Cystic Fibrosis: diagnosis and management

Appendix F

Main appendix document
Summary of identified studies
04 May 2017
Disclaimer
The recommendations in this guideline represent the view of NICE, arrived at after careful consideration of the evidence available. When exercising their judgement, professionals are expected to take this guideline fully into account, alongside the individual needs, preferences and values of their patients or service users. The recommendations in this guideline are not mandatory and the guideline does not override the responsibility of healthcare professionals to make decisions appropriate to the circumstances of the individual patient, in consultation with the patient and/or their carer or guardian.
Local commissioners and/or providers have a responsibility to enable the guideline to be applied when individual health professionals and their patients or service users wish to use it. They should do so in the context of local and national priorities for funding and developing services, and in light of their duties to have due regard to the need to eliminate unlawful discrimination, to advance equality of opportunity and to reduce health inequalities. Nothing in this guideline should be interpreted in a way that would be inconsistent with compliance with those duties.
NICE guidelines cover health and care in England. Decisions on how they apply in other UK countries are made by ministers in the Welsh Government, Scottish Government, and Northern Ireland Executive. All NICE guidance is subject to regular review and may be updated or withdrawn.

Copyright
© National Institute for Health and Care Excellence 2017
## Contents

### Appendices

Appendix F: Summary of identified studies ................................................................. 5

- F.1 Diagnosis of cystic fibrosis ................................................................................. 5
- F.2 Information and support .................................................................................... 5
- F.3 Service delivery .................................................................................................. 6
  - F.3.1 Service configuration .................................................................................... 6
  - F.3.2 Multidisciplinary teams .............................................................................. 7
- F.4 Transition ............................................................................................................ 7
- F.5 Complications of CF .......................................................................................... 8
- F.6 Pulmonary monitoring ....................................................................................... 8
- F.7 Airway clearance .................................................................................................. 9
- F.8 Mucoactive agents ............................................................................................. 9
- F.9 Pulmonary infection ............................................................................................ 10
- F.10 Immunomodulatory agents ............................................................................. 11
- F.11 Nutritional interventions ................................................................................ 11
- F.12 Exocrine pancreatic insufficiency ..................................................................... 12
- F.13 Distal ileal obstruction syndrome .................................................................... 12
- F.14 Liver disease .................................................................................................... 13
- F.15 Ursodeoxycholic acid ....................................................................................... 13
- F.16 Cystic fibrosis related diabetes ......................................................................... 14
- F.17 Bone Mineral density ...................................................................................... 14
- F.18 Exercise ............................................................................................................ 15
- F.19 Psychosocial assessment ................................................................................ 15
- F.20 Cross-infection control ................................................................................... 16
- F.21 Health economics ........................................................................................... 16
Appendices

Appendix F: Summary of identified studies

F.1 Diagnosis of cystic fibrosis

Figure 1: Flow diagram of clinical article selection for when to suspect CF review

- Titles and abstracts identified, N=1330
  - Full copies retrieved and assessed for eligibility, N=44
    - Excluded, N=1286 (not relevant population, design, intervention, comparison, outcomes, unable to retrieve)
    - Publications included in review, N=5
    - Publications excluded from review, N=39 (refer to excluded studies list)

F.2 Information and support

Figure 2: Flow diagram of clinical article selection for information and support review

- Titles and abstracts identified, N=2535
  - Full copies retrieved and assessed for eligibility, N=100
    - Excluded, N=2435 (not relevant population, design, intervention, comparison, outcomes, unable to retrieve)
    - Publications included in review, N=31
    - Publications excluded from review, N=69 (refer to excluded studies list)
F.3 Service delivery

F.3.1 Service configuration

Figure 3: Flow diagram of clinical article selection for service delivery - models of care review

- Titles and abstracts identified, N= 1438
- Full copies retrieved and assessed for eligibility, N= 126
- Excluded, N=1312 (not relevant population, design, intervention, comparison, outcomes, unable to retrieve)
- Publications included in review, N=13 (2 Cochrane SR (including 2 trials); 9 primary studies)
- Publications excluded from review, N= 113 (refer to excluded studies list)
F.3.2 Multidisciplinary teams

