QUEEN SQUARE CNMD
Centre for Neuromuscular Diseases

MRC Centre for Neuromuscular Diseases

UCL

Physiotherapy approaches to supporting people with SBMA

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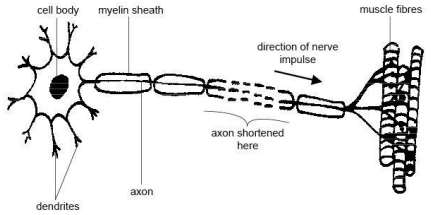
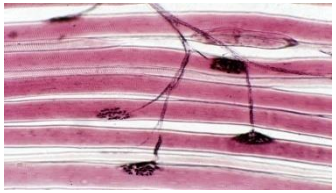
Neuromuscular Rehabilitation Research Team

Neuromuscular conditions

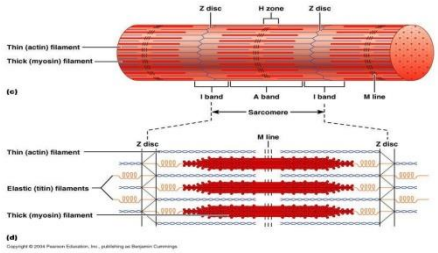
Anterior horn cell

Peripheral Nerves



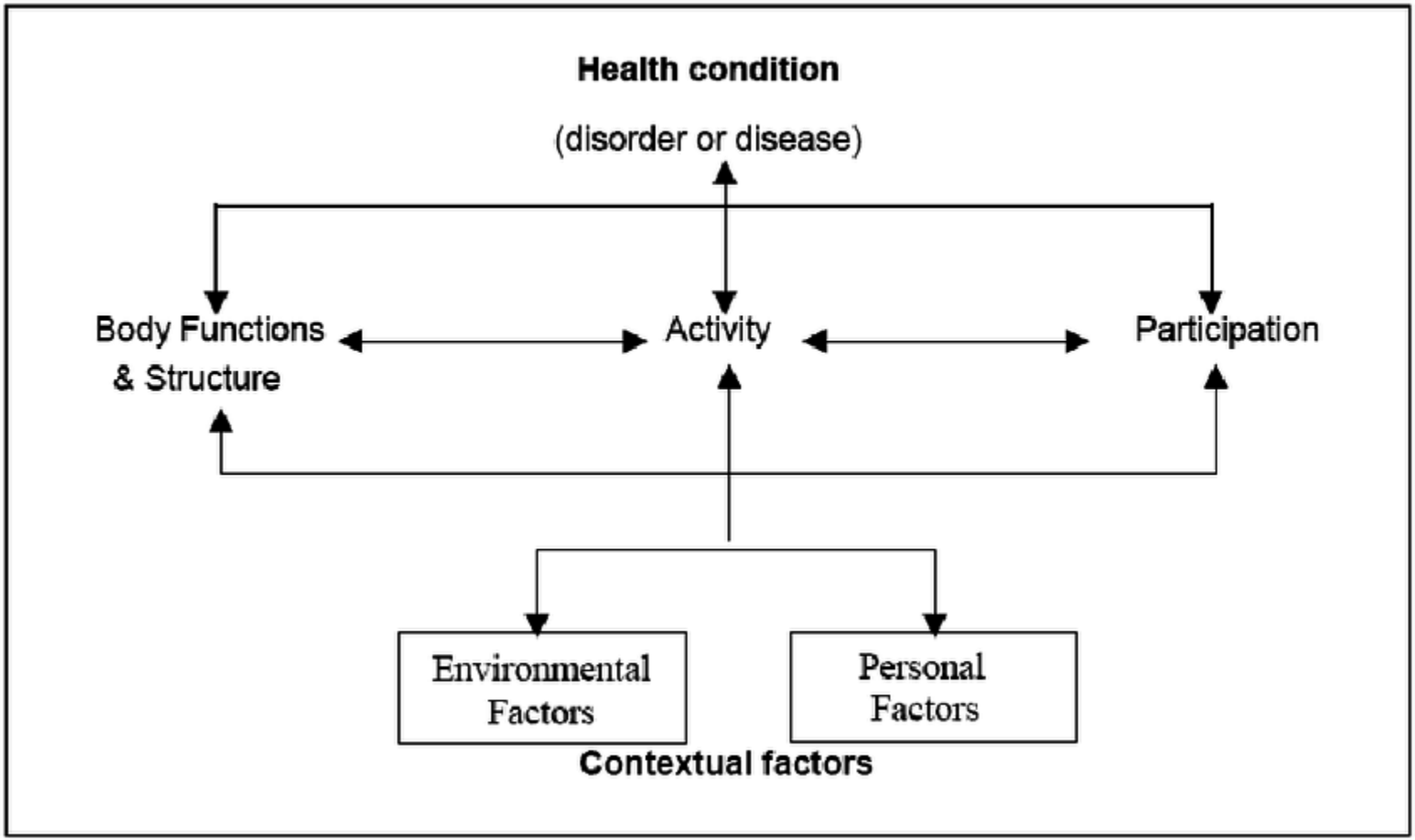


Neuromuscular junction



Muscle

WHO International Classification of Function, Disability and Health (ICF)



