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I. Introduction

Family caregivers of long-term mechanical ventilation (LTMV) users may face the greatest challenges of any group of caregivers. Their attitudes and how they cope with day-to-day problems greatly affect the well-being, quality of life, and survival of their ventilator-dependent loved ones. Family perspectives may also directly affect their loved ones' desire to live or die, and the decisions they make. Moreover, the attitudes and circumstances of ventilator users, and the quality of their relationships with their loved ones, can make a significant difference in the ability and willingness of family members, or significant others, to take on the responsibilities of long-term, round-the-clock care.

Survival beyond respiratory failure may last months or years [when mechanical ventilation (MV) is used] (1–3). When planning optimal management of care, physicians and health care providers may unintentionally overlook crucial problems regarding the needs of family caregivers who are held hostage in their homes. Those who are severely disabled and ventilator dependent rarely visit their physicians. Furthermore, health care professionals may have limited experience in observing LTMV patients in the home setting.

The focus of this chapter is on the perspectives of family caregivers of amyotrophic lateral sclerosis (ALS) patients using LTMV. ALS is a disabling neurologic disease that can strike any adult. Progressive disability and ongoing changes and losses ensue. ALS has a profound effect on the family. Survival of ALS patients using MV can vary considerably. For some patients, survival may exceed five years for users of nasal positive pressure ventilation (NPPV) (2,4), and more than 10 or 15 years when tracheostomy positive pressure ventilation (TPPV) is used (1–3). Thus, the use of LTMV in ALS may result in severe disability (2,5), a high burden of care (2), and high costs if caregivers are hired (2).

This chapter consists of the following:

1. Personal observations of family caregivers of ventilator users with ALS over a 22-year time frame
2. Common misconceptions of family caregivers, 1984–1999
3. Impact of slow communication on the burden of care
4. Impact of totally locked-in patients on families
5. Impact of immobility on the burden of care
6. Hired caregivers
7. Motivating factors for continuing LTMV

8. Need for social interaction
9. Life satisfaction of family caregivers, 1984 to the present
10. The impact of strife on family caregivers
11. Burden of care
12. Common observations of each caregiver group
13. What physicians and nurses can do to help family caregivers achieve life satisfaction and optimal outcomes

II. Background

Since 1984, my nursing practice has focused on serving as a nurse consultant and clinical care investigator to help improve standards of care and survival of ALS patients, particularly for MV users. This long-term investigation of family caregivers of ALS patients using home mechanical ventilation (HMV) was predominantly based on personal observation and interviews with patients and their families. Over the years, I have followed and consulted with several thousand ALS patients and their family caregivers to help make a significant difference in their lives. Serving in the community-based setting, I have visited hundreds of ALS families at their homes or care facilities. Because two hours was the average length of time per visit, there was ample opportunity to observe closely how LTMV users and their families lived day to day. Follow-up consultation and patient family support were provided via the phone on almost a daily basis. However, I have learned firsthand that phone or online assessment of patients and families is an inadequate substitute for actual visits to the home.

Because of the enormous number of patient families that I have closely followed for many years, along with the numerous variables involved, it was not possible to database every observation. The following text includes data from my personal observations of family caregivers whose loved ones used LTMV.

III. Common Misconceptions of Family Caregivers: 1984–1999

In my original investigation of 93 ALS patients using TPPV from 1984 to 1999, 68 (73%) of the 93 patients and their family caregivers believed MV would be a short-term treatment (6). However, 20 patients survived for 8 to 17 years after they commenced TPPV (7). They were unaware that the outcome would be severe disability, high costs, and high burden of care. Most of these patient families were referred for nursing consultation after they began TPPV. My initial visits to the majority of these cases began in the 1980s and early 1990s, an era in which the majority commenced MV during emergency hospitalizations. Many were still able to walk or talk and not ready to die, when TPPV was initiated (2). Families later shared with me their common experiences at the hospital, since I was not present when the MV decision was made. While their loved ones gasped for air, the families tended to panic at the bedside. They did not consider that the future outcome could be severe disability. At that time, 86 (93%) of the 93 commenced MV use when both the patients and the families perceived that MV was an immediate treatment for the respiratory crisis (6). Because they were told “ALS is fatal,” they believed death would occur due to the progression of paralysis and that quadriplegia was the predictor of death. Family members perceived that emergency MV was only temporary treatment (6).

