

Cochrane Database of Systematic Reviews - - Cochrane Review

Vitamin D supplementation for cystic fibrosis

Code: CD007298

Year: 2014 Date: 2010 - updated: 12 OCT 2020

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

All randomised and quasi-randomised controlled trials where dornase alfa was compared to placebo, standard therapy or another mucolytic.

List of included studies (19)

Adde 2004; Amin 2011; Ballmann 2002; Castile 2009; Dodd 2000; Frederiksen 2006; Fuchs 1994; Laube 1996; McCoy 1996; Minasian 2010; Paul 2004; Quan 2001; Ramsey 1993; Ranasinha 1993; Robinson 2000; Robinson 2005; Shah 1995a; Suri 2001; Wilmott 1996

Participants

Children and adults, of any age, with CF diagnosed clinically and by sweat or genetic testing. Participants with all stages of lung disease were included.

Interventions

Dornase alfa

Outcome measures

Absolute mean % change in FEV1; Absolute mean % change in FVC; Adverse event - chest pain; Adverse event - conjunctivitis; Adverse event - dyspnoea (shortness of breath); Adverse event - facial oedema; Adverse event - haemoptysis (blood stained sputum); Adverse event - laryngitis; Adverse event - pharyngitis; Adverse event - pneumothorax; Adverse event - pneumothorax (in participants with acute exacerbations); Adverse event - rash; Adverse event - voice alteration; Adverse event - voice alteration (in participants with acute exacerbations); Adverse event - voice alteration (od versus bd treatment); Adverse event - wheeze; Mean % change in FEV1 (dornase alfa od vs dornase alfa on alternate days); Mean % change in FEV1 (dornase alfa od vs hypertonic saline); Mean % change in FVC (dornase alfa od vs dornase alfa on alternate days); Mean % change in FVC (dornase alfa od vs hypertonic saline); Mean % change in quality of life score (dornase alfa od vs hypertonic saline); Mean % change in quality of life score (dornase alfa od vs dornase alfa on alternate day); Mean change in weight (kg) from baseline (dornase alfa vs dornase alfa on alternate day); Mean change in weight (kg) from baseline (dornase alfa vs hypertonic saline); Mean change in weight from baseline; Mean number of days inpatient treatment; Mean number of days inpatient treatment (dornase alfa od vs dornase alfa alternate day); Mean number of days inpatient treatment (dornase alfa od vs hypertonic saline); Mean number of days IV antibiotics used; Number of deaths; Number of people experiencing exacerbations; Relative mean % change in FEV1; Relative mean % change in FEV1 (in participants with acute exacerbations); Relative mean % change in FEV1 - subgroup analysis at one month (by disease severity); Relative mean % change in FVC; Relative mean % change in FVC - subgroup analysis at one month (by disease severity)

Main results

The searches identified 74 trials, of which 19 (2565 participants) met our inclusion criteria. 15 trials compared dornase alfa to placebo or no dornase alfa (2447 participants); two compared daily dornase to hypertonic saline (32 participants); one compared daily dornase alfa to hypertonic saline and alternate day dornase alfa (48 participants); one compared dornase alfa to mannitol and the combination of both drugs (38 participants). Trial duration varied from six days to three years. - Dornase alfa compared to placebo or no treatment. - Dornase alfa probably improved forced expiratory volume at one second (FEV1) at one month (four trials, 248 participants), three months (one trial, 320 participants; moderate quality evidence), six months (one trial, 647 participants; high quality evidence) and two years (one trial, 410 participants). Limited low quality evidence showed treatment may make little or no difference in quality of life. Dornase alfa probably reduced the number of pulmonary exacerbations in trials of up to two years (moderate quality evidence). One trial that examined the cost of care, including the cost of dornase alfa, found that the cost savings from dornase alfa offset 18% to 38% of the medication costs. - Dornase alfa: daily versus alternate day. - One crossover trial (43 children) found little or no difference between treatment regimens for lung function, quality of life or pulmonary exacerbations (low quality evidence). - Dornase alfa compared to other medications that improve airway clearance. - Results for these comparisons were mixed. One trial (43 children) showed dornase alfa may lead to a greater improvement in FEV1 compared to hypertonic saline (low quality evidence), and one trial (23 participants) reported little or no differences in lung function between dornase alfa and mannitol or dornase alfa and dornase alfa plus mannitol (low quality evidence). One trial (23 participants) found dornase alfa may improve quality of life compared to dornase alfa plus mannitol (low quality evidence); other comparisons found little or no difference in this outcome (low quality evidence). No trials in any comparison reported any difference between groups in the number of pulmonary exacerbations (low quality evidence). -

When all comparisons are assessed, dornase alfa did not cause significantly more adverse effects than other treatments, except voice alteration and rash.

Authors' conclusions

There is evidence to show that, compared with placebo, therapy with dornase alfa may improve lung function in people with cystic fibrosis in trials lasting from one month to two years. There was a decrease in pulmonary exacerbations in trials of six months or longer, probably due to treatment. Voice alteration and rash appear to be the only adverse events reported with increased frequency in randomised controlled trials. There is not enough evidence to firmly conclude if dornase alfa is superior to other hyperosmolar agents in improving lung function.

<http://onlinelibrary.wiley.com/doi/10.1002/14651858.CD007298.pub4/abstract>

See also

Ferguson JH, Chang AB. Vitamin D supplementation for cystic fibrosis. Cochrane Database of Systematic Reviews 2014, Issue 5. Art. No.: CD007298. DOI: 10.1002/14651858.CD007298.pub4

Keywords

Adolescent; Child; Deoxyribonuclease; Airway clearance drugs -expectorants- mucolytic- mucociliary-; hydration; Hypertonic Solutions; Infant; Inhalation OR nebulised; nebuliser; pharmacological_intervention; Respiratory System Agents; Dornase alpha; Pulmozyme;