

Home mechanical ventilation for amyotrophic lateral sclerosis: nasal compared to tracheostomy-intermittent positive pressure ventilation

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Abstract

People with amyotrophic lateral sclerosis (ALS) usually die from respiratory failure unless they use mechanical ventilation (MV). Many die of respiratory failure without being adequately informed about the available options, such as MV, that can provide symptomatic relief and prolong survival. The traditional method of MV used for persons with ALS has been tracheostomy-intermittent positive pressure ventilation (IPPV). However, the advent of nasal-IPPV has provided a new option for relieving respiratory symptoms and prolonging survival among selected individuals. The ALS Association Data on 75 ALS patients using MV is reviewed. Twenty-five patients used nasal-IPPV, all started electively. Survival with nasal-IPPV ranged from 6 to 64 months for non-bulbar patients, or until the onset of severe bulbar dysfunction when nasal-IPPV no longer was effective. Fifteen of these non-bulbar patients used nasal-IPPV from 20 to 24 h daily; one of these patients used nasal-IPPV continuously for 24 h daily for 24 months. One hundred percent of the users indicated they were glad they chose nasal-IPPV. In contrast, 50 have used tracheostomy-IPPV, usually as a result of emergency hospitalization without advance decision making. Twenty-five patients (50%) lived in a sub-acute skilled nursing facility (SNF) and only 18 of these (72%) were satisfied with their quality of life. Patients using tracheostomy-IPPV with good care are able to live many years: 27 of the 50 (54%) are still living, including one patient who is still living after 14 years of MV. In conclusion, home mechanical ventilation with nasal or tracheostomy-IPPV are options for selected people with ALS. Nasal-IPPV offers many advantages; it was only used when MV was planned and desired. Nasal-IPPV can be used unless bulbar impairment is severe.

Keywords: Amyotrophic lateral sclerosis; Motor neuron disease; Mechanical ventilation; Non-invasive ventilation; Decision making

1. Introduction

Progression of amyotrophic lateral sclerosis (ALS) eventually results in ventilatory failure (Braun, 1987). As respiratory muscle weakness advances, alveolar hypoventilation, hypercapnia, poor cough and aspiration usually ensue (Sivak, 1980). Associated symptoms include excessive fatigue, disrupted sleep, orthopnea, and daytime somnolence (Strumpf et al., 1990a). Visible signs of early respiratory compromise may go unnoticed until impaired gas exchange occurs, often triggered abruptly by pulmonary complications (Bach and Alba, 1990). Unless me-

chanical ventilation (MV) is used, CO₂ retention, acidosis, poor cough and sepsis cause death.

The majority of ALS patients succumb to respiratory muscle weakness, often precipitated by pulmonary complications related to bulbar paralysis in which aspiration, pneumonitis, and atelectasis are common (Hill et al., 1983; Oppenheimer, 1993; Norris and Fallat, 1994). Respiratory distress may arise unexpectedly (Bach, 1993a,b). Although respiratory failure can be avoided if MV is used successfully, ALS patients generally do not use ventilatory support and, therefore, die (Moss et al., 1993). In many instances, patients are not informed that breathing support options are available, nor are they adequately educated to make informed decisions, especially prior to respiratory failure which may arise suddenly (Bach, 1992, 1993a; Bach and Barnett, 1994). Some patients experiencing acute

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distress, accompanied by anxiety and panic, may go to the hospital seeking relief of symptoms and be treated with endotracheal ventilation, often forcing a hasty decision on whether to withdraw life-support treatment and die or to have tracheostomy and intermittent positive pressure ventilation (tracheostomy-IPPV) (Oppenheimer, 1993). Methods for non-invasive ventilation are usually not offered (Bach and Barnett, 1994).

In the United States, the traditional method of MV used for persons with ALS has been tracheostomy-IPPV (Sivak, 1982; Bach and Barnett, 1994). Survival after starting tracheostomy-IPPV can be for many years. Some patients are living 10 or more years (Iwata, 1987; Bach, 1993b; Oppenheimer, 1993). The consequences of prolonged survival and the burdens of care are sometimes not realized by patients and family caregivers until after MV is instituted (Sivak, 1982).

