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Correlations of Disease Severity Outcome Measures in Inclusion Body Myositis

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Abstract

This study aimed to evaluate the correlation between various outcome measures used to assess disease severity and progression in inclusion body myositis (IBM) clinical trials. A cross-sectional study, involving 51 IBM patients meeting the European Neuromuscular Center (ENMC) 2011 criteria for clinically defined or probable IBM, was completed at the University of California, Irvine. Clinical details, demographic data, and functional data including timed get up (TGU), manual muscle testing, hand grip, pinch dynamometry, as well as IBM functional rating scale (IBMFRS), modified Rankin score, forced vital capacity (FVC), and modified ocular bulbar facial respiratory scale (mOBFRS) were collected on all patients. Descriptive statistics and Pearson's r correlation were performed to analyze the data. Age of the patient did not correlate with any of the outcome measures. Greater severity of IBMFRS scores correlated with longer disease duration as well as greater severity for FVC, strength outcomes, TGU, modified Rankin, and mOBFRS. Additionally, TGU strongly correlated with muscle strength measurements, modified Rankin, and mOBFRS. mOBFRS moderately correlated with IBMFRS, muscle strength, FVC, TGU and modified Rankin score. We demonstrate moderate to strong correlations among the disease severity outcome measures in this study.

Keywords

inclusion body myositis; outcome measures; clinical trials; IBMFRS

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1. INTRODUCTION

Inclusion body myositis (IBM) is an idiopathic muscle disease of insidious onset which afflicts patients over the age of 45, causing slowly progressive asymmetric muscle weakness preferentially affecting the quadriceps and finger flexors, resulting in severe disability and morbidity.[1] While the precise pathophysiologic mechanism has not been elucidated, IBM is often characterized as both an inflammatory and degenerative muscle disease, with the muscle histopathology showing features divided into two categories: 1) inflammatory pathology of endomysial cellularity, focal invasion and upregulation of immune markers and 2) myodegenerative pathology with protein aggregates, rimmed vacuoles and mitochondrial features.[2,3] Despite the pronounced inflammatory features found on muscle biopsies of IBM patients, several standard immunosuppressive and immunomodulatory agents have failed to show sustained efficacy in IBM; thus far, no effective pharmacological therapy has been identified.[4] Improved insight into the pathomechanism of this complex muscle disorder, may help identify a potential therapeutic target.

Success in drug development is often dependent on demonstrating a clinically meaningful change for the patient. However, the optimal measure of disease severity and disease progression for a clinical trial endpoint is not known for IBM. There is only limited data on the rates of progression of outcome measures within placebo arms of clinical trials, and the influence of factors such as age-related morbidities, the presence of anti-NT5c1A antibodies, and influence of immunological perturbations seen in IBM on disease severity and disease progression is not clear. The IBM functional rating scale (IBMFRS) is an increasingly commonly used outcome measure in IBM placebo-controlled clinical trials [5] and appears to correlate well with measures of muscle strength.[6–10] While several additional outcome measures are used in IBM clinical trials to evaluate disease progression, there is limited data on how well other measures such as timed get up, forced vital capacity, pinch dynamometry, and swallow function correlate. Thus, in this study we aimed to further understand how the currently used outcome measures in IBM clinical trials correlate with one another.

2. MATERIALS AND METHODS

2.1 Patients

The data presented here was collected as part of a cross-sectional study (KILL-IBM Study), involving 51 IBM patients meeting the European Neuromuscular Center (ENMC) 2011 criteria for clinically defined or probable IBM, conducted at a single site (University of California, Irvine). Data on the immunophenotype in these patients has been recently published.[11] The study was approved by The Institutional Review Board at University of California, Irvine (HS# 2019-5134). Informed patient consent was obtained according to the Declaration of Helsinki and all subjects additionally provided Health Insurance Portability and Accountability Act (HIPAA) authorization. All patients were seen at a specific study visit, during which the same investigator and evaluator performed all the assessments. Clinical details and demographic data were abstracted from the medical records and re-verified with the subjects upon direct questioning (including age of onset and disease duration).

