Measurement of Physical Activity in Adults with Muscular Dystrophy: A Systematic Review

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Abstract There is little consensus about measurement of physical activity in adults with muscular dystrophy. This systematic review summarizes evidence for measurement properties of direct and indirect measures of physical activity in adults with muscular dystrophy. A two-phase search for peer-reviewed articles identified firstly, studies which measured physical activity in this population and secondly, studies reporting the measurement properties of activity measures. Methodological quality was assessed using COSMIN guidelines and a best evidence synthesis conducted. Phase 1 included 53 studies identifying 63 measures including accelerometers, direct observation, heartrate monitors, calorimetry, positional sensors, activity diaries, single scales and questionnaires. Phase 2 included 26 studies of measurement properties for 32 measures. Methodological quality of the included studies was low, only 2 were rated good. There was insufficient evidence to robustly recommend any physical activity measures and further research is required to validate measures of physical activity for adults with muscular dystrophy. Based on the findings of this review, measures with potential for further study have been highlighted.

Keywords: measure, physical activity, muscular dystrophy, adult, validity

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1. Introduction

The aim of this review was to appraise measures of physical activity for the assessment of adults with muscular dystrophy. Effective physical activity measurement is important to evaluate outcomes in randomised controlled trials (RCTs), to monitor disease progression and to make recommendations for optimising physical activity [1]. For adults with muscular dystrophy, physical activity has been linked to health benefits, such as improved fitness and self-management [2,3]. However, more research reporting quantified physical activity levels is required to determine optimal activity for adults with muscular dystrophy and to evaluate potential risks, such as exercise-induced damage to dystrophic muscle [4,5].

Figure 1. FITT framework for measurement of physical activity. Diagram created by author (SRL) based on common clinical practice and physical activity measurement analysis frameworks applied in the literature [8,9]

Physical activity is defined as behaviours involving bodily movements and energy expenditure [6,7]. Measurement of physical activity can be defined using a well-recognised conceptual framework [8,9] which considers Frequency, Intensity, Timing and Type (FITT) of activity or overall measurement encompassing these parameters (see Figure 1).

The qualities of measurement tools can be defined in terms of measurement characteristics and properties, according to the COnsensus-based St andards for the selection of health Measurement INstruments (COSMIN) taxonomy [10] (see Figure 2).

Measurement characteristics (generalizability and interpretability) of physical activity measures are variable because they depend on population and setting, and because there are numerous diverse ways to measure physical activity. These include indirect self-report tools, such as diaries and questionnaires, and direct tools which record the physiological consequences of activity, including bodily movements, metabolism and cardio-respiratory responses [11,12]. The characteristic pros and cons of these measurement tools (such as ease of use, burden, range and ability to capture FITT parameters) have been discussed in healthy individuals [8,12,13,14,15] older adults [9], wheelchair users [16] and people with neuromuscular diseases [17]. However, it is not known which measures might be most suitable for the assessment of physical activity specifically in adults with muscular dystrophy which is characterised by progressive weakness, heterogeneous presentations and variable function. It is therefore important to ascertain the generalizability and interpretability of physical activity measures in adults with muscular dystrophy to aid selection of appropriate measurement tools.
The measurement properties (reliability, responsiveness and validity) of physical activity measures have also been investigated in multiple studies in various populations, including other neurological, rheumatological, oncological or pulmonary conditions and healthy, elderly, disabled or cognitively impaired individuals [9,12,13,17]. However, cumulative evidence is inconclusive due to conflicting reports, varied study design, diversity of measures and lack of consensus about gold standard criterion measures. Furthermore, reliability, responsiveness and validity of physical activity measures established in other populations may not be transferable to adults with muscular dystrophy who may have very different muscle, metabolic and cardiac functioning [18,19,20,21]. Thus, the measurement properties of physical activity measures when used with adults with muscular dystrophy remain unclear. To the authors’ knowledge, this is the first review to examine population specific evidence for the reliability, responsiveness and validity of physical activity measurement in adults with muscular dystrophy.

The objectives of this review were, firstly, to identify direct and indirect physical activity measures used to assess adults with muscular dystrophy in a range of study designs and to describe their generalizability and interpretability. Secondly, to appraise the evidence of reliability, responsiveness and validity for physical activity measures in studies which included adults with muscular dystrophy remain unclear. To the authors’ knowledge, this is the first review to examine population specific evidence for the reliability, responsiveness and validity of physical activity measurement in adults with muscular dystrophy. The objectives of this review were, firstly, to identify direct and indirect physical activity measures used to assess adults with muscular dystrophy in a range of study designs and to describe their generalizability and interpretability. Secondly, to appraise the evidence of reliability, responsiveness and validity for physical activity measures in studies which included adults with muscular dystrophy. Finally, based on a narrative synthesis, to make recommendations, where possible, for the selection of suitable physical activity measurement tools for use with adults who have muscular dystrophy.

### 2. Methods

The protocol was registered on PROSPERO in July 2017 (Registration Number CRD42017070514) and follows the Preferred Reporting Items for Systematic Reviews and Meta-Analyses guidelines [22].

#### 2.1. Search

The search was conducted in 2 phases. Phase 1 was designed to identify physical activity measures used to assess adults with muscular dystrophy and to describe their generalizability and interpretability. Phase 2 was designed to identify and appraise evidence for their reliability, responsiveness and validity. In July 2017 (phase 1) and January 2018 (phase 2) the following databases were searched: Medline (1946-2017/8), Embase + Embase Classic (1947-2017/8), Global Health (1973-2017/8), PsycINFO (1806-2017/8), HMIC Health Management Information Consortium (1979-2017/8) and Journals @ Ovid Full Text. In phase 1, Ovid search terms were expanded from ‘muscular dystrophy’, ‘physical activity’ and ‘measure’. In phase 2, the search strategy was informed by the previous search results and additional ‘measurement property’ search terms were added. (For full search see supplementary material appendix I.)

#### 2.1.1. Study Selection

Studies were selected by 3 reviewers using the eligibility criteria listed in Table 1. (In phase 1: SRL and CW (10% sample); in phase 2: SRL, FS and CW (10% sample)). Disagreements were resolved by consensus discussion at this stage and throughout (arbitrated by CW).

Studies of any design were included if they had measured physical activity in any adult(s) with muscular dystrophy. Only studies where the measurement of activity spanned more than 10 minutes were included as shorter bouts of activity are not considered to contribute to recommended daily activity tallies [23,24,25]. The FITT framework (see Figure 1) was applied to ensure that only those intending to quantify physical activity overall or in 3 or more FITT parameters were included. When several reports pertained to the same study, the most recent or comprehensive article in terms of physical activity measurement was selected. In phase 2, inclusion was further limited to full text articles that evaluated reliability,
responsiveness and validity. A scarcity of physical activity measurement evaluation studies in adults with muscular dystrophy was anticipated. So, inclusion encompassed not only studies that overtly reported validity, reliability and responsiveness but also those that included hypothesis testing which incidentally indicated measurement properties of physical activity measures.

2.1.2. Data Extraction

The data extraction form was developed a priori, customised from previously published extraction tools [26,27] (see supplementary material appendix II). In phase 1, descriptive data were extracted by a single reviewer (SRL). In phase 2, 2 reviewers (SRL and FS) independently extracted the data.

2.1.3. Methodological Quality

In phase 1, methodological quality assessment was unnecessary because data were descriptive only. In phase 2, methodological quality was independently assessed by 2 reviewers (SRL and FS) using the COSMIN guidelines [28] to rate evidence supporting measure reliability, responsiveness or validity as excellent, good, fair or poor.

2.1.4. Synthesis

In phase 1, physical activity measures identified were described, listed and categorised. Their generalizability was quantified in terms of number of studies, number of participants, demographics (including age range, gender, diagnoses and mobility) and environment. Their interpretability was considered in terms of FITT measurement scope (i.e. capture of how often and how long different activities were carried out for and at what intensity, for example light, moderate or vigorous), timeframe, mode, metric and range of scores. In phase 2, evidence, and its methodological quality rating, for the validity, reliability and responsiveness of physical activity measures was listed for each included study. A narrative synthesis was carried out, considering the strength and consistency of evidence.

Table 1. Inclusion and exclusion criteria

| Inclusion | Exclusion |
|-----------|-----------|
| **Participants** | |
| Study cohorts including at least 1 adult (aged over 18 years*) with a diagnosis of Muscular Dystrophy including: | Non-human subjects or in vitro cell cultures |
| • Limb Girdle Muscular Dystrophy (LGMD) | Cognitive impairment, pediatric* |
| • Myotonic Myopathy (MM) | Other causes of muscle wasting including: |
| • Becker’s Muscular Dystrophy (BMD) | • Inflammatory myopathies |
| • Fascioscapulohumeral Muscular Dystrophy (FSHD) | • Metabolic myopathies |
| • Oculopharyngeal Muscular Dystrophy (OPMD) | • Other neuromuscular conditions (e.g. Spinal Muscular Atrophy, bulbar spinal muscular atrophy (Kennedy’s disease), Cerebral Palsy, Spina Bifida, Multiple Sclerosis) |
| • Myotonic Dystrophy or Dystrophia Myotonica (DM) | • Muscle wasting secondary to other conditions (e.g. Amputation, Cancer, Stroke, Spinal Cord Injury, Rheumatoid Conditions, immobility) |
| • Unspecified Muscular Dystrophy (MD) | |
| • Duchenne Muscular Dystrophy (DMD) | |
| • Adult Congenital Muscular Dystrophy (CMD) | |
| **Intervention** | |
| Physical activity including free-living activities or prescribed exercise, lasting 10 minutes or more, where physical activity can be defined as: | No physical activity |
| • “Behaviour that involves human movement, resulting in physiological attributes including increased energy expenditure and improved physical fitness” (page S11) [7] or | The following types of activities: |
| • “Any bodily movement produced by skeletal muscles that requires energy expenditure beyond resting expenditure” (page 3109) [6] and | • activity lasting less than 10 minutes (or for an undisclosed time period) |
| “the execution of a task or action by an individual” (page 577) [121] | • Activities during therapy sessions |
| **Comparison and outcomes** | |
| Studies reporting measurement of physical activity. Measurement encompassing 3 or more FITT parameters by a battery of measurement approaches or encompassing overall physical activity measurement for: | Studies with no measurement of physical activity reported, or measurement period lasting under 10mins. |
| • An evaluation of measurement properties | Studies reporting only about measures: |
| • A primary or secondary outcome | • Of activity limitation i.e. ease/difficulty of performing activities (including quality of life measures) |
| • Assessment of compliance | • Of fitness (e.g. from a single time point, such as maximal oxidative capacity, maximal workload or exertion rating) |
| • An independent variable | • Used to capture only resting activity without corresponding active measurement |
| • A demographic characteristic | |
| **Study Design** | |
| Peer reviewed articles† of any methodological design including: | Bibliographies of relevant full texts were hand searched for pertinent articles and authors, but the following were excluded: |
| Measure evaluation studies | Reviews (systematic or narrative) |
| Experimental studies (randomised designs) | Journal letters/ correspondence/ consensus summaries |
| Observational analytic studies (pre/post, comparison) | Unpublished data |
| Descriptive (longitudinal, cross-sectional, case studies) | Articles not available in English language |
| Prospective or retrospective | |

* If age range was not reported, a conservative estimated range was extracted as mean ±1 standard deviation to account for skewed distribution towards lower ages in pediatric studies. † In phase 1 abstracts and protocols were included unless superseded by a full text article; in phase 2 only full text articles were included.
3. Results

3.1. Phase 1

Study selection is summarised in Figure 3a. Agreement between reviewers (SRL and CW) was 90% and 87% for abstract and full text screening respectively, with full agreement after consensus discussion. Included articles are listed and described in Table 2; 63 physical activity measures were identified (see Table 3 and 3a supplement).

