Tables and Figures

Table 1 Neuromuscular disorders (NMD) of children completing study

	NMD (n=30)	
	Unsupported (22)	**NIV(8)**
Duchenne muscular dystrophy (DMD)	13	4
Congenital muscular dystrophy	2	0
Spinal muscular atrophy (SMA2/3)	2	2
Congenital myopathy	3	2
Myasthenia gravis	1	0
Myotonic dystrophy	1	0

Table 2 Characteristics of participants in children with neuromuscular disease (NMD) and controls

Parameter	NMD	Controls
Age mean (±SD) years	11.5 (±3)	12 (±2)
Gender (M/F)	21/9	18/12
Weight median(range) (kg)	39 (19.8 to 60)	43(24 to 54)
Height median(range) (cm)	140.3 (114 to 166)	143 (123 to 166)
BMI median (range) (Kg/m2)	20.8(10 to 28)	17.6(15.1 to 19)

Table 3 Pulmonary Function Tests

NMD (n=30)		Controls	P Value*

	Unsupported (n=22)	NIV (n=8)	All	(n=30)	
Age mean year (±SD)	11.5 (±2.8)	14 (±2.1)	12.3 (±3.0)	12 (±2)	NS
Gender (M/F)	16/6	6/2	21/9	17/13	NS
BMI kg/m2 median(range)	19.9 (11 to 28)	21.7 (10 to 25)	20.8 (10 to 28)	17.6 (15.1 to 19)	0.024
FEV_1(si)mean % (±SD)	83 (±21.9)	72 (±17.9)	78 (±21.9)	95 (±5.2)	<0.001
FVC (si) mean% (±SD)	78 (±20.5)	70 (±24.5)	75 (±20.5)	98 (±4.8)	<0.0001
FEV_1(su)%	83 (±24.1)	53 (±22.2)	72 (±22.1)	94 (±6)	<0.0001
FVC(su)% mean (SD)	70 (±22)	58 (±18.8)	66 (±21)	94(±4)	<0.0001
FEV_1/FVC% mean (SD)	86 (±7)	85 (±7)	86 (±7)	95	0.003
%ΔFVC(si–su) mean(SD)	7 (±8)	12 (±6)	9 (±11)	4 (±3)	0.03
% diff ΔFVC(si–su)	8.9	17	12	4	0.03
PSQ-22 Median (range)	0.28 (0.12 to 0.5)	0.31 (0.2 to 0.5)	0.30 (0.12 to 0.5)	0.25	NS
mESS median(range)	5 (0 to 6)	9 (0 to 7)	7 (0 to 6)	5	NS

Comparison was made using paired t tests between children with NMD (all) and controls

Abbreviations NMD = Neuromuscular disorders, NIV= Nocturnal non-invasive ventilation, FEV1(si) = Forced expiratory volume 1 sec in sitting position, FVC(si)= Forced vital capacity in sitting position, FVC(su)= Forced vital capacity in supine position, ΔFVC(si-su)= Absolute change change in spirometry, PSQ-22= Pediatric sleep questionnaire, % diff ΔFVC=% change mESS= Modified Epworth sleepiness scale

Table 4 Polysomnography data from children with neuromuscular disease

Polysomnography data (n=30)			
	All NMD (n=30)	**SB (n=22)**	**NIV (n=8)**
TST min mean(±SD)	351(±8.8)	371(±10.2)	323(±8.6)
Non-REM% mean(±SD)	77(±15)	76(±14)	81±(8.8)
REM% mean(±SD)	23±(7.8)	24±(12)	19±(10)
Arousal index /hr (±SD)	7.6(±11)	5.4(±5)	10.2(±10.2)
Baseline saturation(±SD)	96 (±5)	98(±4)	94(±4)
Minimum mean (±SD)saturation	88.4 (±4.2)	91(±3.9)	87(±2.2)
AHI (total)/hr mean(±SD)	6.9(±5.9)	7.4(±5.8)	11.4(±6.0)
OAHI mean(±SD)	5.2(±4.0)	4.2(±3.9)	8.9(±6.5)
Central AHI mean(±SD)	3.3(±2.2)	3.2(±1.2)	2.5(±3.6)
REM AHI/hr mean(±SD)	14.1(±5.3)	14.8(±10.2)	18.3(±7.0)
CO_2 baseline mean (±SD)	44 (±3.5)	42(±4.7)	43(±2.1)
CO_2 REM mean (±SD)	50(±4.4)	47(±4.7)	52 (±1.3)
CO_2 rise mean (non-REM to REM mmHg)	6(±2.7)	5(±1.8)	9(±2.0)

Results presented as mean (± SD) unless otherwise stated

TST =total sleep time, SWS= Slow wave sleep,AHI=Apnoea hypopnea index,OAHI=obstructive apnoea hypopnea index

SB- spontaneously breathing, NIV= non-invasive ventilation

Figure 1 FVC% in supine spirometry vs TcCO2 from baseline to REM sleep

FVC % change from sitting to supine versus difference CO2 (non-REM to REM) in SB/NIV