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A survey on self-assessed wellbeing in chronic locked-in syndrome: happy majority, miserable minority

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Abstract

Objectives: Locked-in syndrome (LIS) consists of anarthria and quadriplegia, while consciousness is preserved. Classically, vertical eye movements or blinking allow coded communication. Given appropriate medical care, survival may reach decades. We here studied the self-reported quality of life (QoL) of chronic LIS patients.

Design: 168 LIS members of the French Association for LIS were invited to answer a questionnaire on medical history, current status and end-of-life issues. They self-assessed their global subjective wellbeing with the Anamnestic Comparative Self Assessment (ACSA) scale, whose +5 and -5 anchors were their memories of the best period in their life *before* LIS and their worst period *ever, respectively*. **Results:** 91 patients (54%) responded and 26 were excluded because of missing data on QoL. 47 patients professed happiness (median ACSA +3) and 18 misery (median ACSA -4).

Variables associated with unhappiness included anxiety and dissatisfaction with mobility in the community, recreational activities and with recovery of speech production. A longer time in LIS was correlated with happiness. 58% declared not to wish resuscitation in case of cardiac arrest and 7% expressed the wish for euthanasia.

Conclusions: Our data stress the need for extra palliative efforts directed at mobility and recreational activities in LIS and the importance of anxiolytic therapy. Recently-struck LIS patients who wish to die should be assured that they have a considerable probability of regaining a happy meaningful life. End-of-life decisions, including euthanasia, should not be avoided, but a moratorium allowing the reaching of steady state should be proposed.

Article summary

Article focus

The objective of this study is to

- describe chronic locked-in patients' subjective well being and identify factors that are associated with high or low overall subjective well being.
- evaluate the degree to which locked-in patients are able to return to a normal life
- assess the views of locked-in patients on end-of-life issues

Key messages

- Most chronic locked-in patients self-report severe restrictions in community reintegration. Nevertheless, the majority (72%) of our sample professes “good” subjective wellbeing. This is in line with the notion that patients with severe disabilities may report a good quality of life despite being socially isolated or having major difficulties in activities of daily living
- 28% of our locked-in patients declared unhappiness. Variables associated with unhappiness were dissatisfaction with mobility in the community, with recreational activities and with capacity to face up to life events. Shorter time in locked-in, anxiety and non-recovery of speech production were also associated with unhappiness.
- The principal clinical conditions for requests for euthanasia or physician-assisted death to be legally valid are “unbearable” suffering and irreversibility of the situation. Whereas the first condition may apply in some locked-in patients, irreversibility cannot be ascertained until, after the acute setting and rehabilitation, their subjective well being has reached steady state, which may take as long a year.

Strengths and limitations of this study

- This study is the largest survey of chronic locked-in syndrome patients ever performed and assesses patients' own self-assessed quality of life, general wellbeing and end-of-life wishes. The clinical and ethical implications are evident and important for the medical community at large.
- We here also identify variables associated with unhappiness that can be improved and permit evidence-based policy changes in the management of these challenging and vulnerable patients.
- Our study may suffer from a low response rate and selection bias and results might hence not be representative for chronic LIS patients in general as all participants were member of a patient association (i.e., French Association of LIS), indicating a stable condition and possibly a degree of social integration. Nonetheless, as discussed in the paper, quality of life research has many methodological pitfalls, especially in this low-incidence pathology with limited and difficult communication.

Introduction

Locked-In Syndrome (LIS) is defined by quadriplegia (or quadriparesis) and aphonia (or severe hypophonia) with a primary mode of communication by eye movements or blinking.¹ Most often LIS is caused by an acute (vascular) anterior pontine brainstem lesion. The syndrome can be subdivided on the basis of motor disability²: “*classic*” LIS is characterized by quadriplegia and aphonia with coded communication by vertical eye movement or blinking; “*incomplete*” LIS patients have remnants of voluntary motion other than vertical eye movement; and “*total*” LIS is defined by complete immobility, including all eye movements. Once a patient is medically stabilized in LIS, the 10-year survival is more than 80%.³ With intensive rehabilitation⁴, many “*classic*” LIS patients may evolve to “*incomplete*” LIS, with voluntary control of head, fingers or foot and sometimes recovery of speech production.⁵ Nearly all chronic LIS patients remain dependent on others for activities of daily living. Physicians and caregivers may tend to consider that LIS patients will die anyway³ or would prefer to die if they knew what the clinicians (think they) know.⁶ On the other hand, it is known that people with severe persistent disability tend to self-report good subjective wellbeing (SWB) – the “*disability paradox*”.⁷ A previous study assessing the QoL in fifteen LIS patients showed that they reported normal mental and personal general health despite maximal restriction in physical activities.⁸ The objective of this study is to describe chronic LIS patients’ SWB and their views on end-of-life issues, and to identify factors that are associated with high or low overall SWB.

