

Postural breathing pattern changes in patients with myotonic dystrophy

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Abstract

We recorded by pneumotachography the breathing in nine patients with myotonic dystrophy (MD), both seated and supine and with eyes open in both positions. Irregular breathing (coefficient of variation > 20% for V_T and TTOT) was observed in six of the patients, two of whom showed irregularity in both positions whilst the remaining four had irregular breathing only when supine. In addition, in this latter group, irregularities first appeared in V_T and only after a few minutes in TTOT. Whereas in the group exhibiting irregular breathing in both seated and supine positions, irregularities were observed throughout the recording. However, no significant difference in any ventilatory variable was observed as between the two postures. Rib cage (RC) and abdomen (AB) motions were recorded by uncalibrated respiratory inductance plethysmography. Although for MD patients the mean values of the RC/AB ratio lay within the normal range the relative decrease in value as between seated (0.78 ± 0.52) and supine (0.31 ± 0.13) position was less than in healthy subjects. These observations suggest that MD may cause deficiencies in several mechanisms. Analyses of the respiratory pattern in each patient may provide information leading to the identification of the impaired respiratory mechanisms. © 2000 Elsevier Science B.V. All rights reserved.

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1. Introduction

Although a similar resting ventilation has been observed in patients with myotonic dystrophy (MD) to that in control subjects, high respiratory frequency and low tidal volume have been reported in the former (Bégin et al., 1982; Serisier et

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al., 1982; Jammes et al., 1985; Bogaard et al., 1992; Ververs et al., 1996). Myotonia of the respiratory muscles (Bégin et al., 1982; Rimmer et al., 1993), and altered afferent output from diseased muscles (Bégin et al., 1980; Serisier et al., 1982; Jammes et al., 1985) may offer an explanation for both tachypnea and the lower tidal volume in patients with MD.

Another feature of the breathing pattern in patients with MD is the occurrence of irregular breathing (Gillam et al., 1964; Coccagna et al., 1975; Serisier et al., 1982). Such irregularities in breathing pattern have been described (Gillam et al., 1964) as consisting of periods of irregular respiration occurring at more or less regular intervals and often separated by periods of apnea in the case of patients in a semi-recumbent position. For patients when awake, a significantly greater variability in tidal volume and respiratory cycle time was found than in control subjects (Bogaard et al., 1992; Gibson et al., 1992; Veale et al., 1995; Ververs et al., 1996). In the view of several authors (Gillam et al., 1964; Bogaard et al., 1992; Ververs et al., 1996), this irregularity may be considered to result to a large extent, from influences during consciousness from the higher centers which are insufficiently corrected for by the chemical control mechanism. A 'behavioural' influence also seemed to be present in patients with MD, since irregularity in breathing pattern, present in the wakeful state and during light sleep decreased noticeably during slow-wave sleep (Gibson et al., 1992; Veale et al., 1995). It was suggested (Bogaard et al., 1992) that the uncoordinated action of the expiratory and inspiratory intercostal muscles may provide a partial explanation of breathing irregularity in patients with MD. Another contributory factor would then be a deficiency in respiratory control related to disordered afferent information from the diseased muscles, since abnormalities in the muscle spindles of patients with MD have been documented (Swash and Fox, 1975) and there is evidence of impaired afferent activity from these muscles (Stranock and Newsom Davis, 1978).

The impairment of respiratory muscle function may also be studied by examining breathing patterns in different postures. Indeed, the movements

associated with changes in posture, common in everyday life, call into play forces which alter the operating length of the respiratory muscles and induce changes in force distribution in the respiratory muscle and in the activity of various inspiratory muscles (Vellody et al., 1978).

The aim of the present study was to examine a possible deficiency in the mechanisms responsible for adjusting respiratory muscle activation to different body positions. Resting breathing in patients with MD was recorded by pneumotachography for both the seated and supine position, with eyes open in both postures. In addition, rib cage and abdominal motions in the supine and seated positions were recorded by inductance plethysmography, as patients with neuromuscular diseases have been reported to display in general abnormal thoracoabdominal patterns of breathing (Perez et al., 1996).

2. Methods

2.1. Patients

Nine patients (five male) with MD, seen at the hospital for a routine visit, participated in the study. According to muscular disability rating scale described by Bégin et al. (1997), these patients may be classified as III (moderate proximal weakness, ambulatory). Informed consent was obtained from all patients. The experimental protocol was examined and approved by the Institutional Ethics Review Board. Five patients had a passive pacemaker. Patient characteristics are summarized in Table 1. No patient exhibited severe respiratory deficiency but mild restrictive respiratory disorder was observed in three patients. Pulmonary function data, Pa_{O_2} , Pa_{CO_2} and respiratory CO_2 response (Infrared analyser, Gould) are shown in Table 2. Predicted values for VC, FEV_1 , and FEV_1/VC were those of the European Community (Quanjer et al., 1993) and for PI_{max} and PE_{max} those of Black and Hyatt (1969). A CO_2 rebreathing test was performed according to the method described by Read (1967).

