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European Reference Network
for rare or low prevalence complex diseases
Network Neuromuscular Diseases (ERN EURO-NMD)

8th ERN EURO-NMD
Annual Meeting

Exercise in neuromuscular disorders

5th – 7th March 2025

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Rehabillitation physician
Radboudumc Nijmegen
Klimmendaal Arnhem,
the Netherlands

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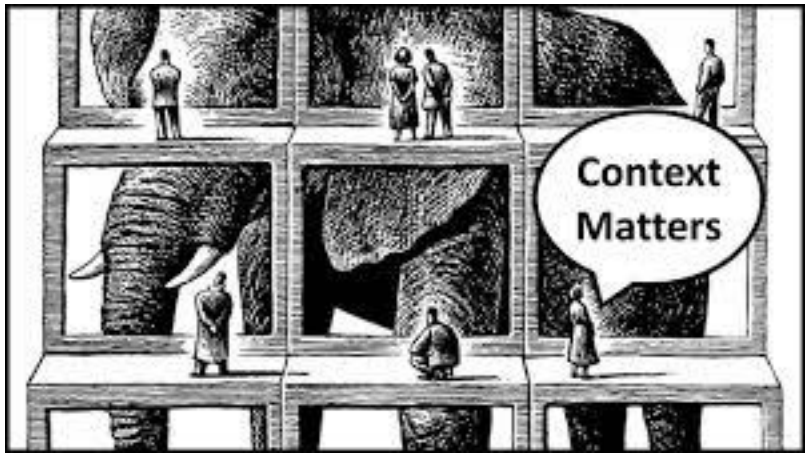
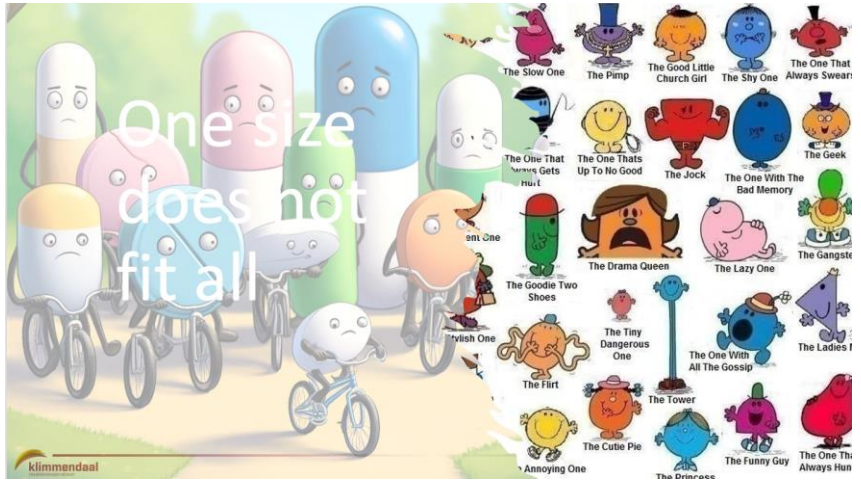
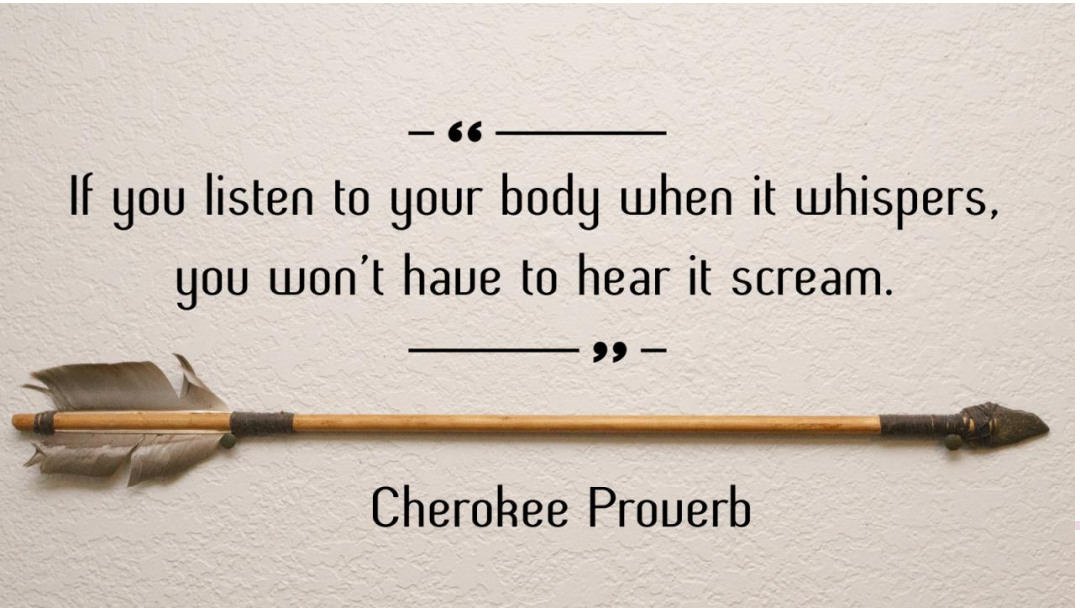
Take home messages



- Start low and go slow
- One size does not fit all
- Context matters: training is more than just exercise
- Listen proactively to your body





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Fatigue in NMD

On average, more than 60% of patients with NMD experience severe and chronic fatigue

Usually an indirect symptom, the result of behavioral change

Not so long recognized and acknowledged as a problem

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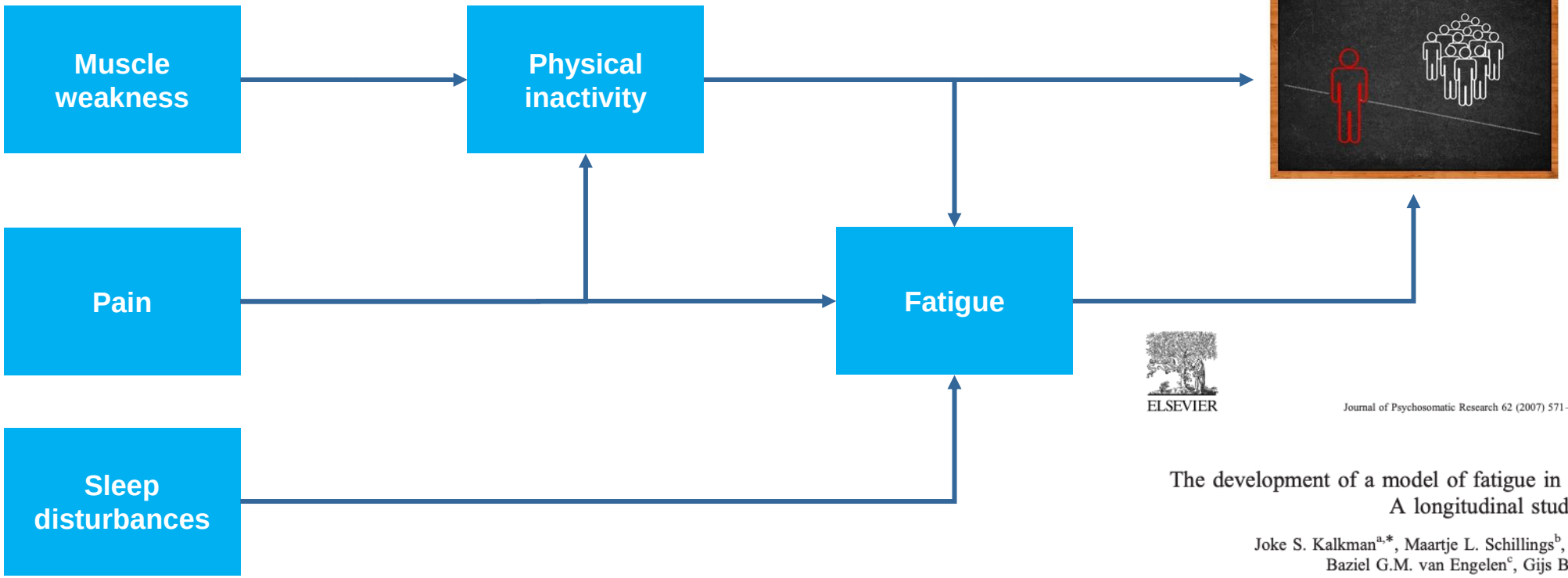
Perpetuating factors of experienced fatigue in FSHD and CMT



ELSEVIER

Journal of Psychosomatic Research 62 (2007) 571–579

Journal of Psychosomatic Research

The development of a model of fatigue in neuromuscular disorders: A longitudinal study

Joke S. Kalkman[a,*], Maartje L. Schillings[b], Machiel J. Zwarts[b], Baziel G.M. van Engelen[c], Gijs Bleijenberg[a]

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Chronic versus acute fatigue

