

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 remacuños 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.)

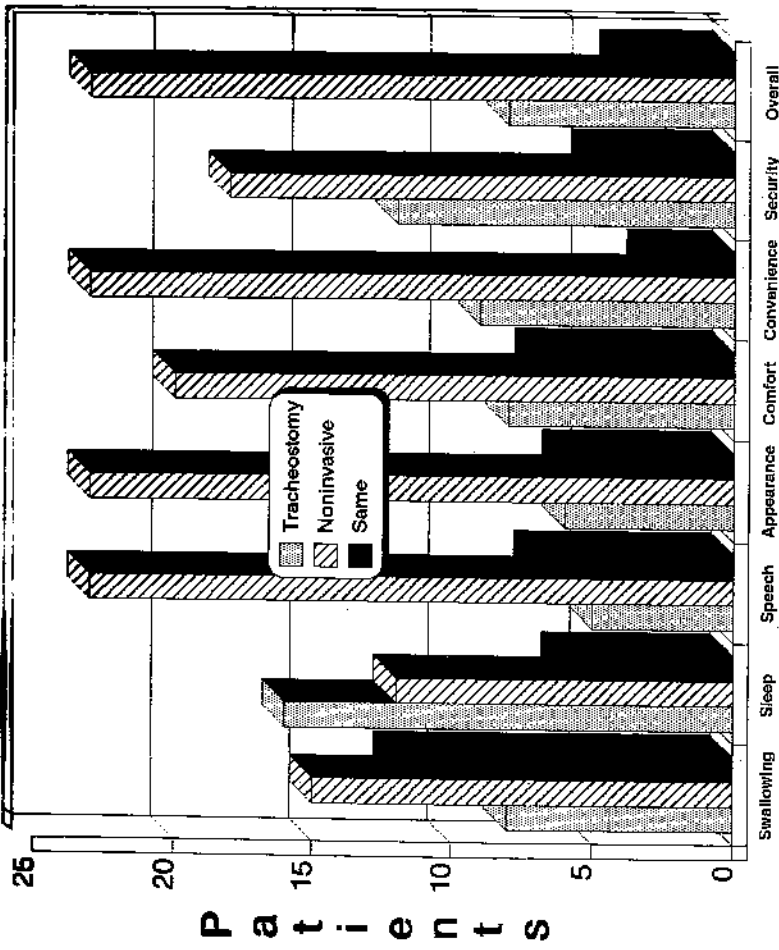


Figure 2. Preferences of 35 patients switched from a regimen of body ventilators and/or noninvasive IPPV to tracheostomy (group 1). (From Bach JR: A comparison of long-term ventilatory support alternatives from the perspective of the patient and care giver. Chest 1993;104:1702-06; with permission.)

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)

For preferring tracheostomy

- Facility in clearing airway secretions during respiratory tract infections: 57 (31%)
- Greater mobility by comparison with body ventilator use: 8 (9%)
- Better speech than by using mouth IPPV: 1 (1%)

For preferring noninvasive aids

- Greater independence and control of breathing: 24 (14%)
- Facilitation of management in the community: 17 (10%)
- Suctioning no longer needed/absence of secretions: 17 (10%)
- Greater portability (less equipment, supply needs, and upkeep): 14 (8%)
- Greater mobility: 11 (7%)
- Fewer infections: 11 (7%)
- More natural: 9 (5%)
- Compatibility with glossopharyngeal breathing/air-stacking: 3 (2%)

noninvasive aid users.¹⁴ The fact that the lowest incidence was in the noninvasive aid users with the greatest need for ventilatory support can be explained by the following factors: noninvasive aid users avoid the complications associated with indwelling tracheostomy tubes; nighttime-only ventilator users are often underventilated during daytime hours and, when not using a ventilator, may not increase insufflation volumes for optimal elimination of airway secretions, as needed; and nighttime-only ventilator users are less likely to have been introduced to MI-E.

In addition to the fact that noninvasive IPPV is associated with fewer respiratory complications and pneumonias for long-term ventilator users, a prospective epidemiologic survey from a single ICU recently demonstrated that noninvasive IPPV is also associated with a significantly lower incidence of ventilator-associated pneumonias compared with tracheal intubation in the acute setting.¹⁰⁷⁰ The groupings taken into account in this study were patients going from noninvasive IPPV to IPPV via tracheal intubation, patients going from tracheal intubation to noninvasive IPPV, users of IPPV via tracheal intubation only, and users of noninvasive IPPV only.