https://www.who.int/classifications/icf/en/

Applying the ICF to people with SBMA

Body Structure & Functions

Primary weakness

Secondary/disuse weakness

Fatigue & fatigability

Reduced fitness and de-conditioning

Pain

Reduced balance reactions

- Loss of sensation
- Muscle weakness

Activities and Participation

Difficulty climbing stairs

Reduced efficiency of walking

Daily tasks & work activities

Increased risk of falls

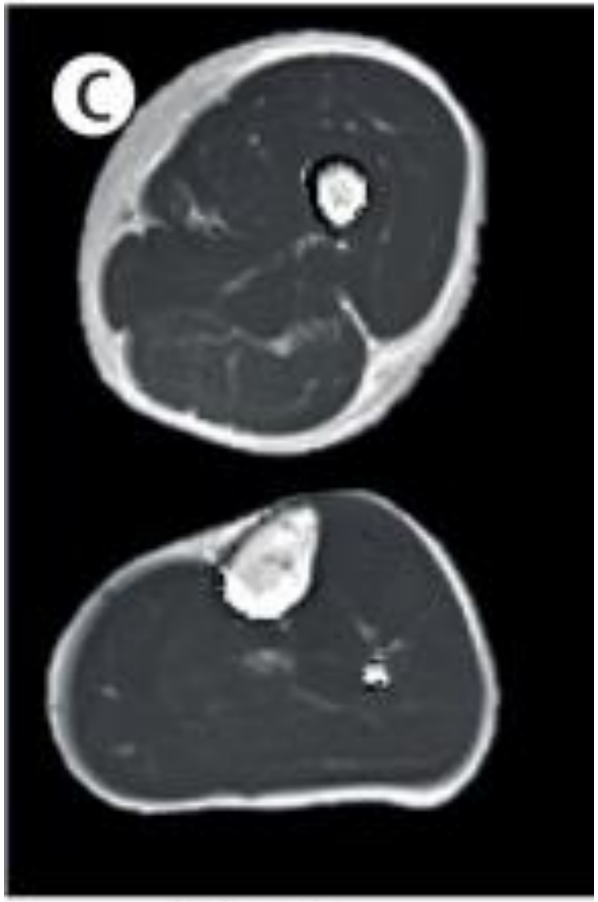
Community mobility

Maintaining work and family roles

