



QUEEN SQUARE  
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Neuromuscular Diseases



**UCL**



# Physiotherapy approaches to supporting people with SBMA

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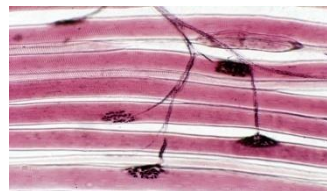
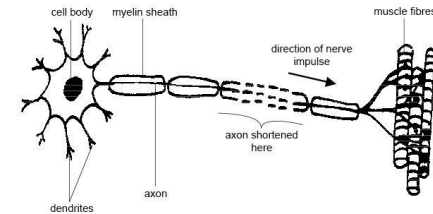
Neuromuscular Rehabilitation  
Research Team

# Neuromuscular conditions

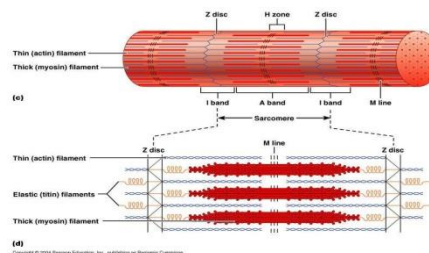
Anterior horn cell



Peripheral Nerves

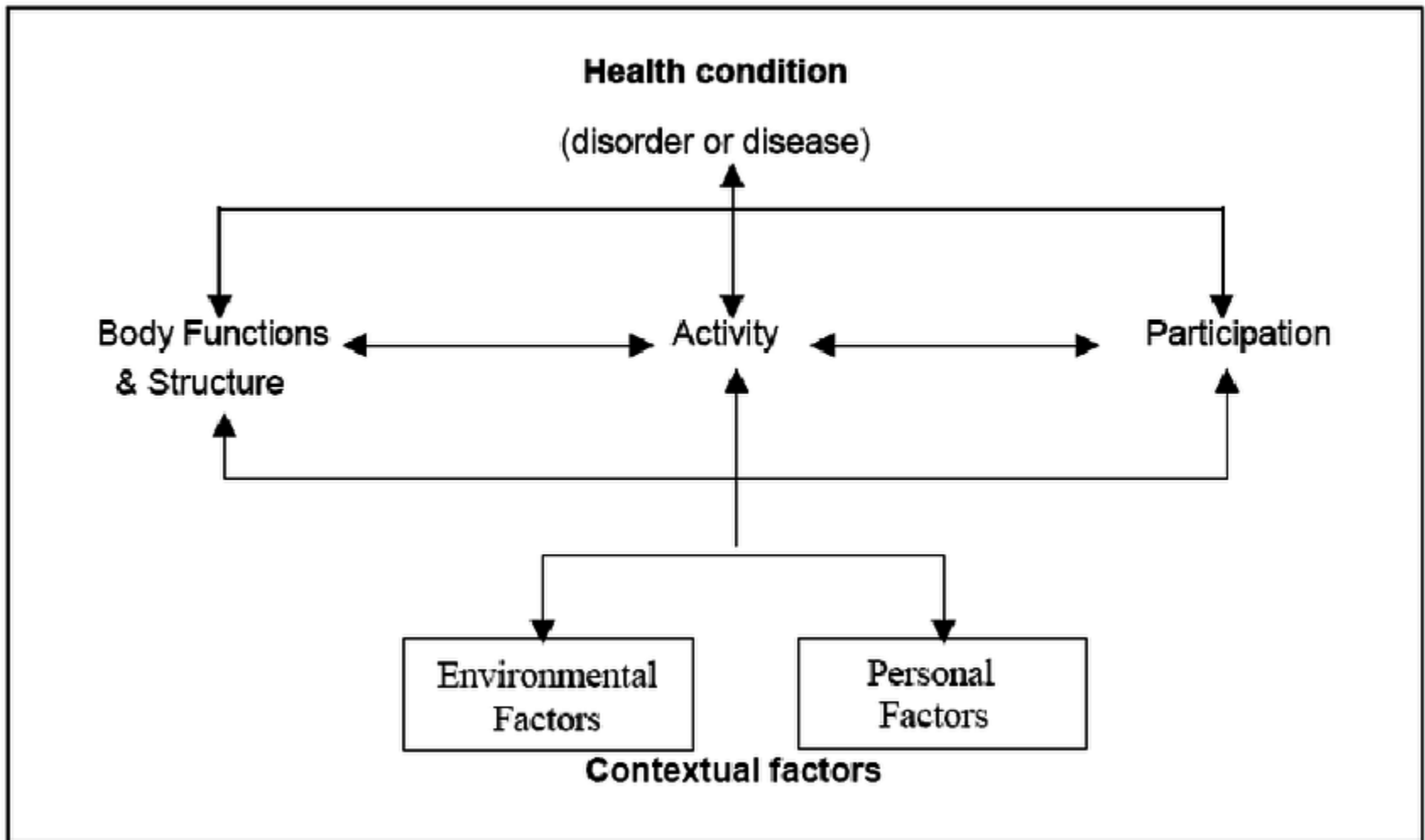


Neuromuscular junction



Muscle

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



# 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



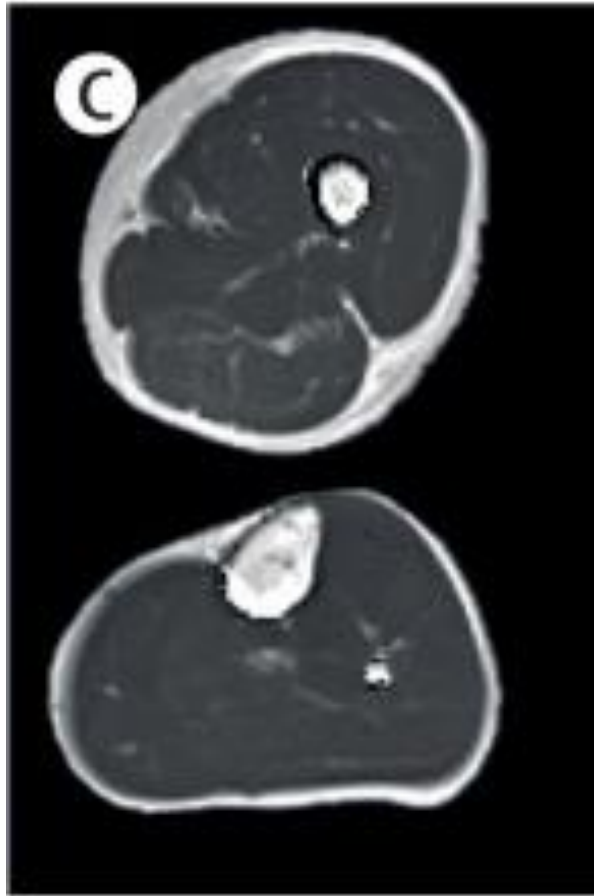
## Activities and Participation

Difficulty climbing stairs  
Reduced efficiency of walking  
Daily tasks & work activities  
Increased risk of falls



