Smartphone Application–Mediated, Supervised, At-Home Telespirometry Identifies Statistically Significant Differences in Erect and Supine Slow Vital Capacity and ALSFRS-R Decline as a Function of Non-invasive Ventilation Treatment Status in Multicenter, Prospective, Longitudinal, Observational Clinical Study [NCT05106569]

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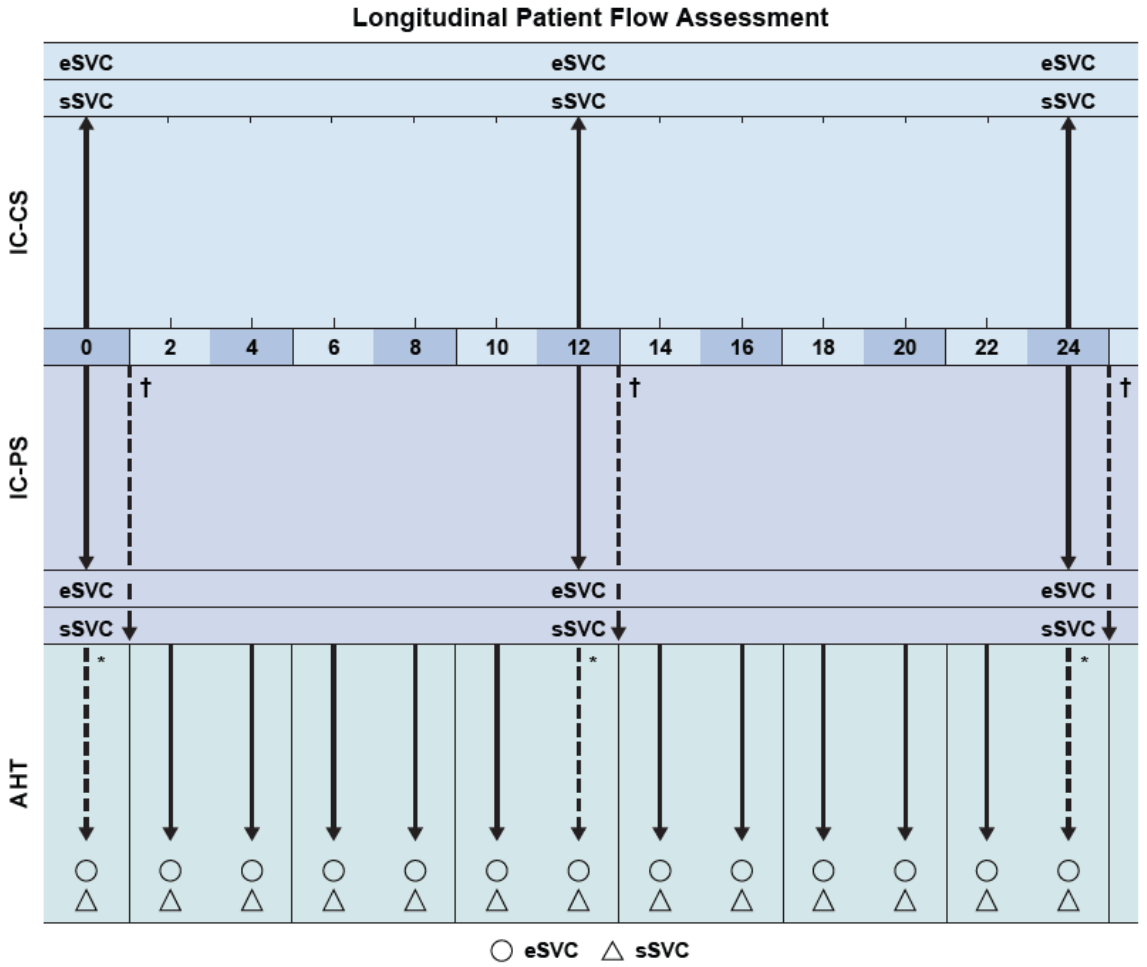
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Background

- A pilot feasibility study to assess both erect and supine forced vital capacity (eFVC, sFVC) longitudinally between clinic visits in subjects with amyotrophic lateral sclerosis (ALS)[1-3] led to the development of the first multi-center implementation study of smartphone application–mediated at-home telespriometry (AHT) feasibility study to assess both erect and supine slow vital capacity (eSVC, sSVC) longitudinally between clinic visits.[4,5] Analysis in subjects with ALS as a function of non-invasive ventilation (NIV) treatment status identified that eSVC and sSVC declined slower in subjects with ALS who were NIV non-users during the 6-month observational study or were on NIV at entry compared with those who started NIV during the observational study[6]

Figure 1. Study Design



AHT, at-home telespirometry; eSVC, erect slow vital capacity; IC-CS, In-Clinic Conventional Spirometry; IC-PS, In-Clinic Portable Spirometry; sSVC, supine slow vital capacity. *AHT at baseline, week 12 and week 24 done in lieu of IC-PS, same day. †AHT at baseline, week 12 and week 24 done in lieu of IC-PS, within 7 days.