In comparison, 82 (87%) of 94 patients preplanned NPPV use, and 12 (13%) of 94 began NPPV during an emergency (6). Survival with NPPV ranged from 6 to 77 months in patients who were nonbulbar when they began NPPV (6). Although family caregivers believed that NPPV may help treat symptoms or improve sleep, many were oblivious to the potential of survival beyond respiratory failure with optimal use of NPPV. Family caregivers of patients who became severely disabled were surprised at the long-term outcome.

Overall, family caregivers of MV users perceived that the progression of immobility was the predictor of death, despite the use of MV. Also, hired caregivers, particularly nurses from hospices and home care agencies, have generally perceived this misconception. Often, families did not comprehend that respiratory failure is the primary cause of mortality of ALS and that MV can prevent respiratory failure and death.

Misconceptions, therefore, either resulted in unexpected outcomes of survival that families did not anticipate, or early mortality due to implementation of hospice services as disability progressed. The perspective of the home care/hospice nurses greatly influenced the views of families (observations on the perspectives of nurses, however, were not databased).

In my opinion, the prevalent misconception of the progression of immobility as the signal for end-of-life care is the reason morphine sulfate and oxygen were given as a substitute for NPPV. I observed that these two treatments were often administered by hospices in successful users of NPPV. This was based on their perspective that since "ALS is fatal," and that their protocols are the treatment of choice, rather than the optimal use of NPPV and airway clearance. In addition, some hospices consider that NPPV should be used intermittently to provide comfort only and not to extend survival. Furthermore, the use of bi-level ventilators by hospices, without backup rates, has increased for ALS patients. The hospice nurses had based their opinion on the belief that backup rates promote survival and had not understood that a backup is necessary for optimal relief of hypoventilation.

In a number of incidences, I learned of previously successful NPPV users with ALS who were given a prognosis of "24 hours to live." During these emergency hospitalizations, however, I was permitted to educate the health care professionals and patient families on the comfortable use of NPPV, the choices for survival, and that respiratory failure could be treated, if desired. As a result, by using NPPV appropriately, many patients had a dramatic reversal of symptoms and lived up to 24 months or more.

IV. Impact of Slow Communication on the Burden of Care

Severe disability was a usual outcome for ALS patients who used LTMV for two to five years or more. Although suctioning of the tracheostomy increased the burden and intensity of home care (2), most family caregivers reported that slow communication and immobility were the primary factors in the burden of care, rather than caring for the tracheostomy.

Methods of communication varied widely for patients who did not have intelligible speech. They included writing, push button devices, alphabet boards, augmentative communication systems, or computers with voice synthesizers that are activated by switches. Unless patients had the ability to write or push buttons speedily, communication devices usually require diligent effort and concentration. This process was extremely slow, despite the use of the most sophisticated communication technology. Some patients were unable to grasp a pen but had sufficient arm mobility to move their hand and some could write legibly and quickly with a writing device (8). The fastest method of communication that I have ever

observed, among people who were immobile and unable to articulate, is a system using alphabet letters on a card or a board. After dividing and numerating the alphabet into five or six rows, patients can readily select, by eye blinks, the rows and letters to spell words.

Overall, family members indicated that slow communication caused the greatest frustration. During home visits, caregivers were observed demonstrating impatience and anger. When patients struggled to communicate their wishes, the process was often slow and unintelligible. Most sophisticated communication devices took a great deal of time and effort to use. When tempers flared, patients were often reduced to tears. This further hampered their communication efforts to become even slower. Immobile patients often indicated that impaired speech exacerbated their feelings of helplessness. If rapid methods of communication were not used, family caregivers, in turn, indicated that this compounded their feelings of hopelessness and depression. In contrast, family caregivers of patients who were nonbulbar and who could articulate effectively had a significantly reduced burden of care. Patients who were able to talk could express their wishes quickly. This greatly expedited the providing of care. The ability to express their feelings reduced misunderstandings and strife in patient-family relationships. The consensus of almost all patients and families is that the ability to communicate is the most essential factor. Effective communication is the primary requirement for maintaining quality of life. When communication became exceedingly difficult, the burden of care increased and the quality of life decreased.

