

# 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.<sup>1,2,3</sup>
- 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.

# 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.



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<sup>TM</sup> and BalanSens<sup>TM</sup> (BioSensics, Newton MA).
- Physical activity was monitored continuously for 14 days at home using PAMSys<sup>TM</sup> pendant sensor.

		CMT-FC	M		DANKS	
Initial Evaluati Patient Profile	ion 🛛 Re-Evaluatior	n 🗖 Date:	_ Time:	_Evaluator:	PAMSys	
ID:		D.O.B:	Age (yrs):	Sex: 🗅 Male 🗅 Female		
Height (m):	Weight (kg):	Dominant Side: L 🗆 R 🗆	Diagnosis:			
Ankle-Foot Orthoses (AFOs):       Yes       No       Footwear:       Barefoot       Walking shoes       Study shoes         Type:       Solid       Hinged       Leaf Spring       Carbon       Other:       Other:         Other Assistive devices:       Yes       No       Required for testing:       Cane       Walking stick       Wheelchair       Other:						



**LEGSys™** 

Gait assessment

Objective 6MWT

BalanSens™

Postural sway

Ankle and hip angles

Single, double, or tandem stance

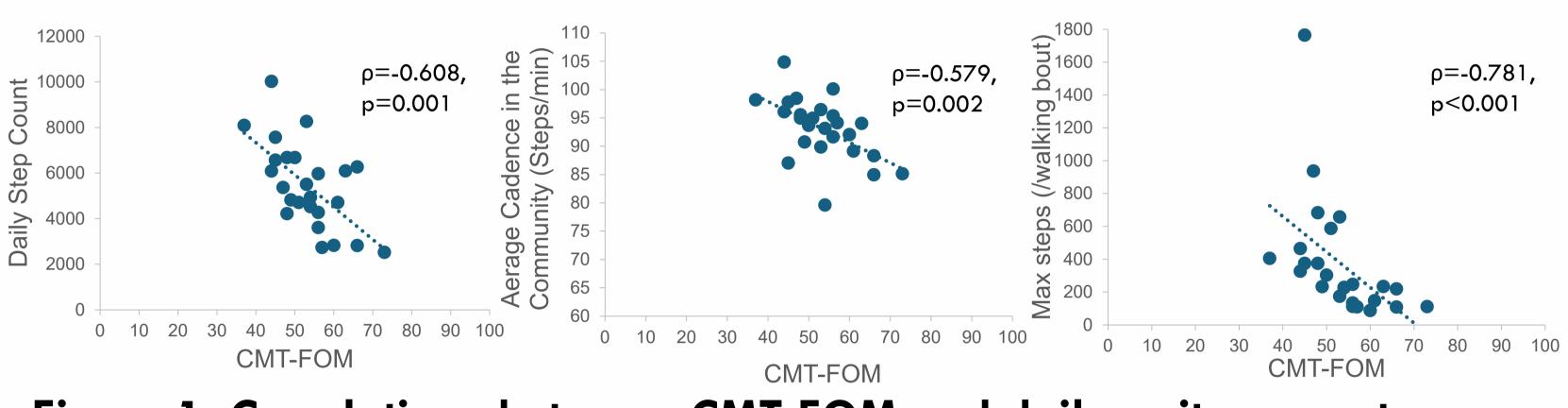
• Fall risk screening

• Timed-Up-And-Go (TUG)

