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Systematic Review of Nasal Endoscopy Scores in Cystic Fibrosis Patients Treated With Cystic Fibrosis Transmembrane Conductance Regulator Modulators.

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

Randomised controlled trials (RCTs) or quasi-RCTs in people with CF

Participants

People with cystic fibrosis

Interventions

Digital technology

Outcome measures

Prediction of pulmonary exacerbations; improved health outcomes; burden of treatment. The primary outcomes were 1. pulmonary exacerbations and 2. quality of life (QoL). The secondary outcomes were 3. lung function, 4. hospitalisations, 5. intravenous (IV) antibiotics, 6. microbiology, 7. cost-effectiveness and 8. adverse events.

Main results

Three studies included (415 participants) in people with CF aged 15 to 41 years over a 12-month period. One was a multicentre RCT, whilst two were single-centre RCTs. The three studies were mostly similar in their risk of bias, having low or unclear risk of selection bias but a high risk of detection bias, due to the unblinded design of these studies. The studies used a variety of digital technologies to monitor symptoms such as a digital symptom diary either with or without home spirometry monitoring. As the trials only included adults and older children, we are not certain that the results would apply to younger children. One of our primary outcomes was to assess time to detection of pulmonary exacerbation and number of pulmonary exacerbations identified between the intervention and routine care groups. We were largely unable to pool results in a meta-analysis due to the variety of methodologies and ways of reporting data. Two studies noted a shorter time to detection of exacerbations in the intervention group and one of these also reported that the intervention group had a shorter time to first exacerbation (hazard ratio for time to first exacerbation 1.45, 95% confidence interval (CI) 1.09 to 1.93), whilst a further study reported a shorter time to detection of exacerbations in the intervention group requiring oral or IV antibiotics compared to the control group (median: 70 (interquartile range (IQR) 123) days with intervention versus 141 (IQR 140) days with control; $P = 0.02$). However, all three studies were concordant in finding no probable effect on spirometry in the intervention groups when compared with their routine care groups over a 12-month period. We found that there is probably no difference between groups with regard to QoL scores across most domains except for Weight and Body Image, which favoured the usual care group. There is also probably no difference in the number of days of additional IV antibiotics needed or newly detected pathogens. No studies reported serious adverse events directly linked to the intervention and one study reported their smartphone application was generally well received.

Authors' conclusions

Pulmonary exacerbations are universally accepted to be detrimental to progression of CF-related lung disease, therefore, it is intuitive that early detection and intervention would help to improve outcomes. Digital technology provides an opportunity to detect physiological and symptomatic changes to identify exacerbations early. Our review found that digital technologies based on recording physiological change (spirometry) and symptoms probably allow earlier identification of exacerbations as a group. However, this may not reduce the number of exacerbations warranting IV antibiotics and there is probably no effect on lung function. This may be partly due to inconsistent definitions of pulmonary exacerbations and discrepancy in the management strategies for pulmonary exacerbations. Overall, the intervention may make little or no difference to QoL scores. The adherence to and uptake of digital technologies, especially those which include physiological measurements, are not well sustained and the costs of these need to be balanced against the clinical efficacy.

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See also

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Keywords

Adolescent; Adult; information; non pharmacological intervention - psycho-soc-edu-org; Self-Management; Psychoeducation; Behavioural interventions; exercise;