ABSTRACT: Objective: The interactions between sleep, neck muscle activity, and cervical spinal pain were examined in a controlled study with nine patients suffering from idiopathic cervical dystonia (ICD; also referred to as spasmodic torticollis), and nine gender- and age-matched controls. Methods: From each participant, two all-night polysomnograms with additional electromyographic recordings from the sternocleidomastoid and upper trapezius muscles were obtained. The first night was for habituation to the laboratory environment; the second night for experimental data collection. Visual analogue scales were used to collect intensity and unpleasantness ratings of cervical spinal pain before and after the second sleep recording. Results: None of the standard sleep variables showed statistically significant differences between average values of both groups of participants. However, a significantly larger variance in sleep latency was obtained for the ICD patients. In general, abnormal cervical muscle activity decreased immediately when lying down without the intention to go to sleep. Subsequently, abnormal muscle contractions were gradually abolished in all ICD patients during the transition from relaxed wakefulness to light NREM sleep. Following this transition phase, no more abnormal EMG activity was found in any of our patients. Finally, cervical spinal pain intensity and unpleasantness were reduced by about 50% overnight. Conclusions: Both supine position and sleep can be associated with an improvement of symptoms of ICD, and this disorder does not induce any sleep perturbations.

RÉSUMÉ: Interrelation entre sommeil, activité musculaire et douleur dans le cas de dystonie cervicale. Objectif: Le but de cette étude était de mettre en evidence les liens possibles entre le sommeil, l’activité musculaire (trapeze et sterno-cléido-mastoïdien) et la douleur cervicale chez neuf patients présentant une dystonie cervicale idiopathique (torticollis spasmodique) par rapport à neuf sujets témoins, appariés pour l’âge et le sexe. Méthode: L’activité polysomnographique et musculaire ainsi que la douleur ont été étudiées. Seules les données de la deuxième nuit sont présentées, la première nuit servant d’adaptation aux conditions expérimentales. Résultats: Aucune des variables de sommeil n’a permis de différencier les deux groupes. Toutefois, la variance de la variable “latence du sommeil” était significativement plus importante chez les patients ayant une dystonie cervicale idiopathique. Ces mêmes patients ont démontré une chute de l’activité électromyographique cervicale anormale, ce en position couchée sans intention de dormir. Cette activité diminuait graduellement au début de l’endormissement et aucune activité anormale ne subsistait lorsque la première période de sommeil “lent léger” a été atteinte. Enfin, il est à noter qu’une diminution d’environ 50% de la douleur a été rapportée, sur une échelle visuelle analogue, en relation avec la baisse d’activité musculaire en cours de sommeil. Conclusions: La position couchée et le sommeil permet un allègement des symptômes associés à la dystonie cervicale idiopathique et le sommeil de ces patients n’est pas perturbé.

At present, there is no cure for ICD. However, several management strategies can be used to relieve its clinical symptoms (e.g., dystonia, pain). Treatment modes include physical therapy, pharmacotherapy, and surgical intervention. Descriptive studies indicate that improvement of symptoms of ICD may also be achieved simply by lying down, whereas symptoms may even disappear while asleep. On awakening, many patients notice a short-lasting absence of their symptoms Indeed (“honeymoon period”), especially early in the course of the disorder. However, in spite of this sleep-related amelioration, preliminary EMG data indicate a persistence or occasional recurrence of abnormal muscle activity during sleep throughout the night. Moreover, clinical observations show that in about 10% of cases, an activation of involuntary muscle contractions occurs during sleep, which may cause a disturbance of sleep organization and, consequently, a worsening of symptoms. These latter observations hamper an unequivocal explanation of the reported capability of lying down and sleeping to improve clinical symptoms of ICD.

The present study aimed to examine the relationship between sleep, neck muscle activity, and cervical spinal pain in patients suffering from ICD, so that more insight could be gained into the pathophysiological processes underlying this three-way interaction. To our knowledge, no complete studies on sleep and ICD have been published so far. Therefore, the present study is the first controlled polysomnographic study on ICD.

**METHODS**

**Patients and Controls**

Nine ICD patients and nine controls were included in the present study. With the help of the “Fondation Médicale en Dystonie du Québec”, patients were selected from those being referred to the Département de Neurologie of the Hôpital du Sacré-Cœur de Montréal. Controls were recruited through announcements published in local journals. All patients and controls gave informed consent to procedures approved by the Human Subjects Ethics Committee. They received a financial compensation for the inconvenience related to their participation.