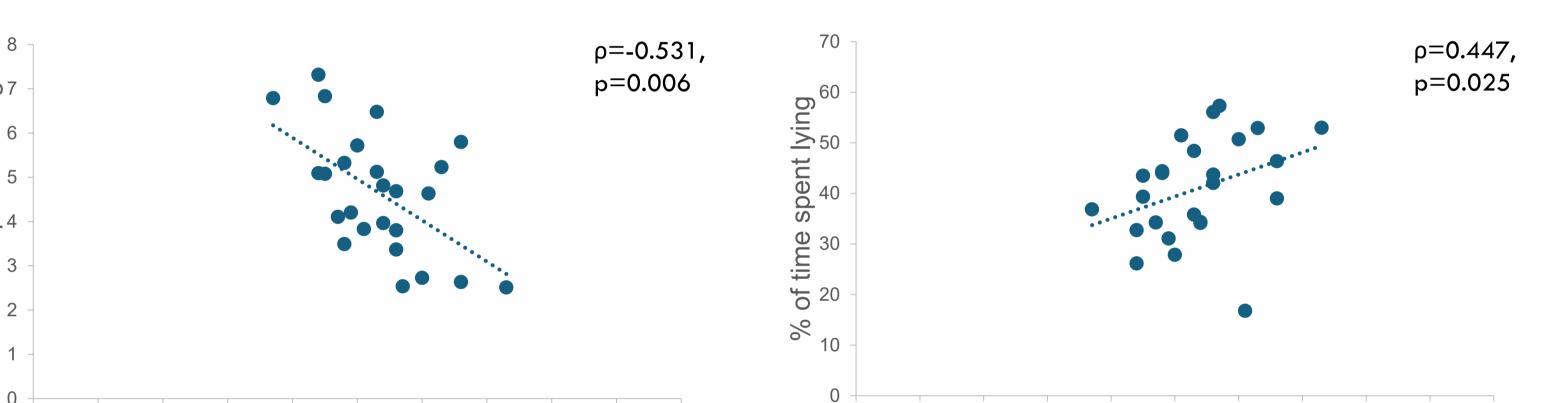
**BioSensic** 

 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



	Hand wea		Hand							g, numbness,		
Lunge test (degrees)							Left:		Right:		_	
Foot Posture Index Talar head palpation												
			e and below lateral malleolus									
			n/eversion of the calcaneus									_
		-	e region of the talonavicular joint								_	
			ction of forefoot on rearfoot (too-many-toes)									
		Total (-12 to 1										- 11
Gait Foot dr		Some 🗆 Ye		Difficulty heel	walking:				, too walk	ing: No 🗆 So	me 🗆 Ves 🗆	<u> </u>
1 Oot die			5 <b>u</b>	Difficulty field	waiking.		u res		/ toe waik	ing. No 🖬 30		
n	0	1		2			3			4	Scor	e
Pinprick No		Decreased bel at ankle bones		Decreased a below midline		Decreased up to and		e calf midline		sed above kn top of patella		1.0
Vibration No	F	Reduced at firs	st	Reduced at a		Reduced a	at knee		Absent	at knee and	,	
	r	metatarsal bor	e	Reduced at a		tuberosity	)		ankle			_
Strength				Trial 1		Trial 2		Trial 3			erage	
1. Hand grip (N	·									×	2:	
2. Foot plantar	flexion (	(N)										
3. Foot dorsifle	exion (N	)										
Hand Dexterity	1											
4. Functional D	Dexterity	/ Test (sec)				5. Nine-Ho	ole Peg	Test (sec)				100
Lower Extremit	tv Funct	tion Assis	tive de	vice required	:			D 🗆 Yes.	Describe	:		_
6. Stair climb (s				Handrailu	read2 🗆	Yes 🗆 No		-second Chai				
	3007						(#)					
Balance		Ass	stive o	levice require		🗖 No	<b></b>	s, Describe:				_
8. Stance with f				9. Stance v line with ey			T1:			stance with	T1:	_
line with eyes o	open (10	D sec) T2:		sec)			T2:	eyes	closed (2	20 sec)	T2:	
Mobility		Ass	istive o	device require	ed:	🗆 No	🗆 Yes	s, Describe:				
11. Timed Up and Go (sec)				12. Six-Minute Walk Test (m)								
												$\dashv$
Total Score (0- <sup>.</sup>	100)											



# Results

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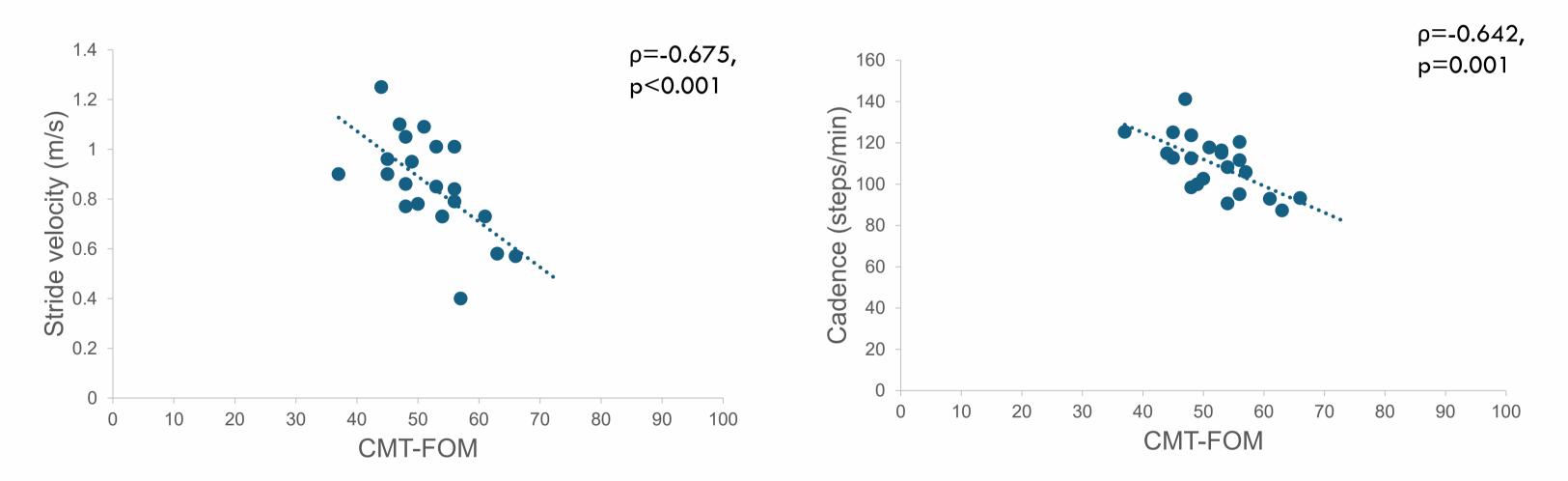
care, advocacy, research, education

- 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).