3.1.1. Generalizability

Activity measurement was generalizable across gender, mobility from independent walking to wheelchair use, age range from teenager to elderly and amongst different muscular dystrophy diagnoses. Myotonic Dystrophy and Facioscapulohumeral Dystrophy were the most commonly assessed. Indirect measures were used to assess larger numbers of participants than direct measures, particularly standardised questionnaires (n=1567). Of the direct measures, the greatest number of participants were assessed using accelerometry (n=731). Many studies used standardised questionnaires, non-standardised diaries, heartrate monitoring, direct observation and accelerometry (15 studies [29,33,43] including 3 RCTs [29,33,41], 18 studies [29,33,40-41,44-57] including 4 RCTs [33,41,44,48], 15 studies [29,33,40,44-45,48-49,50-55,58-59] including 5 RCTs [29,33,44,48,58], 13 studies [29,44,48,51,58-66] including 4 RCTs [29,48,58,65] and 13 studies [29,33,37,41,44,46,53,61,67-71] including 6 RCTs [29,33,41,44,67,68] respectively). Whereas, single and sub-scale activity measures, calorimetry and positioning systems were the least widely used in the fewest participants.

Free-living physical activity was most usually assessed, especially by questionnaires, diaries, continuous heartrate monitoring and accelerometry. However, prescribed
activities were also assessed at home and in other environments, including the gym, sports pitch and laboratory, where activity was monitored by indirect calorimetry, periodic heart rate monitoring and some training logs.

The most generalizable tools within each category, used in the most studies and participants with the widest spectrum of demographics, included 2 standardised questionnaires (the International Physical Activity Questionnaire (IPAQ), the Physical Activity Scale for Individuals with Physical Disabilities (PASIPD)), activity logs, Polar heart rate monitors and triaxial, ankle accelerometers (although, the only accelerometer used to assess non-ambulant participants was wrist-worn [46]).

### Table 2. Description of 53 included studies

| Design (n) / Description* | Citation | Condition (n) | Age range | Gender (m/f) | Phase |
|---------------------------|----------|---------------|-----------|--------------|-------|
| **Experimental (9)**      |          |               |           |              |       |
| RCT                       | Andersen et al., 2017 [29] | FSHD (12) | 26-67 | 9/3 | 1 & 2 |
|                           | Andersen et al., 2015 [33] | FDHD (41) | 22-65 | 21/20 | 1 |
| RCT (crossover)           | Voel et al., 2014 [44] | FSHD (77) | 20-79 | 37/40 | 1 & 2 |
|                           | van der Kooi et al., 2007 [65] | FSHD (65) | 27-51 | 40/25 | 1 & 2 |
| RCT (protocol)            | Veenhuizen et al., 2015 [58] | FSHD (estimated 17) | N/R | N/R | 1 |
|                           | Van Engelen et al., 2015 [68] | DM (296) | N/R | N/R | 1 |
| RCT (pilot)               | Kierkegaard et al., 2011 [48] | DM (35) | 20-65 | 15/20 | 1 & 2 |
|                           | Davies et al., 2006 [41] | BMD (4), FSHD (3), LGMD (4), CMD (1), DM (4) | 18-81 | 10/6 | 1 & 2 |
| RCT (2nd analysis)        | Janssen et al., 2016 [67] | FSHD (31) | 39-67 | 22/9 | 1 & 2 |
| **Observational analytic (26)** |          |               |           |              |       |
| Cross-sectional           | Jacques et al., 2017 [30] | BMD (21) | 26-52 | 21/0 | 1 & 2 |
|                           | Heutnick et al., 2017 [72] | DMD (estimated 30) | 16-26 | 30/0 | 1 & 2 |
|                           | McCorry et al., 1998 [55] | DM (9), FSHD (3), BMD (1) | 29-63 | 8/5 | 1 & 2 |
|                           | Brun et al., 2017 [122] | LGMD (41) | 12-72 | 20/21 | 1 |
|                           | Smith et al., 2016 [31] | MD (282) | N/R | N/R | 1 & 2 |
|                           | Morse et al., 2016 [32] | FSHD (11), BMD (11), LGMD (9), DMD (11) | 18-59 | 42/1 | 1 & 2 |
|                           | Andersen et al., 2015b [62] | DM (7), FSHD (6), LGMD (2), BMD (2) | 18-52 | 10/7 | 1 & 2 |
|                           | Vissing et al., 2014 [45] | LGMD (4), MM (2) | 39-54 | 4/2 | 1 |
|                           | Dahlqvist et al., 2014 [61] | BMD (4) | 26-32 | 4/0 | 1 |
| Cohort                    | Andersen et al., 2013 [64] | BMD (5), FSHD (5), LGMD (4) | 20-58 | 12/2 | 1 & 2 |
|                           | Sven et al., 2008 [49] | BMD (11) | 28-36 | 11/0 | 1 |
|                           | Sven et al., 2005 [50] | LGMD (9) | 43-53 | 6/3 | 1 |
|                           | Wintzen et al., 2007 [123] | DM (13) | 29-57 | 58/1 | 1 & 2 |
|                           | Wiles et al., 2006 [30] | DM (13) | 44-88 | N/R | 1 & 2 |
|                           | Aitkens et al., 2005 [54] | DM (4), LGMD (3), FSHD (1) | 47-54 | N/R | 1 & 2 |
|                           | Kilmer et al., 2005 [53] | LGMD (8), DM (4), FSHD (2), BMD (2) | 37-63 | 10/6 | 1 & 2 |
|                           | Orrgreen et al., 2005 [52] | DM (12) | 21-58 | 9/3 | 1 |
|                           | Olsen et al., 2005 [51] | FSHD (8) | 18-55 | 4/4 | 1 |
|                           | Taivassalo et al., 1999 [66] | LGMD (4), MD (3), OMPD (2), DM (1) | 29-63 | 5/5 | 1 |
|                           | Wright et al., 1996 [56] | DM (5), LGMD (1) | 28-40 | 4/2 | 1 |
|                           | Aitkens et al., 1993 [57] | DM (12), LGMD (3), FSHD (2) | 25-55 | 11/6 | 1 & 2 |
|                           | Florence and Hargreave, 1984 [74] | LGMD (3), CMD (2), FSHD (1) | 21-40 | 6/9 | 1 |
| Cohort (protocol)         | Lassche et al., 2013 [37] | FSHD (predicted 14), OMPD (predicted 14) | N/R | N/R | 1 |
| Case control              | Brady et al., 2014 [124] | DM (63) | 26-57 | 26/37 | 1 |
|                           | Angelini et al., 2011 [125] | MM (13), LGMD (3) | 16-50 | 12/4 | 1 |
|                           | Kalkman et al., 2007 [69] | DM (70), FSHD (60) | 22-61 | 75/55 | 1 & 2 |
| **Descriptive (18)**      |          |               |           |              |       |
| Cross-sectional (Evaluative) | Martin and Whalen, 2012 [75] | MD (4) | 13-64 | 2/2 | 1 & 2 |
|                           | Buss et al., 2004 [71] | MD (10) | 39-65 | 6/4 | 1 & 2 |
|                           | Washburn et al., 2002 [42] | MD (estimated 25) | N/R | N/R | 1 & 2 |
|                           | Kimura et al., 2014 [46] | DMD (12) | 10-19 | 12/0 | 1 & 2 |
|                           | Chen et al., 2014 [47] | MD (1) | 46 | 0/1 | 1 & 2 |
| Cross-sectional           | Barfield et al., 2016 [60] | MD (4) | 7-63 | N/R (4) | 1 & 2 |
|                           | Matsuda et al., 2015 [34] | MD (332) | 18-65 | 140/192 | 1 |
|                           | Rosenberg et al., 2013 [36] | MD (321) | 20-85 | 136/185 | 1 & 2 |
|                           | Shimizu-Fujiiwara et al., 2012 [126] | DMD (6) | 18-28 | 6/0 | 1 |
|                           | Komaki et al., 2011 [127] | DMD (26) | 14-38 | 26/0 | 1 |
|                           | Kierkegaard et al., 2011a [38] | DM (70) | 19-70 | 29/41 | 1 & 2 |
|                           | Harris et al., 2010 [73] | MD (1) | 36-50 | N/R | 1 |
|                           | Phillips et al., 2009 [40] | FSHD (5), DM (5), LGMD (1) | 32-56 | 3/8 | 1 & 2 |
|                           | Longmuir and Bar-Ori, 2000 [43] | MD (12) | 15-20 | 6/6 | 1 & 2 |
| Cross-sectional abstracts | Eichinger et al., 2013 [35] | DM (107) | N/R | 57/30 | 1 |
|                           | Phillips and Marr, 2009 [39] | DM (97), LGMD (82), FSHD (68) | 24-63 | N/R | 1 |
|                           | Meilleur et al., 2015 [61] | CMD (11) | N/R | N/R | 1 |
| Case study                | Pasotti et al., 2014 [59] | FSHD (1) | 43 | 0/1 | 1 |