V. Impact of Totally Locked-in Patients on Families

As immobility progressed, it was not uncommon for patients using TPPV to become totally locked in (after using TPPV for more than five years). I describe this state as having no reliable ability to move a single skeletal muscle, including the eyes. Consequently, the patient who is totally locked in has no ability to communicate or respond to any question. During my investigation from 1984 to 1999, 16 (17%) of 93 TPPV patients became totally locked in (6). Prior to losing all ability to blink or move their eyes, 8 of the 16 indicated that they also lost their vision or became blind (perhaps the inability to blink for a long term led to corneal damage and the permanent loss of vision). Hence, before becoming totally locked in, the impaired vision precluded the use of alphabet boards or communication devices because the patients could not see them. Besides the totally locked-in patients, I have observed many others who almost became locked in. They still had one reliable muscle that slightly moved in response to questions. Thus, I define this as a pre-totally locked-in state and final opportunity to communicate.

As patients became locked in, family caregivers indicated that the burden of care subsided significantly. The frustration with communication ceased because there was no longer an effort to communicate. Patients who had previously been demanding became despondent and silent forever. Family caregivers often referred to their loved ones as "living corpses." Many family members, especially children, became terrified by the physical appearance of someone they loved who had become locked in. They could no longer endure either visiting or assisting with care of their loved one.

In Ohio, the incidence of ventilator users becoming totally locked in since 2000 is significantly less than what I observed in the 1980s and 1990s. Because of increased education about life choices and advance directives, most patients are now indicating their wishes in advance and withdrawing from MV if they can no longer communicate. During visits to Japan, however, I have observed a high number of LTMV users who were severely paralyzed.

VI. Impact of Immobility on the Burden of Care

After impaired communication, immobility was the second most significant unpreventable challenge faced by family caregivers. Immobility was a leading factor for the high burden of care. If patients were quadriplegic, the constant positioning and repositioning of the limbs, trunk, and head were ongoing round-the-clock, never-ending tasks. Patients who had lost the use of their upper extremities were totally dependent on others for care. Those families reported a higher burden of care than families whose members were non-ambulatory, but still retained the use of their upper extremities.

Family members who received adequate help when it was needed had experienced significantly less burden of care than those without help. Some family caregivers reported no burden of care, despite having loved ones with severe disability, if they had the finances and benefits to receive hired help at home everyday. They either had built-in family support or were forced to hire caregivers. Life satisfaction of family caregivers also depended on whether their loved ones were satisfied. Patients who had acquired the ability to cope with the physical and emotional aspects of the disease often had peace of mind. That enhanced the family caregivers' ability to cope and still enjoy life. Despite severe disability, many families continued to respond positively to the patient.

Immobile patients using TPPV could never be left alone, unlike MV users with mobility. If a patient could execute self-care of a tracheostomy, they were occasionally left home alone.

Some patients had private duty nursing insurance benefits in which licensed nurses provided care for at least one shift per day. This had greatly minimized the burden of care for the family, and made it possible for the majority of MV patients to live at home rather than in care facilities. In the 1980s and 1990s, benefits for private duty nursing by third party payers were more widely available (2) than in the previous five years. Since 2000, the number of patients who were forced to hire caregivers without financial aid, if round-the-clock care and supervision were needed, has risen sharply. The lack of private duty nursing benefits has also caused more patients to move to skilled nursing facilities than in the previous years. Thus, the reduced healthcare benefits for caregiver assistance and the increased "out-of-pocket" expenses have resulted in higher direct costs and the depletion of life savings. The hiring of caregivers, not the purchase or rental cost of equipment and supplies, was the primary factor for the high costs of using MV (2). The significant costs and the high burden of care were the most common causes for patients with ALS to withdraw from MV.

On the basis of close observation, most family caregivers provided meticulous care, usually as good as hired nurses (2). Initially, family caregivers were trained on tracheostomy care and were supervised by a registered nurse. Most school-age children and teens assisted with care. Older children usually provided tracheostomy care (2,9), while younger children were often educated on intervening in the event of an emergency.