Tracheostomy IPPV vs. Noninvasive Physical Medicine Alternatives

Whether switched to or from noninvasive ventilatory support regimens that include noninvasive IPPV, the great majority of ventilatory users considered noninvasive methods to be superior to tracheostomy IPPV for every item on the survey, including safety, convenience, swallowing, speech, appearance, comfort, and overall satisfaction.¹⁴ Both ventilator users and their principal caregivers unanimously preferred the noninvasive methods overall to tracheostomy IPPV. Except for sleep, those switched from a noninvasive aid regimen, including noninvasive IPPV, to tracheostomy also preferred the noninvasive regimen for every item evaluated.

There are many reasons that patients prefer noninvasive aid and that noninvasive aid users express greater life satisfaction than tracheostomy IPPV users. Tracheostomy IPPV

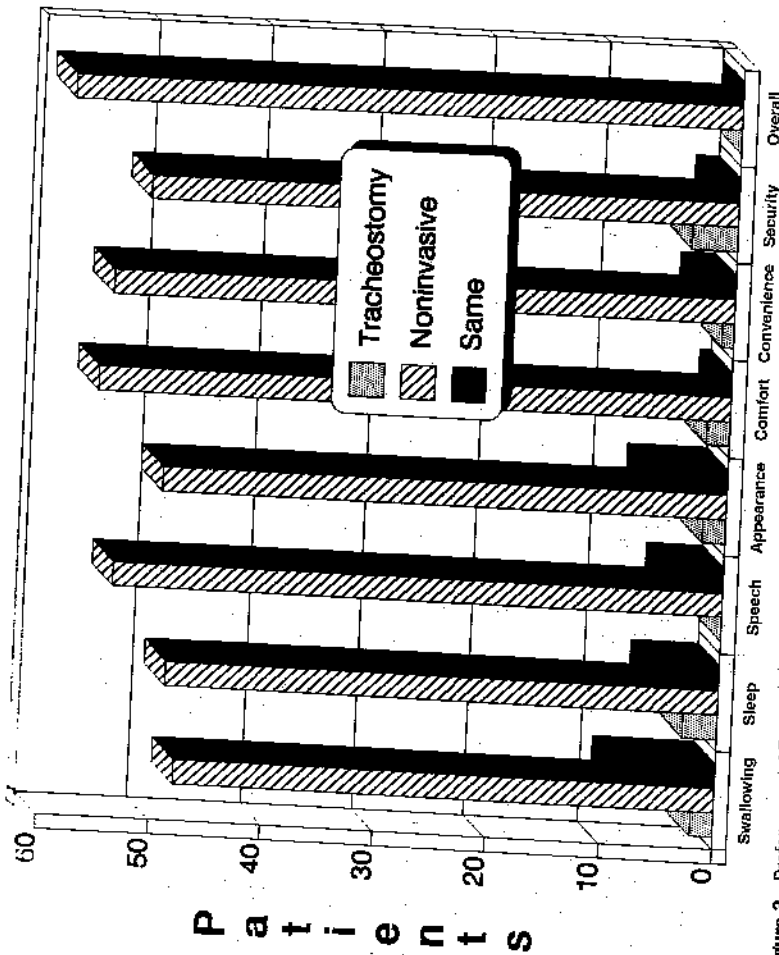


Figure 3. Preferences of 59 patients switched from tracheostomy to noninvasive aids (group 2), including 8 who switched to body ventilators, 18 to noninvasive IPPV methods, and 33 to a combination of body ventilators and noninvasive IPPV methods. (From Bach JR: A comparison of long-term ventilatory support alternatives from the perspective of the patient and care giver. *Chest* 1993;104:1702-06; with permission.)

during use of indwelling tracheostomy tubes and daily IPPV. The groups were controlled for level of extremity function. All comparisons were statistically significant. The highest incidence of hospitalizations for respiratory complications was in patients receiving oxygen therapy, followed by those using tracheostomy IPPV and those using noninvasive aids predominantly overnight. The lowest incidence was for the full-time

