Original articles

Respiratory failure and sleep in neuromuscular disease

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Abstract

Sleep hypoxaemia in non-rapid eye movement (non-REM) and rapid eye movement (REM) sleep was examined in 20 patients with various neuromuscular disorders with reference to the relation between oxygen desaturation during sleep and daytime lung and respiratory muscle function. All the patients had all night sleep studies performed and maximum inspiratory and expiratory mouth pressures (PI and PEmax), lung volumes, single breath transfer coefficient for carbon monoxide (KCO), and daytime arterial oxygen (Pa_{O_2}) and carbon dioxide tensions (Pa_{CO_2}) determined. Vital capacity in the erect and supine posture was measured in 14 patients. Mean (SD) PI max at RV was low at 33 (19) cm H_2O (32% predicted). Mean PE max at TLC was also low at 53 (24) cm H_2O (28% predicted). Mean daytime Pa_{O_2} was 67 (16) mm Hg and Pa_{CO_2} 52 (13) mm Hg (8·9 (2·1) and 6·9 (1·7) kPa). The mean lowest arterial oxygen saturation (Sa_{O_2}) was 83% (12%) during non-REM and 60% (23%) during REM sleep. Detailed electromyographic evidence in one patient with poliomyelitis showed that Sa_{O_2}% during non-REM sleep was maintained by accessory respiratory muscle activity. There was a direct relation between the lowest Sa_{O_2} value during REM sleep and vital capacity, daytime Pa_{O_2}, Pa_{CO_2}, and percentage fall in vital capacity from the erect to the supine position (an index of diaphragm weakness). The simple measurement of vital capacity in the erect and supine positions and arterial blood gas tensions when the patient is awake provide a useful initial guide to the degree of respiratory failure occurring during sleep in patients with neuromuscular disorders. A sleep study is required to assess the extent of sleep induced respiratory failure accurately.

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There have been extensive studies on the mechanical properties of the lung in patients with respiratory muscle weakness,[1 2] and reports of sleep hypoxaemia in several specific neuromuscular disorders.[3–7] Arterial oxygen and carbon dioxide tensions in these patients when awake have been shown to improve after nocturnal assisted ventilation.[6–9] There have been few systematic analyses of ventilation and oxygenation throughout the night in patients with a wide range of neuromuscular disorders.

In patients with kyphoscoliosis Sawicka and Branthwaite[10] found that the degree of nocturnal desaturation was related to arterial oxygen saturation when the patient was awake and was more prominent in those with paralytic kyphoscoliosis, who had the greatest reduction in vital capacity. In a further study of patients with kyphoscoliosis Midgren *et al* [11] found that those with a low vital capacity and a large fall in vital capacity when they changed from sitting to the supine posture were more likely to have arterial hypoxaemia during sleep. In this study all the patients with kyphoscoliosis had lower than predicted values for maximum respiratory muscle pressures measured at the mouth, but there was no difference in maximum pressures between those who did and those who did not become hypoxaemic at night. By contrast, Smith *et al* [3] reported that the severity of sleep disordered breathing in patients with Duchenne muscular dystrophy could not be reliably predicted from daytime pulmonary function, though maximum static expiratory pressure measured at the mouth was lower in those who developed desaturation during sleep.

The aim of our study was to examine arterial oxygenation and breathing pattern during sleep in patients with a wide variety of neuromuscular disorders. We have related these measurements to blood gas tensions measured when they were awake during the day and to assessment of respiratory muscle strength and lung function, particularly spirometric measurements.