2.2 Disease severity outcome measures

Manual muscle testing, timed get up, hand grip and pinch dynamometry, IBM functional rating scale, modified Rankin score, and forced vital capacity were collected on all subjects.

Motor testing: A single investigator/neurologist (JC) performed all motor testing. Standardized manual muscle testing (MMT) was performed on the following 12 muscle groups: bilateral shoulder abductors, wrist flexors, finger flexors, hip flexors, knee extensors and ankle dorsiflexors. Muscle strength was graded using the Medical Research Council (MRC) score. The MRC scores were then converted to a Kendall scale (0-10 point scale) and the MRC scores summated to a MMT12 score, with the maximum total score of 120.[12]

The timed get up test (TGU) was administered as a timed test asking the subject to arise from a sitting position in a standardized armchair to a standing position. The test score was based on the time it took (in seconds) for the subject to perform the task. If needed by the subject, use of the armrest was allowed to perform the timed get up test; however, for the purpose of analyses, any individual unable to get up from a chair, even with the use of arm rests, was scored at 20 seconds (the maximal score).

Handgrip strength was measured using a JAMAR grip dynamometer (Sammons Preston Rolyan, Chicago, IL) and pinch strength was measured with the thumb over index finger using a JAMAR pinch dynamometer (Sammons Preston Rolyan, Chicago, IL). Additionally, we also compared the performance of a tip pinch strength measurement using the Piezo pinch meter [piezoelectric FlexiForce A201-25 sensor (Tekscan, Inc. South Boston, MA)]. This particular pinch meter uses a sensing area of 9.53 mm and thickness of 0.203 mm to convert pressure to electric charge. The Piezo pinch meter is a thin 0.203 mm plate that may allow for more sensitive reading of finger flexor strength in patients with the lower end of strength since the Piezo meter is a thinner plate in contrast to the thicker plate of the JAMAR pinch. The electric charge was then read out through a FlexiForce QuickStart Board (Tekscan, Inc. South Boston, MA) programmed to express the pressure in 0-11 kilograms. Pinch was measured with the elbow at a right angle and forearm in neutral position.

IBMFRS and modified Rankin Scale: The same neurologist administered the IBMFRS in all patients. The IBMFRS is a validated tool used in both interventional and observational clinical trials of IBM to monitor disease progression and has a maximum score of 40 which equals normal function.[6] The modified Rankin Scale (mRS) was assessed to measure the degree of disability, (0= no symptoms, 5= severe disability).

Pulmonary Function Testing: Spirometry (forced vital capacity, FVC) was performed on all subjects by a single trained certified clinical coordinator (VL) using a Microloop Spirometer (Vyaire, Yorba Linda, CA). A facemask was used for subjects with significant facial weakness.

Bulbar Evaluation: A speech therapist (NA) blinded to symptoms of bulbar dysfunction, administered the modified oral bulbar facial respiratory scale (mOBFRS) on all subjects. The mOBFRS is a tool assessing bulbar, facial, and respiratory dysfunction, validated in

myasthenia gravis with possible scores ranging from 0 (no dysfunction) to a maximum score of 17 (severe dysfunction).[13]

2.3 Statistical Analysis

Descriptive statistics were used to characterize the baseline demographic data. Statistical analyses were performed by TM and AP. To measure the strength of association between the IBM-related outcomes with IBMFRS, we constructed a matrix of Pearson's Rank correlation coefficients (r) estimated between each pair of outcomes. All statistical analyses were conducted using PRISM Graphpad v8.3 (San Diego, CA) and R v4.0.1 (Figure 1 and Figure 2). For the purpose of analyses, correlation measures were defined as "poor or no correlation" for r=0-0.30, "fair correlation" for r = 0.30-0.50, "moderately strong correlation" for r=0.60-0.80, and "very strong correlation" for r=0.80-1.0 (r expressed in absolute values).[14]

3. RESULTS

Fifty-one patients with clinically defined or probable IBM (ENMC 2011 criteria) were enrolled and completed all assessments in the study. Baseline demographics are summarized in Table 1. The mean age of the patients was 65.7 years and mean disease duration was 10.8 years, with a majority being male (71%) and seropositive for the NT5c1A antibody (69%). Disease characteristics of the patients included: mean IBMFRS of 26, mean FVC percent predicted of 72%, and a mean modified Rankin score of 2.5.