In the group of patients suffering from ICD, there were five men and four women. Their ages ranged from 28 to 53 yr (mean = 42.0 yr; S.D. = 4.5 yr), whereas the duration of the disorder ranged from 1 to 12 yr (mean = 4.8 yr; S.D. = 3.4 yr). The diagnosis was made by the consultant neurologist (G.R.) on the basis of the outcome of self-report questionnaires, oral history taking, and an extensive clinical, laboratory, and radiological examination.

For each patient, the deviation of the head was described according to Fahn. The direction of head turning (i.e., the direction to which the chin points) was to the right side in four patients and to the left side in five of them. In all patients, the deviation was constant (i.e., the head was deviated for more than 75% of the time while sitting upright with the headunsupported). The severity of head deviation varied from moderate (i.e., the head was rotated one-third to two-thirds of the maximum possible movement about the coronal, longitudinal, or sagittal axis) to severe (i.e., the head was rotated more than two-thirds of the maximum possible movement). In six patients, a rotational torticollis was present due to a rotation about the longitudinal axis. In three patients, the head was primarily moved about the sagittal axis, resulting in a laterocollis. In two patients with laterocollis, a mild flexion of the head was part of the resultant head deviation. A mild extension was involved in the resultant position of the head in three patients with rotational torticollis.

All of the ICD patients reported that they suffered cervical spinal pain regularly, whereas none reported other neurological or sleep-related disorder. The mode of onset of the disorder was sudden in one of the patients and progressive in eight of them. Six patients had a history of minor trauma to the head, neck, or shoulder. Two patients held work-related events responsible for the onset of their disorder. One patient reported emotional shock antecedent to ICD. In eight patients, the family history was negative and in the ninth unknown because of adoption. Six of the nine patients were receiving pharmacologic therapy (e.g., benzodiazepines, muscle relaxants), which was discontinued at least one week before data collection. Three of the nine patients had previously received injections with botulinum A toxin, with minimal subjective or objective improvement.

The ICD patients were compared to five male and four female gender- and age-matched controls, ages 36-49 yr (mean = 42.1 yr; S.D. = 4.5 yr), without a history of neurological disorders. None of the controls reported cervical spinal pain. None was taking medication at the time of the study.

**Procedure and Data Analysis**

From each participant, polysomnographic recordings were obtained in our sleep laboratory for two consecutive nights. Recording took place in a dark, sound-attenuated, and temperature-controlled room. Sleep recording started at 10:30 pm and terminated upon participant’s spontaneous awakening or at 08:00 am. The first night was for habituation to sleeping in a laboratory environment and to rule out other sleep-related disorders. The second night was for experimental data collection.

Sleep was recorded on polygraphic paper, using a standard method. In addition, EMG was recorded bilaterally from the sternocleidomastoid and upper trapezius muscles with bipolar surface disc electrodes. Videotape recordings were taken throughout the night with an infrared camera focused on the head-neck-shoulder area. Sleep staging was performed by an experienced technologist blind to diagnostic status in 20-s epochs, and standard sleep variables were calculated (see Table).

For the ICD patients, the right and left sternocleidomastoid and trapezius EMG activity levels were scored by another experienced technologist on a four-point rating scale in 20-s epochs. The scorer was not aware of the sleep stage during which a certain muscle activity occurred. Muscle activity comparable to that in controls was scored as “normal” cervical EMG activity level. All other muscle activities were considered abnormal (i.e., ICD-related) and were scored as either “low”, “medium”, or “high”. An example of each score was available to the rater throughout. The intra-observer reliability of this scoring method, estimated as the Kappa coefficient, was found to be 0.83, indicating an almost perfect agreement between occasions.

Prior to the start of the sleep recording, sternocleidomastoid and trapezius EMG activity of the ICD patients was recorded in two positions: (I) while sitting upright without headsupport and (II) while lying down without the intention to go to sleep. In both positions, the eyes were kept open, facing in the same direction as the head. These EMG recordings were graded ordinally on the above-described four-point rating scale as well.
The ICD patients were asked to rate the sensory (“intensity”) and affective (“unpleasantness”) dimensions\(^{19}\) of their current cervical spinal pain before and after the second sleep recording. Pain intensity was rated on a 100-mm visual analogue scale (VAS; anchor words: “pas intense” [not intense at all] on the left side and “le plus intense que l’on puisse imaginer” [worst imaginable intensity] on the right side). Pain unpleasantness was rated on a 100-mm VAS as well (anchor words: “pas désagréable” [not unpleasant at all] on the left side and “le plus désagréable que l’on puisse imaginer” [worst imaginable unpleasantness] on the right side). Only VAS ratings of more than five mm were taken into account, because smaller values have been reported to be within the error of this measuring scale.\(^{20}\)

In addition, the ICD patients indicated the occurrence and intensity of their current cervical spinal pain before and after the second sleep recording on a four-point category scale (0 = “pas du tout” [not at all]; 1 = “un peu” [a little]; 2 = “modérément” [moderately]; 3 = “beaucoup” [a lot]).

