

IMACS STUDIES

Over the last decade IMACS has undertaken a number of projects aimed at achieving its goals of improving the lives of children and adults who suffer from myositis by discovering better therapies and understanding the causes of these diseases. Central to these efforts have been a number of studies to develop consensus and standards on the conduct and reporting of adult and juvenile myositis investigations, including outcome measures, clinical trial design and classification of patients with myositis. Listed here is information on the studies that IMACS has reviewed and endorsed. Feel free to contact the Lead Investigators to learn more about or participate in these studies.

Completed IMACS Research Projects

IMACS Project I. Developing and validating core set measures of myositis disease activity and damage

Primary Objectives: Develop and validate new tools to assess disease activity and damage for patients with adult and juvenile dermatomyositis and polymyositis

Start Date: 2000

Completion Date: 2010

Lead Investigators:

- **Lisa Rider, M.D.**
Deputy Chief, Environmental Autoimmunity Group
NIEHS
riderl@mail.nih.gov
- **Frederick W. Miller, M.D., Ph.D.**
Deputy Chief, Clinical Research Branch and Principal Investigator
millerf@mail.nih.gov
- **David A. Isenberg, M.D., F.R.C.P., M.D.**
ARC Diamond Jubilee Professor of Rheumatology
d.isenberg@ucl.ac.uk

Publications:

Publications:

Miller FW, Rider LG, Chung YL, Cooper R, Danko K, Farewell V, Lundberg I, Morrison C, Oakley L, Oakley I, Pilkington C, Vencovsky J, Vincent K, Scott DL, Isenberg DA, Int Myositis Outcome Assessment C.

Proposed preliminary core set measures for disease outcome assessment in adult and juvenile idiopathic inflammatory myopathies. *Rheumatology*. 2001; 40(11):1262-1273. [[Pub Med](#)]

Huber AM et al. Validation and clinical significance of the childhood myositis assessment scale for assessment of muscle function in the juvenile idiopathic inflammatory myopathies. *Arthritis Rheum*. 2004; 50(5):1595-603. [[Abstract](#)]

Isenberg DA et al. International Consensus Outcome Measures for patients with idiopathic inflammatory myopathies: Development and initial validation of myositis activity and damage indices in patients with adult onset disease. *Rheumatology, Oxford*, 2004; 43(1): 49-54. [[Abstract](#)]

Rennebohm RM et al. Normal scores for nine maneuvers of the Childhood Myositis Assessment Scale. *Arthritis Rheum*. 2004; 51(3):365-70. [[Abstract](#)]

Huber AM, Dugan EM, Lachenbruch PA, Feldman BM, Perez MD, Zemel LS, Lindsley CB, Rennebohm RM, Wallace CA, Passo MH, Reed AM, Bowyer SL, Ballinger SH, Miller FW, Rider LG; Juvenile Dermatomyositis Disease Activity Collaborative Study Group. The Cutaneous Assessment Tool (CAT): development and reliability in juvenile idiopathic inflammatory myopathy. *Rheumatology (Oxford)*. 2007 Oct;46(10):1606-11. [[Abstract](#)]

Isenberg DA et al. International Consensus Outcome Measures for patients with idiopathic inflammatory myopathies: Development and initial validation of myositis activity and damage indices in patients with adult onset disease. *Rheumatology, Oxford*, 2004; 43(1): 49-54. [[Abstract](#)]

Huber AH et al. Preliminary validation and clinical meaning of the cutaneous assessment tool (CAT) in juvenile dermatomyositis. *Arthritis Rheum*. 2008; 59:214-221. [[Abstract](#)]

Huber AH et al. Alternative scoring of the cutaneous assessment tool (CAT) in juvenile dermatomyositis: Results using abbreviated formats. *Arthritis Rheum*. 2008; 59:352-356. [[Abstract](#)]

Rider LG et al. Damage extent and predictors in adult and juvenile dermatomyositis and polymyositis using the Myositis Damage Index (MDI). *Arthritis Rheum*. 2009;60:3425-35. [[Abstract](#)]