Methods

We studied 20 patients with various neuromuscular disorders. Their mean age was 47 (range 21–71) years and body mass index 22 (range 15–33; normal range 20–25). Five patients were current smokers and eight ex-smokers, and six had never smoked; no smoking history was obtained for one patient. The diagnoses are given in table 1. All patients had been assessed by a neurologist and most had been investigated with muscle biopsy and electromyographic studies. Three patients also had lung disease. Patient 1 had mild and well controlled asthma. Patient 15 had bronchiectasis, treated with regular postural drainage and antibiotics; severe respiratory muscular weakness was the predominant problem. Patient 7 had polymyositis and associated pulmonary interstitial fibrosis. Other patients had experienced

Table 1 Diagnosis, daytime arterial oxygen and carbon dioxide tensions (Pa_{O_2}, Pa_{CO_2}), and arterial oxygen saturation (Sa_{O_2}) during wakefulness and sleep

Patient No	Diagnosis	Pa_{O_2} (mm Hg)	Pa_{CO_2} (mm Hg)	Sa_{O_2}†				
					Non-REM		REM	
				Awake	Minimum	Typical	Minimum	Typical
1	Spinal muscular atrophy	57	53	92	52	58	30	36
2	Pompes disease	49	83	95	88	89	50	89
3	Guillain-Barré variant	101	39	98	97	97	95	98
4	Poliomyelitis	79	53	96	92	93	65	68
5	Dystrophia myotonia	85	46	97	95	97	89	97
6	Muscular dystrophy	–	45	80	78	90	69	80
7	Polymyositis	85	39	95	83	84	90	91
8	Central core myopathy	44	84	87	76	84	44	65
9	Acid maltase deficiency	74	49	89	88	90	61	80
10	Poliomyelitis	53	61	88	*	*	33	55
11	Dystrophia myotonia	66	46	96	91	94	84	93
12	Poliomyelitis	56	44	87	78	84	57	75
13	Poliomyelitis	67	48	95	64	87	32	55
14	Poliomyelitis	83	38	94	93	94	80	91
15	Dystrophia myotonia	58	53	94	80	86	51	64
16	Motor neurone disease	74	46	95	94	95	78	89
17	Muscular dystrophy	51	53	90	66	77	44	60
18	Motor neurone disease	70	39	97	92	95	91	95
19	Muscular dystrophy	79	50	80	*	*	31	57
20	Dystrophia myotonia	45	68	76	79	84	28	55
	Mean	67	52	91	83	88	60	74
	SD	16	13	6	12	9	23	18

*Sleep stage unable to be determined.
†Corrected according to ref 17 when measured value < 50%.
Conversion: Traditional to SI units—Blood gas tensions: 1 mm Hg ≈ 0·133 kPa.

pulmonary complications related to their respiratory muscle weakness—recurrent atelectasis, lobar collapse, pneumonia, and even respiratory arrest.

Forced expiratory volume in one second (FEV_1) and forced vital capacity (FVC) were obtained with a Godard expirograph (Utrecht, Holland) and lung volumes by closed circuit helium dilution (Pulmotest, Godart). Measurements were performed with the patient in the sitting posture. For 14 patients VC was also measured with the patient supine, to assess diaphragm weakness.[12 13]

The transfer coefficient for carbon monoxide (K_{CO}) was measured by the single breath technique described by Ogilvie and coworkers.[14] Arterial oxygen and carbon dioxide tensions (Pa_{O_2}, Pa_{CO_2}) were measured with a blood gas analyser (Corning, Medfield, Massachusetts, USA) from samples taken from the radial artery with the patient sitting erect. Maximum inspiratory mouth pressure at residual volume ($P_I max$) and maximum expiratory mouth pressure at total lung capacity ($P_E max$) were measured with a Hewlett-Packard pressure transducer (HP-267BC). The mouthpiece contained a fixed leak to avoid artefacts caused by the cheek muscles. Values were compared with the normal values of Black and Hyatt.[15] Patients were taught the maximum inspiratory and expiratory mouth pressure manoeuvres by the respiratory technician on the day before the formal study. The values quoted are the highest obtained from five tests.

All night sleep studies were performed on all patients in a sound attenuated room. The sleep variables included values derived from two electroencephalographic channels (C4/A1, C3/A2), a submental muscle electromyogram, and two ocular channels. Sleep was staged according to standard criteria[16] and was broadly separated into non-rapid eye movement (non-REM) and rapid eye movement (REM) phases.