2.2. Measurement and protocol

Airflow was measured with a pneumotachograph (Fleish head No. 1) and a differential pressure transducer (163PC01D36, Micro Switch) mounted on a face mask. An uncalibrated respiratory inductive plethysmograph (RIP) was used to obtain rib cage (RC) and abdomen (AB) motion signals. The sensitivity and calibration factors were set at the same values for both channels (Respirtrace, model 150, Studley Data).

Two series of 10-min recordings were performed corresponding to the seated and the supine position respectively. In each case patients were instructed to keep their eyes open during the entire recording period.

2.3. Data analysis

Data acquisition was performed with a recorder (TA11, Gould Electronic) on a PCMCIA card at a sampling rate of 200 Hz. A paper trace was also obtained. The PCMCIA card files were converted into compatible Macintosh microcomputer text files for further analysis. The flow signal was analyzed breath-by-breath in order to obtain tidal volume (V_T) by integration of the flow signal, breath duration (T_{TOT}), and inspiratory (T_I) and expiratory (T_E) durations, for each breath. Minute ventilation, V_T/T_I and T_I/T_{TOT} were calculated for each breath. Mean values of these variables were then calculated for each recording.

Uncalibrated inductance plethysmography signals were also processed breath-by-breath using the cycle delimitation of the flow signal. This processing consists in calculating the Fourier coefficients for each rib cage and abdomen cycle. For each set of recordings, these coefficients were averaged over all the breathing cycles and used for the construction of the mean RC and mean AB cycle. Mean RC was plotted versus mean AB for each subject in both positions. In addition, the RC/AB ratio was calculated for each breath and mean values were calculated for each set of recordings.

2.4. Statistical analyses

Values are expressed as mean \pm standard deviation. Student's t-test and paired t-test were used where appropriate to compare the two conditions: seated or supine. Statistical significance was set at $P = 0.05$.

3. Results

3.1. Comparison of breathing pattern between seated and supine positions

The investigation of breathing patterns in seated and supine positions in patients with myotonic dystrophy revealed the occurrence of expiratory pauses in six out of nine patients. In the

Table 1
Patients characteristics

	Height (cm)	Weight (kg)	Age (years)	Sex	Pacemaker
#1	164	73	38	F	No
#2	155	64	57	F	Yes
#3	182	85	36	M	Yes
#4	159	82	37	F	Yes
#5	162	82	56	F	Yes
#6	164	68	47	M	No
#7	173	92	55	M	No
#8	177	68	35	M	Yes
#9	167	77	65	M	No
Mean	167	77	48		
SD	9	10	12		

Table 2
Lung function tests and CO₂ sensitivity^a

	VC (%pred)		FEV ₁ (%pred)		FEV ₁ /VC (%pred)		P _I _{max} (%pred)		P _E _{max} (%pred)		Pa _O ₂ (kPa)	Pa _{CO} ₂ (kPa)	CO ₂ test
	Seated	Supine	Seated	Supine	Seated	Supine	Seated	Supine	Seated	Supine			
#1	85	83	93	90	109	108	86	70	86	76	12.05	6.35	
#2	86	87	72	74	84	85	62	53	42	71	11.40	4.90	1.00
#3	107	110	87	77	83	77	118	120	38	46			0.88
#4	71	64	65	56	91	87	59	46	39	33	10.50	5.40	0.80
#5	62	61	64	60	103	99	37	27	31	31	11.30	5.30	0.36
#6	83	88	70	74	84	84	73	73	44	40	8.03	6.18	1.81
#7	58	58	62	61	107	102	38	28	62	63	8.21	5.61	0.70
#8	85	81	67	62	79	77	44	33	32	27	13.10	5.52	1.40
#9	107	109	111	106	109	109	43	44	46	47			1.31

^a VC, vital capacity; FEV₁, forced expiratory volume in 1 sec; P_I_{max} and P_E_{max}, maximum static inspiratory and expiratory pressures; CO₂ test, slope of \dot{V}_1 versus PET_{CO₂} in seated position, l·min⁻¹ mmHg⁻¹. For VC, FEV₁, and FEV₁/VC predicted values were those of the European Community (Quanjer et al., 1993). For P_I_{max} and P_E_{max} predicted values were those of Black and Hyatt (1969). The CO₂ rebreathing test was performed according to the method described by Read (1967).