Socialising

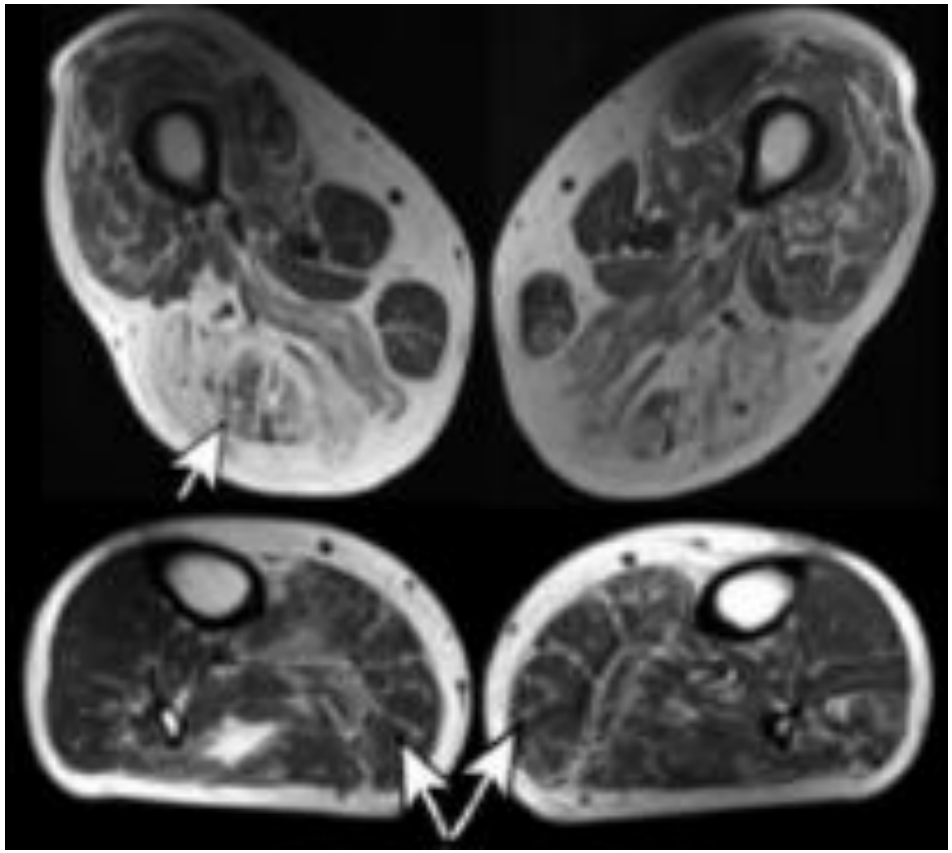
Leisure & sports performance

Primary and Secondary weakness



Control

(Morrow et al. Lancet Neurol, 2015 Nov 5. pii: S1474-4422(15)00242-2).



(Dahlqvist, J.R., Fornander, F., de Stricker Borch, J., Oestergaard, S.T., Poulsen, N.S. and Vissing, J. (2018), Ann Neurol., 84: 754-765.)

Reduced walking capacity

Takeuchi Y, Katsuno M, Banno H et al. Muscle Nerve. 2008 Aug;38(2):964-71

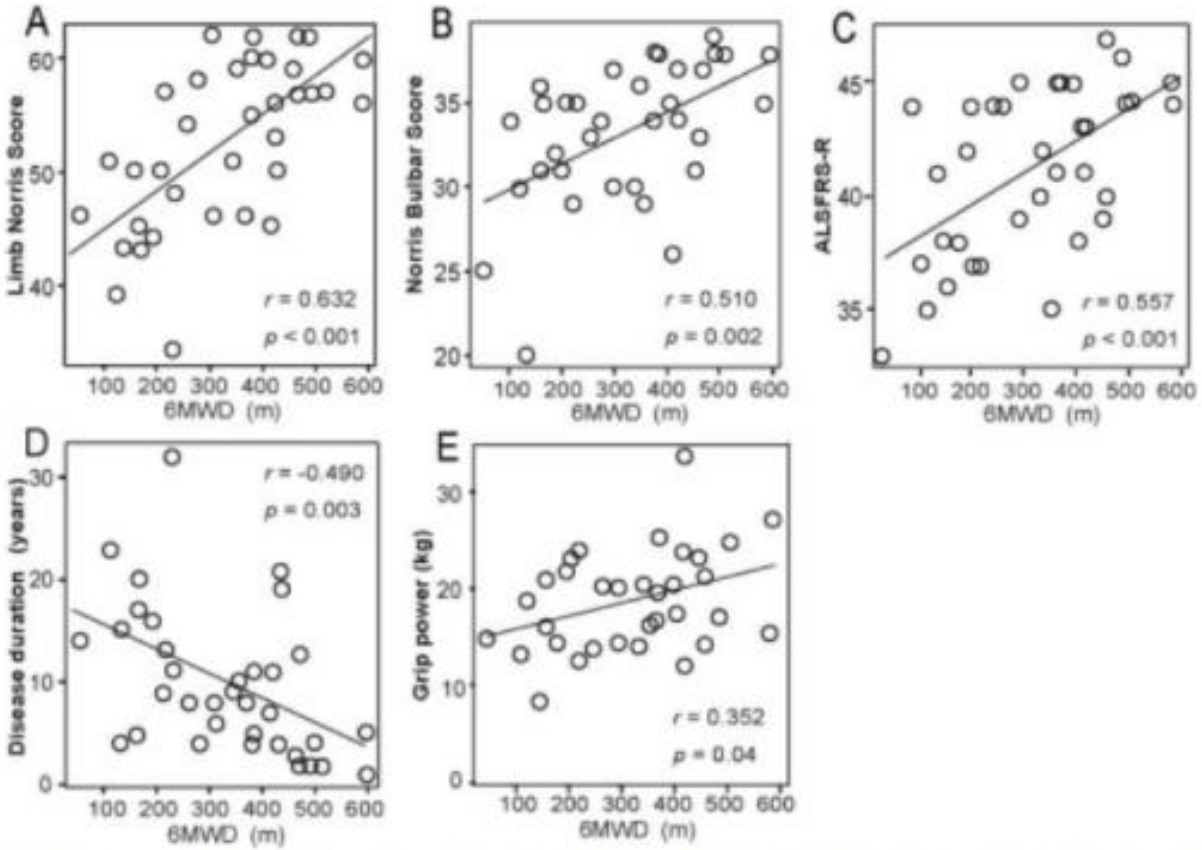
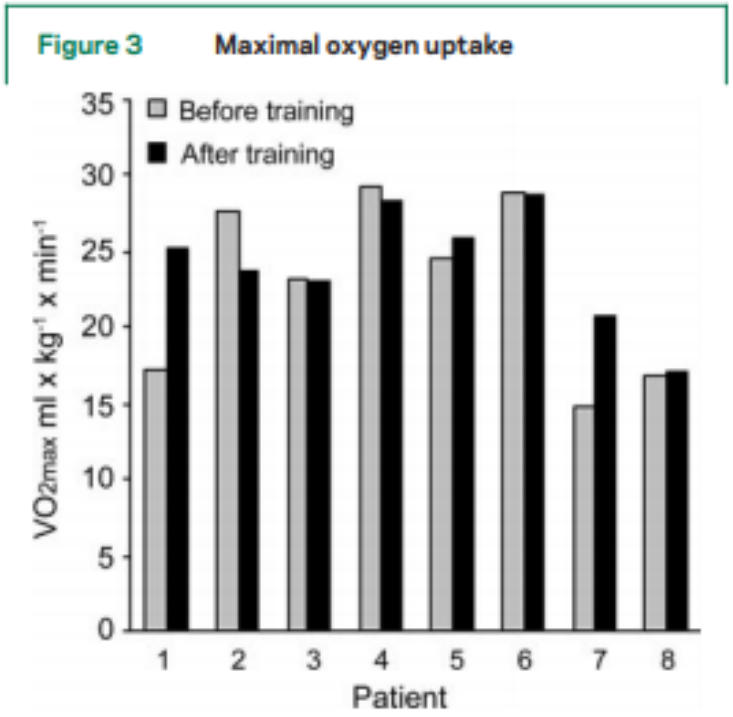


