

BiLevel therapy for the MND patient

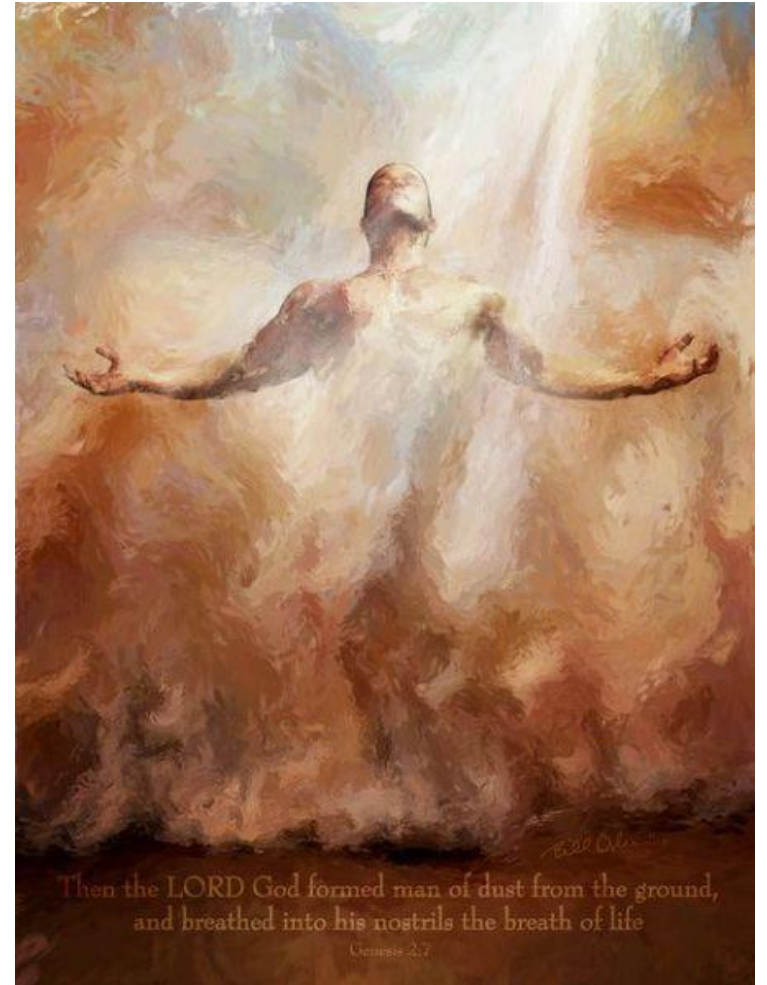
Bilevel ventilation is a mode of biphasic ventilation that allows the patient to breathe spontaneously together with delivered ventilator breaths.

WHY?

WHAT?

WHEN?

HOW?



WHY?

Non-invasive ventilation (NIV) has become an important cornerstone of symptomatic treatment in ALS, improving survival and quality of life.

- Recent retrospective studies with large numbers of patients ($n = 929$, $n = 114$, $n = 140$) found **survival benefits of 13 months,⁴ 11 months,⁵ and 15.5 months,⁶** respectively.
- It has also been shown that NIV **delays deterioration of respiratory function**
- Bourke and colleagues found that **quality of life**, as measured by a general and a sleep-dependent scale, **improved in patients with NIV**
- NIV **reduced daytime fatigue and depression**
- NIV **improved sleep**

Non-invasive ventilation in amyotrophic lateral sclerosis – Johannes Dorst and Albert C Ludolph – Ther Adv Neurol Disord 2019, Vol 12: 1-14

Bourke SC, Tomlinson M, Williams TL, et al. Effects of non-invasive ventilation on survival and quality of life in patients with amyotrophic lateral sclerosis: a randomised controlled trial. *Lancet Neurol* 2006; 5: 140–147. [[PubMed](#)]



WHAT?

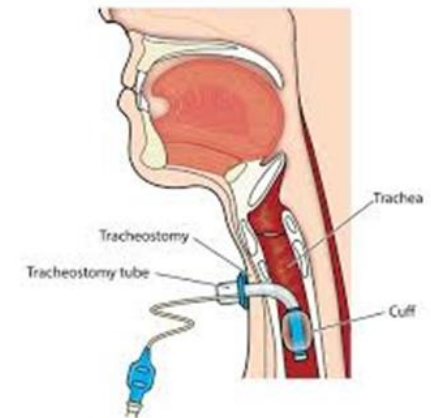
NIV vs IPPV

The aim of NIV is to compensate diaphragm weakness, alleviate hypercapnic symptoms and improve patients' general condition and quality of life. Since respiratory insufficiency limits survival, NIV also prolongs life considerably in ALS. In view of its non-invasiveness and good tolerability, NIV is regarded as an essential therapeutic component in ALS.

Dorst & Ludolph



INVASIVE VENTILATION	
ADVANTAGES	DRAWBACKS
Increases survival time	Increased bronchial secretions
Prevents aspiration	Increased risk of infection
More effective ventilator pressures	Risk of tracheo-oesophageal fistula
Better gas exchange	RO tracheal stenosis
	RO tracheomalacia
	Increased costs
	Increased carer & family burden
	Ethical issues - discontinuation



WHEN?

Respiratory complications are the main cause of death in ALS

EJN P.M. Anderson et al

- **DISCUSS** – As soon after initial diagnostic discussion as possible
- **INITIATE** - Taking into account recent findings, it seems advisable to initiate NIV as soon as any abnormalities in respiratory diagnostics are evident Dorst & Ludolph
- **USE** – Initially at night with intermittent day time use eventually leading to 24/7 EJN P.M. Anderson et al
- **DISCONTINUE** – At patient request

Proposed criteria for NIPPV

Symptoms/signs related to respiratory muscle weakness.

At least one of the following:

Dyspnoea; Tachypnoea; Orthopnoea; Disturbed sleep due to nocturnal desaturation/arousals; Morning headache; Use of auxiliary respiratory muscles at rest; Paradoxical respiration; Daytime fatigue; Excessive daytime sleepiness (ESS > 9)

Abnormal respiratory function tests.

At least one of the following:

Forced vital capacity <80% of predicted value
Sniff nasal pressure <40 cmH₂O
PI max <60 mmH₂O
Significant nocturnal desaturation on overnight oximetry
Morning blood gas pCO₂ > 45 mmHg

HOW?

MASK – The important question about which masks should be used has been completely neglected in clinical studies so far. Dorst & Ludolph

DEVICE - Few studies so far have investigated the effect of different ventilation modes and parameters on blood gases, clinical symptoms, and survival. Dorst & Ludolph

Ventilation parameters should be comfortable for the patient to ensure an acceptable quality of life, as well as sufficient compliance.

The goal of NIV in the MND patient is not to provide life support ventilation, but to positively affect QOL and yet allow for the natural death process.



References

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