Methods

Participants and Procedures

In collaboration with the French association for LIS (ALIS; <http://alis-asso.fr> – a non-profit association created in 1997 to help LIS patients and their families), 168 member LIS patients were invited (in January 2008) by letter to fill in a structured questionnaire, aided by the patient's proxy. The questionnaire included items about socio-demographic (age, gender, educational level, place and condition of living, religiosity, net monthly household income), clinical (etiology and duration of LIS, level of speech production and motor recovery) and quality of life (QoL) and SWB variables. We used the postal French version of the Reintegration to Normal Living Index (RNLI)⁹⁻¹², which evaluates the degree to which a patient has been able to return to a normal life. The RNLI is an 11-item scale that covers areas such as participation in recreational and social activities, movement within the community, and how comfortable the individual is in his or her role in the family and with other relationships. Given the specific constraint of eye-coded communication in the surveyed LIS patients, instead of the visual analogue scale¹¹, a 4-point Likert scale was used as described elsewhere¹³ where a value of 1 was assigned to “no”, 2 to “rather no”, 3 to “rather yes” and 4 to “yes”. The scores were normalized to 100, with a score of 100 indicating that the participants were fully satisfied, scores of 60 through 99 indicating mild to moderate restrictions in self-perceived community reintegration, and scores less than 60 indicating severe restrictions in self-perceived community reintegration, as previously reported.¹⁴ Overall SWB was rated by means of the Anamnestic Comparative Self-Assessment (ACSA)¹⁵ scale, whose biographical +5 and -5 scale anchors were the patients' memories of the best period in their life *before* LIS and their worst period *ever* (Figure 1). Participants were also asked about

the presence of depressive symptoms (yes, no), pain and anxiety (none, moderate, extreme), end-of-life issues: suicidal thoughts (never, occasionally, often), resuscitation in case of cardiac arrest (yes, no) and euthanasia (envisaged, never envisaged). Completion of the anonymous questionnaire was voluntary and taken as consent for participation in the survey. The study was approved by the ethics committee of the Faculty of Medicine of the University of Liège.

INSERT FIGURE 1 ABOUT HERE

Statistical analysis

Data were analyzed using Stata 10.0 (StataCorp. 2007 Stata Statistical Software: Release 10. College Station, TX). The normality distribution of continuous variables was assessed using Shapiro-Wilk tests. For the descriptive analyses, we used subject counts and percentages for categories, calculating mean \pm SD or median with interquartile range (IQR) for continuous variables. Only questions with a response rate above 70% were considered representative of the population and considered for further analyses. ACSA ratings were divided into happy (ratings 0 to 5) and unhappy (ratings -1 to -5). Univariate associations between the dependent variable happy/unhappy and the RNLI and end-of-life questions co-variables were assessed using Student t-test, Wilcoxon or Chi-square tests as appropriate. Multivariable backward stepwise logistic regression was used to assess the associations between happiness status and the significant covariates selected by the univariate analysis. Analyses were performed using casewise deletion. Differences were considered as significant at $p < 0.05$.

