

Quality of Life and Ethical Issues

N'oubliez jamais que chaque être est un monde en soi, que l'on connaît en grande partie, mais seulement dans la mesure où nous possédons cette même partie en nous-mêmes. Le reste, pourtant, demeurera toujours secret pour nous. Si l'on veut établir des lois nouvelles, elles ne sont valables que pour cette partie des autres que nous connaissons, parce qu'elle existe en nous.

Don't ever forget that each person is a world unto himself and that we can only understand that part of the character of each individual that is in ourselves. The rest will always remain incomprehensible for us. If you want to establish new laws, they can only be valid for that part of others that we understand in ourselves.

— Gustav Mahler to Justi Mahler, *Briefe*, 1891¹

It is now clear that life can be greatly prolonged by ventilator use for many people with respiratory muscle weakness or paralysis and that such people can be happy, productive members of society. However, few health care professionals seem to appreciate this fact.⁶¹⁵ Indeed, there is little written in medical literature about the productivity, social reintegration, and life satisfaction of ventilator users with NMDs. This chapter will review these issues along with ventilator style preferences, complications of ventilator use, and the related ethical issues.

Survey of 700 Ventilator Users

Comment donc parvient-on à connaître les hommes, qui sont bien plus profonds et plus complexes que leurs oeuvres? Il faut les observer avec attention et tendresse. How can we understand men, who are much more profound and complex than their works? One must observe them with attention and affection.

— Gustav Mahler

From a mailing to about 1000 community-based users of equipment provided by a single portable ventilator manufacturer (Respironics, Inc.), 695 users responded to a survey. Four hundred ninety-four of the respondents were supported by noninvasive means and 92 by tracheostomy IPPV; 35 did not indicate the means by which they were ventilated or did not fill out the Likert satisfaction scales; and 74 had used both tracheostomy and noninvasive methods of ventilatory support and responded only to questions concerning ventilator-style preferences. Excluding the 74 who did not respond to the satisfaction items, the remaining 621 respondents were wheelchair users, of whom 585 were

completely unable to walk and were dependent on attendant care for virtually all activities of daily living (ADL). The seven ventilator users with intrinsic lung disease could walk short distances using a rolling walker with a ventilator tray. The diagnoses, mean ages, residual function, hours per day and years of ventilator use, and life satisfaction indices are listed in Table 1. Respondents included 313 males with a mean age of 46.5 years, 306 females with a mean age of 52.2 years, and two whose gender was not indicated. The 621 ventilator users had been dependent on ventilatory support for a mean of 21.1 years and, at the time of the survey, used aid for a mean of 15.7 hours per day. An additional 46 autonomously breathing patients with spinal cord injury (SCI), who were randomly identified from the medical records of a rehabilitation facility, were also included in the survey.

Table 1. Duration of Ventilator Use and Life Satisfaction Index

Diagnosis	No. of Patients	Age (yrs) ^a	Residual Function	Years of Ventilator Use	Hours ^b per day	Life Satisfaction ^c
Individuals using noninvasive aids (n = 494)						
Pollomyelitis	336	54.5 ± 8.9	1.2 ± 0.9	27.8 ± 12.6	15.4 ± 9.7	5.2 ± 1.6
Duchenne dystrophy	47	27.3 ± 7.2	0.6 ± 0.5	7.6 ± 5.0	18.0 ± 6.8	5.1 ± 1.5
Unknown ^d	29	54.3 ± 16.2	1.6 ± 1.3	14.2 ± 12.7	13.9 ± 6.7	4.8 ± 1.3
Traumatic SCI	23	33.7 ± 11.0	0.3 ± 0.7	8.8 ± 6.4	17.5 ± 8.1	4.0 ± 1.3
Non-Duchenne myopathy	23	40.2 ± 17.1	1.5 ± 1.2	8.7 ± 10.4	15.9 ± 6.7	4.6 ± 1.9
ALS	8	48.3 ± 12.5	0.4 ± 0.5	7.0 ± 7.8	19.3 ± 8.1	4.1 ± 1.7
Intrinsic ^e	7	57.1 ± 10.2	2.3 ± 1.2	6.3 ± 7.7	11.6 ± 6.8	4.6 ± 1.4
Myasthenia gravis	5	57.6 ± 11.0	2.0 ± 1.0	18.4 ± 12.1	9.4 ± 2.5	4.6 ± 2.3
Kyphoscoliosis	4	60.3 ± 13.7	2.8 ± 0.5	9.5 ± 3.0	8.5 ± 1.3	4.7 ± 1.9
Polymyositis	3	46.3 ± 11.0	1.7 ± 1.2	11.3 ± 7.8	18.0 ± 5.6	5.0 ± 0.9
Obesity/hypoventilation syndrome	3	39.3 ± 11.5	1.3 ± 0.6	7.0 ± 5.6	7.7 ± 2.5	4.0 ± 2.6
Myelopathy	3	54.7 ± 13.7	0.0 ± 0.0	10.9 ± 10.1	18.2 ± 10.1	5.5 ± 1.5
Multiple sclerosis	2	59.0 ± 24.0	0.5 ± 0.7	20.5 ± 12.0	5.8 ± 6.0	4.7 ± 1.1
Arthrogryposis	1	26.0	2.5	11.0	8.0	5.5
Tracheostomy IPPV users (n = 92)						
Pollomyelitis	44	53.1 ± 10.0	1.4 ± 0.9	24.1 ± 11.8	16.0 ± 6.6	4.6 ± 1.8
Duchenne dystrophy	13	30.8 ± 4.8	0.6 ± 0.5	5.0 ± 3.0	20.8 ± 5.5	4.8 ± 1.3
Non-Duchenne myopathy	12	34.2 ± 13.6	1.3 ± 1.0	9.4 ± 6.5	19.9 ± 6.7	4.9 ± 1.6
ALS	5	51.8 ± 21.7	0.6 ± 0.9	11.4 ± 10.5	20.2 ± 6.1	2.5 ± 1.5
Unknown ^d	5	50.8 ± 20.5	2.4 ± 0.5	8.2 ± 1.5	11.8 ± 7.1	5.5 ± 2.6
Traumatic SCI	4	31.3 ± 9.1	0.0 ± 0.0	30.8 ± 18.2	20.0 ± 8.0	5.4 ± 1.7
Polymyositis	2	41.0 ± 9.9	1.0 ± 1.4	4.0 ± 1.4	23.0 ± 0.0	3.0 ± 2.8
Charcot-Marie-Tooth disease	2	53.5 ± 0.8	1.5 ± 0.7	6.5 ± 4.9	24.0 ± 0.0	4.5 ± 3.5
Kyphoscoliosis	1	47.0	3.0	11.0	8.0	5.5
Myelopathy	1	40.0	1.0	27.0	8.0	5.5
Multiple sclerosis	1	62.0	0.0	6.0	24.0	3.0
Spinal muscular atrophy	1	40.0	1.0	26.0	16.0	7.0
Polynuropathy	1	17.0	0.0	7.0	24.0	5.5