Duration: > 3 months

It does not (always) go away after rest

An unpleasant feeling

No relationship between activities, rest, and the level of fatigue

Multidimensional problem

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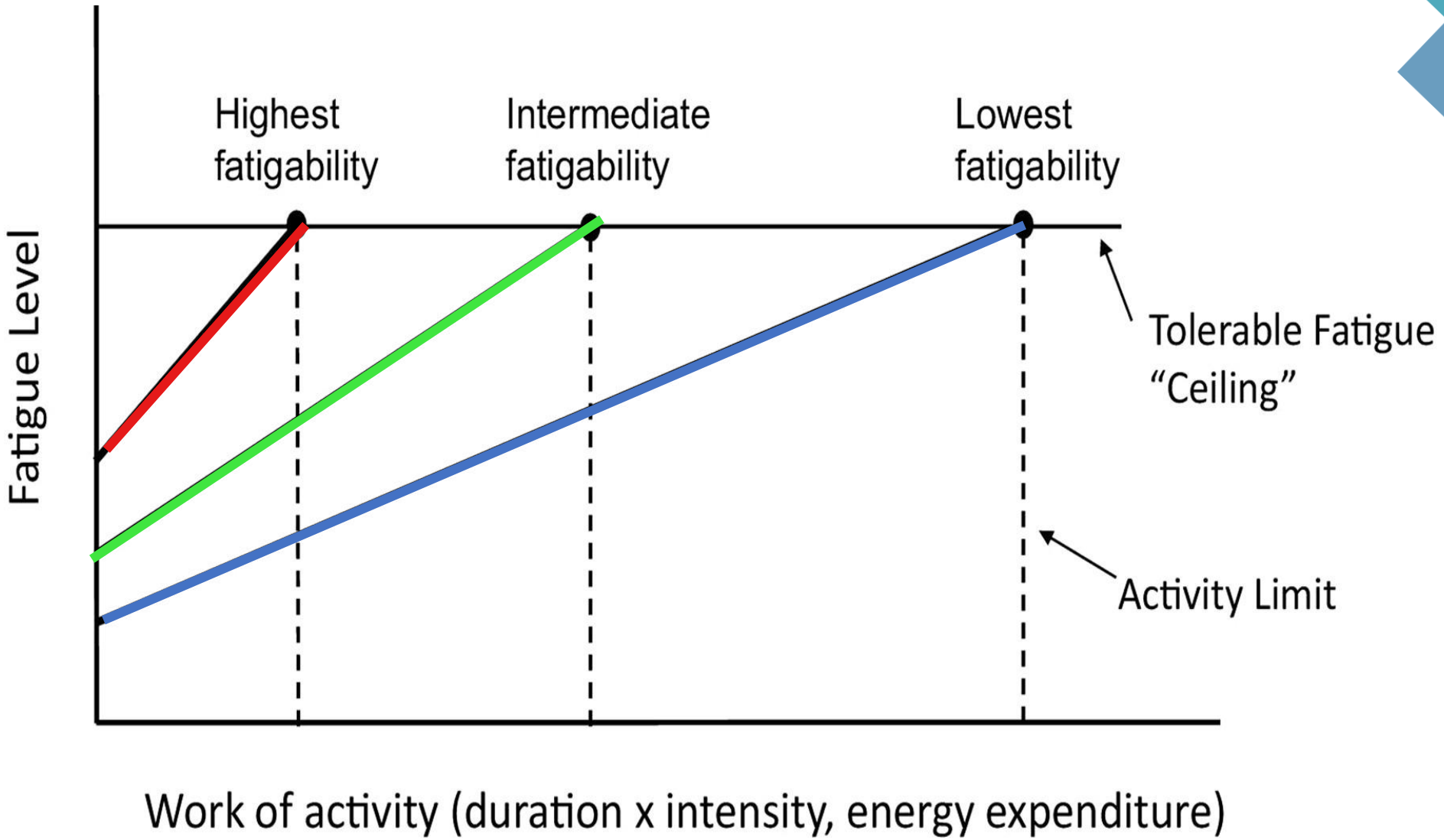
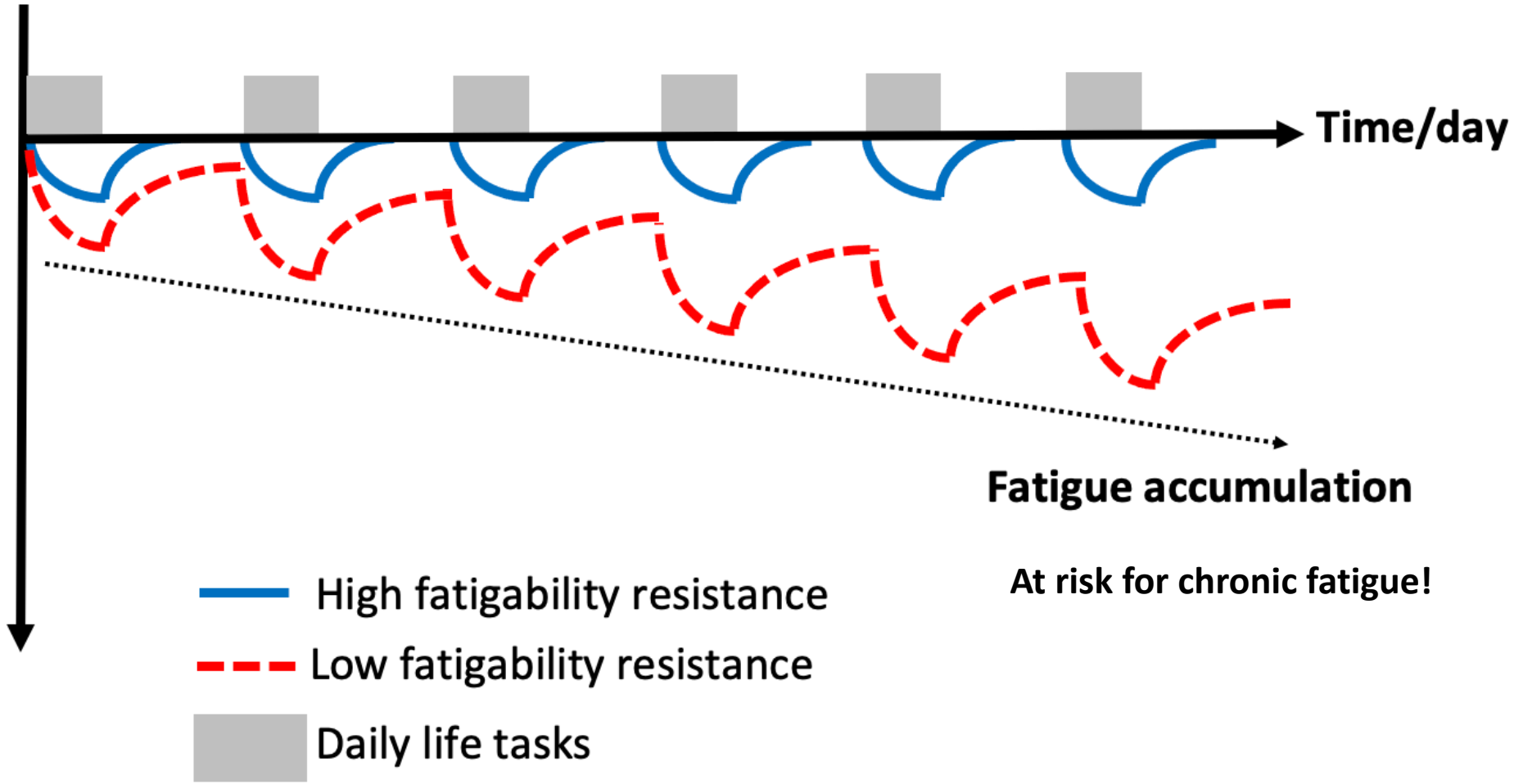

Highest fatigability
Intermediate fatigability
Lowest fatigability
Fatigue Level
Tolerable Fatigue "Ceiling"
Activity Limit
Work of activity (duration x intensity, energy expenditure)



At risk for chronic fatigue!

Both aerobic exercise and cognitive-behavioral therapy reduce chronic fatigue in FSHD

An RCT

Nicoline Voet, MD
Gijs Bleijenberg, PhD
Jan Hendriks, PhD
Imelda de Groot, MD, PhD
George Padberg, MD, PhD
Baziel van Engelen, MD, PhD*
Alexander Geurts, MD, PhD*

Correspondence to Dr. Voet: Nicole.Voet@RadboudUMC.nl

ABSTRACT

Objective: To investigate the effect of aerobic exercise training (AE
apy (CBT) on chronic fatigue in patients with facioscapulohumeral

Methods: We performed a multicenter, assessor-blinded, rando
seven patients with FSHD type 1 with severe chronic fatigue w
CBT, or usual care (UC). Outcomes were assessed before treatme
vention, and after a 12-week follow-up. A linear mixed model fo
used to study the estimated group differences.

Results: Following treatment, both the AET (28 participants) and
tion groups had less fatigue relative to the UC group (24 particip
for AET (95% confidence interval [CI] −12.4 to −5.8) and −13
−10.2). These beneficial effects lasted through follow-up, with
(95% CI −12.4 to −5.8) and −10.2 for CBT (95% CI −14.
received CBT had an increase in registered and experienced p
and social participation. The patients who received AET had an
activity only. The increase in registered physical activity in both
social participation following CBT were still present at follow-up.

Conclusions: This RCT shows that AET and CBT can ameliorate
FSHD.

Classification of evidence: This study provides Class III evidence t
1 and severe chronic fatigue, AET or CBT reduces the severity

2014;83:1914-1922

Quantitative MRI reveals decelerated fatty infiltration in muscles of active FSHD patients

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Nicoline Voet, MD*
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Baziel van Engelen, MD, PhD
Arend Heerschap, PhD

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ABSTRACT

Objective: To investigate the effects of aerobic exercise training (AET) and cognitive-behavioral therapy (CBT), directed towards an increase in daily physical activity, on the progression of fatty infiltration and edema in skeletal muscles of patients with facioscapulohumeral muscular dystrophy (FSHD) type 1 by T2 MRI.

Methods: Quantitative T2 MRI (qT2 MRI) and fat-suppressed T2 MRI of the thigh were performed at 3T on 31 patients, 13 of whom received usual care (UC), 9 AET, and 9 CBT. Muscle-specific fat fractions (%), derived from qT2 MRI, were recorded pretreatment and posttreatment. Intervention effects were analyzed by comparing fat fraction progression rates of the UC with the treated groups using Mann-Whitney tests, and intermuscle differences by a linear mixed model. Edematous hyperintense lesions were identified on the fat-suppressed T2 MRI.

Results: The intraclass correlation coefficient for reproducibility of qT2 MRI fat assessment was 0.99. In the UC group, the fat fraction increased by 6.7/year (95% confidence interval [CI] 4.3 to 9.1). This rate decreased to 2.9/year (95% CI 0.7 to 5.2) in the AET ($p = 0.03$) and 1.7/year (95% CI −0.2 to 3.6) in the CBT group ($p = 0.00015$). The treatment effect differed among individual muscles. Fewer new edematous lesions occurred after therapy.

Conclusions: Fat fraction derived from qT2 MRI is a reproducible and sensitive biomarker to monitor the effects of increased physical activity in individual muscles. This biomarker reports a favorable effect of AET and CBT on the rate of muscular deterioration in FSHD as reflected in decelerated fat replacement.

Classification of evidence: This study provides Class II evidence that for patients with FSHD type 1, both AET and CBT decrease the rate of fatty infiltration in muscles. *Neurology*® 2016;86:1700-1707

The ERN EURO-NMD is funded by the European Commission under the EU4Health programme (EURO-NMD 23-27 — 101156434 — EU4H-2023-ERN2-IBA)

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("Neuromuscular Diseases"[Mesh]) AND "Exercise"[Mesh]

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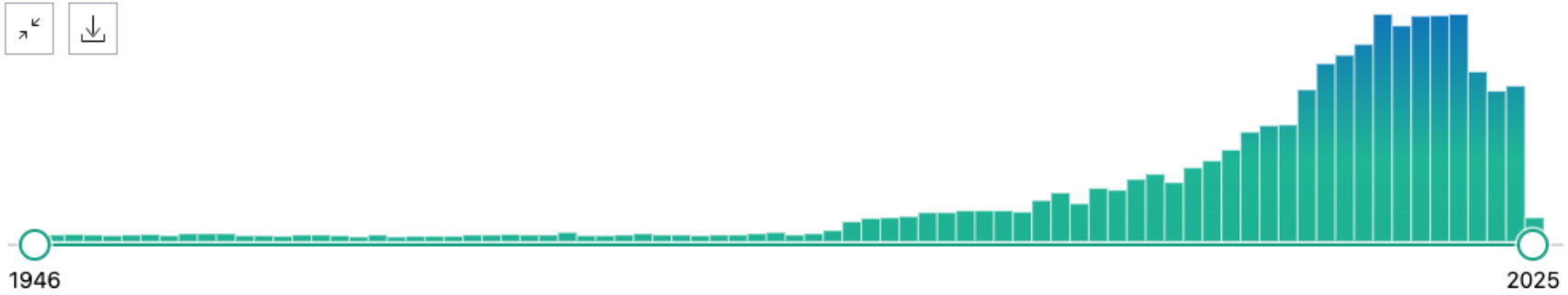
RESULTS BY YEAR

4,804 results

Page 1 of 481



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Pain in neuromuscular disorders

On average, more than 80% of patients with NMD experience severe and chronic fatigue

