Table 2. Characteristics of included studies reporting on targeted therapies for IIM.

Abbreviations: A.: analysis (corresponds to number below in analysis), AZA: azathioprine, CDASI: Cutaneous Dermatomyositis Disease Area and Severity Index, DM: dermatomyositis, IMACS: International Myositis Assessment and Clinical Studies Group, i.v.: intravenously, HAQ: Health Assessment Questionnaire, MDAAT: Myositis Disease Activity Assessment Tool, MMT: manual muscle test, MTX: methotrexate, PM: polymyositis, PROMIS-29: Patient-Reported Outcomes Measurement Information System-29,RCT: randomized controlled trial, s.c.: subcutaneously, S1P: sphingosine-1-phosphate, TIS: Total improvement score, 3TUG: triple Timed Up and Go test, 6MWD: 6-minute walking distance. TNF = TLR = AZA =

A.	Medication	Study	Study design	Population (n)	Intervention	Control	Follow-up	Outcomes
5	**Rituximab**	Oddis 2013 (RIM study)	RCT	Adults and children >5 years with definite or probable refractory DM or PM (n = 200)	Rituximab infusions in week 0 and 1, dosing based on patient's body surface area (750mg-1g/m^2 per infusion). Placebo infusions at week 8 and 9	Placebo infusions at week 0 and 1 (not further specified). Rituximab at week 8 and 9	44 weeks (*results 8 weeks used for analysis)*	Improvement (IMACS) Skin symptoms (MDAAT)
6	**Abatacept**	NCT-683	RCT	Adults with DM or PM (n = 148)	Abatacept 125 mg weekly s.c. for 24 weeks In combination with standard treatment	Placebo to match abatacept s.c. In combination with standard treatment	24 weeks	Function (HAQ) Muscle strength (MMT8) Improvement (IMACS) Serious adverse events
		Tjärnlund 2018 (ARTEMIS)	RCT	Adults with refractory DM or PM (n = 20)	Abatacept 500-1000mg i.v. (depending on body weight), for 7 infusions over 6 months (week 0, 2, 4, 8, 12, 16, 20)	Abatacept 500-1000mg i.v.; initiation after 12 weeks. 7 infusions (week 12, 14, 16, 20, 24, 28, 32)	6 months *(results 3 months used for analysis)*	Function (HAQ) Muscle strength (MMT8) Improvement (IMACS) Serious adverse events
7	Zilucoplan (**Complement 5 inhibitor)**	(NCT-632)	RCT	Adults with immune-mediated necrotizing myopathy (n = 27)	Zilucoplan 0.3 mg/kg/day s.c. for 8 weeks	Matching placebo doses s.c. for 8 weeks	8 weeks	Function (HAQ, 3TUG) Muscle strength Improvement (TIS)
8A	**Anti-TNF-alpha inhibitors**	Muscle study group 2011	RCT	Adults with DM, newly diagnosed or refractory (n = 16)	Etanercept 50mg s.c. weekly	Placebo prefilled liquid syringes consisting of 25mM Na phosphate, 25mM L-arginine-HCl, 100mM. NaCl, 1% sucrose per syringe	24 and 52 weeks	Function (HAQ) Muscle strength (MMT) Improvement (IMACS) Skin symptoms (CDASI) Serious adverse events
		Schiffenbauer 2018	RCT	Adults with DM and PM, using corticosteroids with MTX or AZA (n = 13)	Infliximab 4 infusions of 5mg/kg at week 0, 2, 6, and 14	Placebo 4 infusions at week 0, 2, 6 and 14	16 weeks	Function (HAQ) Muscle strength (MMT8) Improvement (IMACS) Serious adverse events
8B	**Gevokizumab (IL-1-β inhibitor)**	EUCT-34	RCT	Adults with PM, DM, or necrotizing autoimmune myopathy (n = 27)	Gevokizumab 60 mg s.c. every 4 weeks, for 24 weeks	Placebo s.c. every 4 weeks, for 24 weeks	24 weeks	Muscle strength (MMT8) Serious adverse events

8C	**Bazlitoran (TLR 7/8/9 inibitor)**	NCT-857	Three-arm RCT	Adults with DM (n = 30)	1. Bazlitoran 0.6 mg/kg s.c. injections once weekly for 24 weeks 2. Bazlitoran 1.8 mg/kg s.c. injections once weekly for 24 weeks Concomitant prednisone or DMARD was allowed	Placebo in the form of saline injections s.c. for 24 weeks Concomitant prednisone or DMARD was allowed	28 weeks	Muscle strength (MMT8) Skin symptoms (modified CDASI) Serious adverse events
8D	**Lenabasum (cannabinoid agonist)**	EUCT-10 (DETERMINE)	RCT	Adults with DM (n = 175)	1. Lenabasum 20 mg orally, twice daily as hard capsule, for 52 weeks 2. Lenabasum 5 mg orally, twice daily as hard capsule, for 52 weeks	Placebo s a powder-in-capsule containing microcrystalline cellulose and magnesium stearate	28 and 52 weeks	Improvement (IMACS) Skin symptoms (CDASI) Serious adverse events
		NCT-243	RCT	Adults with DM (classic or amyopathic) (n = 22)	Lenabasum 20 mg once daily on days 1-28, then twice daily on days 29-84	Placebo once daily on days 1-28, then twice daily on days 29-84	16 weeks	Function (PROMIS-29) Skin symptoms (CDASI) Serious adverse events
8E	**Sifalimumab (anti-IFN-α)**	NCT-091	RCT	Adults with DM or PM (n = 51)	Sifalimumab for 6 months dosed at 0.3 mg/kg, 1 mg/kg, 3 mg/kg or 10 mg/kg (4 treatment arms), dosing every other week (14 doses total)	Placebo for 3 months, then switched to sifalimumab for 3 months	52 weeks	Adverse events (reported after 14 weeks)
8F	**Siponimod (S1P receptor modulator)**	NCT-274	RCT	Adults with DM (n = 17)	1. 0.5 mg siponimod daily 2. 2 mg siponimod daily 3. 10 mg siponimod daily For 24 weeks	Matching placebo for 24 weeks	48 weeks	Function (6MWD) Muscle strength (MMT24) Serious adverse events
		NCT-810	RCT	Adults with DM or PM (n = 18)	Siponimod 10mg once daily (2 tablets of 5 mg) for 12 weeks With 10-day dose up-titration	Placebo 2 tablets once daily	12 weeks	Improvement (IMACS) Serious adverse events
		NCT-917	RCT	Adults with PM (n = 14)	1. 2 mg siponimod daily (1 tablet + 4 tablets placebo) 2. 10 mg siponimod daily (5 tablets)	Placebo (5 tablets)	12 weeks	Function (6MWD) Muscle strength (MMT24) Serious adverse events
8G	**Tocilizumab (IL-6 inhibitor)**	Oddis 2022 (TIM)	RCT	Adults with refractory DM or PM (n = 36)	Tocilizumab 8mg/kg i.v. every 4 weeks for 24 weeks	Placebo i.v. infusions every 4 weeks for 24 weeks	24 weeks	Function (HAQ) Muscle strength (MMT8) Improvement (TIS) Serious adverse events