Motor neuron disease (MND) is a progressive neurodegenerative disease characterized by progressive weakness of the limb, axial, bulbar, and respiratory muscles. The survival duration for patients with MND is usually as short as 2–3 years from the onset of symptoms. Although physicians cannot cure or halt the progression of MND, there is much that can be done to improve patient quality of life by managing the physical and emotional symptoms of MND.1

A previous study investigating the causes of death in 302 patients with amyotrophic lateral sclerosis, the major type of MND, demonstrated that the most frequent cause of death was respiratory failure.2 Therefore, the existence of dysphagia or dyspnea is commonly understood as an indicator to commence the palliative care plan for patients with MND. However, little evidence is available in the published literature that identifies the optimal approaches for patient end-of-life care in MND.3

Owing to the differences in pathogenesis and therapeutic options of various diseases, the timetables available to discuss end-of-life care planning for patients and their families may vary depending on the patient’s diagnosis. In contrast to other terminal or neurodegenerative diseases, patients with MND usually retain sufficient mental clarity to make decisions during the end of life stage. Astrow et al4 examined a documented discussion about the prognosis of patients with advanced cancer and MND. They found that MND patients were more likely than end-stage cancer patients to have a chance to deliberate and state their intentions in wills that address such issues as ventilation support, artificial nutrition, and resuscitation. Therefore, physicians have the obligation to provide adequate information to help patients make better decisions in these challenging phases of life.

Bulbar symptoms, such as dysphagia and dysarthria, usually are signs indicating the progression to the final stage of MND. Nutrition becomes an important issue in patient care when dysphagia starts. Failure to maintain adequate nutritional status is an established adverse prognostic factor in MND, and gastrostomy is a safe and practical alternative to oral feeding. Although there is no clear evidence that life can be extended, evidence has shown that gastrostomy may improve the quality of life of MND patients with bulbar symptoms.5

Respiratory muscle function may develop incipiently over an extended period and rarely presents with catastrophic symptoms in an emergency situation. Therefore, patients usually can take some time to make their choice about ventilation support. In a randomized controlled trial, noninvasive ventilation (NIV) improved the survival and quality of life of MND patients without severe bulbar dysfunction. In patients with severe bulbar impairment, NIV still could improve sleep-related symptoms but was unlikely to confer a substantial survival advantage.6

Although evidence has strongly supported the advantage of using gastrostomy or NIV, the number of patients with MND receiving these interventions varies from country to country. In this issue of the Journal of the Chinese Medical Association, Kang et al7 report the utilization of hospice care among patients with MND in Taiwan. Based on the National Health Insurance Research Database during 2005–2010, the utilization of hospice care was extremely low among MND patients in Taiwan. There were only 30 MND patients receiving palliative care during this 5-year period.

Similar to reports in Caucasian populations, respiratory problems were the major causes of death in Taiwanese MND patients. However, there was no patient receiving gastrostomy, and only 13 (23.6%) patients received NIV intervention. According to a study conducted in the United Kingdom, there is a 3.4-fold increase in the rate of applying NIV in MND patients from 2000 to 2009.8 Furthermore, NIV also can be used in home care to improve the quality of life of end-stage MND patients.9 Therefore, there is still considerable leeway for physicians to make an effort to promote hospice and palliative care of MND patients in Taiwan.

By contrast, Kang et al noted that family physicians provided most (78.2%) of the inpatient hospice care for MND patients in Taiwan. Specialists with different backgrounds may cause significant variation in palliative care plans.10 Quality communication and cooperation between working staffs with different medical background can greatly improve the quality of palliative care.11 It is important to assemble a multidisciplinary team to develop a collaborative approach in the palliative care of MND.

Taiwan National Health Insurance has, for many years, approved MND for hospice care payment; however, very few MND patients utilize this service in Taiwan. Consequently, physicians involved in palliative care should make a greater effort to promote the quality of end-of-life care for MND patients.
References


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