Usually an indirect symptom, the result of compensation mechanisms

Not so long recognized and acknowledged as a problem

No effect of exercise on pain in NMD; on the contrary…

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Chronic versus acute pain

Duration: > 3 months

It does not (always) go away after rest

No relationship between activities, rest, and the level of fatigue

Multidimensional problem

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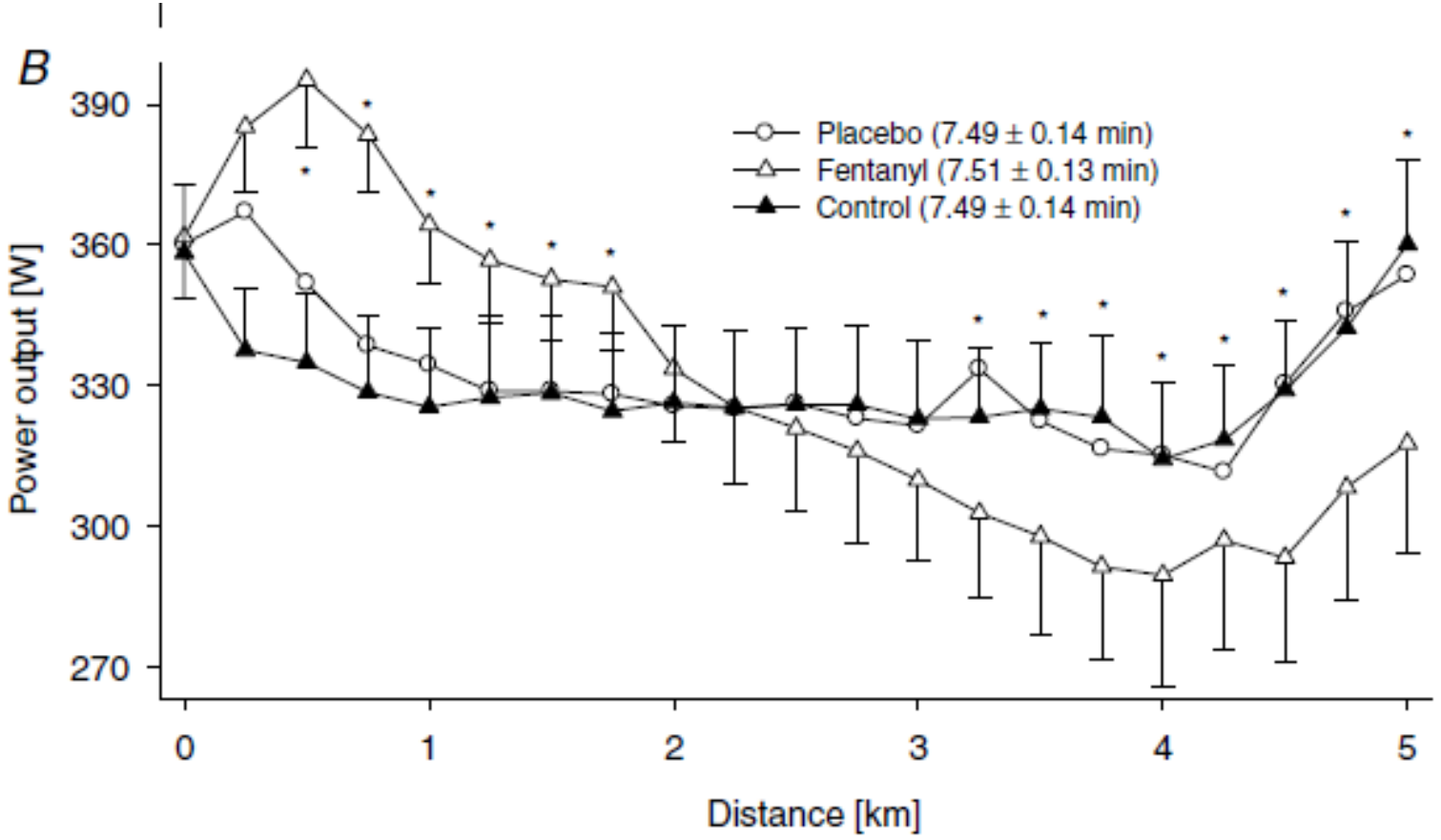
Don’t ignore pain!



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Side effects and adverse effects of exercise

Side effects: mild and temporary

Muscle soreness (more pronounced and longer), (more quickly) increased heart rate and shortness of breath, acute fatigue

Side effects can become **adverse effects** in NMD when:

- recovery time is not taken into account
- improper exercise selection
- contextual factors need a lot of energy
- the exercise intensity is too high

Recovery time is compromised and prolonged in NMD

Examining Recovery from Maximal Exercise Testing in Patients with Neuromuscular Disorders

Constance de Monts 1, Yair Blumberg 2, Sam Montalvo 2, Paxton Ataide 1, Whitney J Tang 1, Sally Dunaway Young 1, Noirin Ni Ghiollagain 1, Dana Parker 1, JW Day 1, Matt Wheeler 2, Jeff Christle 2, Tina Duong 1

1. Department of Neurology, Stanford University, Palo Alto, CA, USA 2. Department of Medicine (Biomedical Informatics Research), Stanford University
2. Division of Cardiovascular Medicine, Department of Medicine, Stanford University Palo Alto CA, USA

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Tour de France 2024
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(Im)proper exercise selection

J Rehabil Med 2019; 51: 518–524

ORIGINAL REPORT

Check for updates

AEROBIC EXERCISE IN ADULT NEUROMUSCULAR REHABILITATION: A SURVEY OF HEALTHCARE PROFESSIONALS

Eric L. VOORN, PhD, Fieke KOOPMAN, MD, PhD, Frans NOLLET, MD, PhD, and Merel A. BREHM, PhD
From the Amsterdam UMC, University of Amsterdam, Department of Rehabilitation, Amsterdam Movement Sciences, Amsterdam, The Netherlands

Clinicians

Safety concerns

- Lack of knowledge about prescribing exercise (screening and dosing) 77%
- Overwork weakness 45%

Patients

Inability to perform training level 73%

Poor motivation 55%

Fatigue 45%

Voorn et al 2019

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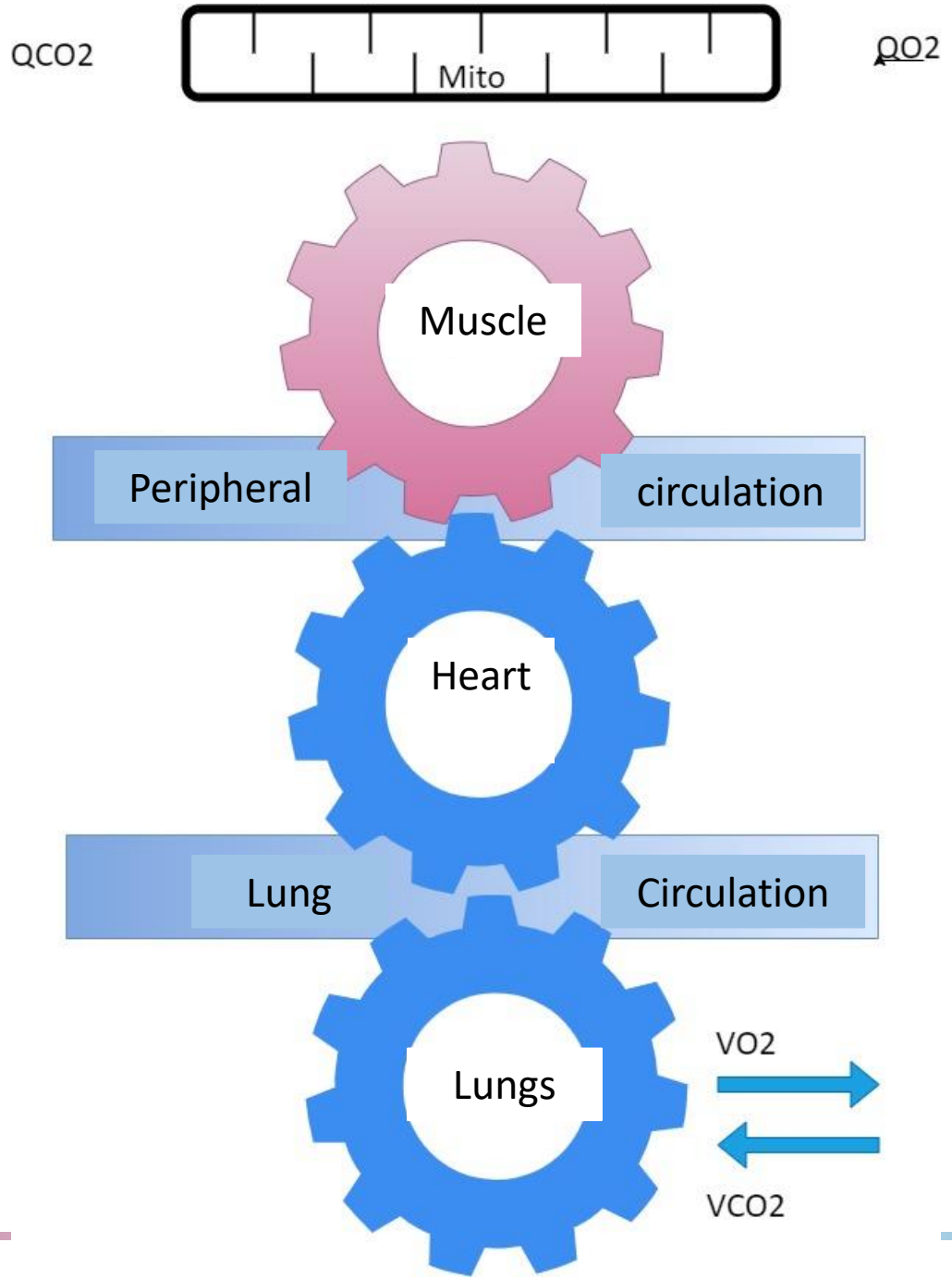

QCO2
Mito
QO2
Muscle
Peripheral
circulation
Heart
Lung
Circulation
Lungs
VO2
VCO2

Fatigue thresholds

frontiers in Physiology

ORIGINAL RESEARCH
published: 17 March 2022
doi: 10.3389/fphys.2022.821584

Check for updates

Surface Electromyography Thresholds as a Measure for Performance Fatigability During Incremental Cycling in Patients With Neuromuscular Disorders

Nicoline B. M. Voet[1,2], Christiaan G. J. Saris[3], Dick H. J. Thijssen[4], Vincent Bastiaans[5], David E. Sluijs[5] and Mariska M. H. P. Janssen[1,2]*

The ERN EURO-NMD is funded by the European Commission under the EU4Health programme (EURO-NMD 23-27 — 101156434 — EU4H-2023-ERN2-IBA)

Methods

32 NMD patients

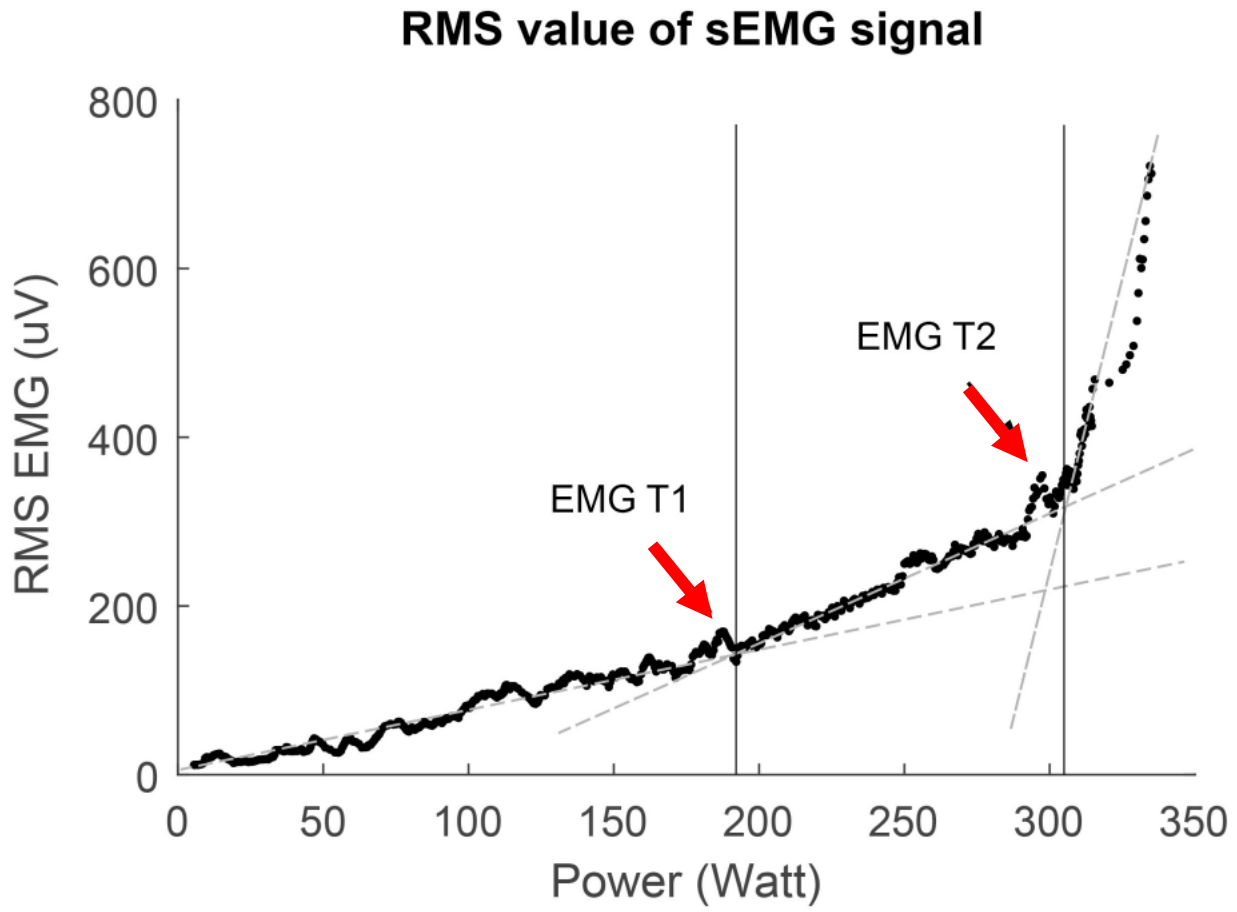
24 healthy controls

CPET (steep ramp protocol)

