Application of wearable sensors to assess disease severity in adults with Charcot-Marie-Tooth disease

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Introduction

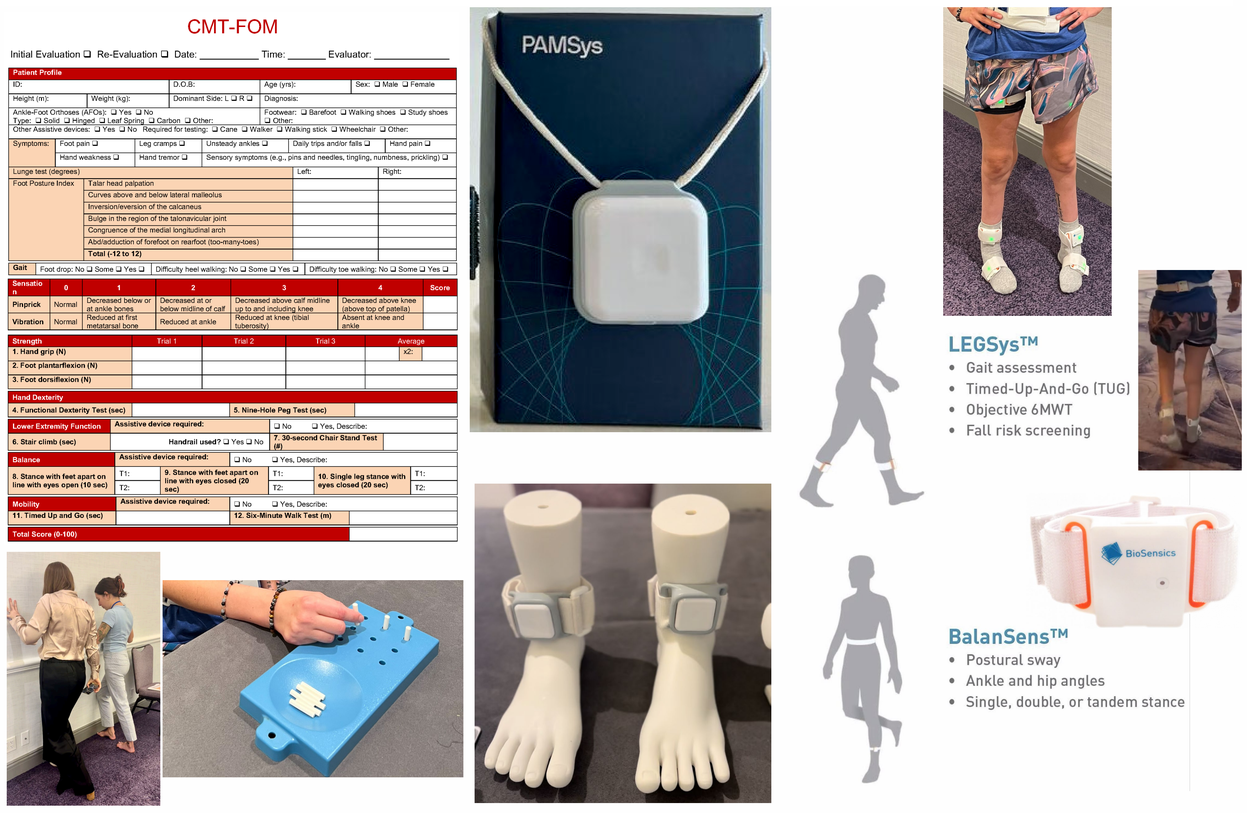
- Reliable, valid, and sensitive clinical outcome assessments (COAs) are essential to monitor disease progression and measure therapeutic interventions.[1,2,3]
- Traditional COAs often capture a single day's performance, which may not reflect a person's true abilities.
- Wearable sensors offer continuous monitoring in a home environment, providing real-world evidence of function and more sensitive assessments of gait and balance.
- Validating wearable sensors could enhance our ability to track disease progression and evaluate therapeutic interventions effectively.

Aims

- To evaluate the feasibility of remote data collection of physical activity and instrumented gait and balance measures in adults with Charcot-Marie-Tooth disease (CMT).

