

Online supplement, efficacy in the open-label part of the study

During the open label follow-up, the mean sitting SVC value declined from the baseline value of 73.7% points (n = 66) to 70.5 points at 1 month (n = 43), 66.4 at 3 months (n = 40), and 63.3% at 6 months (n = 30). The average decline in sitting SVC was 1.75 % points/month during the 9 month study.

The mean supine SVC declined from the baseline value of 73.3% points (n = 66) to 64.9 (n = 43) at 1 month, 63.6 (n = 35) at 3 months, and 61.8% points (n = 28) at 6 months. The average decline in supine SVC was 1.93 % points/month during the 9 month study.

The mean ALSFRS-R total score declined from the baseline value of 36.7 points (n=66) to 32.6 (n=46) at 1 month, 30.2 (n=43) at 3 months, and 28.9 (n = 36) at 6 months. The average decline in ALSFRS-R total score was 1.03 points/month during the 9 month study.

The mean SNP value declined from the baseline value of 56.7 cmH<sub>2</sub>O (n = 66) to 52.2 (n = 46) at 1 months, 53.4 (n = 41) at 3 months, and 51.1 (n = 34) at 6 months. The average decline in SNP was 1.76 cmH<sub>2</sub>O /month during the 9 month study. During the open label follow-up, the mean overnight SpO<sub>2</sub> did not change from the baseline value. Worsening EQ-5D-5L and SF-36 results were seen throughout the study period (data not shown).