^a Age at the time of the survey.

^b Hours per day of ventilator use.

^c Patient satisfaction with life in general, where 1 is very dissatisfied and 7 is very satisfied.

^d Not reported in the survey.

^e Intrinsic lung disease, including pulmonary fibrosis and chronic obstructive pulmonary disease.

Marriage/Divorce

Those who deem me unworthy at a glance and pass me on by, have my blessing to keep walking for they have a long way to go. They have not reached the point in their journey where they are able to see and appreciate me for who I am.

—Terri McPherson, *Words from a Simple Heart*, terri@wischeara.com

Two hundred seventy-seven of the 621 respondents (45%), 157 men and 120 women, had not married. One hundred eighty-six, 97 men and 89 women, were married before requiring ventilatory support and remained married and living with their spouses. This figure includes four respondents who were widowed before requiring ventilatory support and later remarried while using support and two men who were divorced before requiring support and remarried while using support. These 186 people had been using ventilatory support for a mean of 22.7 years and required 13.7 hours of support per day. Twenty other respondents were married before requiring ventilatory aid and subsequently widowed. Thirty-six respondents, 10 men and 26 women, were married before requiring ventilatory support and have been divorced and remained so while using ventilatory aids. An additional 60 respondents (20%), 32 men and 28 women, who were single before requiring ventilator support were married as ventilator users and live with their spouses. Forty-two people did not respond to this question. Therefore, only 16.2% of the ventilator users who were married before onset of ventilator dependence were divorced subsequently and have not remarried over a mean period of 22.7 years (individual ages: 28–51 years) of ventilator use. This group became ventilator users at the mean age of 28 years. The general nondisabled population has a divorce rate of 30% for people married at the mean age of 28 years.^{109c}

In one report, 12 women with SMA with onset from 8 months to 29 years delivered a total of 17 infants. Complications included premature labor, prolonged labor, and delayed postpartum recovery; cesarean section was performed for 3 cases. There were no deleterious effects on fetal outcome. Exacerbation of muscle weakness was noted by 8 women during the second trimester. Three had recovery in strength in the puerperium.¹⁰⁷ As noted in Chapter 1, a 27-year-old woman with SMA type 1 also had a successful pregnancy.⁶⁶

Employment

We must all—people with disabilities in particular—be allowed the opportunity to satisfy the need we all have to prove ourselves, to contribute to our chosen field, and to our quality of life. Anything to the contrary sends the message that we no longer have any contribution to make, that—for example in my case—unless I am physically capable of transporting myself to the seat of justice, I am incapable of rendering it.^{60z}

—Justice Sam Miller

Two hundred thirty-four ventilator users (134 men, 99 women, and 1 respondent whose gender was not indicated) were gainfully employed (Table 2). Seventeen other ventilator users reported being active on a daily basis as volunteers for various philanthropic causes, and 24 were students. In addition, 32 married women ventilator users