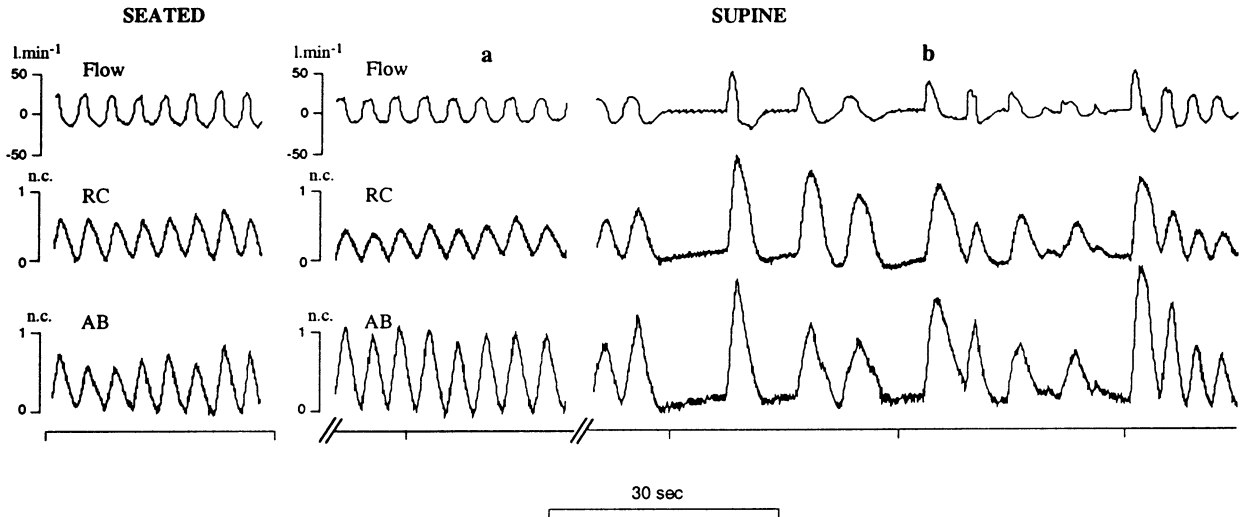


Fig. 1. An example of airflow, rib cage (RC) and abdomen (AB) motion signals recorded seated and supine (at the beginning (a) and towards the end (b)) in subject #9.

case of four patients expiratory pauses were only observed in the supine position and occurred in the later part of the recording a few minutes after the patients had lain down. An example of such recording is shown in Fig. 1. The two remaining patients exhibited expiratory pauses in both positions.

Expiratory pauses were associated with high values of V_T . Indeed, Table 3 shows greater variability in V_T , T_E and T_{TOT} . Irregular breathing was present only in the supine position in subjects # 1, 4, 6 and 9 but in both positions for subjects # 5 and 8. The remaining three patients showed less irregular breathing, i.e. the coefficients of variation of V_T and T_{TOT} were less than 20% in both supine and seated position. Series of breath duration (T_{TOT}) and tidal volume (V_T) are shown in Fig. 2, in both positions, for one representative patient of each group.

However, the pattern of irregular breathing in those patients with irregularities in both postures differed from that of those with irregularities only in the supine position. Indeed, in this latter group,

irregularities are first visible in V_T and only after a few minutes of being supine in T_{TOT} whereas in the former group (i.e. of irregular breathing both seated and supine), irregularities were observed throughout the recording.

The comparison of the respiratory variables T_{TOT} , T_I , T_E , V_T , \dot{V}_I , V_T/T_I and T_I/T_{TOT} within subjects in the seated and supine position, is illustrated in Fig. 3, where mean values and standard deviations are shown for each subject. Identity lines were drawn to better illustrate the changes as between postures. Results show \dot{V}_I , V_T and V_T/T_I to be lower in the supine position except in the case of subjects # 3 and 9 for \dot{V}_I , subject # 1 for V_T and # 9 for V_T/T_I . As regards breathing time variables, T_{TOT} , T_I and T_E , and the T_I/T_{TOT} ratio values, no consistent trend was found to the changes.

Results of comparisons (paired t-test) of the mean values of the respiratory variables for all nine patients are reported in Table 3. No significant difference in any variable as between positions was observed.