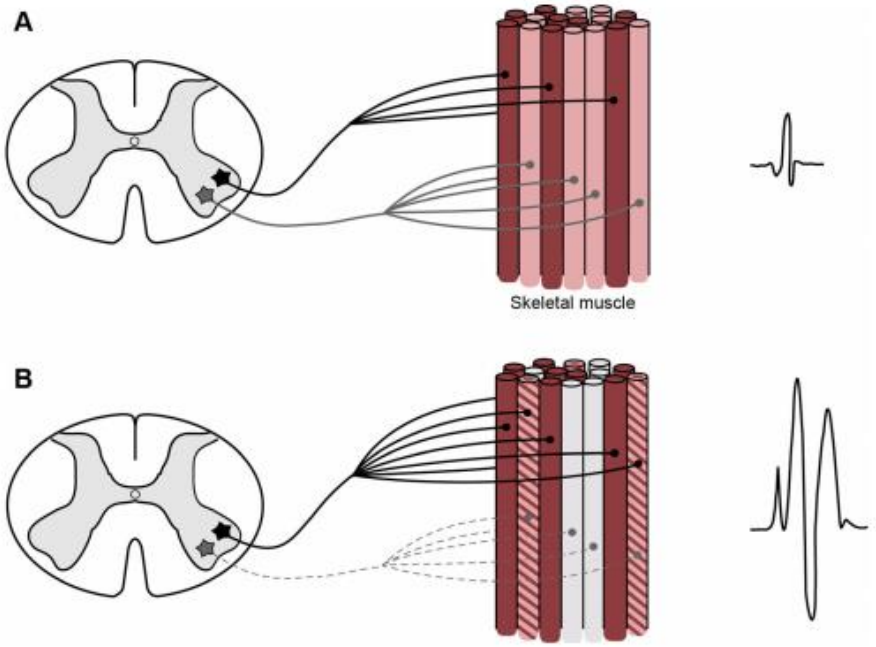
FIGURE 2. Correlation between the 6MWD and other measurements of motor function. **A–C:** The correlation between the 6MWD and general motor function. There was a significant correlation between the 6MWD and motor functional scales such as the Limb Norris Score **(A)**, the Norris Bulbar Score **(B)**, and the ALS functional rating scale-revised (ALSFRS-R, **C**). **D:** The value of the 6MWD was inversely correlated with disease duration. **E:** There was no correlation between the 6MWD and grip power. The value of grip power is shown as the average of left and right hands. 6MWD, six-min walk distance.