Results

One hundred and sixty eight LIS patients were invited to fill in the structured questionnaire. Seventy seven patients did not reply (46%). Among the 91 LIS patients who replied, 26 responses (29%) had missing RNLi and/or ACSA data and were excluded: 5 subjects gave ACSA ratings but failed to answer RNLi questions and 21 failed to report ACSA ratings. The included sample hence consisted of 65 patients (figure 2). LIS patients who failed to report QoL and/or SWB (n=26) were lower-educated ($p=.009$) and had more physical pain ($p=.009$) as compared to LIS patients included in the QoL and SWB study sample (n=65). Other socio-demographic, physical and functional variables including age, duration in LIS, living at home or with partner, income, recovery of speech production or limb mobility were not different between QoL/SWB non-respondents and respondents (Table 1). The socio-demographic characteristics of the 65 LIS patients included in our sample are shown in Table 2. All had chronic LIS (> 1 year after the insult; median 8 years, range 1-28 years) following a brainstem vascular accident. The majority lived at home (64%), had a spouse or partner (64%) and were religious (70%). About half (55%) had recovered some speech and 70% recovered some limb mobility.

INSERT FIGURE 2 ABOUT HERE

Table 1 : Respondents versus non-respondents to Reintegration to normal Living Index (RNLI) and Anamnestic Comparative Self-Assessment Scale (ACSA) locked-in syndrome (LIS) patients' socio-demographic and disease characteristics.

	LIS patients included in our sample (N=65)	LIS patients who failed to report RNLI/ACSA (N=26)	P value
Mean age \pm SD (years)	49 \pm 11	52 \pm 12	0.46
Gender : Men	43/63 (68%)	16/26 (61%)	0.72
Mean duration in LIS (years): median (IQR)	8 (5 - 10)	7 (4 - 9)	0.60
Educational level : university or college (vs high school or lower)	25/56 (45%)	3/21 (14%)	0.009
Net monthly household income \geq 2500 € (vs < 2500 €)	20/53 (38%)	5/24 (21%)	0.13
Place of living : home (vs institution)	38/59 (64%)	18/26 (69%)	0.65
Living with spouse or partner (vs single)	36/56 (64%)	15/25 (60%)	0.93
Religious (vs non-religious)	40/57 (70%)	21/26 (81%)	0.3
Recovery of speech production			0.99
None	26/58 (45%)	11/23 (48%)	
Words	11/58 (19%)	5/23 (22%)	
Sentences	21/58 (36%)	7/23 (30%)	
Recovery of some limb mobility	39/56 (70%)	14/23 (61%)	0.59
Suicidal thoughts			0.56
Never	40/59 (68%)	16/20 (80%)	
Occasionally	14/59 (24%)	3/20 (15%)	
Often	5/59 (8%)	1/20 (5%)	
Anxiety			0.47
No	20/61 (33%)	8/22 (36%)	
Moderate	33/61 (54%)	9/22 (41%)	
Extreme	8/61 (13%)	5/22 (23%)	
Pain			0.009
None	32/59 (54%)	4/21 (19%)	
Moderate	25/59 (42%)	14/21 (67%)	
Extreme	2/59 (4%)	3/21 (14%)	
Euthanasia			0.22
Envisaged	31/59 (53%)	6/19 (32%)	
Never envisaged	28/59 (47%)	13/19 (68%)	
Depression			0.66
Yes	8/60 (13%)	3/17 (18%)	
No	56/60 (87%)	14/17 (82%)	

Table 2 : LIS patients' socio-demographic, disease characteristics quality of life and end-of-life data in unhappy (ACSA ratings <0) versus happy (ACSA ≥0) respondents. Note: ACSA = Anamnestic Comparative Self-Assessment Scale; IQR = inter-quartile range; * significant variables identified by the univariate analyses comparing unhappy versus happy with related p values.

