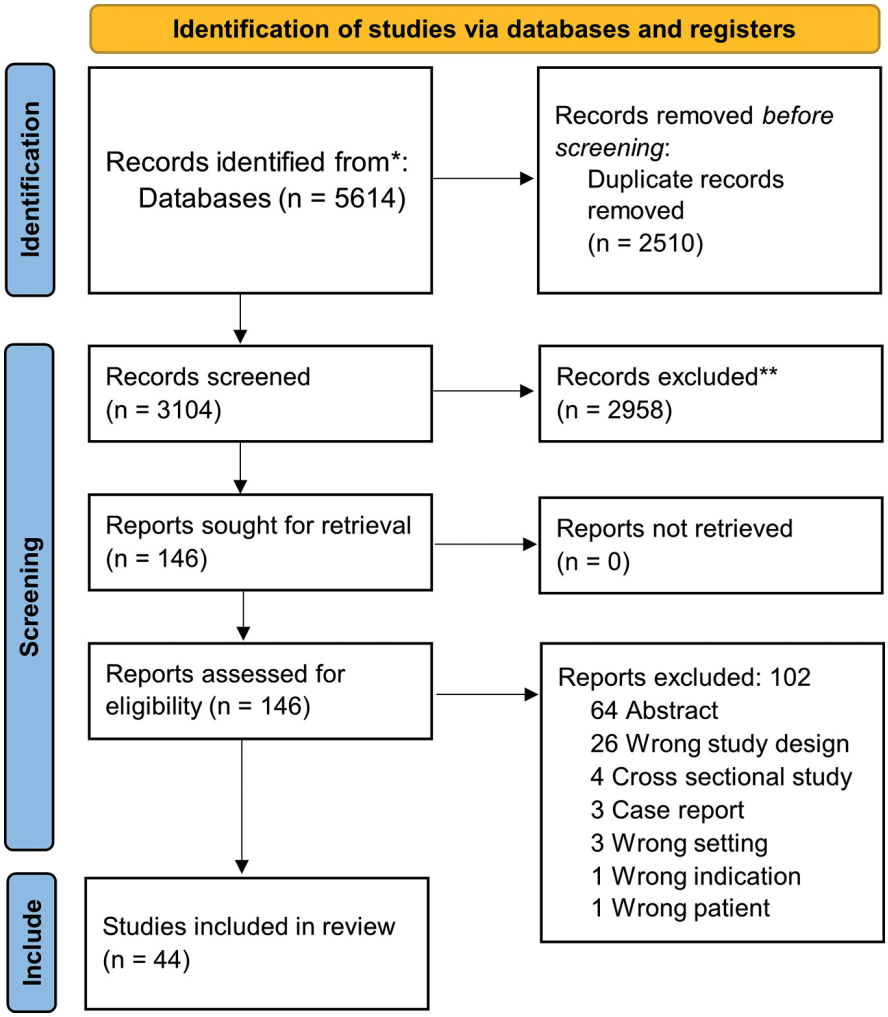

Supplementary Fig. S1. PRISMA 2020 flow diagram for new systematic reviews which included searches of databases and registers only.
*Consider, if feasible to do so, reporting the number of records identified from each database or register searched (rather than the total number across all databases/registers).
**If automation tools were used, indicate how many records were excluded by a human and how many were excluded by automation tools.
From: PAGE MJ, MCKENZIE JE, BOSSUYT PM *et al*.: The PRISMA 2020 statement: an updated guideline for reporting systematic reviews. *BMJ* 2021; 372: n71. https://doi.org/10.1136/bmj.n71
For more information, visit: http://www.prisma-statement.org/

Appendix 1. Search strategy Ovid MEDLINE(R) ALL.

1	exp Myositis, Inclusion Body/
2	inclusion body myositis.tw,kw.
3	1 or 2
4	limit 3 to English language

Supplementary Table S1. Open labelled trials in IBM.

