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Title: Long-term adherence to domiciliary NIV and its relation to survival in patients with chronic respiratory failure

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Body: Few objective data are available about home noninvasive ventilation (NIV) adherence and its relation to survival in patients with chronic respiratory failure (CRF). This observational cohort study was aimed to determine NIV compliance, its predictive factors and its relation to long-term survival in CRF patients. We have included 480 consecutive CRF patients who were initiated to long-term NIV between January 2000 and December 2008. 156 (32%) patients had obesity hypoventilation syndrome (OHS), 112 (23%) had restrictive pulmonary disease including neuromuscular disease (NM, n=30), amyotrophic lateral sclerosis (ALS, n=31) or kyphoscoliosis (KS, n=51), 91(19%) had chronic obstructive pulmonary disease (COPD), 92 (19%) had overlap syndrome (OS) and 29 (6%) had others CRF etiologies (ORF). The mean compliance was 6.8±0.3 h/d. Daily adherence was similar between etiologic subgroups and not related to sociodemographic, baseline clinical data or conditions of NIV initiation. Compliance did not vary significantly during the mean 32.2±2.9 months of follow-up. Median survival was 4.7 yrs for COPD; 9.3 for OHS; 6.2 for NM; 0.6 for ALS; 6.9 for KS and the 75th percentile was 6.35 for OS. Independent predictors of mortality were a higher age and the following CRF causes: COPD, NM, ALS and ORF (p<0.01). In contrast, a higher adherence to NIV was a mortality protector factor in COPD (OR=0.70), OHS (OR=0.81), NM (OR=0.54) and KS (OR=0.77) subgroups (p<0.05). Neither NIV initiation conditions nor home oxygen use was linked to survival. In conclusion, this study did not identify predictive factors of adherence to NIV but showed a significant impact of adherence on long-term survival.