

Non-invasive ventilation (NIV)

There are indications that early non-invasive positive pressure ventilation increases survival compared to late NIPPV (Chio and others 2009). Non-invasive ventilation (NIV) provides relief from symptoms such as fatigue, breathlessness and disturbed sleep patterns, but does not prevent progressive weakening of the respiratory muscles (Andersen and others 2007).

Orrell 2010

An important intervention, which clinical experience suggests is beneficial for patients with ALS/MND, is non-invasive ventilation.

Miller and others 2009a

Does NIV improve respiratory function or increase survival?

- In a randomized controlled study, patients using NIV experienced a median survival benefit of 205 days (Class I). NIV was initiated based on orthopnea with an MIP <-60 cm or symptomatic hypercapnia. No survival benefit was seen in patients with poor bulbar function.
- "Early" intervention with NIV (nocturnal oximetry demonstrating >15 desaturation events/hour) resulted in 11 months longer survival compared to controls, with some beneficial effect in bulbar patients (Class III). Patients who used NIV >4 hours/day survived 7 months longer than patients using the device <4 hours/day (Class III).
- FVC declined more slowly after introducing NIV (pre -2.2% /month compared to post -1.1% /month) (Class I/III)² and the decline was slower in those who used NIV >4 hours/day (Class III). A survival benefit of 20 months was observed in NIV-tolerant patients vs 5 months in NIV-intolerant patients (Class III).
- *Conclusion*
 - NIV is probably effective in prolonging survival (1 Class I, 3 Class III studies) and in slowing the rate of FVC decline (1 Class I, 1 Class III study).
- *Recommendation*
 - NIV should be considered to treat respiratory insufficiency in ALS, both to lengthen survival and to slow the rate of FVC decline (Level B).

How do invasive and noninvasive ventilation affect quality of life?

- NIV had a positive impact on quality of life (QOL) in 4 Class III studies. There was improvement in energy, vitality, shortness of breath, daytime somnolence, depression, concentration problems, sleep quality, and physical fatigue for 10 months or more. In one Class III study, patients using NIV had increased duration of QOL above 75% of baseline and increased time-weighted mean improvement in QOL.
- There was no difference in QOL between those using NIV and patients with tracheostomy invasive ventilation (TIV) (Class III). Most patients using either NIV (94%) or TIV (81%) would choose ventilation again. Caregivers of patients using TIV, however, rated their own QOL lower than that of their patient. Another series of 7 patients using TIV rated their general health as good based on the SF-12[®] Health Survey and none of the patients regretted his/her decision (Class III).
- *Conclusions*
 - NIV is possibly effective in raising QOL for patients with ALS who have respiratory insufficiency (5 Class III studies).
 - TIV is possibly effective in preserving QOL for patients with ALS, but possibly with a greater burden

for their caregivers (2 Class III studies).

- *Recommendations*
 - NIV may be considered to enhance QOL in patients with ALS who have respiratory insufficiency (Level C).
 - TIV may be considered to preserve QOL in patients with ALS who want long-term ventilatory support (Level C).

What factors influence acceptance of invasive and noninvasive ventilation?

- Compliance with NIV was improved when treatment was initiated early based on the presence of at least 15 desaturation events per hour (Class III). A randomized pilot trial of early NIV intervention (2 desaturation events <90% for 1 cumulative minute, mean FVC = 77%) showed that 7 of 7 patients were compliant (Class III). This degree of compliance is much higher than prior reports (Class III) of less than 50% when NIV was initiated based on prior recommendations.
- Noncompliance with NIV was seen in 75% of patients with ALS and frontotemporal dysfunction vs 38% in patients with classic ALS (relative risk 2.0) (Class III). There was low compliance in bulbar patients (Class III) but cognitive/executive function was not described.
- Orthopnea was a strong predictor of benefit and also better compliance with NIV (Class III). NIV use correlated with symptoms of orthopnea and dyspnea as well as with the use of PEG, speech devices, and riluzole (Class III). Young age and preserved upper limb function also predicted better compliance.
- *Conclusions*
 - Nocturnal oximetry is possibly effective in detecting early respiratory insufficiency and the early use of NIV possibly increases compliance (2 Class III studies).
 - Bulbar involvement and executive dysfunction possibly lower compliance with NIV (2 Class III studies).
- *Recommendation*
 - NIV may be considered at the earliest sign of nocturnal hypoventilation or respiratory insufficiency in order to improve compliance with NIV in patients with ALS (Level C).

Chio and others 2009

Although two controlled studies have demonstrated that NIPPV confers a significant advantage to ALS patients in terms of median survival (increase of 205 days) and quality of life, surprisingly NIPPV has not been widely studied as a prognostic factor. In a population-based study NIPPV did not modify overall survival, but this finding was explained by the low number of patients who underwent this intervention. There are indications that early NIPPV increases survival compared to late NIPPV.

MND Australia 2008

Life support via continuous ventilation with a tracheostomy (invasive ventilation) is available but is not an easy decision to make. Detailed discussions should be held with regard to this early in the progression of the disease, considering the significant quality of life issues in the face of progressive weakness.

It is important to discuss the process and timing of withdrawal of NIPPV or invasive ventilation. Implications to consider:

- quality of life issues
- power of attorney/guardianship, financial affairs, medical treatment/advance medical directives, end of life care
- cost of and access to assisted ventilation

- increased dependency on family and carers - the carer should be included in discussions and decision making.

Few patients choose the option of invasive mechanical ventilation but some achieve good palliation of symptoms of CO₂ retention and shortness of breath using non invasive ventilation. These benefits are often sustained for many months or years. Ultimately, however, the breathing difficulties become life threatening