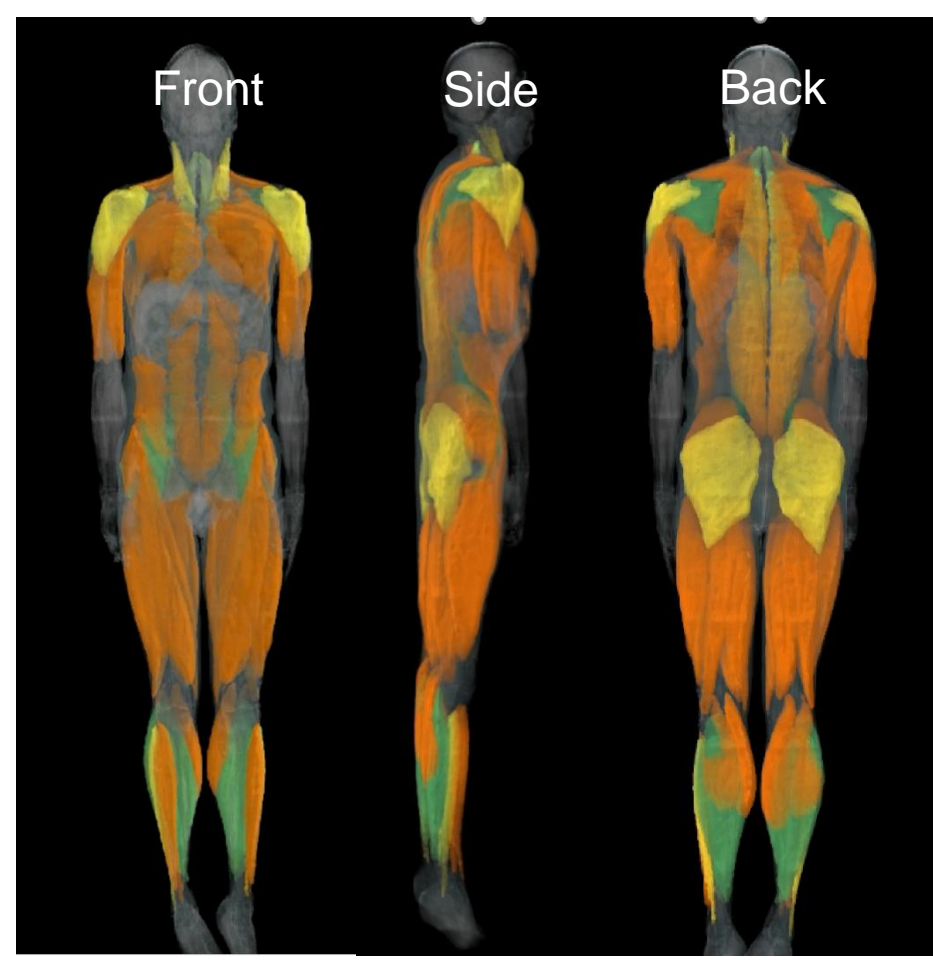




1. Background – Facioscapulohumeral Dystrophy and Emerald



- FSHD is a serious, rare, progressive and disabling disease.
- Second most common muscular dystrophy.
- Affects muscles in the face, shoulders, arms, abdomen, pelvic girdle, hips, and lower legs.

The Challenge

- Clinical trials in FSHD suffer from the lack of accurate, low patient burden metrics that continuously track disease severity and correlate with clinical outcomes.
- Current clinical outcome assessments may not be sensitive enough to detect disease progression over short periods of time.

Green = not affected
Yellow = small increases reported in some studies
Orange = high fat replacement in almost all studies

2. Emerald

- Emerald is a contactless radio-wave-based home monitoring system that measures gait speed¹, time in bed², sleep, and vital signs.
- The device infers patient physiology by analyzing low-power radio reflections (10,000x less power than Wi-Fi) using machine learning algorithms.
- Emerald obtains its measurements passively, as patients go about their normal lives in their homes.



Figure 1. Emerald shown in a subject's home

3. Methods

Patient Characteristics

- 10 FSHD patients
- Observed in their homes 24/7 for 3 months
- 7 females and 3 males
- Aged 31-52
- Clinical Severity Scale: 1.5-4

This study is a sub-study; for full details please refer to poster entitled "An Optimized Timed Up and Go (TUG) for the Facioscapulohumeral Dystrophy (FSHD) Community"; V. Chan, M. Hatch, M. Mellion, D. Cadavid, and J. Han.

The following metrics were measured using Emerald:

- eTUG**, the Emerald measured timed up and go, when the patient naturally gets up from a sleeping position in bed and walks for two meters.
- In-Home Gait Speed**
- Sleep Schedule Variability (SSV)**, the variability across days in the time at which the patient goes to sleep.

