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 (10)

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

Ten trials met the inclusion criteria with a total of 191 participants. Seven trials evaluated single treatment sessions, one evaluated a two-week intervention, one evaluated a six-week intervention and one a three-month intervention. It is only possible to blind trials of airway clearance and overnight ventilatory support to the outcome assessors. In most of the trials we judged there was an unclear risk of bias with regards to blinding due to inadequate descriptions. The six-week trial was the only one judged to have a low risk of bias for all other domains. One single intervention trial had a low risk of bias for the randomisation procedure with the remaining trials judged to have an unclear risk of bias. Most trials had a low risk of bias with regard to incomplete outcome data and selective reporting. Six trials (151 participants) evaluated non-invasive ventilation for airway clearance compared with an alternative chest physiotherapy method such as the active cycle of breathing techniques or positive expiratory pressure. Three trials used nasal masks, one used a nasal mask or mouthpiece and one trial used a face mask and in one trial it is unclear. Three of the trials reported on one of the review's primary outcome measures (quality of life). Results for the reviews secondary outcomes showed that airway clearance may be easier with non-invasive ventilation and people with cystic fibrosis may prefer it. We were unable to find any evidence that non-invasive ventilation increases sputum expectoration, but it did improve some lung function parameters. Three trials (27 participants) evaluated non-invasive ventilation for overnight ventilatory support compared to oxygen or room air using nasal masks (two trials) and nasal masks or full face masks (one trial). Trials reported on two of the review's primary outcomes (quality of life and symptoms of sleep-disordered breathing). Results for the reviews secondary outcome measures showed that they measured lung function, gas exchange, adherence to treatment and preference, and nocturnal transcutaneous carbon dioxide. Due to the small numbers of participants and statistical issues, there were discrepancies in the results between the RevMan and the original trial analyses. No clear differences were found between non-invasive ventilation compared with oxygen or room air except for exercise performance, which significantly improved with non-invasive ventilation compared to room air over six weeks. One trial (13 participants) evaluated non-invasive ventilation on exercise capacity (interface used was unclear) and did not reported on any of the review's primary outcomes. The trial found no clear differences between non-invasive ventilation compared to no non-invasive ventilation for any of our outcomes. Three trials reported on adverse

effects. One trial, evaluating non-invasive ventilation for airway clearance, reported that a participant withdrew at the start of the trial due to pain on respiratory muscle testing. One trial evaluating non-invasive ventilation for overnight support reported that one participant could not tolerate an increase in inspiratory positive airway pressure. A second trial evaluating non-invasive ventilation in this setting reported that one participant did not tolerate the non-invasive ventilation mask, one participant developed a pneumothorax when breathing room air and two participants experienced aerophagia which resolved when inspiratory positive airway pressure was decreased.

Authors' conclusions

Non-invasive ventilation may be a useful adjunct to other airway clearance techniques, particularly in people with cystic fibrosis who have difficulty expectorating sputum. Non-invasive ventilation, used in addition to oxygen, may improve gas exchange during sleep to a greater extent than oxygen therapy alone in moderate to severe disease. The effect of NIV on exercise is unclear. These benefits of non-invasive ventilation have largely been demonstrated in single treatment sessions with small numbers of participants. The impact of this therapy on pulmonary exacerbations and disease progression remain unclear. There is a need for long-term randomised controlled trials which are adequately powered to determine the clinical effects of non-invasive ventilation in cystic fibrosis airway clearance and exercise.

http://onlinelibrary.wiley.com/doi/10.1002/14651858.CD002769.pub5/abstract

See also

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Keywords

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