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Title: Non-invasive ventilation in neuromuscular diseases. A 15-year retrospective study

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Body: Non-invasive ventilation (NIV) has been shown to reduce respiratory morbi-mortality in neuromuscular disorders (NMD). We conducted a retrospective study including NMD patients in whom long term NIV was instituted in our centre over the last 15 years. Methods: All NMD patients (p) in whom NIV was started between 1997 and 2011 were included. They were classified in 3 groups regarding the pattern of NMD: rapidly progressive (RP), early-onset myopathies (EOM) and late-onset myopathies (LOM). Clinical, epidemiological and lung function data at the time of NIV were collected. Criteria leading to introduce NIV and prognosis were analyzed. Results: Eighty seven patients (63% male, age 58±16 y) were included in the study; 55 (63%) had RP (age 65±11 y), 24 (28%) LOM (age 55±12 y) and 8 (9%) EOM (age 21±4 y). Time (in years) between diagnosis and NIV was 3.2±4 (RP), 26±13 (LOM) and 18±6 (EOM). In 17 (19.5%), NIV was begun during an acute episode (13 RP, 3 LOM, 1 EOM). In the remaining cases, NIV decision was based on at least one of the following criteria: hypercapnia (38 p, 44%), symptoms (35 p, 40%), lung test abnormalities (27 p, 32%), nocturnal SpO2 (28 p, 32%) or tcPCO2 (16 p, 18%) abnormalities. In sixty patients (69%) NIV decision was based on more than one abnormality. In eighty six patient (99%) NIV was initiated by using a pressure mode. Length of time on NIV (months) before death or tracheostomy was 17±15 (RP), 78±47 (LOM) and 84±45 (EOM). Conclusion: NIV is still initiated during an acute episode in 19.5% of our patients. In the remaining cases, symptoms and diurnal hypercapnia were the main factors leading to NIV institution; but in most cases NIV decision was based on more than one abnormality.