Table 3
Respiratory variables for each subject seated and supine^a

	T _{TOT} (sec)		T _I (sec)		T _E (sec)		V _T (l)		\dot{V}_I (l·min ⁻¹)		V _T /T _I (l·sec ⁻¹)		T _I /T _{TOT}	
	Seated	Supine	Seated	Supine	Seated	Supine	Seated	Supine	Seated	Supine	Seated	Supine	Seated	Supine
# 1	3.54	4.68	1.22	1.58	2.32	3.09	0.38	0.45	6.51	6.30	0.31	0.29	0.35	0.37
%	16.5	46.2	13.6	20.6	21.5	66.6	32.8	42.8	24.6	46.1	21.0	50.4	12.3	25.4
# 2	3.98	3.78	1.50	1.52	2.48	2.25	0.39	0.34	5.83	5.39	0.26	0.22	0.38	0.40
%	15.6	13.3	14.6	14.7	19.1	15.1	29.7	20.2	20.2	13.6	21.7	15.7	9.7	7.9
# 3	3.93	3.42	1.60	1.46	2.33	1.95	0.65	0.58	9.90	10.20	0.40	0.40	0.41	0.43
%	16.8	13.5	17.8	15.3	21.3	17.6	21.1	20.9	15.9	18.1	15.1	16.2	12.4	10.7
# 4	4.62	4.25	1.42	1.56	3.20	2.69	0.47	0.43	6.20	6.13	0.33	0.27	0.31	0.38
%	18.8	26.9	13.2	18.0	24.2	37.0	21.7	26.5	15.4	19.5	12.9	19.9	14.4	18.4
# 5	4.56	5.37	1.48	1.45	3.08	3.92	0.67	0.65	8.89	7.38	0.45	0.45	0.33	0.28
%	26.8	21.1	22.9	19.4	33.1	25.8	28.3	20.2	26.8	24.1	18.9	12.8	17.6	22.9
# 6	5.14	6.57	2.28	2.34	2.87	4.23	0.66	0.57	7.73	5.26	0.29	0.24	0.44	0.37
%	14.8	29.0	16.1	24.9	18.1	44.3	13.0	39.1	10.7	31.9	11.1	28.0	9.9	24.7
# 7	4.13	3.98	1.47	1.53	2.66	2.45	0.66	0.63	9.69	9.53	0.45	0.41	0.36	0.39
%	16.9	14.2	19.1	14.6	21.0	18.2	17.0	17.9	16.9	15.1	13.9	12.4	14.4	9.5
# 8	6.40	5.03	1.26	1.13	5.14	3.90	0.92	0.63	9.18	8.78	0.73	0.55	0.21	0.26
%	33.1	46.4	16.3	16.1	39.3	60.5	23.4	28.3	28.8	43.3	15.5	20.7	26.6	39.7
# 9	6.05	6.46	3.13	1.82	2.91	4.65	0.74	0.71	7.37	8.30	0.24	0.38	0.52	0.35
%	16.8	69.5	21.6	32.2	18.6	99.6	24.3	62.4	20.5	74.1	30.9	59.2	12.3	40.1
P	0.668		0.526		0.488		0.093		0.213		0.338		0.728	
Mean	4.70	4.84	1.71	1.60	3.00	3.24	0.61	0.55	7.92	7.48	0.39	0.36	0.37	0.36
%	19.6	31.1	17.2	19.5	24.0	42.7	23.5	30.9	20.0	31.7	17.9	26.1	14.4	22.1

^a Mean values and coefficients of variation (%) of the respiratory variables for each subject seated and supine. T_{TOT}: breath duration; T_I and T_E: inspiratory and expiratory time; V_T: tidal volume; \dot{V}_I : minute ventilation; P: paired t-test results of comparisons of the mean values of the respiratory variables for all nine patients; Mean values and mean coefficients of variation calculated for all patients in both positions are represented on the bottom lines.