EMG on leg muscles

Ventilatory thresholds

sEMG thresholds



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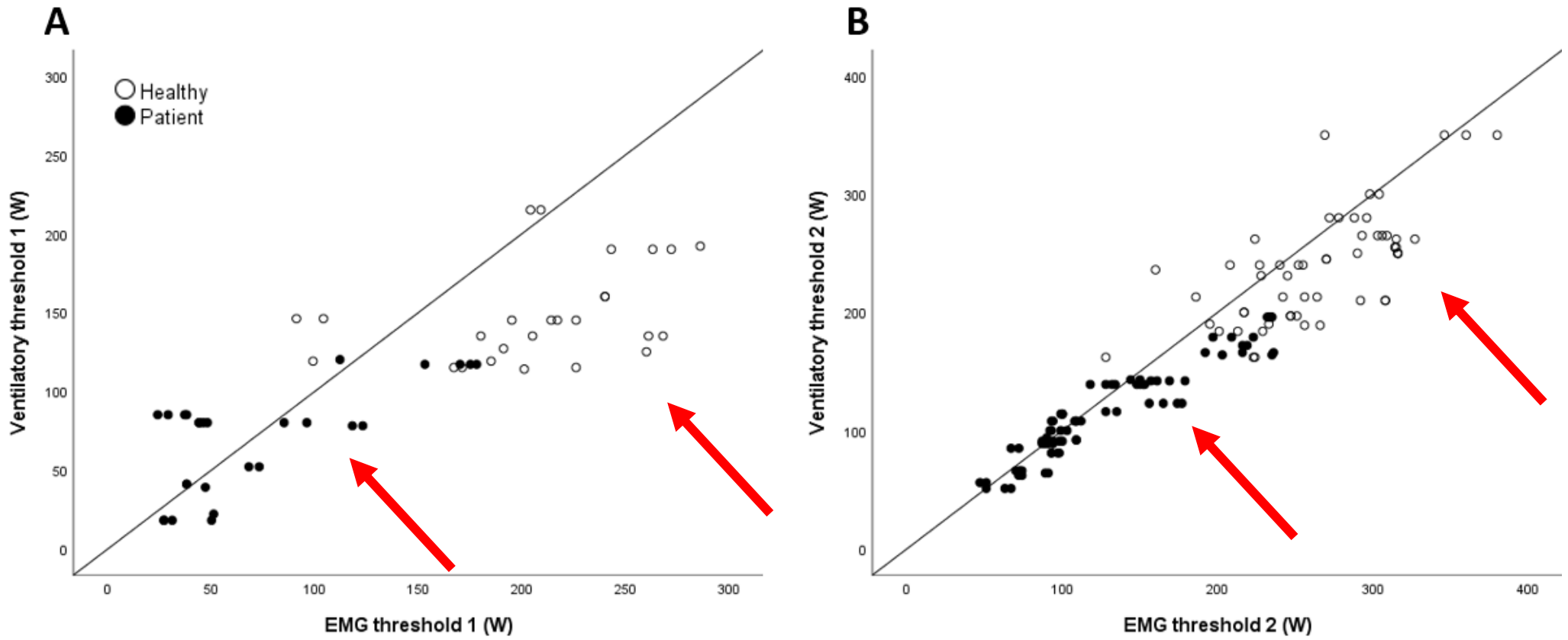
Muscle fatigue relatively earlier in time



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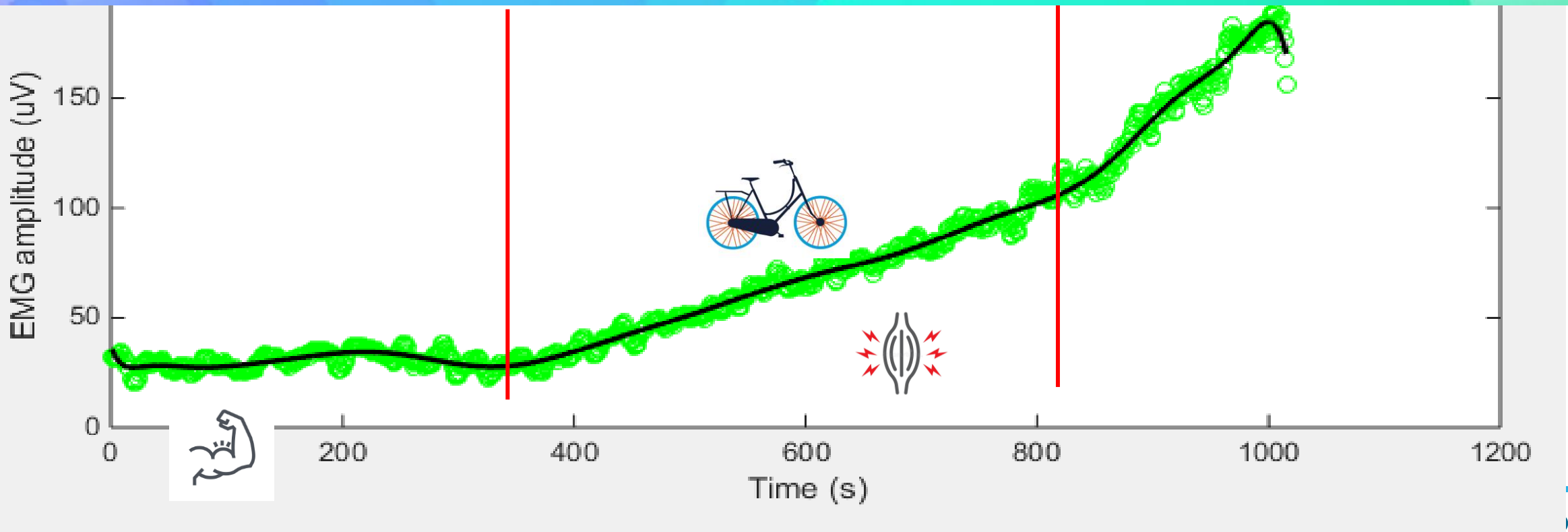
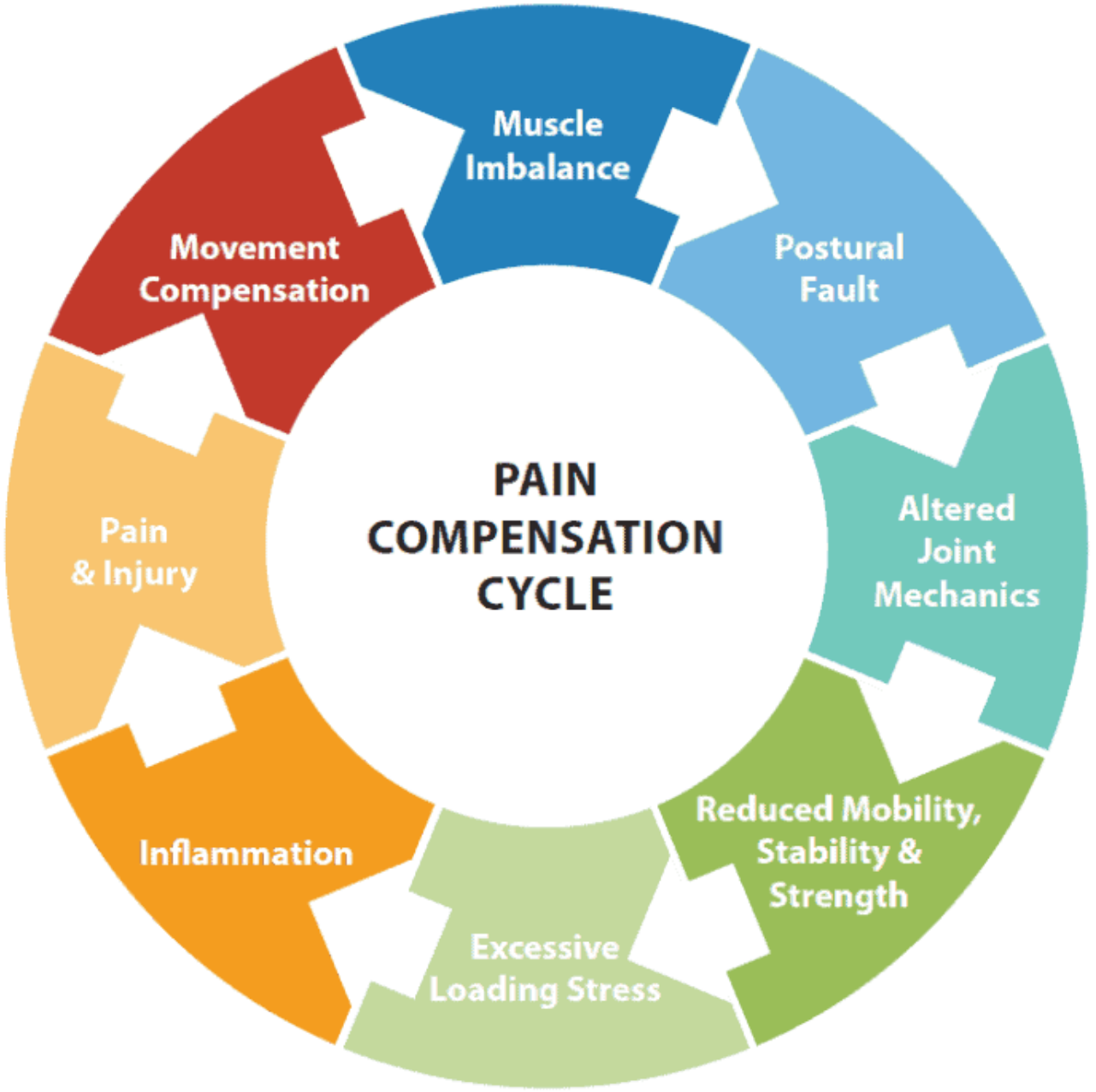
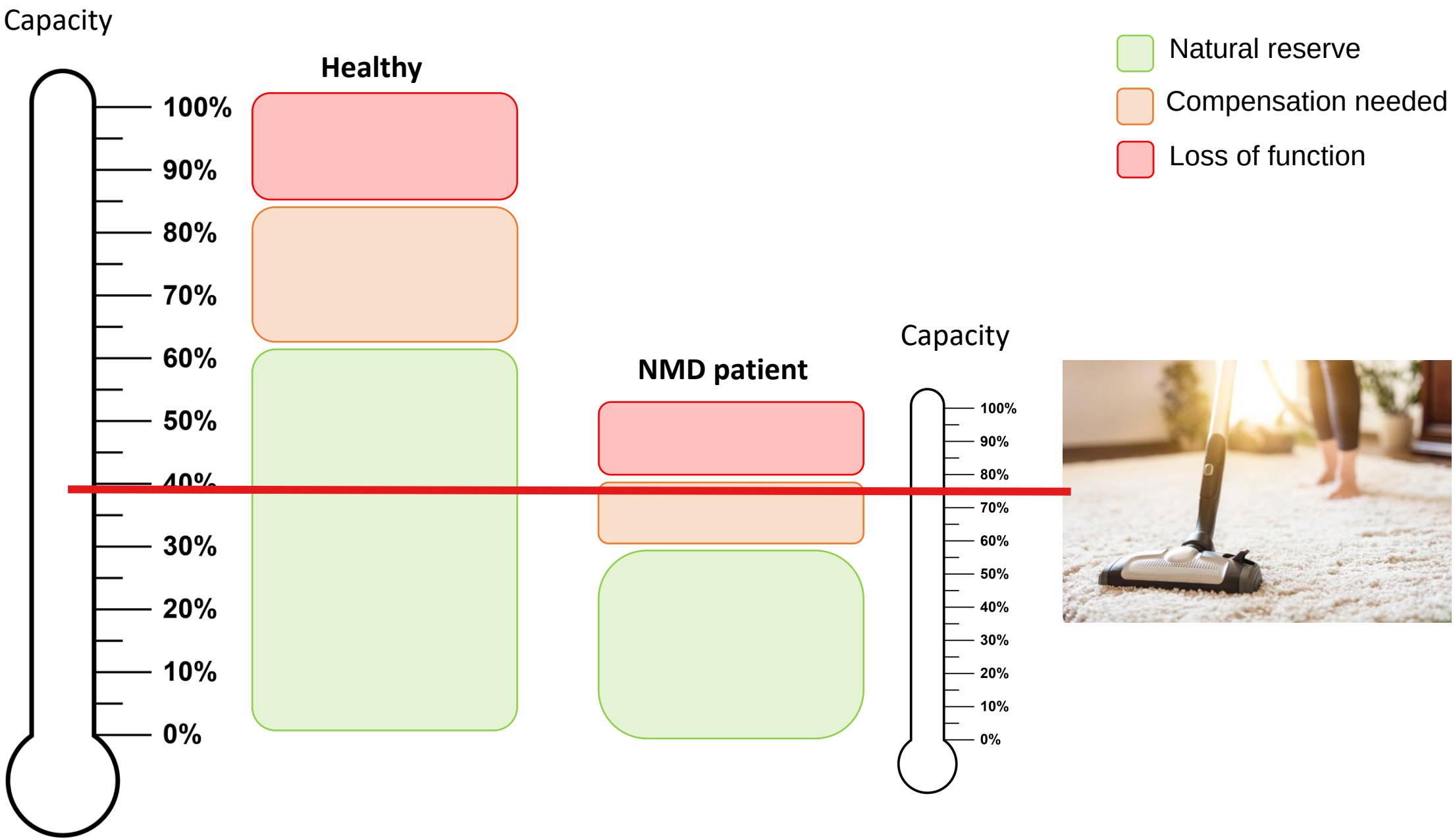
START LOW,
GO SLOW.