Methods

- Cross-sectional cohort study at the Hereditary Neuropathy Foundation CMT Clinical Trial Readiness Summit.
- CMT-FOM and instrumented gait and balance assessments using LEGSys™ and BalanSens™ (BioSensics, Newton MA).
- Physical activity was monitored continuously for 14 days at home using PAMSys™ pendant sensor.



Results

- 26 Participants (81% female, 58% CMT1A) participated in this study.
- Compliance with PAMSys for at home monitoring was excellent with 16 participants having perfect compliance (mean non-compliance: 22 mins/day).

Table 1: Participant Characteristics

	Mean ± SD	Range
Age (yrs)	49.5 ± 14.9	19 - 70
Height (m)	1.68 ± 0.10	1.55 – 2.03
Weight (kg)	78.9 ± 23.6	46.5 – 135.2
CMT-FOM	53.2 ± 8.0	37 - 73

Table 2: Physical Activity Data

	Mean ± SD	Range
Daily Step Count	5433 ± 1882	2522 – 10023
Max Steps (/bout)	370 ± 361	87 – 1764
Cadence (steps/min)	93 ± 5	80 – 105
% time walking	4.6 ± 1.4	2.5 – 7.3
% time lying	40.9 ± 10.1	16.8 – 57.3
% time sitting	36.2 ± 8.8	23.9 – 62.3
% time standing	16.8 ± 4.2	9.3 – 28.7

Significance

- Participants were compliant with the wearable pendant sensor.
- These wearables provided real-world evidence by continuously capturing long-term data that reflects patients' natural environments and daily activities.
- Sensor-derived metrics were validated against established COAs.
- In future trials, the sensitivity of these measures to change should be explored in longitudinal studies.

Results Continued

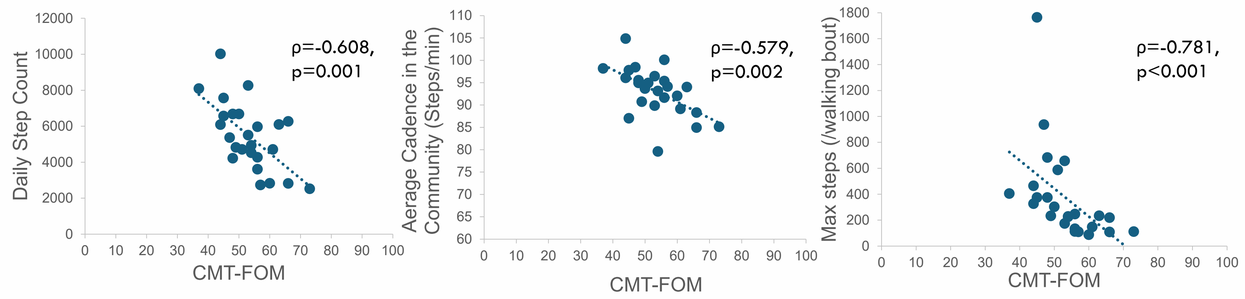


Figure 1: Correlations between CMT-FOM and daily gait parameters

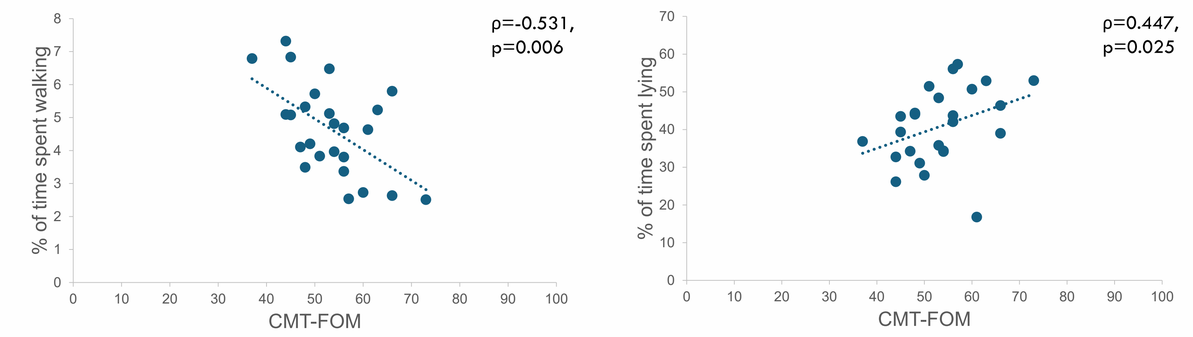


Figure 2: Correlations between CMT-FOM and daily activity and postures

No significant correlations between CMT-FOM and % of time spent sitting or standing.