Patient age and CK did not strongly correlate with any of the outcome measures (Pearson's r ranging between −0.36 to 0.38) (Figure 2). While disease duration showed fair correlation with greater disease severity based on IBMFRS (r=−0.40) (Figure 1), disease duration did not correlate with other outcome measures (r ranging between −0.36 to 0.30) (Figure 2). Of all the outcome measures, IBMFRS showed strongest correlations to modified Rankin score (r=−0.81), MMT 12 (r=0.80), and timed get up (r=−0.78). Modest correlations of IBMFRS were also seen with mOBFRS (r=−0.62), FVC (r=0.50) and grip/pinch outcome measures [handgrip, JAMAR pinch and Piezo pinch on either side (r ranging between 0.41 to 0.66)]. Timed get up also strongly correlated with MMT12 (r=−0.76). Modified Rankin score negatively correlated with several additional outcome measures including: FVC, MMT12, right handgrip and JAMAR pinch on both sides ($r \leq -0.52$). In addition to the correlation with IBMFRS, the mOBFRS showed moderate correlations with several outcome measures: timed get up (r=0.62), modified Rankin score (r=0.59), FVC (r=−0.57) and MMT12 (r=−0.52). Additionally, grip dynamometry strongly correlated to the JAMAR pinch for each side (r= 0.77 on the right, r=0.80 on the left) and the JAMAR pinch showed moderate to strong correlations to the Piezo pinch for each respective side ($r \geq 0.61$). Several additional outcome measures showed modest correlations (Figure 2).

4. DISCUSSION

Inclusion body myositis is a disabling disease with high rates of morbidity and disability, yet there are no effective therapies to halt or even slow the rate of progression of the disease. Several recent clinical trials in IBM that have been completed[5,15], (NCT02753530),

or are underway (NCT04659031, NCT04789070) use similar outcome measures as the ones we examined, and these outcome measures have been historically accepted as valid measures to evaluate for drug efficacy. While frequently used in IBM trials, only a select few of the common outcome measures (primarily IBMFRS and muscle strength) have been formally compared to one another in observational studies. The earliest of these studies, Jackson and colleagues in 2008 described good correlations of the IBMFRS with MMT (r=0.73), maximal voluntary isometric contraction testing (MVICT) (r=0.60), and hand grip dynamometry (r=0.56).[6] In a study of 22 IBM patients, Allenbach at el. demonstrated a strong correlation between muscle strength by dynamometry (whole body composite score) and IBMFRS (r =0.883).[7] Similarly, Hogrel et al. reported a strong correlation between IBMFRS and composite QMT score (r= 0.926) in 13 IBM subjects,[9] while Cortese et al. found a moderate correlation between IBMFRS and MMT (r=0.6) in 51 IBM subjects.[8] The largest study to date (up to 181 IBM patients) by Sangha and colleagues, found strong correlations between IBMFRS and MMT (r=0.70) and moderate correlations between IBMFRS and MVICT (r=0.54).[10] Additionally, similar to our study, they noted moderate correlation of the IBMFRS to disease duration (r=−0.47).[10] The IBMFRS lower limb components (IBMFRS-LL) has also shown good correlation to the mean fat fraction of thigh muscle MRI (r=−0.64).[16] Only a few natural history studies have evaluated the correlation of other functional outcome measures to one another including: whole body composite muscle strength and 6MWT (r=0.741),[7] quantitative muscle testing (QMT) and MMT of the quadriceps (r=0.8),[8] knee extension strength and 6MWT (r=0.858),[9] and MVICT and MMT (r=0.60).[10] However, the optimal outcomes measures to study therapeutic effect or disease progression are yet to be determined.[17]