Muscle Imbalance
Postural Fault
Altered Joint Mechanics
Reduced Mobility, Stability & Strength
Excessive Loading Stress
Inflammation
Pain & Injury
Movement Compensation
PAIN COMPENSATION CYCLE



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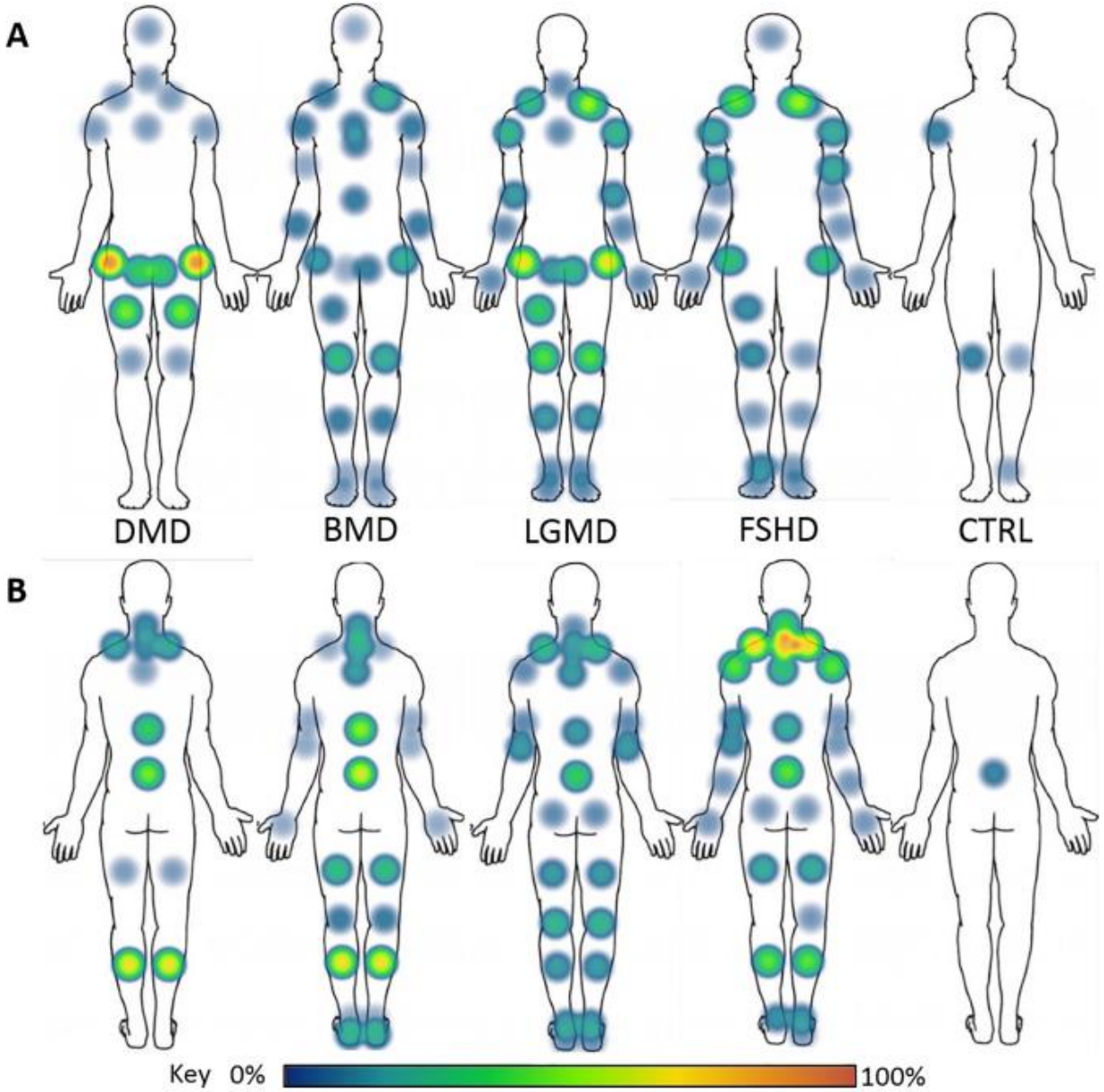

Fig 3. Body maps. Topographic presentation of reported pain frequency across four types of Muscular Dystrophy using a localised method. A = Anterior; B = Posterior; DMD = Duchenne Muscular Dystrophy; BMD = Beckers Muscular Dystrophy; LGMD = Limb-Girdle Muscular Dystrophy; FSHD = Facioscapulohumeral Muscular Dystrophy; CTRL = Control

RESEARCH ARTICLE

Frequency of reported pain in adult males with muscular dystrophy

Matthew F. Jacques[1]*, Rachel C. Stockley[2], Emma I. Bostock[1], Jonathon Smith[3], Christian G. DeGoede[4], Christopher I. Morse[1]

1 Musculoskeletal Science & Sports Medicine Research Centre, School of Healthcare Science, Faculty of Science and Engineering, Manchester Metropolitan University, Manchester, United Kingdom, **2** School of Nursing, University of Central Lancashire, Preston, United Kingdom, **3** The Neuromuscular Centre, Winsford, Cheshire, United Kingdom, **4** Department of Paediatric Neurology, Royal Preston Hospital, Preston, United Kingdom

* matthew.jacques@stu.mmu.ac.uk

Fatigue > change in movement

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Muscle fatigue and compensatory movements

