



## Using MRI to understand muscle changes in Limb Girdle Muscular Dystrophy (LGMD2A/R1) and Becker Muscular Dystrophy

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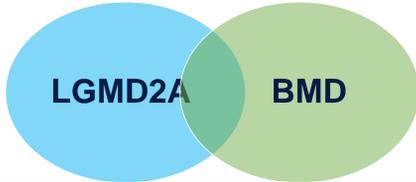
### INTRODUCTION – WHY?



The goal of this project is to demonstrate that MRI techniques can measure muscle changes in diseases such as Limb-girdle muscular dystrophy type 2A/R1 (LGMD2A) and Becker muscular dystrophy (BMD).



One major limitation to the success of treatment studies (clinical trials), including gene therapy and disease-modifying medications, is that the measurements commonly used are not great at measuring small changes or stabilization of diseases. Additionally, because LGMD2A/R1 and BMD are rare diseases, it is important to report what changes in muscles we see in the “natural history” of the progression of people with these diseases.



These diseases have different genetic causes but are both rare and progressive diseases that present as muscle weakness during late adolescence or adulthood. Both diseases result in muscle damage and fat gradually replaces healthy muscle tissue.



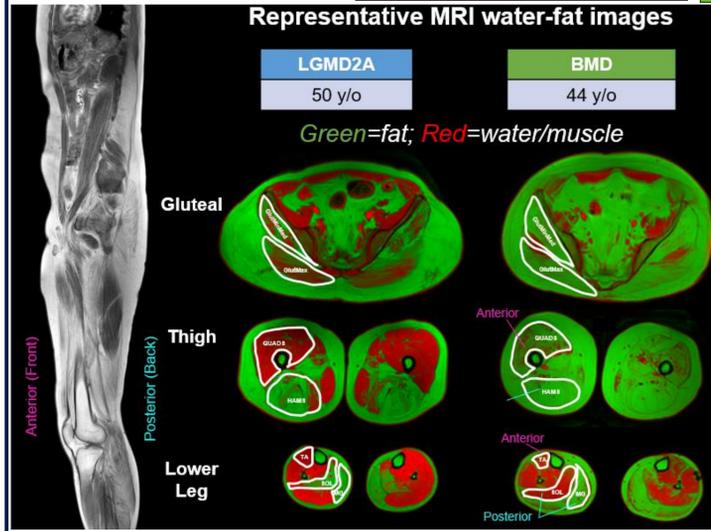
MRI techniques, developed in our lab, can measure the amount of fat infiltration in muscle, which we call “fat fraction”. We can detect small changes in muscles, including fat fraction, which can help design clinical trials and help to determine if the treatment works. Using these methods, we can highlight some major similarities and differences between these two diseases.