Lachenbruch PA et al. On determining the effects of therapy on disease damage in non-randomized studies with multiple treatments: a study of juvenile myositis. *Communications in Statistics – Theory and Methods*. 2009;38:3268-81. [[Abstract](#)]

Dugan EM, Huber AM, Miller FW, and Rider LG; Photoessay of the idiopathic inflammatory myopathies. *Dermatology Online Journal*. 2009;15(2):1. [[Abstract](#)] [[Dermatology Online Journal](#)]

Rider LG, Koziol D, Giannini EH, Jain MS, Smith MR, Whitney-Mahoney K, Feldman BM, Wright SJ, Lindsley CB, Pachman LM, Villalba ML, Lovell DJ, Bowyer SL, Plotz PH, Miller FW, Hicks JE; Validation of manual muscle testing and a subset of eight muscles for adult and juvenile idiopathic inflammatory myopathies. *Arthritis Care Res (Hoboken)*. 2010 Apr;62(4):465-72. [[Abstract](#)]

Rider LG, Werth VP, Huber AM, Alexanderson H, Rao AP, Ruperto N, Herbelin L, Barohn R, Isenberg D, Miller FW. Measures of adult and juvenile dermatomyositis, polymyositis, and inclusion body myositis: Physician and Patient/Parent Global Activity, Manual Muscle Testing (MMT), Health Assessment Questionnaire (HAQ)/Childhood Health Assessment Questionnaire (C-HAQ), Childhood Myositis Assessment Scale (CMAS), Myositis Disease Activity Assessment Tool (MDAAT), Disease Activity Score (DAS), Short Form 36 (SF-36), Child Health Questionnaire (CHQ), physician global damage, Myositis

Damage Index (MDI), Quantitative Muscle Testing (QMT), Myositis Functional Index-2 (FI-2), Myositis Activities Profile (MAP), Inclusion Body Myositis Functional Rating Scale (IBMFRS), Cutaneous Dermatomyositis Disease Area and Severity Index (CDASI), Cutaneous Assessment Tool (CAT), Dermatomyositis Skin Severity Index (DSSI), Skindex, and Dermatology Life Quality Index (DLQI). Arthritis Care Res (Hoboken). 2011 Nov;63 Suppl 11:S118-57. doi: 10.1002/acr.20532. Review. [Abstract]

IMACS Project II. Determining clinically meaningful change in core set activity measures and developing preliminary definitions of improvement

Primary Objectives:

1. Determine clinically meaningful change in core set activity measures
2. Develop preliminary definitions of improvement as response criteria for adult and juvenile dermatomyositis and polymyositis

Start Date: 2002

Completion Date: 2004

Lead Investigators:

- **Lisa Rider, M.D.**
Pediatric Rheumatology
NIEHS
riderl@mail.nih.gov
- **Frederick W. Miller, M.D., Ph.D.**
Deputy Chief, Clinical Research Branch and Principal Investigator
millerf@mail.nih.gov
- **Ann Reed**
ann.reed@duke.edu

Publications:

Rider LG, Giannini EH, Brunner HI, Ruperto N, James-Newton L, Reed AM, Lachenbruch PA, Miller FW. International consensus on preliminary definitions of improvement in adult and juvenile myositis. Arthritis Rheum. 2004; 50(7):2281-90. [Abstract] [Full Text]

Rider LG, Giannini EH, Harris-Love M, Joe G, Isenberg D, Pilkington C, Lachenbruch PA, Miller FW; International Myositis Assessment and Clinical Studies Group. Defining Clinical Improvement in Adult and Juvenile Myositis. J Rheumatol. 2003 Mar;30(3):603-17. [Abstract]

IMACS Research Project III. Consensus guidelines for the design and conduct of myositis clinical trials

Primary Objectives:

Develop consensus in the design of myositis clinical trials, including classification criteria for myositis

Start Date: 2002

Completion Date: 2005

Lead Investigators:

Chet Oddis M.D.

cvo5@pitt.edu

- **Lisa Rider, M.D.**

Pediatric Rheumatology

NIEHS

riderl@mail.nih.gov

- **Frederick W. Miller, M.D., Ph.D.**

Deputy Chief, Clinical Research Branch and Principal Investigator

millerf@mail.nih.gov

- **Ann Reed**

ann.reed@duke.edu

Publications:

Oddis CV et al. International consensus guidelines for therapeutic trials in the idiopathic inflammatory myopathies. *Arthritis Rheum.* 2005; 52(9):2607-2615. [[Abstract](#)]

IMACS Project IV. Dyslipidemia in Myositis Survey

Primary Objectives:

To examine the frequency of lipid profile abnormalities in patients with myositis

Start Date: 2009

Completion Date: 2010

Lead Investigators:

- **Christina Charles-Schoeman**

ccharles@mednet.ucla.edu

Publications:

Charles-Schoeman C, Amjadi SS, Paulus HE; International Myositis Assessment and Clinical Studies Group. Treatment of dyslipidemia in idiopathic inflammatory myositis: results of the International Myositis Assessment and Clinical Studies Group survey, Clin Rheumatol. 2012 Aug;31(8):1163-8. [Abstract]

IMACS Project V. Identification of a candidate core-set of fitness and strength tests for patients with childhood or adult idiopathic inflammatory myopathies

Primary Objectives:

To develop consensus on a candidate core-set of fitness and strength tests for children and adults with myositis

Start Date: 2013

Completion Date: 2015

Lead Investigators:

- **Djamilla K.D. van der Stap**
djamillavanderstap@gmail.com
- **Tim Takken Ph.D.**
T.Takken@umcutrecht.nl

Publications:

van der Stap DK, Rider LG, Alexanderson H, Huber AM, Gualano B, Gordon P, van der Net J, Mathiesen P, Johnson LG, Ernste FC, Feldman BM, Houghton KM, Singh-Grewal D, Kutzbach AG, Munters LA, Takken T. Proposal for a Candidate Core Set of Fitness and Strength Tests for Patients with Childhood or Adult Idiopathic Inflammatory Myopathies. J Rheumatol. 2016; 2016 Jan;43(1):169-765 [Abstract]

Ongoing IMACS Research Projects

IMACS Project VI. IMACS Outcomes Data Repository

Primary Objectives:

Develop a repository of databases of myositis natural history studies and therapeutic trials that have all collected the IMACS disease activity and damage core set measures, as well as core demographic and clinical data, for use by myositis researchers.

For further information on the IMACS Outcomes Data Repository, including use of the data that has been deposited, please see: [IMACS Outcomes Repository](#) [Data Use Guidelines](#).

Start Date: 2004

Completion Date: Ongoing

Lead Investigators:

- **Lisa Rider, M.D.**
Pediatric Rheumatology
NIEHS
riderl@mail.nih.gov
- **Frederick W. Miller, M.D., Ph.D.**
Deputy Chief, Clinical Research Branch and Principal Investigator
millerf@mail.nih.gov

Publications:

None

IMACS Project VII. International Myositis Classification Criteria Project

Primary Objectives:

To develop new preliminarily validated classification criteria for adult and juvenile dermatomyositis, adult polymyositis and inclusion body myositis.

For further information on the International Myositis Classification Criteria Project, please see [International Myositis Classification Criteria Project](#).

Start Date: 2004

Completion Date: Ongoing

Lead Investigators:

- **Ingrid Lundberg, M.D., Ph.D.**
Ingrid.Lundberg@medks.ki.se

Publications:

None

IMACS Project VIII. Standards of Treatment for Adults with Myositis and different Phenotypes - STAMP

Primary Objectives:

To define standard approaches to treat myositis phenotypes

Start Date: 2011

Completion Date: Ongoing

Lead Investigators:

Lead Investigators:

- **Lisa Christopher-Stine**
lchrist4@jhmi.edu
- **Hector Chinoy**
Hector.Chinoy@srft.nhs.uk
- **Neil McHugh**
Neil.McHugh@rnhrd.nhs.uk
- **Sarah Tansley**
sarah.tansley@rnhrd.nhs.uk
- **Lyubo Dourmishev**
l_dourmishev@yahoo.com

Publications:

Tansley S, Shaddick G, Christopher-Stine L, Sharp C, Dourmishev L, Maurer B, Chinoy H, McHugh N. [Developing standardised treatment for adults with myositis and different phenotypes: an international survey of current prescribing preferences.](#) Clin Exp Rheumatol. 2016 May 31. [Epub ahead of print] [Abstract]

IMACS Project IX. ACR-EULAR Project to Revise the Definition of Improvement and Major Clinical Response for Adult and Juvenile Dermatomyositis and Adult Polymyositis

Primary Objectives:

To develop new response criteria for adult and juvenile dermatomyositis and polymyositis, including criteria for minimal and major response for use as endpoints in myositis therapeutic trials.