Exercise interventions: aerobic training



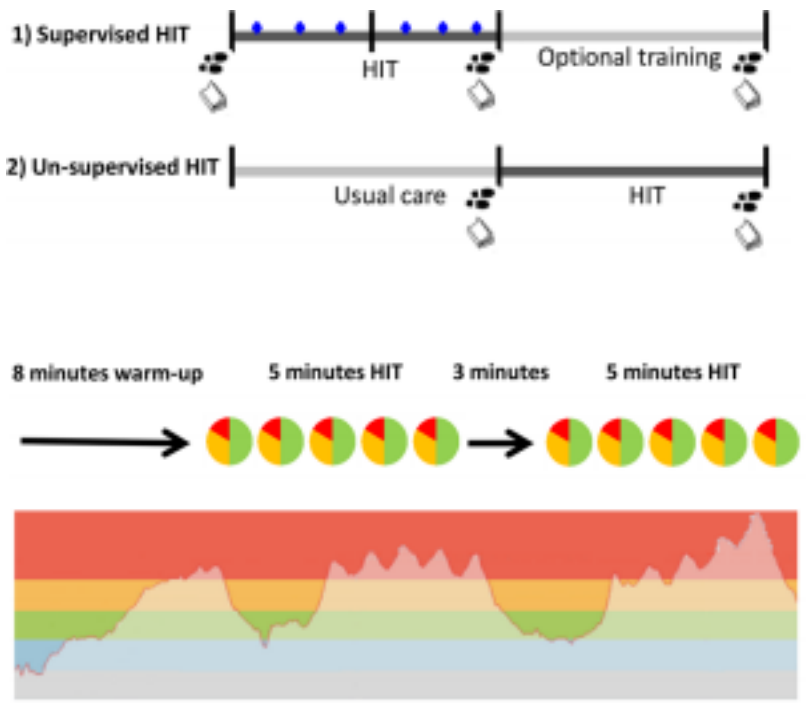
Maximal oxygen uptake (Vo_{2max}), before and after 12 weeks of training, shown individually for eight patients (1–8) with spinal and bulbar muscular atrophy. Patient number in this figure corresponds with patient number in the table.

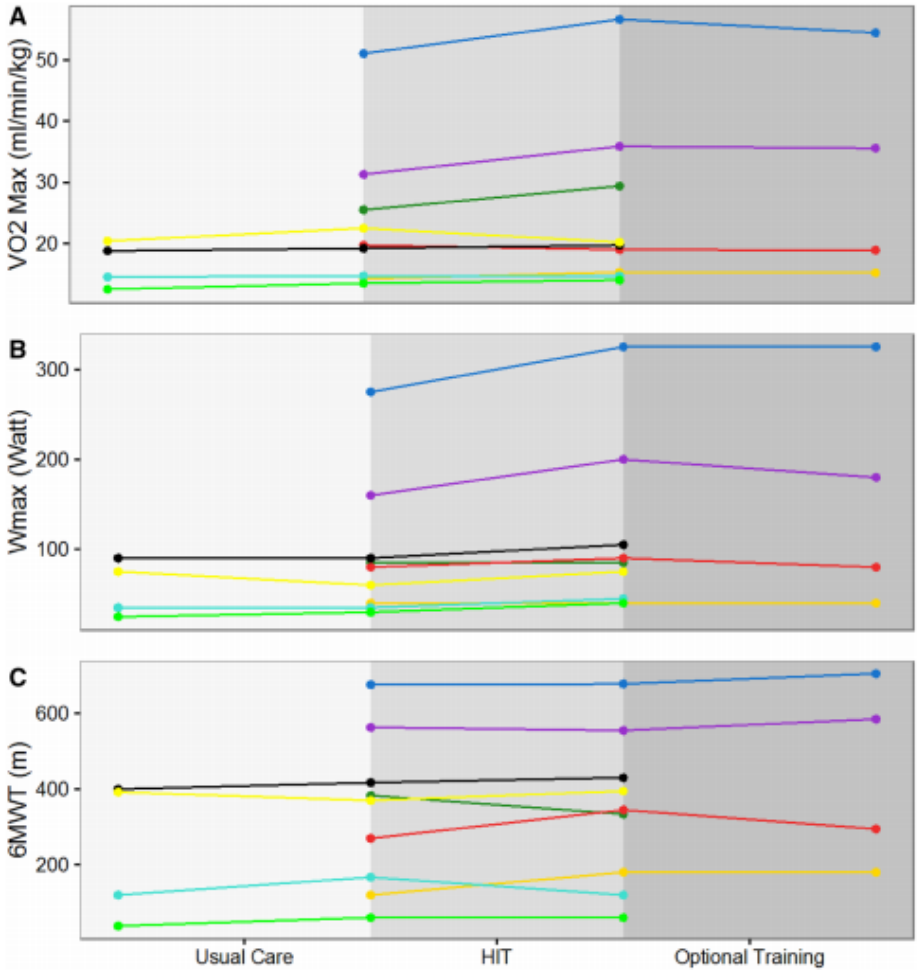
(N. Preisler, G. Andersen, F. Thøgersen, C. et al. Neurology Jan 2009, 72 (4) 317-32)