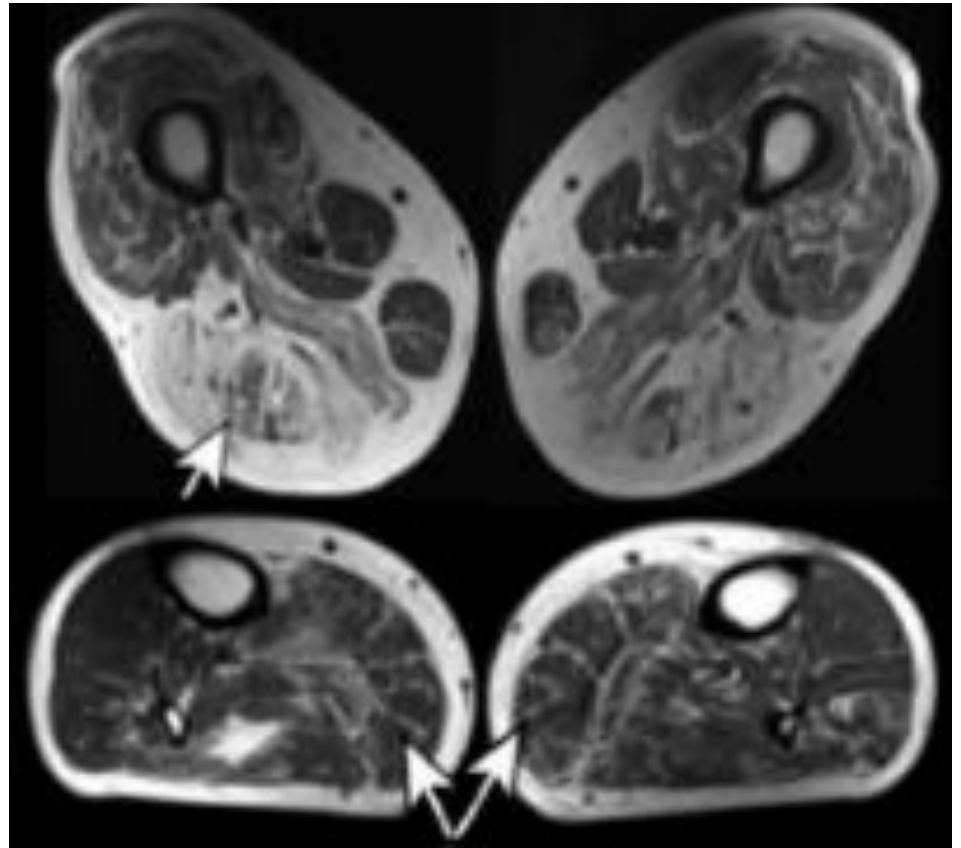
Community mobility  
Maintaining work and family roles  
Socialising  
Leisure & sports performance