Figure 4: Flow diagram of clinical article selection for service delivery: multi-disciplinary teams review

- Titles and abstracts identified, N= 618
- Full copies retrieved and assessed for eligibility, N= 63
  - Excluded, N=555 (not relevant population, design, intervention, comparison, outcomes, unable to retrieve)
- Publications included in review, N= 0
- Publications excluded from review, N= 63 (refer to excluded studies list)

F.4 Transition

Figure 5: Flow diagram of clinical article selection for the transition review

- Titles and abstracts identified, N= 701
- Full copies retrieved and assessed for eligibility, N=64
  - Excluded, N=637 (not relevant population, design, intervention, comparison, outcomes, unable to retrieve)
- Publications included in review, N=11
- Publications excluded from review, N=53 (refer to excluded studies list)
F.5 Complications of CF

Figure 6: Flow diagram of clinical article selection for complications of CF review

F.6 Pulmonary monitoring

Figure 7: Flow diagram of clinical article selection for pulmonary monitoring review
**F.7  Airway clearance**

*Figure 8: Flow diagram of clinical article selection for airway clearance techniques review*

- Titles and abstracts identified, N=2032
  - Full copies retrieved and assessed for eligibility, N=109
    - Excluded, N=1923 (not relevant population, design, intervention, comparison, outcomes, unable to retrieve)
  - Publications included in review, N=18 (4 Cochrane systematic reviews including 14 RCTs)
  - Publications excluded from review, N=91 (refer to excluded studies list)

**F.8  Mucoactive agents**

*Figure 9: Flow diagram of clinical article selection for mucoactive agents review*

- Titles and abstracts identified, N=898
  - Full copies retrieved and assessed for eligibility, N=116
    - Excluded, N=782 (not relevant population, design, intervention, comparison, outcomes, unable to retrieve)
  - Publications included in review, N=32 (1 TA; 3 Cochrane reviews (including 16 trials reported in 19 publications); 9 primary studies)
  - Publications excluded from review, N=84 (refer to excluded studies list)
F.9 Pulmonary infection

Figure 10: Flow diagram of clinical article selection for antimicrobial prophylaxis review

- Titles and abstracts identified, N=2658
- Full copies retrieved and assessed for eligibility, N=133
- Excluded, N=2525 (not relevant population, design, intervention, comparison, outcomes, unable to retrieve)

Publications included in review, Total = 52
- Review 1. Prophylaxis N=4 (1 Cochrane systematic review including 3 trials)
- Review 2. Acute infection N=21 (6 Cochrane systematic reviews (including 15 trials))
- Review 3. Chronic infection N=27 (1 TA (including 3 trials); 5 Cochrane systematic reviews (including 9 trials); 9 primary studies)

Publications excluded from review, N= 81 (refer to excluded studies list)
F.10 Immunomodulatory agents

Figure 11: Flow diagram of clinical article selection for Immunomodulatory agents review

- Titles and abstracts identified, N=1007
- Full copies retrieved and assessed for eligibility, N=61
- Excluded, N=946 (not relevant population, design, intervention, comparison, outcomes, unable to retrieve)
- Publications included in review, N=20 (4 Cochrane SR (including 14 RCTs); N=1 RCT; N=1 observ. Study)
- Publications excluded from review, N=41 (refer to excluded studies list)*

F.11 Nutritional interventions

Figure 12: Flow diagram of clinical article selection for nutrition review

- Titles and abstracts identified, N=823
- Full copies retrieved and assessed for eligibility, N=76
- Excluded, N=747 (not relevant population, design, intervention, comparison, outcomes, unable to retrieve)
- Publications included in review, N=18 (5 Cochrane SR (including 10 RCTs); 3 primary studies)
- Publications excluded from review, N=58 (refer to excluded studies list)
F.12 Exocrine pancreatic insufficiency

Figure 13: Flow diagram of clinical article selection for PERT review

F.13 Distal ileal obstruction syndrome

Figure 14: Flow diagram of clinical article selection for DIOS review
F.14 Liver disease

Figure 15: Flow diagram of clinical article selection for ultrasound review

- Titles and abstracts identified, N=652
- Full copies retrieved and assessed for eligibility, N=69
- Excluded, N=583 (not relevant population, design, intervention, comparison, outcomes, unable to retrieve)
- Publications included in review, N=13
- Publications excluded from review, N=56 (refer to excluded studies list)