(Dahlqvist JR, Vissing J. J Mol Neurosci. 2016 Mar;58(3):388-93)

Exercise interventions: HIIT training





*(Heje, K., Andersen, G., Buch, A. et al. J Neurol **266,** 1693–1697 2019)*

Balance

Neuroscience Letters 705 (2019) 94–98

Contents lists available at ScienceDirect

Neuroscience Letters

journal homepage: www.elsevier.com/locate/neulet

Research article

Disentangling balance impairments in spinal and bulbar muscular atrophy

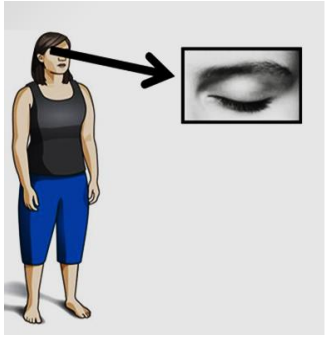
Anagnostou E*, Zachou A, Breza M, Kladi A, Karadima G, Koutsis G

Department of Neurology, University of Athens, Eginition Hospital, Athens, Greece

Standing balance data.

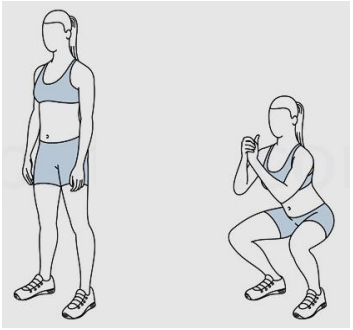
	SUBJECTS	Firm surface / eyes open (mm^2)	Firm surface / eyes closed (mm^2)	Foam pad / eyes open (mm^2)	Foam pad / eyes closed (mm^2)	RQ	vRQ
Kennedy patients	1	831.7	2591.4	609.8	7973.1	3.1	13.1
	2	182.2	2384.3	703.8	6006.9	13.1	8.5
	3	312.1	181.9	131.9	625.9	0.6	4.7
	4	408.9	2875.1	278.5	3681.5	7.0	13.2
	5	210.3	313.7	638.2	1048.9	1.5	1.6
	6	114.9	413.7	187.4	460.5	3.6	2.5
	7	335.9	2591.4	609.8	7973.1	2.1	2.7
	Mean ± SD	342.3 ± 237.9	1350.7 ± 1202.8**	410.1 ± 234.6	2953.2 ± 3017.4*	4.4 ± 4.3*	6.6 ± 5.0
Control subjects	Mean ± SD	224.9 ± 89.9	368.5 ± 171.4	259.8 ± 139.2	1160.7 ± 562.1	1.81 ± 0.81	5.7 ± 3.6
	Upper normal limit[a]	347.8	583.8	426.3	1075.9	2.7	7.0

