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# Proposal for a candidate core-set of fitness and strength tests for patients with childhood or adult idiopathic inflammatory myopathies

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## Abstract

**OBJECTIVES—**Currently there are no evidence-based recommendations regarding which fitness and strength tests to use for patients with childhood or adult idiopathic inflammatory myopathies (IIM). This hinders clinicians and researchers in choosing the appropriate fitness- or muscle strength-related outcome measures for these patients. Through a Delphi survey, we aimed to identify a candidate core-set of fitness and strength tests for children and adults with IIM.

**METHODS—**Fifteen experts participated in a Delphi survey that consisted of five stages to achieve a consensus. Using an extensive search of published literature and through the expertise of the experts, a candidate core-set based on expert opinion and clinimetric properties was developed. Members of the International Myositis Assessment and Clinical Studies Group (IMACS) were invited to review this candidate core-set during the final stage, which led to a final candidate core-set.

**RESULTS—**A core-set of fitness- and strength-related outcome measures was identified for children and adults with IIM. For both children and adults, different tests were identified and selected for maximal aerobic fitness, submaximal aerobic fitness, anaerobic fitness, muscle strength tests and muscle function tests.

**CONCLUSIONS—**The core-set of fitness and strength-related outcome measures provided by this expert consensus process will assist practitioners and researchers in deciding which tests to use in IIM patients. This will improve the uniformity of fitness and strength tests across studies, thereby facilitating the comparison of study results and therapeutic exercise program outcomes among patients with IIM.

### Keywords

exercise; fitness; Myositis; outcome measures; instruments

## Introduction

Childhood and adult Idiopathic Inflammatory Myopathies (IIM) are rare systemic autoimmune diseases that are characterized by chronic muscle inflammation and weakness (1). Patients with the subtypes of IIM – juvenile dermatomyositis (JDM), juvenile polymyositis (JPM), adult dermatomyositis (DM), adult polymyositis (PM) and inclusion body myositis (IBM) – frequently experience anaerobic and aerobic exercise intolerance and fatigue (2-4), and may be limited in their daily physical functioning, which can lead to a poorer quality of life (5). This has led to an exploration of interventions such as exercise training programs (6). To assess the clinical status of IIM and to quantify changes in physical functioning over time, validated fitness and strength tests are essential.

Fitness tests can be divided into three general fitness categories: maximal aerobic fitness tests, submaximal aerobic fitness tests and anaerobic fitness tests. Maximal aerobic fitness

tests determine the maximal oxygen uptake ($VO_{2max}$). Submaximal aerobic fitness predict aerobic fitness using a submaximal exercise protocol (i.e., an exercise test that does not require the participant's maximal effort) (7, 8). Anaerobic fitness tests measure anaerobic performance and/or estimate the capacity of anaerobic energy pathways (e.g., phosphorylcreatine system and anaerobic glycolysis) during short-duration, high-intensity maximal exercise (usually < 30 seconds) (9). Muscle strength tests measure neuromuscular performance and can be measured with either static or dynamic muscle contractions. The neuromuscular performance measured is specific to the muscle group that is tested, which means that, for a comprehensive assessment of muscle strength, several major muscle groups must be assessed. A person's maximum strength for a given muscle group is corresponding to the maximum force that they can generate. For muscle strength tests, a distinction was made between tests that explicitly measure muscle force generation capacity, from now on referred to as *muscle strength tests*, and those that measure performance based functional capacity, from now on referred to as *muscle function tests*.

While fitness and strength testing over time can provide a quantitative assessment of the improvement or decline in the physical condition and strength of the patient with IIM, no clear recommendations are currently available for clinicians and researchers regarding which fitness or strength tests should be used in patients with IIM (1). As a result, a large variety of outcome measures have been used to evaluate the fitness and muscle strength of patients with IIM; however, most of these instruments have not been validated for this patient group (1). With a core-set of fitness- and strength-related outcome measures, the uniformity of fitness and strength tests across studies would improve, thereby facilitating the comparison of study results, allowing for a better comparison of the effects of therapeutic exercise programs.

The aim of this study is to provide a list of evidence-informed fitness- and strength-related outcome measures for patients with IIM, and thus facilitate clinicians and researchers to make better decisions about which tests to use for this patient group. This core-set of fitness and strength tests will serve as a candidate core-set and will be a basis for future research.

## Methods

### Design

The Delphi survey method was used in this study. The Delphi method has been developed to assess opinions and judgments, rather than objective facts, to reach consensus among a group of individuals (10). With the use of questionnaires, a panel of informed individuals, subsequently called experts, was asked to give feedback about a particular issue to achieve a group consensus. Experts were selected based on their activities within the International Myositis Assessment and Clinical Studies Group (IMACS) or because of their expertise in this research area (see Appendix A for the characteristics of the participants).