The electrocardiogram and heart rate were monitored continuously on a 16 channel polygraph (Grass 78, Quincy, Massachusetts). Arterial oxygen saturation (Sa_{O_2}) was monitored by ear oximeter (Hewlett-Packard 47201A) during sleep and for 30–60 minutes before the onset of sleep. A slow recording of Sa_{O_2} was obtained on a strip chart recorder, to obtain both minimum and "typical" Sa_{O_2} values. Minimum Sa_{O_2} was the lowest value recorded during sleep; "typical" Sa_{O_2} was the estimated minimum value of Sa_{O_2} that was sustained for the entire non-REM or REM period. A correction factor was applied to all minimum values for Sa_{O_2} less than 50%.[17] In most patients Sa_{O_2} stabilised during any given epoch of sleep so "typical" oxygen saturation could be measured accurately. Transcutaneous carbon dioxide tension (Ptc_{O_2}) (Hewlett-Packard 47210A capnometer) was recorded on the strip chart recorder during sleep in eight patients. Chest wall and abdominal movements were monitored throughout sleep by inductance plethysmography (Respitrace, Ambulatory Monitoring Inc, New York). Airflow through the nose was measured by a differential pressure transducer linked to a pair of nasal prongs.

In one patient with poliomyelitis (No 4) electromyograms of inspiratory and expiratory muscle activity were obtained with surface electrodes over several nights by a scientist with considerable expertise in electromyographic techniques. Recordings were obtained from the sternomastoid (mid belly), genioglossus (submental), abdominal (2 cm lateral to umbilicus) and intercostal muscles (second right intercostal space 2 cm lateral to the sternum). Two electrodes were positioned in the right subcostal region in the midclavicular line, 2 cm apart to record diaphragm activity. It is recognised that the signal from such electrodes may include activity from the surrounding muscles.

Table 2 Vital capacity (VC), fall in VC from the erect to the supine posture, and maximum inspiratory and expiratory mouth pressures

Patient No	VC		Maximum pressures: Inspiratory		Maximum pressures: Expiratory	
	% pred	% fall	cm H_2O	% pred	cm H_2O	% pred
1	32	39	27	31	53	35
2	33	28	24	32	46	21
3	89	3	35	41	52	34
4	39	42	44	36	60	27
5	62	14	46	52	30	20
6	36	33	25	20	75	33
7	76	0	95	90	60	28
8	37	2	30	24	60	26
9	67	46	70	57	115	51
10	39	35	26	33	70	47
11	63	12	35	24	50	23
12	51	5	20	19	30	23
13	34	28	27	38	42	30
14	81	5	25	25	95	51
15	34	-	28	24	20	14
16	75	-	25	24	65	32
17	18	-	30	24	35	15
18	42	-	18	17	32	16
19	12	-	16	13	36	16
20	27	-	12	15	32	22
Mean	47	21	33	32	53	28
SD	22	17	19	18	24	11

Table 3 Mean (SD) lung volumes (including fall in vital capacity (VC) from the erect to the supine posture) and transfer coefficient for carbon monoxide (KCO)

TLC (% pred) n=20	FRC (% pred) n=20	RV (% pred) n=20	VC (% pred) n=20	VC (% fall) n=14	KCO (% pred) n=15	FEV_1/FVC% n=20
66	76	103	47	21	82	85
15	20	37	22	17	20	9

TLC—total lung capacity; FRC—functional residual capacity; RV—residual volume; FVC—forced vital capacity.

ANALYSIS

Correlation coefficients were obtained by linear regression analysis. Mean values with standard deviations in parentheses are given unless otherwise stated.

Results

Clinical diagnosis, daytime arterial blood gas tensions, and "typical" and minimum Sao_2% during non-REM and REM sleep are shown in table 1; vital capacity (VC), percentage fall in VC when the patient changed from the erect to supine posture, and PImax and PEmax in table 2; and mean values for lung volumes and KCO in table 3. Mean (SD) PImax at residual volume was low at 33 (19) cm H_2O (32% predicted). There was also considerable expiratory muscle weakness with a mean PEmax measured at total lung capacity of 53 (24) cm H_2O (28% predicted). Mean VC (% predicted) was 47 (SD 22), TLC 66 (15), and RV 103 (37).