*Classified according to Centre for Evidence-Based Medicine study design guidelines [128]. † Range calculated by mean (or median) ± 1 standard deviation (or confidence interval) in the absence of reported range. ‡ Estimated age range from proportion of larger sample. Abbreviations: RCT Randomized Controlled Trial; FSHD – Facioscapulohumeral Dystrophy; DM – Dystrophia Myotonica (Myotonic Dystrophy); BMD – Becker’s Muscular Dystrophy; DMD – Duchenne’s Muscular Dystrophy; LGMD – Limb Girdle Muscular dystrophy; MD – Muscular Dystrophy (type unspecified); MM – Miyoshi-like Myopathy; OMPD – Oculopharyngeal Muscular Dystrophy; CMD – Congenital Muscular Dystrophy; N/R – not reported.
| Indirect Measures (27) | Direct Measures (36) |
|------------------------|----------------------|
| IPAQ                   | Unspecified (waist)  |
| PASIPD                | Pulsmo (motion monitor) |
| B-PAR                  | Pulsmo (motion monitor) |
| EPIC                  | Pulsmo (motion monitor) |
| PASE                  | Pulsmo (motion monitor) |
| GLTEQ                 | Pulsmo (motion monitor) |
| CFS                   | Pulsmo (motion monitor) |
| BPAQ                  | Pulsmo (motion monitor) |
| FAI                   | Pulsmo (motion monitor) |
| PSDQ – activity       | Pulsmo (motion monitor) |
| ACS – participation   | Pulsmo (motion monitor) |
| FPACQ – transport     | Pulsmo (motion monitor) |
| MAQ – activity        | Pulsmo (motion monitor) |
| PAGS                  | Pulsmo (motion monitor) |
| ACR                   | Pulsmo (motion monitor) |
| AI 1-5                | Pulsmo (motion monitor) |
| DOA                   | Pulsmo (motion monitor) |
| Diary                 | Pulsmo (motion monitor) |
| Diary (electronic)    | Pulsmo (motion monitor) |
| Dairy + DQ            | Pulsmo (motion monitor) |
| Log                   | Training log [29,33,48-49,51-52] |
| Log + Q               | Training log with a self-report quantifier (rating of perceived exertion) [41] |
| Log + D               | Training log with direct quantifiers (heartrate, revolutions per minute) [45,50] |
| Log + Q + D           | Training log with rating of perceived exertion and direct quantifiers (HR, workload) [44,46] |
| Log + Q (’phone)      | Training log by telephone with self-report quantifiers (rating of perceived exertion and repetitions) [57] |
| Interview             | Study specific interviews [33,37,124,125] |
| Survey                | Study specific survey [122] |
| Direct Measures (36)  | Unspecified [33,58] |
| Pedometer             | Pulsmo (motion monitor) |
| Pedometer             | Pulsmo (motion monitor) |
| Pedometer             | Pulsmo (motion monitor) |
| Actometer             | Pulsmo (motion monitor) |
| Actometer             | Pulsmo (motion monitor) |
| Actometer             | Pulsmo (motion monitor) |
| Actometer             | Pulsmo (motion monitor) |
| Heart monitor         | Pulsmo (motion monitor) |
| Counting              | Directly observed step counts [61] |
| Spectating            | Directly observed competitive sport [60] |
| Video + D             | Supervision via Skype with a direct quantifier (workload) [59] |
| Supervision           | Directly observed supervised exercise [29,51,58,65] |
| Supervision + R       | Directly observed exercise with self-report quantifier (rating of perceived exertion) [48] |
| Supervision + R + D   | Directly observed exercise with a direct quantifier (heartrate) [66,74] |
| Indirect calorimetry  | Gaseous exchange analyser (K,4b2) [60] |
| Indirect calorimetry  | Gaseous exchange analyser (Oxycon Mobile) [60] |
| Indirect calorimetry  | Gaseous exchange analyser (Quark b2, Cosmed) [64] |
| TEE by DLW            | Gaseous exchange analyser (2900 Metabolic System) [53-55] |
| TEE by diet           | Gaseous exchange analyser (2900 Metabolic System) [53-55] |
| TEE by HR             | Gaseous exchange analyser (2900 Metabolic System) [53-55] |
| PAL                   | Gaseous exchange analyser (2900 Metabolic System) [53-55] |
| GPS                   | Gaseous exchange analyser (2900 Metabolic System) [53-55] |

Table 3. List of 63 measures used to assess adults with muscular dystrophy

For full measure descriptions and categorizations, see Table 3a in the supplementary material.
3.1.2. Interpretability

Indirect measures collected activity spanning 3 days to a year (or lifetime), some in real-time, including activity diaries of 3 days to 6 months, and others by recall, including standardised questionnaires often over 7 days. Whereas, all direct measures recorded activity in real-time from 10 minutes to 6 months. Most recording periods lasted 1-14 days, except for direct observation, periodic heartrate monitoring and indirect calorimetry which were conducted over shorter timeframes of between 10-90 minutes. There was great variability in the metrics of activity measures, making it difficult to compare activity measurement ranges (see table 3 and 3a supplement).

Standardised questionnaires and diaries had the best scope to quantify discrete FITT parameters; whereas direct measures like accelerometry and energy expenditure calculations were usually concerned with overall physical activity.

Interpretability was boosted by using multiple physical activity measures in 32 included studies [29,32-33,36-38, 40-41,44-46,48-60,62-66,72-74], including 7 of the 9 RCTs [29,33,41,44,48,58,65]. Multiple measures increased the scope of physical activity measurement in terms of FITT in 29 studies [29,33,38,40-41,44-46,48-60,62-66,72-74] and complementary indirect and direct measures were employed in 29 studies [29,33,37,40-41,44-58, 60, 62-66,72-74].

Table 4. Study characteristics of studies and evaluated measures included in phase 2

| Author / COSMIN | Study Description | Measure(s) | Measurement properties |
|-----------------|-------------------|------------|------------------------|
| Washburn et al., 2002 GOOD | Observational descriptive cross-sectional study for measure evaluation of the Physical Activity Scale for Individuals with Physical Disabilities in 372 disabled people [42] | PASIPD (Physical Activity Scale for Individuals with Physical Disabilities) | Reliability |
| Martin and Whalen, 2012 FAIR | Observational descriptive cross-sectional study for measure evaluation of the Physical Self-Description Questionnaire in 50 disabled people [75] | PSDQ – Activity (Physical activity subscale of the Physical Self-Description Questionnaire) | Reliability |
| Kimura et al., 2014 POOR | Observational descriptive cross-sectional study for measure evaluation of the utility of a wrist actigraph for estimating muscle strength in 22 people with Duchenne’s Muscular Dystrophy [46] | Actometer (Motionlogger Watch: triaxial wrist actigraph, Ambulatory Monitoring, Ardsley, NY, USA) | Construct validity |
| Chen et al., 2014 POOR | Observational descriptive cross-sectional study for measure evaluation of a novel movement tracking system in 5 disabled people [47] | UbiTrak (GPS) (Global Positioning System and Wireless Fidelity tracker) | Reliability |
| Busse et al., 2004 POOR | Observational descriptive cross-sectional study for measure evaluation of the StepWatch in 10 people with primary muscle disease and comparison to healthy controls [71] | Actometer (StepWatch™, biaxial, ankle step activity monitor (Cymatech, USA) | Reliability |

For incidental measurement properties see table 4a in the supplementary material. Abbreviations: ICC – intraclass correlation coefficient; DMD – Duchenne’s Muscular Dystrophy; ZCM – zero crossing mode; PIM – proportional integration mode; 6MWD – 6-minute walk distance.
3.2. Phase 2

Study selection is summarised in Figure 3b. Agreement between the 2 reviewers (SRL and FS) was 86%, 87%, 91% and 86% for abstract, full text screening, data extraction and COSMIN ratings respectively, with full agreement after consensus discussion. Evidence for the reliability, responsiveness and validity of 32 physical activity measures is listed in Table 4 (and 4a supplement). Only 5 included studies [42,46,47,71,75] had as their primary objective the evaluation of measurement properties of a physical activity measure; the remaining 21 articles were included for incidental measurement properties from hypothesis testing relating to other objectives. No studies were rated as excellent; 2 were rated as good [36,42], 12 as fair and 11 as poor. This was largely due to low sample sizes and incidental measure evaluation.

3.2.1. Reliability and Responsiveness

There was very little evidence for reliability or responsiveness testing of any physical activity measures. Of the indirect measures, there was good quality evidence of internal consistency of the PASIPD from an evaluative study including 372 participants, an estimated 7% of whom had muscular dystrophy [42]. There was fair quality evidence of internal consistency of the Physical Self-Description Questionnaire (PSDQ-S) from an evaluative study including 50 participants, 8% of whom had muscular dystrophy [75]. There was also incidental report of moderate to high test-re-test reliability of the Canada Fitness Survey (CFS) [43,76].

Of the direct measures, there was poor quality evidence of good test-re-test reliability of the StepWatch [71] accelerometer and moderate measurement error of Ubirtrak (a Wi-Fi and GPS (Global Positioning System) movement tracker) [47]. There was poor quality, incidental evidence of inter-rater reliability between indirect calorimetry gaseous analysers, K4 b2c and Oxycon Mobiled [60] and responsiveness of a pedometer compared to the Physical Activity Questionnaire (BPAQ), which had consistent, fair quality evidence of discriminative validity for an activity diary [54,55] and convergent validity of a training log [48].

There was no good quality evidence supporting the validity of any direct measures. However, there was some collective, low-quality evidence concerning accelerometer and heartrate monitoring. There was cumulative, predominantly incidental, evidence of discriminative and convergent validity of accelerometry, which was stronger for triaxial accelerometers (Omron [29], Motionlogger [46] and Actilog [67,69]) than biaxial (StepWatch [70,71]) or uniaxial devices (Yamax digi-walker [53]) and for ankle [44,67,69,70,71] rather than trunk [29] or wrist [46] placement. There was incidental, consistent evidence for discriminative validity [40,54] and convergent validity [48] of Polar heartrate monitors and mixed evidence for [54,55] and against [53] discriminative validity of heartrate monitoring used with indirect calorimetry equivalence to estimate total energy expenditure.

4. Discussion

The main finding of this systematic review is that physical activity has been measured in numerous and various ways in a range of 53 studies assessing adults with muscular dystrophy. There is no consensus about the most generalizable or interpretable activity measurement tools for this group. Furthermore, evidence is limited about measure reliability, responsiveness and validity for the assessment of physical activity in adults with muscular dystrophy. Only 5 studies overtly evaluated the measurement properties of physical activity measures and none have provided high quality evidence of reliability, responsiveness and validity.

4.1. Direct Measures

Despite the paucity of evidence for reliability, responsiveness and validity of direct measures of physical activity in adults with muscular dystrophy, tools like accelerometry and heartrate monitoring might have potential. As demonstrated in the literature [9,14,15,16,77] and by the studies identified in this review, accelerometry and heartrate monitoring are both fairly generalizable and interpretable. Accelerometry can capture free-living activity over the medium (days/weeks) to long-term (months) and can detect frequency, absolute intensity, and timing, also yielding an overall quantification of physical activity. Although accelerometry cannot discern relative exertion or type of activity, it is adaptable, relatively inexpensive, and unobtrusive. In terms of measurement properties, multi-plane movement detection, although not integral for regular walking, may be more suited to irregular torsions [78], characteristic of abnormal mobility in adults with muscular dystrophy [79]. Furthermore, for healthy people and those with chronic diseases, multi-axial devices have also demonstrated stronger criterion validity and lower measurement error than uniaxial devices [80]. Similarly, the triaxial GENEActiv has been validated over 6 minutes or less in adults with myotonic dystrophy [81] with construct validity supported incidentally in a high quality RCT [82] (too recent for inclusion in this systematic search) and the biaxial StepWatch has been
extensively validated in ambulant people with Multiple Sclerosis, Parkinson’s Disease and children with Duchenne’s Muscular Dystrophy [71,83,84,85]. In contrast, criterion validity was reportedly low and measurement error unacceptably high for the uniaxial Digi-walker over 2 minutes, in ambulant adults with neuromuscular diseases, including muscular dystrophy [86]. In this review, there was more evidence for generalizability of accelerometer placement on the ankle than the trunk or wrist, although it came only from ambulant participants; whereas, wrist placement better encompassed a range of mobility including wheelchair users [46]. In the literature, wrist accelerometry has been linked to non-ambulant assessment [87] and lower measurement error at slow walking speeds [88] which may become relevant as muscular dystrophy progresses [79]. Thus, triaxial accelerometry, placed at the ankle or wrist, represents a potential tool for the assessment of physical activity in adults with muscular dystrophy, subject to establishing robust reliability, responsiveness and validity in both ambulant and non-ambulant.