### METHODS – HOW?

Who?: People aged 18 to 62 years who can walk

Unaffected Adults = 5    **LGMD2A = 14**    **BMD = 40**

High Functioning    Lower Functioning    High Functioning    Lower Functioning

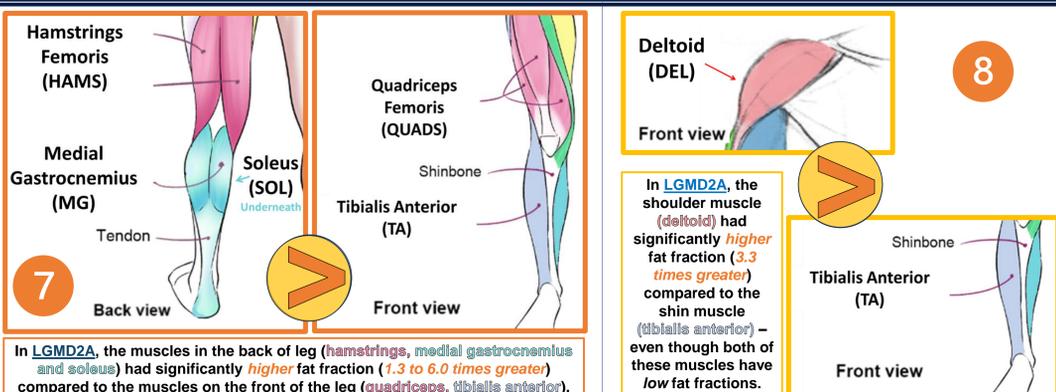
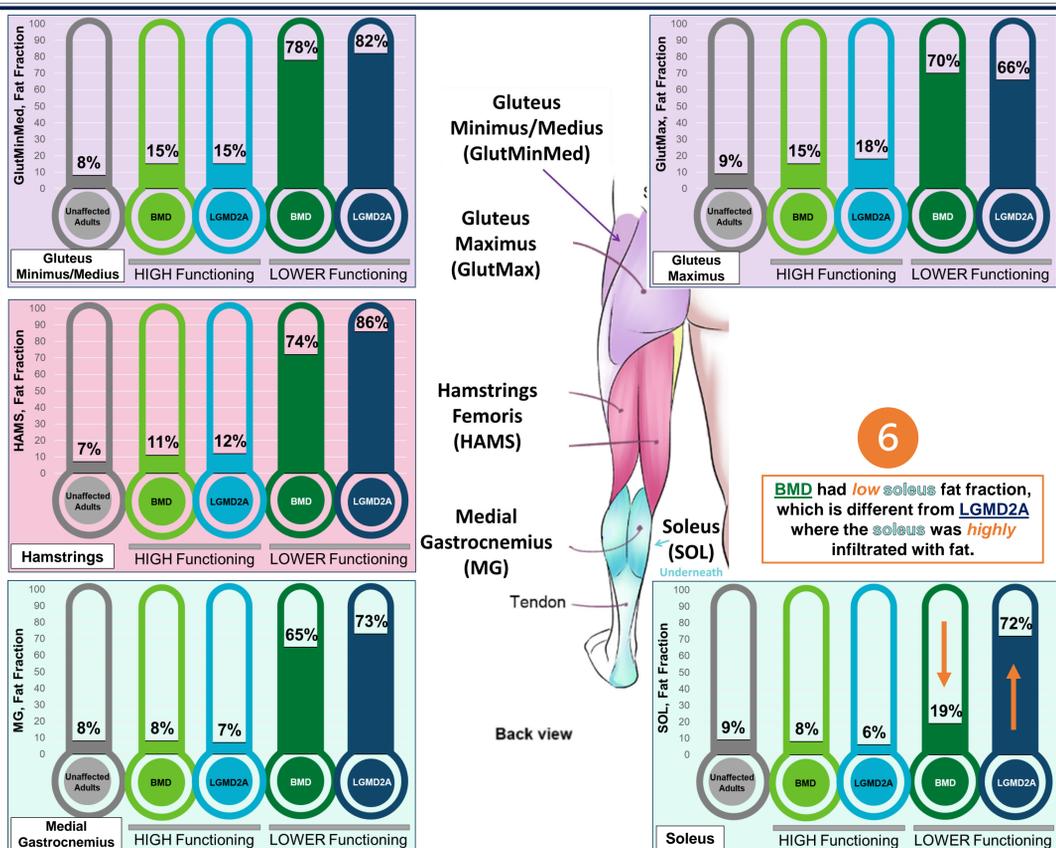
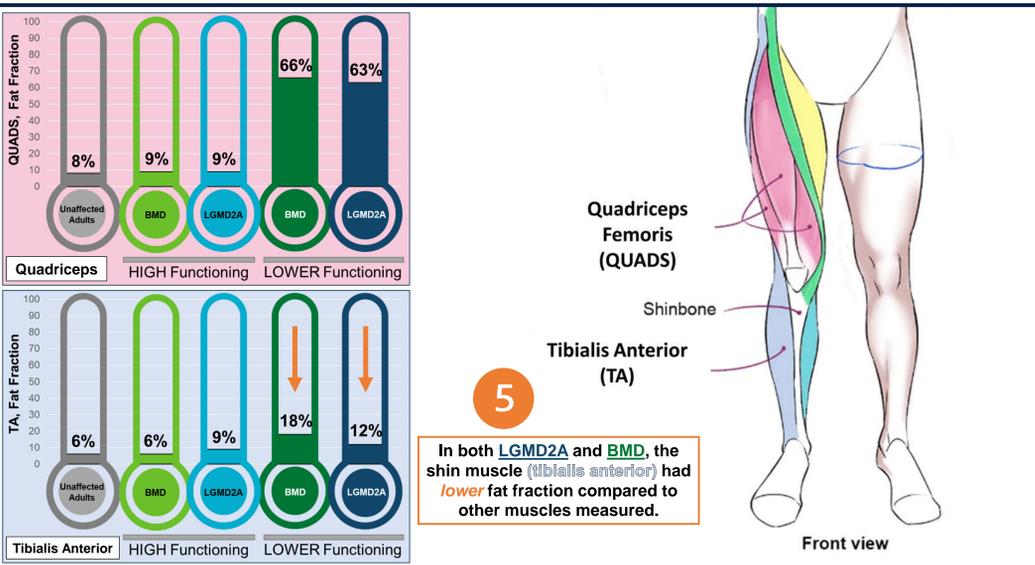
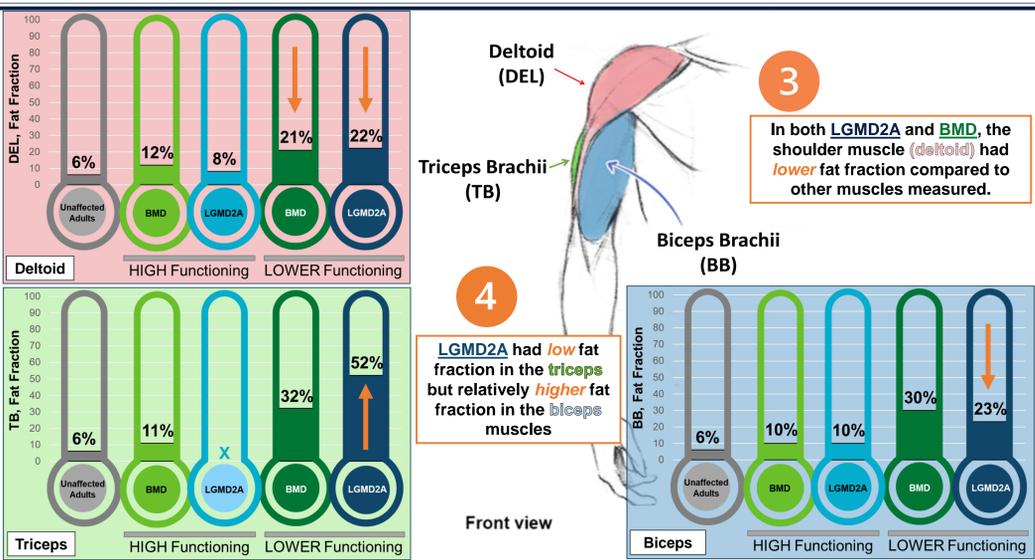


What?: Dixon MRI method to calculate the amount of fat in muscles  
**HIGH Fat Fraction =**

### RESULTS – WHAT WE FOUND

1 People with **high function** (unaffected adults, BMD, and LGMD2A) showed **very low fat fraction values** (median values = less than 20% fat fraction).

2 People with **BMD or LGMD2A** and who had difficulty climbing stairs but were able to walk (with or without assistance) showed **higher fat fraction values** in most of the muscles.



### CONCLUSIONS – SO WHAT?

- These findings demonstrate that muscle fat fraction, measured by MRI, is increased in people with LGMD2A/R1 and BMD – especially in those that have problems with physical function.
- We also demonstrate that these MRI methods can highlight important differences in muscles and muscle groups in LGMD2A/R1 and BMD.
- These findings can help clinicians and researchers know which muscles to monitor to determine whether their treatment helped to stabilize or prevent progression.

### Acknowledgements

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