F.15 Ursodeoxycholic acid

Figure 16: Flow diagram of clinical article selection for UDCA review

- Titles and abstracts identified, N=202
- Full copies retrieved and assessed for eligibility, N=24
- Excluded, N=178 (not relevant population, design, intervention, comparison, outcomes, unable to retrieve)
- Publications included in review, N=4 (1 Cochrane SR (including 3 trials))
- Publications excluded from review, N=20 (refer to excluded studies list)
F.16 Cystic fibrosis related diabetes

Figure 17: Flow diagram of clinical article selection for monitoring for the onset of cystic fibrosis related diabetes review

F.17 Bone Mineral density

Figure 18: Flow diagram of clinical article selection for monitoring of bone mineral density review
F.18 Exercise

Figure 19: Flow diagram of clinical article selection for exercise review

F.19 Psychosocial assessment

Figure 20: Flow diagram of clinical article selection for assessment of psychological and behavioural problems review
F.20 Cross-infection control

Figure 21: Flow diagram of clinical article selection for cross-infection control review

Titles and abstracts identified, N=1837

Full copies retrieved and assessed for eligibility, N=94

Publications included in review, N=16

Publications excluded from review, N=78 (refer to excluded studies list)

Excluded, N=1743 (not relevant population, design, intervention, comparison, outcomes, unable to retrieve)

F.21 Health economics

Figure 22: Flow diagram of selection for economic evaluations

Titles and abstracts identified, N=1,812

Full copies retrieved and assessed for eligibility, N=38

Publications included N=16

Publications excluded from review, N=22 (refer to excluded studies list)

Excluded, N=1,774 (not relevant population, design, intervention, comparison, outcomes, unable to retrieve)

Table 1: Number of included studies by area

<table>
<thead>
<tr>
<th>Area</th>
<th>Include</th>
</tr>
</thead>
<tbody>
<tr>
<td>Airway clearance</td>
<td>1</td>
</tr>
<tr>
<td>Monitoring pulmonary disease</td>
<td>2</td>
</tr>
<tr>
<td>Area</td>
<td>Include</td>
</tr>
<tr>
<td>----------------------------------------------</td>
<td>---------</td>
</tr>
<tr>
<td>Monitoring liver disease</td>
<td>0</td>
</tr>
<tr>
<td>DIOS</td>
<td>0</td>
</tr>
<tr>
<td>PERT</td>
<td>0</td>
</tr>
<tr>
<td>Nutrition</td>
<td>0</td>
</tr>
<tr>
<td>Mucoactive or mucolytic agents</td>
<td>6</td>
</tr>
<tr>
<td>Antimicrobials</td>
<td>4</td>
</tr>
<tr>
<td>Service configuration</td>
<td>3</td>
</tr>
<tr>
<td>Cross-infection</td>
<td>0</td>
</tr>
<tr>
<td>Immunomodulatory agents</td>
<td>0</td>
</tr>
<tr>
<td>UDCA</td>
<td>0</td>
</tr>
<tr>
<td>Psychological and behavioural assessment</td>
<td>0</td>
</tr>
<tr>
<td>Exercise</td>
<td>0</td>
</tr>
<tr>
<td>BMD</td>
<td>0</td>
</tr>
<tr>
<td>CFRD</td>
<td>0</td>
</tr>
<tr>
<td>Clinical manifestations</td>
<td>0</td>
</tr>
<tr>
<td>Information and support</td>
<td>0</td>
</tr>
<tr>
<td>Complications of CF</td>
<td>0</td>
</tr>
<tr>
<td>Transition</td>
<td>0</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td><strong>16</strong></td>
</tr>
</tbody>
</table>

(a) BMD, bone mineral density; CF, cystic fibrosis; DIOS, distal ileal obstruction syndrome; PERT, Pancreatic enzymes for exocrine pancreatic insufficiency; UDCA, ursodeoxycholic acid

The methods and results for each of those 16 economic evaluations will be presented in the appropriate sections below, whilst data extraction tables and quality assessments can be found in Appendix L and M, respectively.