Heart rate monitoring may also have potential, particularly for monitoring compliance with, and recording intensity of, prescribed exercise interventions in adults with muscular dystrophy. In this review there were tentative indications for construct validity of Polar devices. They are generalizable and can record frequency, timing and relative intensity of exertion, which is particularly useful for quantifying prescribed activity [89]. However, heart rate monitors cannot differentiate between activity and emotional heart rate responses, thus there is a requirement to collect supplementary information, such as a diary or predetermined personal activity zone heart rates [54,55]. In addition, reduced heart rate variability in muscular dystrophies [20] might impact the interpretation of heart rate comparisons, especially relative to predicted values. Higher resting and lower maximal heart rates have been reported in Duchenne’s Muscular Dystrophy [83] and increased sympathetic drive with progressive parasympathetic dampening in Facioscapulohumeral Dystrophy [21]. Similar caution is required for energy expenditure extrapolations from heart rate or accelerometer data due to potentially altered metabolic functioning in adults with muscular dystrophy [18,19,90,91]. Thus, it is advisable to report actual recorded heart rate in beats per minute, or absolute step counts, and to treat extrapolated values with circumspection.

4.2. Indirect Measures

The same reservations about energy expenditure extrapolations must be applied to indirect measures that estimate metabolic expenditure. Additional caution is also necessary when interpreting questionnaire scores due to the potential for self-report, re-call and/or social desirability bias, which usually produce overestimations [8]. However, indirect, self-report measures of physical activity for adults with muscular dystrophy are widely generalizable, inexpensive, acceptable and easy to use [9,11,12].

Several standardized questionnaires were identified as having potential in this review. The PASIPD had the strongest evidence supporting its reliability and validity which is consistent with evidence from other populations including strong test-re-test reliability [92,93], discriminative validity [94] and low [92,93,95,96] to moderate [94] criterion validity. However, significant overestimation measurement error has been reported [95]. In terms of interpretability, PASIPD comprehensively covers FITT and is sensitive to disabled and low-level activities, although it is unsuitable for comparisons with non-disabled populations. The IPAQ, BPAQ and PSDQ-S are suitable for comparison with other populations; the BPAQ and PSDQ-S are situation specific to bone health [97] and self-perception [98] respectively. The IPAQ is the most generalizable questionnaire identified in this review and various versions are available including short, long and modified versions (more sensitive to lower activity intensities and non-ambulant mobility [99,100]). In this review, the validity of the IPAQ was inconclusive. However, measurement properties established in other populations include strong test-re-test reliability [23,101,102] moderate responsiveness [103,104], low [23,100,101,103,104] to moderate [23,105] criterion and convergent [105] validity and predominantly overestimation measurement error [104,106]. Thus, if acceptable reliability, responsiveness and validity can be established and energy expenditure scores are treated circumspectly, both the PASIPD and IPAQ have potential for the assessment of physical activity in adults with muscular dystrophy.

Activity diaries also have potential as generalizable and interpretable activity measures, especially those designed to span FITT which are often used for prescribed activity monitoring. In addition, diaries might have potential as an adjunctive activity measure. Supplementary activity logs have been shown, for example, to mitigate IPAQ overestimation [107] and to improve criterion validity and measurement error [104]. Diaries have also been advocated alongside direct activity measures [14,15] and, in this review, diaries appeared to strengthen interpretation of heart rate monitoring and indirect calorimetry equivalence [53,54,55]. Activity diaries are, therefore, not only useful for monitoring prescribed activity, they may have an application as adjuncts to enhance interpretability of free-living physical activity measurement.

4.3. Implications

Clearly, all physical activity measures have limitations, both general and specific to adults with muscular dystrophy. These must be considered in study design and some authors have compiled checklists to aid measure selection [17,108]. There is also an argument, reflected by the findings of this review, for a multi-measurement approach, where multiple, complementary activity measures are employed, to improve the interpretation of physical activity measurement [14,15,16,80] and potentially improve measurement properties [104,107]. Recall bias can be neutralised by triangulation with real-time measurement and social desirability responding can be minimised by the knowledge that responses will be verified directly [109]. Recording both relative and absolute activity, by heart rate monitoring and accelerometry or GPS, can enrich physical activity data interpretation and has also been shown to improve measurement properties [110-113]. Thus, diaries, heart rate monitoring and, possibly, GPS might be suitable adjuncts to standardised questionnaires or accelerometry. A multi-measurement approach is recommended for the assessment of physical activity in adults with muscular dystrophy.
The current lack of research evaluating measurement properties of physical activity measurement in adults with muscular dystrophy means that authors should be encouraged to report study level reliability and validity of the measures employed in trials or observational studies.

In addition, measure evaluation studies are required to determine the validity, reliability and responsiveness of physical activity measures for use with adults with muscular dystrophy. The evidence, both evaluative and incidental, compiled in this review was predominantly low quality-rated, often linked to sample sizes below the 50-100 participant threshold set by COSMIN for high quality-ratings [27]. Sample size challenges include the rarity of adults with muscular dystrophy and restricting study designs to single diagnoses and/or separating ambulant and non-ambulant [1,17]. In larger samples, it is also difficult to find an activity measure suitable to encompass activity heterogeneity within and between muscular dystrophic diagnoses [114,115] and stages of disease progression [116,117,118]. Restrictive sampling is advocated for experimental designs [1]. However, to optimise statistical power, a larger, heterogeneous sample (with whole and sub-group analyses) is recommended for future evaluative studies where measurement properties are to be elucidated.

For evaluative research, it is also difficult to identify a gold-standard criterion measure of physical activity. In the wider physical activity literature, criterion measures include calorimetry, accelerometry and direct observation [8,12,13,119]. Due to burden and cost, direct observation and indirect calorimetry are limited to smaller samples and short timeframes (<1 day). Calorimetry by double-labelled water is suitable over a timeframe of 1-2 weeks, but burdensome. Energy expenditure calculations should also be viewed with caution because calorimetry is likely to be impacted by metabolic abnormalities and progressive physiological changes in muscular dystrophy [18,19,90]. Whereas, direct observation has inherent content validity [119] and, in this review, it was interpretable and generalizable in 13 studies. Thus, it represents a suitable, initial gold-standard criterion for short-term validation. Accelerometry is generalizable in larger samples and over various timeframes. Thus, accelerometry, with prior validation against direct observation, might represent a suitable criterion against which to validate other activity measures for adults with muscular dystrophy.

4.4. Strengths and Limitations

To the authors’ knowledge, this is the first systematic review about measurement characteristics and properties of physical activity measures specifically for adults with muscular dystrophy. The review employed a broad, sensitive search strategy, 3 independent reviewers and rigorous COSMIN appraisal. However, there are some limitations. These include, firstly, exclusion of non-English language articles which means relevant articles published in other languages may have been missed. Nevertheless, a recent review of physical activity measures in adults and children with neuromuscular diseases [17] did not identify additional measurement approaches beyond those identified in this review; which suggests no pertinent measures were missed. Secondly, there is potential for bias in phase 1 as only a 10% sample was second reviewed and there was no methodological appraisal. However, the descriptive nature of phase 1 was straightforward, and the methodological quality of the studies did not impact description of the tool. Thirdly, COSMIN methodology was developed for patient-reported outcome measures, and as such, the participant number cut offs may be too stringent for direct measure evaluation where smaller participant numbers can be statistically robust [120]. Finally, risk of reporting bias was introduced by the inclusion of incidental hypothesis testing (indicative of discriminative or convergent relationships for which null findings are less frequently reported) thus the case for construct validity might have been artificially inflated.

5. Conclusions

Accelerometry, heartrate monitoring, direct observation, calorimetry, GPS, questionnaires and diaries have been used to assess physical activity in adults with muscular dystrophy. They were largely generalizable for adult age ranges, both genders and ambulant and non-ambulant people with a range of muscular dystrophy diagnoses. However, interpretability varied between measures and there was insufficient evidence to support their reliability, validity or responsiveness for use with adults who have muscular dystrophy. Measures identified as having most potential in this review included multi-axial accelerometry and the PASIPD questionnaire. Future evaluative studies of these, and/or other, physical activity measures for use with adults with muscular dystrophy are required. Future evaluative study design should consider direct observation as a fundamental criterion and maximizing sample size. Study design should include an awareness of activity measure limitations (in general and specific to muscular dystrophy) and the potential for improved interpretability by multi-measurement.

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Appendix 1. Full search

1. EXP Muscular Dystrophies
2. Muscular dystrophy
3. Facioscapulohumeral
4. Limb girdle muscular dystrophy
5. Becker’s muscular dystrophy
6. Myotonic dystrophy
7. Sarcoglycanopathy
8. Duchene muscular dystrophy
9. or 10 or 2 or 3 or 4 or 5 or 6 or 7 or 8
10. Wheelchair
11. 9 or 10
12. Physical activity
13. EXP human activities/ or "activities of daily living"/ or EXP social participation/ or EXP exercise/ or EXP circuit-based exercise/ or EXP cool-down exercise/ or EXP muscle stretching exercises/ or EXP physical conditioning, human/ or EXP plyometric exercise/ or EXP resistance training/ or EXP running/ or EXP swimming/ or EXP walking/ or EXP warm-up exercise/ or leisure activities/ or recreation/ or dancing/ or gardening/ or EXP sports/ or EXP athletic performance/ or EXP physical endurance/ or EXP physical fitness/ or baseball/ or basketball/ or EXP bicycling/ or boxing/ or football/ or golf/ or gymnastics/ or hockey/ or martial arts/ or mountaineering/ or racquet sports/ or return to sport/ or running/ or jogging/ or skating/ or snow sports/ or soccer/ or EXP sports for persons with disabilities/ or swimming/ or "track and field"/ or volleyball/ or weight lifting/ or wrestling/ or youth sports/
14. EXP Motor Activity
15. Energy expenditure
16. Exercise
17. Exercise movement techniques/ or breathing exercises/ or dance therapy/ or tai ji/ or yoga/
18. Exercise Test/ or Warm-Up Exercise/ or Cool-Down Exercise/ or Exercise Therapy/ or Plyometric Exercise/ or Circuit-Based Exercise/ or Exercise Tolerance/ or exp Exercise/ or Exercise Movement Techniques/
19. Aerobic
20. Training
21. 12 or 13 or 14 or 15 or 16 or 17 or 18 or 19 or 20
22. Measures
23. Self report
24. Test
25. Score
26. Scale
27. Ergometer
28. Accelerometer
29. Actometer
30. Pedometer
31. Treadmill
32. Questionnaire
33. Assess
34. Index
35. Level
36. MET
37. Week
38. Frequency
39. Intensity
40. 22 or 23 or 24 or 25 or 26 or 27 or 28 or 29 or 30 or 31 or 32 or 33 or 34 or 35 or 36 or 37 or 38 or 39
41. Evaluation.mp. or EXP evaluation study/
42. EXP methodology/ or EXP validation process/ or valid$.mp.
43. Sensitivity.
44. Specificity.
45. Reliability
46. EXP reliability/ or EXP observation/ or direct observation.mp.
47. Responsiveness
48. Validity
Appendix 2. Data extraction tool

