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ORIGINAL COMMUNICATION

Quality of life of ALS and LIS patients with and without invasive mechanical ventilation

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Abstract There are very few studies where quality of life (QOL) is assessed in patients with complete physical and functional disability and dependence to invasive mechanical ventilation (IV). We compared QOL of amyotrophic lateral sclerosis (ALS) and locked-in-syndrome (LIS) patients with invasive mechanical ventilation to ALS and LIS patients without mechanical invasive ventilation. Thirty-four patients, 27 with ALS and seven with LIS (vascular or tumoral aetiology) were included in the study. Twelve had invasive ventilation, 22 had non-invasive ventilation, and in the non-invasive ventilation group, five of them had ventilation via mask. The following scales were used for patients: ALS Functional Rating Scale (ALSFRS), McGILL, Short-Form 36 (SF36), Beck Depression Inventory-II, the Toronto Alexithymia Scale and the anxiety inventory of Spielberger. Mean ALSFRS scores were significantly lower in the invasive ventilation group (IV) than in the non-invasive ventilation group. McGILL and SF36 were not significantly different between the IV group and the non-invasive ventilation group; there were no significant differences between the two groups for others scales either. Comparison between IV group and LIS without invasive mechanical ventilation revealed no significant difference for SF36 and McGILL QOL scores. QOL was not significantly different between the IV and not invasively ventilated patients, but ALSFRS was significantly lower in the IV group, and comparison of QOL scores between non-ventilated LIS patients who had the same score of dependence that invasively ventilated patients did not show any difference. Invasive mechanical ventilation for patients who accept tracheotomy allows life prolongation and their QOL is not affected; medical teams should be aware of that.

Keywords Mechanical ventilation · ALS · LIS · Quality of life

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Introduction

In ALS, respiratory muscle weakness leads to hypoventilation and death is usually due to respiratory failure [1]. The mean survival time without invasive ventilation is 3–5 years [2]. ALS has wide ranging effects on individuals' health-related quality of life. As the disease progresses, patients are confronted with increasing physical impairment that can culminate in locked-in state. Non-invasive ventilation improves quality of life [3]. Invasive mechanical ventilation for patients who accept tracheotomy allows life prolongation [4, 5]. Outcome of ALS patient dependent on mechanical ventilation is similar to locked-in-syndrome (LIS) patients dependent on mechanical ventilation. Both categories of patients have a healthy brain locked into a paralysed body. In both affections intellectual function and personality are usually preserved. In general, physical status in ALS patients seems poorly correlated with QOL [6–8]. Despite severe and progressive paralysis of muscle function caused by ALS, it is a common clinical experience that a significant proportion of patients with ALS report an unexpectedly high level of QOL [9].

We designed this study to compare QOL of ALS and LIS patients with invasive mechanical ventilation to ALS and LIS patients without invasive mechanical ventilation. The goal of this study was to assess and compare QOL for patients with and without invasive mechanical ventilation. As invasive mechanical ventilation is the only means to

prolong survival in ALS patients, very few studies have assessed QOL of ventilator dependent patients.

Methods

We enrolled 34 patients in this study, 27 with probable or definite diagnosis of ALS according to the El Escorial criteria [10], and seven patients with locked-in-syndrome of vascular or tumoral origin according to the description by Plum and Posner in 1983 [11]. Cause of LIS was basilar artery ischemia for six of them; one of them had a pontine neurofibroma. Eight ALS and four LIS patients had invasive mechanical ventilation. All subjects were enrolled in our hospital which is specialised in neurological rehabilitation. Patients with previous history of psychiatric disease, clinically overt frontotemporal dementia, clinically evaluated, were excluded from the study. Mean age was 56.7-years-old (range 24–82 years), 22 men, 12 women. Disease duration had a median of 29 months (range 5–204 months). Twelve patients had invasive ventilation (via tracheotomy) (IV group), 22 patients had non-invasive mechanical ventilation (no ventilation group). In the non-invasive ventilation group, 17 patients had no ventilation at all, five study participants had intermittent non-invasive ventilation (ventilation via nasal or facial mask) (NIV group).