During stage 1 of the Delphi survey, literature was searched for fitness- and muscle strength-related outcome measures that were used in IIM. PubMed and Google Scholar were searched up to April 2013 using the following search terms: ‘physical fitness’, ‘exercise testing’, ‘exercise’, ‘exercise capacity’, ‘exercise tolerance’, ‘muscle strength’, ‘muscle

force', 'dermatomyositis', and 'myositis'. All articles that were available at the time of the search and that matched the inclusion criteria were included in this Delphi survey. Studies were included if: 1) the study population of the articles consisted of IIM patients; and 2) the studies included an examination of a fitness and/or strength test. No other inclusion and exclusion criteria's were applied. Furthermore reference lists of all the selected articles were searched for additional studies. The outcome measures were listed and categorized as follows: 1) maximal aerobic fitness tests, 2) submaximal aerobic fitness tests, 3) anaerobic fitness tests, 4) muscle strength tests and 5) muscle function tests. Afterwards, the experts were asked to list additional tests from their clinical practice or from the unpublished or published literature, that were not yet listed, in order to make the list as complete as possible.

In stage 2, the experts were asked to rate the completed list of fitness and muscle strength tests that were identified in the first Delphi stage. All the tests were rated separately for children and adults. All experts rated each outcome measure on a 10-point scale for four different topics (i.e., safety, suitability, user friendliness, and overall rating). These four topics were chosen based on their previous use in a comparable Delphi of fitness outcomes for children with cerebral palsy (11). Moreover, the experts were asked for additional information about the clinimetric properties of the listed tests. When a test was not studied in the IIM population, clinimetric properties were noted as unavailable.

A draft core-set was identified in stage 3 of the Delphi survey. For each category a test, or several tests, were selected based on expert ratings (median scores were provided) and clinimetric properties of the tests collected during stages 1 and 2. This core-set was presented to the experts together with the complete list of outcome measures and their ratings. Subsequently, the experts were asked whether they agreed or disagreed with each measures inclusion in the suggested core-set.

Based on the comments made by the experts during stage 3, a final draft candidate core-set was presented in stage 4. The experts were asked if they agreed or disagreed with the suggested core-set for both adults and children. Furthermore a conference call was organized to discuss the comments of the experts and to reach consensus among experts.

In stage 5 of the Delphi-survey, the final draft candidate core-set was sent out to all the IMACS members through an internet survey to reach consensus. The IMACS members could choose to complete the questionnaire anonymously or include their name (see Appendix B for the participants who have chosen to include their name). IMACS members were asked whether they agreed or disagreed with the tests included in of the core-set, or to state if they were unfamiliar with a selected test. Furthermore, they were requested to report any important test that in their opinion was missing from the final draft candidate core-set. Based on the outcome of the internet survey the final draft candidate core-set was once more revised. Consensus was achieved when at least 75% of the IMACS members agreed on the inclusion of a given test in the core-set; otherwise, the test was removed from the core-set. This pre-defined cut-off score was selected as a comparable cut-off that has been used previously by the IMACs (12).

### Statistical Analysis

The experts' rating of the fitness and strength tests during each of the Delphi stages was summarized with descriptive statistics using IBM SPSS Statistics for Windows, version 20.0.

## Results

Fifteen experts participated in this Delphi survey, but only 12 completed the entire survey. One expert dropped out during stage 1 due to a lack of time, while the other 2 experts, both adult physicians, dropped out during stage 2 as they felt they did not have enough expertise to score all tests. The experts' characteristics can be found in Appendix A.

### Results of Delphi stage 1

In the first stage of the Delphi survey, lasting from the 1st of February until 10 June 2013, 22 tests were identified in the literature search. Furthermore, the experts suggested 16 additional tests, though 7 of these tests were excluded as they did not measure an outcome of interest. See Figure 1 for a flow diagram showing the identification of the tests. For the complete list of tests identified, see Appendix C.

For maximal aerobic fitness tests, the incremental cycle ergometer test (intraclass correlation coefficient ICC>0.95) was the only test with published reliability data in IIM (12). No data on reliability and/or validity of submaximal aerobic fitness tests in IIM were available. For anaerobic fitness tests, the Wingate cycle test (ICC>0.85) was the only test with published reliability data in IIM (12).

The isometric dynamometer, Manual Muscle Testing (MMT) and the 1 kilogram arm lift test were the only muscle strength tests with available reliability and concurrent validity data. The isometric dynamometer as used by Stoll et al. (13) showed strong and significant intra and interobserver correlations, as well as significant and strong correlations between measurements of the left and right side. MMT was highly correlated with total and proximal MMT scores and with the Childhood Myositis Assessment Scale (CMAS), and moderately correlated with physician global activity, functional disability, magnetic resonance imaging, and axial and distal MMT scores, and, in adults, with creatine kinase level (14). The 1 kilogram arm lift test showed excellent test-retest reliability, and correlated inversely with serum creatinine kinase (15).