The 14 patients whose VC was measured when they were erect and supine had a mean fall of 21% VC on changing from the seated to the supine posture. Seven patients showed a fall in VC on assuming the supine posture of over 25% and three of under 5%. Each of the seven patients with a fall in VC of 25% or more had paradoxical abdominal movement when breathing in the supine posture—that is, inward abdominal movement during inspiration.

When the patients were awake the mean (SD) Pao_2 was 67 (16) mm Hg and $Paco_2$ 52 (13) mm Hg.* KCO was usually preserved (mean 82% (20%) predicted).

In almost all patients Sao_2 was stable during wakefulness; the mean value was 91%. During non-REM sleep the minimum Sao_2 was 83% (12%) and "typical" Sao_2 was 88%; this was usually steady during non-REM sleep.[9] Three patients had a minimum Sao_2 during non-REM sleep of under 75%. During REM sleep some patients experienced repetitive profound falls in Sao_2 to below 50% (mean minimum Sao_2 60% (23%); "typical" Sao_2 74% (18%)). There were more repetitive falls in Sao_2 during REM sleep than in non-REM sleep. The sleep pattern was that of grossly disorganised and fragmented sleep. In two patients (Nos 10 and 19) the sleep patterns could not be scored in the usual way; sleep has been recorded as REM sleep as the pattern resembled this most closely. The mean maximum value for $PtcCO_2$ (n=8) was 80 (23) mm Hg during non-REM sleep and 90 (22) mm Hg during REM sleep (see example in fig 1). Four patients (Nos 3, 5, 7, and 18) maintained an adequate Sao_2 during REM sleep. For PImax their respective % predicted values were 41, 52, 90, and 17, and for percentage fall in VC from the erect to the supine posture 3, 14, and 0% (not measured in one).

The pattern of respiratory muscle activity during both non-REM and REM sleep was examined in patient 4, who had longstanding poliomyelitis and a weak diaphragm (fall in VC from erect to supine posture 42%). During non-REM sleep adequate ventilation was maintained by the combined use of abdominal and intercostal muscles (figs 2*a* and 2*b*). Accessory muscle activity disappeared during REM sleep and returned only on arousal. Sleep hypoxaemia occurred during REM sleep in this patient with poor diaphragm function (fig 2*c*).

PImax % predicted and minimum or "typical" Sao_2 during REM sleep were not significantly related. There was, however, a strong correlation between minimum Sao_2 during REM sleep and vital capacity % predicted

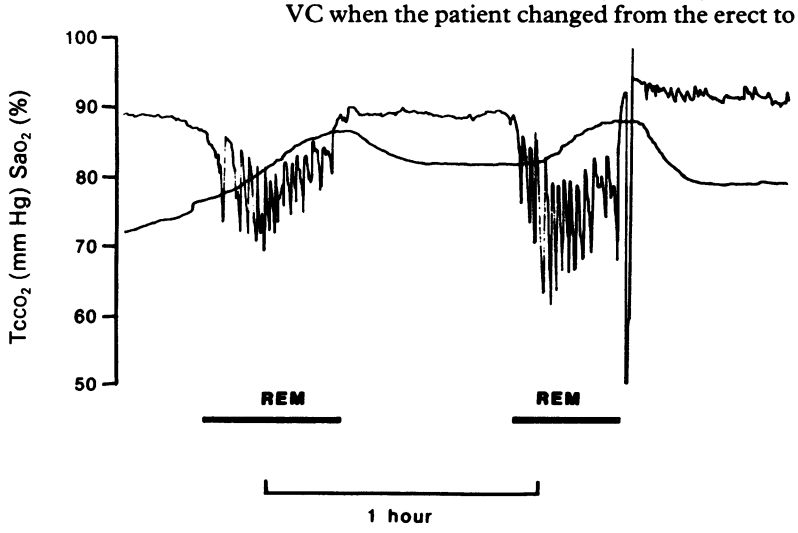
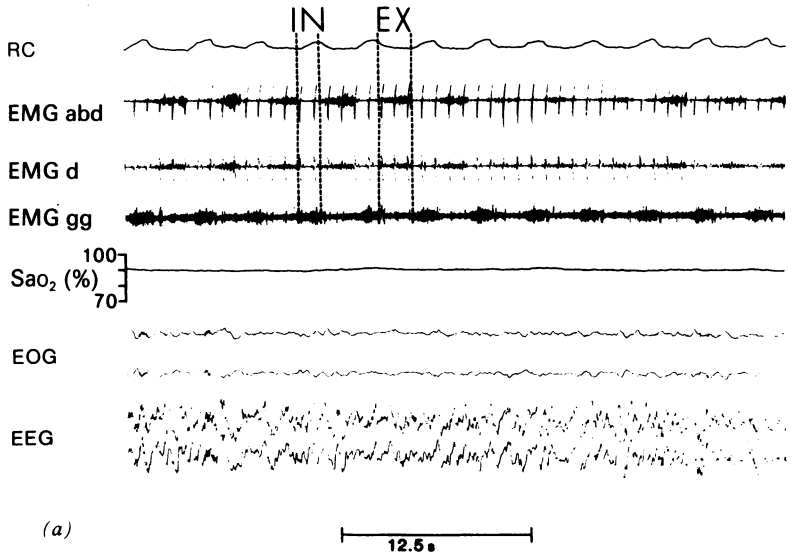