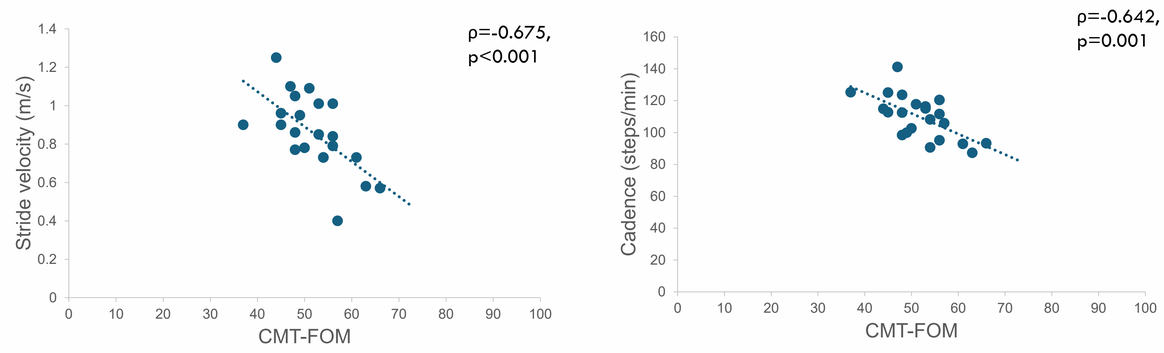


Figure 3: Correlations between CMT-FOM and LEGSys Gait Parameters

No significant correlations between CMT-FOM and joint angles measured during gait.

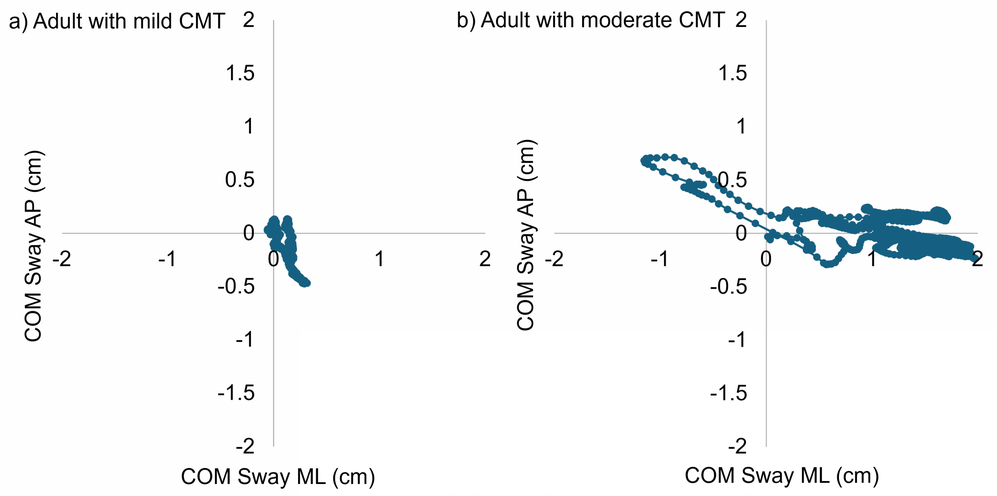


Figure 4: BalanSens Sway data in mild and moderate CMT

a)CMT-FOM=37; age=70 yrs. B)CMT-FOM=66; age=34 yrs.

References

1.Burns at al, Annals of Neurology 2012
2. Mandarakas et al, Brain 2018
3. Mandarakas et al, Neurology 2024

Acknowledgements

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