Objective

- Extend analysis to evaluate ALS Functional Rating Scale-Revised (ALSFRS-R) twice monthly up to 6 months via AHT in subjects with ALS between quarterly in-clinic eSVC and sSVC assessments in a multicenter, prospective, longitudinal, observational clinical study [NCT05106569] using ZEPHYRx™ Breathe Easy software and Remote Respiratory Monitoring™/Provider Dashboard

Methods

- eSVC and sSVC were measured longitudinally via AHT as previously reported[4-6]
- ALSFRS-R changes, evaluated by research personnel, were examined longitudinally together with eSVC and sSVC in-clinic or via smartphone and analyzed in this report
- Delta eSVC and sSVC, and delta ALSFRS-R_{total} (ALSFRS-R total score) trajectories were analyzed with a linear mixed methods model involving baseline covariates according to NIV usage
- Linear mixed model delta eSVC and sSVC least square (eSVC %–predicted/month) and delta ALSFRS-R least square (ALSFRS-R_{total} points/month) calculation included the covariates: sex, site of onset, age of onset, onset of diagnosis interval, baseline ALSFRS-R at study entry

Results: SVC Monthly Decline Rate According to NIV Usage

- eSVC and sSVC declined slower in subjects with ALS not requiring NIV or subjects already treated with NIV compared with subjects with ALS who started NIV after baseline (**Table 1**)

Table 1. SVC Monthly Decline Rate According to NIV Usage Based on a Linear Mixed Model Analysis

Portable eSVC, %predicted		Not Used Yet	Used After Baseline	Used at Baseline	*P* value
By NIV Use	**n**	39	36	20	0.025
	Baseline	76.7	70.5	67.7	
	LS Mean (SD)	−2.4 (2.5)	−3.9 (2.2)	−2.7 (2.3)	
	Median (IQR)	−2.7 (3.1)	−4.1(3.2)	−2.1 (3.8)	
	Min, Max	−8.3, 3.5	−11.1, −0.4	−6.9, 0.7	

Portable sSVC, %predicted		Not Used Yet	Used After Baseline	Used at Baseline	*P* value
By NIV Use	**n**	32	33	13	0.007
	Baseline	79.4	66.8	67.1	
	LS Mean (SD)	−2.2 (2.4)	−3.9 (2.2)	−2.1 (2.1)	
	Median (IQR)	−2.3 (2.7)	−3.9 (3.3)	−1.9 (2.7)	
	Min, Max	−7.4, 3.6	−12.1, −0.7	−6.8, 1.0	

eSVC, erect slow vital capacity; IQR, interquartile range; LS, least squares; NIV, non-invasive ventilation; sSVC, supine slow vital capacity.

Results: ALSFRS-R_{total} Monthly Decline Rate According to Baseline eSVC

- ALSFRS-R_{total} declined faster in the baseline seated eSVC ≤80%–predicted cohort (−1.9 per month) vs the baseline seated eSVC >80%–predicted cohort (−1.1 per month) (**Table 2**)

Table 2. ALSFRS-R_{total} Decline in Subjects With ALS With Baseline eSVC ≤80%–predicted Compared With Baseline eSVC >80%–predicted

ALSFRS-R_{total}		Baseline eSVC ≤80%–predicted	Baseline eSVC >80%–predicted	*P* value
By Baseline eSVC %–predicted	**n**	57	40	0.001
	Baseline	28	40	
	LS Mean (SD)	−1.6 (1.8)	−0.9 (1.2)	
	Min, Max	−5.5, 0.4	−3.5, 0.9	
	Q1, Q3	−2.7, −0.9	−1.6, −0.4	

ALSFRS-R_{total}, Amyotrophic Lateral Sclerosis Functional Rating Scale-Revised total; eSVC, erect slow vital capacity; LS, least square.

Results: ALSFRS-R_{total} Monthly Decline Rate According to NIV Usage

- ALSFRS-R_{total} decline was slower in subjects with ALS who did not require NIV or subjects with ALS already treated with NIV compared with subjects who started NIV after baseline (**Table 3**)

Table 3. ALSFRS-R_{total} Monthly Decline Rate According to NIV Usage Based on a Linear Mixed Model

