

# Long-term noninvasive ventilation in muscular dystrophy: Need planning of future services

Chronic Respiratory Disease  
2017, Vol. 14(2) 194–195  
© The Author(s) 2016  
Reprints and permission:  
sagepub.co.uk/journalsPermissions.nav  
DOI: 10.1177/1479972316679680  
journals.sagepub.com/home/crd



Giuseppe Fiorentino<sup>1</sup> and Antonio M Esquinas<sup>2</sup>

Dear Editor,

No curative treatment for muscular dystrophy (MD) is yet known, but advances in management over the last two decades have altered the natural history of MD. Patients with MD often remain on noninvasive ventilation (NIV) for longer than other patient should be taken into account when planning service requirements. The improvements in general care and the frequent provision of NIV from 1990s have improved the mean survival of patients with Duchenne Muscular Dystrophy (DMD).<sup>1</sup> However, in order to provide an adequate service for patients with MD, there is very little epidemiological data in the current literature on which to base any assessment of service requirements. We have read with great interest, results of long-term NIV in MD in use over 25 years in a home ventilation unit.<sup>2</sup> This highlights great experience and results that how NIV may improve outcome; however, we consider that some key practical points are essential to take into account for a proper clinical extrapolation of this results.

First, regarding survival rate and new referrals need are key issues. In this article, Kinnear et al. examined the trend in MD use of home-NIV in their unit over the last 25 years; however, the number of new referrals appears to be stabilizing at around 20–25 over a 5-year period, equivalent to approximately 0.5 per 100,000 of population per year. This estimate is higher than the incidence figure of 0.2 per 100,000 which we derived previously from national death certification data.<sup>2</sup> The mean age at commencement of home-NIV is now 37.5 years, with 5-year survival rates of 70–75%. Ten-year survival rates are just over 40%.

Second, few data have been published on the quality of life of patients with MND receiving home

mechanical ventilation<sup>3</sup> and their caregivers.<sup>4</sup> Progression in DMD is inevitable, leading to a state of severe physical dependence. When the patient gets older and the need for assisted ventilation increases, does more patient-centered care become necessary. NIV is usually more demanding than invasive ventilation, in particular when the patient has a broncho-pulmonary infection.<sup>4</sup> In selected neuromuscular disease (NMD) patients (no amyotrophic lateral sclerosis) with severe respiratory tract infection, we can consider “Hospital at Home” treatment. It is a service that provides active treatment by health-care professionals in the NMD patient’s home for a limited period.<sup>5</sup> This kind of treatment is very important that family members have become an integral part of severely disabled NMD patient with an improvement of quality of life and cost containment. This is a controversial hot topics that need more precise evaluation in this study such as trends in adaptations to housing, electric beds, and wheelchairs; access to the internet and computers is essential for independent and functional life. Further prospective long-term trials need to define or confirm best planning and future services for MD.

## References

1. Simonds AK. Recent advances in respiratory care for neuromuscular disease. *Chest* 2006; 130: 1879–1886.

<sup>1</sup> Respiratory Unit, AO Ospedali dei Colli Monaldi, Naples, Italy

<sup>2</sup> Intensive Care Unit, Hospital Morales Meseguer, Murcia, Spain

## Corresponding author:

Giuseppe Fiorentino, Respiratory Unit, AO Ospedali dei Colli Monaldi, Naples, Italy.

Emails: giuseppe.fiorentino@ospedaliideicolli.it;

giuseppEFIorentino1@gmail.com

2. Kinnear W, Colt J, Watson L, et al. Long-term non-invasive ventilation in muscular dystrophy: trends in use over 25 years in a home ventilation unit. *Chron Respir Dis* 2016. Epub ahead of print 21 June 2016. DOI: 10.1177/1479972316654285.
3. Lloyd-Owen SJ, Donaldson GC, Ambrosino N, et al. Patterns of home mechanical ventilation use in Europe: results from the Eurovent survey. *Eur Respir J* 2005; 25(6):1025–1031.
4. Calvert LD, McKeever TM, Kinnear WJ, et al. Trends in survival from muscular dystrophy in England and Wales and impact on respiratory services. *Respir Med* 2006; 100: 1058–1063.
5. Vianello A, Savoia F, Pipitone E, et al. “Hospital at home” for neuromuscular disease patients with respiratory tract infection: a pilot study. *Respir Care* 2013; 58(12): 2061–2068. Epub ahead of print 21 May 2013. DOI: 10.4187/respcare.02501.