



INTRODUCTION

- Becker Muscular Dystrophy (BMD) is a progressive neuromuscular disorder and leads to impaired upper extremity (UE) function.
- The Performance of Upper Limb (PUL) assessment can reliably measure UE function in Duchenne muscular dystrophy but has shown a ceiling effect in high functioning individuals.
- Quantitative magnetic resonance imaging (qMRI) has been used to measure disease progression across dystrophinopathies.

OBJECTIVES

- The goal of this study is to:
 1. Evaluate the ability of both the PUL and qMRI measures of fat fraction (FF) to discriminate between functional groups of men with BMD and controls.
 2. Assess the relationship between PUL scores and qMRI FF.

METHODS

- Cross-sectional study of 65 participants:
 - 56 men with BMD – 47 ambulatory; 9 Non- ambulatory
 - 9 controls
 - ages 18-63 years
- Data collected as part of ImagingNMD at:
 - University of Florida & Oregon Health and Science University
- **Functional measures:**
 - PUL 2.0 – a 22-item scale designed to capture UE motor performance in D/BMD with a maximum total score of 42.
- **MRI Assessment:**
 - qMRI FF using whole-body 3P Dixon imaging (Figure 1)
- **qMRI Endpoints:**
 - Composite UE FF - calculated as an average of upper arm and shoulder muscle: (Figure 1A + 1B)
 - Composite whole-body FF – calculated as an average multiple muscles (Figure 1A + 1B + 1C + 1D + 1E + 1F)