Study	Type of the study	No of patients	Inclusion criteria	Dose/Mode of intervention	Duration	Outcome measures
Bimagrumab (BYM338) 2020 (46) (NCT01925209)	Open label multicenter extension study	10	- Definite sIBM following ENMC diagnostic criteria (1997) -40-80 years	bimagrumab 10 mg/Kg IV infusion every 4 weeks	Up to 104 weeks (early termination considering the results of RESILIENT trial)	- TMV, IMAT and SCAT assessed by muscle MRI - LBM assessed by DXA - Handgrip - Quadriceps muscle strength assessed by dynamometry - 6MWD
Canakinumab 2019 (41)	Proof of principle, open label study	5	Not clearly specified.	150 mg subcutaneous canakinumab every 8 weeks	5–33 months	- Bilateral grip force with dynamometry - Total muscle strength sum score of 6 muscle groups
Follistatin Gene Therapy 2017 (43)	Open label	6	Griggs criteria	Follistatin gene therapy (rAAV1. CMV.huFS344)	Median 1 year	- 6MWT - Muscle biopsy
Anakinra 2013 (40) EudraCT, number: 2010-019030-27	Pilot study, open label	4	Biopsy proven sIBM (Griggs criteria)	100 mg subcutaneous anakinra daily	5 to 12 months	- Bilateral grip force with dynamometry MMT with MRC scale in 6 muscle groups
IVIg long-term follow up 2012 (37)	Open label	16	Griggs criteria	IVIg (2 gm/Kg over 3-5 days)		- Muscle strength testing (Kendall) - Dynamic studies of the esophagus by barium x-ray - CK level - Sedimentation rate - Self reported dysphagia and muscle strength
Simvastatin 2011 (45)	Pilot study, open label	14	Griggs criteria; disease duration <10 years; steroids at stable dosage allowed; not allowed immunosuppressants	40 mg oral simvastatin daily	12 months	-IMACS disease activity core set in adult and juvenile idiopathic inflammatory myopathies (2011), including MMT, MDI, SF-36 - IBM-FRS
Stance control orthosis 2010 (35)	Pilot study, open label	9		Stance control orthosis use	6 months	- Gait related measures, including velocity, cadence, step width
Alemtuzumab 2009 (36) (NCT00079768)	Proof of principle, open label study	13	Diagnostic criteria not specified; no immuno-modulating or immuno-suppressive therapies for at least a year	0.3 mg/kg/day alemtuzumab for 4 days	12 months	- QMT, total summed score of strength in kilograms - MMT using the modified MRC scale (0–10) over 30 muscle groups bilaterally in arms and legs
Etanercept 2006 (34)	Pilot trial, open label	9	Griggs criteria	etanercept: 25 mg subcutaneously twice weekly	6 and 12-months evaluations (treatment duration 17±6.1 months)	QMT by measuring MVIC
Anti-T-lymphocyte globulin 2003 (42)	Randomized between Methotrexate and ATG	11	Histopathological findings	Initial dose of ATG 5 mg/kg 1st day, 4 mg/kg 2nd day, thereafter to keep the T-lymphocyte count between 50-150 x 10^6/L.	12 months	- Muscle strength by myometry - Muscle biopsy CK, T-lymphocyte subsets, T-cell - functions
IVIG 1994 (32)	Open label	9	Clinical and histologic features of sIBM	2 g/kg IVIG monthly	Not fully specified. 3-6 months.	- MMT using the modified MRC scale (0-10) over 36 muscle groups: results expressed as AMS - Disability score (from 0 as no impairment to 5 as confined to wheelchair)
Prednisone and Azathioprine/Methotrexate (48)	Open label	14	Not specified	Prednisone, Methotrexate, or Azathioprine, or in combination	Not clearly specified	- Muscle strength testing - creatine kinase [CK], lactate dehydrogenase, aldolase, and/or transaminases
Home exercise program (33)	Open label	7	Griggs criteria	Home based exercise program	12 weeks	- MMT - Maximal voluntary knee extension and flexion - serum CK - muscle biopsy

Study	Type of the study	No of patients	Inclusion criteria	Dose/Mode of intervention	Duration	Outcome measures
Home exercise program (38)	Open label	7	Clinical and histologic features of sIBM (Lotz, Brain 1994); immunotherapies at stable dosages allowed	Home based exercise program	16 weeks	- MMT - maximal voluntary knee extension - functional index in myositis - muscle biopsy histopathology
Exercise Program (39)	Open label	7	Clinical and histological findings	Aerobic, resistance, and stretching exercises	12 weeks	- Aerobic capacity - Muscle strength by dynamometry - CK, Lactate level - Body mass - Self reported RPE
Expiratory muscle strength training (44)	Open label	12	ENMC criteria with abnormal upper esophageal sphincter function and dysphagia	Expiratory Muscle Strength	12 weeks	- Abridged Dysphagia Handicap Index (DHI) - SF-36 - Video fluoroscopic swallow study (VFSS) - Flexible Endoscopic Evaluation of Swallowing (FEES)
Strength training (47)	Open label	7	Clinical and histological characteristics	Exercise training program	12 weeks	- Fatigue severity scale - Barthel index - Isometric, dynamic, and manual muscle strength testing - Muscle size based on MRI - Serum CK and lymphocyte subpopulation in muscle biopsy