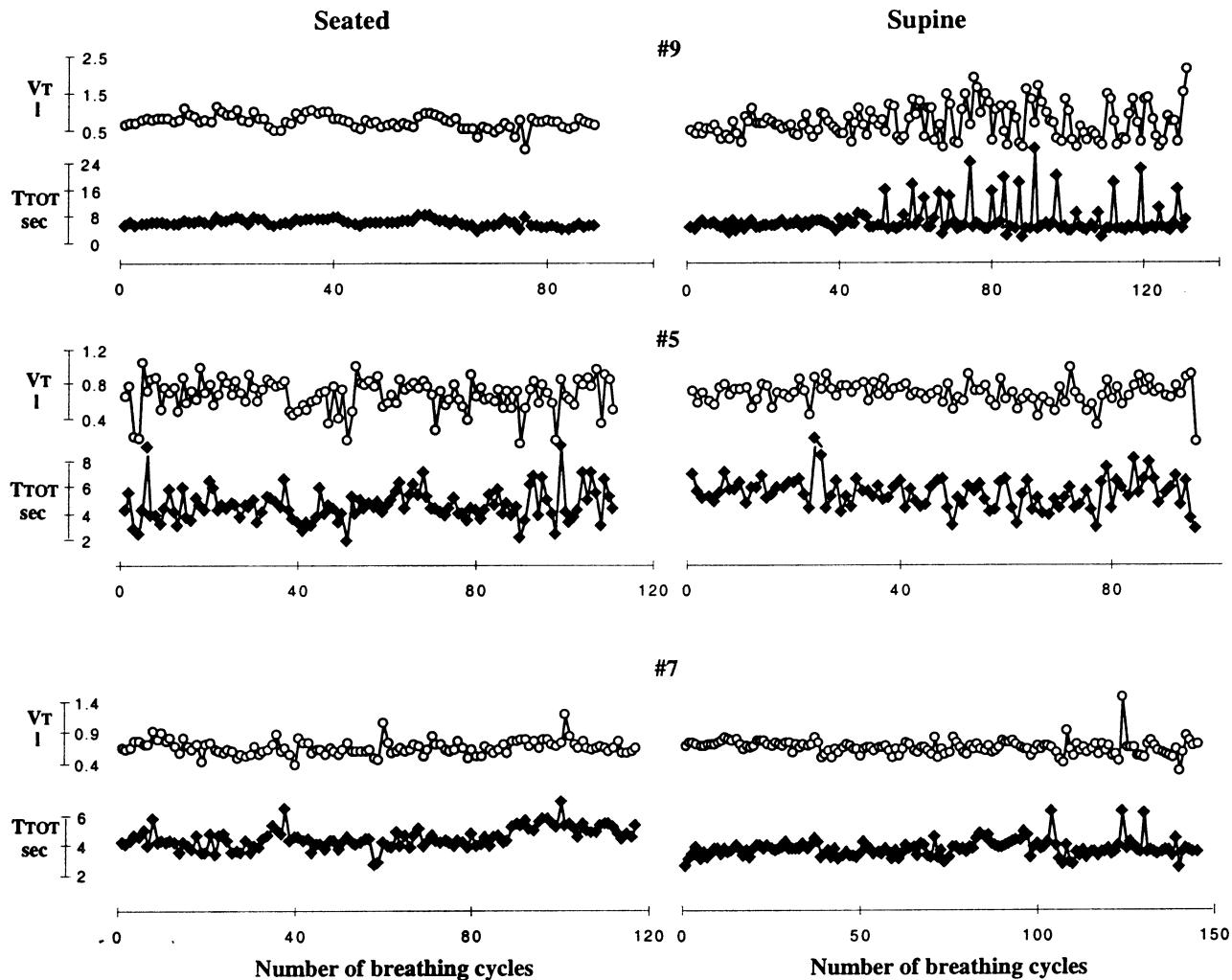
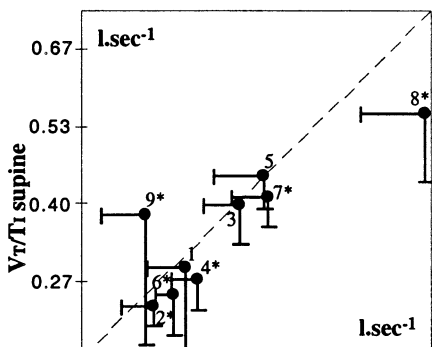
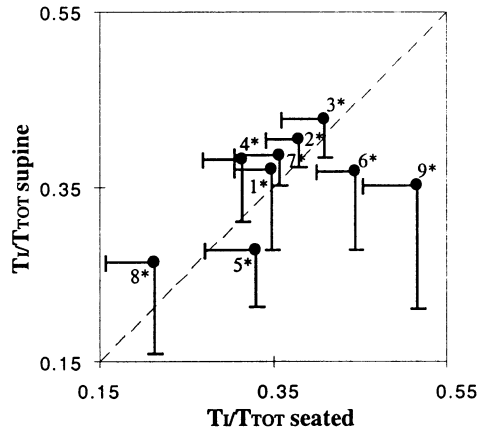
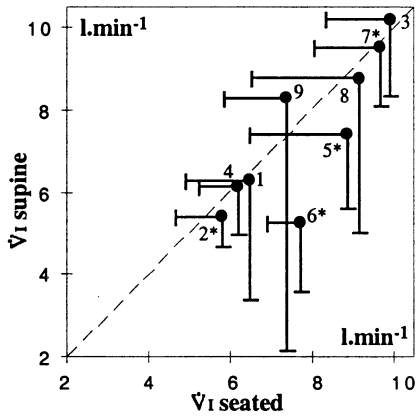
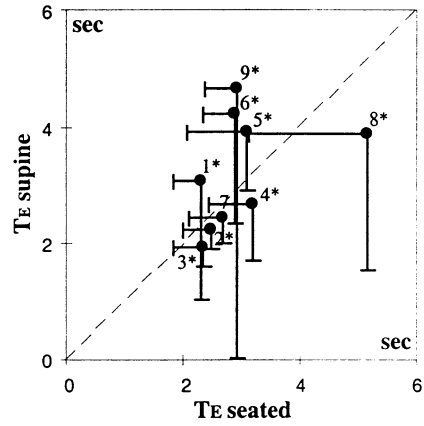
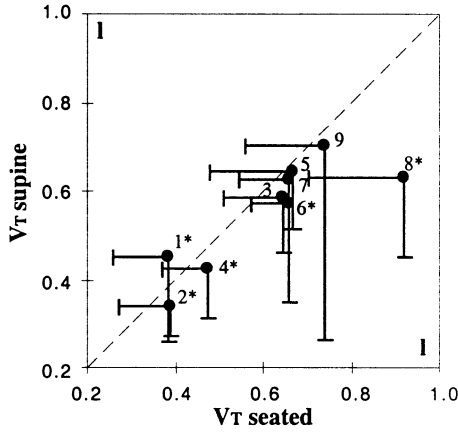
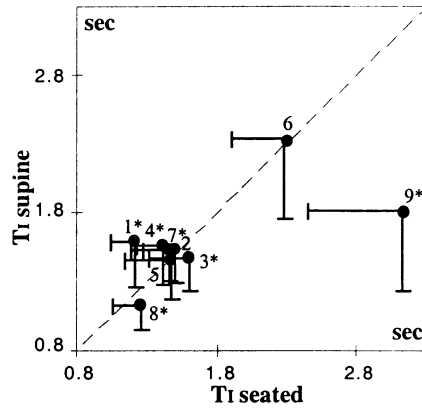
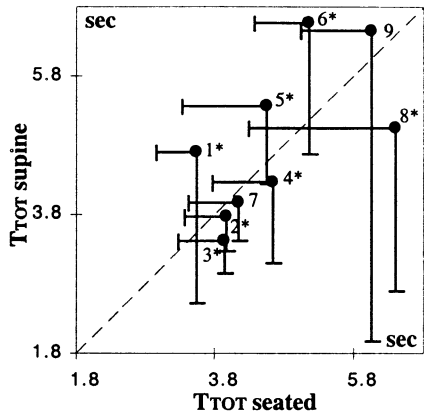


