

















### What do we know?

- Disease progression
  - Breathing difficulties are more common in people living with LGMD R9 than other forms of LGMD

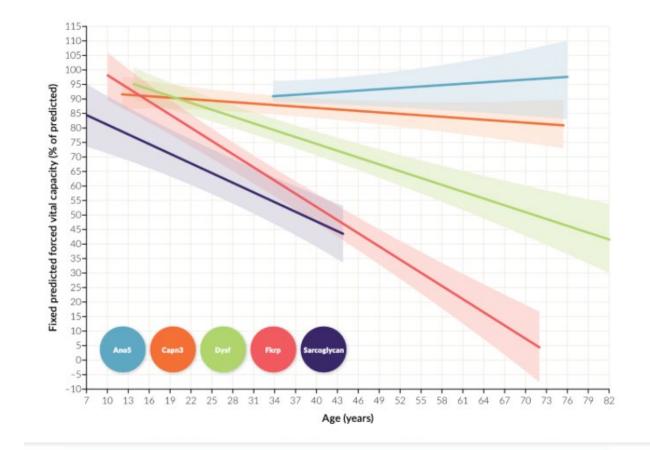
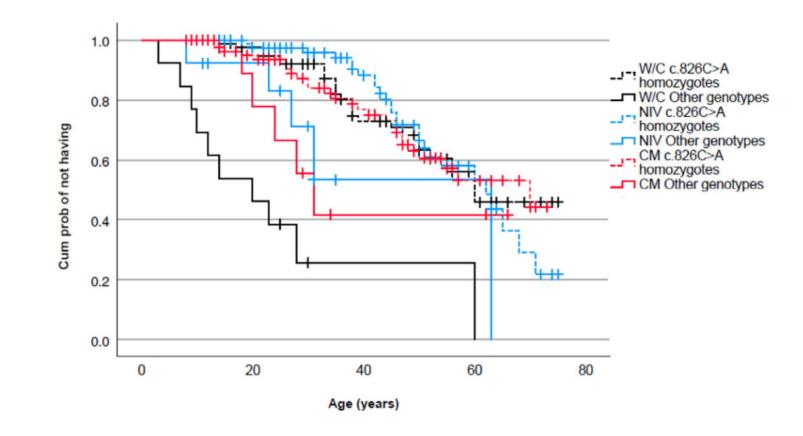


Figure 1: Progression of forced vital capacity percentage predicted values during the follow-up

Muni-Lofra R, Juanola-Mayos E, Schiava M, Moat D, Elseed M, Michel-Sodhi J, et al. Longitudinal Analysis of Respiratory Function of Different Types of Limb Girdle Muscular Dystrophies Reveals Independent Trajectories. Neurol Genet [Internet]. 2023 [cited 2023 Aug 22];9(4):1–12. Available from: https://pubmed.ncbi.nlm.nih.gov/37440793/

## Respiratory Management Guidelines

- Disease progression
  - Having a specific genotype will result in later respiratory difficulties



Jensen SM, Müller KI, Mellgren SI, Bindoff LA, Rasmussen M, Ørstavik K, et al. Epidemiology and natural history in 101 subjects with FKRP-related limb-girdle muscular dystrophy R9. The Norwegian LGMDR9 cohort study (2020). Neuromuscular Disorders. 2023 Feb 1;33(2):119–32.

## What do we know?

- Disease progression
  - Difficulties clearing secretions can be part of the consequences of limited breathing capacity

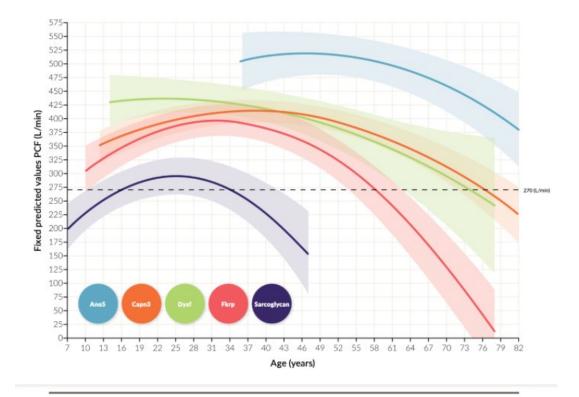


Figure 2: Progression of peak cough flow percentage predicted values during the follow-up

Muni-Lofra R, Juanola-Mayos E, Schiava M, Moat D, Elseed M, Michel-Sodhi J, et al. Longitudinal Analysis of Respiratory Function of Different Types of Limb Girdle Muscular Dystrophies Reveals Independent Trajectories. Neurol Genet [Internet]. 2023 [cited 2023 Aug 22];9(4):1–12. Available from: https://pubmed.ncbi.nlm.nih.gov/37440793/

## How can we monitor this?

#### **Check your breathing function**



- Symptoms:
  - Fatigue, headaches (specially morning), concentration difficulties, school/work performances difficulties, memory changes, shortness of breath, weakness, snoring/gasping/witness apneas, chest infections requiring hospital admission

#### Frequency

- At least once a year
- Every 6 months if you have any difficulties

# What should we do?

- Stay active
  - Aerobic exercise
  - Breathing exercises
- Non-invasive ventilation
- Cough augmentation
  - Secretion management





### WITH THANKS