VII. Hired Caregivers

The number of patient families with hired caregivers varied widely, and was dependent on their health care benefits and/or their incomes. Surprisingly, many families were unaware of their specific health care benefits, both before and after commencement of LTMV. In the United States, patients with poverty status actually obtained the most health care benefits.

This usually included coverage for care by the government for either a part-time hired caregiver at home or full-time admission to a care facility.

All families who hired private duty nurses or caregivers did not like the care attendants in their homes continuously, despite the critical need for help. Some felt that the presence of care attendants in the home invaded their privacy. In addition, married couples did not often have time alone. When families lacked sufficient home care benefits and were forced to pay "out-of-pocket" for care, the majority preferred hiring nonagency nurses. Nurses from home care agencies were significantly higher in cost than nonagency nurses (6). Also, families often complained that agency nurses followed their own protocols, not always the wishes of patients and families.

VIII. Motivating Factors for Continuing LTMV

A motivating factor for continuing long-term MV was the hope for a miracle. In my previous study of 84 patients using TPPV, 26 hoped to stay alive because they were waiting for a cure. Four had faith that God would heal them, while 22 patients and their family members were sustained by the hope that a cure for ALS would be found. On the basis of "time for a cure" campaigns and news on ALS research, they all believed if a cure for ALS was discovered, their paralysis would be reversed. They believed this could happen "almost anytime."

IX. Need for Social Interaction

Despite their immobility, MV patients who maintained a method of communication and experienced regular social interaction could achieve life satisfaction (6,7). Patients and caregivers were satisfied with their quality of life on LTMV as long as social interaction continued, and resources were available. Social interaction included regular contact with people and participation in activities with family, friends, church groups, or caregivers. With the use of portable ventilators, powered wheelchairs, accessible vans, ramps, and laptop computers for communication, these ventilator users were able to go outdoors. They could travel, attend church, school activities, sporting events, and the theater. Others who were homebound enjoyed frequent interaction with their hired caregivers. They could enjoy social or church meetings at their homes every week and also interact with online support groups. This communication greatly minimized the feelings of social isolation, abandonment, and helplessness.

X. Life Satisfaction of Family Caregivers: 1984 to the Present

Studies have shown that the majority of MV patients were "glad to be alive" and that both patients and family caregivers "would choose MV again if given the choice" (2,9,10). The majority of family caregivers indicated that despite the heavy burden, they often continued to be willing caregivers to keep their loved ones alive (2). Use of MV (survival beyond respiratory failure) ranged from one to more than ten years. This included both TPPV and NPPV users and their families (2,10). In the investigation by Moss et al. (10), I visited 31 patient families, or majority of the study subjects. One consideration of this study (10)

was that the patient/family visits occurred only once at any given moment of time. Months or years later, during a follow-up investigation, I learned that perspectives of many caregivers changed as circumstances and conditions of their loved ones using MV progressed. Thus, perspectives of family caregivers may change if disability increases significantly, burden of care becomes significantly higher, life savings become depleted, health status of family caregivers markedly deteriorates, and survival of those who become severely disabled exceeds five years.

Over the years, I have found that those patients who rated their quality of life as high and were glad to be alive had happier family caregivers. In contrast, the patients with severe disability, who suffered extreme depression and no longer desired to live, had family caregivers who shared their depression.

Family caregivers have often indicated that the most stressing period for using MV was from the time of commencement of TPPV through the following six months after their loved one became ventilator dependent. Thereafter, as family caregivers mastered ventilator care procedures, the anxiety and fear subsided.

Male caregivers seemed to have been more creative in their approach to care and tended to "invent" devices that made life easier. Women tended to purchase commercial products to simplify tasks.

Learning the skills for providing care and enhancing mobility, comfort, and safety made a significant difference among family caregivers. There are helpful educational resources on the art of caregiving (11). Some communities offer volunteer services to help family caregivers. One such program is "Share the Care." (12).

The ability to anticipate and engage in pleasurable activities and to maintain an environment where family members live in harmony with one another are essential components for life satisfaction. Family caregivers and LTMV patients need to know they have a purpose for living (13), and must have a desire to live their lives to the fullest potential, despite their circumstances (14).

