Appendix SI. Definitions and rating of measurement properties

Domain	Measurement property	Definition	Measurement property rating
Reliability	Reliability	The extent to which scores of an aerobic capacity measure are the same in patients who have not changed for repeated measurement under several conditions: e.g. comparing different scores of aerobic capacity over time (test-retest); by different persons on the same occasion (interrater); or by the same persons (i.e. raters or responders) on different occasions (intra-rater).	+ ICC or weighted kappa ≥ 0.70 – ICC or weighted kappa < 0.70
	Measurement error	The systematic and random error of a patient's aerobic capacity score that is not attributed to true changes in aerobic capacity.	+ SDC or LoA < MIC – SDC or LoA > MIC
Validity	Content validity	The degree to which the content of a measurement property is an adequate reflection of the construct to be measured. For a maximal exercise test, content validity is assessed by the degree to which the results of the test protocol reflect true VO_{2peak}. Here we determined which percentage of patients reached pre-set criteria for achieving maximal aerobic exercise.	+ ≥ 75% of patients meet VO_{2peak} criteria – ≤ 75% of patients meet VO_{2peak} criteria
	Criterion validity	The degree to which the aerobic capacity score is an adequate reflection of a "gold standard". E.g. for VO_{2peak}, the gold standard is GXT with respiratory gas exchange measurements on a treadmill or bicycle/arm ergometer.	+ Correlation with gold standard ≥ 0.70 – Correlation with gold standard < 0.70
	Construct validity	The degree to which the scores of an aerobic capacity measure are consistent with hypotheses (e.g. the scores of an aerobic capacity measure discriminate between physically active and physically inactive patients).	+ The result is in accordance with the hypothesis – The result is not in accordance with the hypothesis
Responsiveness	Responsiveness	The ability of a measure of aerobic capacity to detect changes in aerobic capacity over time.	+ ≥ 75% of the results are in accordance with the hypotheses OR AUC ≥ 0.70 – ≤ 75% of the results are in accordance with the hypotheses OR AUC ≥ 0.70

+: sufficient rating; −: insufficient rating; ICC: intraclass correlation coefficient; SDC: smallest detectable change; LoA: limits of agreement; MIC: minimal important change; VO_{2peak}: peak oxygen consumption; GXT: maximal effort graded exercise testing; AUC: area under the curve.