In recent years the advent of nasal-IPPV has provided a new option that relieves respiratory symptoms by reversing alveolar hypoventilation and can prolong survival among selected individuals (Ellis et al., 1987; Bach and Alba, 1990; Strumpf et al., 1990b; Goldstein et al., 1991; Waldhorn, 1992; Bach, 1993a; Hill, 1993; Robert et al., 1993; Leger et al., 1994; Meyer and Hill, 1994). Reports in the literature on the successful use of nasal-IPPV in patients with ALS have been limited (Alba et al., 1976; Bach et al., 1987; Ellis et al., 1987; Fallat et al., 1987; Bach, 1992; Norris and Fallat, 1994; Sherman, 1994; Pinto et al., 1995).

The purpose of our review is to report our findings on 75 patients with ALS using MV which includes 50 patients using tracheostomy-IPPV and 25 patients using nasal-IPPV. We compare our findings on the use of non-invasive ventilation to long-term tracheostomy ventilation.

2. Patient population and methods

A prospective study of 25 individuals was conducted from 1990 to 1995, to evaluate the value of nasal-IPPV for ALS patients, and to compare its use to the alternative of tracheostomy-IPPV for long-term life support. This study particularly focused on: relief of symptoms of respiratory failure, length of survival, decision making by the patient and family, the cost for equipment and personal care, and the quality of care provided by family members, unlicensed attendants and nurses.

Fifty people with ALS using tracheostomy-IPPV were identified during the 7 years, 1988–1995. Twenty-five people lived at home and their families usually served as primary caregivers. The majority employed licensed nurses for assistance with care. Their health insurance coverage required using nurses. The other 25 people lived in subacute skilled nursing facilities (SNF), because they were widowed, single or lacked willing, competent or available caregivers at home. Forty-six (92%) of the 50 people had

tracheostomy-IPPV as a result of emergency hospitalization for respiratory failure and distress, and had not decided in advance. All 75 MV users were initially evaluated and diagnosed by their neurologist. The findings were confirmed at an ALS center or neuromuscular clinic (sponsored by the ALS Association, the Muscular Dystrophy Association, or a university medical center).

The 50 tracheostomy-IPPV patients and 25 nasal-IPPV were seen for evaluation, patient care consultation and follow-up by one of the authors (P.A.C.). The patients were initially identified by the ALS Association's Eastern Ohio Chapter, Western Ohio Chapter and Western Pennsylvania Chapter; the Visiting Nurse Society of Stark County, Ohio; and an ALS support group in West Virginia. All patients were visited at their homes or SNF care facilities. Periodic follow-up telephone calls and visits to patients' homes and SNFs were made to determine quality of care and perceived quality of life of both patients and family caregivers. Patients and families were interviewed to evaluate: management of care and costs involved; tracheostomy and home care techniques performed by family members, nurses, and unlicensed attendants; and the use of MV. Evaluation included: relief of symptoms, the hours the ventilator was used daily, how well it was tolerated, problems with nasal interfaces, and bulbar involvement. Further contacts with patients and families were made frequently, when caregivers telephoned from time to time for advice on managing care or regarding caregiving. Some of these patients and family members were also seen at bi-monthly ALS support group meetings, sponsored by the Visiting Nurse Society of Stark County in which one of the authors (P.A.C.) is a facilitator.

3. Results

3.1. Fifty ALS patients who used tracheostomy-IPPV

The mean age of the 50 tracheostomy-IPPV patients was 60 years (range: 31–89; 66% male, 34% female). As of 1995, 23 had died and 27 were still living. Only four (8%) of the 50 patients had chosen tracheostomy ventilation in advance, before emergency hospitalization. Of the 25 patients (50%) who lived at home, 23 were satisfied with their quality of life. Of the 25 patients (50%) who lived in a SNF, seven (28%) were not satisfied with their quality of life, and depression was common. Forty-one (82%) of the 50 patients indicated that they were glad they used tracheostomy-IPPV, and that when they began its use, the alternative was death. Some patients were still ambulatory, or had use of upper extremities, or were non-bulbar for many months after they began tracheostomy-IPPV. Some of these patients reported they did not want to die at the time of respiratory failure when they began MV. One patient used MV for 5 years and eventually stopped MV after seriously considering this for 1 year after he lost