Table 4. Occupations of Ventilator Users with Neuromusculoskeletal Conditions

Accounting/banking	28
Social work/counseling	27
Business owners	21
Teachers	18
Engineers/scientists	13
Business/corporation executives/administration	12
Journalism/freelance writers	10
Computer work (programming, systems analysis)	10
Lawyers	10
College professors	8
Artists (including mouthstick)	8
Insurance sales	6
Investment brokers and analysts	4
Real estate brokers	4
Physicians	4
Architects	2
College administrators	2
Mail order sales	2
Dispatchers (police, trucking)	2
Speech-language pathologist	2
Clergy	1
Receptionist	1
Librarian	1
Travel agent	1
Not specified	1
	42

reported being homemakers. In another report of employment profiles in NMDs, 40% of 154 people with progressive NMDs, including SMA and FSH, Becker, limb-girdle, and myotonic muscular dystrophies, were employed in the competitive market at the time of the study; 50% had been employed in the past; and only 10% had never been employed. The major barrier to employment was education. Intellectual capacity, psychosocial adjustment, and the belief by most people that their physical disability was the only or major barrier to obtaining a job were also identified. Physical impairment and disability were not associated with level of employment. Physical impairment and muscular dystrophy diagnostic groups had significantly lower employment rates, lower educational levels, and fewer employed professional, management, and technical workers.¹⁰⁹⁸ However, a recent study of 1000 Dutch patients with myotonic dystrophy, hereditary motor and sensory neuropathy, SMA, and myasthenia gravis noted that the majority were employed. Employment levels decreased after age 34, but the availability of work adaptations made it possible to prolong employment. Limiting factors were less effective communication, limited cognitive function, and the effect of weakness on

facial expression. ~~1099~~ ¹⁰⁹⁹ ~~It is~~ likely that, with the influence of the Internet, employment rates for ventilator users will increase.

Life Satisfaction

Six hundred fifteen of the 621 ventilator users who responded to a 1-7 Likert scale for overall life satisfaction had a mean response of 5.1. The 586 respondents whose methods of ventilatory support were known had a mean age of 49.4 ± 14 years (range = 16-84 years) and reported a mean life satisfaction index of 4.98 ± 1.68 . In addition, 242 nondisabled health care professionals with an average age of 33.0 ± 8 years (range = 21-59 years) reported scores of 5.33 ± 1.2 for satisfaction with their own lives, with no one reporting a score of 1. This score was significantly higher than the mean 4.98 score of the ventilator users ($p < 0.005$). When asked how ADL-dependent ventilator users would respond to this question, the mean estimate of health care professionals was 2.42 ± 1.37 . This score was significantly worse than the ventilator users' actual responses ($p < 0.0001$).

Differences arose between respondents using noninvasive ventilatory aids and respondents using tracheostomy IPPV. Both groups were compared for level of upper extremity function in the following manner: 0, for no upper extremity function; 1, for sufficient finger movement to operate a motorized wheelchair; 2, for adequate function to feed oneself; and 3, for normal or near-normal function. The ages, levels of function, years of ventilator use, hours per day of use, and life satisfaction index for the tracheostomy and the noninvasive groups are listed in Table 3. The noninvasive group was older than the tracheostomy group (50 vs. 45.8 years; $p < 0.001$), had significantly less upper extremity function (1.13 vs. 1.21; $p < 0.05$), and had used ventilatory support for fewer hours per day (15.5 vs. 17.7; $p < 0.05$) but for more years (22 vs. 17; $p < 0.005$). However, the tracheostomy IPPV group had a mean satisfaction index of 4.68 as opposed to 5.04 for the noninvasive group ($p < 0.05$). These figures are compared for various diagnostic subgroups in Table 1.

Table 3. Characteristics of Ventilator Users

Variable	Mean \pm Standard Deviation
Using tracheostomy IPPV (n = 92)	
Age	45.8 \pm 15.33
Function	1.21 \pm 0.97
Ventilator use (yr)	17.05 \pm 14.74
Ventilator use (hr/day)	17.65 \pm 6.81
Satisfaction index	4.64 \pm 1.85
Using noninvasive methods of ventilatory support (n = 494)	
Age	50.03 \pm 13.69
Function	1.13 \pm 0.97
Ventilator use (yr)	22.01 \pm 14.29
Ventilator use (hr/day)	15.47 \pm 8.95
Satisfaction index	5.03 \pm 1.64

None of the variables studied were significantly different at the 95% confidence level for any of the diagnostic groups except for the post-polio myelitis ventilator users. For this subgroup, age, years of ventilator use, hours per day of use, and level of function were not significantly different for the tracheostomy and noninvasive groups. However, the life satisfaction index for the noninvasive group was significantly greater than that of the tracheostomy group (5.20 vs. 4.55, $p < 0.02$).

Quality of Life and Satisfaction Domains

Dios, que es proveedor de todas las cosas, no nos faltara. No les falta a los mosquitos del aire, ni a los gusanillos de la tierra, ni a los remacuajos del agua. Es tan piadoso que hace salir su sol sobre los buenos y los malos, y llueve sobre los injustos y justos.