In-clinic assessments included:

- Clinical Severity Score (CSS)
- Timed Up and Go (TUG)
- FSHD-TUG (optimized TUG that includes truncal and lower limb impairments)
- PROMIS
- Neurology Upper and Lower Extremities (NeuroUE, NeuroLE)
- Performance of Upper Limbs (PUL)
- Reachable Work Space (RWS)

4. Sleep Schedule Variability and eTUG

Sleep Schedule Variability and eTUG are novel metrics that are extracted by Emerald:

- Sleep Schedule Variability**: Standard deviation of the time at which a subject goes to sleep across the duration of the study.
- eTUG**: Inferred from patients' normal act of getting up and walking out from the bed. Emerald measures the spatial location of the subject when in their bed area and extracts the time from motion initiation within the bed to moving 2m away from the bed edge.

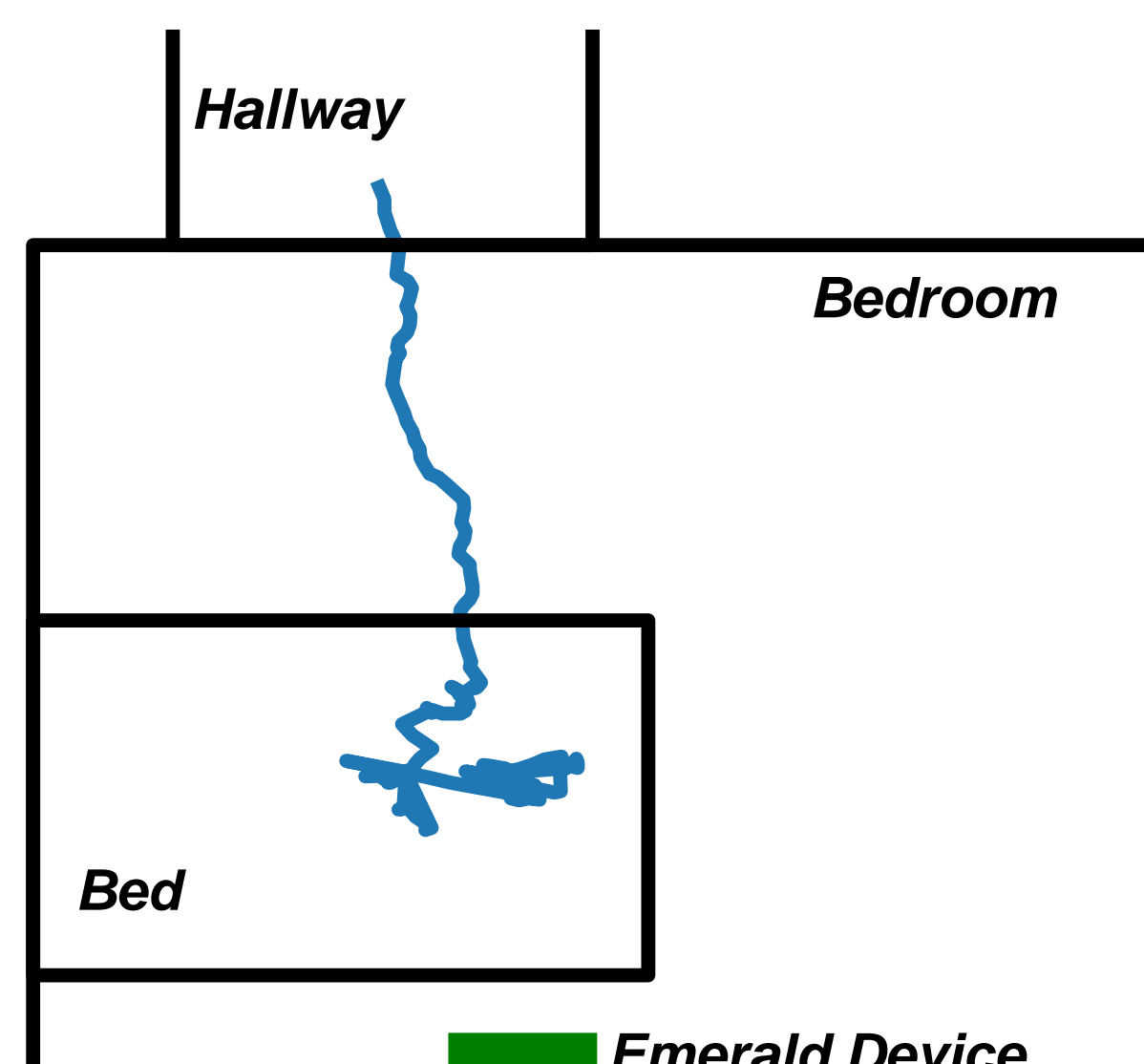


Figure 2. Emerald's device maps the location of the subject as they exit the bed. The blue trajectory shows subject moving in bed and eventually getting up and walking to the hallway.

