

Cochrane Database of Systematic Reviews - - Cochrane Review

Nebulized and oral thiol derivatives for pulmonary disease in cystic fibrosis

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

Randomized controlled studies

List of included studies (2)

Wainwright 2011

Participants

Children and adults with CF

Interventions

Therapies guided by the results of bronchoscopy (including bronchoalveolar lavage or protected bronchial brush sampling) vs therapies guided by the results of any other type of sampling (including cultures from sputum, throat swab and cough swab).

Outcome measures

Primary outcomes: lung function (FEV1, FVC), infant lung function (FEV0,5, FVC, FEF25-75, FEF75), LCI; CT; weight (kg or percentile), BMI (percentile)

Main results

We included two studies in this updated review. One study enrolled 170 infants under six months of age who had been diagnosed with CF through newborn screening. Participants were followed until they were five years old, and data were available for 157 children. The study compared outcomes for pulmonary exacerbations following treatment directed by BAL versus standard treatment based on clinical features and oropharyngeal cultures. The second study enrolled 30 children with CF aged between five and 18 years and randomised participants to receive treatment based on microbiological results of BAL triggered by an increase in lung clearance index (LCI) of at least one unit above baseline or to receive standard treatment based on microbiological results of oropharyngeal samples collected when participants were symptomatic. We judged both studies to have a low risk of bias across most domains, although the risk of bias for allocation concealment and selective reporting was unclear in the smaller study. In the larger study, the statistical power to detect a significant difference in the prevalence of *Pseudomonas aeruginosa* was low because *Pseudomonas aeruginosa* isolation in BAL samples at five years of age in both groups were much lower than the expected rate that was used for the power calculation. We graded the certainty of evidence for the key outcomes as low, other than for high-resolution computed tomography scoring and cost-of-care analysis, which we graded as moderate certainty. Both studies reported similar outcomes, but meta-analysis was not possible due to different ways of measuring the outcomes and different indications for the use of BAL. Whether antimicrobial therapy is directed by the use of BAL or standard care may make little or no difference in lung function z scores after two years (n = 29) as measured by the change from baseline in LCI and forced expiratory volume in one second (FEV1) (low certainty evidence). At five years, the larger study found little or no difference between groups in absolute FEV1 z score or forced vital capacity (FVC) (low certainty evidence). BAL-directed therapy probably makes little or no difference to any measure of chest scores assessed by computed tomography (CT) scan at either two or five years (different measures used in the two studies; moderate certainty evidence). BAL-directed therapy may make little or no difference in nutritional parameters or in the number of positive isolates of *P aeruginosa* per participant per year, but may lead to more hospitalisations per year (1 study, 157 participants; low certainty evidence). There is probably no difference in average cost of care per participant (either for hospitalisations or total costs) at five years between BAL-directed therapy and standard care (1 study, 157 participants; moderate certainty evidence). We found no difference in health-related quality of life between BAL-directed therapy and standard care at either two or five years, and the larger study found no difference in the number of isolates of *Pseudomonas aeruginosa* per child per year. The eradication rate following one or two courses of eradication treatment and the number of pulmonary exacerbations were comparable in the two groups. Mild adverse events, when reported, were generally well tolerated. The most common adverse event reported was transient worsening of cough after 29% of procedures. Significant clinical deterioration was documented during or within 24 hours of BAL in 4.8% of procedures.

Authors' conclusions

This review, limited to two wellâ€•designed randomised controlled studies, shows no evidence to support the routine use of BAL for the diagnosis and management of pulmonary infection in preschool children with CF compared to the standard practice of providing treatment based on results of oropharyngeal culture and clinical symptoms. No evidence is available for adults.

<http://onlinelibrary.wiley.com/doi/10.1002/14651858.CD007168.pub3/abstract>

See also

Nash EF, Stephenson A, Ratjen F, Tullis E. Nebulized and oral thiol derivatives for pulmonary disease in cystic fibrosis. Cochrane Database of Systematic Reviews 2013, Issue 7 Art. No.: CD007168. doi: 10.1002/14651858.CD007168.pub3

Keywords

Anti-Bacterial Agents; pharmacological_intervention; Respiratory Tract Infections; Respiratory Tract Diseases; Infection; Bacterial Infections;