

	Diagnostic sleep study Median (IQR, range)	NIV titration sleep study Median (IQR, range)	Difference (p value)
Age (years)	14.04 (2.67, 10.47-17.89)	14.66 (2.35, 10.47-17.96)	0.33
Total AHI	8.2 (12.8, 1.8-137)	1.9 (2.8, 0-15.9)	<0.0005*
AHI REM	26.7 (32.5, 2.5-117.6)	0.4 (3.3, 0-30.8)	<0.0005*
AHI NREM	1.8 (5.3, 0-15.1)	1.6 (3.6, 0-19.7)	0.15
Total Central AHI	3.1 (5.8, 0-38.3)	1.2 (3, 0-15.9)	0.06
Central AHI REM	7.1 (20.7, 0-116.4)	0 (3.3, 0-30.8)	0.0002*
Central AHI NREM	0.9 (1.5, 0-14)	0.9 (4.2, 0-19.7)	0.88
Total Obstructive AHI	2 (6.1, 0-30.3)	0 (0.4, 0-4.3)	0.0001*
Obstructive AHI REM	2.7 (22, 0-117.6)	0 (0, 0-3.4)	<0.0005*
Obstructive AHI NREM	0.5 (1.6, 0-14.9)	0 (0.4, 0-4.8)	0.02*
Maximum TcCO ₂	51.8 (8.70, 43.6-78.2)	46.5 (6, 38.6-60.3)	0.004*
Maximum REM TcCO ₂	51.8 (9.30, 43.6-78.2)	45.8 (6.70, 37.7-60.3)	0.0006*
Maximum NREM TcCO ₂	49 (6.40, 0-70.4)	46 (7.10, 0-59.6)	0.07
Minimum SpO ₂	83 (13, 59-92)	90 (6, 0-95)	<0.0005*
Min SpO ₂ REM	81 (13, 50-94)	93 (4, 83-96)	<0.0005*
Min SpO ₂ NREM	90 (6, 0-96)	90 (6, 0-96)	0.21
Respiratory arousal index	0.87 (0.99, 0.12-16)	0.58 (0.94, 0-16)	0.40
Total arousal index	4.535 (4.72, 0.43-13.47)	4.455 (3.89, 0-13.47)	0.58

Table 1S: Changes in sleep study parameters between diagnostic and NIV titration sleep studies. NIV – non-invasive ventilation, AHI – apnoea hypopnea index, REM – rapid eye movement sleep, NREM – non-rapid eye movement sleep, TcCO₂ – transcutaneous carbon dioxide level, mmHg – millimeters of mercury, SpO₂ – oxygen saturation level.

	Total Group		Difference (p value)	Steroid Users		Difference (p value)	Steroid Naïve		Difference (p value)
	Mean (95% CI)			Mean (95% CI)			Mean (95% CI)		
	Pre-NIV	Post-NIV		Pre-NIV	Post-NIV		Pre-NIV	Post-NIV	
FEV ₁ (l)	-0.05 (-0.07, -0.02)	-0.06 (-0.09, -0.04)	<0.001*	-0.03 (-0.07, 0.004)	-0.07 (-0.12, -0.01)	0.11	-0.04 (-0.09, 0.006)	-0.07 (-0.13, -0.01)	<0.001*
FEV ₁ z-score	-0.59 (-0.66, -0.52)	-0.37 (-0.45, -0.30)	<0.001*	-0.53 (-0.67, -0.39)	-0.63 (-0.78, -0.47)	<0.001*	-0.64 (-0.76, -0.51)	-0.31 (-0.43, -0.19)	<0.001*
FEV ₁ % predicted	-7.86 (-8.78, -6.94)	-4.78 (-5.74, -3.83)	<0.001*	-6.66 (-8.37, -4.94)	-7.86 (-9.77, -5.95)	<0.001*	-8.41 (-9.95, -6.87)	-4.24 (-5.82, -2.66)	<0.001*

Table 2S: Summary of annual rates of decline in lung function for the Total Group, Steroid User and Steroid Naïve groups before and after NIV initiation. FEV₁ – forced expiratory volume in one second.

	Total Group	Steroid Users	Steroid Naïve	Difference between Steroid Users and Steroid Naïve (p value)
n	29	9	8	
Age (years) median (IQR, range)	14.66 (2.35, 10.47-17.96)	14.04 (2.67, 11.68-17.39)	14.33 (2.44, 10.18-16.71)	0.96
FVC (l) median (IQR, range)	1.41 (0.67, 0.34-2.61)	1.63 (0.71, 1.38 - 2.28)	1.36 (0.99, 0.67 - 2.38)	0.41
FVC z-score median (IQR, range)	-5.24 (2.65, -8.60 - -1.16)	-3.26 (1.21, -5.24 - -1.85)	-5.41 (1.87, -7.0576 - -1.16)	0.02*
FVC % predicted median (IQR, range)	40.83 (2.77, 9.50 – 86.60)	62.95 (13.52, 40.83 - 79.90)	39.3 (19.95, 21.43 - 86.60)	0.03*
FEV ₁ (l) median (IQR, range)	1.27 (0.56, 0.33 – 2.33)	1.46 (0.5, 1.01-1.87)	1.22 (0.94, 0.57-2.33)	0.84
FEV ₁ z-score median (IQR, range)	-5.00 (1.95, -6.81 - -1.07)	-3.16 (0.82, -5.34- -1.87)	-4.78 (1.89, -6.22- -1.07)	0.11
FEV ₁ % predicted median (IQR, range)	38.2 (27.21, 10.40 – 88.00)	61.25 (9.64, 32.91-78.30)	41 (25, 20.94-88.00)	0.10

Table 3S: Summary of lung function prior to beginning NIV Total Group, and Steroid Users compared to Steroid Naïve subjects. FVC – forced vital capacity, FEV₁ – forced expiratory volume in 1 second.

FVC ₁ (l) rate of decline (pa) mean (95% CI)	-0.082 (-0.10, -0.06)	0.003 (-0.03, 0.3)	-0.09 (-0.12, -0.06)	<0.001*
FVC z-score rate of decline (pa) mean (95% CI)	-0.65 (-.70, -0.61)	-0.58 (-0.67, -0.50)	-0.68 (-0.75, -0.60)	<0.001*
FVC % predicted rate of decline (pa) mean (95% CI)	-7.17 (-7.65, -6.69)	-6.60 (-7.55, -5.65)	-7.39 (-8.23, -6.54)	<0.001*
FEV ₁ (l) rate of decline (pa) mean(95% CI)	-0.06 (-0.08, -0.05)	-0.005 (-0.32, -0.02)	-0.07 (-0.11, -0.04)	<0.01*
FEV ₁ z-score rate of decline (pa) mean(95% CI)	-0.53 (-0.57, -0.49)	-0.52 (-0.60, 0.43)	-0.57 (-0.66, -0.49)	<0.001*
FEV ₁ % predicted rate of decline (pa) mean(95% CI)	-7.07 (-7.60, --6.54)	-6.55 (-7.57, -5.54)	-7.67 (-8.73, -6.62)	<0.001*

Table 4S: Summary of rates of decline throughout adolescence (independent of NIV use) for the *Total Group*, and *Steroid Users* compared to *Steroid Naïve* subjects. FVC – forced vital capacity, FEV₁ – forced expiratory volume in 1 second.