The following scales were administered to all patients: ALS Functional Rating Scale (ALSFRS). This scale assesses the patients' physical status and dependence: bulbar, limb, and respiratory function. The total score range from 0 (worst) to 40 (normal). McGILL QOL is a patient-generated measure of individual QOL perceived by the patient. Short-Form 36 (SF36) contains 36 items over eight domains, from which physical (PCS) and mental (MCS) component summary scores may be calculated.

The Beck Depression Inventory-II (BDI-II), is a self report scale that measures depression severity (range from 0 = no depression to 63 = severe depression). The Toronto Alexithymia Scale (TAS) is a 20-item instrument that is one of the most commonly used measures of alexithymia. Alexithymia refers to people who have trouble identifying and describing emotions and who tend to minimize emotional experience and focus attention externally. Equal to or less than 51 = no-alexithymia, scores of 52–60: possible alexithymia, equal to or greater than 61 = alexithymia.

Anxiousness was evaluated with the anxiety inventory of Spielberger (STAI-Y); this test can determine basal and reactive anxiousness.

Statistics

We used Epi Info version 6 (EPI6) to calculate the differences between the IV group and the NIV using the Mann–Whitney U test. The significance level was set at 0.05.

Results

Nineteen patients had gastrostomy, five had intermittent non-invasive ventilation, and there were five bulbar forms of ALS (all in the IV group). The mean age of the IV group was lower than that of the no ventilation group (49 years vs. 61 years, range: 24-64 years and 35-82 years, respectively; $p = 0.02$). Mean duration of evolution was 38 months range (5–204), there was no statistical difference between groups for duration of evolution. IV patients were dependent on invasive ventilation for a mean of 43.5 months (range 2–204 months). The overall score of the ALSFRS ranged from 0 to 40 with a mean of 10. In the non-invasive mechanical ventilation group, mean ALSFRS score was 15, mean SF36 score was 75.1, mean McGILL score was 92.8, mean BDI-II score was 18.8, 22.7% patients had no depression, 31.8% patients had mild depressive symptoms, 27.3% patients had moderate depressive symptoms and 18.2% patients had severe depressive symptoms. Mean TAS score was 60.3. Mean Spielberger score was 53.6 for basal anxiousness and 52.2 for reactive anxiousness (Table 1).

In the IV group, mean ALSFRS was 0.6, mean SF36 score was 74.6, mean McGILL score was 79, mean BDI-II score was 16.3, 33.3% patients had no depression, 25% patients had mild depressive symptoms, 33.3% had moderate depressive symptoms and 8.4% had severe depressive symptoms. Mean TAS score was 56.9. Mean Spielberger score was 53 for basal anxiousness and 52 for reactive anxiousness (Table 1).

Mean ALSFRS scores were significantly lower in the IV group than in the no ventilation group ($p = 0.0001$). Comparison of McGILL and SF36 (total and MCS or PCS) scores between NIV patients did not differ from patients who were invasively ventilated via tracheotomy.

Comparing between the IV and non-invasive ventilation groups, there were no significant differences between the two groups for McGILL, total SF36, TAS, Spielberger and BDI-II (Table 1). Comparing between the IV group and LIS without invasive mechanical ventilation revealed no significant difference for SF36 and McGILL QOL scores.

Discussion

Previous studies suggested that non-invasive mechanical ventilation leads to a longer survival time and better QOL in ALS patients [3–5, 12]. Invasive mechanical ventilation for ALS patients is rarely proposed by medical teams

Table 1 Comparison between invasively ventilated patients and non invasively ventilated patients

	IV group ($N = 12$) (8 ALS, 4 LIS)	Non-invasive mechanical ventilation ($N = 22$) (5 NIV, 17 no ventilation at all)	p values
ALSFRS	0.66 (2.3)	15.3 (11.2)	<0.01
SF36	74.6 (16.4)	75.1 (4.6)	0.94
McGILL	79 (21.4)	92.8 (17.2)	0.08
BDI-II	16.3 (9.9)	18.8 (8)	0.32
TAS	56.9 (11.6)	60.3 (11.3)	0.85
STAI-Y			
Basal anxiousness	53 (13.5)	53.6 (10.2)	0.97
Reactive anxiousness	52 (13.2)	52.2 (10.8)	0.59

The values in bracket represent SD

because, while it increases survival as patients are in a nearly locked-in state, it is with supposedly poor QOL for these patients. QOL assessment in ALS poses a particular challenge because recent studies have shown that the individual, patient-perceived QOL in ALS patients does not correlate with physical function [6–8, 13–21].