	All patients (N=65)	Unhappy (ACSA <0) (N=18)	Happy (ACSA ≥0) (N=47)	P value
Mean age ± SD (years)	49±11	50±10	49±12	0.571
Gender : Men	43/63 (68%)	12/18 (67%)	31/45 (69%)	0.864
Mean duration in LIS*: median (IQR)	8 (5 - 10)	7 (3 - 8)	9 (5 - 13)	0.005
Educational level : university or college (vs high school or lower)	25/56 (45%)	5/16 (31%)	20/40 (50%)	0.197
Net monthly income ≥ 2500 €(vs < 2500 €)	20/53 (38%)	4/13(30%)	16/40 (40%)	0.547
Place of living : Home (vs Institution)	38/59 (64%)	9/17 (53%)	29/42 (69%)	0.247
Living with spouse or partner vs single	36/56 (64%)	9/16 (56%)	27/40 (67%)	0.431
Religious (vs non-religious)	40/57 (70%)	13/16 (81%)	27/41 (66%)	0.240
Recovery of speech production*				
None	26/58 (45%)	10/16 (63%)	16/42 (38%)	0.049
Words	11/58 (19%)	4/16 (25%)	7/42 (17%)	
Sentences	21/58 (36%)	2/16 (12%)	19/42 (45%)	
Recovery of some limb mobility	39/56 (70%)	10/17 (59%)	29/39 (74%)	0.252
Reintegration to normal living index items				
I move around my living quarters as I feel necessary				
Yes	23/60 (38%)	6/17 (35%)	17/43 (40%)	0.453
Rather yes	13/60 (22%)	2/17 (12%)	11/43 (26%)	
Rather no	10/60 (17%)	3/17 (18%)	7/43 (16%)	
No	14/60 (23%)	6/17 (35%)	8/43 (18%)	
I move around my community as I feel necessary*				
Yes	6/59 (10%)	0/18 (0%)	6/41 (15%)	0.042
Rather yes	16/59 (27%)	4/18 (22%)	12/41 (29%)	
Rather no	14/59 (24%)	3/18 (17%)	11/41 (27%)	
No	23/59 (39%)	11/18 (61%)	12/41 (29%)	
I am able to take trips out of town as I feel necessary				
Yes	17/57 (30%)	3/17 (18%)	14/40 (35%)	0.298
Rather yes	13/57 (23%)	3/17 (18%)	10/40 (25%)	
Rather no	7/57 (12%)	2/17 (11%)	5/40 (13%)	
No	20/57 (35%)	9/17 (53%)	11/40 (27%)	
I am comfortable with how my self-care needs (dressing, feeding, toileting, bathing) are met				

Yes	36/59 (61%)	10/18 (56%)	26/41 (64%)	0.292
Rather yes	16/59 (27%)	4/18 (22%)	12/41 (29%)	
Rather no	3/59 (5%)	1/18 (6%)	2/41 (5%)	
No	4/59 (7%)	3/18 (16%)	1/41 (2%)	
I spend most of my days occupied in work activity that is necessary or important to me				
Yes	8/60 (13%)	1/18 (5%)	7/42 (17%)	0.390
Rather yes	5/60 (8%)	2/18 (12%)	3/42 (7%)	
Rather no	7/60 (12%)	1/18 (5%)	6/42 (14%)	
No	40/40 (67%)	14/18 (78%)	26/42 (62%)	
I am able to participate in recreational activities (hobbies, crafts, sports, reading television games, computers etc.) as I want to*				
Yes	35/61 (57%)	8/18 (44%)	27/43 (63%)	0.028
Rather yes	18/61 (31%)	4/18 (22%)	14/43 (33%)	
Rather no	2/61 (3%)	1/18 (6%)	1/43 (2%)	
No	6/61 (9%)	5/18 (28%)	1/43 (2%)	
I participate in social activities with family friends and or business acquaintances as is necessary or desirable to me				
Yes	22/60 (37%)	5/18 (28%)	17/42 (40%)	0.113
Rather yes	14/60 (23%)	4/18 (2%)	10/42 (24%)	
Rather no	9/60 (15%)	1/18 (6%)	8/42 (19%)	
No	15/60 (25%)	8/18 (44%)	7/42 (17%)	
I assume a role in my family which meets my needs and those of my family members				
Yes	25/59 (42%)	8/18 (44%)	17/41 (41%)	0.396
Rather yes	21/59 (36%)	4/18 (22%)	17/41 (41%)	
Rather no	5/59 (8%)	2/18 (12%)	3/41 (8%)	
No	8/59 (14%)	4/18 (22%)	4/41 (10%)	
In general I am comfortable with my personal relationships				
Yes	32/61(52%)	9/18 (50%)	23/43 (53%)	0.884
Rather yes	18/61 (30%)	5/18 (28%)	13/43 (30%)	
Rather no	4/61 (7%)	1/18 (5%)	3/43 (7%)	
No	7/61 (11%)	3/18 (17%)	4/43 (10%)	
In general I am comfortable with myself when I am in the company of others				
Yes	27/60 (45%)	7/18 (39%)	20/42 (48%)	0.293
Rather yes	19/60 (31%)	4/18 (22%)	15/42 (36%)	
Rather no	7/60 (12%)	3/18 (17%)	4/42 (9%)	
No	7/60 (12%)	4/18 (22%)	3/42 (7%)	
I feel that I can deal with life events as they happen*				
Yes	20/60 (33%)	6/18 (33%)	14/42 (33%)	0.022
Rather yes	18/60 (30%)	3/18 (17%)	15/42 (36%)	
Rather no	12/60 (20%)	2/18 (11%)	10/42 (24%)	
No	10/60 (17%)	7/18 (39%)	3/42 (7%)	
<u>Depression, anxiety and pain</u>				
Depression*				
Yes	8/60 (13%)	5/18 (28%)	3/42 (7%)	0.040