While the IBMFRS is a reliable and validated measure of disease severity that has shown significant correlations with muscle strength, the novel aspects of our study include identifying correlations between several other IBM clinical outcome measures such as FVC and other functional measures: timed get up, modified Rankin and mOBFRS for swallow dysfunction, especially given that respiratory insufficiency and dysphagia are leading causes of morbidity in IBM. Our study showed that significant correlations were seen with several of the commonly used IBM outcome measures with one another. Furthermore, there appears to be good concordance with severity of the disease based on the IBMFRS score. Specifically, the patients in this study who had less severe disease (noted by a higher IBMFRS score), also showed strong correlations to measurements of FVC, strength scores and functional assessments, demonstrated by correlations to higher FVC (less respiratory insufficiency), higher strength scores (noted on MMT 12, grip and Jamar pinch), less time to get up from a chair, less disability (modified Rankin score), and less oral, bulbar, facial weakness (mOBFRS). Additionally, strong correlations were seen with handgrip and pinch strength, an important finding given that finger flexion weakness is often a significant disabling feature of IBM. While standard dynamometers for handgrip and pinch strength are not sensitive enough to detect changes in strength in severely weak patients resulting in a floor effect, we tested a new device called the Piezo pinch meter designed with a thin, more sensitive plate and sensor, and an ability to measure tip pinch strength of even severely weak patients that were unrecordable with the standard pinch dynamometer, addressing the floor effect in some patients with severe finger flexor weakness. The findings in this

study indicate that several of these clinical measures correlate very well with another and are well able to evaluate disease progression. Even though the IBMFRS does not have extensive coverage of bulbar function, IBMFRS correlated well with mOBFRS. These findings support that IBMFRS tracks disease severity and should be able to track disease progression well.

The high correlation of IBMFRS with other outcome measures supports its validity as a measure of disease severity. The rate of decline for several IBM outcome measures have been described by some recent studies. Sangha and colleagues published a longitudinal observational study in up to 181 IBM patients for up to 7.3 years evaluating manual muscle testing, quantitative muscle testing and the IBMFRS score and noted an average decline of 3.7%, 3.8%, and 6.3% (respectively) per year characterizing the rate of disease progression.[10] Even though this is a large study examining a large number of patients, the results presented in this paper reflect data from three separate studies done over different time points at three different UK sites, with only 130 of the 181 patients having two or more visits, measurements done through different techniques, and the median follow up of only 1.3 years. Oldroyd et al. collected longitudinal data on 75 IBM patients (348-person-years follow up) and described the greatest loss of annual strength in pinch (−10%), followed by knee extension (−4%) with a minimal annual decline seen in IBMFRS (−1%).[18] These findings are limited by the fact that this study was a “real-world” experience with data collected as part of their clinical care at non-standard intervals, rather than a scientifically designed prospective study. The variability in the annual decline in the IBMFRS from these two studies, as well as the lack of significant change in the annual score of IBMFRS by Oldroyd et al. is an important finding since many therapeutic interventional studies in IBM aim to complete a clinical trial within 1-2 years and may not be able to successfully depict the impact of drug efficacy if using the IBMFRS alone.

It is notable that in a heterogenous population of patients, such as can be seen in IBM, significant correlations can be found between any of the outcome measures. In our study, we demonstrate that other outcome measures correlate well with IBMFRS and may potentially be useful adjunctive measures that can be used in IBM trials to detect drug efficacy, but at this point we don’t have sufficient longitudinal data to understand if they are interchangeable. Given the clinical heterogeneity of the disease within IBM patients, with some patients having more severe leg involvement and in others predominant grip or even bulbar involvement, all the different outcome measures tested in this study such as timed get up, pinch, FVC, and mOBFRS may reveal distinct facets of the disease. The data from this study may give insight into which outcome measures play an important role in building stronger clinical trials especially given the strong correlations that we saw with IBMFRS, MMT12, and timed get up. Limitations of this study include the use of a cross-sectional design, which is unable to measure disease progression within individuals and unable to answer if these outcome measures are sensitive to change in a similar way or are dependent on the clinical status of patients. The above quoted longitudinal studies, also highlight the need for a well-designed prospective natural history that would evaluate these outcome measures at regular intervals, through highly trained evaluators and in a standardized fashion. One such prospective natural history study has been funded by the National Institute for Arthritis, Musculoskeletal, and Skin Diseases (NIAMS) and has

recently started (NCT05046821). Through this NIAMS study and similar prospective studies we should finally be able to learn the rate of changes in these outcome measures (and thus disease progression) in IBM which will be helpful to design appropriate pharmacological intervention studies.

