

A motor function measure scale for neuromuscular diseases. Construction and validation study

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Abstract

A new scale for motor function measurement has been developed for neuromuscular diseases. The validation study included 303 patients, aged 6–62 years. Seventy-two patients had Duchenne muscular dystrophy, 32 Becker muscular dystrophy, 30 limb-girdle muscular dystrophy, 39 facio-scapulo-humeral dystrophy, 29 myotonic dystrophy, 21 congenital myopathy, 10 congenital muscular dystrophy, 35 spinal muscular atrophy and 35 hereditary neuropathy. The scale comprised 32 items, in three dimensions: standing position and transfers, axial and proximal motor function, distal motor function. Agreement coefficients for inter-rater reliability were excellent ($\alpha=0.81-0.94$) for nine items, good ($\alpha=0.61-0.80$) for 20 items and moderate ($\alpha=0.51-0.60$) for three items. High correlations were found between the total score and other scores: Vignos ($r=0.91$) and Brooke ($r=0.85$) grades, Functional Independence Measure ($r=0.91$), the global severity of disability evaluated with visual analog scales by physicians ($r=0.88$) and physiotherapists ($r=0.91$). This scale is reliable, does not require any special equipment and is well-accepted by patients. Its sensitivity to change is being assessed to permit its use in clinical trials of neuromuscular diseases.

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1. Introduction

In neuromuscular diseases, identification of new genotypes and hope for gene therapy have obliged scientists and clinicians to collaborate in order to classify phenotypes more precisely and to link them to specific genetic defects. Evaluation of motor deficit, and in particular measurement of muscle force by muscle testing or instrumental measures, is the most frequently used evaluation [1,2]. However, this muscle testing does not reflect the subject's functional abilities. These depend on the heterogeneity of the muscle defect, the muscular compensations and the limitations of the joints.

Several tests for the measurement of motor function have been proposed: the Functional Motor Scale for spinal

muscular atrophy (SMA) [3]; the ALS score [4], the Tufts Quantitative Neuromuscular Exam [5] and the Amyotrophic Lateral Sclerosis Functional Rating Scale [6] for amyotrophic lateral sclerosis; the Hughes Functional Score [7] for Guillain-Barre syndrome and the Hammersmith Motor Ability Score [8] for Duchenne muscular dystrophy (DMD). Some tests focus on the function of one part of the body: the Zupan Functional Test [9] or the Brooke Upper Extremity Scale [11] for the upper limbs and the Vignos Lower Extremity Scale for the lower limbs [10]; other tests specifically address a single medical question such as the Diagnostic Motor Performance Test [11] for establishing the differential diagnosis between myopathy and neuropathy. Some have not been validated: Timed tasks [1] and others are non-specific such as the Jobben Hand Function Test [12], or have not been adapted or validated for neuromuscular diseases, like the Gross Motor Function Measure which has been validated for cerebral palsy [13]. At the moment, there is no well validated test which is easy to administer and which has been adapted for the objective

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Patricia A. Bober, Sandra L. Corbett



A Motor Function Measure Scale For Neuromuscular Diseases:

Muscular Dystrophy Madhuri Hegde, Arunkanth Ankala, 2012-05-09 With more than 30 different types and subtypes known and many more yet to be classified and characterized muscular dystrophy is a highly heterogeneous group of inherited neuromuscular disorders This book provides a comprehensive overview of the various types of muscular dystrophies genes associated with each subtype disease diagnosis management as well as available treatment options Though each different type and subtype of muscular dystrophy is associated with a different causative gene the majority of them have overlapping clinical presentations making molecular diagnosis inevitable for both disease diagnosis as well as patient management This book discusses the currently available diagnostic approaches that have revolutionized clinical research Pathophysiology of the different muscular dystrophies multifaceted functions of the involved genes as well as efforts towards diagnosis and effective patient management are also discussed Adding value to the book are the included reports on ongoing studies that show a promise for future therapeutic strategies