	Patient and Principal Caregiver Ventilator Method Preferences	
	Noninvasive	No Preference
Group 1		
Patients	55	42
Caregivers	50	38
Group 2		
Patients	59	0
Caregivers	48	1

users with little breathing tolerance are fearful of sudden apnea resulting from tracheostomy disconnection or ventilator failure. Unlike noninvasive ventilation, IPPV via tracheostomy tends to cause continuous ventilator dependence (Chapter 8). Tracheostomy IPPV users also require ongoing tracheostomy care, including tracheal suctioning and regular uncomfortable tube changes. Sudden detachment of mucus plugs from the tube or cuff can cause acute respiratory distress. It is more difficult to obtain attendant care in the home for tracheostomized patients. In addition, tracheostomy IPPV users cannot use the sense of smell to taste their food and require supplemental humidification because air does not pass through the nasal passages and upper airway. They often complain of unpleasant taste sensations, which appear to be associated with chronic pathogenic bacterial colonization of the tracheostomy site and upper airways.

Patients using noninvasive aids, on the other hand, maintain more normal speech, do not require deep airway suctioning, and usually require assisted elimination of airway secretions only during respiratory tract infections. The fear of tracheostomy disconnection and sudden ventilator failure is eliminated by decannulation, use of noninvasive IPPV, and mastery of GPB (Chapter 10). Physiologic use of the upper airway permits the noninvasive ventilator user to maintain normal airway humidification and olfactory and taste sensations. All of these factors facilitate self-confidence and more active community living and may have contributed to the noninvasive aid users' greater life satisfaction.

Patients who were switched from body ventilator use-only to tracheostomy IPPV preferred the former for appearance, convenience, and speech and the latter for comfort, security, swallowing, sleep, and overall satisfaction. Most of these ventilator users were never introduced to noninvasive IPPV methods. Nocturnal mouthpiece IPPV with a lipseal was used almost exclusively in one center,⁷³² and nasal IPPV was not comprehensively described until 1987, long after most of these ventilator users had been switched to tracheostomy.⁶⁷⁴ Their general preference for tracheostomy IPPV was understandable because body ventilators are less effective and more restrictive than IPPV; airway secretion clearance can be impossible without use of noninvasive IPPV and MI-E; and, except for the chest shell ventilator and LAPV, body ventilators must be used while the patient is reclining.²⁹⁵ Of the ventilator users who "preferred" tracheostomy, none had mastered GPB and none had had access to MI-E. It is not surprising, therefore, that 31% of the ventilator users who preferred tracheostomy IPPV indicated that they did so because of the use of tracheal suctioning to clear airway secretions during respiratory tract infections. When more than nocturnal ventilatory support was required, resort to tracheostomy IPPV appeared to be desirable because noninvasive IPPV and MAC were unavailable.

The preferences for noninvasive ventilation are contrary to the views of physicians who think that IPPV via tracheostomy can facilitate voice production or swallowing or delivered volumes and a convenient portal for airway suctioning. On the contrary, as noted in Chapters 4 and 8, tracheostomy tubes tie down the strap muscles in the neck and impede swallowing.^{228,286,1119} Even when a cuffless tracheostomy tube or a deflated cuff is used, speech is of poorer quality than when noninvasive aids are used. Rhythm and voice tone are less natural than when the patient uses an LAPV with GPB or noninvasive IPPV. The use of GPB for deep breaths to increase voice volume, improve cough efficacy, and ensure safety in the event of ventilator failure is not possible during tracheostomy IPPV.⁷⁹² Indeed, a lower percentage of effective glossopharyngeal breathers

would be expected and was found in the group that was ultimately switched to tracheostomy IPPV. All of these factors have considerable bearing on quality of life.

Ethics

Beware of bringing the hospital into the home.

Beware of astronomical costs involved with ventilation or society will choose the more economical alternative: no ventilators.

Beware of underestimating the limitless potential of individuals with disability.

Independence for a ventilator user means that one can say, "So I use a ventilator, I am in charge of it. I direct my own life and take my own risks. I am the one who decides about my quality of life."¹¹²⁰

—Gini Laurie's Ventilator User Caveats

Purtilo¹¹²¹ observed that most discussion of ventilator use has focused on the critically ill patient maintained by tracheostomy IPPV in ICUs. She pointed out that this focus has "fostered misconceptions and stereotypes" about other appropriate uses of ventilators. There is, likewise, great potential for "misunderstanding of the ethical issues involved in treating patients whose chronic maintenance depends on either positive or negative long-term ventilator support."