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Declaration of Interests:

S.A.G. is an inventor of intellectual property related to myositis diagnostics and therapeutics, owned and managed by Brigham and Women's Hospital (BWH); he is a founder of Abcuro, Inc. Partners HealthCare, the owner of BWH, and S.A.G. have financial interests in Abcuro. The financial interests were reviewed and managed in accordance with the conflict-of-interest policies of Partners HealthCare. A.M.P. was supported by the National Center for Research Resources and the National Center for Advancing Translational Sciences, National Institutes of Health, through Grant UL1 TR001414. S.A.V. was supported by PHS grants R01NS120060, R21NS114918, and KL2TR001416. T.M. was supported by PHS grants R01AR078340, UL1TR001414 and U24NS107210. The content is solely the responsibility of the authors and does not necessarily represent the official views of the NIH. The rest of the authors report no relevant disclosures.

ABBREVIATIONS

6MWT	6-minute walk test
CK	Creatine Kinase
FVC	Forced Vital Capacity
IBM	Inclusion body myositis
IBMFRS	Inclusion body myositis functional rating scale
MVICT	Maximal voluntary isometric contraction testing
MMT	Manual Muscle Testing
MRC	Medical Research Council
mOBFRS	Modified oral bulbar facial respiratory scale
QMT	Quantitative muscle testing
TGU	Timed get up

REFERENCES

[1]. Weihl CC. Sporadic Inclusion Body Myositis and Other Rimmed Vacuolar Myopathies. Continuum (Minneapolis, Minn) 2019;25:1586–1598. 10.1212/CON.0000000000000790. [PubMed: 31794461]

[2]. Weihl CC, Mammen AL. Sporadic inclusion body myositis - a myodegenerative disease or an inflammatory myopathy. Neuropathology and applied neurobiology 2017;43:82–91. 10.1111/nan.12384. [PubMed: 28111778]

[3]. Pestronk A. Acquired immune and inflammatory myopathies: pathologic classification. Current opinion in rheumatology 2011;23:595–604. 10.1097/BOR.0b013e32834bab42. [PubMed: 21934500]

[4]. Schmidt K, Schmidt J. Inclusion body myositis: advancements in diagnosis, pathomechanisms, and treatment. Current opinion in rheumatology 2017;29:632–638. 10.1097/BOR.0000000000000436. [PubMed: 28832349]

[5]. Benveniste O, Hogrel JY, Annoussamy M, Bachasson D, Rigolet A, Servais L. Rapamycin vs. placebo for the treatment of inclusion body myositis: improvement of the 6 min walking distance, a functional scale, the FVC and muscle quantitative MRI [abstract]. Arthritis Rheumatol 2017;69 (Suppl 10), 5L.

[6]. Jackson CE, Barohn RJ, Gronseth G, Pandya S, Herbelin L, Group M. Inclusion body myositis functional rating scale: a reliable and valid measure of disease severity. Muscle & nerve 2008;37:473–476. 10.1002/mus.20958. [PubMed: 18236463]

[7]. Allenbach Y, Benveniste O, Decostre V, Canal A, Eymard B, Herson S, et al. Quadriceps strength is a sensitive marker of disease progression in sporadic inclusion body myositis. Neuromuscular disorders : NMD 2012;22:980–986. 10.1016/j.nmd.2012.05.004. [PubMed: 22738680]

[8]. Cortese A, Machado P, Morrow J, Dewar L, Hiscock A, Miller A, et al. Longitudinal observational study of sporadic inclusion body myositis: implications for clinical trials. Neuromuscular disorders : NMD 2013;23:404–412. 10.1016/j.nmd.2013.02.010. [PubMed: 23489664]

[9]. Hogrel JYY, Allenbach Y, Canal A, Leroux G, Ollivier G, Mariampillai K, et al. Four-year longitudinal study of clinical and functional endpoints in sporadic inclusion body myositis: implications for therapeutic trials. Neuromuscular disorders : NMD 2014;24:604–610. 10.1016/j.nmd.2014.04.009. [PubMed: 24857365]

[10]. Sangha G, Yao B, Lunn D, Skorupinska I, Germain L, Kozyra D, et al. Longitudinal observational study investigating outcome measures for clinical trials in inclusion body myositis. Journal of neurology, neurosurgery, and psychiatry 2021. Apr 13:jnnp-2020-325141. 10.1136/jnnp-2020-325141.

