A pilot study investigating the impact of glycogen storage disease (type III) on skeletal muscle properties and habitual physical activity patterns in young and middle-aged adults.

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Glycogen storage disease type III (GSDIII) is an autosomal recessive condition, characterised by skeletal muscle (SM) impairment leading to functional limitations. Previous research has identified both handgrip strength to decline following the age of 37 and sit-to-stand time to increase post 30 years old (Decostre et al., 2017, *Molecular genetics and metabolism*, 122, 108-16). However, the age-related drop in SM strength is noted to be more prominent in loaded antigravity SM in comparison to the upper extremities (Delmonico et al., 2009, *American Journal Clinical Nutrition*, 90, 1579-1585) and may not adequately detail the impact of GSDIII on SM and physical function. Therefore, the aim of this study is to preliminary detail the impact of GSDIII on antigravity SM properties and investigate habitual physical activity (PA) patterns that may contribute to a loss strength. With institutional and NHS ethics approval, five participants were recruited and categorised into two age groups (Mean±SD: **<30yrs** (*n*=2) age: 27±1; body mass 76.9±5.2kg; **≥30yrs** (*n*=3) age: 49±3; body mass 76.5±6.1kg). Group differences were examined between body composition assessed by bioelectric impedance, SM structural properties by ultrasonography (vastus lateralis muscle thickness (VLMT), pennation angle and fascicle length), knee extensor strength by isokinetic dynamometry (isometric maximum voluntary contraction (iMVC) at 80°) and PA patterns using thigh-mounted 3D accelerometry undertaken for 7 days. Primarily, the ≥30yrs group had 52% higher body fat% and 16% lower lean mass, supporting the theory of a drop in physical function post 30yrs of age. Similarly, SM structural differences, revealed the ≥30yrs group to have 17.7% lower VLMT (2.03cm vs. 1.67cm), 5.6% decrease in pennation angle (15.9° vs. 15°), yet 7.5% greater fascicle length (6.7cm vs. 7.2cm). Interestingly though, knee extensor iMVC was 78% lower (225Nm vs. 49Nm) in the ≥30yrs group, in conjunction with 75% lower relative strength (iMVC/body mass) and 68% lower muscle quality (iMVC/VLMT). Similar habitual PA patterns were observed between age groups with an average of 10.9hrs spent being sedentary (>8 hours classed as sedentary) and no medium/vigorous intensity PA bouts lasting >10mins, potentially explained through substrate utilisation and availability. In conclusion, preliminary data suggests that there is a substantial drop in muscle strength post 30 years of age in antigravity SM. However, whilst further research is needed confirm these findings, future investigations should focus on neuromuscular and structural differences that may contribute to the substantial drop in strength that was observed.