# Primary and Secondary weakness



Control

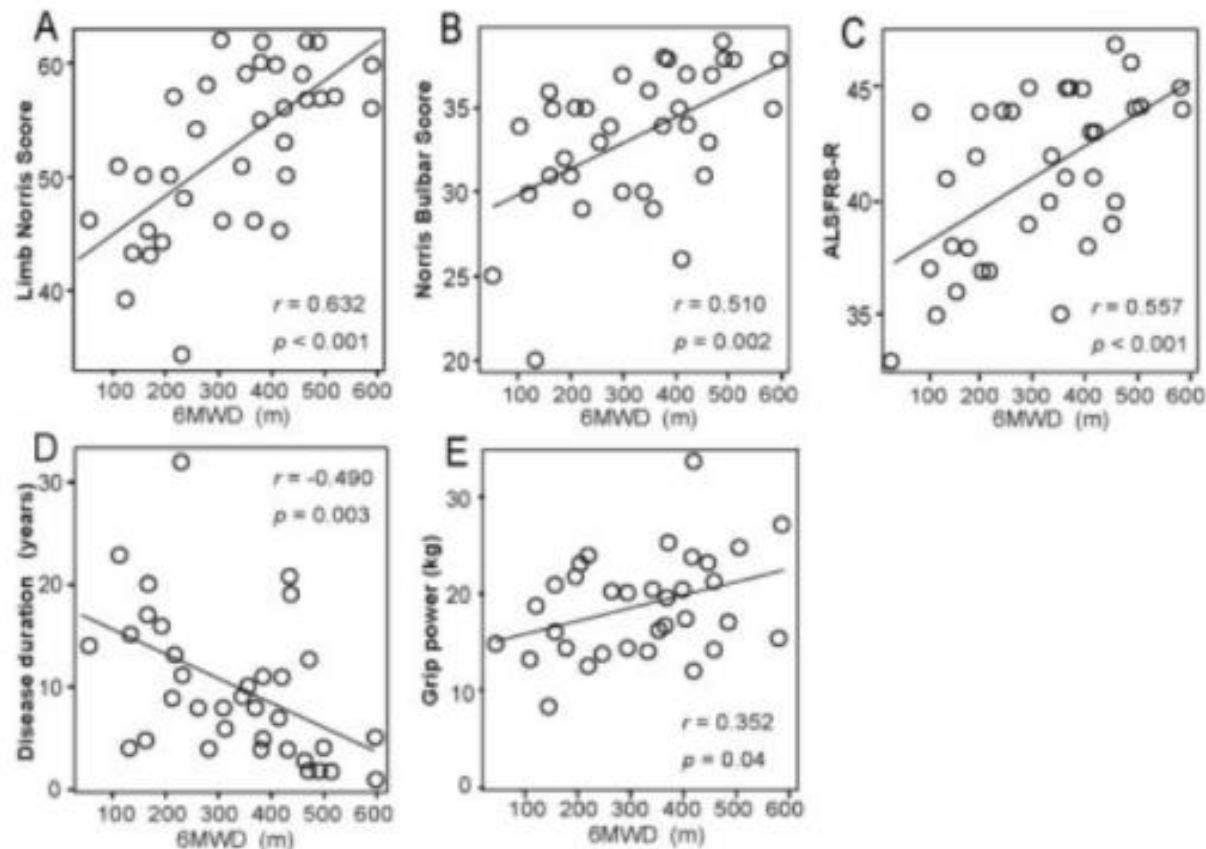
(Morrow et al. *Lancet Neurol*, 2015  
Nov 5. pii: S1474-4422(15)00242-2).



(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

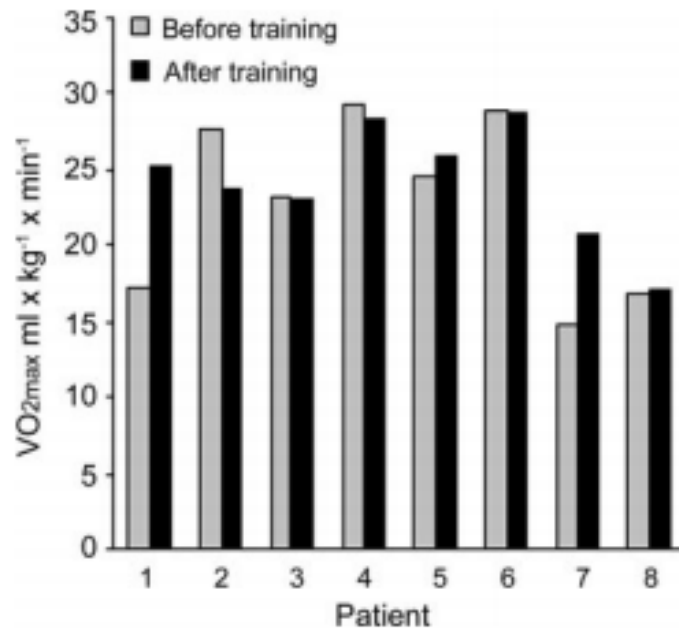


**FIGURE 2.** Correlation between the 6MWD and other measurements of motor function. **A–C:** The correlation between the 6MWD and general motor function. There was a significant correlation between the 6MWD and motor functional scales such as the Limb Norris Score (**A**), the Norris Bulbar Score (**B**), 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.



