

Cochrane Database of Systematic Reviews - - Cochrane Review

# Newborn screening for cystic fibrosis

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

randomised controlled trials (RCTs) and quasi-RCTs of parallel design assessing any intervention aimed at enhancing adherence to physiotherapy in people with CF versus no intervention, another intervention or usual care

## Participants

People with CF

## Interventions

interventions to enhance adherence to airway clearance treatment and exercise therapy

## Outcome measures

Adherence to airway clearance treatment and exercise. Pulmonary exacerbations, exercise capacity, health-related QoL and healthcare costs.

## Main results

Two RCTs (77 participants with CF; age range 2 to 20 years; 44 (57%) males) met the inclusion criteria of this review. One study employed an intervention to improve adherence to exercise and the second an intervention to improve adherence to ACT. Both studies measured outcomes at baseline and at three months, but neither study formally assessed our primary outcome of adherence in terms of our planned outcome measures, and results were dependent on self-reported data. Adherence to ACTs: One RCT (43 participants) assessed using specifically composed music alongside ACTs compared to self-selected or no music (usual care). The ACT process consisted of nebuliser inhalation treatment, ACTs and relaxation or antibiotic nebuliser treatment. We graded all evidence as very low certainty. This study reported adherence to ACTs using the Morisky-Green questionnaire and also participants' perception of treatment time and enjoyment, which may influence adherence (outcome not reported specifically in this review). We are uncertain whether participants who received specifically composed music may be more likely to adhere at six and 12 weeks compared to those who received usual care, risk ratio (RR) 1.75 (95% confidence interval (CI) 1.07 to 2.86) and RR 1.56 (95% CI 1.01 to 2.40) respectively. There may not be any difference in adherence when comparing specifically composed music to self-selected music at six weeks, RR 1.21 (95% CI 0.87 to 1.68) or 12 weeks, RR 1.52 (95% CI 0.97 to 2.38); or self-selected music to usual care at six weeks, RR 1.44 (95% CI 0.82 to 2.52) or 12 weeks, RR 1.03 (95% CI 0.57 to 1.86). The music study also reported the number of respiratory infections requiring hospitalisation at 12 weeks, with no difference seen in the risk of hospitalisation between all groups. Adherence to exercise: One RCT (24 participants) compared the provision of a manual of aerobic exercises, recommended exercise prescription plus two weekly follow-up phone calls to reinforce exercise practice over a period of three months to verbal instructions for aerobic exercise according to the CF centre's protocol. We graded all evidence as very low certainty. We are uncertain whether an educational intervention leads to more participants in the intervention group undertaking increased regular physical activity at three months (self-report), RR 3.67 (95% CI 1.24 to 10.85), and there was no reported difference between groups in the number undertaking physical activity three times per week or undertaking at least 40 minutes of physical activity. No effect was seen on secondary outcome measures of spirometry, exercise capacity or any CF quality of life domains. This study did not report on the frequency of respiratory infections (hospitalised or not) or adverse events.

## Authors' conclusions

We are uncertain whether a music-based motivational intervention may increase adherence to ACTs or affect the risk of hospitalisation for a respiratory infection. We are also uncertain whether an educational intervention increases adherence to exercise or reduces the frequency of respiratory infection-related hospital admission. However, these results are largely based on self-reported data and the impact of strategies to improve adherence to ACT and exercise in children and adolescents with stable CF remains inconclusive. Given that adherence to ACT and exercise therapy are fundamental to the clinical management of people with CF, there is an urgent need for well-designed, large-scale clinical trials in this area, which should conform to the CONSORT statement for standards of reporting and use appropriate, validated outcome measures. Studies should also ensure full disclosure of data for all important clinical outcomes.

<http://onlinelibrary.wiley.com/doi/10.1002/14651858.CD001402.pub2/abstract>

## See also

Southern KW, Mérelle MME, Dankert-Roelse JE, Nagelkerke A. Newborn screening for cystic fibrosis. Cochrane Database of Systematic Reviews 2009, Issue 1. Art. No.: CD001402. DOI: 10.1002/14651858.CD001402.pub2.

## Keywords

Adult; Caregivers; Child; non pharmacological intervention - psycho-soc-edu-org; Psychoeducation; training; Self-Management; Systemic interventions; Behavioural interventions; Cognitive analytic therapy; information; Acapella; Active Cycle of Breathing Technique -ACBT-; Adolescent; Aerobic training; Airway clearance drugs -expectorants- mucolytic- mucociliary-; Airway clearance technique; Autogenic drainage; Chest physiotherapy; Chest Wall Oscillation; Combined Modality Therapy; Drainage; exercise; flutter; forced expiration technique; High Frequency Chest Wall Oscillation -HFCWO-; Inhalation OR nebulised; inspiratory muscle training; Intrapulmonary Percussive Ventilation; non pharmacological intervention - devices OR physiotherapy; oscillating devices; Percussion; pharmacological\_intervention; Positive-Pressure Respiration- PEP- pep mask; Postural Drainage; Respiratory Tract Diseases; strength training; VEST Airway Clearance System; Vibration;