Table 1
Data on use of nasal-IPPV by living patients ($n = 8$)

Patient	Gender	Age	Ventilator	Maximum hours per day	Duration of use (months)	Bulbar signs
1	M	57	BiPAP [®] S/T	24	64	No
2	F	61	BiPAP [®] S/T	23	30	No
3	F	71	BiPAP [®] S/T	24	30 (then tracheostomy)	2 months prior to tracheostomy IPPV
4	M	54	PLV-100	23	26	No
5	M	35	BiPAP [®] S/T	16	15	No
6	M	73	PLV-100	23	16	No
7	M	39	BiPAP [®] S/T	24	16	No
8	M	48	BiPAP [®] S/T	12	1 (then tracheostomy)	When MV began

ability to use his extremities; he wanted to relieve his wife, who had failing health, of the burden of care. The patient's physician supported his decision to withdraw MV, and ordered palliative treatment through hospice services. The patient died peacefully. Another patient used MV for 2 years and developed sudden complications, resulting in brain anoxia and permanent unconsciousness. The patient's spouse requested withdrawal from MV; his physicians honored this; tracheostomy-IPPV was then discontinued.

The primary caregivers of 22 patients were family members. Suctioning of tracheostomy increased the burden and intensity of home care provided by family caregivers. Healthcare insurance reimbursed only licensed nurses for home care. In 1995, the usual cost for a licensed practical nurse was \$25 per hour from an agency. Twenty (80%) of these patients used nurses to supplement the family caregivers. Five patients never hired licensed nurses for home care. None of the patients were eligible for hospice services because hospices in these communities exclude peo-

ple who use tracheostomy-IPPV. All family members (including older children in some cases) provided tracheostomy care as skillfully as trained respiratory care practitioners and registered nurses. Unlicensed paid attendants were employed by some families; when properly trained they provided excellent quality care.

Tracheostomy-IPPV survival data ($n = 50$) after starting MV: (A) Twenty-three have died: 8 lived 1–2 years; 4 lived for 2–4 years; 5 lived for 5–6 years; 2 lived for 7–8 years; 4 lived for 9–12 years. (B) Twenty-seven are alive: 11 living for 1–2 years; 8 living for 2–4 years; 3 living for 5–6 years; 2 living after 7–8 years; 3 are alive after 11–14 years of MV.

3.2. Twenty-five ALS patients who used nasal-IPPV

The mean age of the 25 nasal-IPPV patients was 64 years (range: 35–86; 64% male, 36% female). By 1995, 17 nasal-IPPV users had died and eight were still living. All

Table 2
Prior use of nasal-IPPV by deceased patients ($n = 17$)

Patient	Gender	Age	Ventilator	Maximum hours per day	Duration of use (months)	Onset of bulbar signs	Using MV when died	Reason for withdrawal
9	M	65	BiPAP [®] S/T	24	32	2 months prior to death	Yes	
10	F	77	BiPAP [®] S/T	22	30	2 months prior to death	Yes	
11	F	63	BiPAP [®] S/T	21	25	2 months prior to death	Yes	
12	F	63	BiPAP [®] S/T	21	19	2 months prior death	Yes	
13	F	74	PLV-100	20	15	2 weeks prior to death	Yes	
14	M	65	PLV-100	21	14	2 months prior to death	No	Did not relieve symptoms
15	F	56	BiPAP [®] S/T	6	10	When MV began	No	Did not relieve symptoms
16	M	65	BiPAP [®] S/T	23	9	No	No	Did not relieve symptoms
17	M	76	PLV-100	Intervals	8	1 month prior to death	No	Did not like alarm
18	M	72	PLV-100	Intervals	7	No	No	Did not like alarm
19	M	86	BiPAP [®] S/T	20	6	No	Yes	
20	M	78	PLV-100	10	3	1 month prior to death	No	Did not relieve symptoms
21	F	71	BiPAP [®] S/T	Intervals	2	When MV began	No	Did not relieve symptoms
22	M	61	BiPAP [®] S/T	Intervals	3	When MV began	No	Did not relieve symptoms
23	F	52	BiPAP [®] S/T	Intervals	2	No	No	Did not want to live
24	M	72	PLV-100	Intervals	1	No	No	Did not want to live
25	M	60	PLV-100	Intervals	1	When MV began	No	Did not relieve symptoms