For several muscle function tests reliability and concurrent validity data were available in IIM: the Childhood Myositis Assessment Scale (CMAS) in children (ICC=0,89; highly correlated with the Childhood Health Assessment Questionnaire score and with MMT scores, and moderately correlated with physician-assessed global disease activity and skin activity, parent-assessed global disease severity and muscle magnetic resonance imaging) (16); the Functional Index (FI-2) for adults (ICC=0.86-0.99; moderately correlated with the shoulder flexion task of the preliminary revised FI and isokinetic measurements of shoulder flexion endurance) (14); and the 30-second chair stand test (excellent test-retest reliability; correlated inversely with serum creatinine kinase) (15).

### Results of Delphi stage 2

In the second Delphi stage, all experts were asked to rate the complete list of outcome measures (Appendix C) for safety, suitability, user friendliness and overall rating for both children and adults. The email to the experts was sent out on June 10 and the experts were given 7 weeks to complete the survey. The experts had no additional information about the clinimetric properties.

### Results of Delphi stages 3 and 4

On the 3rd of September 2013 a draft core-set of fitness and strength measures was presented to the experts. The tests that made it into the draft core-set were selected because they had the highest median scores in their category and/or because they had good reliability and validity data. However, because of the lack of available clinimetric properties, most tests were included based on expert's opinion, rather than on reliability and validity. The median scores and interquartile ranges of the selected tests in the proposed core-set are presented in Table 1. Experts got another 7 weeks to respond on the draft core-set.

Based on this draft core-set, a conference call was organized on November 12, 2013 with the expert panel, including the three experts who dropped out during previous stages. Using the comments from the third Delphi stage and from the conference call, a revised core-set was developed (See Table 2). In this revised core-set, two major changes were made; the Åstrand cycle test was added in the core-set for the adults, while the CMAS was removed for the adults. These changes were discussed during the conference call and consensus about these changes and the revised core-set amongst experts was reached.

### Results of the final Delphi stage

The online questionnaire was sent out on March 28, 2014, and there were 88 additional IMACS members that responded to the online questionnaire, 57 anonymously and 31 IMACS members that chose to include their name, which was listed in the appendix of contributors (Appendix B). Based on the consensus scores (Table 2), some tests were removed from the draft candidate core-set as they did not meet the requirement of the ≥ 75% consensus agreement. There were no additional tests added, as none of the additional tests were mentioned more than twice, which was not enough to reach consensus. A final candidate core-set was made, as shown in Table 3. The final candidate core-set includes 5 tests for children and 6 for adults of which the MMT, CMAS & FI-2 have been validated and shown to be reliable in people with IIM.

## Discussion

Although several other groups have identified and proposed core-sets of outcome measures for the IIM population (17-20), none of these efforts have specifically focused on a core-set for fitness- and strength-related outcome.

### Maximal aerobic fitness tests

The modified Bruce protocol (2, 21-23) and the incremental cycle ergometer test (24) were the two maximal aerobic fitness tests that were included in the core-set for both adults and

children after the first four stages of the Delphi survey. Both tests were chosen based on expert opinion rather than on their clinimetric properties. There was reliability data available for maximal cycle ergometry (25). During the final stage of the Delphi survey there was no consensus reached about whether or not to use the modified Bruce Protocol in children. Therefore this test was removed from the candidate core-set.

For adult IIM patients, both the modified Bruce protocol and the incremental cycle ergometer protocol were included in the final core-set. The experts decided it would be better to use the modified Bruce protocol, as it makes the test more accessible for IIM patients with reduced physical function. The incremental cycle ergometer test includes workload increases dependent on disease activity and body height with 10, 15 or 20 watts/minute (24, 25). In addition, the expert panel advised to use a 5 watts/minute increase for very weak patients.

There was no data available regarding the clinimetric properties of the (modified) Bruce protocol or the incremental cycle ergometer test, and therefore further research is needed to validate these tests for IIM. Further research is also needed to identify maximal aerobic field-tests to include in the core-set.

### Submaximal aerobic fitness tests

During the first four Delphi stages, the 6-minute walk test (6-MWT) was included in the core-set for both children and adults. As an addition to the core-set for adults, the Åstrand cycle test was also included. Both tests were included based on expert opinion rather than on clinimetric properties, as these were not available. However, during the final stage of the Delphi survey there was clearly no consensus about the use of the Åstrand cycle test in adults. Therefore, this test was removed from the final candidate core-set.

The 6-MWT is a practical and simple test that is inexpensive and easy to administer, and it allows the individual to set their own pace and voluntarily stop if necessary. The 6-MWT is currently one of the core outcome measures in trials involving patients with muscle disease (26). All experts agreed that the 6-MWT should be included in the core-set. The Åstrand cycle test was initially added as a result of the feedback received in the third Delphi stage based on the clinical experience of one of the expert physical therapists. This could be an optional test for patients that have difficulty walking. The Åstrand cycle test was added for adults only, as it has been found to have large measurement errors in children (27). Even though this test is removed from the final candidate core-set, it could still be recommended in adult patients that have difficulty walking. Further research is needed to validate these tests for IIM.

