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Advanced Amyotrophic Lateral Sclerosis

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Introduction

Amyotrophic lateral sclerosis (ALS) is a progressive disease that causes respiratory muscle paralysis, eventually leading to death or the use of mechanical ventilation for survival. The median ventilation-free survival time for ALS patients in Japan is four years. Non-invasive and invasive ventilation, on the other hand, can increase survival time; a recent national surveillance report in Japan found that tracheostomy and invasive ventilation (TIV) increased survival time in patients with ALS by more than ten years [1].

Description

Using death or tracheostomy as endpoints, researchers identified several prognostic factors for survival in ALS patients. These factors include onset age, body region affected, diagnostic delay, progression rate of the revised ALS functional rating scale (ALSFRS-R) score from onset, and respiratory, bulbar, nutritional, and psychological factors [2]. In terms of nutrition, a low BMI at diagnosis, a rapid decline in body weight, and hypermetabolism in the early stages predicted a short survival time or the need for a tracheostomy. Recent evidence suggests that the hypothalamus is involved in lesions that cause weight loss or hypermetabolism.

In contrast to early-stage prognostic factors, there have been few studies on the predictors of functional prognosis in patients with advanced ALS who are undergoing TIV. The rapid progression rate from onset to TIV initiation foreshadows the development of severe communication impairments, including a completely locked-in state. With long-term TIV use, rapid weight loss until tracheostomy predicts a functional decline in the advanced stages [3]. In clinical settings, however, we frequently encounter ALS patients who gain significant weight while using TIV. Assuming that the weight loss in the early stages is the result of non-motor system neurodegeneration, including in the hypothalamus, weight gain or other nutritional problems in the advanced stage of TIV could also be the result of advanced non-motor neurodegeneration in ALS [4].

Long-term TIV use increased body weight in ALS patients. Additionally, weight gain after TIV was linked to an initial weight loss prior to TIV and the disease stage (communication impairment) during TIV use. Patients who developed the most severe communication impairment (totally locked-in state) during TIV had the fastest progressive weight loss prior to TIV, the lowest BMI during TIV use, and the greatest weight gain at the end of TIV use. Despite a significant weight gain, these patients had low energy intake and serum albumin levels [5].

Conclusion

Because the skeletal muscles in patients with ALS are thought to be almost completely destroyed during a long-term TIV, the main component of the weight gain could be fat. Previous studies of energy metabolism in ALS patients found that those using TIV had significantly lower energy expenditure (700 to 1000 kcal/day), indicating "hypometabolism," which could lead to fat accumulation and weight gain. Patients with the most advanced stage of communication impairment (stage V) had the lowest energy intake in this study. They did, however, show the greatest increase in weight, implying that weight gain is pathognomonic for ALS and is associated with central metabolic dysregulation.

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