The primary muscle becomes fatigued

More powerful muscle compensates

Change in kinematics

Compensation fails

Muscle fatigue as measured with EMG

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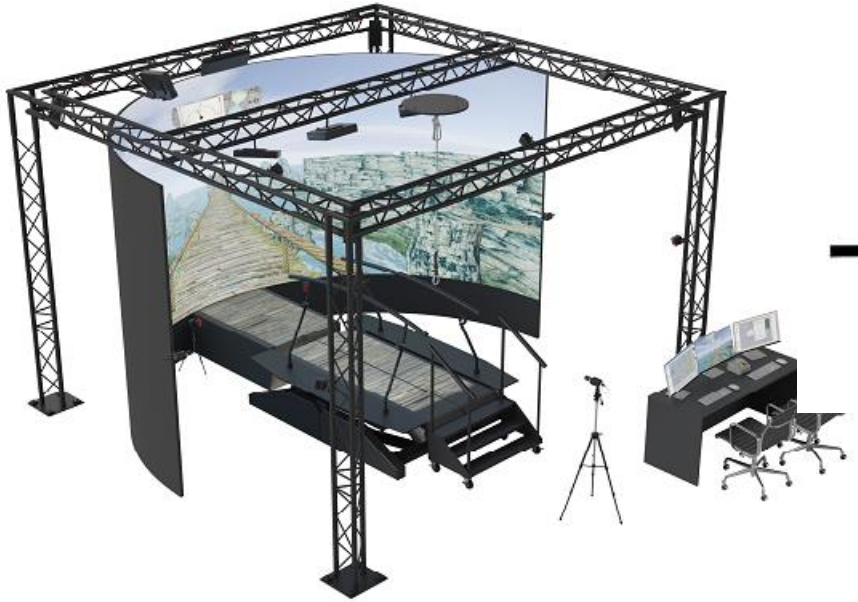
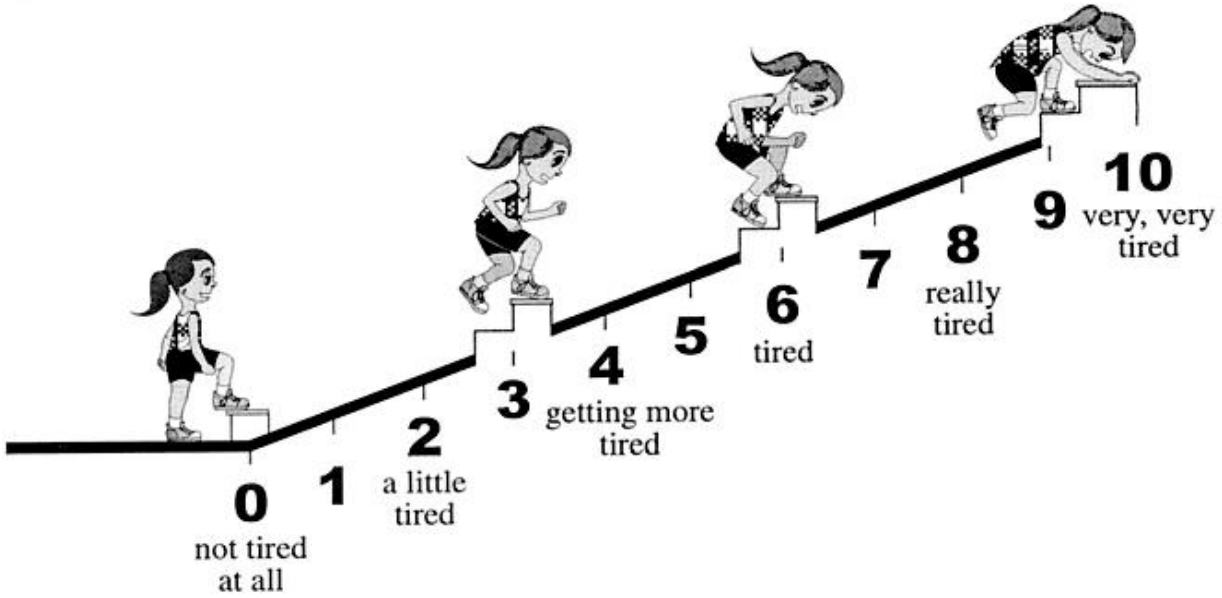
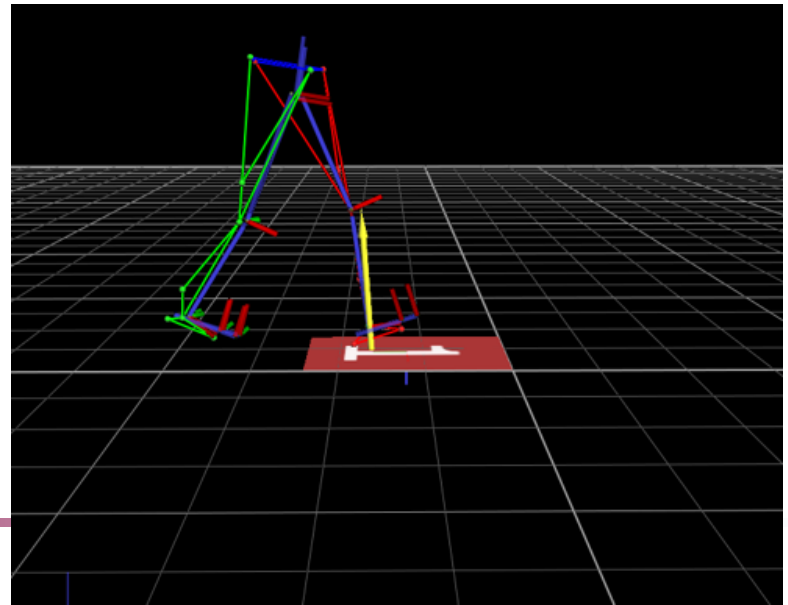
Methods





OMNI- scale for experienced fatigue

Kinematics



EMG measurements

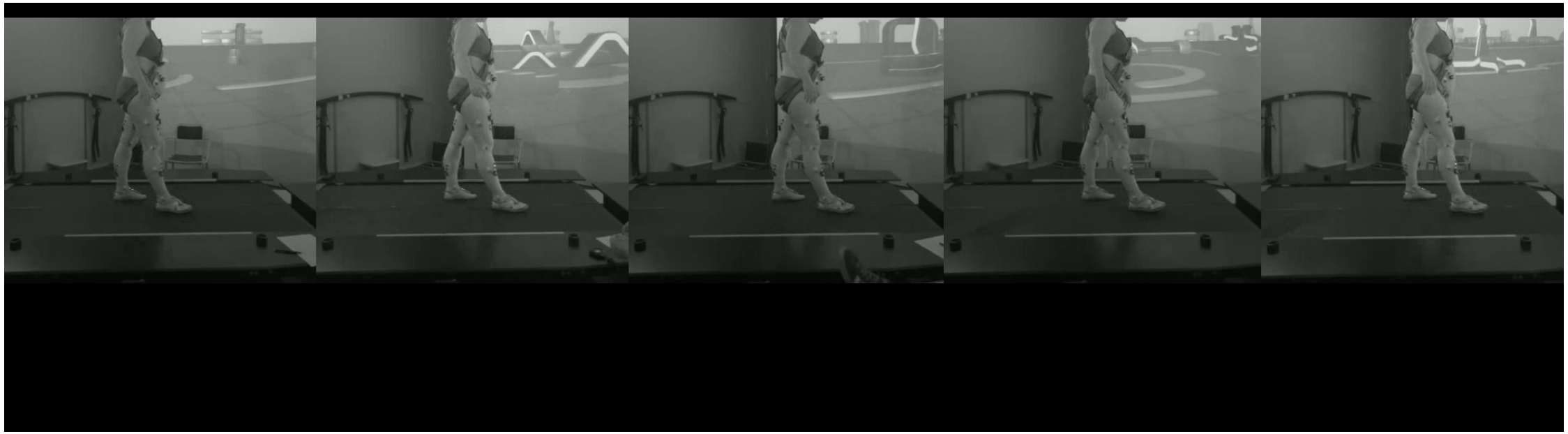
Methods

OMNI score after 8 to 30 minutes walking

Baseline measurement

Long-term measurement

Final measurement



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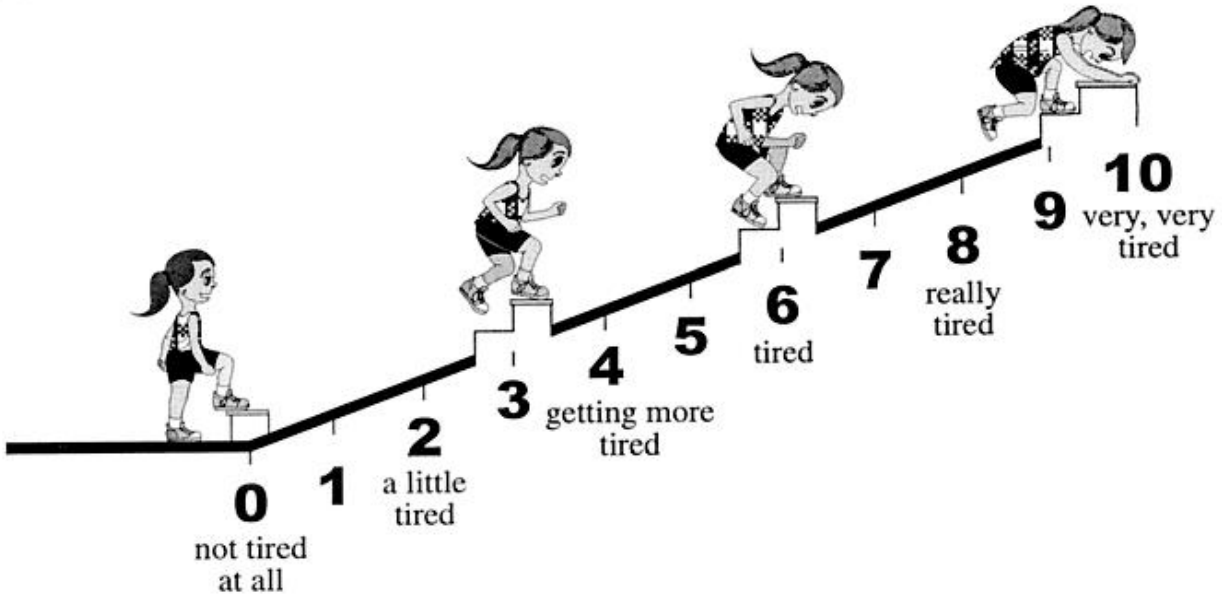
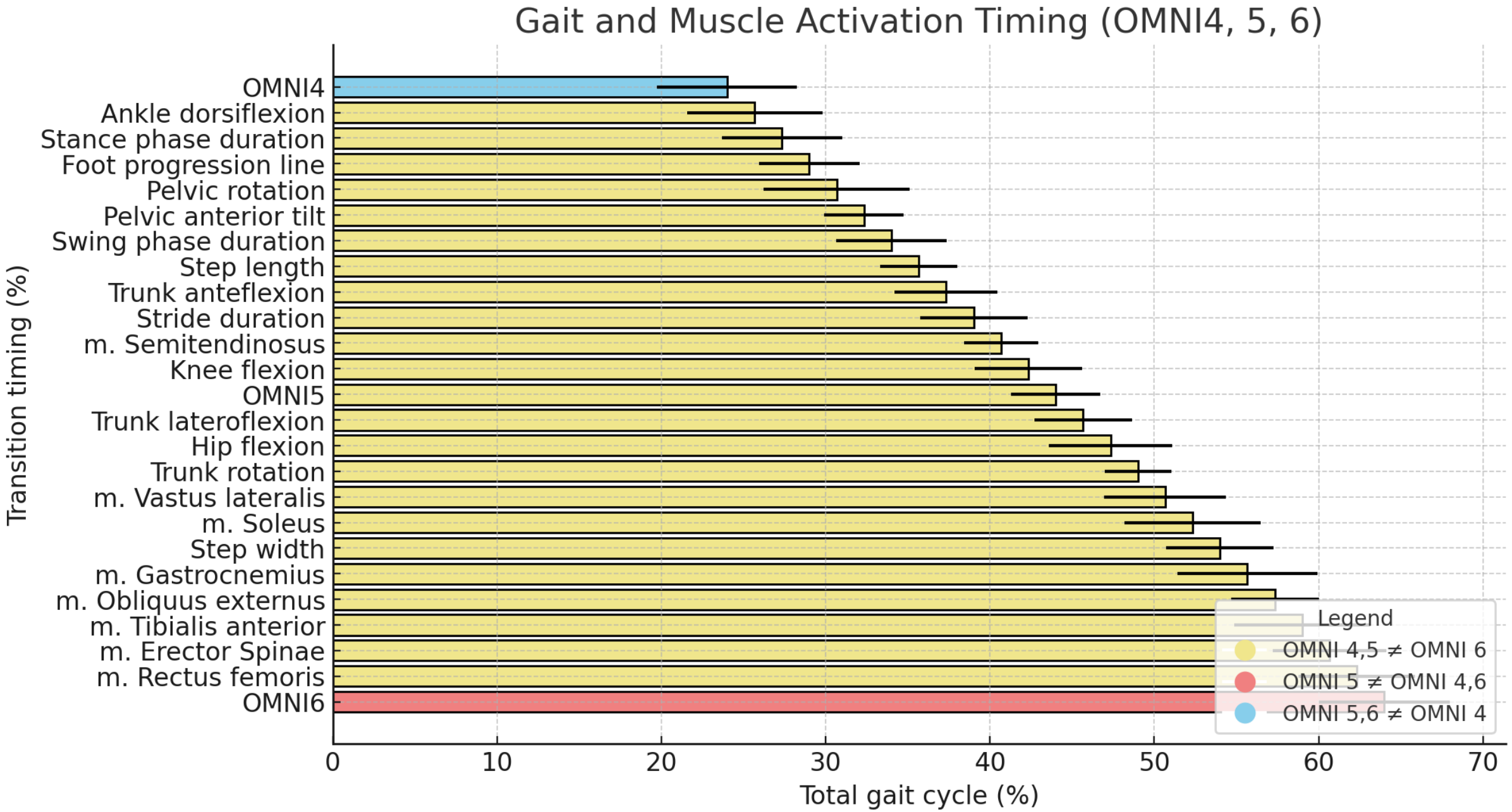
Results