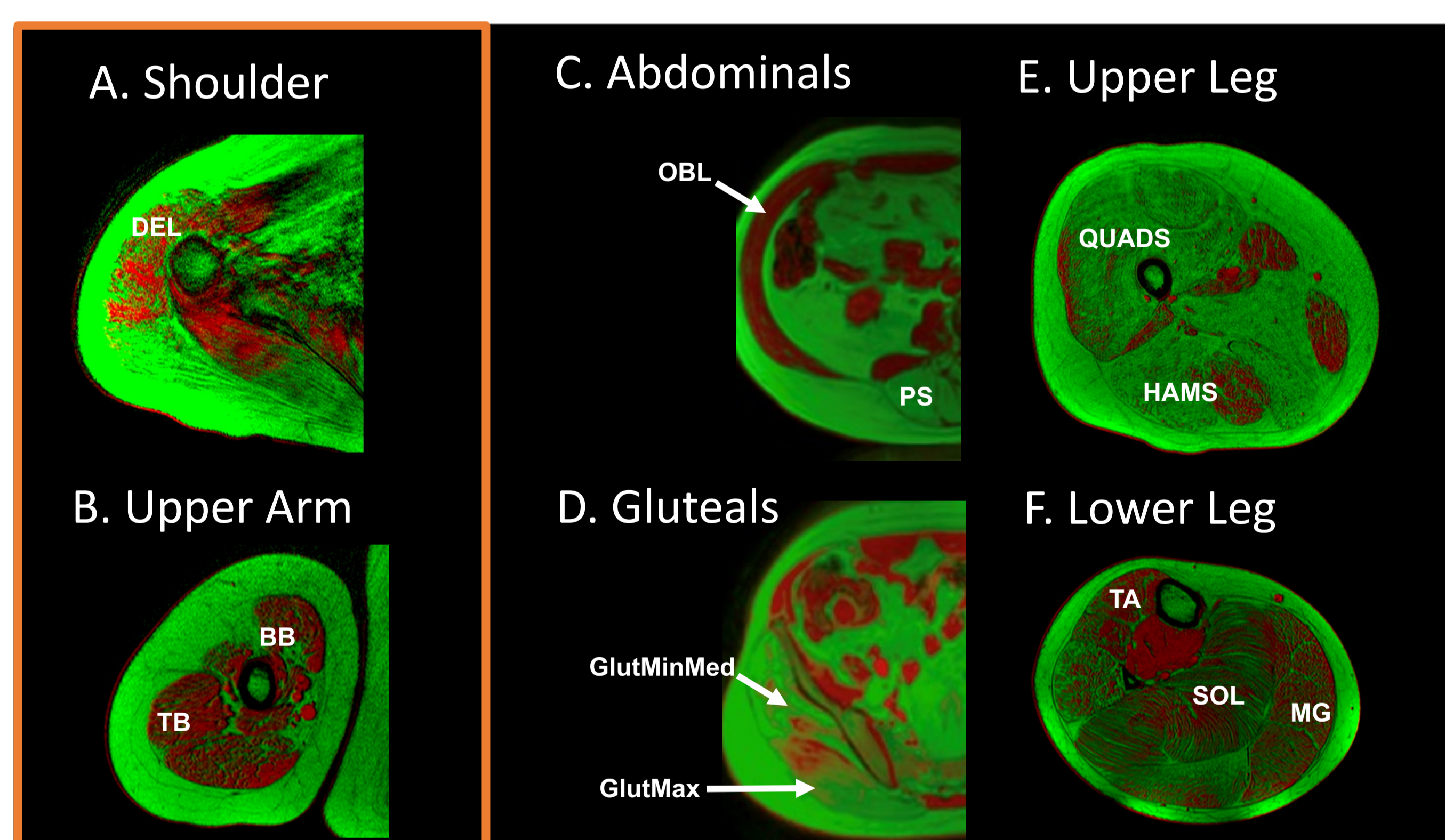


FIGURE 1: Example Dixon (fat-water fusion) images of BMD participants. Green = fat; Red = water (muscle).

- BMD participants were grouped based on performance during the 10-m run/walk test into 4 groups:

Lower-functioning groups:		Higher-functioning groups:	
A	Unable to walk 10m	C	Fast walk or jog or ability to increase walking speed
B	Walks, with no increased in speed possible	D	Runs- both feet off the ground, achieves flight

- **Statistical Analyses:**
 - One-way ANOVA/Kruskal Wallis and Spearman's rho were used to assess differences between groups and correlations, respectively.

REFERENCES

- Comi et al., 2022.
- Forbes et al., 2020.
- Brogna et al., 2021.
- Mayhew et al., 2020.
- Barnard et al., 2018.
- No Conflict of Interests to disclose.