Don Quijote de La Mancha

The mean score of the overall life satisfaction in Campbell's survey of 2134 random subjects responding to a 1-7 Likert scale was 5.54 compared with 5.36 for our 273 health care professionals. Although the 5.1 mean score of the ventilator users was significantly less than the scores of the physically able population, it was still very positive (> 4.0) and significantly greater than the 2.42 anticipated by the health care professionals. In addition to the overall life satisfaction item, 380 ventilator users with post-polio myelitis syndrome,¹¹⁰⁰ 60 ventilator users with DMD,¹¹⁰¹ and 273 health care professionals (controls) were asked to respond to various Life Domain Satisfaction Measures and to 1-7 Semantic Differential Scales of General Affect, with 7 being the most positive response.¹¹⁰² The controls were also asked how they thought ventilator users with little or no extremity function would respond. A summary of the responses is presented in Tables 4 and 5.

Ninety-three percent of Campbell's 2134 subjects, 91% of our 273 health care professional controls, 85% of ventilator users with post-polio syndrome, and 87.5% of ventilator users with DMD reported being satisfied with their lives (response of 4 or greater). The ventilator users with post-polio syndrome, DMD, and spinal cord injury were significantly less satisfied with their transportation, education, health, social lives, sexual lives, and life in general than were the controls. They were significantly more satisfied with their housing. There were no significant differences in satisfaction with family life and employment. Except for health (post-polio ventilator users) and sexual activity (DMD ventilator users), the ventilator users were generally satisfied in each domain and reported means greater than 4.0.

The controls felt that their lives were significantly easier, more worthwhile, fuller, more hopeful, freer, and more rewarding than the ventilator users. However, the controls significantly misjudged that the ventilator users would give negative responses for each semantic differential (see Table 5). In fact, the ventilator users' mean responses were greater than 4 for each differential, except "hard—easy" (polio myelitis and DMD ventilator users), and "tied down—free" (DMD ventilator users). The post-polio ventilator users even judged their lives to be more interesting and friendly than did the controls, although the differences did not reach statistical significance. There was no significant difference in the "miserable—enjoyable" differential between the two groups.

Ventilator Users	Post-polio		DMD		SCI	
	n	Mean ± SD	n	Mean ± SD	n	Mean ± SD
Housing	386	5.7 ± 1.7	78	5.6 ± 1.4	42	5.6 ± 2.1
Transportation	351	5.3 ± 2.1	77	4.7 ± 2.0	41	4.5 ± 2.3
Education	388	5.2 ± 1.9	82	5.2 ± 1.5	42	4.6 ± 2.1
Job	216	5.2 ± 1.9	29	4.6 ± 1.7	15	5.2 ± 2.2
Health	384	3.9 ± 1.9	82	3.8 ± 2.0	42	4.1 ± 2.1
Family life	364	5.6 ± 1.8	77	5.6 ± 1.7	41	5.7 ± 1.7
Social life	360	5.8 ± 1.8	75	4.4 ± 1.9	41	4.5 ± 2.0
Sexual life	324	4.1 ± 2.2	51	3.6 ± 2.3	36	3.1 ± 2.2
Life in general	380	5.1 ± 1.7	80	4.9 ± 1.3	42	4.4 ± 1.8
Life in general†	273	2.5 ± 1.7				

Controls

	Controls		SCI Controls	
	n	Mean ± SD	n	Mean ± SD
Housing	263	5.2 ± 1.5	47	5.0 ± 1.5
Transportation	268	5.7 ± 1.6	47	4.6 ± 2.0
Education	266	5.5 ± 0.1	46	4.8 ± 1.8
Job	269	5.2 ± 1.4	33	3.5 ± 2.0
Health	269	5.7 ± 1.2	47	3.9 ± 1.7
Family life	268	5.6 ± 1.4	47	4.9 ± 1.8
Social life	268	5.4 ± 1.4	47	4.4 ± 1.9
Sexual life	227	5.5 ± 1.5	46	3.2 ± 2.0
Life in general	259	5.4 ± 1.2	47	4.1 ± 1.7

DMD = Duchenne muscular dystrophy, SCI = spinal cord injury, SD = standard deviation.

* The ventilator users were asked to rate their satisfaction with the dimension under question from 1 to 7, where 1 indicates extreme dissatisfaction and 7 indicates extreme satisfaction. The controls were autonomously breathing.

† Controls' responses assessing the ventilator users' satisfaction with life.

Social Integration

Compter...

Apprendre à compter sur soi-même

A compter pour ceux qui vous aiment

Pour faire aussi partie du nombre

Pouvoir enfin sortir de l'ombre

Apprendre by Yves Duteil

Home mechanical ventilation has proved to be safe and to optimize quality of life. A total of 307 ventilator users, or about one-half of the 621 in our study, maintained active and productive lives in their communities, as seen by their social and occupational

Il ne suffit pas de lire que les sables des plages sont doux; je veux que mes pieds nus le sentent. It does not suffice to read that the sands of the beaches are smooth, I want to feel them with my bare feet.

André Gide, *Les Nourritures Terrestres*, 1897, ed. Gallimard.

Assessment Issues

Campbell et al. recognized the difficulty in dealing with subjective perceptions of well-being in that reports of "excellent," "good," or "poor" overall quality of life may have different meanings for different people. He concluded, nonetheless, that these subjective parameters yielded consistent results when compared between different populations and that they are essential for assessing individual personal values and for self-assessment of quality of life.¹¹⁰³ He found that the single-item measure of overall life satisfaction closely fit the measures of satisfaction with the specific domains of life, and as such, it was one of the most important measures. Kammann et al. also demonstrated that the items with the highest validities were those which had a global frame of reference, such as feeling that life is going well in general.¹¹⁰⁴ Not one subject who expressed satisfaction with life in general was dissatisfied with his or her family life. The strongest association for all ventilator user groups was between general life satisfaction and satisfaction with social life.