| Phase 1/2 |  |
|-----------|---|
| Author(s) |  |
| Title |  |
| Year |  |
| Study Design | Pilot, feasibility, RCT, pre-post, measure evaluation, observational, cross sectional |
| Study main purpose | was study primarily aimed at testing an outcome measure of PA? Y/N |
| Sample population being measured | BMD, FSHD, LGMD, DM, DMD |
| | unspecified MD |
| | Rare (Myoshi, Bethlem, Laminin alpha, collagen 6, Emery-Dreifus etc) |
| Sample size | Number with MD |
| | Details of participants not diagnosed with MD |
| Aspect of physical activity (PA) measured | Indirect (standardised questionnaires or diaries, non-standardised diaries, PA subsection, non-standardised questionnaire or interview, single scale PA level) Direct (accelerometry, heart rate monitoring, direct observation (including supervised exercise testing like VO2 and workload), positioning sensors (like GPS), calorimetry (double labelled water, indirect calorimetry) Interpretability (scope in terms of FITT framework) |
|-------------------------------|--------------------------------------------------------------------------------|
| Measure(s) of PA | Measurement tool(s) Exact wording from paper |
| Measure administration | Was the measure used to assess free-living PA? Is the administrative burden discussed (time frame, training, cognitive ability required by participant, cost, expertise, equipment, copyright, other) and was this burden considered acceptable? |
| Tool development | Concept to be measured stated (rationale and description, use of theoretical framework, intended population described, user involvement, item determination method appropriate)? Track back to referenced development paper |
| Reliability | Reliability (relative agreement) (i) test re-test: ANOVA/t-test (or non-parametric equivalent) plus ICC, Kappa or Pearson's, Spearman's correlation coefficient, Bland and Altman relative agreement (ii) inter or intra-rater: % agreement, ICC, Kappa or Krippendorff's alpha or other statistic? Measurement Error (absolute agreement) (i) test re-test: ANOVA/t-test (or non-parametric equivalent), Kappa, Bland and Altman absolute agreement (ii) inter or intra-rater: % agreement, Kappa or Krippendorff's alpha or other statistic? Internal consistency (for scale measures): Cronbach's alpha or other statistic (Kuder-Richardson formula 20, KR-20) |
| Validity | Was the test robust? (interval between tests, adequate sample size, appropriate statistics if appropriate, appropriate statistics ICC or kappa ≥ 0.4, Krippendorff's alpha ≥ 0.8, alpha or KR-20 > 0.7) |
| Responsiveness | What was the reported sensitivity to change or responsiveness? Cohen's d effect size, standardised response mean (SRM), Guyatt's. If scoring or cut-offs are part of data analysis, does the paper provide sufficient detail including information on response options, scoring cut offs and interpretation? Was this test robust? (interval between tests, change over time or change following intervention clearly reported, standardised response means, effect size, other statistic, adequate results reported) |
| COSMIN generalizability | 1. Median or mean age (with standard deviation or range) 2. Distribution of sex (conservatively estimated by mean ±1 SD to account for potentially skewed lower age distributions, particularly in DMD) 3. Important disease characteristics (e.g. severity, status, duration) and description of treatment 4. Setting(s) in which the study was conducted (e.g. general population, primary care or hospital/rehabilitation care) 5. Countries in which the study was conducted 6. Language in which the HR-PRO instrument was evaluated 7. Method used to select patients (e.g. convenience, consecutive, or random) 8. Percentage of missing responses (response rate) |
| COSMIN Interpretabililty | 1. Percentage of missing items 2. Description of how missing items were handled 3. Distribution of the total scores 4. Percentage of participants with lowest possible score (floor effect) 5. Percentage of participants with the highest possible score (ceiling effect) 6. Scores and change scores (i.e. means and SD) for relevant (sub) groups, e.g. for normative groups, subgroups of patients, or the general population 7. Minimal Important Change (MIC) or Minimal Important Difference (MID) Cohen's d effect size, standardised response mean (SRM), Guyatt's |
| COSMIN checklist | All relevant boxes A-I |
| COSMIN score | Excellent, good, fair or poor, Lowest score counts |
| Comments | |
Table 3a. List of 63 measures used to assess adults with muscular dystrophy (27 indirect and 36 direct measures)