[11]. Goyal NA, Coulis G, Duarte J, Farahat PK, Mannaa AH, Cauchii J, et al. Immunophenotyping of Inclusion Body Myositis Blood T and NK Cells. Neurology 2022. Mar 29;98(13):e1374–e1383. 10.1212/WNL.0000000000200013. [PubMed: 35131904]

[12]. Kendall FP ME, Provance PG. Muscles: Testing and function. 4 ed Baltimore: Williams and Wilkins; 1993.

[13]. Farrugia ME, Harle HD, Carmichael C, Burns TM. The Oculobulbar Facial Respiratory score is a tool to assess bulbar function in myasthenia gravis patients. Muscle & nerve 2011;43:329–334. 10.1002/mus.21880. [PubMed: 21305562]

[14]. Chan YH. Biostatistics 104: correlational analysis. Singapore medical journal 2003;44:614–619. [PubMed: 14770254]

[15]. Hanna MG, Badrising UA, Benveniste O, Lloyd TE, Needham M, Chinoy H, et al. Safety and efficacy of intravenous bimagrumab in inclusion body myositis (RESILIENT): a randomised, double-blind, placebo-controlled phase 2b trial. The Lancet Neurology 2019;18:834–844. 10.1016/S1474-4422(19)30200-5. [PubMed: 31397289]

[16]. Morrow JM, Sinclair CD, Fischmann A, Machado PM, Reilly MM, Yousry TA, et al. MRI biomarker assessment of neuromuscular disease progression: a prospective observational cohort study. The Lancet Neurology 2016;15:65–77. 10.1016/S1474-4422(15)00242-2. [PubMed: 26549782]

[17]. Hogrel JYY. Still seeking the holy grail of outcome measures in inclusion body myositis. Journal of neurology, neurosurgery, and psychiatry 2021. Apr 13:jnnp-2021-326460. 10.1136/jnnp-2021-326460.

[18]. Oldroyd AGS, Lilleker JB, Williams J, Chinoy H, Miller JAL. Long-term strength and functional status in inclusion body myositis and identification of trajectory subgroups. Muscle & nerve 2020;62:76–82. 10.1002/mus.26859. [PubMed: 32134516]

HIGHLIGHTS

- IBMFRS strongly correlated to Rankin, MMT12, and timed get up (|r| =0.78 to 0.81).
- IBMFRS modestly correlated to mOBFRS, FVC and grip/pinch (r = 0.41 to 0.66).
- Timed get up strongly correlated with MMT12 (r=−0.76).
- Several IBM outcome measures showed significant correlations with one another.

