Cochrane Database of Systematic Reviews - - Cochrane Review

Non-invasive ventilation for cystic fibrosis

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Study design (if review, criteria of inclusion for studies)

Randomised controlled trials comparing a form of pressure preset or volume preset non-invasive ventilation to no non-invasive ventilation in people with acute or chronic respiratory failure in cystic fibrosis.

List of included studies (14)

Fauroux 1999; Gozal 1997; Holland 2003; Kofler 1998; Milross 2001; Placidi 2006; Young 2008

Participants

People with CF, of any age, diagnosed on the basis of clinical criteria and sweat testing or genotype analysis with any type of acute and chronic respiratory failure.

Interventions

NIV in overnight ventilation; Non-invasive ventilation; Non-invasive ventilation in overnight ventilation

Outcome measures

ABG: HCO3 (mmol/L); ABG: PaCO2 (mmHg); ABG: PaO2 (mmHg); ABG: pH; ABG: SaO2 (%); Airway resistance % predicted; Breathlessness; CF QoL chest symptom score; CF QoL traditional dyspnoea index score; CFQoL chest symptom score; CFQoL transitional dyspnoea index; Exercise performance (metres); Exercise performance (MSWT) (metres); Hypopneas; Lung function - chest physiotherapy including directed cough; Lung function - chest physiotherapy including PEP; Lung function during sleep; Lung function while awake; Mean respiratory rate; Mean Respiratory Rate (breaths/min); Nocturnal oxygen saturation (%); Nocturnal TcCO2 (mmHg; Nocturnal TcCO2 (mmHg); Nocturnal TcCO2 TST (mmHg); Nocturnal TcCO2(mmHg); Oxygen saturation after airway clearance (SpO2) - chest physiotherapy including directed cough; Oxygen saturation after airway clearance (SpO2) - chest physiotherapy including PEP; Oxygen saturation during airway clearance (%); Oxygen saturation during airway clearance (change in SpO2 % during treatment); REM sleep architecture; Respiratory muscle strength (cmH20); Respiratory rate (breaths/min); Respiratory rate(breaths/min) during sleep; Sleep latency; Sleep latency (min); Sputum dry weight (g) - chest physiotherapy including directed cough; Sputum dry weight (g)- chest physiotherapy including PEP; Sputum wet weight (g) - chest physiotherapy including directed cough; Sputum wet weight (g)- chest physiotherapy including PEP; Symptoms of Sleep Disordered Breathing; Total sleep time (min)

Main results

NIPPV for pwCF experiencing a pulmonary exacerbation: The evidence is very uncertain regarding the acute effects of BiPAP (bilevel positive airway pressure), both in comparison to the PEP (positive expiratory pressure) mask (mean difference (MD) â€•0.06, 95% confidence interval (CI) â€•0.46 to 0.34) and to directed coughing (MD â€•0.09, 95% CI â€•0.56 to 0.38) on the amount of dry sputum expectorated. Regarding patientâ€•reported tiredness, participants reported feeling less tired after BiPAP than after the PEP mask (limited data for analysis). Evidence is also very uncertain regarding the shortâ€•term effects of BiPAP combined with airway clearance techniques on length of hospital stay (MD â€•0.5, 95% CI â€•3.06 to 2.06) and on adverse events. Similarly, the evidence is very uncertain about the acute effect of BiPAP compared to oxygen therapy on patientâ€•reported comfort (MD 1.00, 95% CI â€•0.75 to 2.75). NIPPV for people with stable CF: The evidence is very uncertain regarding the acute effect of BiPAP compared to the PEP mask on FEVâ,• (forced expiratory volume in the first second) and FEF25â€•75 (forced expiratory flow between 25% and 75% of FVC) (limited data for analysis). The evidence is also very uncertain regarding adverse events (limited data for analysis). In the short term, the evidence is very uncertain about the effect of BiPAP combined with other treatments compared to PEP with the same treatments on predicted FEVâ,• % (MD â€•13.00, 95% CI â€•21.32 to â€•4.68) and predicted FVC (forced vital capacity) % (MD â€•17.00, 95% CI â€•26.80 to â€•7.20). The same applies to CPAP (continuous positive airway pressure) combined with airway clearance techniques compared to techniques alone, with very low certainty of evidence for predicted FEVâ,• % (MD â€•0.90, 95% CI â€•17.41 to 15.61), predicted FVC % (MD 0.40, 95% CI â€•13.46 to 14.26) and predicted FEF25â€•75 % (MD â€•6.00, 95% CI â€•28.03 to 16.03). Also in the short term, the evidence is very uncertain regarding the effect of BiPAP compared to room air on sleep quality (MD â€•1.0, 95% CI â€•4.04 to 2.04), predicted FEVâ,• % (MD 1.00, 95% CI â€•8.62 to 10.62), predicted FVC % (MD 4.00, 95% CI â€•10.32 to 18.30) and patientâ€•reported acceptability (withdrawal due to mask). Similarly, the evidence is also very uncertain regarding the effect of BiPAP compared to oxygen therapy on sleep quality (MD 0.0, 95% CI â€•2.62 to 2.62), predicted FEVâ,• % (MD 1.00, 95% CI â€•8.13 to 10.13), predicted FVC % (MD 4.00, 95% CI â€•11.22 to 19.22) and acceptability (withdrawal due to mask). And the evidence is very uncertain regarding the mediumâ€•term effect of BiPAP combined with oxygen therapy compared to oxygen therapy alone on sleep

quality (MD 1.00, 95% CI â€•1.77 to 3.77), predicted FEVâ‚ % (MD 2.00, 95% CI â€•7.52 to 11.52) and predicted FVC % (MD â€•1.00, 95% CI â€•19.11 to 17.11). The risk of bias limitations involved period and carryover effects (from crossâ€•over), deviations from interventions, outcome measurement, and outcome selection. All outcomes presented very low certainty of the evidence due to methodological limitations, limited data for analysis, and imprecision. The other prespecified outcomes that were considered priorities in this review were not assessed in the included studies.

Authors' conclusions

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 studies included few participants, assessed few critical outcomes, and presented methodological limitations, resulting in substantial uncertainties. Highâ€•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.

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See also

Sousa AS, Rocha-Filho CR, Rocha A, Piva SR, Jahnke N, Trevisani VFM, Pinto ACPN. Nonâ€•invasive ventilation for cystic fibrosis. Cochrane Database of Systematic Reviews 2025, Issue 11. Art. No.: CD002769. DOI: 10.1002/14651858.CD002769.pub6. Accessed 25 November 2025.

Keywords

Artificial Ventilation; non pharmacological intervention - devices OR physiotherapy; Ventilators; NIV;