Campbell noted that different life domains have different importance to different people. The downgrading of importance of any particular life domain can be explained by denial or adaptation to situations.¹¹⁰² Likewise, it appears that family life and housing issues—domains less affected by physical disability—take on the greatest significance for ventilator users. Campbell found that satisfaction with family life was one of the most effective predictors of general life satisfaction in his study population and correlated even more significantly with general life satisfaction for ventilator users in his study.

More recently it has become evident that there are more relevant ways of assessing the impact of disability, including disability associated with ventilator use, on the quality of a person's life. The impact of disability on the activities or domains most important to individual patients can be determined by asking patients to list favorite activities in order of importance to them and then to consider the impact of the disability on each one.^{1105,1106}

Physicians' Consideration of Quality of Life

Physicians' perceptions of the patients' quality of life are extremely variable.¹¹⁰⁷ Surveys have demonstrated the extent to which health care professionals underestimate the satisfaction with life of severely disabled, ventilator-assisted people. This point is important because physicians' assessment of patients' quality of life and the relative desirability of certain types of existence determine the likelihood that patients will receive therapeutic interventions.^{1108,1109} Physicians consider patients' quality of life more often to support decisions to withhold therapy than to support decisions to use mechanical ventilation ($p < 0.01$).¹¹⁰⁷ This situation is further revealed by the fact that despite the widespread use of nocturnal bilevel PAP for patients with ALS, less than 10% of patients with ALS ever use ventilatory support, and virtually none are offered the respiratory muscle aids that could spare them morbidity and mortality.¹⁰⁰ Some neurologists openly profess their "therapeutic nihilism."⁷⁶⁴

Psychometric Properties of the Ventilator Users' Quality of Life Scale of General Affect*

Ventilator Users	Post-Polio		DMD		SCI	
	368 > 360	Mean ± SD	256 > 249	Mean ± SD	41 > 39	Mean ± SD
Number of respondents	368		256		41	
Responses to each item	> 360		> 249		> 39	
Boring—Interesting	5.6 ± 1.6	4.5 ± 1.6	4.5 ± 1.6	4.7 ± 2.0	4.7 ± 2.0	
Miserable—Enjoyable	5.6 ± 1.5	4.9 ± 1.5	4.9 ± 1.5	4.9 ± 1.9	4.9 ± 1.9	
Hard—Easy	3.8 ± 1.8	2.9 ± 1.7	2.9 ± 1.7	3.2 ± 1.8	3.2 ± 1.8	
Useless—Worthwhile	5.9 ± 1.7	5.0 ± 1.9	5.0 ± 1.9	5.0 ± 2.1	5.0 ± 2.1	
Lonely—Friendly	5.9 ± 1.7	5.3 ± 1.7	5.3 ± 1.7	5.1 ± 2.3	5.1 ± 2.3	
Empty—Full	5.5 ± 1.6	5.0 ± 1.5	5.0 ± 1.5	4.8 ± 1.8	4.8 ± 1.8	
Discouraging—Hopeful	5.4 ± 1.8	4.9 ± 1.5	4.9 ± 1.5	4.9 ± 2.1	4.9 ± 2.1	
Tied down—Free	4.0 ± 2.0	3.5 ± 2.1	3.5 ± 2.1	3.2 ± 2.1	3.2 ± 2.1	
Disappointing—Rewarding	5.3 ± 1.8	4.5 ± 1.4	4.5 ± 1.4	4.1 ± 2.3	4.1 ± 2.3	
Number of respondents	256		46		239	
Responses to each item	> 249		> 43		> 232	
Boring—Interesting	5.4 ± 1.5	4.7 ± 2.2	4.7 ± 2.2	2.4 ± 1.5	2.4 ± 1.5	
Miserable—Enjoyable	5.7 ± 1.2	4.5 ± 1.6	4.5 ± 1.6	2.4 ± 1.4	2.4 ± 1.4	
Hard—Easy	4.1 ± 1.5	3.4 ± 1.7	3.4 ± 1.7	1.8 ± 1.1	1.8 ± 1.1	
Useless—Worthwhile	6.2 ± 1.1	5.0 ± 2.0	5.0 ± 2.0	2.8 ± 1.7	2.8 ± 1.7	
Lonely—Friendly	5.8 ± 1.4	4.9 ± 1.9	4.9 ± 1.9	3.0 ± 1.9	3.0 ± 1.9	
Empty—Full	5.8 ± 1.2	4.6 ± 1.8	4.6 ± 1.8	2.7 ± 1.4	2.7 ± 1.4	
Discouraging—Hopeful	5.9 ± 1.2	4.8 ± 1.8	4.8 ± 1.8	2.8 ± 1.7	2.8 ± 1.7	
Tied down—Free	5.0 ± 1.6	3.8 ± 2.0	3.8 ± 2.0	1.8 ± 1.2	1.8 ± 1.2	
Disappointing—Rewarding	5.8 ± 1.1	4.5 ± 1.8	4.5 ± 1.8	2.4 ± 1.5	2.4 ± 1.5	

DMD = Duchenne muscular dystrophy, SCI = spinal cord injury, SD = standard deviation.