Quality of life is difficult, if not impossible, to measure by objective criteria that can be applied to all people. Most efforts at measurement, however, appear to have been based on factors relevant for physically able people.¹¹²² Jonsen et al. noted that the important point is the subjective satisfaction experienced by a person in physical, mental, and social situations, even though these experiences may be deficient in some manner.¹¹²³ Thus, potential satisfaction with life and with the various aspects of existence should be considered in questioning the appropriateness of vital therapeutic options. Informed decisions about ethically and financially complex matters such as long-term ventilator use should be made by examining the life satisfaction of competent people who have already chosen these options. The great majority of severely disabled ventilator users with NMD are satisfied with their lives despite the inability to achieve many of the "usual" goals associated with quality of life in the physically able population. Their principal life satisfaction derives from their social relationships, reorganization of goals, and immediate environment.

In a study of 98 ventilator users, most of whom had neuromuscular weakness or chest wall disease, the impact of using a mechanical ventilator was found to be overwhelmingly positive (87%) in regard to sustaining life, facilitating mobility, improving physical symptoms, and overall satisfaction. Tracheostomy IPPV users volunteered significantly fewer positive statements than users of noninvasive ventilation. Whereas 53% of users indicated that they had initially experienced difficulties in coping with the ventilator, only 11% identified difficulties at the time of the survey.¹¹²⁴ Although the willingness of and hardship imposed on care providers and community resources need to be taken into consideration, it is unethical not to consider the life satisfaction of people who have already chosen to use mechanical ventilation and who are living with the consequences of their decisions when decisions about about long-term ventilator use are made for others.^{1108,1125} Eighty to 97% of tracheostomy continuous IPPV users with ALS are glad

to have chosen mechanical ventilation, are satisfied with their lives, and would use mechanical ventilation again if they had to do so.^{97,100,1126-1128} The longer that totally ADL-dependent patients with ALS/MND use continuous ventilatory support (from 1 to 15 years with only the ability to blink), the more life satisfaction they report.¹¹²⁸ Only about 8% ever seriously consider discontinuing ventilatory support.¹¹²⁶ Yet many physicians inappropriately judge the "lack of quality" of their patients' lives to justify withholding mechanical ventilation, even during episodes of acute respiratory failure. Indeed, less than 3% of patients with ALS undergo tracheotomy in the U.S.,⁷⁴⁶ less than 10% are offered the option, and death from respiratory complications is almost ubiquitous.¹⁰⁰

The right of the disabled person to live freely is often jeopardized by health care and societal issues. Zola described the "dehumanizing indignity in safety"¹¹²⁹: "Life entails risk and to try to create an environment without risk ultimately devalues the person with the disability by suggesting that he/she is not capable of coping except in the most restrictive and supportive environment."¹¹³⁰ What is often appropriate and necessary in the protective environment of the acute care setting is not appropriate in the rehabilitation setting and has led many patients to express "intense frustration at the double messages given by rehabilitation professionals: you must learn to be independent as a disabled person, but we will make all your decisions for you."¹¹³⁰

A common reason for constraining the actions of institutionalized patients is to avoid risk to them and liability to the institution. However, this controlling focus does not always change once the person returns home because the daily care of "patients" is often directed by licensed nurses contracted by professional nursing agencies. Thus, the activities of the severely disabled person continue to be circumscribed by the rules of institutional health care organizations that hamper the client's self-determination and individuality. The solution for this problem lies in the formation of client-maintained personal assistance services (PAS) and the provision of adequate PAS for the severely disabled, self-directed people who require them. In the words of Gill:

Our people are still being warehoused without hope of ever having a home or family or lifestyle of their own. When you think about it, in contrast to the vast numbers of us delicately sought death. Yet... laws are being passed at record speed to ease the way to our demise. Prominent "experts" [argue] for our right to die... as defenders of our freedom, dignity, humane treatment, and even "independence" ... [yet it] has taken almost 20 years of exhausting struggle to get a basic civil rights law for disabled people.¹¹³¹

Although the Americans with Disabilities Act (ADA) is seen as an important step to prevent discrimination against disabled people, it does little or nothing for the self-directed disabled person who, like most patients with neuromuscular disorders, is not informed by physicians about potentially vital therapeutic options,¹¹⁰⁸ nor does it help those who are warehoused in institutions because of lack of a national PAS policy. The false beliefs that prohibitively expensive continuous nursing care is required and that disabled ventilator users are unable to take responsibility for and manage their own care are the greatest obstacles to returning them to the community. This position is ironic since the provision of PAS for the home care of ventilator users can greatly reduce cost¹⁰⁶⁷ as well as enhance quality of life.