### Anaerobic fitness tests

Only the Wingate cycle test was included in the draft candidate core-set to measure anaerobic fitness (28, 29). There were only 2 anaerobic fitness tests identified in the first Delphi stage, and the clinimetric properties of the Wingate cycle test in JDM have previously been published (29). Therefore, based on expert opinion and clinimetric properties, the Wingate cycle test was initially included. However, only 55.6% of the IMACS members agreed with this test to be in the core-set for children, and only 61.5% of

the IMACS members agreed with this test to be included in the core-set for adults. Unfortunately, no other alternative tests of anaerobic fitness have been identified, and therefore the final candidate core-set does not include an anaerobic fitness test. There is also no field-based anaerobic fitness test available yet for the IIM population. Future studies should investigate the clinimetric properties of a field-based anaerobic fitness test, such as the muscle power sprint test (9), to determine its potential utility as a fitness test for the IIM population.

### Muscle strength tests

Based on the first four stages of the Delphi survey, handgrip strength (21-23, 30-34) and MMT (31, 34-43) were identified as common measures of muscle strength in both adults and children. The IMACS members agreed on these two tests for both children and adults, as consensus scores of at least 75% were reached. For a better understanding of patients' muscle strength, isometric, isokinetic or isotonic strength could be measured, depending on the available equipment and the patients' abilities. However, consensus amongst IMACS members was not reached on these tests (consensus scores ranged between the 40 to 55%). Therefore, these tests were removed from the final candidate core-set (Table 3).

Handgrip strength was included in the final candidate core-set because the consensus scores were above the pre-defined 75% cut off. However, as handgrip strength does not always capture post-exercise changes in muscle performance, the MMT has been included in the core set as an addition to the handgrip strength (1). The panel of experts advised to follow the valid and reliable protocol of Rider et al. and test the described 8 muscle groups using the Kendall 0-10 scale (44).

Initially, the suggestion of the expert panel was to perform an additional test when a MMT score of ≥6 is achieved in a particular muscle group. One could choose to test isometric, isokinetic or isotonic strength for that muscle group as well, as they may be a more sensible mode of assessment than MMT in patients with little to no severe muscle involvement. For isometric and isokinetic strength, this could be done with a dynamometer, and for isotonic strength, the panel advised to measure 1 or 10-15 repetition maximum. Some clinimetric properties have been found for the 1 kg arm lift test (15), but the experts rated the 1 or 10-15 repetition maximum higher, and this test was therefore selected instead. The choice between isometric, isokinetic or isotonic strength appears to be dependent on the equipment available, the abilities of the patient, and the competencies of the clinician.

Even though consensus was not reached on these three additional muscle strength tests, it would still be advised to do one of these tests if an additional muscle strength test is needed. The low consensus scores of these three additional muscle strength tests could possibly be explained by the lack of information the respondents of the Internet survey received. The respondents were not aware that these three tests were only listed in addition to the handgrip strength and MMT, and were to be considered as an adjunct measure should a clinician need more insight in the patient's strength. Another explanation for these low consensus scores may be the limited availability of testing equipment in the centers.

There were no clinimetric properties available for the selected handgrip, isokinetic and isotonic strength tests. Further research is needed to determine whether these muscle strength tests and corresponding assessment equipment are valid and reliable to use in patients with IIM.

### Muscle function test

For muscle function, it is advised to perform the CMAS in children and the FI-2 in adults as both tests have been found to be valid and reliable in patients with IIM. Clinimetric properties have been demonstrated in the 30 seconds chair stand test (15), but this test was not included in the core-set as it was found to be redundant after the inclusion of the CMAS and/or FI-2. Consensus scores of both the CMAS in children and the FI-2 in adults were high (both 95%).

## Limitations

One of the limitations of this Delphi was the fact that the IMACS members that participated in the final Delphi round do not have extensive experience with exercise physiologic studies and exercise tests. This could have resulted in a final list of tests biased towards what tests the panelists are familiar with, and not what might be the most appropriate tests for patients with IIM.

## Recommendations for future research and clinical practice

Future clinical trials studying the effects of rehabilitation or exercise for patients with IIM are advised to incorporate the outcome measures listed in this core-set, since this will facilitate the comparability between studies. A recent review on the efficacy of exercise training in patients with IIM reported that a large variety of outcome measures were used in the studies included, impeding data pooling and meta-analysis (6).

Further, the current report identified a major gap in the knowledge regarding the clinimetric properties of many outcome measures in juvenile and adult IIM patients. In the final candidate core-set presented in this article MMT, CMAS and FI-2 are the only tests with good reliability and validity data. Therefore, more research in this area is warranted.

Availability of the tests in centers and the expertise needed to carry out these tests were not identified in this Delphi as leading arguments to have a test included in the core-set. However, for implementation in clinical practice and research we advise researchers and clinicians to obtain experience in carrying out these tests before use.

## Conclusion

We have presented a candidate core-set of fitness and strength tests for patients with childhood and adult idiopathic inflammatory myopathies. The core-set will help standardize the conduct and reporting of clinical trials of exercise therapies, and assist practitioners in deciding which tests to use when assessing patients with IIM in the clinical setting. This will facilitate comparability of results across studies and clinical programs.

