GAIT ANALYSIS IN PATIENTS AFFECTED BY LATE-ONSET POMPE DISEASE (LOPD)

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Background

Late-onset Pompe disease (LOPD) is a rare condition characterized by a progressive accumulation of lysosomal glycogen mainly resulting in muscle weakness and respiratory problems. Proximal muscular weakness in patients affected by LOPD can determine myopathic gait pattern with slower walking speeds and deviations in gait patterns.

Objective

To assess analysis of spatial-temporal parameters in patients affected by LOPD, using a waist-mounted wireless inertial sensor..

Materials and Methods

Four patients (mean age at observation 49,7±13,9 years, two of female gender) affected by LOPD during treatment with enzyme replacement therapies (two patients with alglucosidase alfa and two with avalglucosidase alfa) were assessed with a 6-minute walking test (6MWT) as performed with a 5-month interval (T1 and T2). Spatial-temporal parameters of gait (i.e., step length and duration, along with their symmetry and variability) were recorded using a waist-mounted wireless inertial sensor [1].

Results

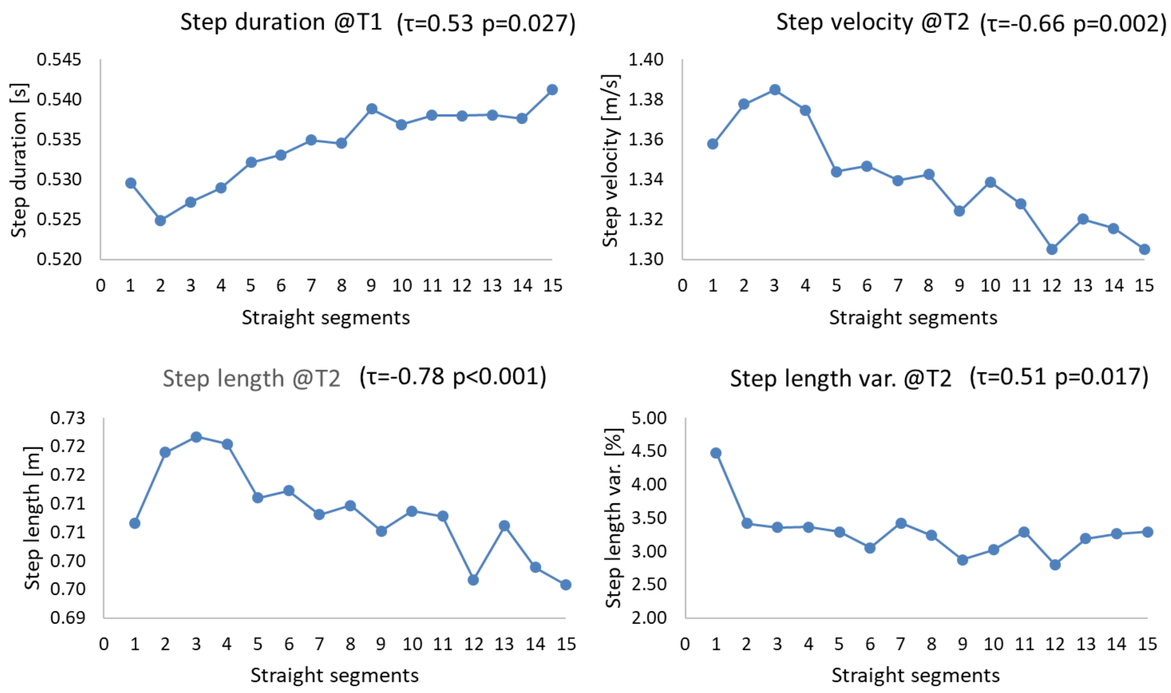
The Wilcoxon test revealed no significant difference between T1 and T2 in any of the considered spatial-temporal parameters ($0.125<p<0.875$) (Table 1). Within each time point, trend analysis of spatial-temporal parameters over the instrumented 6MWT was performed using the Mann-Kendall Test and revealed a significant increase of step duration at T1 ($\tau=0.53$, $p=0.027$), an increase of step length variability ($\tau=0.51$, $p=0.017$) and a decrease of step length ($\tau=-0.78$, $p=0.0003$) and velocity ($\tau=-0.66$, $p=0.002$) at T2.

Table 1

Analysis of spatial-temporal parameters in patients affected by Late-onset Pompe disease (LOPD), performed with a 5-month interval (T1 and T2).

	T1	T2
	median (IQR)	median (IQR)
Straight segments	16	17
Step number (in 25 meters)	33	33
Straight path duration [s]	18.7 (3.4)	17.5 (3.4)
Step length [m]	0.72 (0.08)	0.73 (0.1)
Step duration [s]	0.52 (0.05)	0.52 (0.01)
Step frequency [Hz]	1.91 (0.16)	1.93 (0.05)
Gait speed [m/s]	1.33 (0.21)	1.41 (0.19)
Step duration variability [%]	3.13 (0.93)	3.21 (0.73)
Step duration symmetry [%]	1.76 (2.35)	2.06 (1.96)
Step length variability [%]	3.20 (1)	3.24 (1.02)
Step length symmetry [%]	1.07 (1.29)	1.47 (1.18)

Mann-Kendall trend analysis of variables with statitically signficant tau value



Conclusion

Overall, the instrumented 6MWT in patients affected by LOPD can be useful to objectively monitor both possible fatigue effects during the execution of the test and to evaluate the efficacy of the treatment over a given time-interval.

Reference

[1] Kirk, C., Küderle, A., Micó-Amigo, M.E. et al. Mobilise-D insights to estimate real-world walking speed in multiple conditions with a wearable device. Sci Rep 14, 1754 (2024).