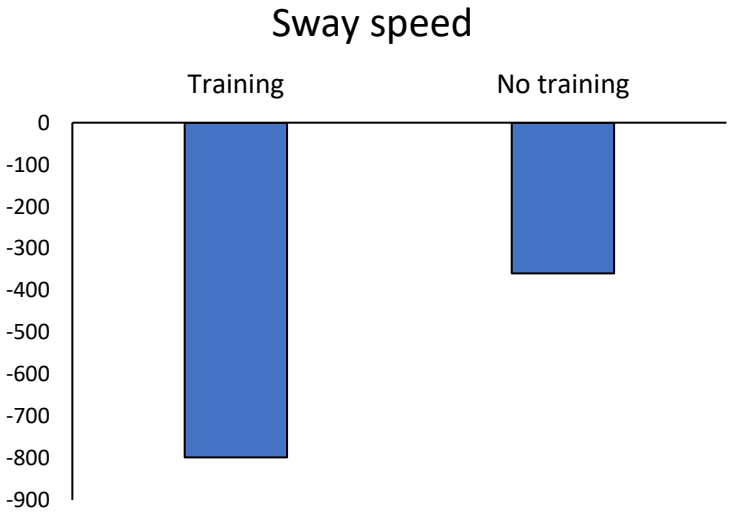
Improvements in balance

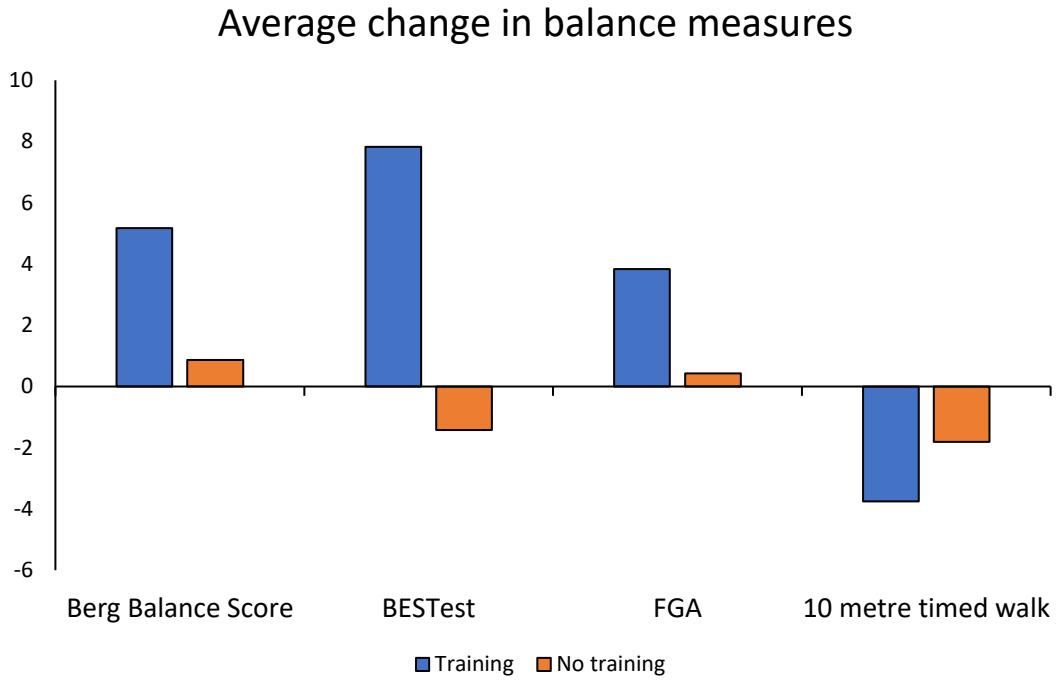


Home based balance training in 15 people with genetic neuropathy

Strength and multi-sensory balance exercises for 12 weeks







(Dudziec et al. *in preparation*)

Pilates for people with Neuromuscular conditions

4 videos • 4,637 views • Last updated on Jun 10, 2020

These Pilates exercises are designed for people with Neuromuscular conditions. Please contact your physiotherapist if you have any questions.