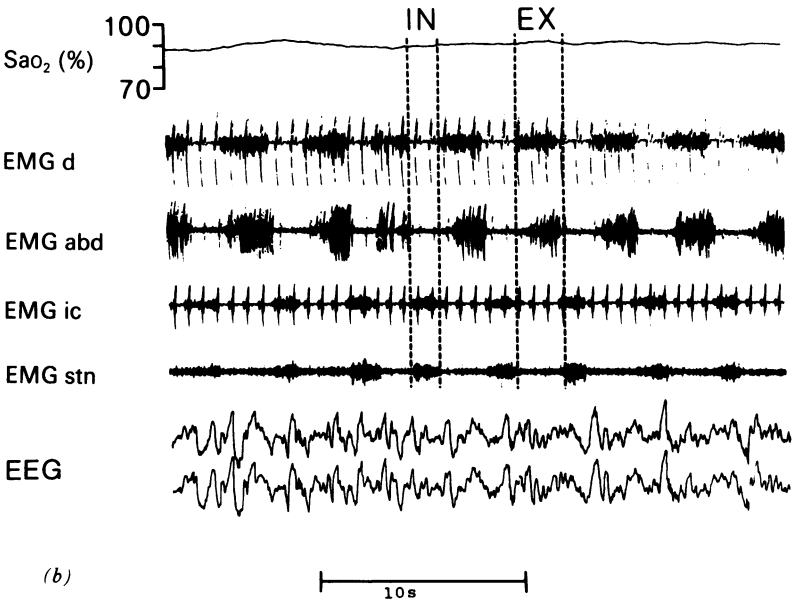
Figure 1 Part of the recording from the all night sleep study in patient 9 showing arterial haemoglobin oxygen saturation (Sao_2) and transcutaneous carbon dioxide ($Tcco_2$). Desaturation in REM sleep and hypercapnia are shown. 1 mm Hg ≈ 0·133 kPa.

*1 mm Hg ≈ 0·133 kPa.

(a) Record showing the pattern of widespread activation of respiratory muscles during non-REM in patient 4, who had longstanding poliomyelitis and a weak diaphragm. IN—inspiration; EX—expiration; RC—rib cage movement; EMG abd—abdominal muscle electromyogram; EMG d—subcostal surface electromyogram recording of respiratory electrical activity (diaphragm); EMG gg—genioglossus electromyogram; Sao_2—arterial oxygen saturation; EOG—electro-oculogram; EEG—electroencephalogram. Note the absence of inspiratory EMG activity in the subcostal diaphragm electrodes as well as substantial expiratory abdominal activation.



(b) Record from a different night, showing widespread activation of accessory muscles during non-REM sleep. This example shows activation of inspiratory parasternal intercostal (EMG ic) and sternomastoid muscles (EMG stn) as well as substantial expiratory abdominal activation. Note the absence of inspiratory EMG activity in the subcostal diaphragm electrodes.