To determine the diurnal variation in pain intensity in more detail, a diagram (modified from Fields)\(^{21}\) was completed with time (in hours; from midnight to midnight) on the abscissa and a 100-mm VAS as the ordinate. In that way, the ICD patients could give a graphic representation of the typical daily changes in the intensity of their cervical spinal pain.

**Statistics**

Statistical analyses were performed with significance set at the 0.05 probability level. For all sleep variables under examination, the Shapiro-Wilk test was used to test the null hypothesis of the input data values being a random sample from a normal distribution. If normality occurred, two-independent-samples \(t\) tests were applied for the analysis of differences in sleep variables between controls and ICD patients. Equality-of-variance was assessed by calculating the \(F\) ratio. If two population variances were equal, the probability of the pooled-variance test statistic (\(T_p\)) was examined. The separate-variance test statistic (\(T_s\)) was analyzed if the population variances were not equal. If the distribution of input data values was not normal, Wilcoxon rank-sum tests for two independent samples were used. The ordinal scores of cervical EMG activity and pain intensity were analyzed by means of Wilcoxon’s signed rank tests for related samples. In case of multiple comparisons, Bonferroni-adjusted probability levels were employed. Finally, univariate and multivariate ANOVA with repeated measures were used to test the null hypothesis of no statistically significant differences between the VAS variables obtained before and after the second sleep recording.

**Results**

For both the controls and the ICD patients, the standard sleep variables are given in the Table. None of the sleep variables showed statistically significant differences between average values. However, a significantly larger variance in sleep latency was found for the ICD patients (\(F_{(0.8)} = 13.893; p < 0.01\)).

Figure 1 illustrates the percentage distribution of the cervical muscles of the ICD patients by ordinal EMG activity score. The values are collapsed across the four cervical muscles under examination. The two bars on the left side represent the distributions of the recordings obtained prior to the start of the sleep recording while sitting upright without head support and while lying down. Following the postural change, a shift occurred towards the lower activity scores, indicating an immediate decrease in abnormal EMG activity. The cervical muscle distribution while sitting upright differed significantly from that while lying down (Z = -2.976; \(p < 0.005\)).

In addition, Figure 1 depicts the distributions of four 20-s epochs that represent the wake-to-sleep transition, viz., those of: (I) the epoch following the start of the sleep recording (“Lights off”; Figure 2A); (II) the last epoch staged as awake (“Last awake”); (III) the first epoch staged as stage 1; and (IV) the first epoch staged as stage 2 (Figure 2B). As can be gathered from Figure 1, a decrease in abnormal cervical muscle activity occurred from “Lights off” to “First stage 2”. This decrease was associated with a compensatory increase in the percentage of cervical muscles displaying normal activity levels. With significance set at the 0.008 probability level (Bonferroni-adjusted), the cervical muscle distribution at “Lights off” differed significantly from that at the other three 20-s epochs (Z = -2.887, -2.972, and -3.272 for comparisons between “Lights off” on the one side and “Last awake”, “First stage 1”, and “First stage 2” on the other side, respectively; \(p < 0.005\)).

At the level of the individual ICD patients, cervical EMG activity did not exceed the normal activity level during the wake-to-sleep transition in two out of nine patients. In six of them, abnormal cervical EMG was completely reduced to normal during “First stage 2”. In one ICD patient, a low contraction level persisted in the left sternocleidomastoid muscle up to and including “First stage 2”. However, a normal activity level was already reached in the subsequent epoch. At this point, it should be noted that from the epoch following “First stage 2”, normal activity levels were maintained throughout the night in all ICD patients. Increases in cervical EMG activity were observed during movement time only, equally in ICD patients and controls.