5. Emerald's in-home monitoring enables a large number of measurements

Over 3 months, Emerald collected:

- 535 eTUG** measurements
- 3797 In-Home Gait Speed** measurements
- 707 Sleep Schedule Variability** measurements

	eTUG (s)	In-Home Gait Speed (m/s)	Sleep Schedule Variability (minutes)
count	10	10	10
mean	11.38	0.85	132.11
std	5.6	0.16	51.18
min	6.12	0.58	54.72
25%	7.48	0.8	112.61
50%	9.53	0.88	128.28
75%	13.99	0.94	156.13
max	22.86	1.11	205.59

Figure 3. Summary Statistics for each of the Emerald metrics

6. Emerald's in-home measurements exhibit strong correlations with clinical metrics

	CSS	TUG	FSHD-TUG	PROMIS	NeuroUE	NeuroLE	PUL	RWS
eTUG	0.88 (0.001)	0.73 (0.017)	0.73 (0.017)	-0.66 (0.038)	0.69 (0.027)	-0.73 (0.017)	-0.62 (0.056)	-0.49 (0.146)
In-Home Gait Speed	-0.56 (0.095)	-0.73 (0.018)	-0.68 (0.032)	0.43 (0.220)	-0.27 (0.443)	0.51 (0.135)	0.73 (0.017)	0.47 (0.174)
Sleep Schedule Variability	0.80 (0.005)	0.41 (0.234)	0.44 (0.204)	-0.68 (0.030)	0.72 (0.018)	-0.54 (0.108)	-0.60 (0.066)	-0.68 (0.032)

Figure 4. The correlation and the corresponding p-value in parenthesis between the Emerald metrics and the clinical scores.

- eTUG** demonstrates very strong correlation with CSS ($\rho=0.88$, $p=0.001$) and good correlation with in-clinic FSHD metrics: TUG ($\rho=0.73$, $p=0.017$), FSHD TUG ($\rho=0.73$, $p=0.017$), NeuroLE ($\rho=-0.73$, $p=0.017$), NeuroUE ($\rho=0.69$, $p=0.027$), and PROMIS ($\rho=-0.66$, $p=0.038$).
- In-Home Gait speed** has good correlation with TUG ($\rho=-0.73$, $p=0.018$), FSHD-TUG ($\rho=-0.68$, $p=0.032$), and PUL ($\rho=0.73$, $p=0.017$).
- Sleep Schedule Variability** demonstrates very strong correlation with CSS ($\rho=0.8$, $p=0.005$) and good correlation with NeuroUE ($\rho=0.72$, $p=0.018$), PROMIS ($\rho=-0.68$, $p=0.03$) and RWS ($\rho=-0.68$, $p=0.032$).

7. Emerald's measurements are sensitive and reliable

As the number of measurements increases, Emerald's metrics become increasingly sensitive and can detect smaller fluctuations in disease progression.

	5 measurements	20 measurements	100 measurements	1000 measurements
eTUG	7.8s	3.9s	1.7s	0.53s
In-Home Gait Speed	0.23 m/s	0.11 m/s	0.05 m/s	0.01 m/s

Figure 5. The Minimum Detectable Change (MDC95)¹ for eTUG and Gait Speed

eTUG and In-home Gait Speed have excellent Intraclass correlations, which indicate high test-retest reliability.

¹The minimum amount of change in a person's observed measurement that with 95% confidence is not the result of measurement error.

8. Discussion and Conclusion

- Emerald's metrics provide **reliable and sensitive** measurement of FSHD patients' disease condition while being **passive and contactless**.
- Emerald's metrics show strong correlations with clinical metrics: clinical severity, clinical outcome assessments of lower (TUG, FSHD-TUG) and upper extremity function (RWS, PUL), and patient reported outcomes currently used in the clinic.
- eTUG may be a meaningful measurement that captures various aspects of subjects' disease condition as indicated by its excellent correlations.
- Emerald enables a large number of measurements in patients' homes, and its metrics become increasingly sensitive to change with increased measurements.
- Emerald may be a sensitive tool to assess clinical progression in FSHD trials.

References

- "Extracting Gait Velocity and Stride Length from Surrounding Radio Signals", C.-Y Hsu et. al., ACM Conference on Human Factors in Computing Systems (CHI), 2017.
- "Zero-Effort In-Home Sleep and Insomnia Monitoring using Radio Signals", C.-Y Hsu et. al., ACM International Joint Conference on Pervasive and Ubiquitous Computing (Ubicomp/IMWUT), 2017.