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Letter to the Editor

Ventilatory support in Japan: A new life with ALS and a positive approach to living with the disease

Although there has been much progress in understanding ALS in recent years no effective cure is yet available. Nonetheless, management of ALS has improved considerably during this time. In Japan there has been a particular emphasis on the provision of ventilatory support and here we report some aspects of the Japanese experience.

In 1972 the Japanese Ministry of Health and Welfare (MHW) published a guideline on "Counter Measures in Incurable Diseases" and the following year a research group led by Tadao Tsubaki was set up to organize care for people with ALS. In October 1974 ALS was categorized by MHW in Japan as an incurable disease. In 1982 MHW reported on the experience of ventilatory support to that date.[1] In a survey 472 patients with ALS responded, of whom 63 (13%) were using a ventilator. The survival rate after one year was 8% but survival beyond one year was rare.

A survey conducted in 1997 by the Japanese Study Group on Care and Management of PALS [people with ALS] and Similar Diseases, headed by Takeshi Sato, estimated that of a total of 4430 people with ALS (PALS) in Japan, 948 (21.4%) were using a ventilator.[2] Of PALS not using a ventilator, 61.3% died within three years. Of those using a ventilator, 52.6% had died after eight years (Figure 1). A nursing investigation on a neurological ward suggested that 52% of PALS hoped to use a ventilator, of whom 60% hoped to use home mechanical ventilation.

The Japanese experience suggests that people with ventilatory support can live with ALS for over 10 years at home. Indeed, many of these people play important roles at the Japanese ALS Association and enjoy a meaningful life, with social participation through the Association; although in several PALS, complications have developed, particularly pneumonia and intestinal obstruction.

The Japanese ALS Association (JALSA) was established in 1986.[3] It has encouraged awareness of the plight of PALS and their families and has encouraged changes in medical policies of Government level in relation to the care of PALS (Table 1). JALSA has worked with MHW in improving care facilities and financial support systems for PALS in Japan. Ventilatory care has become possible by leasing equipment and by the application of medical insurance during the 1990s through MHW for all PALS in the case of home mechanical ventilation, thus making it possible for many PALS to opt for home mechanical ventilation (HMV). The difference in uptake of HMV before and after the costs were covered by medical insurance is demonstrated in Table 2, representing a survey carried out by the Kinki Branch of JALSA.[4]

Attitudes to ALS with respiratory failure

In the past many PALS, and the medical profession itself, in Japan and elsewhere, thought of life with ALS after respiratory failure as impossible. This attitude has been changed by a new positive view of the role of a ventilator in ALS.[5] Much of this change in attitude derived from the experience of PALS with ventilator usage at the Tokyo Metropolitan Neurological Hospital, where a ventilator was introduced in 1987. It has become apparent, as a