11 people with different NMD, from 17 – 7 years of age

Walking duration 10 to 30 minutes

Main result: objective muscle fatigue precedes experienced fatigue





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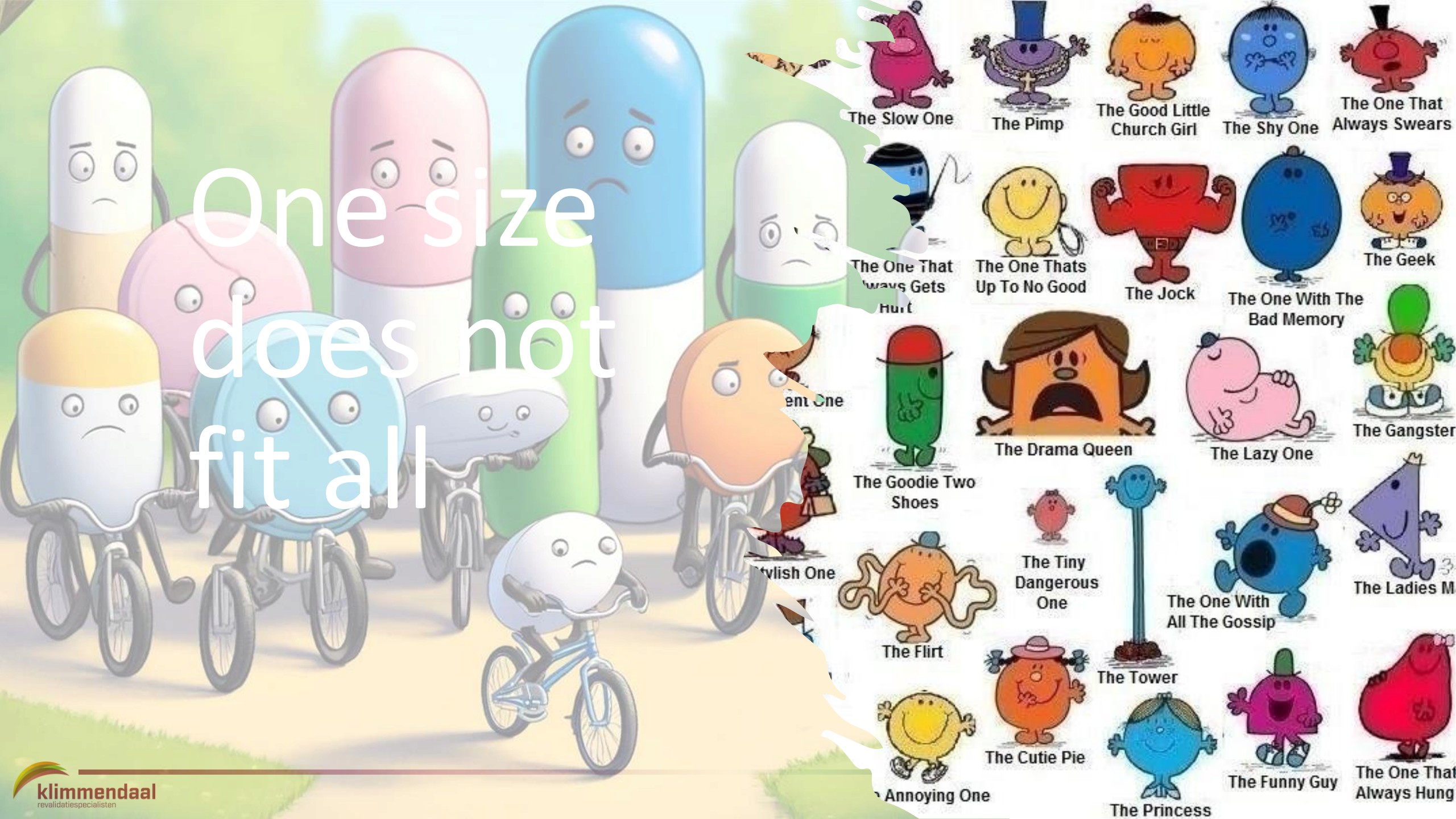
One size does not fit all



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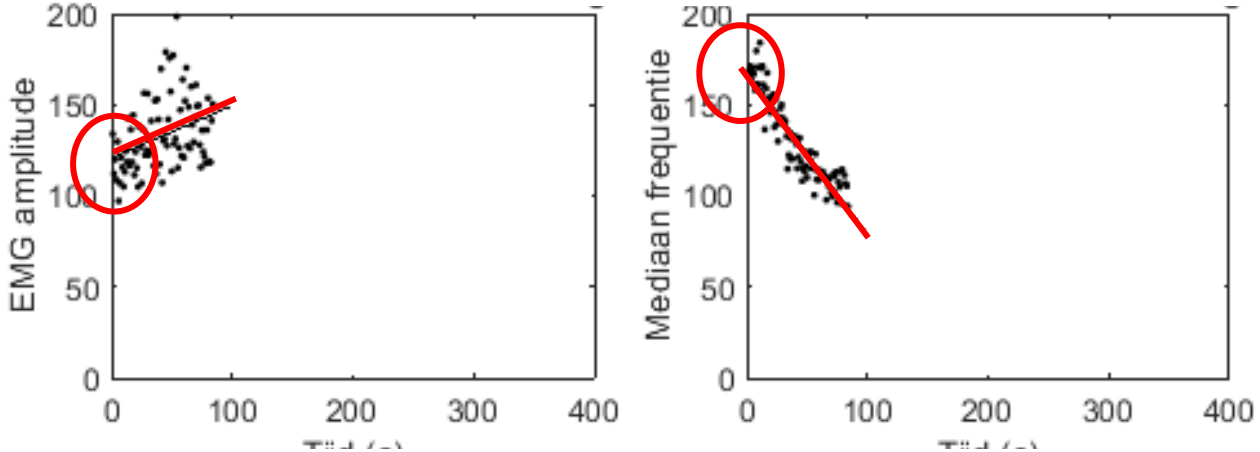
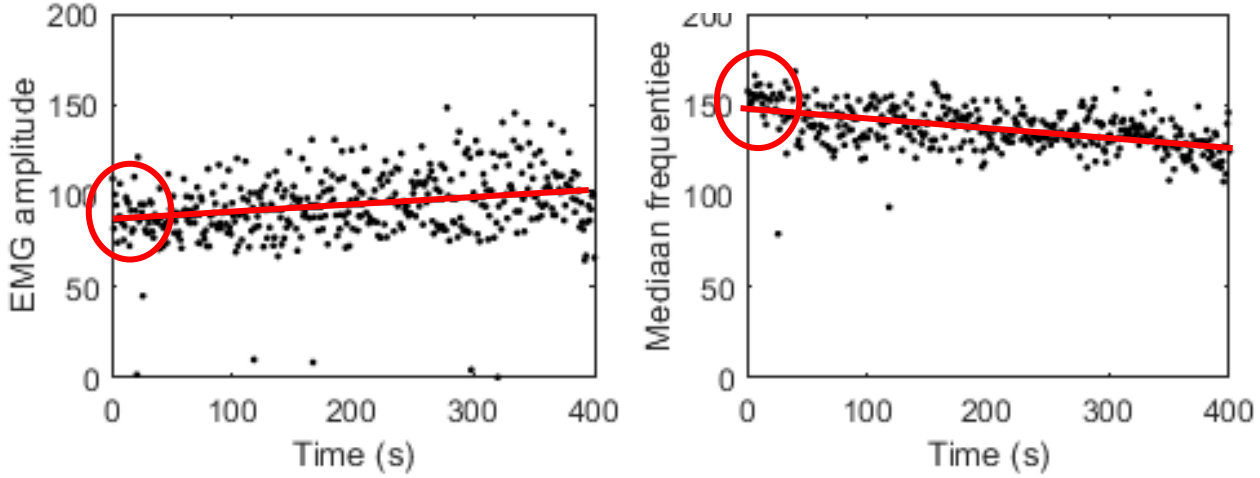
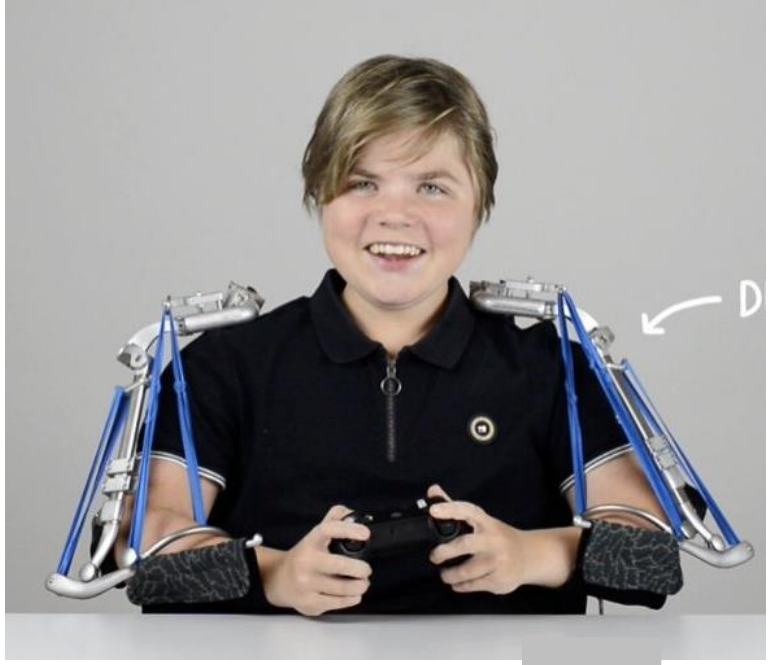
Less fatigue with arm support

Deltoid without support



Deltoid with support





Disability and Rehabilitation: Assistive Technology

ISSN: (Print) (Online) Journal homepage: www.tandfonline.com/journals/iidt20

Evaluation of fatigue and fatigability in people with Duchenne muscular dystrophy using a dynamic arm support – a pilot study

Lonneke Alberts, Nicole Voet & Mariska Janssen

To cite this article: Lonneke Alberts, Nicole Voet & Mariska Janssen (12 Sep 2024): Evaluation of fatigue and fatigability in people with Duchenne muscular dystrophy using a dynamic arm support – a pilot study, Disability and Rehabilitation: Assistive Technology, DOI: 10.1080/17483107.2024.2388284

To link to this article: https://doi.org/10.1080/17483107.2024.2388284

Does it work in clinical practice?