Appendix SII. Literature search

#	Search	Results
	Ovid MEDLINE(R) ALL < 1946 to June 29, 2021 > Search date: 30 June 2021	
1	exp exercise test/	67045
2	(aerobic capacity or maximal exercise test? or submaximal exercise test? or (oxygen adj2 uptake) or (oxygen adj2 consumption) or vo2max or "vo2 max" or "vo(2)max" or "vo(2max)" or vo2peak or "vo2 peak" or "Vo(2)peak" or "vo(2peak)" or ((peak or max*) adj1 (oxygen consumption or oxygen uptake or o2 consumption or o2 uptake)) or "po(peak)" or vo2 kinetics or vent).ab,kf,ti.	81070
3	(yo-yo or Shuttle or "20-MST" or "20-meter" or twenty meter or "20 m" or Chester or "6 minute?" or six minute? or "6 min" or Astrand or physical working capacity or PWC* or Canadian home fitness or Conconi or Bruce or Balke or Cooper).ab,kf,ti.	55224
4	(Ventilatory threshold or lactate threshold or anaerobic threshold).ab,kf,ti.	6041
5	or/1–4 [aerobic capacity]	181914
6	Neuromuscular Diseases/ or Motor Neuron Disease/ or Muscular Atrophy, Spinal/ or Bulbo-Spinal Atrophy, X-Linked/ or Bulbar Palsy, Progressive/ or Muscular Diseases/ or Mitochondrial Myopathies/ or Muscular Disorders, Atrophic/ or Postpoliomyelitis Syndrome/ or Muscular dystrophies/ or Myotonic dystrophy/ or Muscular Dystrophies, Limb-Girdle/ or Muscular Dystrophy, Oculopharyngeal/ or Muscular Dystrophy, Facioscapulohumeral/ or Muscular Dystrophy, Duchenne/ or Muscular Dystrophy, Emery-Dreifuss/ or Distal Myopathies/ or Myopathies, Structural, Congenital/ or yopathies, Nemaline/ or Myopathy, Central Core/ or Myositis/ or Myositis, Inclusion Body/ or Polymyositis/ or Dermatomyositis/ or Myotonic Disorders/ or myotonia congenita/ or Neuromuscular Junction Diseases/ or Myasthenia Gravis/ or Lambert-Eaton Myasthenic Syndrome/ or Myasthenic Syndromes, Congenital/ or Peripheral Nervous System Diseases/ or Giant Axonal Neuropathy/ or Guillain-Barre Syndrome/ or Isaacs Syndrome/ or Mononeuropathies/ or Polyneuropathies/ or Polyradiculoneuropathy/ or Polyradiculoneuropathy, Chronic Inflammatory Demyelinating/ or "Hereditary Sensory and Motor Neuropathy"/ or Charcot-Marie-Tooth/ or Hereditary spastic paraplegia/ or Poliomyelitis/ or Poliomyelitis, Bulbar/ or Postpoliomyelitis Syndrome/ or Glycogen Storage Disease/ or Glycogen Storage Disease Type II/ or Glycogen Storage Disease Type V/ or multifocal motor neuropathy/ or chronic idiopathic axonal polyneuropathy/ or amyotrophic lateral sclerosis/	185281
7	(Amyotrophic Lateral Sclerosis or Amyotrophic sclerosis or Gehri* Disease or Lou-Gehri* Disease or als or "Hereditary Sensory and Motor Neuropath*" or "Hereditary Motor and Sensory Neuropath*" or Hereditary sensory-motor neuropathy or Hereditary sensorimotor neuropathy or Charcot-Marie-Tooth or Charcot Marie Tooth or Charcot-Marie Tooth or Peroneal musc* atrophy* or Charcot Marie Disease or Charcot-Marie Disease or Roussy-Levy or Roussy Levy or Post Poli* or Post-Poli* or Postpoli* or Polio late effects or Spinal Amyotroph* or Spinal muscular atroph* or Spinal muscle degeneration or Bulbospinal muscular atrophy or Spinobulbar muscular atrophy or Kenned* Disease or Mitochondrial Myopath* or Multiple Myositis or Neuromyositis or Polymyositi* or Dermatomyositis or Dermatopolymyositis or Polymyositis-Dermatomyositis or Dermatomucosomyositis or Wagner-Unverricht or Unverricht-Wagner or Glycogen storage disease Type 5 or Glycogen storage disease Type V or Glycogen storage disease V or Glycogen storage disease 5 or Glycogenosis 5 or Glycogenosis V or Glycogenosis type 5 or Glycogenosis type V or GSD 5 or GSD V or GSD type 5 or GSD type V or Mcardl* or Sporadic inclusion body myositi* or Inclusion Body Myositi* or Glycogen storage disease Type 2 or Glycogen storage disease Type II or Glycogen storage disease II or Glycogen storage disease 2 or Glycogenosis 2 or Glycogenosis II or Glycogenosis type 2 or Glycogenosis type II or GSD 2 or GSD II or GSD type 2 or GSD type II or Pomp* disease or Central core diseas* or Central Core Myopath* or Muscle core disease or Muscular central core disease or Shy Magee Syndrome or Shy-Magee Syndrome or Nemaline body disease or Nemaline Myopath* or Nemaline rod disease or Nemaline rod myopathy or Rod body disease or Rod Body Myopathy or Rod Myopath* or Duchenn* muscular dystrophy or Duchenn* Type Progressive Muscular Dystrophy or Duchenne or Pseudohypertrophic Childhood Muscular Dystrophy or Pseudohypertrophic Muscular Dystrophy or Becke* disease or Becke* muscular dystroph* or Becker dystrophinopathy or Facio-Scapulo-Humeral Dystrophy or Facio Scapulo Humeral Dystrophy or Facioscapulohumeral Atroph* or Facioscapulohumeral dystroph* or Facioscapulohumeral Muscular Dystroph* or Facioscapulohumeral myopathy or Landouzy Dejerine Dystroph* or Landouzy Dejerine muscular dystroph* or Landouzy-Dejerine Dystroph* or Landouzy-Dejerine muscular dystroph* or Limb Girdle or Limb-Girdle or Muscular Dystroph* or Myodystroph* or Dystrop* myotonica or Myotonia Atroph* or Myotonia dystroph*OR Myotonic muscular dystroph* or Steiner* disease or Steiner* Myotonic Dystroph* or Steiner* syndrome or Chronic idiopathic axonal polyneuropath* or Oculopharyngeal dystroph* Oculopharyngeal Muscular Dystroph* or Familial spastic parap* or Hereditary spastic parap* or Hypertrophic Motor Sensory Neuropath* or Hypertrophic Motor-Sensory Neuropath* or Spastic Parap* or Spastic congenital parap* or Spastic Parap* or Hypertrophic Motor Sensory Neuropath* or Strumpel* dis*OR Strumpel*-Lorrain dis* or Degenerative motor system dis* or Motor neuro* dis* or MND or Motor System Dis* or Primary lateral sclerosis or Lateral Sclerosis or Adult-onset PLS or Adult-onset primary lateral sclerosis or Multifocal motor neuropath* or Motor neuropath* with multiple conduction block or Congenital myopath* or Myopath* congenital or Distal muscular dystroph* or Distal myopath* or Emery-Dreifuss or Emery Dreifuss or Eaton Lambert or Eaton-Lambert or Lambert Eaton or Lambert-Eaton or Myasthenic syndrome).ab,kf,ti.	115665
8	6 or 7 [NMD]	236357
9	5 and 8	1525
10	("IMproving FItness in NEuromuscular diseases" or "IM FINE" or "B-FIT").ab,kf,ti.	47
11	(NCT04322357 or NCT04187482 or NCT04173234 or NCT03324152 or NCT03293615 or NCT02158156 or NCT02121678 or NCT02020187 or NCT02003937 or NCT01650818 or NCT01501019 or NCT01116570 or NCT01047761 or NCT03326622 or NCT01895283 or ISRCTN99826269 or ISRCTN67078621 or NL7344 or NL7899 or NL7374 or ACTRN12617001244392).ab,ti.	2
12	or/9-11	1571
	Ovid Embase Classic+Embase < 1947 to 2021 June 29 > Search date: 30 June 2021	
1	aerobic capacity/ or exercise test/ or exp walk test/	88628
2	(aerobic capacity or maximal exercise test? or submaximal exercise test? or (oxygen adj2 uptake) or (oxygen adj2 consumption) or vo2max or "vo2 max" or "vo(2)max" or "vo(2max)" or vo2peak or "vo2 peak" or "Vo(2)peak" or "vo(2peak)" or ((peak or max*) adj1 (oxygen consumption or oxygen uptake or o2 consumption or o2 uptake)) or "po(peak)" or vo2 kinetics or vent).ab,kw,ti.	111172
3	(yo-yo or Shuttle or "20-MST" or "20-meter" or twenty meter or "20 m" or Chester or "6 minute?" or six minute? or "6 min" or Astrand or physical working capacity or PWC* or Canadian home fitness or Conconi or Bruce or Balke or Cooper).ab,kw,ti.	82883
4	(Ventilatory threshold or lactate threshold or anaerobic threshold).ab,kw,ti.	8064
5	or/1-4 [aerobic capacity]	245749
6	(Neuromuscular Disease? or Motor Neuron Disease or Spinal Muscular Atrophy or x-linked Bulbo-Spinal Atrophy or progressive Bulbar Palsy or Muscular Diseases or Mitochondrial Myopathies or atrophic Muscular Disorder? or Muscular dystroph* or Myotonic dystroph* or limb girdle Muscular Dystrophies or Oculopharyngeal Muscular Dystrophy or Facioscapulohumeral Muscular Dystrophy or Duchenne or Emery-Dreifuss or Distal Myopathies or congentinal Myopath* or nemaline yopathies or central core Myopathy or Myositis or Myositis or Polymyositis or Dermatomyositis or Myotonic Disorders or myotonia congenita or Neuromuscular Junction Diseases or Myasthenia Gravis or Lambert-Eaton Myasthenic Syndrome or Myasthenic Syndromes or Peripheral Nervous System Diseases or Giant Axonal Neuropathy or Guillain-Barre Syndrome or Isaacs Syndrome or Mononeuropathies or Polyneuropathies or Polyradiculoneuropathy or Polyradiculoneuropathy or "Hereditary Sensory and Motor Neuropathy" or Charcot-Marie-Tooth or Hereditary spastic paraplegia or Poliomyelitis or Postpoliomyelitis Syndrome or Glycogen Storage Disease or multifocal motor neuropathy or chronic idiopathic axonal polyneuropathy or amyotrophic lateral sclerosis).hw,kw.	268827