**Figure 1:** Grading of EMG activity recorded from neck muscles of ICD patients. Activity levels from four muscles were scored as described under Methods. Muscle activity comparable to that in controls was scored as “normal”. The bars represent the percentage distribution of gradings from 36 muscles. Distributions are shown for sitting upright without head support, for lying down without the intention to go to sleep, and for the transition from relaxed wakefulness (“Lights off”) to light NREM sleep (“First stage 2”).
Figure 2: Examples of polysomnographically recorded 20-s epochs, representing the wake-to-sleep transition from “Lights off” (A) to “First stage 2” (B). The following channels are displayed (from top to bottom): (I) EMG from the right (R) and left (L) sternocleidomastoid muscles; (II) EMG from the sub-mental (anterior digastric) muscles; (III) EEG from the C3-A2 and O2-A1 leads; (IV) electro-oculogram (EOG) from the right and left outer canthus; and (V) EMG from the right and left upper trapezius muscles. Note the decrease in abnormal cervical muscle activity from relaxed wakefulness (A) to light NREM sleep (B) to an almost normal activity level.
Figure 3: A typical example of the reported cervical spinal pain intensity between 10:00 pm and 06:00 am. The intensity that has been indicated typically daily changes, and the patient indicated his habitual sleep period within this period reflects this patient's extrapolation and is therefore inconclusive.

For both dimensions of current cervical spinal pain, i.e., intensity and unpleasantness, VAS ratings of the ICD patients were significantly lower in the morning than in the evening ($F_{(88)} = 13.996; p < 0.01$). The average intensity rating in the morning and increases more or less gradually during the day, with a percentage of 54.2 was found (32.6 mm ±27.1 mm in the morning and 60.1 mm ± 17.3 mm in the evening). No significant interaction was found between dimension and moment. Similar findings were obtained with the category scales for pain intensity before and after the second sleep recording: the average rating went down from 2.0 (S.D. = 0.7) in the evening to 1.0 (S.D. = 1.1) in the morning ($Z = -2.271; p < 0.05$).

The typical diurnal variation in pain intensity showed a similar pattern in all ICD patients. As illustrated in the typical example given in Figure 3, reported pain intensity is low in the morning and increases more or less gradually during the day, until a plateau is reached in the evening.

### Table: Sleep variables derived from the second sleep recording of nine controls and nine ICD patients.

<table>
<thead>
<tr>
<th>Sleep variables</th>
<th>Controls</th>
<th>ICD patients</th>
<th>Test statistic</th>
<th>P²</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total sleep time (min)</td>
<td>443.4 ± 40.3</td>
<td>425.0 ± 36.7</td>
<td>$T_p = 1.016$</td>
<td>0.325</td>
</tr>
<tr>
<td>%TST stage 1</td>
<td>11.1 ± 3.9</td>
<td>12.9 ± 7.6</td>
<td>$T_p = -0.610$</td>
<td>0.550</td>
</tr>
<tr>
<td>%TST stage 2</td>
<td>66.1 ± 6.4</td>
<td>61.9 ± 8.6</td>
<td>$T_p = 1.164$</td>
<td>0.262</td>
</tr>
<tr>
<td>%TST stage 3</td>
<td>0.032 ±14.8</td>
<td>0.012 ±19.5</td>
<td>$Z = -0.089$</td>
<td>0.930</td>
</tr>
<tr>
<td>%TST stage 4</td>
<td>0.008 ±0.42</td>
<td>0.000 ±0.96</td>
<td>$Z = 0.201$</td>
<td>0.841</td>
</tr>
<tr>
<td>%TST stage REM</td>
<td>17.6 ± 6.1</td>
<td>17.7 ± 5.4</td>
<td>$T_p = -0.025$</td>
<td>0.981</td>
</tr>
<tr>
<td>Wake time after sleep onset (min)</td>
<td>7.740.31/20.3</td>
<td>10.328.01/58.0</td>
<td>$Z = -0.707$</td>
<td>0.480</td>
</tr>
<tr>
<td>Sleep efficiency (%)</td>
<td>75.495 ±3085.5</td>
<td>69.793 ±3097.7</td>
<td>$Z = 0.884$</td>
<td>0.377</td>
</tr>
<tr>
<td>Sleep latency (min)</td>
<td>8.4 ± 3.3</td>
<td>8.7 ± 12.3</td>
<td>$T_s = 1.467$</td>
<td>0.162</td>
</tr>
<tr>
<td>REM latency (min)</td>
<td>51.3178 ±215.7</td>
<td>54.3148 ±231.7</td>
<td>$Z = 0.971$</td>
<td>0.331</td>
</tr>
<tr>
<td>Number of awakenings</td>
<td>29.2 ± 15.0</td>
<td>24.2 ± 12.2</td>
<td>$T_p = 0.776$</td>
<td>0.449</td>
</tr>
<tr>
<td>Number of sleep stage shifts</td>
<td>191.1 ±70.2</td>
<td>169.6 ±55.0</td>
<td>$T_p = 0.725$</td>
<td>0.479</td>
</tr>
</tbody>
</table>

¹For normally distributed variables, mean and standard deviation are given; for non-normal distributions, the 5th, 50th (=median), and 95th percentiles are given.