(c) Record made during REM sleep. Episodes of progressive desaturation terminated by transient motor arousals. Accessory muscle activity faded out (see abdominal and genioglossus activity) and only returned on arousal. There was virtually no subcostal electrical activity. Sleep hypoxaemia developed when accessory muscle activity was inhibited in REM sleep. 1 mm Hg ≈ 0·133 kPa.

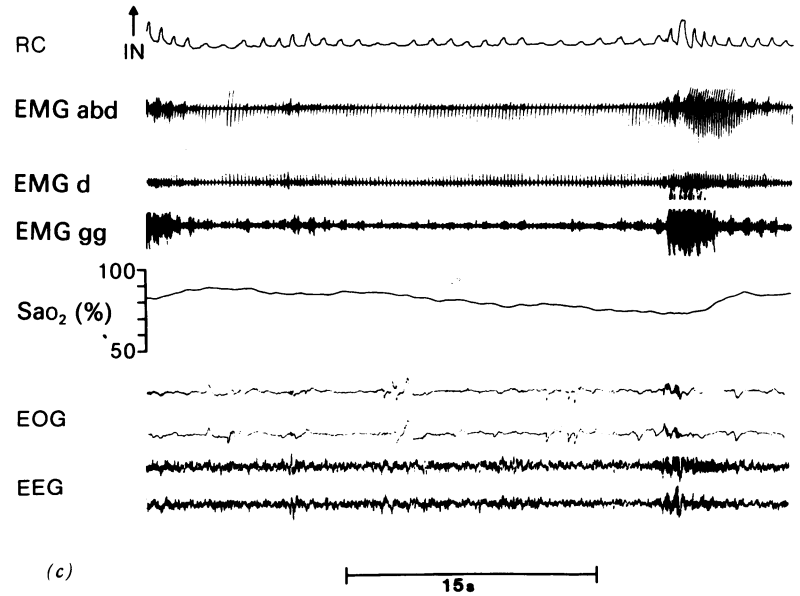


Figure 2 Polygraph recordings from patient 4.