* The subjects were asked to indicate the extent that each heuristic dimension describes them by indicating a number from 1 to 7, where 1 and 7 reflect the extremes of the polar adjective pairs in a seven-point Likert-type scale.

† The controls were 256 health care professionals, who provided at least 250 responses to each item, and 46 autonomously breathing patients after spinal cord injury, who provided at least 44 responses to each item.

‡ Controls' estimates of the ventilator users' responses.

activities. The male/female ratio was equal. Since other activities such as visits with friends, going to restaurants, sporting events, or other spectacles were not surveyed, it is clear that over one-half of this population maintains a considerable degree of mobility despite severe disability and ventilatory dependence. Technologic advances in environmental control systems, personal computers, and robot aids⁷⁵² as well as in ventilators and home health care delivery have also greatly facilitated a more active and higher-quality lifestyle.

Why Life Satisfaction in Ventilator Users?

Happiness = reality + expectations

At first it is surprising that people with such severe disability might be satisfied with their lives. However, habituation tends to produce a decline in the subjective pleasantness or unpleasantness of any input.¹¹⁰⁴ Campbell stated that "where an [unpleasant] situation is fixed for a person over a long term, there may be a tendency toward accommodation to it, reflected in gradual increases in satisfaction."¹¹⁰² Perhaps for ventilator users with ever-greater disability and decreasing expectations, habituation occurs along with maturity and acceptance, thereby decreasing the unpleasantness of the circumstances. Constricted horizons may also lead to satisfaction with the status quo.¹¹⁰² In addition, the ventilator serves as a daily reminder of the tenuousness of human existence. Unless the person uses nontracheostomy aids and is capable of GPP,⁹¹⁷ the ventilator is all that stands between the user and death. In a concrete way these people appear to appreciate that their quality of life is closely tied to their family lives and personal relationships, and their use of a ventilator takes on a positive aspect in permitting continued appreciation of human ties. Campbell described other populations of people with limited horizons who, although severely oppressed by society, remained satisfied until their status in society improved. With less limited horizons, they became aware that more could be obtained, and with new frustration came discontent. Thus, such severe disability may lead to a general scaling down of expectations and shifts in the importance of life domains. In addition, ventilator users may come to experience life satisfaction as a consequence of cognitive dissonance. They overcame the greatest of obstacles and challenges simply to be alive; therefore, life must be meaningful and satisfying.

Whiteneck studied the psychosocial outcomes of people with spinal cord injury at least 20 years after the injury occurred.¹¹¹⁰ Three-fourths of the subjects rated their current quality of life as good or excellent on a 5-point scale. There were no significant differences by level of injury, but satisfaction correlated inversely with age. Whiteneck et al. also demonstrated that ventilator users with spinal cord injury rated their quality of life higher than autonomously breathing traumatic tetraplegics. They reported that fewer of the former than of the latter considered suicide at least once and six times or more and that more of the former were happy to be alive and had greater self-esteem.¹¹¹¹ Our survey also demonstrated that, while ventilator users with spinal cord injury were dissatisfied only with sexual function, autonomously breathing traumatic tetraplegics were dissatisfied (mean responses below 4.0) with the domains of job satisfaction, health, and sexual functioning.¹¹¹² The ventilator users with spinal cord injury reported a significantly greater satisfaction with housing, job, and family life and a greater satisfaction with social life, health, and life overall in comparison with the autonomously breathing traumatic tetraplegics. They also felt that life was somewhat friendlier, more interesting, more enjoyable, fuller, and more hopeful than the latter group. This level of psychosocial adjustment and well-being is remarkable, considering that, in addition to ventilator use, people with spinal cord injury also had less upper extremity function than autonomously breathing tetraplegics. Thus, ventilator use had to be the difference in their more positive perceptions of life. The level of disability may not be as important a factor as aging in determining overall long-term life satisfaction in ventilator users.¹¹¹³

The more positive well-being scores of patients with polio and DMD compared with the scores of ventilator users with ALS may also be explained by the fact that the former were initially managed by noninvasive methods of long-term ventilatory assistance and their ventilator use requirement usually increased gradually. Thus, the patients with polio and DMD generally had more time to adjust to both physical disability and ventilator use than did ventilator users with ALS and other motor neuron diseases (MNDs).

Other studies have confirmed the positive view of life held by the great majority of ventilator users with NMD.¹¹¹⁴ One study reported that 92% of ventilator users with DMD "had positive feelings of life" more than half of the time.¹¹¹⁵ In one study of 19 patients with ALS/MND, over two-thirds were satisfied with their lives; 84% felt that they had made the right choice; and 18 felt that ventilator-assisted breathing had been a worthwhile option and would choose it again. In another study of 92 long-term tracheostomy IPPV users with ALS, 20 lived 8 to 17 years using IPPV and 9 became locked in. Although most wanted to live as long as possible, 14 eventually requested that mechanical ventilation be withdrawn.¹¹¹⁶ In the study by Ganzini et al. of 100 patients with ALS, 84% said they would not consider physician-assisted suicide and looked "forward to the future with hope and enthusiasm."¹¹¹⁷ Although still positive, the views of patients with ALS/MND in these studies were less positive toward assisted ventilation and life satisfaction than were those of patients with DMD.