No	52/60 (87%)	13/18 (72%)	39/42 (93%)	
Anxiety*				
No	20/61 (33%)	5/18 (28%)	15/43 (35%)	0.015
Moderate	33/61 (54%)	7/18 (39%)	26/43 (60%)	
Extreme	8/61 (13%)	6/18 (33%)	2/43 (5%)	
Pain				
No	32/59 (54%)	10/18 (56%)	22/41 (54%)	0.071
Moderate	25/59 (43%)	6/18 (33%)	19/41 (46%)	
Extreme	2/59 (3%)	2/18 (11%)	0/41 (0%)	
End-of-life issues				
Resuscitation in case of cardiac arrest*				
Yes	23/55 (42%)	2/14 (14%)	21/41 (51%)	0.011
No	32/55 (58%)	12/14 (86%)	20/41 (49%)	
Euthanasia*				
Envisaged	31/59 (53%)	12/16 (75%)	19/43 (44%)	0.032
Never envisaged	28/59 (47%)	4/16 (25%)	24/43 (56%)	
Suicidal thoughts*				
Never	40/59 (68%)	9/17 (54%)	31/42 (74%)	0.040
Occasionally	14/59 (24%)	4/17 (23%)	10/42 (24%)	
Often	5/59 (8%)	4/17 (23%)	1/42 (2%)	

Overall SWB, as measured by the ACSA scale, permitted to disentangle two subpopulations: 72% of LIS patients declared happiness (ACSA rating ≥ 0 , median +3) and 28% unhappiness (ACSA rating < 0 , median -4) (Figure 3). As assessed by RNLI, 51% of the sampled LIS patients reported severe restrictions and 49% reported mild to moderate restrictions in self-perceived overall community reintegration. Most (82%) were comfortable with personal relationships, but only 21% were engaged most of the day in activities which they considered important. Only a minority were dissatisfied with their participation in recreational (12%) or social (40%) activities.

The happy and unhappy groups did not differ for socio-demographic, physical and functional variables including religion, living at home or with partner, income, education, physical care, and feeling comfortable in the company of others (Table 2). Depression, suicidal ideas, consideration or wish of euthanasia and the wish not to be

resuscitated in case of cardiac arrest were significantly more frequent in the unhappy group. Variables associated with unhappiness were dissatisfaction with mobility in the community, with recreational activities and with capacity to face up to life events. Shorter time in LIS, anxiety and non-recovery of speech production were also associated with unhappiness. Only half of the respondents stating happiness wished to be resuscitated in case of cardiac arrest and this rate was as low as 14% among the unhappy respondents ($p=.011$) (Table 2). Multivariate logistic regression showed that the variables associated with happiness were time spent in LIS ($p=.007$), absence of anxiety ($p=.032$) and recovery of speech production ($p=.013$) (Table 3).

INSERT FIGURE 3 ABOUT HERE

Table 3: Significant associations between happiness status and variables identified by the univariate analyses (marked by an asterix in Table 2). *Odds ratio per year in LIS.