RESULTS

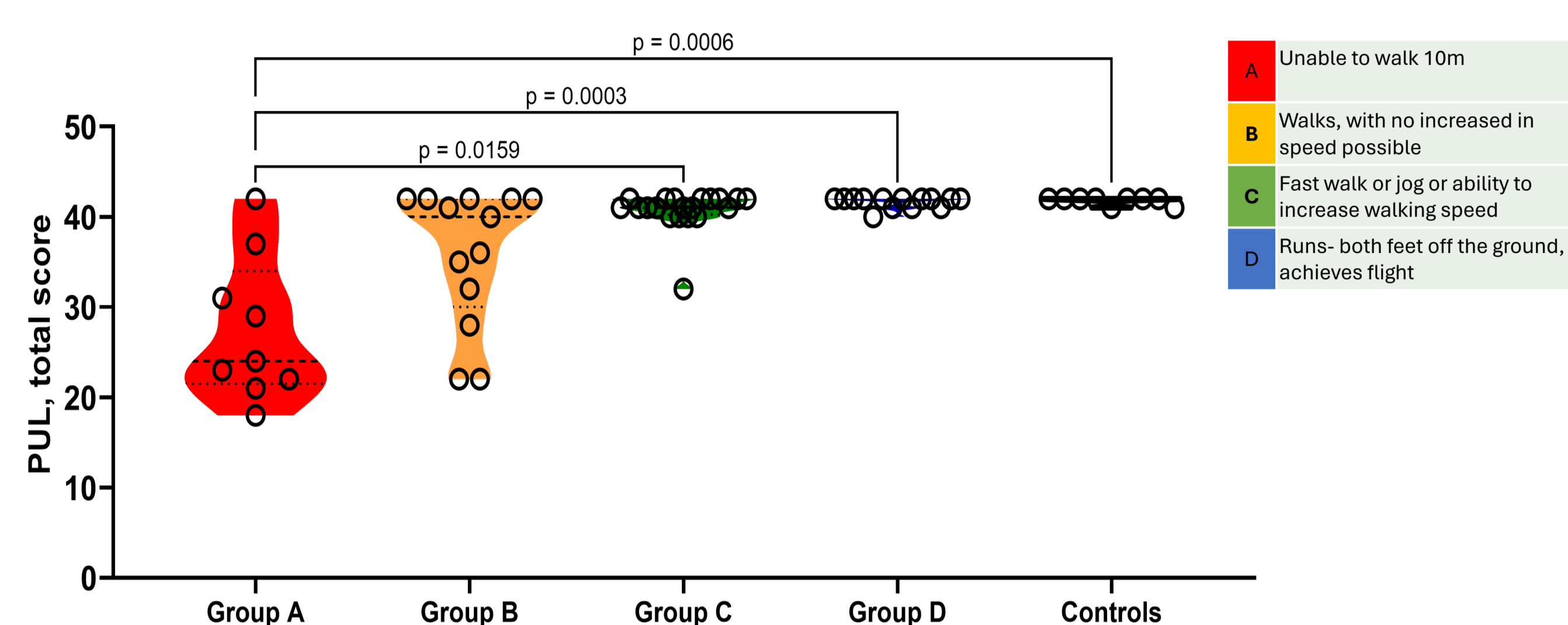


FIGURE 2: Men with BMD scored 18-42 on the PUL. The PUL only detected differences between the non-ambulatory (Group A) and high-functioning groups.