Fig. 2. Series of breath duration (TTOT) and tidal volume (VT) for three subjects. The top panel shows a subject in which irregular breathing appeared only in the supine position. The subject in the middle panel showed irregular breathing in both seated and supine positions while the subject in the bottom panel was considered to breathe regularly because their coefficient of variation was less than 20%.



3.2. Comparison of thoracoabdominal motion between seated and supine positions

Plots of mean RC versus mean AB for all subjects in both positions are represented in Fig. 4 with the same X and Y-axis scales. Fig. 4 shows that the RC versus AB plots are flattened ellipses, which indicate that there were no asynchrony between the rib cage and abdomen motions neither seated nor supine. For subject # 6, the RC versus AB plots crosses in both positions. This suggests that the RC and AB motion have different rate of changes.

The mean RC/AB ratio \pm standard deviation appears in the left-hand corner of each plot for the seated position and in the right-hand corner for the supine position. Significantly lower mean values of this ratio are found in the supine as compared to the seated position in all patients. The change in the RC/AB ratio as between seated and supine position seems to bear no relation to the breathing pattern variability. Indeed, as regards subjects # 2 and # 9 who exhibit the greatest change in the RC/AB ratio from seated to supine position, variability in T_{TOT} is similar in both positions for subject # 2 whilst exhibiting a marked increase from seated to supine in subject # 9.

4. Discussion

In this study on nine wakeful patients with MD, breathing pattern analysis revealed irregularities for four patients in the supine position only whereas for two other patients irregular breathing was exhibited in both positions. Despite the irregular pattern observed in the supine position in these four patients, comparison of the mean values of the respiratory variables as between the seated and supine position revealed no significant difference.

Comparing breathing pattern in control subjects and patients with MD in seated and supine posi-

tion, a significant decrease in supine as compared to seated position was found only in V_T/T_I for patients but also for control subjects (Bégin et al., 1982). Although not significantly different, both V_T and V_T/T_I appeared to be lower in supine as compared to seated posture for most of the patients (Fig. 3) in the present study.

Irregularities in breathing pattern are clearly apparent when breath-by-breath values are plotted for the whole of the recording, as illustrated in Fig. 2. Irregular respiration in patients with MD has already been reported both in individual case studies (Coccagna et al., 1975) and in group studies (Serisier et al., 1982; Bogaard et al., 1992; Gibson et al., 1992; Veale et al., 1995; Ververs et al., 1996) where a significantly more marked irregularity than in control subjects was observed. However, as in the present study, not all patients with MD manifest irregular breathing. In the earlier studies irregular breathing was reported in seven out of 19 myotonic patients with less marked irregularities in six others (Serisier et al., 1982), and in four out of seven patients when awake (Gibson et al., 1992; Veale et al., 1995). In the present study, it is noteworthy that the pattern of irregular breathing in those patients with irregularities in both postures was different from that of those with irregularities in the supine position only. Indeed, in the two patients with irregular breathing in both positions, irregularity was 'continuous' (Fig. 2), whereas in the four patients with irregularities only when supine, periods of irregular breathing appeared after a few minutes in the supine position and occurred at more or less regular intervals.

It has been postulated that breathing irregularity results, to a large extent, from influences from the higher centers that are insufficiently corrected for by the chemical control mechanism (Ververs et al., 1996). Indeed, although statistically not significant, there was a tendency for patients with a normal ventilatory CO_2 response to breathe more regularly than patients with a lowered ventilatory CO_2 response (Bogaard et al., 1992; Ververs et al., 1996).

Fig. 3. Comparison of respiratory pattern between the two postures in all subjects. Seated versus supine plots of mean \pm S.D. values of respiratory variables: Breath duration (T_{TOT}), inspiratory time (T_I), expiratory time (T_E), tidal volume (V_T), minute ventilation \dot{V}_I , T_I/T_{TOT} and V_T/T_I .