One large group of ventilator users with respiratory failure was institutionalized for 24-31 years before obtaining release to enter the community in 1979. The ventilator users were authorized to develop a client-maintained organization, Concepts of Independence, Inc., that permits self-directed clients, themselves and others, to hire, train, direct, and dismiss their own personal care attendants.¹¹³² Virtually all of the current 1156 clients of Concepts of Independence, Inc. require continuous attendant care. This population includes 25 noninvasive and 11 tracheostomy IPPV users. The program has now successfully expanded throughout New York State, and there have been no significant accidents or litigation in its 23-year history. Indeed, the home health care industry has had virtually no history of tort litigation.^{1132,1133} The efforts of these courageous and tenacious ventilator users succeeded in establishing a precedent for humanizing the care of severely disabled, self-directed people in private domiciles and, in doing so, greatly increased the quality of their lives with 70% cost savings to the taxpayers of New York.¹⁰⁶⁷ As Kafka observed, "Without effective community options we lose our humanity. More and more people are choosing to die rather than exist in institutions... warehoused for 'cost' and 'efficiency.'"¹¹³⁴

The institutional control of chronic care, whether in an institution or in the community with PAS managed by nursing organizations, can impersonalize and dehumanize care in the name of safety. It reduces the client's sense of personal control and self-efficacy and suggests inadequacy in coping. Physicians who train their patients in how to manage and take responsibility for their care need to play a more active role in advocating for the procurement of the services needed to permit them to return to or remain in the community.

Typical counterproductive regulations include the prohibition of tracheal suctioning by unlicensed personal care attendants. In 46 states young children who are relatives of the user may perform tracheal suctioning, but third-party payors and home nursing care agencies will not sanction this activity by trained PAS. Such regulations too often serve to protect private interests under the pretext of avoiding liability and ensuring the patient's safety. These restrictions continue despite the fact that the American College of Chest Physicians¹¹³⁵ and others¹¹³⁶ support the use of properly trained PAS for self-directed ventilator users. So far, regulations have been modified to permit this practice in at least Colorado, Massachusetts, and New York.¹¹³⁷

We routinely close tracheostomy sites and convert ventilator users to noninvasive respiratory aids. Because this service is performed by very few institutions,³³² most ventilator users are never made aware of it, and many, without adequate family support, must remain tracheostomized and institutionalized. It is the ethical responsibility of the health care community to inform ventilator users about noninvasive alternatives to tracheostomy before considering advanced medical directives. Patients should be offered training in their use and decannulation to noninvasive supports, especially when these methods can facilitate community living with or without PAS. This approach benefits both the patient and the taxpayer. In the face of calls to limit entitlement spending, it should be noted that a society willing to provide free room, board, health care, legal and educational services, vocational training, and cable television for murderers, rapists, drug dealers, and other felons at a cost of billions of dollars, including \$30,000 per year per inmate just for interment, has the ethical responsibility to provide PAS to those in need, some of whom are crime victims themselves.¹⁰⁶⁷ Further information about the need for and status of PAS is available in various sources.¹¹³⁷⁻¹¹³⁹

Despite the consternation about growing medical costs, little is being done to limit costs and optimize care for users of mechanical ventilation. Instead of preventing the otherwise inevitable episodes of respiratory failure for patients with NMD by using the methods discussed in this book, patients are left to develop respiratory failure and undergo tracheotomy. However, the average length of hospital stay for patients with NMD when they undergo tracheotomy is 72.1 days, most of which are spent in intensive care; for 16.2 of these days the patient is intubated.⁶⁵⁵ The prolonged hospitalization is due to failed weaning and extubation attempts; in addition, the family is often ill prepared to care for a tracheostomy IPPV-dependent member and requires considerable time for acceptance, training, and home preparation. Most of these costly hospitalizations can be avoided with proper patient follow-up, education, training, and equipment.