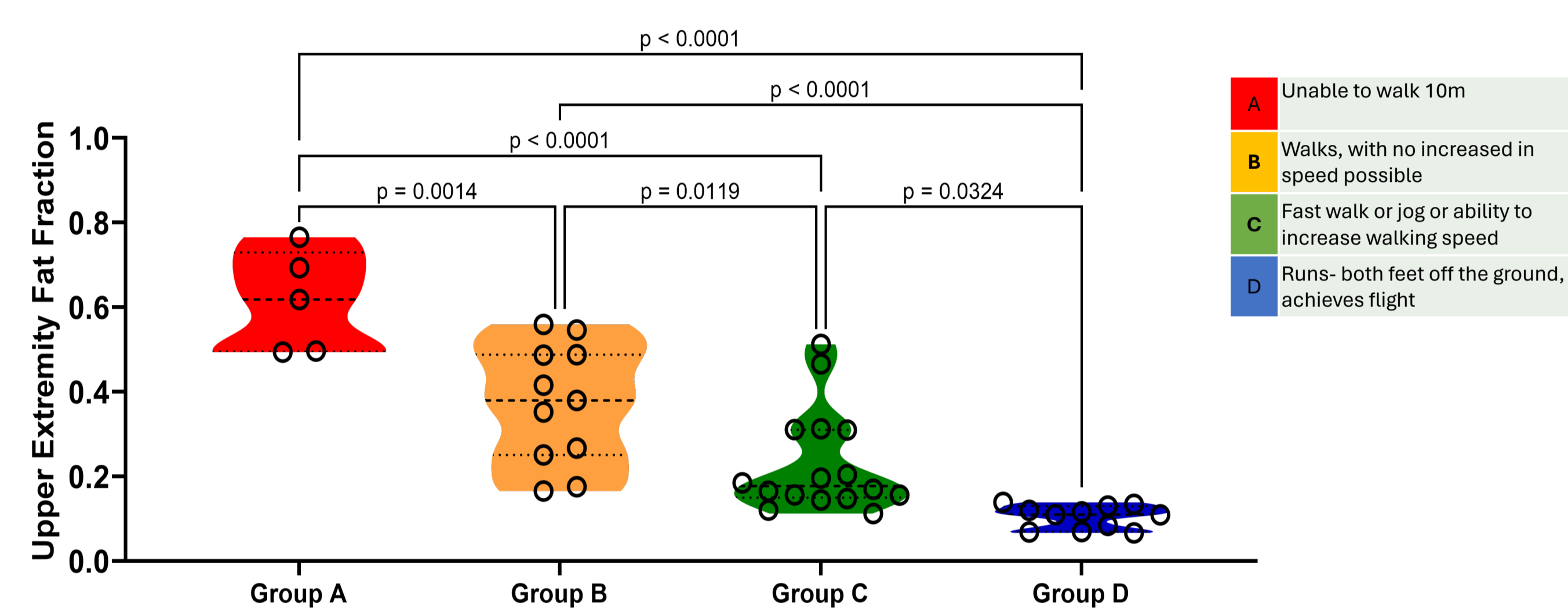


FIGURE 3: qMRI UE FF composite measures ranged from 7 to 77% and UE FF discriminated between all BMD functional groups.

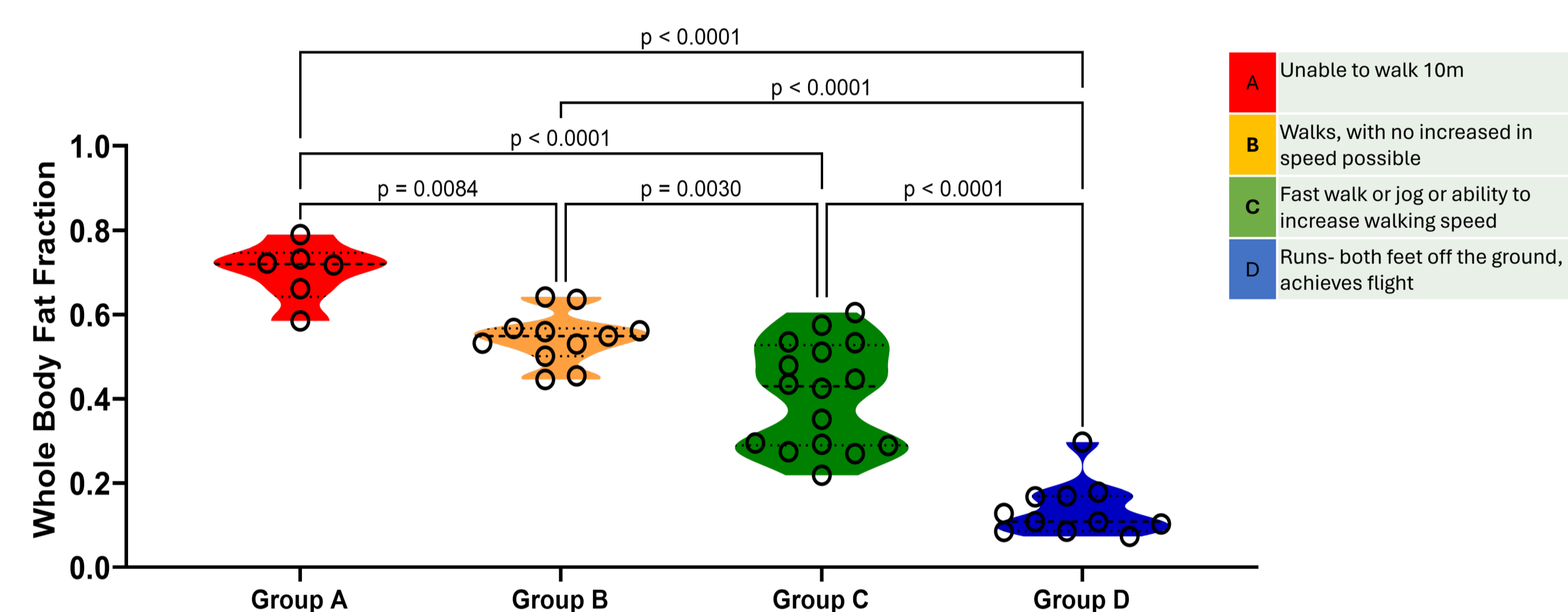


FIGURE 4: qMRI whole-body FF composite discriminated between all BMD functional groups.