While QOL of patients with ALS has been largely investigated, relatively few studies have assessed QOL of ventilator-dependent patients. Given the widely held perception that life, when dependent on artificial ventilation, is not worth living, we examined and compared QOL in ventilated and non-ventilated patients.

We chose to compare QOL of patients with invasive mechanical ventilation and QOL of patients without invasive mechanical ventilation using two different QOL scales, SF36 and a heath-related quality of life questionnaire. SF36 is a surrogate measure for physical function in ALS with two parts, MCS and PCS, and McGILL is a patient-generated measure of individual QOL where the physical and functional aspects of quality of life are not as pronounced. McGILL has been effectively used in patients with ALS and has good validity [7, 13, 21]. In contrast with health-related quality of life questionnaires, scales such as the Schedule for the Evaluation of Individual Quality of Life-Direct Weighting (SEIQOL-DW) and McGILL point out areas of life relevant for patients [9]. In our study there was no difference of QOL between patients without invasive ventilation and patients dependent on invasive mechanical ventilation, this result is in agreement with some previous studies [6–8, 13, 15], except the study of Kaub-Wittemer et al. [22]. Although mean ALSFRS scores in these studies were above 22, in our study, ALSFRS mean scores were very low: 10 with 63% <15.

Our IV patients had the same QOL (McGILL and SF36) as patients without invasive mechanical ventilation in our study, although ALSFRS was significantly lower in the IV group (mean ALSFRS = 0.6) as IV patients were in an advanced stage of the illness or in locked-in state. As the disease advances, patients reported more physical dependency, accompanied by greater stress and anxiety, with no significant impact on QOL. All patients of the IV group (invasively ventilated) communicated with communication aids and it is known that loss of speech leads to emotional suffering and deterioration of QOL [23].

In our work, neither the clinical or demographic characteristics of the patients that we investigated, except for age, were significantly different between the two groups.

Although non-ventilated LIS patients who had the same score of dependency that invasively ventilated patients did not show any difference in QOL score with McGILL and as well as SF36, this emphasis the fact that in patients with the same level of dependence, invasive mechanical ventilation does not affect QOL.

Fifteen percent of our subjects with ALS or LIS had severe depression symptoms and 46% of our patients had any depressive symptoms. This fact agrees with already published data that found 44–75% of their ALS patients exhibited significant signs and symptoms of depression [24–26]. Prevalence of depression was not different between invasively ventilated and non-ventilated patients or between IV and NIV patients, a fact that is coherent with Kübler et al. [27]. Alexithymia scores were not statistically different between invasively ventilated patients and patients without invasive ventilation, and invasive mechanical ventilation does not influence basal and reactive anxiousness evaluated by STAI-Y.

There are, to our knowledge, very few studies where QOL is assessed in patients with complete physical and functional disability and dependence on invasive mechanical ventilation. There is a lack of data referring to QOL in invasively ventilated patients; most data in literature refer to non-invasive ventilation. Invasive mechanical ventilation for patients who accept tracheotomy allows life prolongation and their QOL is not affected; these data can help medical teams to help patients make a decision on tracheotomy. However, it is a real ethical problem because tracheotomy condemns the patients to a total dependence on caregivers who are subjected to a daily and permanent presence. It is then necessary for the caregivers to have a therapeutic education for those special materials and caregiving.

Furthermore, for patients around whom such surroundings cannot be gathered, long-term hospitalization remains the only option, which is expensive for the hospital system. When the ventilator-dependent patients decide to go back home, this situation represents a heavy burden for caregivers and is less secure.

Conflict of interest The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

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