| Measure type (n) | Measure | Description of measurement (FTTT parameters*) | Timeframe (mode) Environment | Metric (reported range) | Participant mobility (n) |
|------------------|---------|---------------------------------------------|-----------------------------|------------------------|------------------------|
| **Indirect**     |         |                                             |                             |                        |                        |
| standardised     |         |                                             |                             |                        |                        |
| questionnaires   |         |                                             |                             |                        |                        |
| and diaries (9)  |         |                                             |                             |                        |                        |
| **Direct**       |         |                                             |                             |                        |                        |
| standardised     |         |                                             |                             |                        |                        |
| **Physical activity diaries (8)** | | | | | |
| IPAQ             | (International Physical Activity Questionnaire) | Collected activity time and frequency in minutes per day for types, including walking, transport, household, leisure and work, and intensity levels (moderate/vigorous) to yield overall estimated total energy expenditure [29,31,34-37]. (FITTO) | 7 days (recall) | Free-living | MET minutes per week (0-15,918) | Ambulant (462) Assisted (78) Wheelchair (125) N/R (417) |
| PASIPD           | (Physical Activity Scale for Individuals with Physical Disabilities) | Collected activity frequency and time in hours/days per week for 13 activity types, including work, exercise and household, each allocated intensity levels multiplied to yield an overall estimated energy expenditure score [32,39,42]. (FITTO) | 7 days (recall) | Free-living | MET hours per day (0-39.4†) | Ambulant (20) Wheelchair (22) N/R (272) |
| B-PAR            | (Bouchard Physical Activity Record) | Recorded activity types (categorised by intensity levels 1-9) at a frequency of every 15 minutes over 24 hours to yield an overall estimated total energy expenditure score [33]. (FITTO) | 3 days (real-time) | Free-living | Kcal per day (1400-4400) | N/R |
| EPIC             | (EPIC-Norfolk Physical Activity Questionnaire) | Collected time and frequency per week of listed activity types, including household, work and recreational categories, to yield hours per week for each activity and category [40]. (FITT) | 1 year (recall) | Free-living | Hours per week (0-8.47 hours recreational activity) | Ambulant (11) |
| PASE             | (Physical Activity Scale for the Elderly) | Collected frequency and time (hours/days per week) for 13 activity types, including work, exercise and household, allocated intensity levels multiplied to yield an overall score [41]. (FITTO) | 7 days (recall) | Free-living | Arbitrary units (43-302) | Ambulant (16) |
| GLTEQ            | (Godin Leisure Time Exercise Questionnaire) | Collected frequency of mild, moderate and strenuous activities lasting 15mins or more in 4 questions, each intensity level allocated metabolic equivalents and multiplied to yield an overall exercise activity score [36]. (FITTO) | 7 days (recall) | Free-living | Arbitrary units (0-119) | Ambulant (267) Wheelchair (54) |
| CFS              | (The Canada Fitness Survey) | Collected frequency, intensity and time for home, transport, work/school and leisure activity types, carried out yearly and seasonally, yielding an overall weighted activity score [43]. (FITTO) | 1 year (recall) | Free-living | Arbitrary units (2.3-21.7†) | N/R |
| BPAQ             | (Bone-specific Physical Activity Questionnaire) | Collected types of activities carried out in each year of life, and frequency per week of activities carried out in the last year. Activities are rated according to bone-load and multiplied by time and frequency to produce the score [30,32]. (FITTO) | Lifetime / 1 year (recall) | Free-living | Arbitrary units (0-4) | Ambulant (29) Wheelchair (34) |
| FAI              | (Frenchay Activities Index) | Collected frequency of lifestyle activities, timing of work activities and intensity of household maintenance, each item rated 0-3 and summed to yield an overall score [38]. (FITTO) | 3-6 months (recall) | Free-living | Arbitrary units (19-32) | N/R |
| **Indirect**     |         |                                             |                             |                        |                        |
| non-standardised |         |                                             |                             |                        |                        |
| physical activity |         |                                             |                             |                        |                        |
| diaries (8)      |         |                                             |                             |                        |                        |
| Electronic Diary | (Activity entered into computer software) | Recorded frequency, time and type of activity by location every 30 minutes throughout the day for triangulation with direct measure [47]. (FITT) | 1 day per week | (real-time) | Free-living | Outdoor activity time per week (14.3 hours) | Wheelchair (1) |
| Activity Diary   |         | Recorded verification of direct measures non-wear time and non-wear activities [46,54-55] or all activities for clarification of true activity versus electrical interference or other events [40]. (FITT) | 3-7 days | (real-time) | Free-living | Non-wear time (16-114† mins) | Ambulant (32) Wheelchair (11) Bedridden (1) |
| Activity Diary + DQ (Direct Quantifier: step count) | Recorded daily walking activity by logging daily step counts in a diary [53]. (FITyO) | 6 months | (real-time) | prescribed exercise at home | Steps per day (estimated 4500-6250) | Ambulant (16) |
| Log (training log) |         | Collected training compliance by duration and frequency per week of training sessions, carried out on an exercise bike [29,33,49,51-52] or walking [48]. (FITT) | 8-14 weeks | (recall) | prescribed exercise at home/gym | Sessions per week (0-4 x 15-35 mins) | Ambulant (54) Assisted (4) N/R (61) |
| Log + Q (training log with self-report Quantifier: RPE) | Recorded compliance by duration, frequency and type of activity (strength exercises and walking) with activity intensity quantified by Borg CR10 rating of perceived exertion (RPE) [41]. (FIT) | 8 weeks | (real-time) | prescribed exercise at home/gym | Sessions per week (3.5 x 20 mins); RPE (light to moderate) | Ambulant (16) |
| Log + DQ (training log with Direct Quantifier: HR/ rpm) | Recorded compliance by duration and frequency of bike training sessions with intensity quantified by heartrate (HR) [50] and ergometer revolutions per minute (rpm) [45] recorded in log. (FITT) | 8 weeks | (real-time) | prescribed exercise at home/gym | Sessions per week (3-47 x 30mins); rpm (60-90) | Ambulant (6) N/R (9) |
| Measure type (n) | Measure | Description of measurement (FITT parameters*) | Timeframe (mode) | Metric (reported range) | Participant mobility (n) |
|-----------------|---------|-----------------------------------------------|-----------------|------------------------|------------------------|
| **Indirect non-standardised physical activity diaries (8)** continued | Log + Q/DQ (training log with mixed Quantifiers: RPE, HR and work) | Recorded compliance by duration and frequency of walking/ bike training sessions with intensity quantified by RPE, heartrate (HR) | 12-16 weeks (real-time) prescribed exercise at home/gym | Sessions per week (0-4×15-31 mins); RPE (11.4-14.1†); Watts (43-99†) | Ambulant (83) |
| | Telephone log + Q (training log with self-report Quantifiers: RPE and repetitions) | Collected compliance by duration and frequency of strength exercises with intensity quantified by RPE (3-point scale 1 minimal, 2 moderate, 3 marked) and exercise resistance, sets and repetitions recorded in log [57]. | 7 days (recall) prescribed exercise at home | Sessions per week (2.5 x 15-20 mins); sets and reps (3 x 4-8) | Ambulant (estimated 10) Assisted (estimated 7) |
| **Indirect subscales of standardised questionnaires (4)** | PSDQ – activity (Physical Self-Description Questionnaire) | Assessed perception of physical activity participation by Likert rating (1 false to 6 true) of statements including examples of frequency, intensity, timing and type of activities, summed to yield an overall score [75]. | Current perception | Free-living | N/R Wheelchair (4) |
| | ACS – participation ranking (Activity Card Sort) | Ranks participation (frequency or duration) in a range of activity types (e.g. social and instrumental) and intensities (e.g. high and low physical demand) depicted on cards [58]. | N/R | Free-living | N/R N/R |
| | FPACO – transport (Flemish Physical Activity Computerized Questionnaire) | Collected duration and type/intensity of transportation (active (e.g. self-propelled wheeling) versus passive (e.g. motorised)) | 7 days (recall) Free-living | Minutes of transport per day (% active (9-226** mins (< 5%)) | Wheelchair (estimated 30) |
| | MAQ – activity (Modifiable Activity Questionnaire) | Collected frequency, duration and type of light and strenuous intensity exercise activities undertaken, including sports, competition and swimming | 2 weeks (recall) Free-living | Sessions per week (0->3 x 0-64 mins) | Wheelchair (estimated 42) |
| **Indirect single-rating activity scales (4)** | PAGS (Physical Activity Grading Scale) | Collected overall activity rating, 1-6, for summer and winter (1-hardly any activity, 5- moderate exercise for 3hrs, 6- hard exercise several times per week e.g. skiing, running) | 6-month seasons / 1 year (recall) Free-living | Arbitrary rating (1-5) | Ambulant (35) N/R (70) |
| | ACR (Activity Change Rating) | Collected relative activity level rating from start to finish of intervention (worsened, unchanged or improved) | 12-20 weeks (recall) Free-living | worsened – improved | N/R |
| | Activity Increment 1-5 | Collected activity changes compared to baseline on a scale of 0 – no change to 3 – new activities undertaken | 14 days (recall) Free-living | N/R | Ambulant (11) Wheelchair (2) |
| | DOA (Daily Observation of Activity) | Recorded 4 times daily a rating of activity level from 0 (no activity) to 4 (extremely active) to yield an overall score out of 16, averaged over 14 days for each participant [65]. | 14 days (real-time) Free-living | Arbitrary units (4.1-8.1†) | Ambulant (65) |
| **Indirect non-standardised questions and interviews (2)** | Interview (study specific) | Collected frequency, time and type of exercise carried out in the year following an intervention [33], between clinic visits [124]; hours of sporting activity (>=1000 hours) pre-onset [125] and prompted contextualisation of GPS tracker data [73]. | 1 week-1 year/ pre-onset (recall) Free-living | Sessions per week (2 x 25-30 mins); n=7 > 1000 hours pre-onset | Ambulant (16) Wheelchair (1) N/R (104) |
| | Survey (study specific) | Collected physical activity level (PAL) on a single Likert scale from 0-10 and types of activity participated in at different times of life during school years and beyond) [122]. | Lifetime (recall) Free-living | PAL (5-9†) | N/R |
| **Direct accelerometric monitoring (10)** | Pedometer (Yamax Digiwalker, uniaxial pedometer) | Recorded ambulatory activities, encompassing frequency, intensity and timing of stepping activities, expressed overall in steps per day and prescribed at 125% baseline [53]. | 6 months (real-time) prescribed exercise at home | Steps per day (estimated 4500-6250) | Ambulant (16) |
| | Pedometer (unspecified) | Recorded ambulatory activities, encompassing frequency, intensity and timing of stepping activities, expressed overall in steps per day | 8 weeks (real-time) prescribed exercise at home | Steps per day (2798-8331) | Ambulant (16) |
| | Pedometer (triaxial, Omron Walking Style Pro) | Recorded ambulatory activities, encompassing frequency, intensity and timing of stepping activities, stride length used to calculate daily overall activity in cumulative distance | 4-7 days (real-time) Free-living | Distance per day (1.0-4.2 km) | Ambulant (8) Assisted (4) |
| Measure type (n) | Measure | Description of measurement (FITT parameters*) | Timeframe (mode) | Metric (reported range) | Participant mobility (n) |
|-----------------|---------|-----------------------------------------------|-----------------|------------------------|------------------------|
| Direct | | | | | |
| | | Ankle actometer | Records ankle movements overall (encompassing frequency, intensity and timing) [37]. (O) | 12 days (real-time) | Free-living | N/R |
| | | Ankle Actometer (Actigil V3.0, triaxial accelerometer) | Recorded ankle movements overall (encompassing frequency, intensity and timing) averaged as accelerations per 5-minute epoch [44,67,69]. (O) | 12 days (real-time) | Free-living | Mean accelerations per 5 minutes (1-74) |
| | | Ankle StepWatch" (Step activity monitor (SAM) biaxial accelerometer) | Recorded ankle movements overall averaged over 7 days to yield mean steps per day, per minute within peak non-consecutive 30 minutes and highest sustained 20, 30 and 60 minutes and inactivity duration percentage total time [70-71]. (TiO) | 7 days and nights (real-time) | Free-living | Steps/day (716 – 5412*) Peak steps per min (21-477); Inactive (71-90%) |
| | | Wrist Actigraph (Motionlogger Watch, triaxial accelerometer) | Recorded wrist movement counts overall (encompassing frequency, intensity and timing), analysed in terms of zero-crossing mode (ZCM) in counts per minute or proportional integration mode (PIM) in bits per second [46]. (O) | 7 days (real-time) | Free-living | ZCM counts per min (5-2209); PIM bits per second (0-90,000) |
| | | Actigraph (GTX3, triaxial accelerometer) | Recorded steps overall (encompassing frequency, intensity and timing) [61]. (O) | 7 days (real-time) | Free-living | N/R |
| | | SenseWare Pro (temperature sensing biaxial accelerometer armband) | Recorded arm movements and temperatures, algorithmically calculated as overall steps, and energy expenditure expressed as average kilocalories, per day [33]. (O) | 3 days (real-time) | Free-living | Steps per day (2200-13400) Kcal per day (1800-3500) |
| | | Kinesense / Gene Active (triaxial accelerometers) | Records movement counts overall (encompassing frequency, intensity and timing) [68]. (O) | 2 weeks (real-time) | Free-living | N/R |
| Direct | | | | | |
| | | Garmin Pulsewatch (Forerunner 50) | Recorded intensity of exercise in heart rate (HR) beats per minute (bpm) and downloads provided a record of frequency and duration of activity sessions [44]. (FITi) | 16 weeks (real-time) | prescribed exercise at home/gym | HR (109-135† bpm); sessions per week (0-3 x 29-317mins) |
| | | Polar Pulsewatch (RS400) | Recorded intensity of exercise in heart rate (HR) beats per minute (bpm) and recorded timing and frequency of warm up/ work out sessions [59]. (FITi) | 6 months (real-time) | prescribed exercise at home | HR (90-130 bpm); sessions per week (2 x 15-25mins) |
| | | Pulsewatch (make and model not specified) | Recorded frequency, timing and intensity of heart rate (HR) training zone (linked to percentage maximal oxidative capacity (%VO2 max) (Cosmed, Quark CPET)19 or percent of predicted maximal heart rate (%HRM)20) [33,58]. (FITi) | 12-16 weeks (real-time) | prescribed exercise at home/gym | HR (50-70% HRM, 70% VO2max); sessions per week (1-3 x 15-90 mins) |
| | | Polar Pulsewatch (model not specified) | Recorded intensity of heart rate (HR) training zone (corresponding to percentage of maximal oxidative capacity (%VO2 max)(CPX MedGraphics)) and recorded timing and frequency of warm up/ work out sessions [51-52]. (FITi) | 32 mins - 12 weeks (real-time) | prescribed exercise at home | HR (65% VO2max) sessions per week (2-5 x 35mins) |
| | | Polar Pulsewatch (S610) | Recorded intensity of heart rate (HR) training zone (corresponding to percentage of measured or predicted maximal heart rate (HRM) and downloads provided a record of frequency and duration of activity sessions [29,48]. (FITi) | 14-16 weeks (real-time) | prescribed exercise at home/gym | HR (60-87% HRM); sessions per week (2-3 x 20-447 mins) |
| | | Polar Pulsewatch (Accurex) | Recorded intensity of heart rate (HR) training zone (corresponding to percentage of maximal oxidative capacity (%VO2 max) (CPE MedGraphics)) and downloads provided a record of frequency and duration of activity sessions [49-50]. (FITi) | 12 weeks (real-time) | prescribed exercise at home | HR (65% VO2max) sessions per week (2-5 x 30mins) |
| | | Polar Pulsewatch (FT40) | Recorded intensity of heart rate (HR) training zone (corresponding to percentage of maximal oxidative capacity (%VO2 max)) and downloads provided a record of frequency and duration of activity sessions [45]. (FITi) | 10 weeks (real-time) | prescribed exercise at home | HR (70% VO2max) Sessions per week (3-4 x 10-30mins) |
| | | Polar Pulsewatch (Vantage XL) | Recorded continuous heart rate, analysed by time, frequency and overall activity above flex heart rate (HR) (established individually from rest and low speed treadmill walking test heart rate + 10 beats per minute) [53-55]. (FITiO) | 3 days (real-time) | Free-living | Active minutes per day (29-237mins) |