For many, spirituality also played a key role in life satisfaction. Family caregivers, who expressed that they had faith in God and prayer, attributed their peace of mind to God. Many believe that they "would not have been able to make it" without God's help. "Courage To Live: The Story of Charlie and Lucy Wedemeyer" (15) is an inspirational DVD about a LTMV user with ALS and his wife. They believe that through the power of faith and the love of family and friends, they have been able to face daily challenges. They believe that there is no obstacle that they cannot overcome. The DVD also gives profound words of encouragement to other LTMV user families. Faith is a predictor for caregivers achieving life satisfaction (Table 1).

XI. The Impact of Strife on Family Caregivers

Family caregivers and/or patients who are quick to anger, who are impatient and demanding, and who have a tendency to display mood swings or temper outbursts from daily situational stress frequently create an atmosphere of strife in the home. These pre-existing characteristics usually become significantly worse after LTMV is initiated. Offensive communication and refusal to forgive may ultimately result in deep bitterness, hatred, and the inability to cope. Patients who are demanding, manipulative, and seldom express appreciation to their family caregivers will foster resentment.

Table 1 Predictors for Caregivers Achieving Life Satisfaction

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- Availability of willing, competent caregivers
 - Adequate assistance with care
 - Adequate finances and healthcare benefits
 - Accessible home environment, including exit and entry of home
 - Accessibility of appropriate durable medical equipment and self-help devices
 - Achieving and maintaining effective methods of communication
 - Regular social interaction and participation in pleasurable activities
 - Supportive family and friends
 - Harmonious family relationships
 - Faith and close relationship with God
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Table 2 Predictors of Caregiver Stress

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- Strife or unresolved conflict between patient, primary family caregiver and/or with relatives of the patient
 - Slow or ineffective method of communication by patient, including use of augmentative communication systems
 - Severe disability of loved one, particularly if upper extremities are immobile
 - Insufficient help with care
 - Depression, hopelessness, despair, and no desire of patient to live
 - Unmanaged anxiety and fear
 - No social interaction or opportunity to enjoy pleasurable activities
 - Lack of sleep
 - High costs for equipment/supplies, hired help and/or depletion of life savings
-

Family caregivers attributed strife as a major stress factor (Table 2). Unresolved conflict robs families of "peace of mind," the ability to sleep, and the attainment of joy in life. Families who harbor strife never achieve their potential life satisfaction. Women, more often than men, reported that strife was often sparked by hypercritical remarks by their in-laws. Adult children have often reported conflicts among siblings when one person takes on all the caregiving or household tasks for a parent, while the others offer little or no assistance. Becoming consumed with anger and unresolved conflict is destructive to both the physical and emotional well-being of patients and their families. It destroys the spirit of unity. Conflicts in the home compound stress, grief, and interfere with the ability to cope. Anger may also trigger respiratory failure and result in early mortality.

XII. Burden of Care

Caregivers without a supportive family or who lack finances to hire help are often held hostage in an environment with little or no opportunity for pleasurable activities. Prolonged isolation and physical or emotional fatigue will result in caregiver burnout. Prolonged inability to cope may lead some to become emotionally dysfunctional to the point of jeopardizing the safety of their ventilator-dependent loved ones. Family caregivers who lack family support may turn to alcohol or tranquilizers for relief.

XIII. Common Observations of Each Caregiver Group

A. Well Spouse Caregivers

In my experience, the majority of primary caregivers of ALS patients using LTMV at home were the patients' spouses. Men or women in their prime of life with a full-time career and/or those who are parents to dependent children living at home have significantly higher caregiver stress levels than those male or female well spouses who are retired and without children living at home. Unless well spouses who work full time have adequate family assistance and finances to hire help, it is difficult, if not impossible, to pursue the task alone when their partners are physically helpless. Slow communication compounds the burden of care. Caregivers without adequate assistance often feel the sense of pending doom. They suffer from frequent headaches and back pain. Their lack of sleep often escalates into outbursts of anger, followed by guilt and depression. These are a few signs of caregiver burnout.

The responsibility of round-the-clock care can result in caregiver breakdown. Because use of LTMV may continue for years, care facility placement may be the only solution to assure the availability of round-the-clock caregivers. In my overall experience, I have observed significantly more caregiver husbands who were less able to provide ongoing care for more than two years than caregiver wives. Feeling the burden of care on their spouses and children, more women than men withdrew from MV, anticipating death.