In a recent study of sexuality in 200 patients who used noninvasive IPPV for a mean of 41 ± 27 months, 34.5% reported that they were sexually active. For 46% "nothing changed" after beginning noninvasive IPPV; 36% were less active; and 13% were more active. The sexually active noninvasive IPPV users reported having intercourse 5.4 ± 4.8 times per month.¹¹¹⁸ The rocking bed ventilator has been used by some to facilitate sexual intercourse (and to conserve energy during intercourse) as well as for ventilatory assistance. The rocking bed also has beneficial effects on alleviating constipation.

Ventilator Style Preference: Tracheostomy vs. Noninvasive Approaches

A survey of a subset of the 695 ventilator users—the 170 with greater than 1 month experience in the use of both tracheostomy and noninvasive methods—was undertaken to compare ventilator use preferences.⁶⁸⁹ Participants had the following diagnoses: post-polio myelitis, 129; non-Duchenne myopathies, 14; spinal cord injury, 13; severe kyphoscoliosis, 3; chronic obstructive pulmonary disease, 3; DMD, 2; Guillain-Barré syndrome, 2; and myasthenia gravis, multiple sclerosis, ALS/MND, and polymyositis, 1 each. This group had a mean age of 54.7 ± 11.4 years, was using ventilatory support for 17.1 ± 6.5 hours per day, and had been ventilator users for 22.7 ± 13.1 consecutive years. Of the 170, 155 required both nocturnal and at least some daytime ventilatory aid. Over 155 had required 24-hour support for some period. The group was divided into two subgroups: the 111 noninvasive ventilation users who were switched to tracheostomy IPPV (group 1) and the 59 ventilator users who were switched from tracheostomy IPPV to noninvasive aids (group 2). There were no significant differences between the groups in age ($p = 0.22$), hours per day of ventilator use ($p = 0.11$), or years of ventilator use ($p = 0.10$). Group 1 respondents used noninvasive aids for 13.7 ± 11.5 years before being switched to tracheostomy IPPV, which they used for another 10.5 ± 10.3 years. Group 2 respondents

used tracheostomy IPPV for 1.6 ± 4.8 years before switching to noninvasive ventilatory aids for another 18.8 ± 14.8 years.

Of the 170 patients who used noninvasive ventilation, 84 used a body ventilator for a mean of 14.3 ± 14.1 years overnight and 10.7 ± 14.1 years during waking hours; 24 used noninvasive IPPV for a mean of 9 ± 8.2 years for nocturnal support and 4.5 ± 7.5 years during waking hours; and 62 used both body ventilators and noninvasive IPPV methods for a mean of 21.7 ± 10.0 years overnight and 21.7 ± 12.9 years during waking hours. Figures 1-3 present the respondents' preferences for the quality-of-life issues included in the survey. Table 6 demonstrates concordance of the principal caregivers' preferences with those of the ventilator users. Other reasons cited for preferring tracheostomy or noninvasive aids are listed in Table 7.

In addition, the ventilator users reported that during use of indwelling tracheostomy tubes they required tracheal suctioning a mean of 7.6 ± 8.3 times per day; 27 patients (16%) reporting 10 or more times per day. Thirteen of the latter group cited numerous respiratory complications before switching to as much as 24-hour use of noninvasive aids. Thirty-five percent of group 1 ventilator IPPV users expressed the desire to return to noninvasive aids, whereas none of the group 2 ventilator users wished to switch back to