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Companion to Clinical Neurology William Pryse-Phillips, 2009-06-03 This book is designed for the neurologist who in this day of unusually strict accountability needs to have at hand an authoritative guide to the diagnostic criteria for all conditions that he or she may be faced with in clinical practice While originally conceived as a compendium of diagnostic criteria the author has felt the need to expand the work to include definitions of practically all terms that are used in neurology today Historical elements are also provided including entries of important neurologists and neurosurgeons who have impacted the field The result is an effective representation of the tools of the trade for the neurologist in training and a concise and precise source for the practicing neurologist The second edition was published in 2003 Since then advances in the definition of many neurological conditions have been made all of which have been incorporated in the third edition There has also been a fine tuning of the definitions and diagnostic criteria of many other conditions The author has collated over 1300 articles since the last edition in order to update many of the entries As such the entries will have the most up to date

definition of diseases symptoms diagnostic tests and pearls of wisdom The third edition remains an invaluable guide to the spectrum of neurological practice and with nearly 7 000 references this truly is the bible of neurological terms and conditions

Muscular Dystrophy Madhuri Hegde, Arunkanth Ankala, 2012-05-09 With more than 30 different types and subtypes known and many more yet to be classified and characterized muscular dystrophy is a highly heterogeneous group of inherited neuromuscular disorders This book provides a comprehensive overview of the various types of muscular dystrophies genes associated with each subtype disease diagnosis management as well as available treatment options Though each different type and subtype of muscular dystrophy is associated with a different causative gene the majority of them have overlapping clinical presentations making molecular diagnosis inevitable for both disease diagnosis as well as patient management This book discusses the currently available diagnostic approaches that have revolutionized clinical research Pathophysiology of the different muscular dystrophies multifaceted functions of the involved genes as well as efforts towards diagnosis and effective patient management are also discussed Adding value to the book are the included reports on ongoing studies that show a promise for future therapeutic strategies *Umphred's Neurorehabilitation for the Physical Therapist Assistant* Rolando Lazaro, Darcy Umphred, 2024-06-01 A comprehensive guide to neurological rehabilitation for physical therapist assistants PTAs Umphred's Neurorehabilitation for the Physical Therapist Assistant Third Edition presents contemporary evidence based principles and techniques for examination and intervention for individuals with neurological conditions Umphred's Neurorehabilitation for the Physical Therapist Assistant Third Edition addresses a wide variety of pediatric and adult neurological disorders including spinal cord injury brain injury stroke Parkinson's disease multiple sclerosis amyotrophic lateral sclerosis Guillain Barr syndrome and more Drs Lazaro and Umphred have updated this classic text to reflect current and emerging trends in physical therapy including The role of the PTA in neurocritical care The role of the PTA in management of clients with lifelong impairments and activity limitations Technology in neurorehabilitation Also included is a new chapter on functional neuroanatomy which provides the foundational background for understanding the relationship between the structure and function of the nervous system The Third Edition also features helpful instructor and student resources Included with the text are online supplemental materials for faculty use in the classroom Umphred's Neurorehabilitation for the Physical Therapist Assistant Third Edition is the definitive resource for any PTA faculty student or clinician interested in the physical therapy management of individuals with neurological conditions *Swaiman's Pediatric Neurology E-Book* Kenneth F. Swaiman, Stephen Ashwal, Donna M Ferriero, Nina F Schor, Richard S. Finkel, Andrea L. Gropman, Phillip L. Pearl, 2017-03-16 Since 1975 Dr Kenneth Swaiman's classic text has been the reference of choice for authoritative guidance in pediatric neurology and the 6th Edition continues this tradition of excellence with thorough revisions that bring you fully up to date with all that's new in the field Five new sections 62 new chapters 4 new editors and a reconfigured format make this a comprehensive and clearly written resource for the experienced clinician as well as the