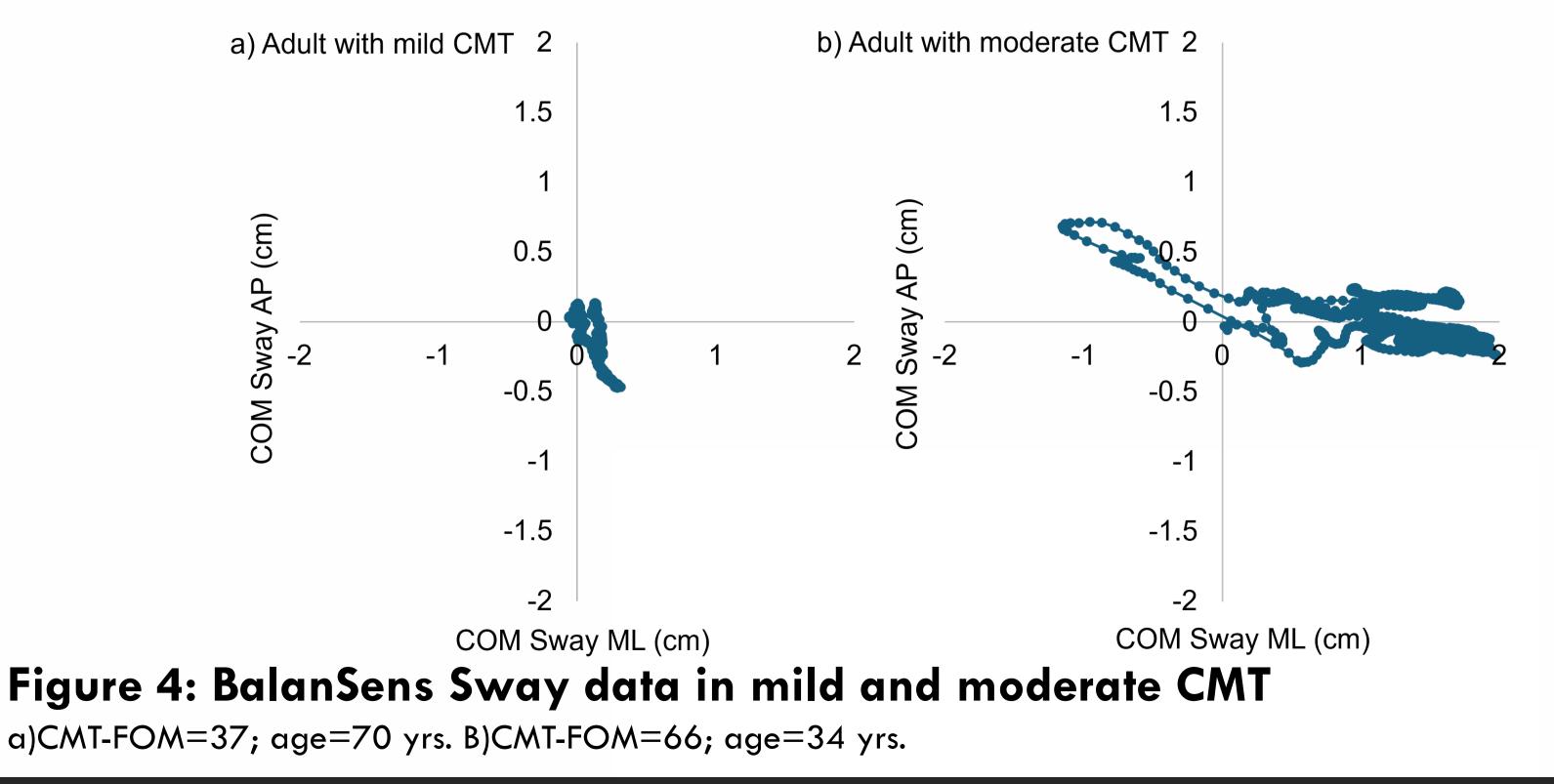
#### Figure 2: Correlations between CMT-FOM and daily activity and postures

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.



#### **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

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	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	$4.6 \pm 1.4$	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				

### References

Burns at al, Annals of Neurology 2012
 Mandarakas et al, Neurology 2024

2. Mandarakas et al, Brain 2018

### Acknowledgements

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