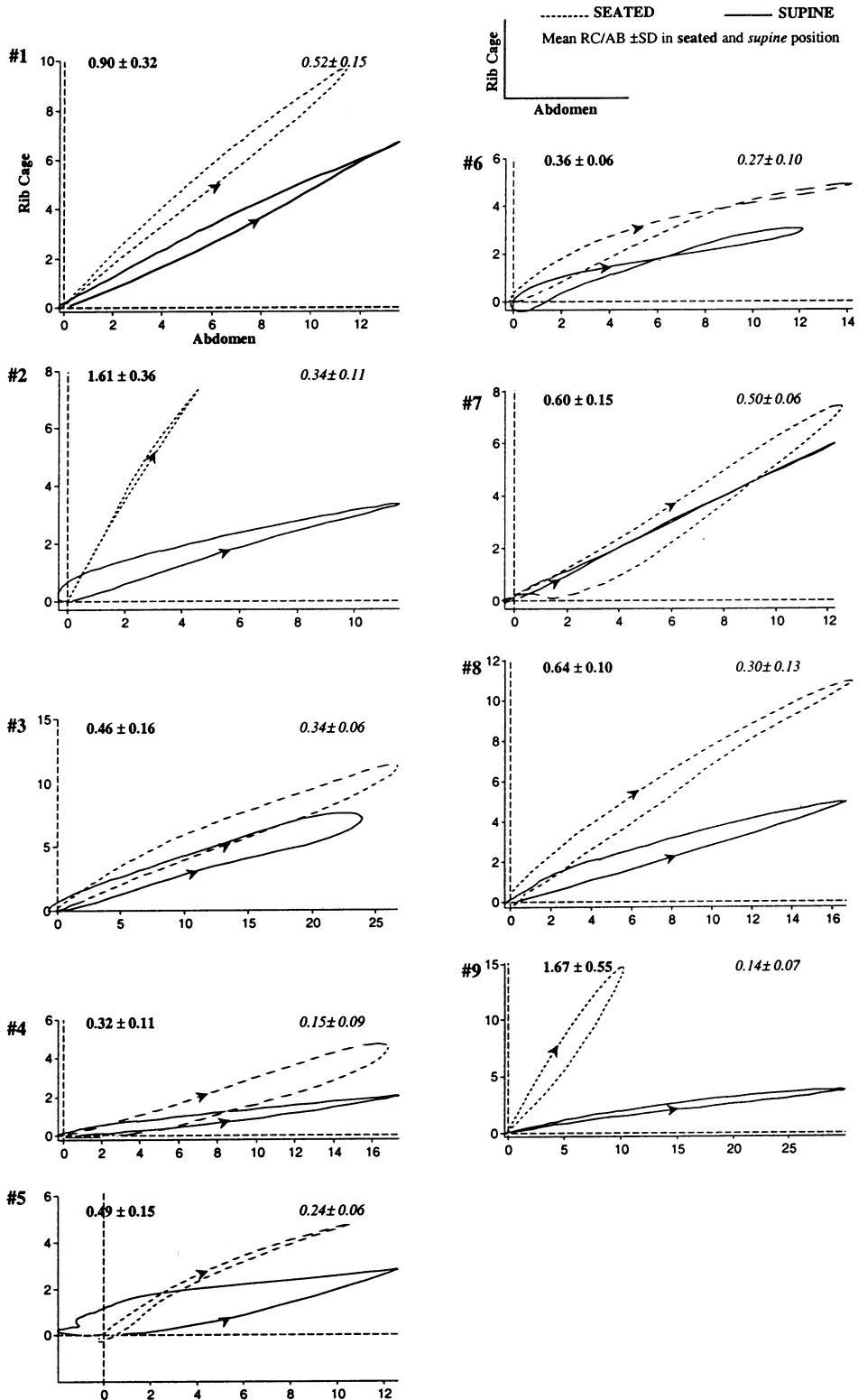


Fig. 4. Mean rib cage (RC) versus mean abdomen (AB) Fourier coefficients, calculated breath-by-breath, for the nine subjects in seated and supine positions. Mean values of the RC/AB ratio \pm S.D. for both positions are reported on each plot.

This hypothesis is also supported by Gillam et al. (1964), who showed breathing irregularities in patients with MD to increase in cases of weakening of CO₂ sensitivity by an anesthetic agent (thiopentone). Although statistically not significant, an inverse relationship was found to exist between the ventilatory CO₂ sensitivity and the irregularity of respiratory cycle time and tidal volume (Bogaard et al., 1992). Such mechanisms may explain irregularities such as those observed in patient # 5, who exhibited irregular breathing in both positions associated with low CO₂ sensitivity. However, patient # 8, with irregular breathing in both positions had, a normal ventilatory CO₂ sensitivity response. Furthermore, we did not observe any correspondence between the value of Pa_{CO₂} and the irregularity of the breathing pattern.