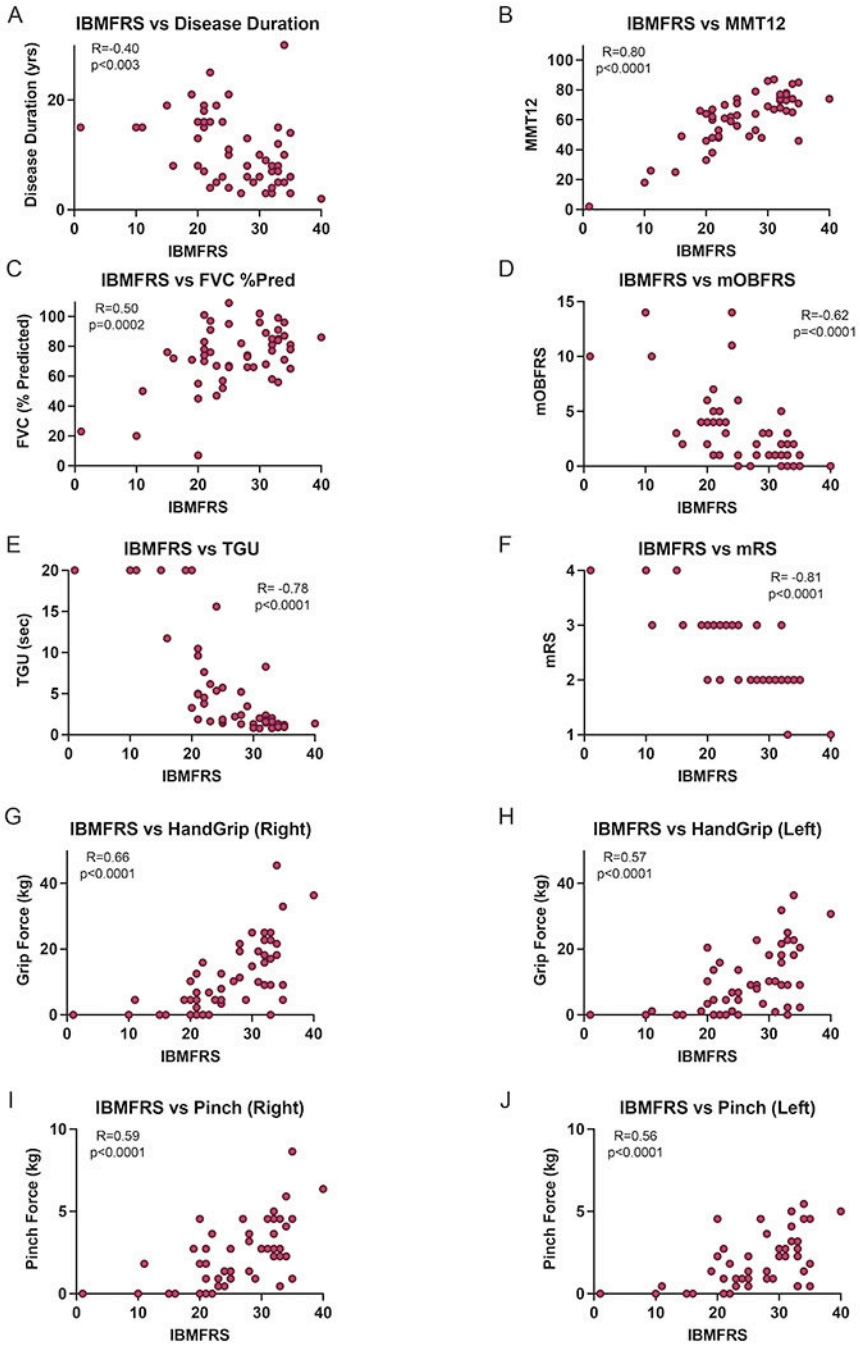

Figure 1: Correlation of IBM disease outcome measures with the IBM Functional Rating Scale (IBMFRS).

Correlations of 10 outcome measures to IBMFRS in 51 IBM patients (Pearson's r) (A) disease duration, (B) MMT12, (C) forced vital capacity (FVC, % predicted), (D) modified oral bulbar facial respiratory scale (mOBFRS), (E) timed get up test (TGU), (F) modified Rankin scale (mRS), (G,H) right and left hand grip dynamometry, and (I,J) right and left pinch dynamometry.