B. Children Caregivers

Adult children who have a disabled parent living home alone have the constant burden of coordinating daily care and household tasks. Often a parent resists moving to a care facility, despite the risk of falling or dependent use of NPPV. Unless the parent has advance directives that are clearly understood, conflicts may arise among siblings on sharing the responsibilities of care or addressing life support issues.

Teenagers with a parent at home using LTMV faced increased responsibilities. They often resented caregiving tasks, especially if the duties interfered with their social activities or studies. Watching their beloved parent wither away was also very devastating. Through the years, I observed more strife and conflict in the home when the TPPV user with ALS was the father, rather than the mother. If the father previously governed discipline and then became inarticulate and helpless, mothers with "burnout" and rebellious teens often encountered compounded stress. Poor grades, discipline problems at school, and teen pregnancies were not uncommon in such households, particularly if there was unresolved conflict. Some teens attempted suicide. Younger children adapted better than older children. Small or school-age children in the home usually assisted with minor tasks.

C. Parent Caregivers

Of all the categories of family caregivers, parents had the most difficulty in adapting to the catastrophic diagnosis and threat to their child's life, regardless of the age of the child. Some parents tended to overprotect their child and hover over hired caregivers to assure that every procedure of care was performed with perfection. In addition, some tended not to support the wishes of their child to withdraw or refuse LTMV. Occasionally, I heard, "I plan to keep my baby alive." Parents, who were elderly, sometimes could not physically

or emotionally endure the caregiving tasks. Some of them kept their adult child bed bound with diapers, and kept visitors away. In general, however, parent caregivers tended to demonstrate the most tolerance in their caregiving role.

XIV. What Physicians and Nurses Can Do to Help Family Caregivers Achieve Life Satisfaction and Optimal Outcomes

1. Provide accurate, understandable, and necessary information on the patient's diagnosis and the possible survival and outcome of LTMV. Inform patient families that (i) immobility will progress; (ii) if TPPV is used, ongoing tracheal suctioning will be required and round-the-clock caregivers will be necessary; (iii) if dependent on NPPV, periodic interface adjustment and supervision will be required; (iv) survival of nonbulbar NPPV users may be long term (2); (v) paralysis is likely irreversible; (vi) quadriplegia is not a predictor of survival; (vii) costs for hiring caregivers are high, and may deplete life savings, if survival is long; (viii) heavy family burdens or nursing home placement may ensue (2); and (ix) the patient has the right to withdraw from MV, if desired.
2. Respect patient family wishes and regard each MV patient's life as precious. Do not perceive that quality of life is over if tracheostomy ventilation is used, nor believe that the patient must live like a "vegetable" and be held hostage to a bed. Be aware that portable, lightweight ventilators are available, as well as technology to enhance mobility. Understand that many MV users are glad to be alive and would choose MV again, if given the choice. Recognize that many patient families have financial resources to hire help or have available, willing, and competent family caregivers to assist with care.
3. Tell families not to promise their loved ones that they will never allow them to live in a care facility. This will help eliminate conflict, guilt, and stress in the event facility placement becomes necessary.
4. Help patient families obtain adequate caregiver assistance. Assess their health-care benefits and financial resources, if necessary. Identify all possible resources for help and arrange respite care services, or care facility placement, periodically, to help prevent caregiver burnout.
5. Help patient families achieve and maintain an effective and rapid method of communication. Although augmentative communication systems can be accessed by the severely disabled, most commercial systems require concentration and are slow. Encourage patients and families to memorize an audible alphabet system for fast communication if the patient has one reliable muscle to signal rows and letters of the alphabet, as the caregiver says the rows and letters out loud. Plan and post charts of lists of the patient's possible wants and needs to spare time and to help caregivers understand what the patient is expressing or requesting.
6. Help family caregivers plan a reliable and easily accessible method for the patient to signal for help, contact their families, or send an alert in the event of an emergency situation.