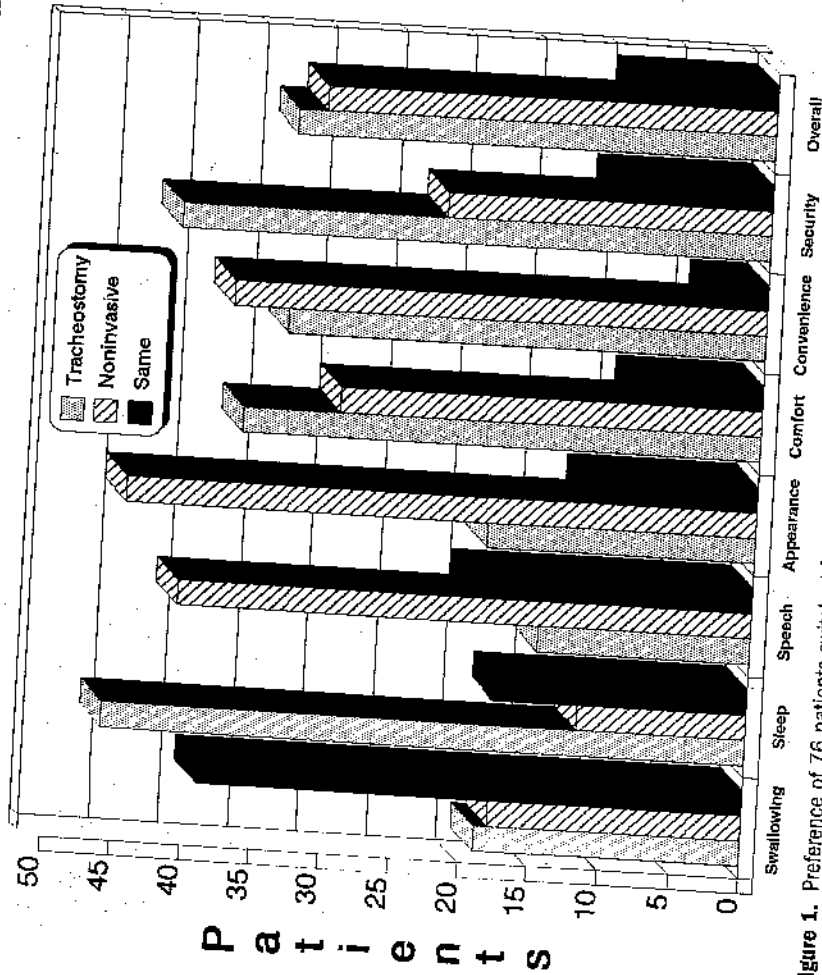


Figure 1. Preference of 76 patients switched from body ventilator use to tracheostomy IPPV (group 1). (From Bach JR: A comparison of long-term ventilatory support alternatives from the perspective of the patient and care giver. Chest 1993;104:1702-06; with permission.)

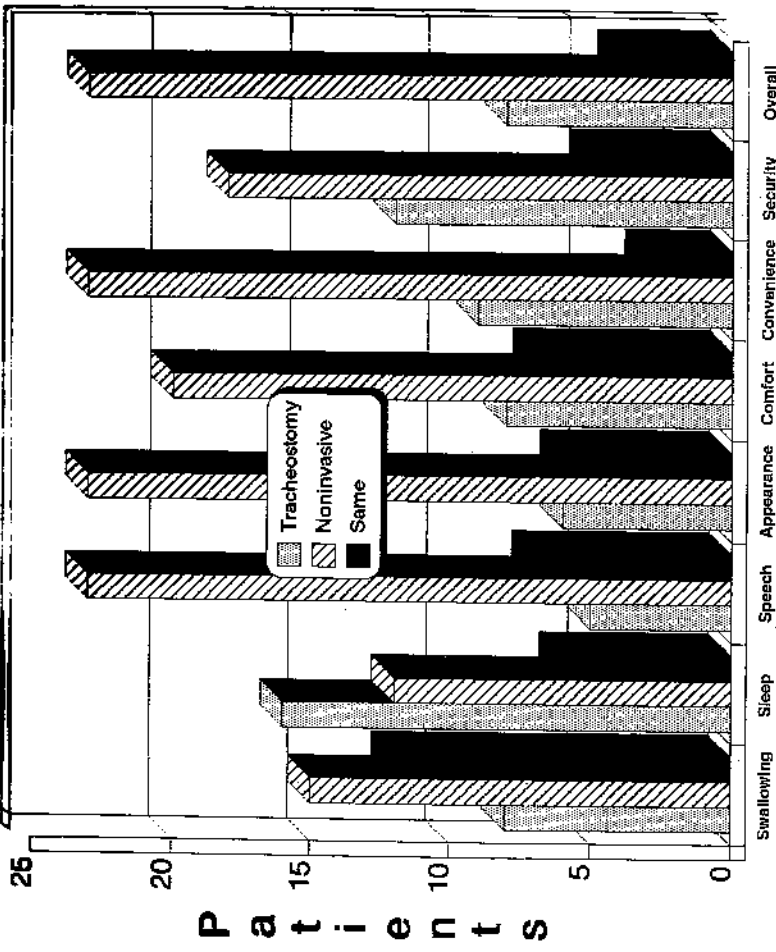


Figure 2. Preferences of 35 patients switched from a regimen of body ventilators and/or noninvasive IPPV to tracheostomy (group 1). (From Bach JR: A comparison of long-term ventilatory support alternatives from the perspective of the patient and care giver. Chest 1993;104:1702-06; with permission.)

tracheostomy IPPV. Two of the ventilator users in group 1 (2%) and 8 in group 2 (1.4%) had regular access to MI-E during respiratory infections. Twenty-eight of the ventilator users in group 1 (25%) and 22 in group 2 (37%) had mastered GPB sufficiently to achieve or increase ventilator-free breathing tolerance while they were using noninvasive ventilation.

In miscellaneous survey questions, every ventilator user without exception indicated that ventilatory assistance should be offered to every person who could benefit from it; that finances should not be a consideration; and, if having to do it over, each ventilator user would have made the same choice.

Respiratory Complications

The 621 ventilator users were surveyed for the number of times that they were hospitalized for respiratory distress after an initial respiratory hospitalization under the following circumstances: (1) before use of ventilators on an ongoing daily basis, (2) during use of ongoing oxygen therapy, (3) during use of noninvasive aids predominantly overnight, (4) during use of noninvasive aids greater than 16 hours per day, and (5)