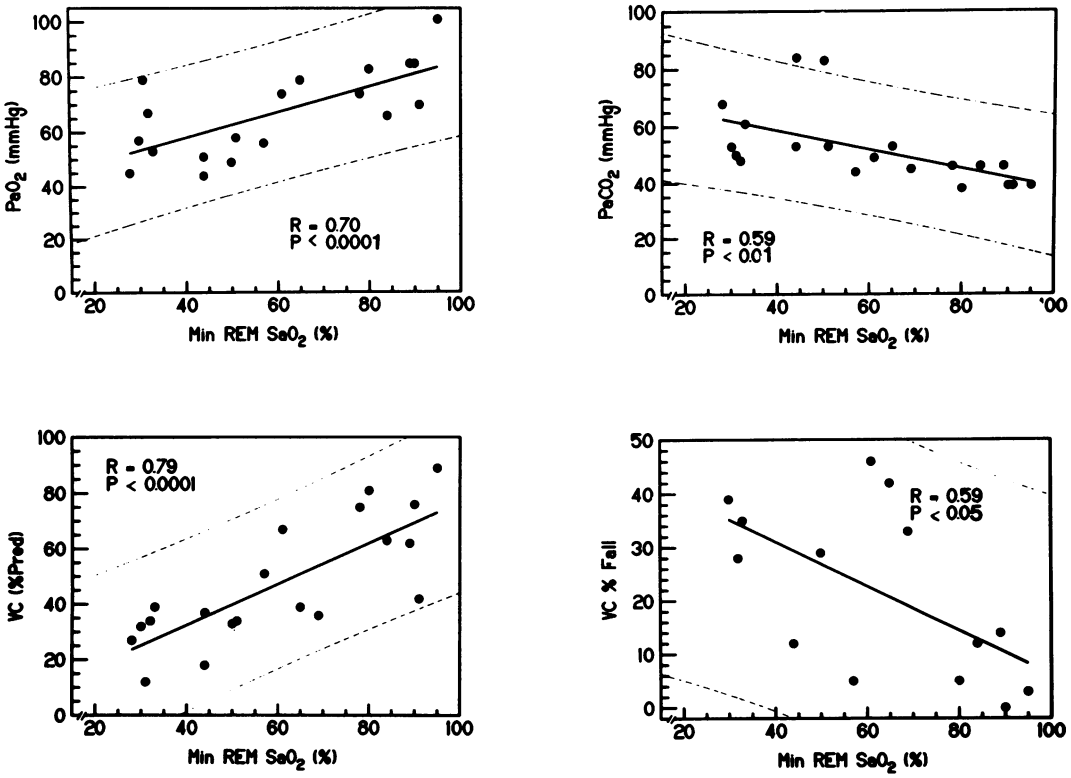


Figure 3 Relation between minimum REM arterial oxygen saturation (Sao_2) and daytime arterial oxygen and carbon dioxide tensions (Pao_2, $Paco_2$), vital capacity (VC) % predicted, and the percentage fall in VC from the erect to the supine posture. The regression line and 95% confidence limits are shown. 1 mm Hg ≈ 0·133 kPa.

($p < 0·001$), daytime Pao_2 ($p < 0·001$), and daytime $Paco_2$ ($p < 0·01$; fig 3). The relation between the minimum Sao_2 during REM sleep and the percentage fall in VC from the erect to the supine position was significant ($p < 0·05$). "Typical" Sao_2 during REM sleep was related to Pao_2 ($p < 0·01$), VC ($p < 0·001$), and percentage fall in VC from the erect to the supine posture ($p < 0·05$).

Discussion

Our results show that patients with neuromuscular disorders and respiratory muscle weakness may maintain oxygenation during non-REM sleep, but typically develop oxygen desaturation during REM sleep. The extent of arterial hypoxaemia and hypercapnia when patients were awake was directly related to the fall in Sao_2. In these patients with neuromuscular disease minimum Sao_2 during REM sleep was related to both VC % predicted and percentage fall in VC when they moved from the erect to the supine position, suggesting that these two relatively simple tests allow some prediction of nocturnal hypoxaemia.

Severe respiratory muscle weakness is clearly an important factor contributing to the development of respiratory failure in patients with neuromuscular problems. In general our results agree with the findings of Braun *et al*,[18] that patients with a VC below 55% predicted and values for maximum respiratory pressures below 30% predicted are likely to have daytime hypercapnia, though in both studies there were some exceptions. The patients studied by Braun *et al* were in general younger and had higher daytime Pao_2 values and stronger respiratory muscles than our patients, and a much smaller proportion of their patients had hypercapnia. In our study 11 of the 13 patients with a VC below 55% predicted and eight of the 11 with a Pimax below 30% predicted had hypercapnia ($Paco_2$ 6 kPa).

Other factors are, however, likely to contribute to the development of respiratory failure. In our study the extent of respiratory muscle weakness was not related to either arterial Pco_2 when patients were awake or the degree of sleep hypoxaemia (neither "typical" nor minimum Sao_2). Braun *et al* did not find a simple relation between respiratory muscle strength and daytime $Paco_2$.[18] In our study four patients maintained an Sao_2 of at least 85% during REM sleep despite having respiratory muscle weakness varying from minimal to severe. We suggest that the extent of sleep induced hypoventilation in patients with neuromuscular disease is a key determinant of daytime $Paco_2$ when they are awake.

The relation between minimum Sao_2 during REM sleep and daytime Pao_2 and $Paco_2$ probably reflects the shape of the oxygen dissociation curve. Subjects with higher $Paco_2$ values will have lower Pao_2 values, which will be closer to the steep part of the oxygen dissociation curve. Hypoventilation during sleep, and any further reduction in ventilation during REM sleep, will cause a proportionately greater fall in Sao_2 in these patients. In the patients with neuromuscular disease we believe that hypoxaemia during REM sleep may relate to a combination of upper airway narrowing, lung stiffness, and ventilation perfusion abnormality to varying extents, in addition to diaphragm weakness. The worsening of arterial hypoxaemia during REM sleep may depress the respiratory drive further and thus worsen the respiratory failure.