7. Arrange a professional assessment of the patient's home environment to obtain recommendations for making the home accessible. Obtain appropriate durable medical equipment, mobility aids, and self-help devices for enhancing mobility, comfort, and safety. Make all activities of daily living as easy as possible. Be sure to see that the home has accessible means for the patient to leave and enter the home.
8. Plan professional counseling services for households with unmanaged emotions, troubled teens, and broken marriages. Also, encourage patient families to regularly engage in pleasurable activities, leave the home, visit friends, and take trips, despite the patient's disability and use of MV. Provide education and refer to helpful resources for traveling with ease.
9. Visit or arrange for a registered nurse or social worker to visit the homebound patient using LTMV periodically, despite the appearance that all is stable. Monthly visits are ideal. I highly recommend that physicians visit their homebound patients at least once a year to assess firsthand the patient/family's well-being, wishes, and other concerns. Family circumstances may change each year.

By understanding the perspectives of your family caregivers, you can help your patient families avoid misconceptions, obtain necessary information for making best choices, and help them to achieve and maintain the best life possible. The outcomes of family caregivers and their loved ones using LTMV may depend on you.

Acknowledgments

This chapter is dedicated to the late Edward A. Oppenheimer, MD, FCCP, who has served as a special colleague, mentor, and advocate of this long-term investigation. His dedication to improving the quality of life of people with ALS and ventilator users will always be remembered. The author also gives special thanks to the many patients and families who participated in the studies, as well as their physicians, nurses, respiratory therapists, and social workers.

References

1. Oppenheimer EA. Decision-making in the respiratory care of amyotrophic lateral sclerosis: should home mechanical ventilation be used? *Palliat Med* 1993; 5(suppl 2):49-64.
2. Cazzolli PA, Oppenheimer EA. Home mechanical ventilation for amyotrophic lateral sclerosis: nasal compared to tracheostomy-intermittent positive pressure ventilation. *J Neurol Sci* 1996; 139(suppl):123-128.
3. Bach JR. Amyotrophic lateral sclerosis. Communication status and survival with ventilatory support. *Am J Phys Med Rehabil* 1993; 72:343-349.
4. Bach J R. Amyotrophic lateral sclerosis: prolongation of life by noninvasive respiratory aids. *Chest* 2002; 122:92-98.
5. Hayashi H, Kato S, Kawada A. Amyotrophic lateral sclerosis patients living beyond respiratory failure. *J Neurol Sci* 1991; 105:73-78.
6. Cazzolli PA, Oppenheimer EA. Use of nasal and tracheostomy positive pressure ventilation in patients with amyotrophic lateral sclerosis (ALS). Abstracts of Papers, 7th International Conference on Noninvasive Ventilation Across the Spectrum from Critical Care to Home Care, Orlando, Florida, March 14-17, 1999.

7. Cazzolli PA, Oppenheimer EA. Use of nasal and tracheostomy positive pressure ventilation in patients with ALS: changing patterns and outcomes. *Neurology* 1998; 50(suppl 4):A417–A418.
8. Writing Angel. Solutions for Living, PO Box 36116, Canton, Ohio, U.S.A., 44735 (888-884-5483) www.solutionsforliving.us.
9. Cazzolli PA, Oppenheimer EA. Home mechanical ventilation for motor neuron disease (MND/ALS): nasal compared to tracheostomy intermittent positive pressure ventilation (IPPV). Abstracts of Papers, 6th International Symposium on ALS/MND, Dublin, Ireland, November 17–19, 1995.
10. Moss AH, Oppenheimer EA, Casey P, et al. Patients with amyotrophic lateral sclerosis receiving long-term mechanical ventilation: advance care planning and outcomes. *Chest* 1996; 110:249–255.
11. Arbuckle C. *A Caregiver's Journey with a Terminal Patient*. Los Angeles: Milligan Books, 2004.
12. Caposella C and Warnock S. *Share the Care: How to Organize a Group to Care for Someone Who is Seriously Ill*. New York, NY: Simon & Schuster, 2006.
13. Warren R. *The Purpose Driven Life: What on Earth Am I Here For?* Grand Rapids, Michigan: Zondervan, 2002.
14. Osteen J. *Your Best Life Now: 7 Steps to Living at Your Full Potential*. New York, NY: Warner Books, 2004.
15. Wedemeyer C, Wedemeyer L. *Courage to Live: The Story of Charlie and Lucy Wedemeyer*. DVD 2006. Available at: www.couragetolive.com.