	Odds Ratio	Std. Err.	Z	$p> z $	95% Conf. Interval
Duration in LIS*	1.5	0.2	2.71	0.007	1.1-2.0
Speech production	20.47	24.87	2.48	0.013	1.89-221.45
Anxiety	0.19	0.15	-2.14	0.032	0.04-0.87

Discussion

It is important to stress that our study may suffer from a selection bias given that only 91 of 168 invited patients participated in the study (54%). The patients who did not return the survey might be the ones showing the lowest QoL. Our results might hence not be representative for chronic LIS patients in general. All participants were member of a patient association (i.e., French Association of LIS), indicating a stable condition and possibly a degree of social integration.¹⁶ Given the dependence of LIS participants on the help of a caregiver for communication of the survey answers, a dependency relationship, social desirability¹⁷ or “self presentation”^{18, 19} may also have biased some responses - despite a written recommendation not to allow helpers to influence responses. Patients with a lower educational level and more pain were under-represented among those answering the QoL questions. This might also have overestimated QoL rates in our LIS patients as low educational level^{6, 20} and presence of pain are inversely associated with satisfaction with QoL.²¹⁻²³ In sum, some methodological constraints may have biased the SWB ratings of our patients and most biases were likely to result in overestimations of SWB.

A recurrent problem in QoL research¹⁹ is the possible relativity bias or response shift, by which, for example, patients with severe chronic conditions tend to assess their QoL relative to peers or given the circumstances. This problem tends to invalidate comparisons of SWB between groups.²⁴ Such relativity biases may result in rather similar responses across objectively very dissimilar disease groups, and even between healthy and diseased people²⁵, including those with spinal cord injury.²⁶ We have therefore chosen to employ the ACSA scale, a self-anchored scale whose upper limit here was the memory of the best period in the patients’ life experience before their LIS state. A strength of the ACSA methodology is that by virtue of its biographical

references it constitutes a practical compromise between the hedonic and the eudaimonic philosophies of QoL, allowing the respondent to choose between the two perspectives, or to take both into account.²⁷ This internal standard of ACSA reduces the likelihood of peer relativity or “under-the-circumstances” responses.²⁸ However, the retrospective anchoring of the ACSA scale also has a drawback. Paraplegic patients may recall their past as happier than did controls, a phenomenon called the “nostalgia effect”.²⁹ If this applies in LIS, it would have tended to depress the ratings of current SWB with ACSA. This said, some authors have played down relativity biases, arguing that “given the circumstances” responses of disabled people must be taken at face value and that there is no such thing as a disability paradox.³⁰

Given these limitations, our results show that most chronic LIS patients self-report severe restrictions in community reintegration, in line with previous studies in paraplegia following spinal cord injury.^{16, 31} Nevertheless, the majority of our sample professes “good” subjective wellbeing. This is in line with the notion that patients with severe disabilities may report a good QoL despite being socially isolated or having major difficulties in activities of daily living.^{7, 32} That some LIS patients self-report happiness may suggest that they have succeeded in adapting to their condition of extreme physical disability. According to Cummins’ theory of SWB, their homeostatic resources may have overcome even the formidable challenge of LIS.³³ Our results hence underscore the strength of homeostatic processes of adaptation to chronic (often definitive) extreme disability.

The “happy” subgroup of LIS survivals may indeed be those capable of high flexibility and plasticity who have fully succeeded in recalibrating, reprioritizing and reorienting their needs and values, whereas the low raters cope poorly because they cannot shed needs and values from their previous life. Because such an adaptation

process is lengthy, this hypothesis is consistent with the observed positive association of SWB with duration of time in LIS, corroborating previous studies on QoL in spinal cord injury patients.^{31, 34} However, the direction of causality might be otherwise if unhappiness and its correlates, by whatever mechanisms, reduce survival. Unhappy patients may be more susceptible to complications, these may be treated less vigorously and more end-of-life decisions may be made. The association between a long time in LIS and happiness would then be an effect of selection by attrition. The second finding is that some LIS patients self-report a state of misery. Depression, suicidal ideas and a wish not to be resuscitated were associated with unhappiness, but are most probably co-variables of unhappiness rather than causal factors. The identified predictors of SWB in LIS differed somewhat from those previously reported in traumatic brain or spinal cord injury.^{35, 36} Living conditions appear less determining for SWB than in spinal cord injury, possibly because in LIS expectations are lower. In our LIS patients, partner relationships also not correlated with SWB, unlike in traumatic spinal cord or brain injury^{37, 38} or in patients with multiple sclerosis.³⁹ Importantly, a shorter time spent in LIS, anxiety and no recovery of speech production were found as possible predictors of unhappiness. Yet, the studied variables and objective characteristics explained only 38% of the variance of overall SWB. Maybe this was because some potentially important variables such as personality traits could not be explored in our study. Other variables associated with unhappiness that can potentially be remediated included dissatisfaction with mobility in the community and with recreational activities, in line with previous studies on QoL in spinal cord injury, multiple sclerosis, cerebral palsy and stroke.^{40, 41}