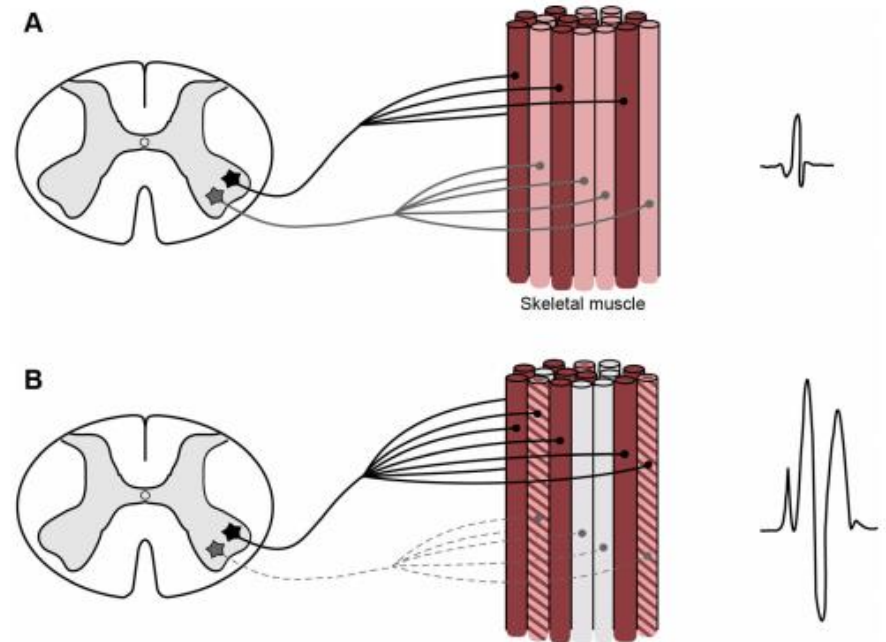
# Exercise interventions: aerobic training

**Figure 3** Maximal oxygen uptake



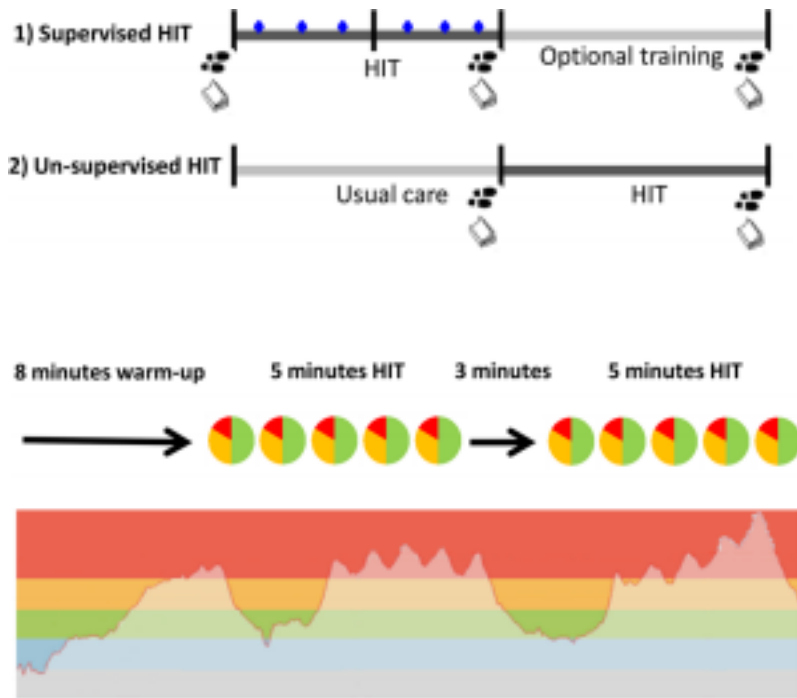
Maximal oxygen uptake ( $VO_{2max}$ ), before and after 12 weeks of training, shown individually for eight patients (1-8) with spinal and bulbar muscular atrophy. Patient number in this figure corresponds with patient number in the table.