In another example of waste of financial resources, the initial costs of placement and training in phrenic nerve pacing exceed \$300,000, and the technique has a realistic long-term success rate of only 33%.⁸⁸² Furthermore, patients using electrophrenic respiration (EPR) most often retain tracheostomy tubes, which create additional expenses for disposable suction catheters, tracheostomy tube changes, related stoma care, and possibly home nursing services. In contrast, removal of the tracheostomy tube, teaching of GPB to free the patient from vital ventilator dependence, and use of noninvasive ventilatory support alternatives are significantly less expensive. Mouthpieces cost \$3-27 each; nasal interfaces, \$30-200 each; and Exsufflation Belts, \$350 each. The \$300,000 initial cost of EPR often comes out of a limited insurance policy. This money often can be used with much better effect to provide ADL-enhancing equipment and personal care. Unfortunately, the profits from pacemaker placement and use and the intensive market- ing of this approach obscure the more reasonable but less immediately profitable alternative of using respiratory muscle aids. One still earns more money by placing tracheostomy tubes and pacemakers than by removing them.

With the growing emphasis on cost containment and reform of the health care delivery system in the United States, quality-of-life issues are being evoked to justify holding life-sustaining medical interventions, including mechanical ventilation.^{1107,1108} Although it would appear that an intelligent, self-directed person should be fully informed about therapeutic options and prognosis, in the frenzy of seeking a less expensive health care delivery system, some physicians have suggested eliminating the patient from the decision-making process. As recently as 1989 it was recommended that the physician's assessment of patients' quality of life should be "independent of the physician's feelings" in determining whether or not to institute mechanical ventilation.¹¹²⁵

Besides the fact that most physicians underestimate the life satisfaction of ventilator users with NMD, the great majority of physicians caring for such people are ignorant of noninvasive methods that offer prolonged survival. It should not be surprising, therefore, that many, if not most, physicians are biased against the long-term use of ventilatory assistance, which they wrongly associate with the need for an indwelling tracheostomy tube. This misconception was documented in a survey of NMD clinic directors.¹¹⁰⁸ Forty-one percent of 273 MDA clinic directors openly discouraged ventilator use, whereas only 30% ever recommended it. Fifty-five percent of the directors cited "poor quality of life" to justify their position. Only two physicians who discouraged ventilator use were familiar with noninvasive IPPV. Not surprisingly, the directors who most underestimated the ventilator users' life satisfaction were also the least likely to encourage

ventilator use and were unfamiliar with noninvasive respiratory aids.¹¹⁰⁸ More recent surveys of MDA clinic directors in the United States⁶¹⁵ and Canada¹¹⁴⁰ unfortunately demonstrated similar results. In the Canadian study, most physicians had a negative bias against assisted ventilation; 53% cited poor quality of life as the reason not to offer assisted ventilation; and 25% of them never even broached the subject of mechanical ventilation. A manuscript demonstrating a 2-year prolongation of survival by the use of strictly noninvasive methods for people with ALS/MND and no breathing tolerance was recently rejected by the *Journal of Neurology* when the reviewer stated that, in his opinion, "these patients did not need to use ventilators." Did his opinion reflect disbelief in the efficacy of the methods, the view that the patients were simply better off dead, or both? Other clinicians have frankly stated that "the use of chronic assisted ventilation should be avoided."¹¹⁴¹ Still others consider the long-term use of assisted ventilation for people with DMD to be "most controversial" and to raise "enormous ethical difficulties."¹¹⁴²

Indeed, some physicians appear to impose hardly any limits on their self-determined right to make life-and-death decisions for their patients. Ventilator use is a situation "in which the support system does not replace the diseased organ. The ventilator assists an organ system that is not primarily diseased. The model of renal dialysis does not apply, since the diseased organ is replaced by the function of the machine."⁷⁴⁰ This rationale has been used despite the fact that the cost of ventilator use is about 30% of the cost of renal dialysis and far less disruptive of the patient's day-to-day life.