25 patients had planned MV in advance and chose nasal-IPPV. They used either the BiPAP[®] S/T System, bi-level positive airway pressure spontaneous/timed mode ventilator (Respironics[®], Murrysville, PA, USA) or a PLV[®]-100 volume cycled ventilator (LIFECARE International, Westminster, CO, USA). Twenty-three patients lived at home and two widows (8%) lived at nursing homes.

Five of the 25 nasal-IPPV users had bulbar symptoms of dysphagia, dysarthria and excessive oral secretions when they first began using nasal-IPPV. Four of these bulbar patients withdrew from nasal-IPPV because it did not relieve their respiratory distress, and accepted death. The fifth bulbar patient also did not experience relief of symptoms using nasal-IPPV; he desired long-term life support and elected tracheostomy-IPPV. He is still living.

Twenty (80%) of the 25 patients were non-bulbar when nasal-IPPV was started, able to talk and swallow, without sialorrhea, with intact pharyngeal muscle function. Seven of the non-bulbar patients are still living and 13 have died (see Tables 1 and 2). The 20 patients who were non-bulbar when they began nasal-IPPV had reported relief of respiratory symptoms. Nasal-IPPV increased the voice volume among successful users. Five of the 20 non-bulbar patients used nasal-IPPV from 1 to 8 months. One of the five patients discontinued use because of inability to adapt to the ventilator. Two patients withdrew because of inability to adapt to the diagnosis; they had progressive immobility, concern about the burden of care, and desired not to live. The fourth and fifth patients developed the onset of bulbar symptoms 1 month prior to their deaths. Before they died, both patients developed difficulty with excessive oral secretions. One reported that nasal-IPPV no longer relieved his symptoms, while the other experienced difficulty with the alarm on the volume cycled ventilator which triggered frequently. They decided to stop using MV; neither patient was using nasal-IPPV when they died. Thus, 15 non-bulbar patients used nasal-IPPV, ranging from 20 to 24 h daily. This included one patient who used it continuously for 24 h daily for 24 months. Survival with nasal-IPPV for these 15 patients ranged from 6 to 64 months. They continued until they developed the onset of severe bulbar dysfunction which made nasal-IPPV no longer effective (see Tables 1 and 2). One patient who successfully used nasal-IPPV for 30 months chose tracheostomy-IPPV after the onset of excessive oral secretions. This patient is still living.

Six of the 15 initially non-bulbar patients had an onset of bulbar symptoms 2 months prior to their deaths. As secretions became excessive, five of the six patients died while using nasal-IPPV. The sixth patient indicated that his ventilator was not relieving his breathing distress. Nasal-IPPV was then discontinued by the family and the patient was taken to the hospital where he died. Two of the 15 non-bulbar patients who remained non-bulbar died shortly after reporting they "weren't getting enough air". One patient was using nasal-IPPV at the time of his death, while the other patient withdrew from the ventilator and

was taken to the hospital where he was given oxygen and died.

4. Discussion

Many who used tracheostomy-IPPV would probably have decided to avoid MV if they had been able to plan in advance, particularly if they had known that they would become a heavy burden to family or be placed in a nursing home (SNF). Most people with ALS do not want to use MV. They need assistance with palliative care and planning for emergencies, so that intubation and MV can be avoided when respiratory failure occurs. In the USA, patients have the right to discontinue MV if it is not desired; this is much easier to do with nasal-IPPV. Once tracheostomy-IPPV is started, people are often reluctant to stop life-support even though this choice is legally and ethically supported in the USA.

Before a decision is made to use tracheostomy-IPPV, the person with ALS (and family) should understand that death related to ALS can usually be prevented with MV and good care; and, that if MV is used, immobility will progress; ongoing tracheal suctioning will be required; round-the-clock caregivers will be necessary; needed resources and costs are high and might result in heavy family burdens or nursing home placement.