(Contined)

Appendix SII. (*Continued*) Literature search

#	Search	Results
	Ovid Embase Classic+Embase < 1947 to 2021 June 29 > Search date: 30 June 2021	
7	(Amyotrophic Lateral Sclerosis or Amyotrophic sclerosis or Gehri* Disease or Lou-Gehri* Disease or als or "Hereditary Sensory and Motor Neuropath*" or "Hereditary Motor and Sensory Neuropath*" or Hereditary sensory-motor neuropathy or Hereditary sensorimotor neuropathy or Charcot-Marie-Tooth or Charcot Marie Tooth or Charcot-Marie Tooth or Peroneal musc* atrophy* or Charcot Marie Disease or Charcot-Marie Disease or Roussy-Levy or Roussy Levy or Post Poli* or Post-Poli* or Postpoli* or Polio late effects or Spinal Amyotroph* or Spinal muscular atroph* or Spinal muscle degeneration or Bulbospinal muscular atrophy or Spinobulbar muscular atrophy or Kenned* Disease or Mitochondrial Myopath* or Multiple Myositis or Neuromyositis or Polymyositi* or Dermatomyositis or Dermatopolymyositis or Polymyositis-Dermatomyositis or Dermatomucosomyositis or Wagner-Unverricht or Unverricht-Wagner or "GSD 5" or "GSD V" or "GSD type 5" or "GSD type V" or Mcardl* or Sporadic inclusion body myositi* or Inclusion Body Myositi* or "GSD 2" or "GSD II" or "GSD type 2" or "GSD type II" or Pomp* disease or Central core diseas* or Central Core Myopath* or Muscle core disease or Muscular central core disease or Shy Magee Syndrome or Shy-Magee Syndrome or Nemaline body disease or Nemaline Myopath* or Nemaline rod disease or Nemaline rod myopathy or Rod body disease or Rod Body Myopathy or Rod Myopath* or Duchenn* muscular dystrophy or Duchenn* Type Progressive Muscular Dystrophy or Duchenne or Becke* disease or Becke* muscular dystroph* or Becker dystrophinopathy or Facio-Scapulo-Humeral Dystrophy or Facio Scapulo Humeral Dystrophy or Facioscapulohumeral Atroph* or Facioscapulohumeral dystroph* or Facioscapulohumeral Muscular Dystroph* or Facioscapulohumeral myopathy or Landouzy Dejerine Dystroph* or Landouzy Dejerine muscular dystroph* or Landouzy-Dejerine Dystroph* or Landouzy-Dejerine or Limb Girdle or Limb-Girdle or Muscular Dystroph* or Myodystroph* or Dystrop* myotonica or Myotonia Atroph* or Myotonia dystroph*OR Myotonic muscular dystroph* or Steiner* disease or Steiner* Myotonic Dystroph* or Steiner* syndrome or Chronic idiopathic axonal polyneuropath* or Oculopharyngeal dystroph* Oculopharyngeal Muscular Dystroph* or Familial spastic parap* or Hereditary spastic parap* or Hypertrophic Motor Sensory Neuropath* or Hypertrophic Motor-Sensory Neuropath* or Spastic Parap* or Spastic congenital parap* or Spastic Parap* or Hypertrophic Motor Sensory Neuropath* or Strumpel* dis*OR Strumpel*-Lorrain dis* or Degenerative motor system dis* or Motor neuro* dis* or MND or Motor System Dis* or Primary lateral sclerosis or Lateral Sclerosis or Adult-onset PLS or Adult-onset primary lateral sclerosis or Multifocal motor neuropath* or Congenital myopath* or Myopath* congenital or Distal muscular dystroph* or Distal myopath* or Emery-Dreifuss or Emery Dreifuss or Eaton Lambert or Eaton-Lambert or Lambert Eaton or Lambert-Eaton or Myasthenic syndrome).ab,kw,ti.	159479
8	6 or 7 [NMD]	316873
9	5 and 8	2900
10	("IMproving FItness in NEuromuscular diseases" or "IM FINE" or "B-FIT").ab,kw,ti.	70
11	(NCT04322357 or NCT04187482 or NCT04173234 or NCT03324152 or NCT03293615 or NCT02158156 or NCT02121678 or NCT02020187 or NCT02003937 or NCT01650818 or NCT01501019 or NCT01116570 or NCT01047761 or NCT03326622 or NCT01895283 or ISRCTN99826269 or ISRCTN67078621 or NL7344 or NL7899 or NL7374 or ACTRN12617001244392).ab,cn,ti.	12
12	or/9-11	2977
	Ebscohost SPORTdiscusSearch date: 30 June 2021	
1	AB ("Amyotrophic Lateral Sclerosis" OR "Amyotrophic sclerosis" OR "Gehri* Disease" OR "Lou-Gehri* Disease" OR als OR "Neuromuscular Disease?" or "Motor Neuron Disease" or "x-linked Bulbo-Spinal Atrophy" or "progressive Bulbar Palsy" or "Muscular Diseases" or "atrophic Muscular Disorder?" or "Postpoliomyelitis Syndrome" or "Muscular dystroph*" or "Myotonic dystroph*" or "Facioscapulohumeral Muscular Dystrophy" or "Duchenne" or "Emery-Dreifuss" or "Distal Myopathies" or "congentinal Myopath*" or "nemaline yopathies" or "central core Myopathy" or "Myositis" or "Myositis" or "Polymyositis" or "Dermatomyositis" or "Myotonic Disorders" or "myotonia congenita" or "Neuromuscular Junction Diseases" or "Myasthenia Gravis" or "Lambert-Eaton Myasthenic Syndrome" or "Myasthenic Syndromes" or "Peripheral Nervous System Diseases" or "Giant Axonal Neuropathy" or "Guillain-Barre Syndrome" or "Isaacs Syndrome" or "Mononeuropathies" or "Polyneuropathies" or "Polyradiculoneuropathy" or ""Hereditary Sensory and Motor Neuropathy"" or "Hereditary spastic paraplegia" or "Poliomyelitis" or "Glycogen Storage Disease" or "chronic idiopathic axonal polyneuropathy" or ""Hereditary Sensory and Motor Neuropath*"" or ""Hereditary Motor and Sensory Neuropath*"" or "Hereditary