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

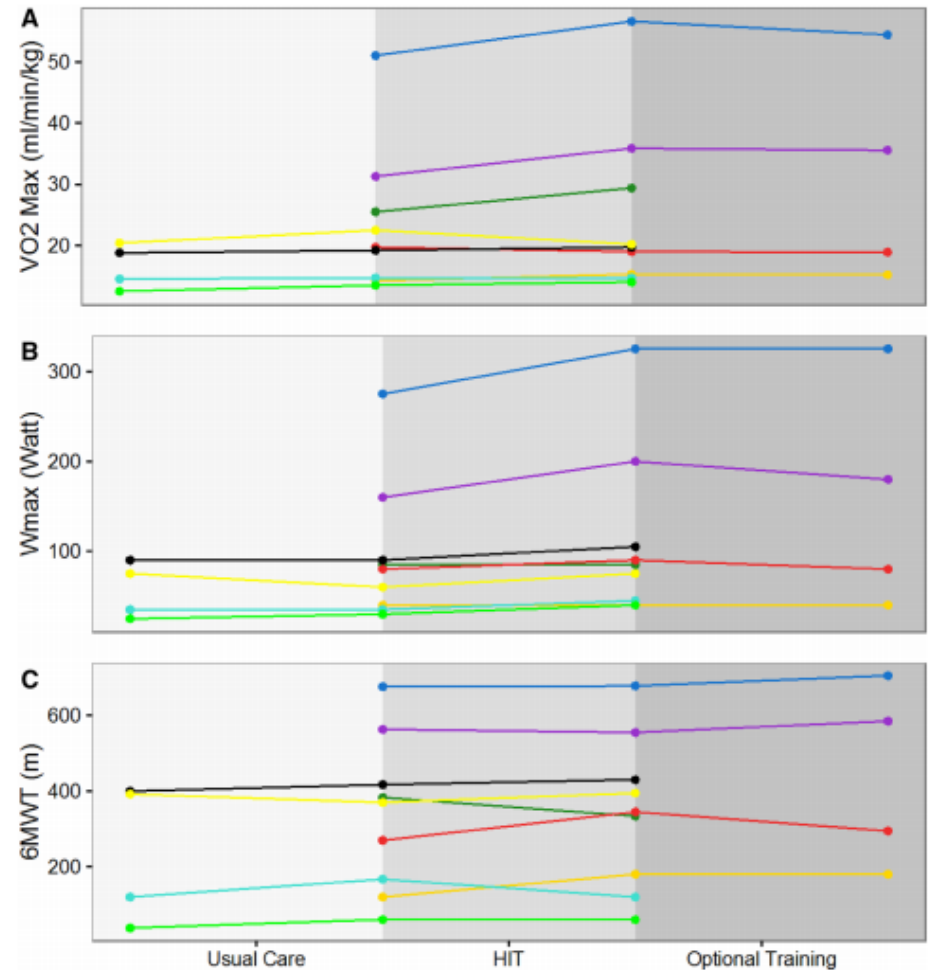


(Dahlgqvist JR, Vissing J. *J Mol Neurosci.* 2016 Mar;58(3):388-93)

# Exercise interventions: HIIT training



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





# Balance



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

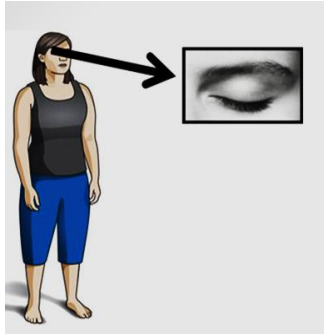


Standing balance data.

