corticosteroids	42 (98)	15 (100)	<b>57 (98)</b>	25 (100)	4 (100)	<b>29 (100)</b>
Methotrexate	39 (91)	2 (13)	<b>41 (71)</b>	21 (84)	3 (75)	<b>24 (83)</b>
IVIG	34 (79)	5 (33)	<b>39 (67)</b>	23 (92)	4 (100)	<b>27 (93)</b>
Mycophenolate mofetil	29 (67)	2 (13)	<b>31 (53)</b>	15 (60)	4 (100)	<b>19 (66)</b>
Hydroxychloroquine	23 (53)	1 (7)	<b>24 (41)</b>	8 (32)	0	<b>8 (28)</b>
Azathioprine	21 (49)	2 (13)	<b>23 (40)</b>	2 (8)	0	<b>2 (7)</b>
Tacrolimus	8 (19)	7 (47)	<b>15 (26)</b>	4 (16)	3 (75)	<b>7 (24)</b>
Cyclophosphamide	2 (5)	13 (87)	<b>15 (26)</b>	4 (16)	1 (25)	<b>5 (17)</b>
Rituximab	11 (26)	2 (13)	<b>13 (22)</b>	14 (56)	2 (50)	<b>16 (55)</b>
Cyclosporine	4 (9)	9 (60)	<b>13 (22)</b>	6 (24)	1 (25)	<b>7 (24)</b>
Plasma exchange	1 (2)	4 (27)	<b>5 (9)</b>	6 (24)	0	<b>6 (21)</b>
Quinacrine	4 (9)	0	<b>4 (7)</b>	0	0	<b>0</b>
Infliximab	2 (5)	0	<b>2 (3)</b>	3 (12)	0	<b>3 (10)</b>
Amlodipine	1 (2)	1 (7)	<b>2 (3)</b>	0	0	<b>0</b>
Abatacept	1 (2)	0	<b>1 (2)</b>	0	2 (50)	<b>2 (7)</b>
Pamidronate	1 (2)	0	<b>1 (2)</b>	2 (8)	0	<b>2 (7)</b>
Adalimumab	1 (2)	0	<b>1 (2)</b>	1 (4)	0	<b>1 (3)</b>
Etanercept	1 (2)	0	<b>1 (2)</b>	1 (4)	0	<b>1 (3)</b>
Lenalidomide	1 (2)	0	<b>1 (2)</b>	0	0	<b>0</b>
Thalidomide	1 (2)	0	<b>1 (2)</b>	0	0	<b>0</b>
Colchicine	1 (2)	0	<b>1 (2)</b>	0	0	<b>0</b>
Ibandronate	0	1 (7)	<b>1 (2)</b>	0	0	<b>0</b>
Etidronate	1 (2)	0	<b>1 (2)</b>	0	0	<b>0</b>
Sirolimus	0	0	<b>0</b>	1 (4)	0	<b>1 (3)</b>

\*Not mutually exclusive.

†Landon-Cardinal *et al.* (2019) (7), Ding *et al.* (2021) (37) and Wang *et al.* (2021) (46) reported prior therapies for their overall analysis rather than for individual patients and are thus not included in this table.

DM: dermatomyositis; ILD: interstitial lung disease; IVIG: intravenous immunoglobulin; JDM: juvenile dermatomyositis; SOC: standard of care.

**Supplementary Table S4.** Summary of the effect of JAK inhibitor therapy in DM.

Organ involvement	DM		JDM	
	N	Improved* (%)	N	Improved* (%)
Skin	61	100	60	95
Muscle	16	100	36	83
Lung	31	94	10 (4) <sup>‡</sup>	100

\*Improvement is based on measured improvement in skin disease activity (*e.g.* CDASI, Skin DAS), muscle strength (*e.g.* CMAS), lung capacity and performance (*e.g.* FVC% or DLCO%), or investigator/physician decision.

‡Four patients out of 10 had reported outcomes; all four improved.

CDASI: Cutaneous Dermatomyositis Disease Area and Severity Index; CMAS: Childhood Myositis Activity Score; DAS: disease activity score; DLCO%: diffusing capacity for carbon monoxide; DM: dermatomyositis; FVC%: forced vital capacity; JAK: Janus kinase; JDM: juvenile dermatomyositis.

**Supplementary Table S5.** Oral JAK inhibitors approved or in late-stage development for rheumatologic autoimmune diseases.

JAK inhibitor	JAK isoform specificity	Latest* US development phase	Indication
XELJANZ (tofacitinib)	JAK1/JAK2/JAK3	Approved	RA, JIA, PsA, UC, AS
OLUMIANT (baricitinib)	JAK1/JAK2	Approved Filed Phase 3	RA AD, AA SLE, JIA, JIA-associated uveitis
RINVOQ (upadacitinib)	JAK1	Approved Filed Phase 3 Phase 2	RA, PsA, AD UC, AS, axSpA CD, GCA Vitiligo, SLE, HS
CIBINQO (abrocitinib)	JAK1	Approved	AD
Deucravacitinib	TYK2	Filed Phase 3 Phase 2	PsO PsA SLE, CLE, CD, UC
Deuruxolitinib	JAK1/JAK2	Phase 3	AA
Filgotinib	JAK1	Phase 3	CD, UC
Ritlecitinib	JAK3/TEC	Phase 3 Phase 2	AA RA, vitiligo, CD, UC
SHR0302	JAK1	Phase 3 Phase 2	AD, UC AA, CD
Brepocitinib	JAK1/TYK2	Phase 2	SLE, CD, HS
Gusacitinib	Pan-JAK/Syk	Phase 2	CHE
INCB54707	JAK1	Phase 2	Vitiligo, HS
NDI-034858	TYK2	Phase 2	PsO, PsA
OST-122	JAK3/TYK2/ARK5	Phase 2	UC
Ropsacitinib	TYK2	Phase 2	HS
TLL018	JAK1/TYK2	Phase 2	UC

\*As of January 21, 2022.

AA: alopecia areata; AD: atopic dermatitis; ARK5: AMPK-related protein kinase 5; AS: ankylosing spondylitis; axSpA: axial spondylarthritis; CD: Crohn's disease; CHE: chronic hand eczema; CLE: cutaneous lupus erythematosus; GCA: giant cell arteritis; HS: hidradenitis suppurativa; JAK: Janus kinase; JIA: juvenile idiopathic arthritis; PsA: psoriatic arthritis; PsO: psoriasis; RA: rheumatoid arthritis; SLE: systemic lupus erythematosus; Syk: spleen tyrosine kinase; TEC: tyrosine kinase expressed in hepatocellular carcinoma; TYK2: tyrosine kinase 2; UC: ulcerative colitis.

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