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## Appendix A: Additional members of the International Myositis Outcome Assessment Collaborative Study Group who participated in the final Delphi survey and who have chosen to include their name

Luis Arboleya

Richard Barohn

Marco van Brussel

Hector Chinoy

Lorinda Chung

Robert Cooper

Mazen Dimachkie

Richard Finkel

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Thomas Griffin

Hidenaga Kawasumi

Raju Khubchandani

Ingrid Lundberg

Frank Mastaglia

Britta Maurer

Liza McCann

Merrille Needham

Marzena Olesinska

Nancy Olsen

Annet van Royen-Kerkhof

Lidia Rutkowska-Sak

Claudia Saad-Magalhaes

Adriana Sallum

Helga Sanner

Albert Selva-O'Callaghan

Clovis Silva

Vetrila Snejana

Yeong-Wook Song

Richard Vehe

Robert Wortmann

## Appendix B: Characteristics of the participants

Fifteen experts participated in this Delphi survey including adult physicians (n=5), pediatricians (n=4), physical therapists (n=4), and exercise physiologists (n=2). Furthermore, five experts were specialized in adult rheumatology and seven experts in pediatric rheumatology. All experts worked with one or more subtypes of IIM: JDM (n=11), JPM (n=7), DM (n=7), PM (n=7) and IBM (n=7). The median (interquartile range) experience with IIM patients was 15 years (7-25), with a median annual number of IIM patients treated of 50 (20-87). The experts were an international group, originating from the Netherlands (n=1), the USA (n=2), Canada (n=3), the UK (n=2), Brazil (n=1), Guatemala (n=1), Sweden (n=2), Denmark (n=1), and Australia (n=2). Most experts were members of IMACS, and participated also in the Rehabilitation and Exercise Special Interest Group, or have a high expertise in this research area.

## Appendix C: Overview of tests that were identified during the first Delphi stage

**Appendix C**

Overview of tests that were identified during the first Delphi stage

| Tests | Children | Adults |
|---|---|---|
| **Maximal aerobic fitness test** | | |
| (Modified) Bruce protocol | + | + |
| Balke protocol[a] | + | + |
| Treadmill test | + | + |
| Cycle test | + | + |

| Tests | Children | Adults |
|---|---|---|
| **Submaximal aerobic fitness test** | | |
| Treadmill test[a] | + | + |
| Åstrand 6 minute cycle test[a] | + | + |
| 6-MWT | + | + |
| RFWT[a] | + | + |
| CT12[a] | + | + |
| **Anaerobic fitness test** | | |
| 30-s Wingate cycle test | + | + |
| Isokinetic Wingate cycle test[a] | + | + |
| **Muscle strength test** | | |
| Handgrip strength | | |
| Hand-grip dynamometer | + | + |
| Isometric strength | | |
| Isometric dynamometer | + | + |
| Sphygmomanometry[a] | + | + |
| MVICT | + | + |
| Modus M393 dynamometer | + | + |
| Isokinetic strength | | |
| Isokinetic dynamometer | + | + |
| Isotonic strength | | |
| Dynamic dynamometer | + | + |
| Variable resistance exercise | + | + |
| 1/10-15 RM | + | + |
| 1kg arm lift test | + | + |
| MMT | + | + |
| **Muscle function test** | | |
| CMAS | + | + |
| Functional Index | + | + |
| Subscale 8 BOT-2 | + | + |
| TUG | + | + |
| 30s chair stand test | + | + |
| MEFT | + | + |
| Squats in 30 sec[a] | + | + |
| Sit-ups in 30 sec[a] | + | + |
| **Other test** | | |
| Houston non-exercise test[a] | + | + |

[a]Test was added by the experts during the first stage. 6-MWT: 6-minute walk test; RFWT: Rockport Fitness Walking Test; CT12: Coopers 12 minute walk/run test; RM: repetition maximum; VRM: voluntary repetition maximum; BOT-2: Bruininks-Osteretsky Test, second edition; MMT: Manual Muscle Testing; CMAS: Childhood Myositis Assessment Scale; TUG: Timed Up and Go test; MVICT: maximum voluntary isometric contraction testing; MEFT: muscle endurance with functional test.

## Appendix D. Short description of test in final candidate core-set

### Bruce Protocol

The Bruce protocol is an treadmill exercise stress test with stages of 3 minutes. The treadmill is started with 2.74 km/hr (1.7 mph) and a gradient of 10% and then increases every 3 minutes in speed and gradient as shown in the table below.

| Stage | Speed (km/hr) | Speed (mph) | Gradient |
|---|---|---|---|
| 1 | 2.74 | 1.7 | 10 |
| 2 | 4.02 | 2.5 | 12 |
| 3 | 5.47 | 3.4 | 14 |
| 4 | 6.76 | 4.2 | 16 |
| 5 | 8.05 | 5.0 | 18 |
| 6 | 8.85 | 5.5 | 20 |
| 7 | 9.65 | 6.0 | 22 |
| 8 | 10.46 | 6.5 | 24 |
| 9 | 11.26 | 7.0 | 26 |
| 10 | 12.07 | 7.5 | 28 |

Reference: Bruce RA, Kusumi F, Hosmer D. Maximal oxygen intake and nomographic assessment of functional aerobic impairment in cardiovascular disease. Am Heart J 1973; 85(4):546-562.