SUBJECTS		Firm surface / eyes open (mm <sup>2</sup> )	Firm surface / eyes closed (mm <sup>2</sup> )	Foam pad / eyes open (mm <sup>2</sup> )	Foam pad / eyes closed (mm <sup>2</sup> )	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 <sup>a</sup>	347.8	583.8	426.3	1075.9	2.7	7.0

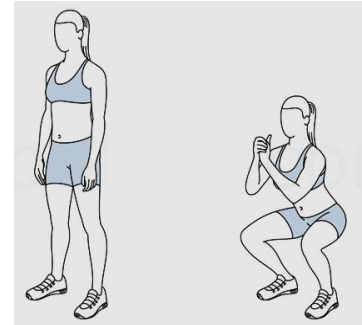


# Improvements in balance



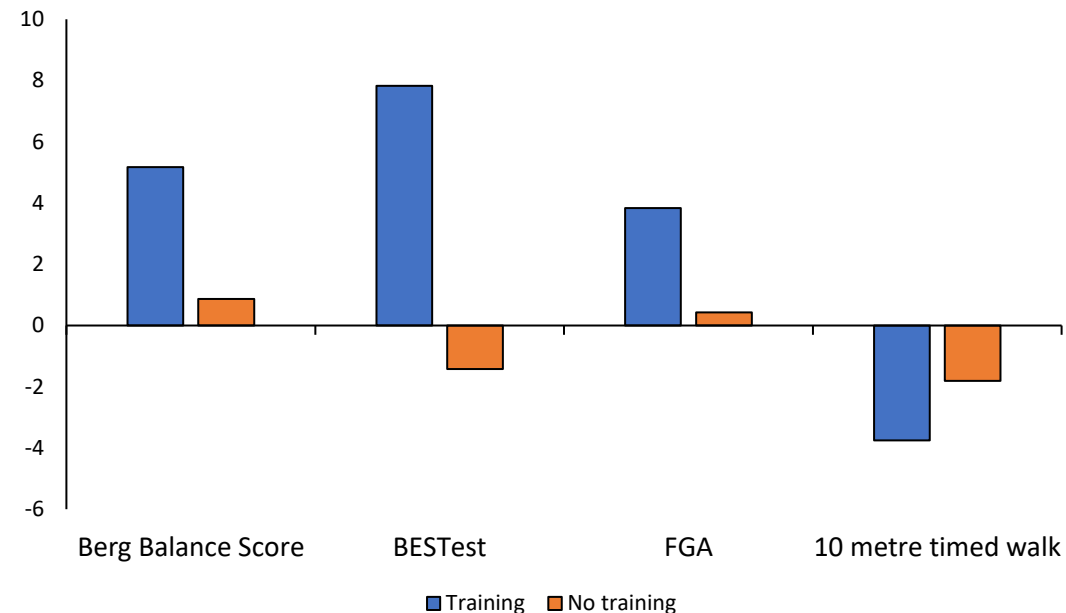
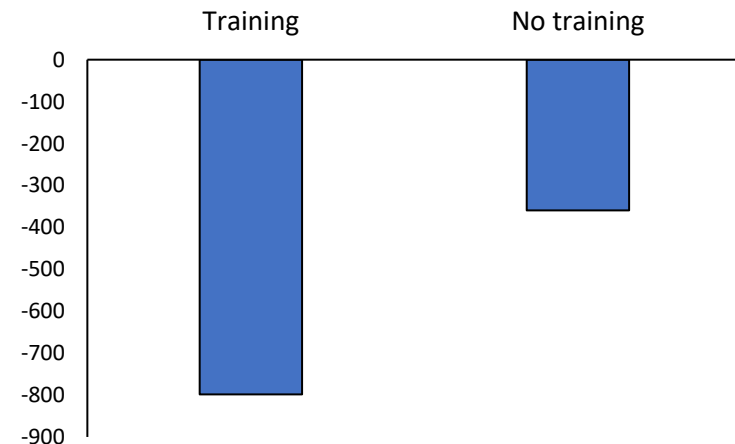
Home based balance training in 15 people with genetic neuropathy

