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Poster Abstract: Diagnostic

Dynamic Change in Muscle Volume after Exercise is Altered in Pompe Disease: A Quantitative, Sequential MRI Study

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BACKGROUND

Resting muscle MRI has been increasingly used to obtain information about muscles in Pompe disease. However, dynamic changes in muscles after exercise, and how muscles are used to achieve a motor outcome in Pompe disease have not been explored. By using sequential quantitative MRI, here we investigated the temporal profile of volumetric changes in the hamstring complex after concentric exercise in patients with Pompe disease.

MATERIALS AND METHODS

Nine patients with late-onset Pompe disease and 10 control subjects were enrolled. Standard exercise protocol was established by using quantitative dynamometry. Quantitative MRI of the hamstring complex was performed at three time points: before exercise, and 30 minutes and 24 hours after exercise. Anatomical cross sectional areas (ACSAs) of the hamstring complex were measured on axial reformations of T1W images at each time points. Data collected 30 minutes and 24 hours after exercise were subtracted from baseline to investigate the rate of change in muscle ACSAs over time in both groups. Repeated-measures ANOVA was performed for the control and Pompe group to compare changes in ACSAs among the three time points.

RESULTS

After Bonferroni's test for multiple comparisons, our data showed significantly higher ACSAs in the control group at both 30 minutes and 24 hours after exercise compared with baseline measurements ($p < 0.001$); in Pompe patients, muscle volume also showed a trend of increase by 30 minutes similar to controls, but this was not significant due to high standard deviation. Despite a similar increase in muscle volume, the rate of change 30 minutes after exercise (30 minutes minus baseline) was significantly higher in the semimembranosus muscle ($p = 0.02$) of controls compared with Pompe patients; however, it showed decreased values in the hamstring complex (semimembranosus, semitendinosus, biceps femoris muscles). Finally, 24 hours after exercise, ACSAs of Pompe patients still depicted higher values, while these were decreasing in control subjects.

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CONCLUSIONS

The concentric type of hamstring exercise has a similar effect in both controls and patients with Pompe disease, indicated by a similar trend of muscle volume increase: despite severe damage, patients activate muscles in comparable ways. Nevertheless, the rate of increase in muscle volume is less prominent in patients, and muscle volumes are still increasing 24 hours after exercise in Pompe disease, while decreasing in controls. This may be due to the altered motor unit activity and abnormal regulation of metabolic changes. Finally, volumetric changes in individual muscles of the hamstring complex differentially increased volume after exercise in controls and patients, which may indicate differential usage of the particular muscles in order to obtain the same motor outcome.