physician in training Nearly 3 000 line drawings photographs tables and boxes highlight the text clarify key concepts and make it easy to find information quickly Spinal Muscular Atrophy Charlotte J Sumner, Sergey Paushkin, Chien-Ping Ko, 2016-10-24 Spinal Muscular Atrophy Disease Mechanisms and Therapy provides the latest information on a condition that is characterized by motoneuron loss and muscle atrophy and is the leading genetic cause of infant mortality Since the identification of the gene responsible for SMA in 1995 there have been important advances in the basic understanding of disease mechanisms and in therapeutic development This book provides a comprehensive accounting of recent advances in basic and clinical research that covers SMA clinical features and standards of care multifaceted aspects of SMN protein functions and SMA disease pathology various animal models and biomarkers as well as current therapeutic development This title is ideal for graduate students postdocs and principal investigators who are already in the SMA field and need to keep updated on recent findings and approaches and for those who are new to or would like to join the field Likewise users will find an excellent source of reading for biotech pharma scientists clinical researchers and practitioners regulators and patients and their advocacy organizations Furthermore this book is a handy reference for researchers and clinicians who may want to apply the research strategies and therapeutic approaches in SMA to other rare diseases Provides comprehensive up to date reviews by leading investigators on diverse topics of SMA including clinical features and patient care SMN genetics and protein functions animal models disease pathology and mechanisms biomarkers current therapeutic development and the role of non profit organizations in therapeutic development Written to bridge multiple disciplines and promote better communications among basic scientists clinical researchers and health care providers on the latest developments in SMA Includes outstanding questions and perspectives for future investigations and key references for additional detailed study

Lifespan Neurorehabilitation Dennis Fell, Karen Y Lunnen, Reva Rauk, 2018-01-02 The neuro rehab text that mirrors how you learn and how you practice Take an evidence based approach to the neurorehabilitation of adult and pediatric patients across the lifespan that reflects the APTA s patient management model and the WHO s International Classification of Function ICF You ll study examination and interventions from the body structure function impairments and functional activity limitations commonly encountered in patients with neurologic disorders Then understanding the disablement process you ll be able to organize the clinical data that leads to therapeutic interventions for specific underlying impairments and functional activity limitations that can then be applied as appropriate anytime they are detected regardless of the medical diagnosis *Case Files in Physical Therapy Pediatrics* Eric S. Pelletier, 2015-11-22 Thirty pediatrics physical therapy cases sharpen students critical thinking skills and prepare them for real world practice This unique review features case studies that help physical therapy students successfully transition from coursework to clinical work Each of the more than thirty cases includes a discussion of the health condition examination evaluation diagnosis plan of care and interventions evidence based practice recommendations and references NPTE style review questions accompany each case reinforcing students

learning These case studies give students practical experience before they actually work with patients and helps build the confidence they need to succeed in real world clinical practice Analysis of the case includes remediation material making the book almost completely self contained Spares instructors from having to create their own cases as is often done