In the words of Goldblatt, "Patients should be encouraged as much as possible to direct their own lives, but, if they are incapable of reaching an unequivocal and seemingly rational decision, they should be able to trust their doctor to act in their best interest...the physician will consult the family and will look as objectively as possible at the quality of the patient's life."¹¹⁴³ Negative physician attitudes make it imperative for patients to have sources other than their health care professionals to learn about therapeutic options. For many the Internet has provided the needed patient-networking capabilities. Health care providers must ethically avoid a paternalistic stance when working with the disabled. Self-directed people, once properly informed, should be treated as competent to make decisions about their own welfare. Paternalism undermines the goals of rehabilitation.

The Futility of Advanced Directives: When Does No Really Mean No?

Advanced directives are valid only if the patient is fully informed about treatment options and, in particular, the likely clinical course if they are not used. Patients with NMD are rarely properly informed because few clinicians are knowledgeable about noninvasive options. In part as a result of this problem, some clinicians argue that "the patient, his or her family, the physician, and other care providers may not be able to adhere to a planned decision to withhold [invasive] ventilatory support in the presence of impending respiratory failure, when the instinctive urge to preserve life supplants the rational conclusion that, in this particular instance, death is preferable to life under such profoundly altered conditions."¹¹⁴³

When asked early in the course of their disease, many patients with ALS/MND indicate that they would rather die than use respiratory support. In a recent study, 38% of 640 physicians also stated that they would prefer to end their lives rather than use ventilators for ALS.¹¹⁴⁴ Many people change their minds during episodes of acute respiratory

failure, however, particularly when dyspnea can be relieved by the simple use of noninvasive respiratory muscle aids. The patient's attitude toward the use of ventilatory aids more closely reflects the physician's attitude and the nature of the treatment options than an informed rational decision.¹¹⁰⁸ There were no differences in scores of depression, hopelessness, overall quality of life, or psychological well-being in patients with ALS/MND who required ventilatory support for 1-120 months and patients with ALS/MND who had less disability and did not require ventilator use.¹¹²⁷ It should be recognized that a decision for ventilatory support is probably the best predictor of an acceptable quality of life using a ventilator.¹¹²⁷

Few if any clinicians would consider the use of respiratory muscle aids to be "heroic" interventions. Yet, to the health care professional offering advance directives, heroic measures to be avoided include "mechanical ventilation," which to most clinicians means intubation and tracheostomy. The belief is that intubation will result in failure to wean from the ventilator; tracheostomy, and long-term dependence on tracheostomy IPPV. In reality, this belief is true only for patients with severe bulbar ALS/MND and a persistent decrease in baseline SpO₂ due to chronic aspiration.

Although most patients say that they would refuse tracheostomy, as VCs and CPF decrease, few if any refuse the occasional use of noninvasive respiratory muscle aids to relieve dyspnea or to facilitate airway secretion elimination. Some patients initially use mouthpiece IPPV only to increase voice volume. With time and further decreases in VC, the patient eventually uses noninvasive IPPV continuously. Once accustomed to it, the patient may be more likely to accept tracheostomy when it is warranted by decreasing SpO₂.

Because of avoidance of noninvasive aids, patients are unfamiliar with ventilator use when respiratory failure unexpectedly occurs. At that time, desperate, intubated, suboptimally managed, and poorly informed patients are given a tracheostomy ultimatum that they accept with fear and anger. Patients and families feel overwhelmed and inadequately counseled;⁷⁴⁰ they rarely understand the wide-reaching financial, social, and physical consequences of acquiescing to tracheostomy. To avoid this scenario, which is often incorrectly thought to be inevitable, some physicians prescribe euthanasia with supplemental oxygen and morphine.^{648,1145}

Should the patient's desire to live be ignored because "death is preferable to life under these conditions," or should morbidity be avoided by using noninvasive aids? Noninvasive aids can postpone or entirely obviate the "crisis" of deciding whether or not to "go on a ventilator" (to undergo tracheostomy). Furthermore, the use of noninvasive methods eliminates the ethical considerations related to suicide by withdrawal of ventilatory support because users of noninvasive methods are not passively attached to a respirator but actively control their alveolar ventilation. The personal sense of controlling one's own life is also better maintained. Physicians have the ethical responsibility to inform patients about noninvasive respiratory aids.

Children and Quality of Life

"Terminal illness" is a self-fulfilling prophecy.