sensory-motor neuropathy" or "Hereditary sensorimotor neuropathy" or "Charcot-Marie-Tooth" or "Charcot Marie Tooth" or "Charcot-Marie Tooth" or "Peroneal musc* atrophy*" or "Charcot Marie Disease" or "Charcot-Marie Disease" or "Roussy-Levy" or "Roussy Levy" or "Post Poli*" or "Post-Poli*" or "Postpoli*" or "Polio late effects" or "Spinal Amyotroph*" or "Spinal muscular atroph*" or "Spinal muscle degeneration" or "Bulbospinal muscular atrophy" or "Spinobulbar muscular atrophy" or "Kenned* Disease" or "Mitochondrial Myopath*" or "Multiple Myositis" or "Neuromyositis" or "Polymyositi*" or "Dermatomyositis" or "Dermatopolymyositis" or "Polymyositis-Dermatomyositis" or "Dermatomucosomyositis" or "Wagner-Unverricht" or "Unverricht-Wagner" or "GSD 5" or "GSD V" or "GSD type 5" or "GSD type V" or "Mcardl*" or "Sporadic inclusion body myositi*" or "Inclusion Body Myositi*" or "Glycogen storage disease Type 2" or "Glycogen storage disease Type II" or "Glycogen storage disease II" or "Glycogen storage disease 2" or "Glycogenosis 2" or "Glycogenosis II" or "Glycogenosis type 2" or "Glycogenosis type II" or "GSD 2" or "GSD II" or "GSD type 2" or "GSD type II" or "Pomp* disease" or "Central core diseas*" or "Central Core Myopath*" or "Muscle core disease" or "Muscular central core disease" or "Shy Magee Syndrome" or "Shy-Magee Syndrome" or "Nemaline body disease" or "Nemaline Myopath*" or "Nemaline rod disease" or "Nemaline rod myopathy" or "Rod body disease" or "Rod Body Myopathy" or "Rod Myopath*" or "Duchenn" or "Becke* disease" or "Becker dystrophinopathy" or "Facio-Scapulo-Humeral Dystrophy" or "Facio Scapulo Humeral Dystrophy" or "Facioscapulohumeral Atroph*" or "Facioscapulohumeral dystroph*" or "Facioscapulohumeral myopathy" or "Landouzy Dejerine Dystroph*" or "Landouzy Dejerine" or "Landouzy-Dejerine Dystroph*" or "Landouzy-Dejerine" or "Limb Girdle" or "Limb-Girdle" or "Myodystroph*" or "Dystrop* myotonica" or "Myotonia Atroph*" or "Myotonia dystroph*" or "Steiner* disease" or "Steiner* syndrome" or "Familial spastic parap*" or "Hereditary spastic parap*" or "Hypertrophic Motor Sensory Neuropath*" or "Hypertrophic Motor-Sensory Neuropath*" or "Spastic Parap*" or "Spastic congenital parap*" or "Spastic Parap*" or "Hypertrophic Motor Sensory Neuropath*" or "Strumpel* dis*OR Strumpel*-Lorrain dis*" or "Degenerative motor system dis*" or "Motor neuro* dis*" or "MND" or "Motor System Dis*" or "Primary lateral sclerosis" or "Lateral Sclerosis" or "Adult-onset PLS" or "Adult-onset primary lateral sclerosis" or "Multifocal motor neuropath*" or "Motor neuropath* with multiple conduction block" or "Congenital myopath*" or "Myopath* congenital" or "Distal myopath*" or "Emery-Dreifuss" or "Emery Dreifuss" or "Eaton Lambert" or "Lambert Eaton" or "Myasthenic syndrome") OR TI ("Amyotrophic Lateral Sclerosis" OR "Amyotrophic sclerosis" OR "Gehri* Disease" OR "Lou-Gehri* Disease" OR als OR "Neuromuscular Disease?" or "Motor Neuron Disease" or "x-linked Bulbo-Spinal Atrophy" or "progressive Bulbar Palsy" or "Muscular Diseases" or "atrophic Muscular Disorder?" or "Postpoliomyelitis Syndrome" or "Muscular dystroph*" or "Myotonic dystroph*" or "Facioscapulohumeral Muscular Dystrophy" or "Duchenne" or "Emery-Dreifuss" or "Distal Myopathies" or "congentinal Myopath*" or "nemaline yopathies" or "central core Myopathy" or "Myositis" or "Myositis" or "Polymyositis" or "Dermatomyositis" or "Myotonic Disorders" or "myotonia congenita" or "Neuromuscular Junction Diseases" or "Myasthenia Gravis" or "Lambert-Eaton Myasthenic Syndrome" or "Myasthenic Syndromes" or "Peripheral Nervous System Diseases" or "Giant Axonal Neuropathy" or "Guillain-Barre Syndrome" or "Isaacs Syndrome" or "Mononeuropathies" or "Polyneuropathies" or "Polyradiculoneuropathy" or ""Hereditary Sensory and Motor Neuropathy"" or "Hereditary spastic paraplegia" or "Poliomyelitis" or "Glycogen Storage Disease" or "chronic idiopathic axonal polyneuropathy" or ""Hereditary Sensory and Motor Neuropath*"" or ""Hereditary Motor and Sensory Neuropath*"" or "Hereditary sensory-motor neuropathy" or "Hereditary sensorimotor neuropathy" or "Charcot-Marie-Tooth" or "Charcot Marie Tooth" or "Charcot-Marie Tooth" or "Peroneal musc* atrophy*" or "Charcot Marie Disease" or "Charcot-Marie Disease" or "Roussy-Levy" or "Roussy Levy" or "Post Poli*" or "Post-Poli*" or "Postpoli*" or "Polio late effects" or "Spinal Amyotroph*" or "Spinal muscular atroph*" or "Spinal muscle degeneration" or "Bulbospinal muscular atrophy" or "Spinobulbar muscular atrophy" or "Kenned* Disease" or "Mitochondrial Myopath*" or "Multiple Myositis" or "Neuromyositis" or "Polymyositi*" or "Dermatomyositis" or "Dermatopolymyositis" or "Polymyositis-Dermatomyositis" or "Dermatomucosomyositis" or "Wagner-Unverricht" or "Unverricht-Wagner" or "GSD 5" or "GSD V" or "GSD type 5" or "GSD type V" or "Mcardl*" or "Sporadic inclusion body myositi*" or	5959

