

## Abstract

### Studies of patients with chronic alveolar hypoventilation.

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Chronic alveolar hypoventilation (CAH) is a condition caused by limitations in the respiratory pump system, resulting in hypercapnic respiratory failure. Mechanical ventilation is the therapy of choice.

*Aims:* to describe the prevalence of patients with CAH treated with home mechanical ventilation in Sweden, explore indications and types and modes of ventilation; to investigate patients with CAH to describe their health-related quality of life (HRQL); to study the effects on HRQL of non-invasive positive pressure ventilation (NPPV); to prospectively investigate survival in patients with CAH, evaluate prognostic importance of HRQL for survival and investigate long-term effects of NPPV on blood gases and HRQL; to investigate if improvements seen in daytime blood gases after NPPV can be explained by improved ventilatory drive.

*Methods:* registration forms were sent to all clinics known to take care of patients with home mechanical ventilation in Sweden.

Consecutive patients (n=44) referred for evaluation of CAH were investigated by HRQL questionnaires, spirometry, blood gases and ventilatory response to CO<sub>2</sub> rebreathing before and after 9 months of treatment with NPPV. At long-term follow up after 8 years, the investigations were repeated, apart from the ventilatory response to CO<sub>2</sub> rebreathing. Preferably non-parametric statistical analysis was used, and long-term survival and its predictors were analysed by conventional methods.

*Results:* the prevalence of home mechanical ventilation in 1996 was 6.1/100 000 inhabitants with great variations among our health care regions. The underlying diagnoses were neuromuscular diseases, "previous polio", chest wall deformities and other disorders in approximately equal proportions. Most patients were ventilated non-invasively and only during night.

Patients with untreated CAH were severely affected in many vital areas of HRQL, especially those relating to physical function and sleep. Age, underlying disease and standard bicarbonate correlated with HRQL measurements. After 9 months treatment with NPPV, improvements were seen in blood gases and HRQL, especially in condition-specific measurements but also in physical function and emotional behaviour. Effectiveness in ventilation correlated with these improvements. 1-, 5- and 8-year survival rates were 89%, 68% and 47%, respectively. Improvements in blood gases and HRQL persisted at long-term follow-up. Survival was predicted by diagnosis, age and HRQL at start of treatment.

Ventilatory response to CO<sub>2</sub> rebreathing was low in patients before treatment. Among patients with substantial improvement in daytime PaCO<sub>2</sub> we found significant improvements in ventilatory response.

*Conclusions:* The prevalence of home mechanical ventilation is increasing but varies between our health care regions. Patients with untreated CAH have impaired HRQL. Age, diagnosis and severity of hypoventilation relate to these impairments. Blood gases and HRQL improve after treatment with NPPV. Improved blood gases and HRQL remain for long-term. Diagnosis and HRQL at start of treatment are predicting long-term survival.

*Key words:* chronic alveolar hypoventilation, non-invasive positive pressure ventilation, health related quality of life, observational prospective longitudinal study, prevalence, ventilatory response to CO<sub>2</sub>-rebreathing, long-term survival.

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