Despite the lack of any relation between minimum Sao_2 during REM sleep and maximum respiratory pressures, the percentage fall in VC from the erect to the supine position was correlated with the increase in hypoxaemia in REM sleep. This may in part reflect the distribution and pattern of respiratory muscle disease in these patients. PImax measures the strength of the combined respiratory muscles and does not distinguish between the contributions of the diaphragm and accessory muscles, including the intercostals. On the other hand, the percentage fall in VC from the erect to the supine position is a stronger indicator of diaphragm weakness.[12 13] The finding of a relation between the percentage fall in VC and minimum Sao_2 during REM sleep supports the hypothesis that the hypoventilation and substantial hypoxaemia during REM sleep is secondary to intercostal inhibition in the patients with severe diaphragm weakness. Presumably patients with intercostal muscle weakness, in whom diaphragm function is preserved to some extent, would be able to maintain Sao_2 during REM sleep if they did not develop upper airway obstruction or have coexisting severe lung disease. Two patients (Nos 3 and 7) probably had well preserved diaphragm function (fall in VC from erect to supine posture zero and 3%) and they were able to maintain adequate oxygenation during REM sleep. The relative distribution of respiratory muscle weakness between the intercostal muscles and the diaphragm could be crucial in determining the adequacy of sleep oxygenation. The electromyographic findings in one patient with poliomyelitis and documented diaphragm weakness support this hypothesis: inhibition of intercostal muscles during REM sleep was associated with hypoventilation and arterial oxygen desaturation. Diaphragm weakness is likely to have been a major determinant of the fall in Sao_2 during REM sleep when intercostal and accessory muscle activity was inhibited. This result is similar to the findings of Johnson and Remmers in patients with chronic obstructive pulmonary disease.[19] They found that loss of accessory muscle activity (scalene and sternocleidomastoid) during REM sleep was associated with reduced ribcage excursion and a fall in Sao_2%.

The extent of sleep hypoxaemia was strongly related to vital capacity (% predicted). A reduction in vital capacity may result from reduced respiratory muscle strength or the lung stiffness that can occur in patients with longstanding neuromuscular disorders, or both.[1 2] There was a weak but significant relation between vital capacity and both PImax and PEmax. The low values for total lung capacity and normal residual volume provide further supporting evidence for respiratory muscle weakness. Nocturnal gas exchange would obviously be further impaired in patients with underlying lung disease in addition to muscle weakness. Diaphragm weakness will also cause gas exchange abnormalities, and deterioration in the relation of ventilation to perfusion in the supine posture in particular.[5] An increased respiratory load—that is, increased airway resistance or increased elastic recoil—in the presence of weak respiratory muscles will clearly worsen hypoventilation during sleep. Increased upper airway resistance during REM sleep is a further factor that might contribute to hypoxaemia during REM sleep. Some patients in this study had bulbar palsy with pharyngeal muscle weakness.

The clinical management of patients with neuromuscular disorders would be simplified if a reliable daytime test were available that could accurately predict the occurrence of sleep induced respiratory failure. In a follow up study of 209 patients with poliomyelitis Howard *et al*[7] found that respiratory deterioration was related to the reduction in forced vital capacity; respiratory deterioration was associated most commonly with the development of nocturnal alveolar hypoventilation. They reported that regular mechanical respiratory assistance was rarely necessary for patients with a forced vital capacity above 1·8 litres. In patients with chronic airflow limitation Connaughton *et al*[20] reported the value of simple measurements (awake Sao_2, $Paco_2$, vital capacity) in determining survival. Our study in patients with neuromuscular disorders provides general support for these findings. There is, however, considerable scatter of the data relating daytime respiratory function to sleep hypoxaemia, making it difficult to provide confident clinical predictions for individual patients.

Serial measurements of VC and daytime blood gas tensions are recommended as the initial method of evaluation for patients with neuromuscular disorders and respiratory muscle weakness. There are limitations to the predictive value of these measurements, however, and a sleep study with measurement of Sao_2 and $Tcco_2$ is necessary to assess the extent of nocturnal respiratory failure in an individual patient. A sleep study is esssential for detecting upper airway obstruction during sleep and selecting an appropriate form of nocturnal ventilatory support.[21 22]

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