(Contined)

Appendix SII. (*Continued*) Literature search

#	Search	Results
	Ebscohost SPORTdiscusSearch date: 30 June 2021	
	"Inclusion Body Myositi*" or "Glycogen storage disease Type 2" or "Glycogen storage disease Type II" or "Glycogen storage disease II" or "Glycogen storage disease 2" or "Glycogenosis 2" or "Glycogenosis II" or "Glycogenosis type 2" or "Glycogenosis type II" or "GSD 2" or "GSD II" or "GSD type 2" or "GSD type II" or "Pomp* disease" or "Central core diseas*" or "Central Core Myopath*" or "Muscle core disease" or "Muscular central core disease" or "Shy Magee Syndrome" or "Shy-Magee Syndrome" or "Nemaline body disease" or "Nemaline Myopath*" or "Nemaline rod disease" or "Nemaline rod myopathy" or "Rod body disease" or "Rod Body Myopathy" or "Rod Myopath*" or "Duchenn" or "Becke* disease" or "Becker dystrophinopathy" or "Facio-Scapulo-Humeral Dystrophy" or "Facio Scapulo Humeral Dystrophy" or "Facioscapulohumeral Atroph*" or "Facioscapulohumeral dystroph*" or "Facioscapulohumeral myopathy" or "Landouzy Dejerine Dystroph*" or "Landouzy Dejerine" or "Landouzy-Dejerine Dystroph*" or "Landouzy-Dejerine" or "Limb Girdle" or "Limb-Girdle" or "Myodystroph*" or "Dystrop* myotonica" or "Myotonia Atroph*" or "Myotonia dystroph*" or "Steiner* disease" or "Steiner* syndrome" or "Familial spastic parap*" or "Hereditary spastic parap*" or "Hypertrophic Motor Sensory Neuropath*" or "Hypertrophic Motor-Sensory Neuropath*" or "Spastic Parap*" or "Spastic congenital parap*" or "Spastic Parap*" or "Hypertrophic Motor Sensory Neuropath*" or "Strumpel* dis*OR Strumpel*-Lorrain dis*" or "Degenerative motor system dis*" or "Motor neuro* dis*" or "MND" or "Motor System Dis*" or "Primary lateral sclerosis" or "Lateral Sclerosis" or "Adult-onset PLS" or "Adult-onset primary lateral sclerosis" or "Multifocal motor neuropath*" or "Motor neuropath* with multiple conduction block" or "Congenital myopath*" or "Myopath* congenital" or "Distal myopath*" or "Emery-Dreifuss" or "Emery Dreifuss" or "Eaton Lambert" or "Lambert Eaton" or "Myasthenic syndrome") OR SU ("Amyotrophic Lateral Sclerosis" OR "Amyotrophic sclerosis" OR "Gehri* Disease" OR "Lou-Gehri* Disease" OR als OR "Neuromuscular Disease?" or "Motor Neuron Disease" or "x-linked Bulbo-Spinal Atrophy" or "progressive Bulbar Palsy" or "Muscular Diseases" or "atrophic Muscular Disorder?" or "Postpoliomyelitis Syndrome" or "Muscular dystroph*" or "Myotonic dystroph*" or "Facioscapulohumeral Muscular Dystrophy" or "Duchenne" or "Emery-Dreifuss" or "Distal Myopathies" or "congentinal Myopath*" or "nemaline yopathies" or "central core Myopathy" or "Myositis" or "Myositis" or "Polymyositis" or "Dermatomyositis" or "Myotonic Disorders" or "myotonia congenita" or "Neuromuscular Junction Diseases" or "Myasthenia Gravis" or "Lambert-Eaton Myasthenic Syndrome" or "Myasthenic Syndromes" or "Peripheral Nervous System Diseases" or "Giant Axonal Neuropathy" or "Guillain-Barre Syndrome" or "Isaacs Syndrome" or "Mononeuropathies" or "Polyneuropathies" or "Polyradiculoneuropathy" or "Polyradiculoneuropathy" or ""Hereditary Sensory and Motor Neuropathy"" or "Hereditary spastic paraplegia" or "Poliomyelitis" or "Glycogen Storage Disease" or "chronic idiopathic axonal polyneuropathy" or ""Hereditary Sensory and Motor Neuropath*"" or ""Hereditary Motor and Sensory Neuropath*"" or "Hereditary sensory-motor neuropathy" or "Hereditary sensorimotor neuropathy" or "Charcot-Marie-Tooth" or "Charcot Marie Tooth" or "Charcot-Marie Tooth" or "Peroneal musc* atrophy*" or "Charcot Marie Disease" or "Charcot-Marie Disease" or "Roussy-Levy" or "Roussy Levy" or "Post Poli*" or "Post-Poli*" or "Postpoli*" or "Polio late effects" or "Spinal Amyotroph*" or "Spinal muscular atroph*" or "Spinal muscle degeneration" or "Bulbospinal muscular atrophy" or "Spinobulbar muscular atrophy" or "Kenned* Disease" or "Mitochondrial Myopath*" or "Multiple Myositis" or "Neuromyositis" or "Polymyositi*" or "Dermatomyositis" or "Dermatopolymyositis" or "Polymyositis-Dermatomyositis" or "Dermatomucosomyositis" or "Wagner-Unverricht" or "Unverricht-Wagner" or "GSD 5" or "GSD V" or "GSD type 5" or "GSD type V" or "Mcardl*" or "Sporadic inclusion body myositi*" or "Inclusion Body Myositi*" or "Glycogen storage disease Type 2" or "Glycogen storage disease Type II" or "Glycogen storage disease II" or "Glycogen storage disease 2" or "Glycogenosis 2" or "Glycogenosis II" or "Glycogenosis type 2" or "Glycogenosis