*FITT parameters: Frequency, Intensity, Time, and Type.
| Measure type (n) | Measure | Description of measurement (FITT parameters*) | Timeframe (mode) | Metric (reported range) | Participant mobility (n) |
|-----------------|---------|-----------------------------------------------|-----------------|------------------------|-------------------------|
| **Direct heart rate monitoring (9) continued** | Polar heart monitor (model not specified) | Recorded continuous heart rate, analysed by time, frequency and overall activity above flex heart rate (HR) (established individually from rest and treadmill walking tests; flex HR = 87-102† bpm) and percentage maximal heart rate (HRM) [40]. (FITT) | 3 days (real-time) | Active per day (8-25 mins); at 40% and 70% HRM (8-24 mins and 1 min) | Ambulant (11) |
| **Direct observation (7)** | Step counting | Observed type (walking) with frequency and intensity indicated by number of steps counted within timeframe [61]. (FITT) | 7 days (real-time) | Free-living | N/R Ambulant (11) |
| | Supervision (Supervised exercise) | Observed frequency, duration and type of exercise (tailored [58], strength [65] or ergometer cycling [51]) carried out in the gym or during home visits, with intensity of cycling rated vicariously as easy, hard or maximal [29] or resistance [65]. (FITT) | 8-52 weeks (real-time) | Cycling intensity (easy-maximal) Sessions per week (3-4 x 21-34 mins) | Ambulant (8) Assisted (4) N/R (estimated 25) |
| | Supervision + Q (Supervised exercise with Quantifier: RPE) | Observed frequency, duration and type of class exercises (warm up, flexibility, strength, balance, aerobic and cool down exercises), with participant-rated intensity between 1 (much too easy), 3 (sufficient) and 5 (far too strenuous) [48]. (FITT) | 14 weeks (real-time) | prescribed exercise at gym | Exercise class intensity (3-4); sessions per week (2 x 60 mins) | Ambulant (35) |
| | Supervision + DQ (Supervised exercise with Direct Quantifier: heart rate (HR)) | Observed frequency, duration and type of exercise (treadmill walking and ergometer cycling) with intensity quantified by ECG HR monitoring (and brachial pulse palpation [74] (corresponding to % HRM [66] and %VO2 max [74]). (FITT) | 12 weeks – 6 months (real-time) | prescribed exercise at home/gym | Walking/cycling HR (70% VO2max / 70-85% HRM); sessions per week (4 x 15-30 mins) | N/R |
| | Supervision + Q/DQ (Supervised exercise with mixed quantifiers: RPE, work, HR) | Observed frequency, duration and type of exercise (cycle ergometer pedalling) with intensity quantified by rating or perceived exertion (RPE), ergometer workload and heart rate (HR) (linked to percentage maximal oxidative capacity (%VO2 max) or maximal HR) [44,62-64]. (FITT) | 40-42 mins – 16 weeks (real-time) | prescribed exercise at home/laboratory | RPE (11-17†); Watts (43-109†); HR (95-177† bpm) sessions per week (0-3 x 20-42 mins) | Ambulant (106) Wheelchair (2) N/R (4) |
| | Video + DQ (Supervision via Skype with Direct Quantifier: workload) | Observed frequency, duration and type of exercise (arm cycling and strength exercises), with intensity quantified by ergometer workload in Kilocalories per minute and Theraband 50% lengthening [59]. (FITT) | 8 weeks (real-time) | prescribed exercise at home | Work in Kcal per minute (4); sessions per week (4 x 15-30 mins) | Ambulant (65) Wheelchair (1) |
| | Spectating + Q (Observation of competitive sport with Quantifier: RPE) | Observed frequency, duration and type of continuous wheelchair soccer play during scrimmages and game halves with intensity measured by rating of perceived exertion (RPE) Borg 6-20 [66]. (FITT) | 2 days (real-time) | competitive sport pitch | Wheelchair soccer RPE (6.9-16.7†) active game time (10-15 mins) | Wheelchair (4) |
| | Indirect calorimetry (by gas analyser (K4b2)) | Recorded frequency and time during training and match play spent at different intensities quantified by oxidative capacity (VO2) in metabolic equivalents (METs) measured using a telemetric portable gas analyser [60]. (FITI) | 10-15 mins (real-time) | competitive sport pitch | VO2 in METs (0.98-2.54†) | Wheelchair (4) |
| | Indirect calorimetry (by gas analyser (Oxycon Mobile)) | Recorded frequency and time during training and match play spent at different intensities quantified by oxidative capacity (VO2) in metabolic equivalents (METs) measured using a telemetric portable gas analyser [60]. (FITI) | 10-15 mins (real-time) | competitive sport pitch | VO2 in METs (0.98-2.54†) | Wheelchair (4) |
| | Indirect calorimetry (by gas analyser (Quark b2, Cosmed)) | Recorded frequency and time during interval cycling spent at different intensities quantified by oxidative capacity (VO2) in millilitres of oxygen per minute per kilogram of body weight, measured using an online gas analyser [64]. (FITI) | 20 mins (real-time) | Laboratory testing | VO2 in ml/min/kg (11-37†) | Ambulant (12) Wheelchair (2) |
| | Indirect calorimetry (by gas analyser (2900 Metabolic System)) | Recorded frequency and time during treadmill walking at 3 speeds, for each reaching 5 minutes of steady state intensity quantified by oxidative capacity (VO2) in millilitres of oxygen per minute, measured using an online gas analyser [53-55]. (FITI) | 15 mins (real-time) | Laboratory testing | VO2 in ml/min (579-1109†) | Ambulant (37) |
| | TEE by DLW (Total Energy Expenditure by Double -Labelled Water) | Recorded overall energy in Kilocalories per day calculated by double-labelled water technique monitoring excretion of stable isotopes (2H2O and H218O) [127]. (O) | N/R (real-time) | Free-living | Energy in Kcal per day (948-1320) | N/R |
Table 4a. Study characteristics of studies and evaluated measures included in phase 2

