

# Non-Invasive Ventilation (NIV) use in MND/ALS: a simple summary of the research evidence

## Introduction

Non-invasive ventilation (NIV) has been shown to help with some of the symptoms associated with MND/ALS. This includes poor sleep, tiredness and breathlessness. To find out how effective NIV could be for people living with MND various research studies have been done. The findings of the key studies are summarised here. Research is currently being done in the UK into the effectiveness of invasive ventilation for MND. As this uses similar ventilation, it is thought to offer similar benefits.

Understanding the research and the potential benefits it shows can help people to make 'evidence-based' choices about using NIV or not. Understanding this research evidence - along with reflecting upon your personal feelings - can help you to make the best choice for you.

## Randomised controlled trial of NIV

There are different types of research studies. The type considered to be the most reliable is called a 'Randomised Controlled Trial' (RCT). This compares a group people who use a new intervention with a group of people who receive standard care. It is randomly chosen which group people are put in.

The only RCT assessing the effectiveness of NIV in people with MND was conducted in Newcastle, UK, 2006 by Bourke and team (see reference 1). This trial established the effectiveness of NIV in MND. Because we know it works, it would be unethical to do another RCT where people are not given NIV.

### What did they do?

- 41 people were chosen at random to receive NIV with standard care, or standard care alone.
- In the analysis, people were separated into those with better 'bulbar function' and those with worse 'bulbar function' (problems with swallowing, eating and speech).

### What did they find?

- **NIV improved quality of life for all those who used it**, with good and poor bulbar function.
- For people with better bulbar function **NIV extended life on average by seven months**.

## Cohort study of people using NIV

A 'cohort study' looks at large numbers of people over several years. It sees how they are affected by interventions. It is considered less reliable than an RCT, as it is less strictly controlled. They are still useful indicators of effect. Berlowitz and team (see reference 2) undertook a cohort study in Victoria, Australia in 2015.

### What did they do?

- Reviewed the medical notes of 929 people with MND using NIV from 1991 and 2011.
- This was done 'retrospectively' - they looked back over time.

## What did they find?

- **NIV extended life on average by 13-19 months.**
- **People with both good and poor bulbar function both had extended life.**

## Systematic reviews of effective NIV use

'Systematic reviews' are when researchers examine several similar research studies together. This means they can see what findings they have in common and also where they differ. This helps to build a bigger picture to inform clinical practice.

Researchers from Sheffield Institute of Translational Neuroscience (SITraN) (UK) conducted two systematic reviews of several types of studies in 2019. These included RCTs and other types of research looking at the effective use of NIV in MND/ALS (see references 3 and 4).

## What did they do?

- Examined the research that looked at several factors affecting people using NIV, their carers, clinicians, services/teams and equipment.
- Included a wide range of study types to provide a complete summary of the available evidence.
- This resulted in some suggestions for clinicians who support people to use NIV in MND.

## What did they find?

- **Multidisciplinary care** (support from a range of collaborating healthcare professionals) is important in promoting good uptake of NIV and supporting its daily usage.
- **People with significant bulbar problems** can gain extended life and quality of life benefits from NIV. They may require greater support to ensure it is effective.
- **Mask selection** should be aimed at maximising comfort and minimising air leakage. Alternatives should be available where necessary.
- The **NIV machine, mode and settings** should be selected to promote comfortable use and effective ventilation. They may need adapting over time to as the disease progresses.
- **Regular monitoring** of people using NIV should help to identify any issues and make changes to the equipment as required.
- **Cough-assist machines** can help people bring up mucus from their lungs. These should be combined with NIV to maximise benefits where appropriate.

## References

1. Bourke SC, Tomlinson M, Williams TL, et al. (2006) **Effects of non-invasive ventilation on survival and quality of life in patients with amyotrophic lateral sclerosis: A randomised controlled trial.** Lancet Neurology. 5(2):140-7.
2. Berlowitz DJ, Howard ME, Fiore Jr JF, et al. (2016) **'Identifying who will benefit from non-invasive ventilation in amyotrophic lateral sclerosis/motor neurone disease in a clinical cohort.'** Journal of Neurology, Neurosurgery and Psychiatry. 87:280-286.
3. O'Brien D, Stavroulakis T, Baxter S, et al. (2019) **'The optimisation of non-invasive ventilation in amyotrophic lateral sclerosis: A systematic review.'** European Respiratory Journal. 54(3):1900261
4. Baxter SK, Johnson M, Clowes M, et al. (2019) **'Optimizing the noninvasive ventilation pathway for patients with amyotrophic lateral sclerosis/motor neuron disease: a systematic review.'** Amyotrophic Lateral Sclerosis and Frontal Degeneration. 20(7-8)461-472