Start Date: 2012

Completion Date: Ongoing

Lead Investigators:

- **Lisa Rider, M.D.**
Pediatric Rheumatology
NIEHS
riderl@mail.nih.gov
- **Nicola Ruperto M.D. MPH**

nicolaruperto@ospedale-gaslini.ge.it

- **Frederick W. Miller, M.D., Ph.D.**

Deputy Chief, Clinical Research Branch and Principal Investigator

millerf@mail.nih.gov

- **Jiri Vencovsky M.D., Dr.Sc.**

venc@revma.cz

- **Rohit Aggarwal, M.D.**

aggarwalr@upmc.edu

Publications:

None

IMACS Project X. Development of an internationally agreed minimal dataset for juvenile dermatomyositis (JDM) for clinical and research use

Primary Objectives:

To develop consensus on minimal elements to include in a JDM dataset to be used for clinical and research purposes

Start Date: 2014

Completion Date: Ongoing

Lead Investigators:

- **Liza McCann**

lizamccann@btinternet.com

Publications:

McCann LJ, Kirkham JJ, Wedderburn LR, Pilkington C, Huber AM, Ravelli A, Appelbe D, Williamson PR, Beresford MW. Development of an internationally agreed minimal dataset for juvenile dermatomyositis (JDM) for clinical and research use. *Trials*. 2015 Jun 12;16:268. [[Abstract](#)]

McCann LJ, Arnold K, Pilkington CA, Huber AM, Ravelli A, Beard L, Beresford MW, Wedderburn LR. Developing a provisional, international Minimal Dataset for Juvenile Dermatomyositis: for use in clinical practice to inform research. *Pediatr Rheumatol Online J*. 2014 Jul 21;12:31. [[Abstract](#)]

IMACS Project XI. Identification of novel biomarkers and clinical or gene signature phenotypes in myositis that predict disease activity, damage, and prognosis

Primary Objectives:

Goals of this project include defining predictive biomarkers of disease course in different myositis subsets.

Start Date: 2015

Completion Date: Ongoing

Lead Investigators:

TBA

Publications:

None

IMACS Project XII. Guidelines for cancer screening and follow-up of myositis phenotypes

Primary Objectives:

Objectives of this project include derivation/definition of consensus guidelines for cancer screening in patients diagnosed with idiopathic inflammatory myopathies.

Start Date: 2015

Completion Date: Ongoing

Lead Investigators:

- **Rohit Aggarwal, M.D.**
aggarwalr@upmc.edu

Publications:

None

IMACS Project XIII. Screening, treatment, and monitoring of myositis- associated interstitial lung disease

Primary Objectives:

Through the development of a combined retrospective/prospective database, the purpose of this project is to devise consensus guidelines for diagnosis, management, and outcomes assessment in myositis-associated ILD.

Start Date: 2015

Completion Date: Ongoing

Lead Investigators:

- **Dana Ascherman, M.D.**
DAscherman@med.miami.edu

Publications:

None

IMACS Project XIV. The role of rituximab and mycophenolate mofetil, without oral steroids, in the treatment of myositis

Primary Objectives:

This purpose of this project is to synthesize retrospective experience involving rituximab and mycophenolate mofetil (MMF) combination therapy in the treatment of myositis subsets (including myositis-associated ILD) as a foundation for future clinical trials using rituximab and MMF in the absence of corticosteroids.

Start Date: 2015

Completion Date: Ongoing

Lead Investigators:

- **David A. Isenberg , M.D., F.R.C.P., M.D.**
ARC Diamond Jubilee Professor of Rheumatology
d.isenberg@ucl.ac.uk

Publications:

None