The irregularities associated with the supine position may involve other mechanisms. Irregular breathing was found in patients sitting or lying undisturbed and it was not limited to periods of drowsiness (Serisier et al., 1982). Although drowsiness has often been reported in patients with myotonia (Kilburn et al., 1959; Phemister and Small, 1961; Coccagna et al., 1975), there is no possible question that our patients did not remain alert as they were instructed to keep their eyes open during the whole of the recordings. Thus, irregular breathing would not appear to be linked to drowsiness. Furthermore, the variability in breathing interval in patients with MD is only significantly higher during wakefulness and light sleep, than that in patients with nonmyotonic muscle disease and in normal subjects, all three groups showing similar variability during slow-wave sleep (Gibson et al., 1992; Veale et al., 1995).

Changes in posture involve the exertion of forces which alter the operating length of the respiratory muscles. In normal subjects there is adjustment of respiratory muscle activation such that any difference in respiratory characteristics between seated and supine position is remedied by the second breath following posture change. This ventilatory compensation during postural change is not obtained via vagal afferent information but rather through respiratory muscle receptors (Kin-

near et al., 1989). In this study irregular T_{TOT} when supine occurs only after a few minutes whereas V_T exhibits higher variability from the outset. One possible explanation could be that the mechanism adjusting ventilation when changing from seated to supine position does not operate as rapidly as in healthy subjects, leading to 'fluctuations' in V_T. The accumulation of deficiencies in this regulating mechanism may then be responsible of the occurrence of irregular breaths with expiratory pauses. On this hypothesis, impairment of the respiratory muscle receptors is assumed to trigger irregular breathing. Indeed, abnormalities in the muscle spindles of patients with MD are well recognised (Swash and Fox, 1975) and evidence of impaired afferent activity has been demonstrated (Stranock and Newsom Davis, 1978). However, the weakness of the respiratory muscle can not be ruled out as a factor in irregular breathing. Weakness of respiratory muscles in patients with MD may be partly responsible for limiting ventilatory performance (Bégin et al., 1982) and for the pathogenesis of hypoventilation (Kilburn et al., 1959; Bégin et al., 1997). A large decrease in maximal mouth pressure in patients with MD as compared to that in normal subjects has been reported in several studies (Gillam et al., 1964; Bégin et al., 1982; Serisier et al., 1982; Bogaard et al., 1992). In addition, these studies reported a more marked impairment of P_{E_{max}} than of P_{I_{max}}, consistent with the data in this study. This reduction is probably the result of weakness of the abdominal muscles which are the major expiratory muscles (Serisier et al., 1982). Myotonia of the respiratory muscles could result in a decrease in compliance of the chest wall, which may in turn increase the work of breathing and threaten muscle fatigue resulting in a change of tidal volume and respiratory frequency in order to prevent this fatigue (Rimmer et al., 1993). Respiratory muscle weakness in patients with MD may induce abnormal motions of rib cage and abdomen, similar to those observed in patients with neuromuscular diseases (Perez et al., 1996), these abnormalities being aggravated by the change from seated to supine position. Unlike Perez et al. (1996) we did not observe (Fig. 4) asynchrony between RC and AB motions. Indeed,

the RC versus AB plots in both positions are elliptic loops similar to those described by Verschakelen and Demedts (1995) in healthy subjects during quiet breathing.

The partition of VT into its thoracic and abdominal volume displacement did not differ as between patients with MD and control subjects, either in the seated or supine position (Bégin et al., 1982). A decrease of 74.4% in the RC/AB ratio has been reported (Vellody et al., 1978) from seated to supine position, mean values being 0.90 seated and 0.23 supine. In another study (Sharp et al., 1975) an 82.7% decrease in the RC/AB ratio from seated to supine was found with however higher values of the RC/AB ratio in both positions (2.08 in seated and 0.36 in supine). Our results indicated a fall from 0.78 ± 0.52 in the seated position to 0.31 ± 0.13 in the supine, i.e. a decrease of 60.3% when changing from seated to supine position. Although the values of the RC/AB ratio were in the range of those already reported (Sharp et al., 1975; Vellody et al., 1978), the percentage decrease appeared to be lower in patients with MD than in healthy subjects. However, it should be noticed (Fig. 4) that there is a great inter-individual difference in the values of the RC/AB ratio in the supine and seated position as well as in the percentage change in value between the positions (top left of plots). This would suggest that rather than using a single value of RC/AB, changes in this ratio monitored at regular time intervals would provide the basis of more meaningful data concerning respiratory muscle impairment.

In conclusion, this study on the breathing pattern of patients with MD corroborates previous observations that deficiencies may exist in several of the respiratory system mechanisms of these patients. Analyses of resting breathing pattern recorded by pneumotachography and inductance plethysmography may provide supplementary information to that obtained from lung function tests on the impaired ventilatory mechanism of each patient. Furthermore, this study suggests that it may be meaningful to compare seated and supine breathing pattern prior to sleep studies in these patients.

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