Supplementary Table S2. Observational and longitudinal studies in IBM.

Study	No of patients	Diagnostic criteria	Duration	Outcome measures
Sangha *et al.* 2021 (31)	181	Griggs, Hilton-Jones, or ENMC criteria	0-7.3 years	- MMT - MVICT - IBMFRS - Grip strength
Oldroyd *et al.* 2020 (16)	75	MRC 2010 criteria	4.3 years (2.6-6.2)	- Muscle strength (dynamometry) - IBM-FRS - NSS (Neuromuscular Symptom Score) - Grip strength - Pinch strength
Alfano *et al.* 2017 (23)	55	MRC clinical criteria (Patients who can walk 6 minutes)	20 months (median) (1-28 months)	- Strength testing - Stair climbing (time to ascend or descend 4 stairs) - Stepping up on a curb - Getting out of a chair - Timed up-and-go - 6MWT
Morrow *et al.* 2016 (29)	20	MRC criteria	12 months	- Muscle strength - SF-36 quality of life - IBM-FRS - Lower extremity myometry - Muscle MRI findings 3-point-Dixon fat fraction quantification Non-fat suppressed T2 measurement MTR imaging
Hogrel *et al.* 2014 (27)	13	All patients had muscle biopsy, but criteria not clearly reported	4 years	- Strength by MMT - Muscle dynamometry - Grip strength - 6MWT - IBM-FRS - Walton, Karnovsky, Rivermean Mobility Index (RMI) - sIBM weakness composite index
Cortese *et al.* 2013(25)	51 patients	Griggs criteria MRC criteria	1 year	- Strength by MMT - IBM-FRS - Quadriceps quantitative muscle strength testing by dynamometry - IBM-FRS

Study	No of patients	Diagnostic criteria	Duration	Outcome measures
Allenbach *et al*. 2012 (24)	16	No clearly reported criteria (diagnosis based on muscle biopsy)	9 months	- Strength by MMT - Walton, Rivermead mobility index - sIBM weakness composite index - IBM-FRS - Dynamometry - 6MWT
Lindberg *et al*. 2012 (28)	66 patients	No clearly reported criteria	61.1 months (11-211 months)	- Muscle strength - Serum CK
Cox *et al*. 2011 (6)	64	ENMC criteria	2 years (median)	- Strength by MMT - Dynamometry - Physical disability score - Rivermead mobility index - Brooke's functional grading scale - Dysphagia standardized questionnaire (Wintzen)
Benveniste *et al*. 2011(5)	136	Griggs criteria Hilton-Jones criteria	Not specified	- Manual muscle strength testing - Walton scale, Rivermead Mobility Index, - Grip strength - IBM weakness composite index
Felice *et al*. 2001 (26)	8	Biopsy proven, Griggs Criteria	Mean 4.1 years, (1-8 years)	- Manual muscle testing - Grip strength
Rose *et al*. 2000(19)	11	Griggs IBM criteria definite	6 months	- MVIC fixed myometry (QMT system) - Six muscle groups bilaterally (shoulder abduction, elbow flexion/extension, knee flexion/extension, ankle dorsiflexion. - Muscle mass based on urine creatinine excretion - Lean body mass using DEXA - Brooke's Grading System - Rivermead Mobility Index - CK level - Patient's own assessment
Peng *et al*. 2000 (30)	78 (6 were clinically examined) Questionnaire based study		Mean 24 months (12-52 months)	- Whole body MMT - Upper and lower extremity MVICT (significant upper extremity MVICT) - Upper extremity MMT - Lower extremity MMT - Get up and go - 6MW - Stair climb (improvement of 1 step) - BMI - Lean Body Mass (increased) - LFT (increased but small) - Lipid profile

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