type II" or "GSD 2" or "GSD II" or "GSD type 2" or "GSD type II" or "Pomp* disease" or "Central core diseas*" or "Central Core Myopath*" or "Muscle core disease" or "Muscular central core disease" or "Shy Magee Syndrome" or "Shy-Magee Syndrome" or "Nemaline body disease" or "Nemaline Myopath*" or "Nemaline rod disease" or "Nemaline rod myopathy" or "Rod body disease" or "Rod Body Myopathy" or "Rod Myopath*" or "Duchenn" or "Becke* disease" or "Becker dystrophinopathy" or "Facio-Scapulo-Humeral Dystrophy" or "Facio Scapulo Humeral Dystrophy" or "Facioscapulohumeral Atroph*" or "Facioscapulohumeral dystroph*" or "Facioscapulohumeral myopathy" or "Landouzy Dejerine Dystroph*" or "Landouzy Dejerine" or "Landouzy-Dejerine Dystroph*" or "Landouzy-Dejerine" or "Limb Girdle" or "Limb-Girdle" or "Myodystroph*" or "Dystrop* myotonica" or "Myotonia Atroph*" or "Myotonia dystroph*" or "Steiner* disease" or "Steiner* syndrome" or "Familial spastic parap*" or "Hereditary spastic parap*" or "Hypertrophic Motor Sensory Neuropath*" or "Hypertrophic Motor-Sensory Neuropath*" or "Spastic Parap*" or "Spastic congenital parap*" or "Spastic Parap*" or "Hypertrophic Motor Sensory Neuropath*" or "Strumpel* dis*OR Strumpel*-Lorrain dis*" or "Degenerative motor system dis*" or "Motor neuro* dis*" or "MND" or "Motor System Dis*" or "Primary lateral sclerosis" or "Lateral Sclerosis" or "Adult-onset PLS" or "Adult-onset primary lateral sclerosis" or "Multifocal motor neuropath*" or "Motor neuropath* with multiple conduction block" or "Congenital myopath*" or "Myopath* congenital" or "Distal myopath*" or "Emery-Dreifuss" or "Emery Dreifuss" or "Eaton Lambert" or "Lambert Eaton" or "Myasthenic syndrome")	
2	AB ("aerobic capacity" or "maximal exercise test?" or "submaximal exercise test?" or (oxygen N1 uptake) or (oxygen N1 consumption) or vo2max or "vo2 max" or "vo(2)max" or "vo(2max)" or vo2peak or "vo2 peak" or "Vo(2)peak" or "vo(2peak)" or ((peak or max*) N1 ("oxygen consumption" or "oxygen uptake" or "o2 consumption" or "o2 uptake")) or "po(peak)" or "vo2 kinetics" or vent) or TI ("aerobic capacity" or "maximal exercise test?" or "submaximal exercise test?" or (oxygen N1 uptake) or (oxygen N1 consumption) or vo2max or "vo2 max" or "vo(2)max" or "vo(2max)" or vo2peak or "vo2 peak" or "Vo(2)peak" or "vo(2peak)" or ((peak or max*) N1 ("oxygen consumption" or "oxygen uptake" or "o2 consumption" or "o2 uptake")) or "po(peak)" or "vo2 kinetics" or vent) OR SU ("aerobic capacity" or "maximal exercise test?" or "submaximal exercise test?" or (oxygen N1 uptake) or (oxygen N1 consumption) or vo2max or "vo2 max" or "vo(2)max" or "vo(2max)" or vo2peak or "vo2 peak" or "Vo(2)peak" or "vo(2peak)" or ((peak or max*) N1 ("oxygen consumption" or "oxygen uptake" or "o2 consumption" or "o2 uptake")) or "po(peak)" or "vo2 kinetics" or vent)	28855
3	AB ("yo-yo" or Shuttle or "20-MST" or "20-meter" or "twenty meter" or "20 m" or Chester or "6 minute?" or "six minute?" or "6 min" or Astrand or "physical working capacity" or PWC* or "Canadian home fitness" or Conconi or Bruce or Balke or Cooper) OR TI ("yo-yo" or Shuttle or "20-MST" or "20-meter" or "twenty meter" or "20 m" or Chester or "6 minute?" or "six minute?" or "6 min" or Astrand or "physical working capacity" or PWC* or "Canadian home fitness" or Conconi or Bruce or Balke or Cooper) OR SU ("yo-yo" or Shuttle or "20-MST" or "20-meter" or "twenty meter" or "20 m" or Chester or "6 minute?" or "six minute?" or "6 min" or Astrand or "physical working capacity" or PWC* or "Canadian home fitness" or Conconi or Bruce or Balke or Cooper)	12402
4	AB ("Ventilatory threshold" or "lactate threshold" or "anaerobic threshold") OR TI ("Ventilatory threshold" or "lactate threshold" or "anaerobic threshold") OR SU ("Ventilatory threshold" or "lactate threshold" or "anaerobic threshold")	5406
5	S2 OR S3 OR S4	41978
6	S1 AND S5	215
7	AB ("IMproving FItness in NEuromuscular diseases" or "IM FINE" or "B-FIT") OR TI ("IMproving FItness in NEuromuscular diseases" or "IM FINE" or "B-FIT") OR SU ("IMproving FItness in NEuromuscular diseases" or "IM FINE" or "B-FIT")	11
8	AB (NCT04322357 OR NCT04187482 OR NCT04173234 OR NCT03324152 OR NCT03293615 OR NCT02158156 OR NCT02121678 OR NCT02020187 OR NCT02003937 OR NCT01650818 OR NCT01501019 OR NCT01116570 OR NCT01047761 OR NCT03326622 OR NCT01895283 OR ISRCTN99826269 OR ISRCTN67078621 OR NL7344 OR NL7899 OR NL7374 OR ACTRN12617001244392)	0
9	S6 OR S7 OR S8	226

