Non Invasive Ventilation

Non-invasive ventilation (NIV)

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Background

Non-Invasive mechanical Ventilation (NIV) has been used since the late 1980s as an alternative first-line intervention to endotracheal intubation in acute and chronic respiratory failure. NIV, that is based on mechanical inflation and deflation of the lungs via a nasal or facial mask, has been shown to improve gas exchange, reduce respiratory muscle work and improve pulmonary function. NIV is the preferred ventilatory modality to treat acute respiratory failure due to chronic obstructive pulmonary disease exacerbations (Boldrini R, 2012) and it is used just at night, or for part of the day in addition, or for 24 hours as clinical status indicates. NIV has been used in CF since the 1990s (Hodson M, 1991, Fauroux B, 1999), both in acute and chronic respiratory failure, mostly as a bridge to lung transplantation.

A role of NIV as an airway clearance technique, particularly in patients who have difficulty expectorating sputum and respiratory muscle weakness, was been already hypothesized some time ago (Fauroux B, 1999) and more recently speculated again (Rodriguez Hortal MC, 2018).

A retrospective analysis (Spoletini G, 2021) about all CF patients who received NIV over a 10 years period in a large UK CF Center, showed that NIV is being used as adjunct therapy for the management of advanced lung disease and that adherence to NIV treatment can stabilize lung function, but does not reduce pulmonary exacerbation.

In the same year another study (Wadsworth LE, 2021) about 94 patients attending a UK adult CF Centre receiving NIV as treatment for hypercapnic respiratory failure over a nine-years period showed that NIV use is associated with improvements in lung function and attenuation of hypercapnia which is maintained for up to three years post NIV initiation.

An observational study (Papale M, 2021) has suggested that nocturnal NIV for one year can counteract the progression of the lung disease also in normocapnic patients.

In 2022 (Jobanputra A, 2022) a review summerized the phisiologic basis for NIV use in CF, described indications and discussed how to monitor patients receiving NIV.

Issues

NIV role in improving acute and chronic respiratory failure condition.

NIV role in overnight ventilation.

NIV role as an airway clearance technique.

CF patients who might benefit to a larger extent from NIV.

NIV impact in terms of practical difficulties, such as uncomfortableness, noise, intrusiveness, and travel restrictions.

What is known

One Cochrane Review (Moran F, 2017), included ten trials with a total of 191 patients showed that NIV can improve gas exchange in lungs, exercise tolerance and dyspnoea in patients with daytime hypercapnia and, used in addition to oxygen, may improve gas exchange during sleep to a greater extent than oxygen therapy alone in moderate to severe disease. Moreover it may be a useful adjunct to other airway clearance techniques. NIV has been proven to be safe and well-tolerated.

One CDSR (Willis LD, 2021) concluded that NIV during sleep improves nocturnal hypercapnia and exercise tolerance, while there is not enough evidence to support the routine use as an adjunct to airway clearance or during exercise.

One CDSR (Sousa AS, 2025) concluded that the current evidence on the effects of NIPPV in patients with CF, both in people experiencing a pulmonary exacerbation and in stable conditions, is still uncertain. The 14 studies that met the incusion criteria included few participants, assessed few critical outcomes, and presented methodological limitations, resulting in substantial uncertainties. High?quality studies with longer interventions are needed to better estimate the effects of NIPPV on airway clearance, during nocturnal ventilation, during exercise, or for other applications in patients with CF.

In a crossover, randomized, clinical trial (Lima CA, 2014) on 13 children and adolescents with CF and pulmonary impairment, NIV has shown to be an effective tool to increase functional capacity on the basis of the results about treadmill 6-min walk distance and regional chest wall volumes.

A RCT (Rodriguez Hortal MC, 2016), data from 32 subjects, mean age 31 years and mean forced expiratory volume in 1 second 47% (+/-14), demonstrated that NIV is a good alternative to PEP in chest physiotherapy for severely ill CF patients.

A study (Sklar MC,2018) in adult CF patients, stabilized after indication for ventilatory support, showed that NIV and a heated

humidified, high flow oxygen delivery system (HFNTH), have similar effects on diaphragmatic work per breath, but HFNT confers additional physiological benefits by decreasing respiratory rate and minute ventilation.

A study (Milross MA, 2019) in adult patients with CF and sleep desaturation that compared 12?months of NIV?with or without oxygen, with low flow oxygen therapy, demonstrated no differences between the two groups.

A cross-over trial (Stanford G,2019) in patients older than 16 years at the end of hospitalisation for a pulmonary exacerbation, showed that there is no difference in treatment effect between NIV-supported airway clearance techniques (ACT) and ACT alone.

An observational study (Wadsworth LE, 2021) about CF adult patients with hypercapnic respiratory failure, followed in a UK Center, showed that outcomes for these kind of patients have significantly improved with fifty per cent of patients expected to survive for approximately five years.

A pilot study (Papale M, 2021) has speculated that, also in normocapnic patients, nocturnal NIV improves clinical condition and can be useful to counteract the progression of lung disease.

Unresolved questions

- NIV impact on pulmonary exacerbations and disease progression.
- CF patients who might benefit from NIV.
- Long-term NIV impact on pulmonary disease and quality of life.

No RCT is ongoing about this issue

Keywords

Respiratory Insufficiency; Artificial Ventilation; Ventilators;