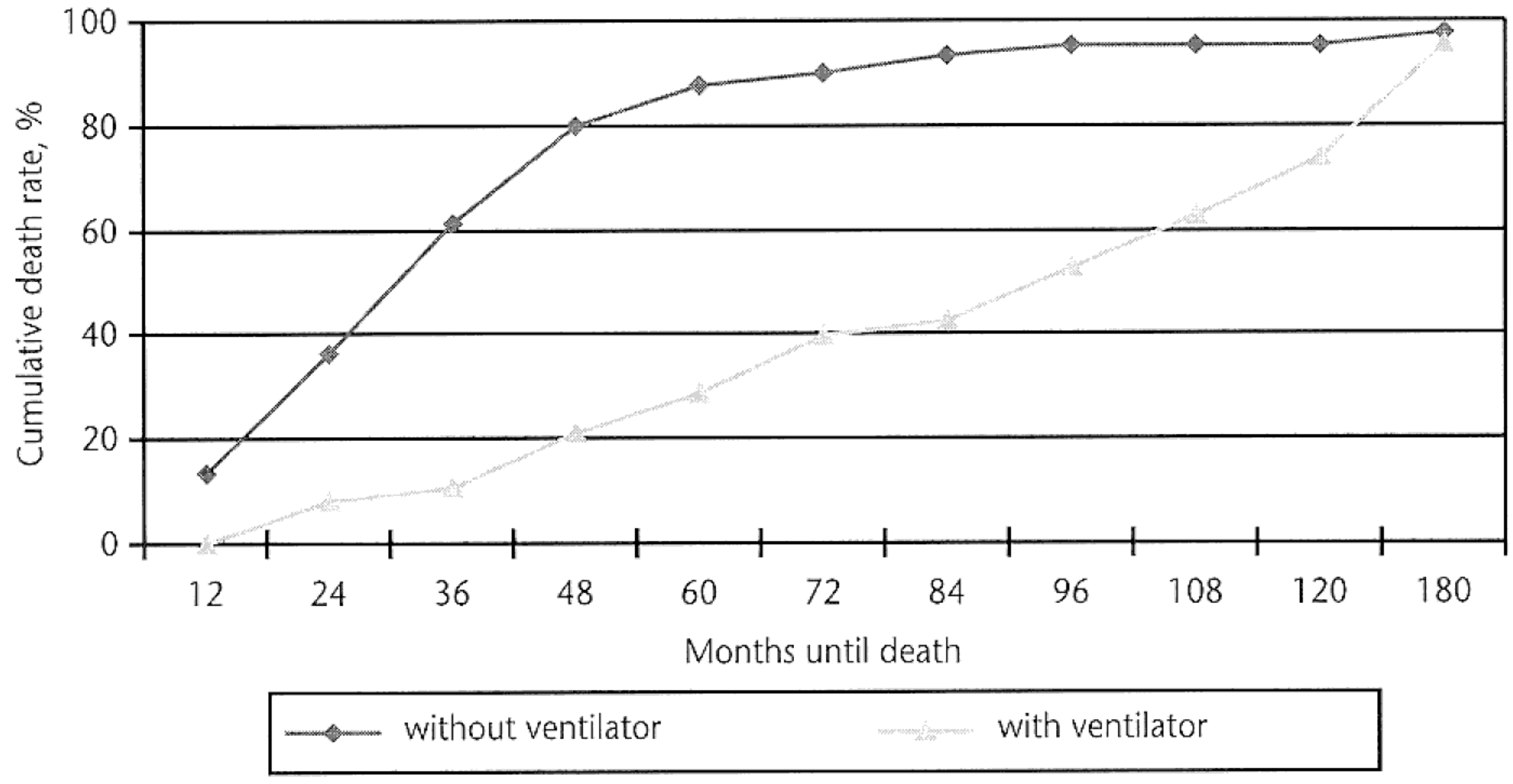

Figure 1
Prolongation of life of PALS with ventilator

Year	'86	'87	'88	'89	'90	'91	'92	'93	'94	'95
Negotiation with government	●	●	●	●	●	●	●	●	●	●
Publication of journal	●	●	●	●	●	●	●	●	●	●
Mutal encouragement	●	●	●	●	●	●	●	●	●	●
Negotiation with political parties								●	●	●
Awareness campaign		●					●	●	●	●
Support of unorganized area	●	●	●	●	●	●	●	●	●	●
Handbook of care						●				
ALS Fund										●

Table 1

Activities of the Japanese ALS Association in its first decade

Time of survey	1992 Nov.(1)	1996 Dec.(2)	2000 Oct.(2)
Responded PALS	72	159	176
Average age, years	57.4	58.7	60.2
PALS in hospital	22 (30.6%)	41 (25.8%)	35 (19.8%)
PALS at home	50 (69.4%)	118 (74.2%)	135 (76.7%)
PALS with ventilator	29 (40.0%)	80 (50.3%)	98 (56.0%)
PALS who selected HMV	13 (18.0%)	50 (31.4%)	71 (40.3%)

Table 2

Comparison of states of health of PALS before (1) and after (2) introduction of medical insurance

consequence of this change in management, that respiratory failure is not the end-point of ALS, but is only one of many impairments that occur in that disease. In Japan, JALSA particularly encourages the view that PALS should be treated not as patients but as severely disabled persons. This postive understanding of ALS has gradually become accepted by physicians and paramedical personnel in Japan and elsewhere during the last 20 years.[6]

Several PALS have made great contributions to the more general acceptance of ventilatory support in the management of ALS in Japan. Not only have such persons proved the effectiveness of HMV in the long term, but they have also shown, as living models, how meaningful life can be after respiratory failure in ALS. Mr X, has lived with ALS, with a ventilator, since 1988. He and his wife have shown that PALS can participate in social activities with MHW, provided there is positive acceptance of the disease and its disability. They have shown that if there is enough care, life with dignity is possible in spite of ALS. Mr X has continued to work, has been a leader and a personal counsellor for many PALS and their families.

On the basis of the experience in Japan of the utilization of mechanical ventilation in ALS we suggest the following.

1. PALS can live even after respiratory failure with appropriate ventilatory support.
2. Meaningful life after respiratory failure in ALS is possible if there is enough care, and there is positive acceptance of the disease and its disabililty.
3. There have been three important factors leading to this outcome.
    a. The role of the Japanese ALS Association in campaigning and providing support.
    b. A positive acceptance of this new view of ALS after respiratory failure.
    c. The contribution of pioneers among PALS.
4. The popularization of ventilatory support in ALS in Japan has implications for the organization of health care. These are insufficiently understood at present. There are also implications for the human rights of those using ventilation, and for those in whom, for various reasons, this management has not been made available.

It is important to recognize that the quality of life of people with ALS using ventilatory support is of cardinal importance. This is currently subject to qualitative judgement rather than quantitative assessment. More research is required in this area.

We hope to report later on these and other aspects of the implications of the usage of chronic ventilatory support in ALS.

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References 1–4 are Japanese only.