²$T_p$ = test statistic for two-independent-samples $t$ test with pooled variances; $T_s$ = test statistic for two-independent-samples $t$ test with separate variances; $Z$ = test statistic for Wilcoxon rank-sum test.

³Bonferroni-adjusted probability level = 0.004.

### Discussion

In accordance with the findings of Odergren et al., who found similar subjective sleep disturbance scores for ICD patients and controls, the results of the polysomnographic recordings obtained in the present study show that in general, ICD patients have a normal sleep organization (see Table). A larger variance in sleep latency was the only abnormality that could be associated with ICD. This larger variance may be related to the report by Rondot et al. that some ICD patients are more troubled by their disorder when first in bed than others, which might be even more pronounced in the sleep laboratory environment (e.g., different mattress and pillow). Although we did not measure pain intensity and unpleasantness in direct relation to the ongoing level of EMG activity, one can assume that a decrease in abnormal cervical muscle activity at the onset of the sleep recording (this study) facilitates the transition from wake to sleep. Indeed, two of our patients in whom the cervical EMG activity did not exceed the normal activity level from “Lights off” showed sleep latencies smaller than the 25th percentile. At the other extreme, the sleep latency value of one of our patients in whom a low contraction level persisted in one muscle up to and including the first epoch staged as stage 2 exceeded the 75th percentile. Accordingly, in patients with dystonia muscularum deformans (a generalized torsion dystonia), Jankel et al. found that severely affected patients exhibited a greater increase in latency to sleep than mildly affected patients.

In contrast with our findings, Rondot et al. found abnormalities in the sleep variables of their ICD patients, including decreased sleep efficiency, lower percentage of total sleep time spent in stage REM, and prolonged REM latency. However, interpretation of the study of Rondot et al. is hampered because the results are given in a descriptive fashion, statistics were not obtained, and information about controls is lacking.

Abnormal cervical muscle activity decreased immediately when lying down without the intention to go to sleep (Figure 1). This is in line with previous observations indicating that many ICD patients gain relief with a supine posture. A further decrease in abnormal EMG activity occurred during the transition from relaxed wakefulness to light NREM sleep (Figure 1;...
Figure 2, A and B). From the 20-s epoch following “First stage 2”, no more abnormal EMG activity was observed in any of our patients. This is in contrast with the findings of Rondot et al., who found no reduction in cervical muscle activity during sleep compared to the waking state. However, as mentioned above, the study of Rondot et al. was not performed in a fully controlled fashion, which makes it hard to compare the present results with theirs. Preliminary data of Forgach et al., showed the presence of brief bursts of cervical muscle activity during REM sleep in half of the ICD patients. As for the results of Rondot et al., this finding could not be confirmed in the present study either. A possible explanation for the discrepancy between these previous observations and our results might be the broad spectrum among ICD patients of presenting symptoms, examination findings, and history findings such as duration of the disorder, age at onset, mode of onset, antecedent risk factors, family incidence, and pharmacological history. Since neither Rondot et al., nor Forgach et al. give any information regarding the profile of their patients, it is hard to tell if similar subgroups of ICD patients have been studied.

Although ICD patients complain more about the abnormal posture of their head than about the pain they experience, in approximately two-thirds of cases the intensity of the pain necessitates some kind of management. Oral medications or localized infiltrations of anesthetics and steroids in tender muscles may be used to diminish pain. Repeated injections with botulinum A toxin may relieve pain due to muscle spasm. Besides these therapeutic interventions, it is a clinically known fact that in ICD patients, pain intensity may be reduced after sleep. This was confirmed in the present study, in which both intensity and unpleasantness of cervical spinal pain were reduced by about 50% overnight in the absence of any other type of intervention aimed to relieve the pain.

In summary, this study shows that abnormal cervical muscle activity disappears during the first sleep cycle of ICD patients. In addition, sleep has an alleviating influence on the cervical spinal pain that accompanies the ICD. Although the underlying mechanisms of both sleep-related ameliorations in ICD symptoms are still unclear, it is interesting to note that comparable effects of sleep are seen in a wide variety of movement disorders. For example, although abnormal muscle contractions are not completely abolished, sleep decreases considerably the abnormal movements seen while awake in Parkinson’s disease, Huntington’s disease, and Gilles de la Tourette’s syndrome.

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REFERENCES