Occupational Therapy and Physical Therapy Patricia A. Bober, Sandra L. Corbett, 2011 **Rehabilitation R & D progress reports 1987** ,1983 *Harrison's Principles of Internal Medicine, Twenty-First Edition (Vol.1 & Vol.2)* Joseph Loscalzo, Anthony S. Fauci, Dennis L. Kasper, Stephen Hauser, Dan Longo, J. Larry Jameson, 2022-04-05 The Voice of Clinical Reason A Doody's Core Title for 2024 2023 Harrison's Principles of Internal Medicine is the world's most trusted clinical medicine text and a superb resource for learning the art and science of clinical reasoning Recognized by healthcare professionals worldwide as the leading authority on applied pathophysiology and clinical medicine Harrison's Principles of Internal Medicine provides the informational foundation you need for the best patient care possible This new edition is fully updated with timely new chapters and essential updates across the spectrum of internal medicine Harrison's Principles of Internal Medicine stands as the benchmark for authoritative practical information on patient care and the pathogenesis and clinical management of symptoms and signs and specific diseases Written and edited by the world's top experts in their respective fields this landmark guide provides the comprehensive accurate and essential coverage of the pathogenesis diagnosis and treatment of disease Harrison's is world renowned as the most authoritative source for Descriptions of disease mechanisms and how the clinician can apply that knowledge for the best patient care and optimal diagnosis and treatment of specific diseases Clear concise schemas that facilitate the generation of differential diagnoses to reason efficiently through complex real world clinical cases The physiologic and epidemiologic basis of signs and symptoms which are covered through a wealth of unsurpassed expert guidance and linked to the disease specific chapters that follow Updated clinical trial results and recommended guidelines Excellent and extensive visual support including radiographs clinical photos schematics and high quality drawings Coverage of both therapeutic approaches and specific treatment regimens Practical clinical decision trees and algorithms Organ specific sections with clinically relevant pathophysiology and practical clinical advice on the approach to the patient strategies towards building a differential diagnosis outstanding clinical algorithms and diagnostic schema a wealth of clinical images and diagrams current clinical guidelines general and specific approaches to therapy Harrison's remains the most trusted resource in a world influenced by endless sources of medical information The most timely and comprehensive updates from the world's top experts are featured in the 21st edition Current coverage of the diagnosis and treatment of diseases from COVID to dementia to sepsis to multiple sclerosis to lung cancer Updated content that reflects new approved therapeutics and new practice changing guidelines and evidence summaries More than 1000 clinical pathological and radiographic photographs diagnostic and therapeutic decision trees and clear schematics and diagrams describing pathophysiologic processes More than a dozen atlases featuring curated collections of visual aspects of

diagnosis and management Complete updated curation and synthesis of primary medical literature which incorporates current data from major studies and clinical trials Clinical reasoning resources and helpful disease presentation schemas Clinically relevant coverage of disease mechanics and pathophysiology and related therapeutic mechanisms

Pediatric Rehabilitation Gabriella E. Molnar, Michael Allen Alexander, 1999 Pediatric Rehabilitation is an important component of both Physiatry and Pediatrics Given the potential survival time of the patient and the enormous emotional social and economic costs involved the rehabilitation management of children is one of the most important areas of both specialties This book will be the definitive text reference on this important area and is a great addition to Hanley and Belfuss impressive program of books in Physical Medicine and Rehabilitation Drs Molnar and Alexander have made a classic reference better than ever in this completely revised and updated work Some of the major names in the field have contributed comprehensive yet highly practical chapters

Pediatric Dosage Handbook Carol K. Taketomo, Jane Hurlburt Hodding, Donna M. Kraus, 2009 The pediatric population is a dynamic group with major changes in pharmacokinetics and pharmacodynamics taking place throughout infancy and childhood Because of these changes the need for the evaluation and establishment of medication dosing regimens in children of different ages is great This book includes 17 drug monographs

Handbook of Neurologic Rating Scales Robert M. Herndon, 1997

Guide to Pediatric Physical Therapy: A Clinical Approach Martha Bloyer, Tricia Catalino, Eric Shamus, Cindy Miles, 2025-01-10 Everything you need to know to perform safe effective physical therapy on babies children and teens Guide to Pediatric Physical Therapy provides pedagogy from top experts in the field to help you master the practice of PT for kids This dynamic easy to follow resource is filled with cases that help you apply concepts to real world situations along with art and illustrations that reinforce what you have learned Each chapter opens with a case which is followed by two or three additional cases presented as boxed features Critical information is presented in tables particularly effective in helping you quickly digest key concepts With more than 75 collective years teaching pediatric physical therapy this author team are masters of the subject matter and know how today's students prefer to learn Key tables highlight high yield information Each case study is followed by open ended questions for to consider Chapter summaries are presented in bullet form to make learning easy and quick Q A following summaries are written in NPTE Exam format

Abstracts, 2003

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available for download *Amyotrophic Lateral Sclerosis and Other Motor Neuron Diseases* Lewis P. Rowland, 1991 A collection of invited papers from a Muscular Dystrophy Association sponsored symposium which highlight findings and theories on the molecular genetics of these diseases assess concepts on immune mediated motor neuron destruction and examine the pathogenesis of motor neuron disease **Lexikon** Joint Commission on Accreditation of Healthcare Organizations, 1998

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