### Incremental cycle ergometer test

This maximal exercise test uses an electronically braked cycle ergometer. The seat height is adjusted to the participants leg length. After 1 minute of unloaded cycling, the workload increases by 10, 15, or 20 watts every minute depending on actual disease activity and body height. Participants maintain a pedal cadence of 60-80 revolutions per minute via feedback from a visual display on the ergometer. This protocol continues until the participant stops due to volitional exhaustion, despite strong verbal encouragements from the investigators.

Reference: Takken T, van der Net J, Helders PJ. Anaerobic exercise capacity in patients with juvenile-onset idiopathic inflammatory myopathies. Arthritis Rheum 2005;53:173-7.

### 6 minute walk test (6MWT)

The 6 minute walk test (6MWT) measures the distance an individual is able to walk over a total of six minutes on a hard, flat surface. The goal is for the individual to walk as far as possible in six minutes. The individual is allowed to self-pace and rest as needed as they traverse back and forth along a marked walkway.

Holland AE, Spruit MA2, Troosters T, Puhan MA, Pepin V, Saey D, McCormack MC, Carlin BW, Sciurba FC, Pitta F, Wanger J, MacIntyre N, Kaminsky DA, Culver BH, Revill

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## Handgrip strength

The subject holds the dynamometer in the hand to be tested, with the arm at right angles and the elbow by the side of the body. The handle of the dynamometer is adjusted if required - the base should rest on first metacarpal (heel of palm), while the handle should rest on middle of four fingers. When ready the subject squeezes the dynamometer with maximum isometric effort, which is maintained for about 5 seconds. No other body movement is allowed. The subject should be strongly encouraged to give a maximum effort.

Reference: Mathiowetz V, Kashman N, Volland G, Weber K, Dowe M, Rogers S. (1985). Grip and pinch strength: normative data for adults. Archives of Physical Medicine and Rehabilitation 66: 69–74.

## Manual Muscle Testing (MMT) with the Medical Research Council 0-10 scale

Different muscle groups are evaluated following the MRC 0-10 scale:

| Scale | Description |
|---|---|
| 0 | No contraction of muscle is felt |
| T | Tendon is visible, but no movement is detectable |
| 1 | Moves trough partial range of motion in the horizontal plane |
| 2 | Moves trough completion of range of motion in the horizontal plane |
| 3 | -Moves trough completion of range of motion against resistance in the horizontal plane<br>-Holds against pressure or moves through partial range of motion in an antigravity position |
| 4 | Gradual release from test position in an antigravity position |
| 5 | Holds test position (no added pressure) in an antigravity position |
| 6 | Holds test position against slight pressure in an antigravity position |
| 7 | Holds test position against slight to moderate pressure in an antigravity position |
| 8 | Holds test position against moderate pressure in an antigravity position |
| 9 | Holds test position against moderate to strong pressure in an antigravity position |
| 10 | Holds test position against strong pressure in an antigravity position |

Reference: Rider LG, Koziol D, Giannini EH, Jain MS, Smith MR, Whitney-Mahoney K, et al. Validation of manual muscle testing and a subset of eight muscles for adult and juvenile idiopathic inflammatory myopathies. Arthritis Care Res (Hoboken) 2010;62:465-72.

## The Childhood Myositis Assesment Scale (CMAS)

The Childhood Myositis Assessment Scale (CMAS) was developed to assess muscle function in the areas of strength and endurance across a wide range of abilities and age groups in children with juvenile dermatomyositis (JDM) and polymyositis. There are 14

ordinal items included which were chosen to assess primarily axial and proximal muscle groups and are ranked with standard performance and scoring methods. The CMAS has a potential range of 0-52 with higher scores indicating greater muscle strength and endurance.

Reference: Lovell DJ, Lindsley CB, Rennebohm RM, Ballinger SH, Bowyer SL, Giannini EH, Hicks JE, Levinson JE, Mier R, Pachman LM, Passo MH, Perez MD, Reed AM, Schikler KN, Smith M, Zemel LS, Rider LG. Development of validated disease activity and damage indices for the juvenile idiopathic inflammatory myopathies. II. The Childhood Myositis Assessment Scale (CMAS): a quantitative tool for the evaluation of muscle function. The Juvenile Dermatomyositis Disease Activity Collaborative Study Group. Arthritis Rheum. 1999 Oct;42(10):2213-9.