Children's physicians often intentionally withhold information about the potentially life-saving use of respiratory muscle aids, feeling that the quality of life of severely disabled

children is too poor to warrant letting them survive and that their parents cannot make the "appropriate" decision to "let nature take its course." In one study, over 70 health care professionals were asked to judge the quality of life of children with SMA type 1 using a Likert scale of 0-10, on which 10 is maximum and 0 is minimum. Their replies averaged less than 2, and most said that they would advise against any ventilator use. Not surprisingly, parents are often told that there is no point in keeping their children alive because their quality of life is too poor and too much effort is required to raise them. Yet, in a study of 104 responding care providers, mostly parents, of 66 children with SMA type 1 (Chapters 1 and 12), the responses were significantly more positive (Table 8) than the estimates of the health care professionals. Except for effort required to raise the child, the responses were not significantly different from the responses of parents of unaffected children. The parents and other care providers of children with SMA type 1 noted that the children were happy and that their lives were worthwhile despite the relatively high effort that it took to raise them. The ongoing use of noninvasive ventilation was clearly not considered an intolerable burden for these children or their care providers.¹¹⁴⁶

On the other hand, the emotional status of the children with SMA needs to be assessed. It has been noted that 5 patients with SMA type 2 and 3 had intense and recurrent anguish about death and the image of "a narcissistically injured self."¹¹⁴⁷ However, the title of the paper, which cites onset of "a terminal disease," can in part explain the anguish. Patients and their families are told that they are terminally ill because their physicians are unaware of how to help them avoid morbidity and mortality. Nevertheless,

Table 8. Responses of 104 Care Providers of 66 Children with SMA Type 1

Likert scale 0 (minimum) to 10 (maximum) except where noted	7.8
Quality of the child's life	8.0
Quality of the provider's life	8.3
Effort in raising the child (compared with 5 for average child)	5.8
Burden in raising the child (compared with 5 for average child)	8.5
Child's happiness	9.6
Child's life worthwhile	
Likert scale 0 (minimum) to 7 (maximum)	6.0
Child's satisfaction with life	
Semantic differential 1 to 7	
Life with your child is:	
Boring—Interesting	6.7
Hard—Easy	3.8
Lonely—Friendly	6.1
Miserable—Enjoyable	6.3
Useless—Worthwhile	6.7
Empty—Full	6.6
Disappointing—Rewarding	5.9
Tied down—Free	4.0

now that even patients with SMA type 1 can live into adulthood, more work is required to evaluate their emotional needs.

Interestingly, children with SMA, including those with type 1, do very well in school. They have been found to have IQs significantly higher than normal (Chapter 12). Even before age 2 they can often cooperate with VC measurements and Cough-Assist use. People with all severities of SMA have gone to college and achieved meaningful social relationships. Those with types 2-5 have achieved professional employment. We are following 5 children with SMA type 1 over the age of 10 years who are continuously dependent on ventilatory support via tracheostomy tubes. All are doing very well in school. One 19-year-old man graduated third in his high school class and now attends college, despite being unable to speak or eat and having only minimal residual finger and no eye movement. In another study of ventilatory support via tracheostomy for patients with SMA, three of the 10 patients over 18 years of age had college degrees, 2 were college students, 3 graduated from high school, and 2 completed eleventh grade. One patient is the mother of a healthy child. Two patients are employed, and 2 others do volunteer work.⁷⁴

Purtillo summarized an article about ethical issues related to the management of ventilator users by saying that misconceptions about the undesirability of "going on a respirator" have far-reaching negative effects for persons now happily being supported on a respirator, and mitigate the positive effects it could have for some types of chronically impaired persons whose quality of life also could be enhanced by the use of a ventilator.^{71,72} Freed stressed the importance of professionals not imposing their own concepts, values and judgments onto the disabled person.¹¹⁴⁸ Clinicians should be cognizant of their inability to gauge disabled patients' life satisfaction and potential for social and vocational productivity and thus refrain from letting inaccurate and unwarranted judgment of subjective factors associated with quality of life in the general population affect patient management decisions.

Surprendre...

Cueillir ses mots comme des fleurs

Semer des graines au long des coeurs

Confier son âme et sa mémoire

A celui qui viendra plus tard

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