Does Tracheostomy Remain an Option in Neuromuscular Patients?

Numerous patients with a progressive neuromuscular disease are characterized by a decline in respiratory muscle performance, which results in progressive respiratory failure, initially during sleep and subsequently during the day.[1] At this point, daytime hypercapnia and symptoms related to sleep-disordered breathing can often be corrected with nocturnal noninvasive ventilation (NIV). As the respiratory muscle weakness progresses, the patient can become more dependent on ventilation, at which time NIV should be extended during daytime.[1] While daytime ventilation can be delivered with the same interface used at night, a mouthpiece is preferred when daytime ventilation becomes continuous. This allows the patient to easily connect and disconnect from the ventilator, depending on social activities and respiratory sensations.[2] The success of this long-term NIV technique depends mainly on the patient's ability to clear secretions from their airways, to avoid aspiration, and to maintain their nutritional status. Indeed, the medical team should be familiar with an array of techniques to optimally tailor cough-enhancement techniques to each patient, to teach these techniques to home caregivers, and, if severe dysphagia is present, to consider a gastrostomy before substantial weight loss occurs.[1]

Accordingly, the improving knowledge of NIV techniques, especially mouthpiece ventilation for diurnal ventilation, has allowed its use among patients who are completely ventilator-dependent. When combined with systematic cough-assistance management, this therapy has been effective at reducing the indication of invasive techniques such as tracheostomy.[2-4]

The experience of Suh et al[5] presented in this issue showed a good tolerance and effectiveness of NIV despite the progressively increasing duration of ventilation for most subjects with neuromuscular disease. One notable exception was amyotrophic lateral sclerosis (ALS) subjects, who were tracheostomised when severe bulbar symptoms may risk maintaining NIV. Although the survival proportion and tracheostomy-free period after NIV application were possibly overestimated considering that 28% of the subjects were lost during the follow-up, this result remains consistent with observations from other industrial countries.[1,6] It was asserted that NIV improved life expectancy in subjects with Duchenne muscular dystrophy compared to invasive ventilation.[4] However, that comparison was made with a historically tracheostomized population, and other studies have found similar life expectancy when tracheostomy remained an option in a similar population.[7,8]

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Another limit of the study by Suh et al[5] is the lack of evaluation of quality of life. While it has historically been argued that quality of life was better in a non-tracheostomized condition because NIV methods were considered to have less negative impact on speech, appearance, and comfort,[9] recent clinical evaluations suggest that invasive mechanical ventilation can improve speech[10,11] and swallowing.[12,13] In a survey of subjects on long-term ventilation, more than two thirds of subjects who had a tracheostomy were satisfied with their lives, and 84% thought they had made the right choice.[14] Moreover, Hutmann et al[15] recently found similar quality of life in noninvasively and invasively ventilated subjects with neuromuscular disorders.

While NIV is undoubtedly the first-line treatment for restrictive respiratory failure, patients may experience difficulty using a mouthpiece over time, decreased tolerance of prolonged nasal ventilation, and upper airway dysfunction or bulbar dysfunction with risks of aspiration despite adequate cough assistance. In these cases, tracheostomy should remain an option.[6] Indeed, recent guidelines retained an indication for tracheostomy in case of the failure of NIV to maintain respiratory function, bulbar problems, an inability of the local medical infrastructure to support NIV, and patient preference.[16-18]

Over the past decades, respiratory care has improved in all industrial countries, largely due to the development of noninvasive techniques of mechanical ventilation. However, the literature suggests that important disparities in respiratory care management remain across the world, which could be explained by differences between teams' experience and health care systems. For instance, in France, medical care is supported by the state health care system, which can finance professional care provided at home for dependent

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patients; moreover, thanks to the efforts of associations such as the Association Française Contre les Myopathies-Téléthon, families are well informed and can be trained in the care necessary to allow ventilation at home. This simplifies the return home of tracheostomized patients. In the United States, tracheostomized patients also can often return home with their family members as caregivers, but some teams consider that they can only do so if they have skilled nursing care 16–24 h per day, which might not be covered by their insurance policy.[19] In such situations, tracheostomy may be an impossibly expensive alternative for the patient and the patient's family; the alternative would be to discharge patients to a long-term acute care facility.[19] Curiously, in France, the use of tracheostomy tends to be avoided in patients with ALS.[20] The fear of a "locked-in syndrome," the high burden placed on caregivers, and the unmasking of cognitive disorders occurring in the evolution of ALS are among the limitations considered for tracheostomy.[20] This is supported more negative or ambiguous opinion of ALS patients toward invasive ventilation and lower life satisfaction scores when compared with patients with Duchenne muscular dystrophy.[14]

Therefore, while NIV is at the forefront of restrictive respiratory failure treatment and has proven to be a valuable and efficient treatment for ventilator-dependent patients, as reported by Suh et al,[5] tracheostomy remains an alternative technique for patients in whom NIV has become inefficient or is not well tolerated. The choice of the ventilation technique depends on multiple complex factors that include, among others, access to health care and the patient's environment and preference; this decision cannot rely on dogma alone and needs to be tailored to a patient's specific situation.

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