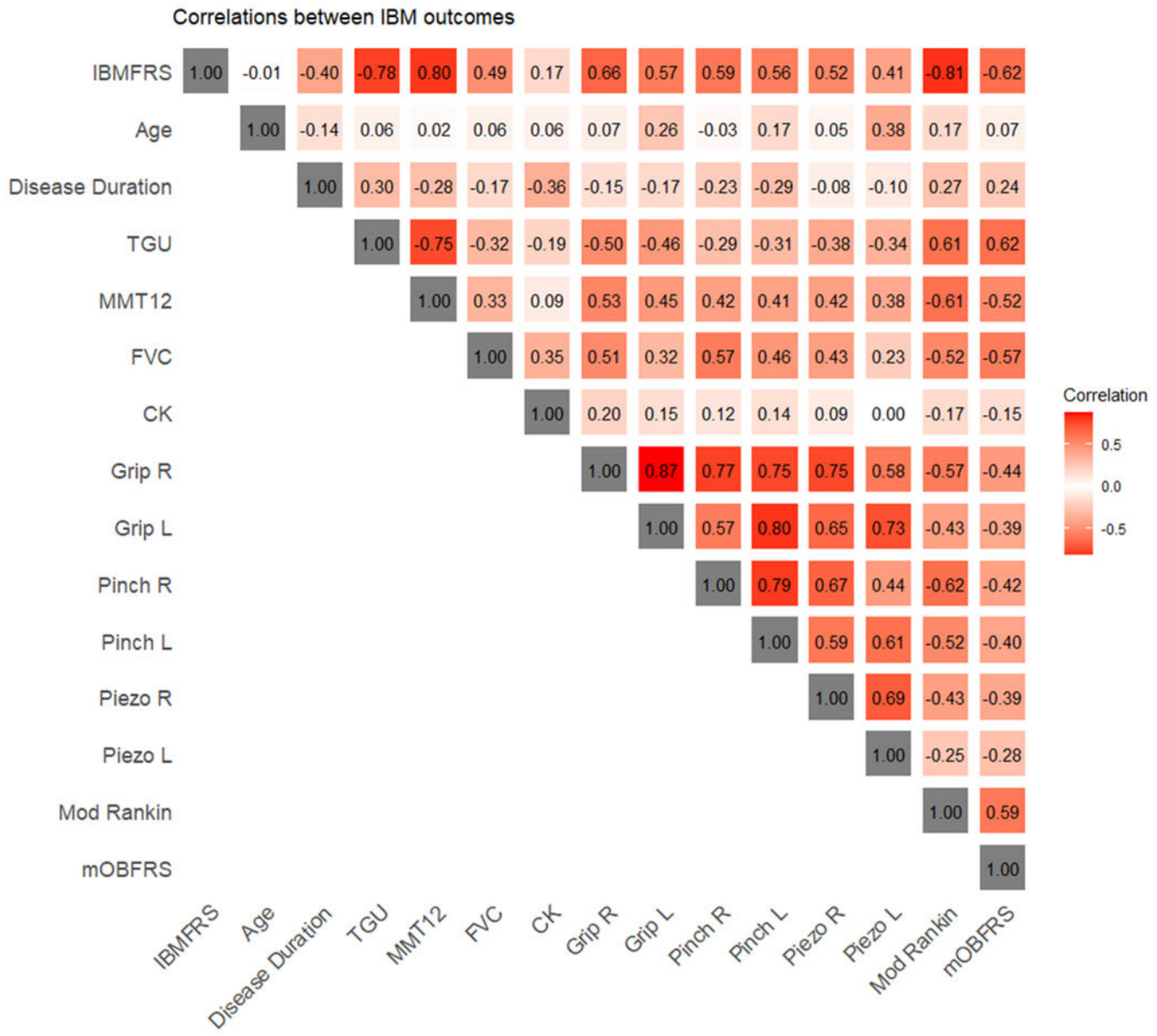

Figure 2: Correlations between IBM Disease Outcome Measures

Correlogram depicting Pearson's r correlations between each pair of outcome measures. The darker highlighted cells indicate stronger Pearson's r correlation coefficients. IBMFRS= Inclusion body myositis functional rating scale; TGU= timed get up test; MMT12= manual muscle testing sum score of 12 muscles; FVC= forced vital capacity; CK= creatine kinase; Mod Rankin= modified Rankin score; mOBFRS= modified oculobulbar facial respiratory scale.

TABLE 1:

Patient demographics and clinical data

Demographics and clinical characteristics of 51 IBM patients are provided.

Variable	IBM Patients Mean (SD)	Median (Ranges)
# of patients (N)	51	
Sex (% female)	29	
Age (years)	65.69 (8.47)	65 (47-84)
Age of Onset (years)	54.84 (7.53)	53 (42-76)
Disease Duration (years)	10.82 (6.37)	9 (2-30)
IBMFRS total score	26.10 (7.55)	27 (1-40)
Modified Rankin Score	2.49 (0.67)	2 (1-4)
FVC % predicted	72.61 (20.75)	74 (7-109)
CK IU/L	443.69 (425.80)	347 (35-2204)
NT5c1A Ab seropositive (%)	69	

IBMFRS=IBM functional rating scale; FVC=forced vital capacity; CK=creatine kinase