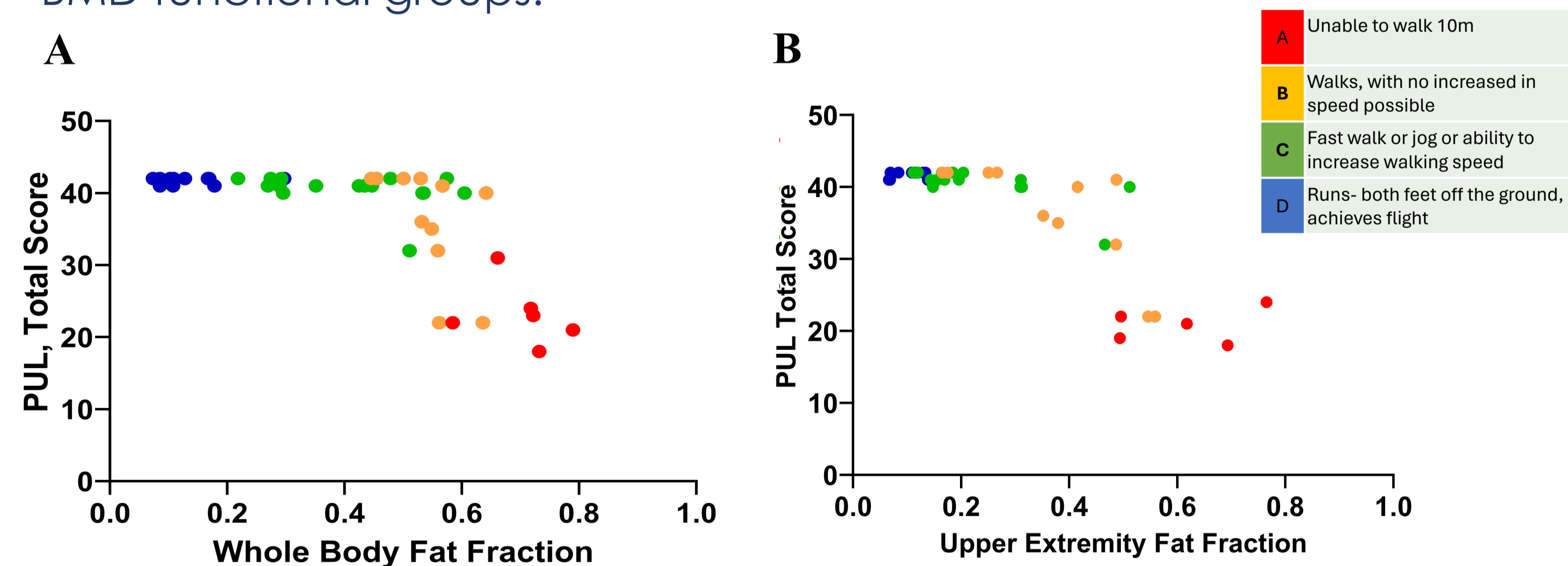


FIGURE 5: The PUL total score remains high until whole-body FF reaches ~50% (A) and UE FF reaches ~40% (B)

CONCLUSIONS

- **PUL has utility** in assessing UE function in **low-functioning men with BMD.**
- **qMRI FF detects muscle deterioration** across all functional levels, offering a **robust measurement of muscle health in BMD.**
- Because the PUL may provide **limited information** about UE impairment in **high-functioning men with BMD**, it should be **used cautiously and other measures, such as qMRI FF, should be considered.**

ACKNOWLEDGEMENTS

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GRATEFUL TO ALL THE STUDY PARTICIPANTS AND THEIR FAMILIES!