## Functional index (FI)

The FI-2 is a functional outcome developed for patients with adult polymyositis or dermatomyositis assessing muscle endurance in seven muscle groups. Each muscle group is scored as the number of correctly performed repetitions with 60 or 120 maximal number of repetitions depending on muscle group. The FI-2 is a further development of the original Functional Index (FI) and has been validated as to content and construct validity and intra- and inter-rater reliability.

Reference: Alexanderson H, Broman L, Tollbäck A, Lundberg IE, Stenström CH. Functional Index-2: validity and reliability of a disease-specific measure of impairment in patients with polymyositis and dermatomyositis. Arthritis Rheum (Arthritis Care Res), 2006;55:114-22.[

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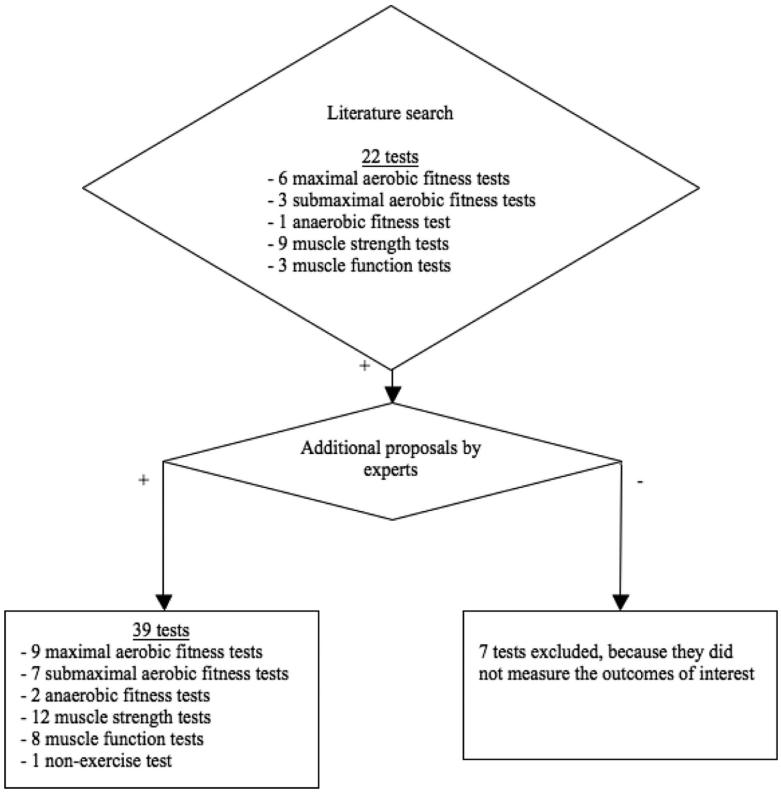


**Figure 1.**
Flow diagram on the identification of the tests.

## Table 1

Overview of tests that were presented during stage 3 of the Delphi survey

| Tests | Safety median score [25-75% IQR] | Suitability median score [25-75% IQR] | User friendliness median score [25-75% IQR] | Overall rating median score [25-75% IQR] |
|---|---|---|---|---|
| **Maximal aerobic fitness test** | | | | |
| (Modified) Bruce protocol | | | | |
| Children: | 8.0 [7.00-10] | 8.0 [7.00-10] | 7.0 [5.00-8.00] | 7.0 [7.00-9.00] |
| Adults: | 8.5 [7.00-10] | 9.0 [7.25-10] | 7.5 [6.25-9.50] | 7.0 [6.50-9.50] |
| Cycle test | | | | |
| Children: | 8.5 [7.00-9.75] | 8.0 [5.00-9.00] | 7.5 [6.25-8.00] | 8.0 [6.00-8.75] |
| Adults: | 9.0 [8.25-9.75] | 8.5 [8.00-9.75] | 7.5 [6.25-8.00] | 8.0 [7.25-8.75] |
| **Submaximal aerobic fitness test** | | | | |
| 6-MWT | | | | |
| Children: | 10 [9.00-10] | 8.0 [7.00-10] | 9.0 [8.00-10] | 9.0 [7.00-10] |
| Adults: | 10 [9.00-10] | 8.0 [5.00-8.00] | 9.0 [8.00-10] | 8.0 [6.75-9.25] |
| **Anaerobic fitness test** | | | | |
| Wingate cycle test | | | | |
| Children: | 8.0 [8.00-8.00] | 8.0 [5.00-9.00] | 8.0 [5.00-8.00] | 8.0 [6.00-8.00] |
| Adults: | 8.0 | 9.0 | 6.0 | 7.0 [6.25-8.50] |
| **Muscle strength test** | | | | |
| Handgrip strength | | | | |
| Handgrip dynamometer | | | | |
| Children: | 10 [10-10] | 6.0 [5.00-8.00] | 6.0 [6.00-7.00] | 6.0 [5.50-7.00] |
| Adults: | 10 [10-10] | 8.0 [6.00-8.00] | 6.0 [6.00-7.00] | 7.0 [5.00-8.25] |
| MMT | | | | |
| Children: | 10 [9.00-10] | 8.0 [7.00-10] | 9.0 [8.00-10] | 8.0 [7.00-10] |
| Adults: | 10 [9.00-10] | 8.0 [7.00-10] | 8.0 [7.50-10] | 8.0 [7.00-8.00] |
| Isometric strength | | | | |
| Isometric dynamometer | | | | |
| Children: | 9.0 [9.00-10] | 8.0 [7.00-9.00] | 8.0 [6.00-9.00] | 8.0 [7.00-9.00] |
| Adults: | 9.5 [9.00-10] | 8.0 [6.25-9.00] | 7.5 [6.00-8.00] | 7.5 [5.50-8.75] |
| Isokinetic strength | | | | |
| Isokinetic dynamometer | | | | |
| Children: | 9.0 [9.00-10] | 7.0 [2.00-8.00] | 5.0 [2.00-6.00] | 6.0 [2.00-7.00] |
| Adults: | 10 [9.00-10] | 8.0 [7.00-9.00] | 5.0 [4.00-6.00] | 7.0 [6.00-8.00] |
| Isotonic strength | | | | |