(Contined)

Appendix SII. (*Continued*) Literature search

#	Search	Results
	Web of ScienceSearch date: 30 June 2021	
1	TS=("Amyotrophic Lateral Sclerosis" OR "Amyotrophic sclerosis" OR "Gehri* Disease" OR "Lou-Gehri* Disease" OR als OR "Neuromuscular Disease?" or "Motor Neuron Disease" or "x-linked Bulbo-Spinal Atrophy" or "progressive Bulbar Palsy" or "Muscular Diseases" or "atrophic Muscular Disorder?" or "Postpoliomyelitis Syndrome" or "Muscular dystroph*" or "Myotonic dystroph*" or "Facioscapulohumeral Muscular Dystrophy" or "Duchenne" or "Emery-Dreifuss" or "Distal Myopathies" or "congentinal Myopath*" or "nemaline yopathies" or "central core Myopathy" or "Myositis" or "Myositis" or "Polymyositis" or "Dermatomyositis" or "Myotonic Disorders" or "myotonia congenita" or "Neuromuscular Junction Diseases" or "Myasthenia Gravis" or "Lambert-Eaton Myasthenic Syndrome" or "Myasthenic Syndromes" or "Peripheral Nervous System Diseases" or "Giant Axonal Neuropathy" or "Guillain-Barre Syndrome" or "Isaacs Syndrome" or "Mononeuropathies" or "Polyneuropathies" or "Polyradiculoneuropathy" or "Polyradiculoneuropathy" or "Hereditary Sensory and Motor Neuropathy" or "Hereditary spastic paraplegia" or "Poliomyelitis" or "Glycogen Storage Disease" or "chronic idiopathic axonal polyneuropathy" or "Hereditary Sensory and Motor Neuropath*" or "Hereditary Motor and Sensory Neuropath*" or "Hereditary sensory-motor neuropathy" or "Hereditary sensorimotor neuropathy" or "Charcot-Marie-Tooth" or "Charcot Marie Tooth" or "Charcot-Marie Tooth" or "Peroneal musc* atrophy*" or "Charcot Marie Disease" or "Charcot-Marie Disease" or "Roussy-Levy" or "Roussy Levy" or "Post Poli*" or "Post-Poli*" or "Postpoli*" or "Polio late effects" or "Spinal Amyotroph*" or "Spinal muscular atroph*" or "Spinal muscle degeneration" or "Bulbospinal muscular atrophy" or "Spinobulbar muscular atrophy" or "Kenned* Disease" or "Mitochondrial Myopath*" or "Multiple Myositis" or "Neuromyositis" or "Polymyositi*" or "Dermatomyositis" or "Dermatopolymyositis" or "Polymyositis-Dermatomyositis" or "Dermatomucosomyositis" or "Wagner-Unverricht" or "Unverricht-Wagner" or "GSD 5" or "GSD V" or "GSD type 5" or "GSD type V" or "Mcardl*" or "Sporadic inclusion body myositi*" or "Inclusion Body Myositi*" or "Glycogen storage disease Type 2" or "Glycogen storage disease Type II" or "Glycogen storage disease II" or "Glycogen storage disease 2" or "Glycogenosis 2" or "Glycogenosis II" or "Glycogenosis type 2" or "Glycogenosis type II" or "GSD 2" or "GSD II" or "GSD type 2" or "GSD type II" or "Pomp* disease" or "Central core diseas*" or "Central Core Myopath*" or "Muscle core disease" or "Muscular central core disease" or "Shy Magee Syndrome" or "Shy-Magee Syndrome" or "Nemaline body disease" or "Nemaline Myopath*" or "Nemaline rod disease" or "Nemaline rod myopathy" or "Rod body disease" or "Rod Body Myopathy" or "Rod Myopath*" or "Duchenn" or "Becke* disease" or "Becker dystrophinopathy" or "Facio-Scapulo-Humeral Dystrophy" or "Facio Scapulo Humeral Dystrophy" or "Facioscapulohumeral Atroph*" or "Facioscapulohumeral dystroph*" or "Facioscapulohumeral myopathy" or "Landouzy Dejerine Dystroph*" or "Landouzy Dejerine" or "Landouzy-Dejerine Dystroph*" or "Landouzy-Dejerine" or "Limb Girdle" or "Limb-Girdle" or "Myodystroph*" or "Dystrop* myotonica" or "Myotonia Atroph*" or "Myotonia dystroph*" or "Steiner* disease" or "Steiner* syndrome" or "Familial spastic parap*" or "Hereditary spastic parap*" or "Hypertrophic Motor Sensory Neuropath*" or "Hypertrophic Motor-Sensory Neuropath*" or "Spastic Parap*" or "Spastic congenital parap*" or "Spastic Parap*" or "Hypertrophic Motor Sensory Neuropath*" or "Strumpel* dis*OR Strumpel*-Lorrain dis*" or "Degenerative motor system dis*" or "Motor neuro* dis*" or "MND" or "Motor System Dis*" or "Primary lateral sclerosis" or "Lateral Sclerosis" or "Adult-onset PLS" or "Adult-onset primary lateral sclerosis" or "Multifocal motor neuropath*" or "Motor neuropath* with multiple conduction block" or "Congenital myopath*" or "Myopath* congenital" or "Distal myopath*" or "Emery-Dreifuss" or "Emery Dreifuss" or "Eaton Lambert" or "Lambert Eaton" or "Myasthenic syndrome")	187090
2	TS=("aerobic capacity" or "maximal exercise test?" or "submaximal exercise test?" or (oxygen NEAR/1 uptake) or (oxygen NEAR/1 consumption) or vo2max or "vo2 max" or "vo(2)max" or "vo(2max)" or vo2peak or "vo2 peak" or "Vo(2)peak" or "vo(2peak)" or ((peak or max*) NEAR/1 ("oxygen consumption" or "oxygen uptake" or "o2 consumption" or "o2 uptake")) or "po(peak)" or "vo2 kinetics" or vent)	108787
3	TS=("Ventilatory threshold" or "lactate threshold" or "anaerobic threshold")	7361
4	TS=("yo-yo" or Shuttle or "20-MST" or "20-meter" or "twenty meter" or "20 m" or Chester or "6 minute?" or "six minute?" or "6 min" or Astrand or "physical working capacity" or PWC* or "Canadian home fitness" or Conconi or Bruce or Balke or Cooper)	110956
5	#2 OR #3 OR #4	219136
6	#1 AND #5	948
7	Refined by: WEB OF SCIENCE INDEX: (WOS.ISTP)	50
8	TS=("IMproving FItness in NEuromuscular diseases" or "IM FINE" or "B-FIT")	54
9	TS=(NCT04322357 OR NCT04187482 OR NCT04173234 OR NCT03324152 OR NCT03293615 OR NCT02158156 OR NCT02121678 OR NCT02020187 OR NCT02003937 OR NCT01650818 OR NCT01501019 OR NCT01116570 OR NCT01047761 OR NCT03326622 OR NCT01895283 OR ISRCTN99826269 OR ISRCTN67078621 OR NL7344 OR NL7899 OR NL7374 OR ACTRN12617001244392)	0
10	#7 OR #8 OR #9	104