| Author                        | Study Description                                                                 | Measure(s)                                                                 | Measurement properties                                                                 | COSMIN rating |
|-------------------------------|-----------------------------------------------------------------------------------|-----------------------------------------------------------------------------|----------------------------------------------------------------------------------------|---------------|
| Rosenberg et al., 2013        | Observational descriptive cross-sectional study testing associations between depression and physical activity in 1676 people with NMD [36] | 1. IPAQ (International Physical Activity Questionnaire) 2. GLTEQ (Godin Leisure Time Exercise Questionnaire) | Incidental Construct validity 1. and 2. Incidental convergent validity was demonstrated for both questionnaires by moderate negative associations with depression (Patient Health Questionnaire-9) (multiple linear regression, IPAQ Beta=0.06, r2=0.004, P=0.012 and GLTEQ Beta=-0.13, r2=0.02, P=0.001) | GOOD          |
| Jacques et al., 2017          | Observational analytic cross-sectional study of resting energy expenditure and comparison to healthy controls [30] | BPAQ (Bone-specific Physical Activity Questionnaire)                       | Incidental discriminative validity was demonstrated by extreme group differences in BPAQ score (current) between healthy controls and adults with BMD (Independent student t-test, p<0.05, mean difference in score 4.8) and between control, ambulatory and non-ambulatory adults with BMD (ANOVA, P<0.01, mean difference in score 1.4) | FAIR          |
| Heustinck et al., 2017        | Observational analytic cross-sectional study of physical activity and comparison to healthy controls [72] | 1. FPACQ - transportation (Subscale of Flemish Physical Activity Computerized Questionnaire) 2. MAQ - activity (Subscale of Modifiable Activity Questionnaire) | Incidental discriminative validity was demonstrated by extreme group differences in FPACQ transportation time and type (active/passive) between healthy controls and people with DMD (independent t-test, p<0.001, mean difference 51 minutes less for healthy controls) at early and late non-ambulatory stages (independent t-test, p=0.003, mean differences 48 and 84 minutes respectively less for healthy controls). In addition, extreme group differences between early ambulatory stage and late ambulatory stage for people with DMD (One-way ANOVA with post hoc tests, p=0.022, mean difference 84 minutes). 2. Incidental discriminative validity was demonstrated by extreme group differences in MAQ light and strenuous activities by people with DMD at various stages of disease progression from early ambulatory to late non-ambulatory (One-way ANOVA with post hoc tests, p<0.01, mean difference 29% and 34% more doing no light and strenuous exercise respectively) | FAIR          |
| Author                  | Study Description                                                                 | Measure(s)                                                                                      | Measurement properties                                                                                                                                                                                                 | COSMIN rating |
|-------------------------|------------------------------------------------------------------------------------|-------------------------------------------------------------------------------------------------| ----------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------| ------------- |
| Janssen et al., 2016    | Randomised, controlled trial secondary analysis for measure evaluation of MRI fat fraction analysis [67] | Ankle Actometer (Actilog V3.0, triaxial)                                                        | Incidental convergent validity of ankle actometer (in average accelerations per 5 minutes over 12 days) with Magnetic Resonance Imaging (MRI) fat fraction of thigh musculature was demonstrated by a moderate negative correlation (Pearson’s r = 0.27, p = 0.0013) | FAIR          |
| Morse et al., 2016      | Observational analytic cohort study of bone health in different muscular dystrophies and comparison to healthy controls [32] | 1. BPAQ (Bone-specific Physical Activity Questionnaire) 2. PASIPD (Physical Activity Scale for Individuals with Physical Disabilities) | 1. and 2. Incidental concurrent validity was demonstrated between BPAQ and PASIPD by a strong association (reported in a later study [33]) between the 2 questionnaires (Pearson’s r = 0.71, p < 0.005)  
Incidental Construct validity  
1. Incidental discriminative validity was demonstrated by extreme group differences in BPAQ score between healthy controls and adults with muscular dystrophy (one-way between groups ANOVA with Bonferroni post hoc, p < 0.05, mean difference in score 24)  
2. Incidental discriminative validity was demonstrated by extreme group differences in PASIPD score between DMD and all other groups (one-way between groups ANOVA with Bonferroni post hoc, p < 0.05, mean difference in score 13)  
2. Incidental convergent validity was demonstrated by low and moderate associations of PASIPD scores with tibialis anterior cross-sectional area and grip strength (Pearson’s r = 0.46, p < 0.01 and r = 0.65, p < 0.05 respectively). | FAIR          |
| Smith et al., 2016      | Observational analytic cohort study of the incidence, prevalence, age of onset and predictors of 5 chronic conditions in 1594 adults with long-term physical disability [31] | IPAQ (International Physical Activity Questionnaire)                                               | Discriminative validity was not demonstrated as there was no significant difference between IPAQ scores for participants with and without new onset of any chronic comorbid condition over the 3.5-year time period (independent student t-test, t = -0.85 to 1.69, df = 1173, p > 0.05, IPAQ mean score differences for hypertension = 188.3, coronary heart disease = 372.6, cancer = 26.2, diabetes = 206.8 and arthritis = 0.0). | FAIR          |
| Voet et al., 2014       | Randomised controlled, crossover trial of aerobic exercise training and cognitive-behavioural therapy on chronic fatigue [44] | Ankle actometer (Actilog V3.0, triaxial)                                                        | Incidental convergent validity of ankle actometer (in average accelerations per 5 minutes over 12 days) with Magnetic Resonance Imaging (MRI) fat fraction of thigh musculature was demonstrated by a moderate negative correlation (Pearson’s r = 0.27, p = 0.0013) | FAIR          |
| Kierkegaard et al., 2011b | Pilot randomised controlled trial of Friskis & Svettis Open doors exercise classes in 35 adults with Myotonic Dystrophy [48] | 1. Training log 2. Direct observation of exercise classes 3. Polar Pulsewatch | 1. Incidental discriminative validity was demonstrated by extreme group differences in average accelerations per 5 minutes between participants receiving Aerobic Exercise Training (AET), or Cognitive Behavioural Therapy (CBT), and those who received usual care (linear mixed model for repeated measurements, p < 0.05, mean differences AET 4.6, CBT 5.6 and at follow up AET 5.5, CBT 7.1) | FAIR          |
| Philips et al., 2009    | Observational descriptive cross-sectional study of activity patterns and barriers in 13 people with NMD and comparison to healthy controls [40] | 1. Polar heartrate monitor (Polar Electro UK ltd) 2. EPIC (Norfolk Physical Activity Questionnaire) | 1. Incidental discriminative validity was demonstrated by extreme group differences in minutes active (above flex heart rate) between people with NMD and healthy controls on 3 week and weekend days (Mann Whitney U test, p < 0.05, median difference 154 minutes active)  
2. Incidental discriminative validity was demonstrated by extreme group differences between people with NMD and healthy controls in hours per week spent in overall physical activity measured by EPIC (Mann Whitney U test, p < 0.004, mean difference 24.1 hours per week) and work-related activities (Mann Whitney U test, p < 0.002, median difference 21.5 hours/week). There was a significant difference in choice of mode of transport to travel 1 mile (Fisher's exact test, p = 0.02, frequency difference 10 more control participants chose walking). There was no significant difference in household activities or recreational hours per week between groups. | FAIR          |
| Author                        | Study Description                                                                 | Measure(s)                                                                                   | Measurement properties                                                                                                                                                                                                 | COSMIN rating |
|------------------------------|-----------------------------------------------------------------------------------|---------------------------------------------------------------------------------------------|------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|---------------|
| Kalkman et al., 2007         | Observational analytic case-control study of predictors of fatigue in 198 people with NMD [69] | Ankle Actometer (Actilog, V3.0, triaxial)                                                   | Incidental convergent validity of average accelerations per 5 minutes from 12 days was demonstrated by low negative correlation with Sickness Impact Profile (Pearson’s r = -0.38, p<0.01, FSHD and r = -0.38, p<0.01, Dystonia Myotonica) and Checklist Individual Strength – fatigue (Pearson’s r= -0.34, p<0.01, FSHD) | FAIR          |
| Longmuir et al., 2000        | Observational descriptive cross-sectional study of habitual physical activity, perceived fitness and participation in 987 disabled youths [43] | CFS (Modified Canada Fitness Survey)                                                        | Reliability Test-re-test reliability of the CFS ranged from 0.66 to 1.00 (reported from an earlier study [76]).                                                                                                      | FAIR          |
| McCrory et al., 1998         | Observational analytic cross-sectional study of energy expenditure and physical activity patterns in 26 ambulatory adults with slowly progressive NMD and comparison to healthy controls [55] | 1. Diary (3-day activity and non-wear log book or Dictaphone logging with energy expenditure by factorial method)  
2. TEE and PAL (established by indirect calorimetry (2900 Metabolic System) with heat rate equivalence)  
3. Polar XL vantage | 1. Incidental discriminative validity was demonstrated by extreme group differences between people with FSHD 1 and healthy controls with NMD, and between genders (2 factor multivariate ANCOVA, respectively p=0.001 and p=0.007, mean differences = 2.61MJ/day and 2.44MJ/day). Similarly, for ACTEE and ACTEE as a percentage of TEE (2 factor univariate and multivariate ANCOVA, p=0.001 and p=0.05, mean differences = 2.19MJ/day and 1.64MJ/day and p<0.001 and p=0.0001, mean differences = 15% and 9%).  
2. Incidental discriminative validity was demonstrated by extreme group differences between genders and compared to healthy controls in diary reported exercise duration, energy expenditure and intensity (light versus moderate) between healthy controls and NMD (2 factor multivariate ANCOVA, p=0.02, p<0.03, mean differences = 20min, 0.5 MJ/day and 2.0 PAL and 55 mins, 1.26 MJ/day and 1.3 PAL respectively)  
3. Incidental discriminative validity was demonstrated by extreme group differences in active minutes and heart rate between genders and between healthy controls and NMD (2 factor multivariate ANCOVA, respectively p=0.001 and p=0.002, mean difference = 132 min/day and 122/min/day less, 11-12bpm). | FAIR          |
| Andersen et al., 2017        | Randomised, controlled, crossover trial comparing 8 weeks of high intensity interval training with usual care and comparison to healthy controls [29] | 1. IPAQ (International Physical Activity Questionnaire) Accelerometer:  
2. Pedometer (Omron Walking Style Pro Pedometer HJ-720FT-E2) | Incidental discriminative validity was demonstrated by extreme group differences in IPAQ (MET hours per week) between people with FSHD 1 and healthy controls (equal variance test and 2 tailed t-test or Mann–Whitney rank-sum test, p<0.05, mean difference in score 13)  
2. Incidental discriminative validity was demonstrated by extreme group differences in Ommron step distance (km per day) between people with FSHD 1 and healthy controls (equal variance test and 2 tailed t-test or Mann–Whitney rank-sum test, p<0.05, mean difference in score 2.4) | POOR          |
| Barfield et al., 2016        | Observational descriptive cross-sectional study of exercise intensity during power wheelchair soccer in 30 people with mobility impairments [60] | 1. Indirect calorimetry (portable gas analyser: K4 b2c)  
2. Indirect calorimetry (Oxycom Mobiled) | Incidental Reliability  
1. and 2. Incidental inter-rater reliability was demonstrated by no significant difference in METS between gas analysers (K4 b2c versus Oxycom Mobiled) (independent group t-tests, p<0.05)  
Incidental Construct validity  
1. and 2. Incidental discriminative validity was demonstrated by extreme group difference in METS between rest and game play exertion (repeated measures t-test, p<0.01, mean difference = 0.46METS) | POOR          |
| Author                        | Study Description                                                                 | Measure(s)                                                                 | Measurement properties                                                                 | COSMIN rating |
|------------------------------|------------------------------------------------------------------------------------|----------------------------------------------------------------------------|----------------------------------------------------------------------------------------|---------------|
| Andersen et al., 2013        | Observational analytic cohort study of response to high intensity aerobic exercise and comparison to sedentary controls [64] | 1. Indirect calorimetry (Online gas analyser: Quark b2; Cosmed) 2. Direct observation of training with Heartrate monitoring via exercise bike telemetry (Monark 939E) and Borg RPE | Incidental Construct validity 1. and 2. Incidental discriminative validity was demonstrated by extreme group differences in activity intensity during continuous (65% and 75% maximal oxidative capacity (VO2max)) or interval (85% and 95% VO2max) cycling, measured by: Indirect calorimetry (mean VO2 ml/min/kg): significant differences between healthy controls and people with LGMD and BMD (unpaired student t-test), p<0.05, mean differences = 11-16 and 14-18 respectively. There was no significant difference between FSHD and healthy controls. Heartrate (bpm), for which there were significant differences between people with LGMD and healthy controls (unpaired student t-test), p<0.05, mean differences = 29-38. There were no significant differences between healthy controls and people with BMD and FSHD. | POOR          |
| Kierkegaard et al., 2011a    | Observational descriptive cross-sectional study of functioning and disability [38] | 1. FAI (Frenchay Activities Index) 2. PAGS (Physical Activity Grading Scale) | Incidental Construct validity 1. Incidental discriminative validity was demonstrated by extreme group differences in FAI score between participants with mild and severe impairment (Mann-Whitney U test, p<0.001, mean difference = 9) 2. Incidental discriminative validity was demonstrated by extreme group differences in PAGS rating between participants with mild and severe impairment (Mann-Whitney U test, p=0.001, mean difference N/R) | POOR          |
| Van der Kooi et al., 2007    | Randomised controlled, crossover trial investigating the effect of strength training and albuteral on pain and fatigue [65] | DOA (Daily Observation of Activity) | Incidental Construct validity | Incidental discriminative validity was not demonstrated, there were no extreme group differences in DOA score between participants who reported pain/severe fatigue and those who did not (t-test, p=0.3 and p=0.33) Incidental convergent validity was not demonstrated there was no correlation between DOA score and pain/fatigue measured using Daily Observation of Pain/Fatigue (Pearson’s, r=0.06, p=0.66 and r = -0.05, p = 0.73 respectively). | POOR          |
| Wiles et al., 2006           | Observational analytic cohort study of falls and associated risk factors and comparison to healthy controls [70] | StepWatch™ (Ankle step activity monitor (SAM, biaxial, Cymatech, USA) | Incidental Construct validity | Incidental discriminative validity was demonstrated by extreme group differences between healthy people and people with DM (mean differences: daily step count = 2876, p<0.001, peak 30-min step rate = 19.8, p<0.001, sustained 60-min step rate = 11.8, p=0.002, time inactive = 8.4%, p=0.01) Incidental convergent validity was not demonstrated for mean daily step count over 7 days with strength (Spearman’s rank correlation not significant, r=0.6, p=0.06). | POOR          |
| Dawes et al., 2006           | A pilot randomised, controlled trial of a home-based exercise programme in 20 adults with NMD [41] | 1. PASE (Physical Activity Scale for the Elderly) 2. Pedometer (unspecified) | Incidental Responsiveness 1 and 2. Incidental specificity of PASE and pedometer counts was demonstrated by no significant changes in either activity measure after the intervention (Wilcoxon, p<0.05, mean difference PASE = -11.64 (SD 38.31) and step count = -1485 (SD 2681)) | POOR          |
| Aitkens et al., 2005         | Observational analytic cohort study of risk factors for cardiovascular disease and diabetes in 11 ambulatory adults with NMD and comparison to healthy controls [54] | 1. Diary (3-day activity and non-wear log book or Dictaphone logging with energy expenditure by factorial method) 2. TEE and PAL (established by indirect calorimetry (2900 Metabolic System) with heartrate equivalence 3. Polar XL vantage | Incidental Construct validity 1. Incidental discriminative validity was demonstrated by extreme group differences in self-reported activity minutes of exercise between healthy controls and adults with NMD at baseline and follow up (Repeated measures ANOVA, p<0.05, mean difference = 34mins/day at baseline and 20mins/day at follow up) 2. and 3. Incidental discriminative validity was demonstrated by extreme group differences in indirect calorimetry derived PAL between healthy controls and adults with NMD at baseline and follow up (Repeated measures ANOVA, p<0.027, mean difference = 0.5 at baseline and 0.3 at follow up). Incidental discriminative validity was not significant extreme group differences for TEE alone (p<0.05). 3. Incidental discriminative validity was demonstrated by extreme group differences in heartrate derived active minutes between healthy controls and adults with NMD at baseline and follow up (Repeated measures ANOVA, p=0.037, mean difference (less for adults with NMD) = 20mins/day at baseline and 160mins/day at follow up) | POOR          |
| Author          | Study Description                                                                                                                                                                                                 | Measure(s)                                                                                                                                                                                                 | Measurement properties                                                                                                                                                                                                 | COSMIN rating |
|-----------------|---------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|----------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|---------------|
| Kilmer et al., 2005 | Observational analytic cohort study testing the efficacy of a home-based activity and diet intervention in 20 people with slowly progressive NMD [53]                                                            | 1. TEE and PAL (established by HR equivalence with indirect calorimetry (2900 Metabolic System) 2. HR (Polar XL vantage) 3. Pedometer (Yamax Digi-Walker)                                                                 | Incidental Construct validity 1. and 2. Incidental discriminative validity was not demonstrated by extreme group differences in TEE and PAL pre and 6 months post intervention (1 way repeated-measures ANOVA, p>0.05). 3. Incidental discriminative validity was demonstrated by extreme group differences in pedometer daily step count pre and post intervention (1 way repeated-measures ANOVA, p=0.001, mean difference 1,250 steps/day). 3. Incidental convergent validity was not demonstrated for pedometer daily step counts, there was no correlation with fitness measured (submaximal heart rate and VO2 testing) at 3 walking speeds (Pearson’s correlation, p>0.05) | POOR          |

Abbreviations: ICC – intraclass correlation coefficient, NMD – neuromuscular diseases; DMD – Duchenne’s Muscular Dystrophy; FSHD – Facioscapulohumeral Dystrophy; LGMD – limb girdle muscular dystrophy; BMD – Becker’s muscular dystrophy; DM – Myotonic Dystrophy (Dystrophia Myotonica); ZCM – zero crossing mode; PIM – proportional integration mode; SAM – step activity monitor; REE – resting energy expenditure; TEE – total energy expenditure; PAL – physical activity level (index=TEE/REE); ACTEE – Active energy expenditure (TEE minus REE); RPE – rating of perceived exertion; HR – heart rate; bpm – beats per minute; VO2 max – maximal oxidative capacity; METs – Metabolic equivalents; CBT – cognitive behavioural therapy; AET – aerobic exercise training; 6MWD – 6-minute walk distance; ANOVA – Analysis of Variance; ANCOVA – Analysis or covariance; N/R – not reported.

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