For clinical practice, our data show that, whatever the physical devastation and the mental distress of LIS patients during the acute stage of the condition, optimal life-

sustaining care and revalidation can have major long-term benefit. Maybe, since low satisfaction with mobility and recreational activities were here associated with poor SWB, extra palliative efforts directed at these problems could be helpful. Also more vigorous treatment of anxiety may be helpful. Finally, our results also bear on existential and ethical issues⁴². Because they are cognitively intact⁴³, LIS patients are competent to make decisions on whether to continue life in LIS or to ask for withholding or withdrawal of treatment or for physician-assisted death.⁴⁴ That half of the respondents professing happiness do not wish to be resuscitated in case of cardiac arrest complicates the interpretation of their statement of happiness. As for current wish of euthanasia, expressed by only 4 of the 59 subjects (7%) responding to this question, it must be taken into account that, unlike in Belgium, the Netherlands, Luxemburg, Oregon, Washington and Montana, euthanasia and physician-assisted suicide are not legally permitted in France, where the study was carried out. What do our data suggest regarding the practice of euthanasia or physician-assisted suicide? The principal clinical conditions for requests for physician-assisted death to be legally valid are “unbearable” suffering and irreversibility of the situation. Whereas the first condition may apply in some LIS patients, irreversibility cannot be ascertained until, after rehabilitation, their SWB has reached steady state, which may take as long a year.³ This is particularly true in view of expected medical progress such as by e.g. brain-computer interfaces (i.e., modes of communication in which commands or messages are emitted directly by the brain without needing motor or verbal mediation).^{45, 46} We suggest that patients recently struck by LIS should be informed that, given proper care, they have a considerable chance of regaining a happy life. In our view, shortening-of-life requests by LIS patients are valid only when the patients have been given a chance to attain a steady state of SWB. Anderson et al. reported

suicidal thoughts in four out of seven LIS patients with long-term survival but all patients nevertheless wanted life-sustaining treatment.⁴⁷ Acute or subacute LIS patients' requests for early death should be received with sympathy, but our data suggest that a moratorium should be proposed.^{48, 49}

Taking into account the methodological challenges and limitations of QoL research, especially when dealing with LIS patients, our data show that a non-negligible group of chronic LIS survivors self-report a meaningful life and their demand for euthanasia is surprisingly infrequent. In our view, these results are important as healthy individuals and medical professionals might assume that the comfort of a LIS patient is so limited that it is not worth living.⁸ Such discrepancies in valuation of disability states between the healthy and those affected raise questions about the validity of utility measures based on valuation of disease states by panels of healthy people using e.g. standard gamble or time trade-off.⁵⁰ Underestimation of patients' self-reported QoL by caregivers and family has previously also been reported for amyotrophic lateral sclerosis patients.^{6, 51, 52} More research is needed to learn about the factors influencing the success or failure of adaptation to LIS. Also, longitudinal studies of LIS patients should throw light on the reversibility of high or low SWB and on when happiness is a consequence or a causal factor of long survival in LIS.

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Competing interests

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Authors' contributions

MAB, SL and JB made were responsible for acquisition, and analysis of the data. MAB and JB interpreted them and drafted the manuscript. DL provided statistical

expertise. JB, SL, DL, FP and AD contributed to critical revision of the manuscript.

All authors read and approved the final manuscript.

Figure 1 : Anamnestic Comparative Self Assessment (ACSA) scale.

Figure 2 : Participation. Note : SWB : subjective well-being ; QoL : quality of life.

Figure 3 : Distribution of Anamnestic Comparative Self Assessment (ACSA) ratings in locked-in syndrome.

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