Appendix SIII. Content validity risk of bias assessment checklist

	Very good	Adequate	Doubtful	Inadequate
1. Was an appropriate protocol used for the maximal exercise test? (Start with warm-up phase, duration of test in general between 6 and 18 min, increase in workload of 5–25 watts per min, protocol well described)	Widely recognized or well justified method used	Assumable that the method was appropriate, but not described in detail	Doubtful whether themethod was appropriate, or protocol not (clearly) described	Method used not appropriate
2. Were appropriate criteria used for assessing the content validity of the maximal exercise test? (Recommended criteria for maximal effort are 1) RER ≥ 1.1, 2) HR >90% predicted max, 3) Patient exhaustion/Borg scale ≥ 9 (range 1–10), ≥ 17 (range 6–20) and 4) a plateau in VO_2 (25, 26)	Multiple widely recognized or well justified criteria used, a minimum of 2 criteria have to be achieved	Multiple widely recognized or well justified criteria used, only one criterion has to be achieved	Doubtful whether thecriteria were appropriate	Criteria used not appropriate or not well described
3. Was the content validity tested in an appropriate number of patients?	≥ 50	≥ 30	< 30 or not clear	-
4. Was skilled personnel used to conduct the test?	An experienced test supervisor conducted the test	The test supervisor had limited experience orwas trained specifically for the study	Not clear if the test supervisor was trainedor not trained and/or had no experience	-
5. Was an appropriate approach used to analyse the data?	A widely recognized orwell justified approach was used	Assumable that the approach was appropriate, but not clearly described	Not clear what approach was used or doubtful whether the approach was appropriate	Approach not appropriate
6. Were there any other important flaws in the design or methods of the study	No other important methodological flaws	-	Other minor methodological flaws	Other important methodological flaws

Appendix SIV. Criterion validity risk of bias assessment checklist

	Very good	Adequate	Doubtful	Inadequate
1. Can the criterion used or employed be considered as a reasonable "gold standard"? (VO_{2peak}: Maximal exercise test with respiratory gas analysis. Anaerobic threshold: Lactate measurement or V-slope method)	Widely recognized or well justified gold standard used	–	Doubtful whether the gold standard was appropriate, or golden standard not clearly described	Gold standard usednot appropriate
2. Were correlations, or the area under the curve calculated?	Correlations or AUC calculated	–	–	Correlations or AUC NOT calculated
3. Were there any other important flaws in the design or methods of the study	No other important methodological flaws	–	Other minor methodological flaws	Other important methodological flaws

Appendix SV. Modified GRADE approach for grading the quality of the evidence

Quality of evidence	Lower if
High	Risk of bias
Moderate	−1 Serious
Low	−2 Very serious
Very low	−3 Extremely serious
	Inconsistency
	−1 Serious
	−2 Very serious
	Imprecision
	−1 total n=50–100
	−2 total n<50
	Indirectness
	−1 Serious
	−2 Very serious
High	We are very confident that the true measurement property lies close to that of the pooled or summarized result of the measurement property reported in the studies
Moderate	We are moderately confident in the pooled or summarized result of the measurement property found in the studies: the true measurement property is likely to be close to the measurement property reported in the studies, but there is a possibility that it is substantially different
Low	Our confidence in the pooled or summarized result of the measurement property reported in the studies is limited: the true measurement property may be substantially different
Very Low	We have very little confidence in the pooled or summarized result of the measurement property reported in the studies: the true measurement property is likely to be substantially different