| Tests | Safety median score [25-75% IQR] | Suitability median score [25-75% IQR] | User friendliness median score [25-75% IQR] | Overall rating median score [25-75% IQR] |
|---|---|---|---|---|
| 1/10-15 RM | | | | |
| Children: | 8.0 [6.00-10] | 8.0 [7.00-9.25] | 7.0 [6.00-8.00] | 7.5 [6.75-8.00] |
| Adults: | 7.5 [5.25-10] | 7.0 [6.25-9.75] | 8.0 [6.25-9.75] | 7.0 [6.00-8.00] |
| **Muscle function test** | | | | |
| CMAS | | | | |
| Children: | 10 [9.00-10] | 9.0 [8.00-10] | 10 [9.00-10] | 9.0 [9.00-10] |
| Adults: | 10 [9.50-10] | 8.0 [0.50-9.00] | 10 [5.00-10] | 8.0 [3.50-9.00] |
| Functional Index | | | | |
| Adults: | 9.5 [8.75-10] | 8.0 [6.50-8.00] | 7.5 [7.00-9.25] | 7.5 [7.00-8.25] |

Legend: All tests were rated on a 10-point scale. IQR: interquartile range; 6-MWT: 6-minute walk test; MMT: Manual Muscle Testing; RM: Repetition Maximum; CMAS: Childhood Myositis Assessment Scale.

## Table 2

Results of the online questionnaire among all IMACS members during Stage 5 of the Delphi survey

| Tests | Children Consensus agreement in % (N) | Adults Consensus agreement in % (N) |
|---|---|---|
| **Maximal aerobic fitness test** | | |
| (Treadmill) Modified Bruce protocol | 58.3 % (12) | 87.5 % (16) |
| (Cycle ergometer) Incremental exercise test | 75.0 % (14) | 83.3 % (18) |
| **Submaximal aerobic fitness test** | | |
| 6-MWT | 85.7 % (21) | 89.3 % (28) |
| Astrand cycle test[a] | NA | 20.0 % (15) |
| **Anaerobic fitness test** | | |
| Wingate cycle test | 55.6 % (9) | 61.5 % (13) |
| **Muscle strength test** | | |
| Handgrip strength | | |
| Handgrip dynamometer | 82.4 % (17) | 81.5 % (27) |
| MMT | 95.0 % (20) | 96.6 % (29) |
| Isometric strength | | |
| Isometric dynamometer | 46.2 % (13) | 54.5 % (22) |
| Isokinetic strength | | |
| Isokinetic dynamometer | 41.7 % (12) | 45.5 % (22) |
| Isotonic strength | | |
| 1/10-15 RM | 53.8 % (13) | 54.5 % (22) |
| **Muscle function test** | | |
| CMAS | 95.0 % (20) | NA |
| Functional Index | NA | 95.0 % (20) |

Legend:

[a]Test was added by the experts during the first stage. The % provided in this table reflects the percentage of the IMACS members that agree on the given test of the core-set. 6-MWT: 6-minute walk test; MMT: Manual Muscle Testing; RM: Repetition Maximum; CMAS: Childhood Myositis Assessment Scale; NA: not applicable.

**Table 3**

Final candidate core-set of fitness and muscle strength tests

| Tests | Children | Adults |
|---|---|---|
| **Maximal aerobic fitness test** | | |
| (Treadmill) Modified Bruce protocol | - | + |
| (Cycle ergometer) Incremental exercise test | + | + |
| **Submaximal aerobic fitness test** | | |
| 6-MWT | + | + |
| **Muscle strength test** | | |
| Handgrip strength | | |
| Handgrip dynamometer | + | + |
| MMT | + | + |
| **Muscle function test** | | |
| CMAS | + | - |
| Functional Index-2 | - | + |

Legend: 6-MWT: 6-minute walk test; MMT: Manual Muscle Testing; CMAS: Childhood Myositis Assessment Scale.