Strength and multi-sensory balance exercises for 12 weeks



Average change in balance measures

Sway speed



(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

8:16

2



### Pilates video in sitting for people with Neuromuscular conditions

University College London Hospitals NHS Foundation Trust

10:23

3



### Pilates video in prone for people with Neuromuscular conditions

University College London Hospitals NHS Foundation Trust

10:21

4




### Pilates video in lying for people with Neuromuscular conditions

University College London Hospitals NHS Foundation Trust


11:16

[https://www.youtube.com/playlist?list=PLazCbfp\\_tqxyve043vSch45aPfMzFfHxX](https://www.youtube.com/playlist?list=PLazCbfp_tqxyve043vSch45aPfMzFfHxX)

# Supported Self-Management



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## Adapting to life with a neuromuscular condition

### Welcome

People who either live with neuromuscular conditions or support family members with a condition share their experiences alongside advice from professionals on this platform. Their stories and advice are intended to give you hope and boost your confidence to live the life you want.

The Centre for Neuromuscular Diseases (NMD) at University College London Hospital developed this platform in partnership with the social enterprise Bridges Self-Management, which is an expert in evidence-based approaches to co-production and self-management support.


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




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### Patient stories


Read and listen to stories from people who live with different neuromuscular conditions. They follow the path from their initial reactions after diagnosis, to adapting their lives, and moving forward with new dreams and ambitions.




**Tatum, 25 years old**  
I have a rare progressive disease called Limb-Girdle Muscular Dystrophy, type 2B (LGMD2B).



**Andrew**  
38 years. I have Type 2A LGMD. I use a wheelchair and enjoy taking my son on adventures around the UK.




**Victoria**  
27 years. I am half Middle Eastern and Asian. I love languages and technology. I also like socialising with others and the diversity that brings.




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### Family stories



**Mother - Hazel**  
My son Will lives with Becker Muscular Dystrophy.



**Husband - Harry**  
My wife Hannah lives with Congenital Muscular Dystrophy (SEPN1).



**Sister - Annabelle**  
My younger sister Donna was diagnosed with Myotonic Dystrophy Type 1, in 2015.

[READ STORY >](#) [READ STORY >](#) [READ STORY >](#)




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### Family advice

#### Family advice topics

- Topic 1: Knowing how much to help
- Topic 2: Being patient – understanding the person
- Topic 3: Being more of a family member than a carer
- Topic 4: Dealing with difficult emotions and negativity




Home Patient stories Family stories Patient advice

### Patient advice

#### Patient advice topics


- Topic 1: Newly diagnosed
- Topic 2: Fatigue and physical weakness
- Topic 3: Practical solutions
- Topic 4: Emotional and other support
- Topic 5: Staying positive
- Topic 6: Friendships, dating and relationships



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### Professional advice

tors, nurses, therapists, and orthotists from the National Hospital for Neurology & Neurosurgery (UCLH) answer questions that they frequently get asked by people who live with neuromuscular conditions. Read or watch videos about handy hints from professionals in neurorehabilitation.



**Question 1: How many other people have got my condition?**

[WATCH A VIDEO](#)

Dr Aisling Carr, Consultant Neurologist, and Dr Gila Ramsharny, Consultant Allied Health Professional at the National Hospital for Neurology & Neurosurgery (UCLH), talk about how many people live with nerve and muscle conditions which can be inherited or inflammatory.

[READ TRANSCRIPT OF THE VIDEO](#)

# 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

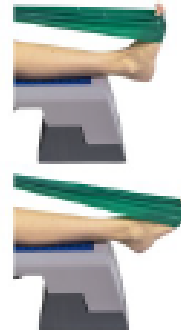
- **Weakness** Left lower limb Unable to walk on toes over the last year
- **CNS involvement:** None
- **Respiratory impairment:** No  
No swallowing issues but eats slowly. No choking. No chest infections.
- **Cardiac Involvement:** Yes  
Left ventricular hypertrophy. On betablockers
- **Fatigue:** Yes Not a good sleeper: wake 3-4 times a night  
Details: Activity related fatigue
- **Pain:** Yes  
Mild arthritis 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





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Research Nurses



Neuromuscular Rehabilitation Research Group  
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