Male, 52 years, mitochondrial myopathy

High level of experienced fatigue

Participated in ‘Energetic’ program, with baseline CPET

Failed in increasing training load, prolonged recovery time

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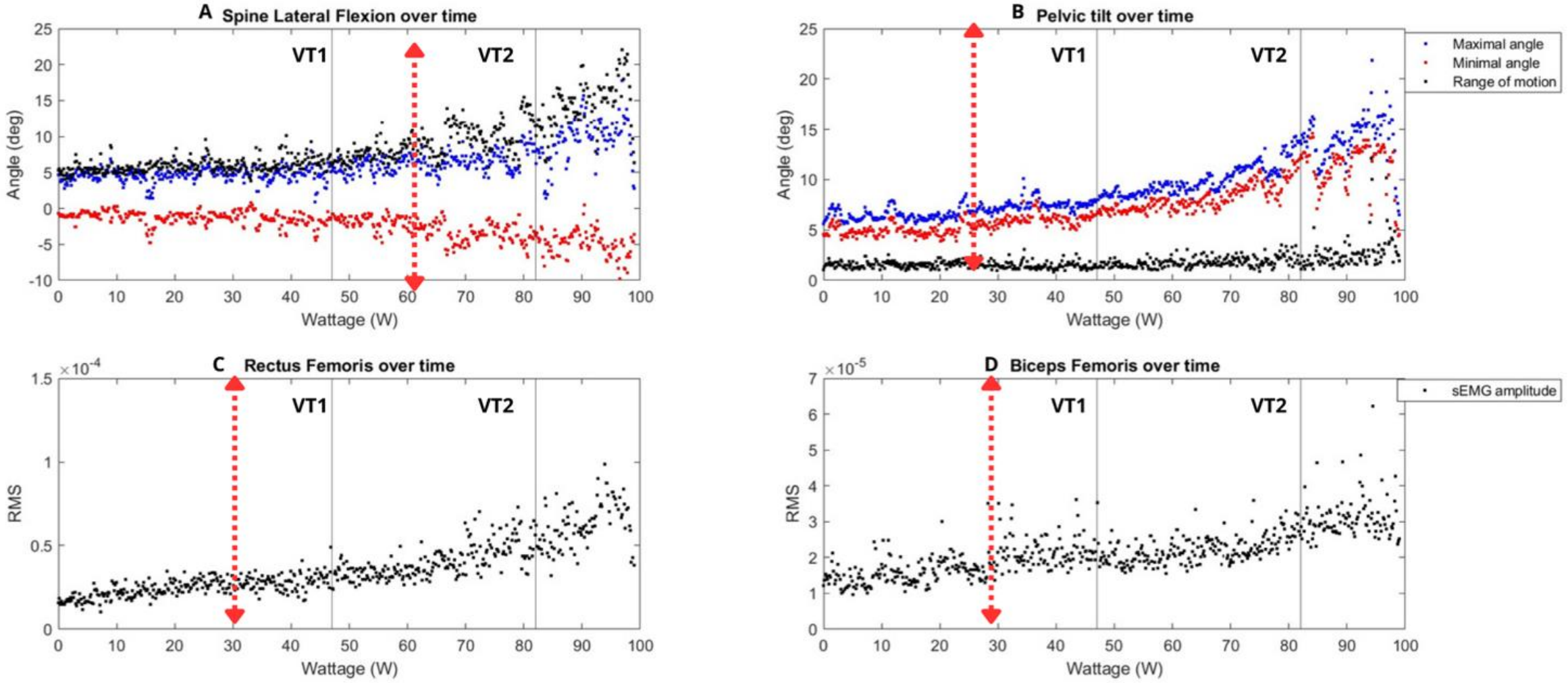

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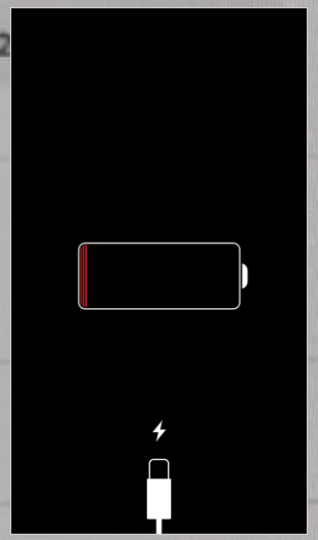
Body Battery



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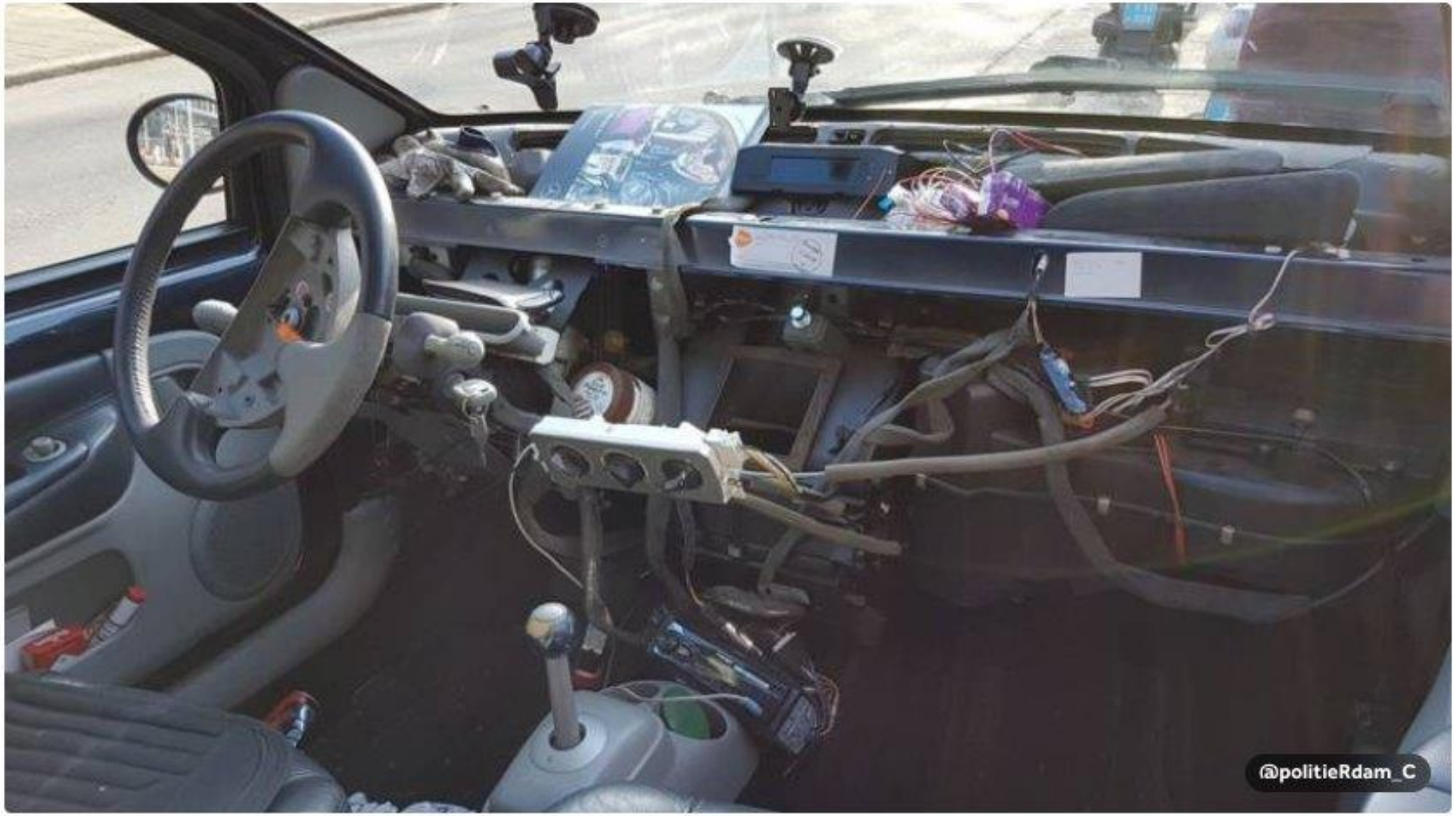
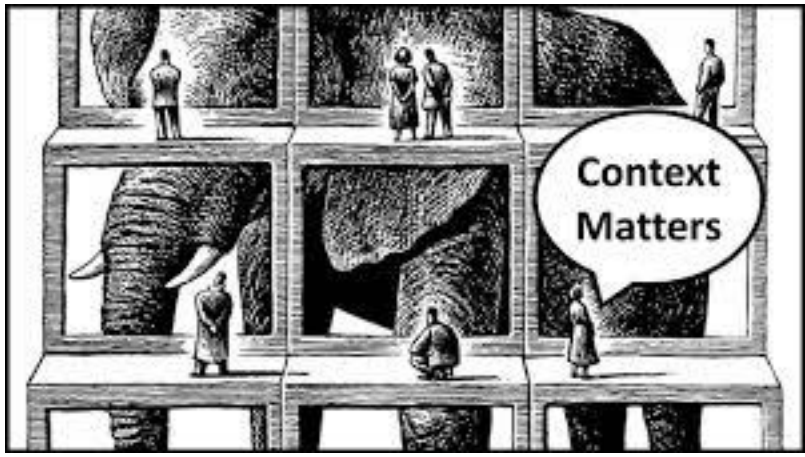


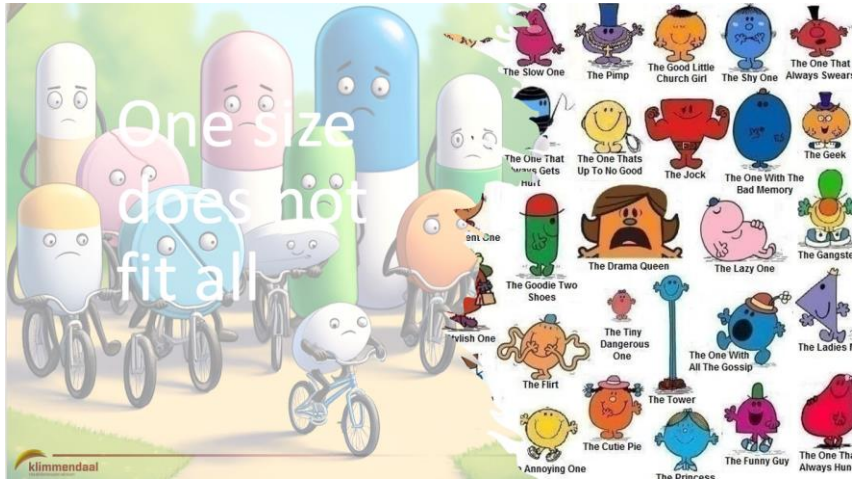
Do-it-yourselfer with a car without a dashboard taken off the road.

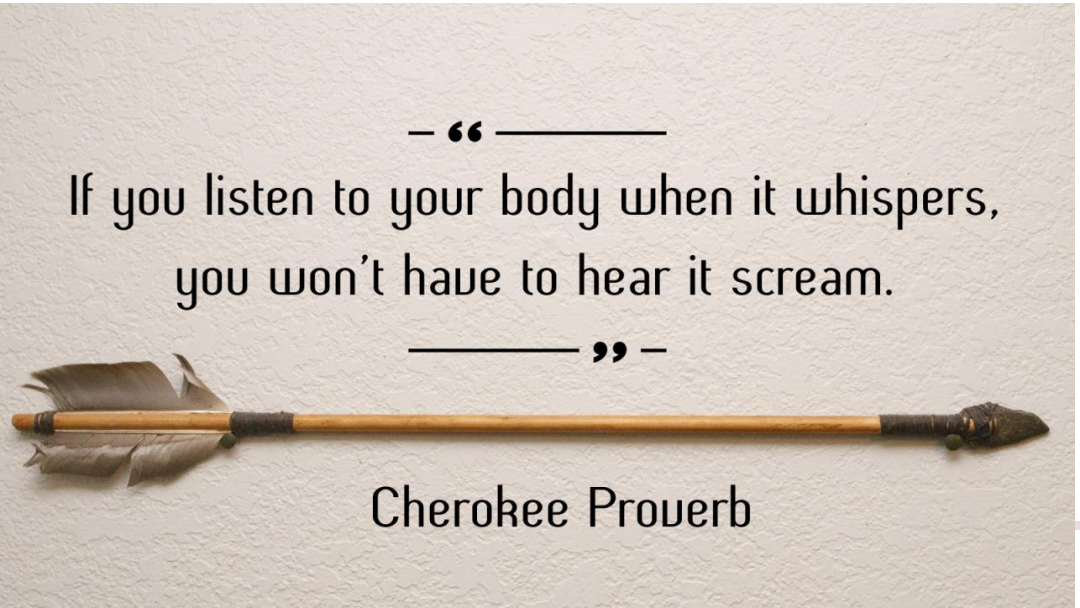
Muscle watch

Take home messages



- Start low and go slow
- One size does not fit all
- Context matters: training is more than just exercise
- Listen proactively to your body





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Network Neuromuscular Diseases (ERN EURO-NMD)

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Exercise in neuromuscular disorders

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