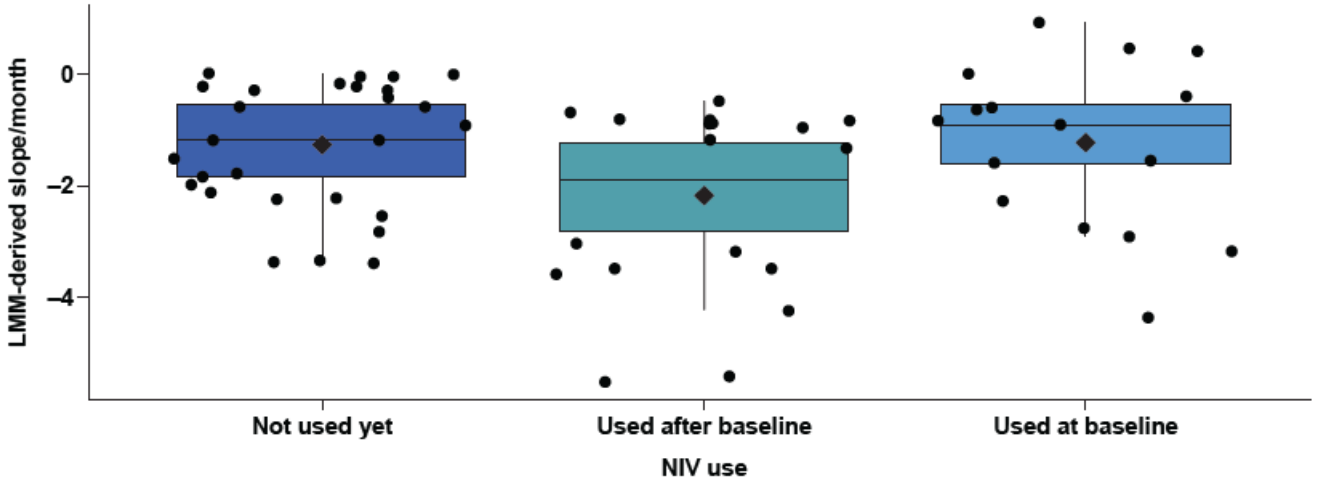
ALSFRS-R_{total}		Not Used Yet	Used After Baseline	Used at Baseline	*P* value
By NIV Use	**n**[a]	40	36	21	0.001
	Baseline	37	33	26.4	
	LS Mean (SD)	−1.3 (1.0)	−2.2 (1.3)	−1.2 (1.3)	
	Median (IQR)	−1.2 (1.3)	−1.9 (1.6)	−0.9 (1.0)	
	Min, Max	−3.4, 0	−5.5, −0.5	−4.4, 0.9	

ALSFRS-R_{total}, Amyotrophic Lateral Sclerosis Functional Rating Scale-Revised total; IQR, interquartile range; LS, least square; NIV, non-invasive ventilation.
[a]Total N=97 (1 participant in “not used yet” NIV cohort did not have an ALSFRS-R_{total} slope).

Results: ALSFRS-R_{total} Monthly Decline Rate According to NIV Usage (continued)

- ALSFRS-R_{total} score declined faster in subjects with ALS that started NIV during the study, compared to the cohort that never used NIV and the cohort that used NIV at baseline (**Figure 2**)
- Subjects with ALS who were on NIV at baseline or never used NIV after baseline had a higher proportion (19/61, 31.1%) of subjects with ALS with delta ALSFRS-R_{total} <0.5 points per month than subjects with ALS who started NIV after baseline (1/36; 2.8%) (Chi-square *P*=0.0008)
- Subjects with ALS who were on NIV at baseline or never used NIV after baseline had a lower proportion (1/61; 1.6%) of subjects with ALS with delta ALSFRS-R_{total} >3.5 points per month than subjects with ALS who had to start NIV after baseline (6/36; 16.7%) (Chi-square *P*=0.0057)

Figure 2. ALSFRS-R_{total} Monthly Decline Rate According to NIV Usage



ALSFRS-R_{total}, Amyotrophic Lateral Sclerosis Functional Rating Scale-Revised total; LMM, linear mixed model; NIV, non-invasive ventilation.

Discussion/Conclusions

- This first prospective, longitudinal, observational study of smartphone-mediated AHT eSVC and sSVC measurement identified statistically significant differences in monthly decline of eSVC and sSVC and ALSFRS-R_{total} score between subjects with ALS not requiring NIV and those on NIV at baseline compared with subjects with ALS not on NIV at baseline who started NIV during the study, which were not previously studied[7,8]
- The proportion of ALS subjects with slower delta ALSFRS-R_{total} progression was 10-fold higher in those subjects who did not progress to NIV utilization during the observational study or who were already on NIV at baseline
- The proportion of ALS subjects with faster delta ALSFRS-R_{total} progression was 8-fold lower in those subjects who did not progress to NIV utilization during the observational study or who were already on NIV at baseline
- NIV use, due to its potential to alter both respiratory and motor function, needs to be more carefully captured in clinical trials and clinical studies to permit correct conclusions regarding pharmacological and device interventions

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- **GS, EIY, DW, DM, BS, UD, LD, MP, JC, EB, JM, BN, AI, GB, DV, SA,** and **PC** have no disclosures to report.
- **TK** is an employee of Tanabe Pharma Corporation.
- **KP,** and **SA** are employees of Shionogi Inc.
- **BRB** is a consultant for Tanabe Pharma America, Inc.
- **EIY** is a consultant for, and her institution has received funding from, Tanabe Pharma America, Inc.
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