CFGD Priority Topics for CF Reviews

Topics identified through the James Lind Priority Setting Partnership in CF (2017)1.

1. What are the effective ways of simplifying the treatment burden of people with CF?

Published reviews

Duration of intravenous antibiotic therapy in people with cystic fibrosis

Home versus hospital intravenous antibiotic therapy for cystic fibrosis

Nebuliser systems for drug delivery in cystic fibrosis

Once-daily versus multiple-daily dosing with intravenous aminoglycosides for cystic fibrosis

2. How can we relieve gastro-Intestinal (GI) symptoms, such as stomach pain, bloating and nausea in people with CF?

Published reviews

Drug therapies for reducing gastric acidity in people with cystic fibrosis

Interventions for preventing distal intestinal obstruction syndrome (DIOS) in cystic fibrosis

Interventions for treating distal intestinal obstruction syndrome (DIOS) in cystic fibrosis

Pancreatic enzyme replacement therapy for people with cystic fibrosis

Published protocol

Probiotics for people with cystic fibrosis

Vacant titles

Dosing regimens for pancreatic enzyme replacement therapy in cystic fibrosis

3. What is the best treatment for non-tuberculous mycobacterium (NTM) in people with CF (including when to start and what medication)?

Published reviews

Antibiotic treatment for nontuberculous mycobacteria lung infection in people with cystic fibrosis

4. Which therapies are effective in delaying or preventing progression of lung disease in early life in people with CF?

Published reviews

Dornase alfa for cystic fibrosis

Elective versus symptomatic intravenous antibiotic therapy for cystic fibrosis

Inhaled anti-pseudomonal antibiotics for long-term therapy in cystic fibrosis

Inhaled corticosteroids for cystic fibrosis

Inhaled mannitol for cystic fibrosis

Macrolide antibiotics for cystic fibrosis

Nebulised hypertonic saline for cystic fibrosis

Oral non-steroidal anti-inflammatory drug therapy for lung disease in cystic fibrosis

Oral steroids for long-term use in cystic fibrosis

Pneumococcal vaccines for cystic fibrosis

Prophylactic anti-staphylococcal antibiotics for cystic fibrosis

Sodium channel blockers for cystic fibrosis

Vaccines for preventing infection with Pseudomonas aeruginosa in cystic fibrosis

Vaccines for preventing influenza in people with cystic fibrosis

Registered title

Inhaled muco-active agents for cystic fibrosis: an overview of Cochrane systematic reviews

5. Is there a way of preventing CF related diabetes (CFRD) in people with CF?

Published protocol

Continuous glucose monitoring systems for screening of cystic fibrosis-related diabetes [Diagnostic Test Accuracy Review]

6. What effective ways of motivation, support and technologies help people with CF improve and sustain adherence to treatment?

Published review

Interventions for promoting physical activity in people with cystic fibrosis

Psychological interventions for individuals with cystic fibrosis and their families

Self-management education for cystic fibrosis

Published protocol

Interventions for promoting participation in shared decision-making for children and adolescents with cystic fibrosis

Vacant titles

Anti-depressant medication for people with cystic fibrosis

Interventions for improving adherence to treatment in cystic fibrosis

7. Can exercise replace chest physiotherapy for people with CF?

Published reviews

Physical exercise training for cystic fibrosis

Published protocol

Airway clearance techniques for cystic fibrosis: an overview of Cochrane systematic reviews

Registered title

Exercise versus airway clearance techniques for respiratory health in people with cystic fibrosis

8. Which antibiotic combinations and dosing plans should be used for CF exacerbations and should antibiotic combinations be rotated?

Published reviews

Antibiotic treatment for Burkholderia cepacia complex in people with cystic fibrosis experiencing a pulmonary exacerbation

Bronchoscopy-guided antimicrobial therapy for cystic fibrosis

Combination antimicrobial susceptibility testing for acute exacerbations in chronic infection of Pseudomonas aeruginosa in cystic fibrosis

Duration of intravenous antibiotic therapy in people with cystic fibrosis

Inhaled antibiotics for pulmonary exacerbations in cystic fibrosis

Intravenous antibiotics for pulmonary exacerbations in people with cystic fibrosis

Once-daily versus multiple-daily dosing with intravenous aminoglycosides for cystic fibrosis

Oral anti-pseudomonal antibiotics for cystic fibrosis

Single versus combination intravenous anti-pseudomonal antibiotic therapy for people with cystic fibrosis

Vacant titles

Continuous infusions of beta-lactam antibiotics for pulmonary exacerbations of cystic fibrosis

9. Is there a way of reducing the negative effects of antibiotics e.g. resistance risk and adverse symptoms in people with CF?

Published reviews

Once-daily versus multiple-daily dosing with intravenous aminoglycosides for cystic fibrosis

Antibiotic adjuvant therapy for pulmonary infection in cystic fibrosis

Published protocol

Strategies to prevent kidney injury from antibiotics in people with cystic fibrosis

Vacant titles

Otoprotective strategies for preventing drug-induced hearing loss in people with cystic fibrosis

Otoprotective drugs for preventing drug-induced hearing loss in people with cystic fibrosis

10. What is the best way of eradicating Pseudomonas aeruginosa in people with CF?

Published reviews

Antibiotic strategies for eradicating Pseudomonas aeruginosa in people with cystic

Published protocol

Treatments for preventing recurrence of infection with Pseudomonas aeruginosa in people with cystic fibrosis

References

1. Rowbotham NJ, Smith S, Leighton PA, Rayner OC, Gathercole K, Elliott ZC, et al. The top 10 research priorities in cystic fibrosis developed by a partnership between people with CF and healthcare providersThorax 2018;73:388-390.