The decision to initiate nasal-IPPV was based on the severity of respiratory symptoms together with reduced simple pulmonary function test data. Symptoms include: disrupted sleep, orthopnea, daytime somnolence, exertional shortness of breath, and ineffective cough. Values that indicate significant respiratory impairment are a forced vital capacity of 50% or less, a maximum inspiratory force of -30 cm H₂O or less, or a maximum expiratory force of 30 cm H₂O or less. Other warning signs may not be obvious. Abnormal arterial blood gas or oximetry values are often late findings.

After the onset of bulbar dysfunction, previously non-bulbar patients using nasal-IPPV died within 2 months as aspiration increased, unless tracheostomy was done. Patients with significant bulbar impairment when they tried to start nasal-IPPV were not able to tolerate its use and respiratory distress was not improved.

Tracheostomy-IPPV can extend survival for more than 10 years despite significant bulbar impairment when effective respiratory care is given. Nasal-IPPV prolonged the survival of the 15 non-bulbar patients using the ventilator 20–24 h daily for 6–64 months. Most of these nasal-IPPV users (90%) decided not to have a tracheostomy when bulbar impairment became severe. Factors determining successful use of nasal-IPPV included absence of excessive oral secretions, properly fitting nasal or oral interfaces, willingness to wear interfaces, desire to live and good caregiver support.

The home ventilator management should be monitored

periodically to determine if the patient is comfortable and properly ventilated. Patients who report they are "not getting enough air" need clinical reevaluation and often require adjustment of the ventilator settings. Oxygen administration is usually neither indicated nor safe unless there is pneumonia. If the ventilator is providing appropriate alveolar ventilation, the oxygen saturation should be 93–95% or greater on room air; PaCO₂ should be 40 mm Hg or less, and the arterial pH should be within normal limits.

The quality of tracheostomy-IPPV care provided by properly trained family members and unlicensed attendants was as good as care provided by licensed nurses. The cost for nurses as paid caregivers was \$25 per hour. Costs for unlicensed paid caregivers who are properly trained and supervised by registered nurses should be reimbursed by health care or community resources to assist with home MV. Use of competent unlicensed attendants helps relieve the burden of care to family members and helps spare many patients from being forced to live in institutional care facilities (SNF). The majority of family caregivers indicated that the burden of care was heavy. However, despite this, family members often continued to be willing caregivers to keep their loved ones alive and at home. In contrast, none of the nasal-IPPV patients used nurses to help with regular home care. Using nasal-IPPV when this is possible is more cost effective than tracheostomy-IPPV due to lower costs for both equipment and caregivers. The main medical cost for nasal-IPPV was the monthly rental of the ventilator. For tracheostomy-IPPV, the ventilator, suction equipment and supplies were more expensive, complex, cumbersome and heavier than was equipment for the nasal-IPPV.

All the non-bulbar users of nasal-IPPV experienced relief of their respiratory symptoms. The non-bulbar patients enjoyed the benefits of talking and swallowing, compared to the bulbar patients who use MV. Seven of the 15 non-bulbar patients are still living, although one has chosen tracheostomy-IPPV. Two non-bulbar patients who successfully used nasal-IPPV, ranging 6–9 months, reported "not getting enough air" shortly before they died unexpectedly. They had not had regular assessment of their ventilator management. Some patients who did not like using nasal-IPPV, withdrew from MV and died. Four patients began use of the nasal-IPPV despite bulbar signs. All four of these patients were unable to tolerate its use except for short intervals.

All nasal-IPPV users were eligible for hospice services. The non-bulbar patients did not require suctioning. The care involved in using nasal-IPPV is less intensive than tracheostomy-IPPV which requires suctioning, even when no bulbar involvement is present.

Those who seriously wish to use long-term MV, after careful evaluation and counseling, benefit from starting nasal-IPPV before respiratory crisis occurs, if bulbar impairment is mild or absent. Compared to tracheostomy-

IPPV, ALS patients using nasal-IPPV: have planned this in advance and initiate it electively; are able to make an informed choice as to whether to stop MV, or to continue nasal-IPPV, or to use tracheostomy-IPPV if significant bulbar impairment occurs; have lower costs and less complex care; are eligible for hospice services; and are usually (92%) able to remain at home.

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