1 

Pilates video in standing for people with Neuromuscular conditions

University College London Hospitals NHS Foundation Trust

2 

Pilates video in sitting for people with Neuromuscular conditions

University College London Hospitals NHS Foundation Trust

3 

Pilates video in prone for people with Neuromuscular conditions

University College London Hospitals NHS Foundation Trust

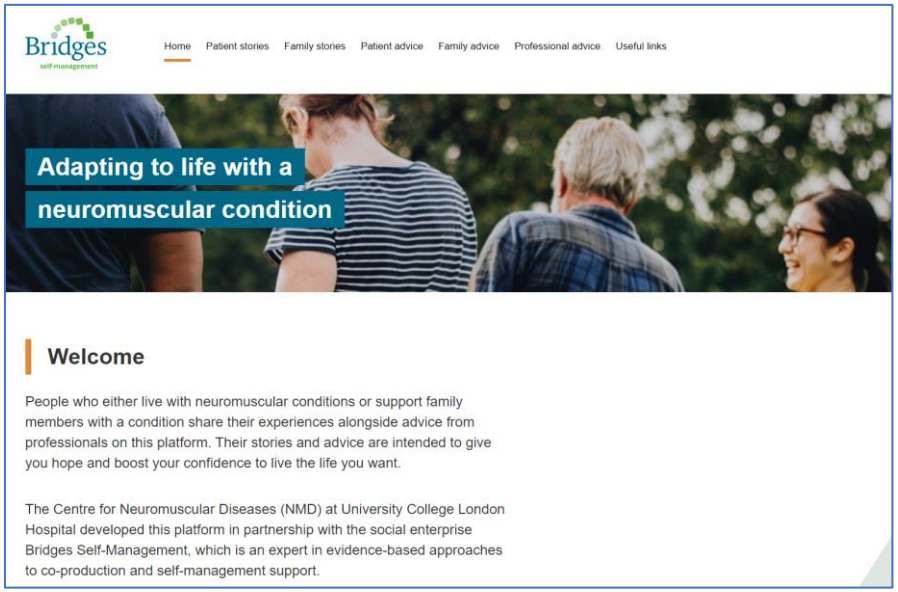
4 

Pilates video in lying for people with Neuromuscular conditions

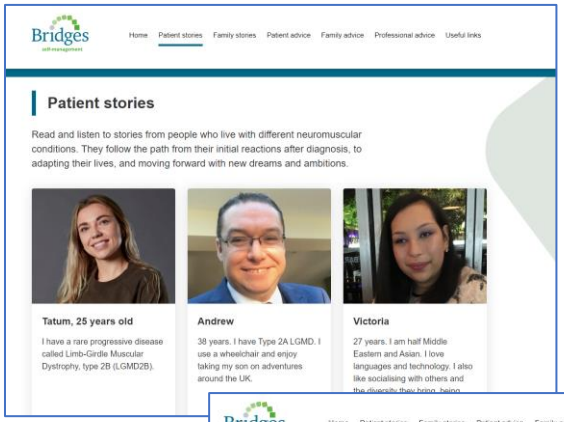
University College London Hospitals NHS Foundation Trust

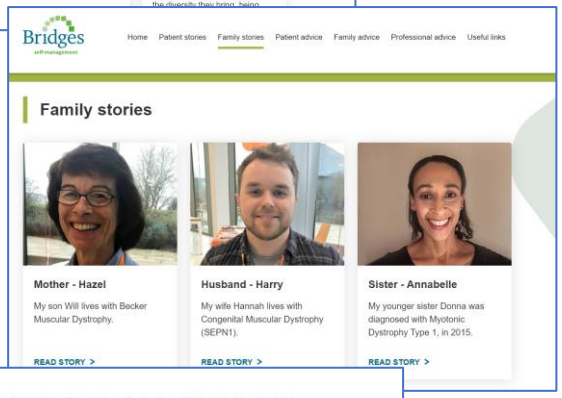
https://www.youtube.com/playlist?list=PLazCbfp_tqxyve043vSch45aPfMzFfHxX

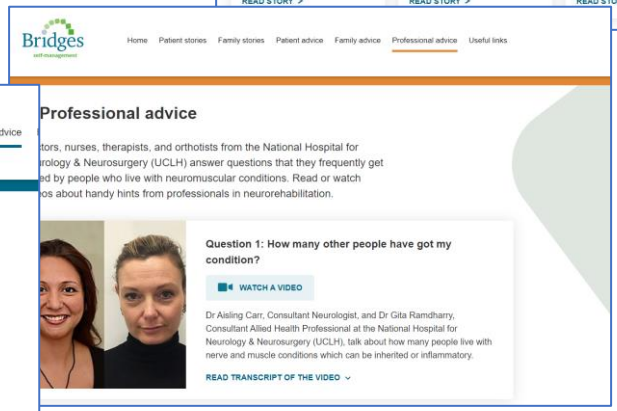
Supported Self-Management



https://nmd.bridgesselfmanagement.org.uk/







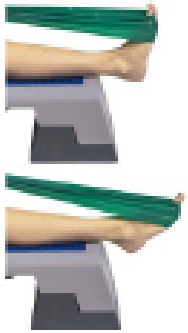
Case:

- **72 year old man,15 year history of slowly progressive disease** (started with cramps)
- **Gait:** Flat gait, short stride length, pelvic drop on stance
- **Balance:** Unable to stand still. Tends to fall backwards
- **Range of motion:** restriction at ankles, 5˚ from plantar grade

- **Weakness** Left lower limb Unable to walk on toes over the last year
- **CNS involvement:** None
- **Respiratory impairment:** No
  No swallowing issues but eats slowly. No choking. No chest infections.
- **Cardiac Involvement:** Yes
  Left ventricular hypertrophy. On betablockers
- **Fatigue:** Yes Not a good sleeper: wake 3-4 times a night
  Details: Activity related fatigue
- **Pain:** Yes
  Mild arthitis in fingers
- **Reduced balance** Yes
  Details: Standing still difficult.
- **Falls**: Yes Falls at least once a month
  Most common causes: Turns and legs give way
  Previous injuries: Broke little toe 2 years ago. Difficulty getting off the floor.
- **Reduced exercise tolerance:** Yes
  Limited by Fatigue
- **CURRENT MOBILITY LEVEL:** Independent
  Details: Can walk for 1-2 miles slowly with trekking poles.
- **Current activity or exercise:** Swims three times a week, gardening and DIY

	Right	Left
Shoulder abduction	5	5
Elbow flexion	5	5
Elbow extension	5	5
Wrist extension	5	5
Finger flexion	5	5
Thumb abduction	5	4
Hip flexion	5	
Hip extension	5	5
Knee flexion	4	4-
Knee extension	4+	4-
Plantarflexion	4-	4-
Dorsiflexion	5	5
	Right	Left
Interossei	5	5
Abductor digiti minimi	5	4

Case:



- Strength/resistance training
  - Baseline strength >4 MRC score
- Reviewing & planning physical activity
- Balance: motor impairment
- Orthotics

Acknowledgements

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