Cystic Fibrosis: diagnosis and management

Appendix F

Main appendix document
Summary of identified studies
25 October 2017

Developed by the National Guideline Alliance, hosted by the Royal College of Obstetricians and Gynaecologists
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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.
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### 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

F.3.2 Multidisciplinary teams

Figure 4: Flow diagram of clinical article selection for service delivery: multidisciplinary teams review
F.4 Transition

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

[Diagram showing the flow from titles and abstracts identified (N=701) to publications included in review (N=11) and publications excluded from review (N=53), with reasons for exclusion and exclusions excluded from review (N=53).]

F.5 Complications of CF

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

[Diagram showing the flow from titles and abstracts identified (N=2952) to publications included in review (N=21) and publications excluded from review (N=89), with reasons for exclusion and exclusions excluded from review (N=89).]
F.6  Pulmonary monitoring

Figure 7: Flow diagram of clinical article selection for pulmonary monitoring review

- Titles and abstracts identified, N= 2,786
- Full copies retrieved and assessed for eligibility, N=119
  - Publications included in review, N= 2
    (Review 1: n=1
    Review 2: n=0
    Review 3: n=1)
  - Publications excluded from review, N=117
    (refer to excluded studies list)
- Excluded, N=2,667
  (not relevant population, design, intervention, comparison, outcomes, unable to retrieve)

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
  - Publications included in review, N=18 (4 Cochrane systematic reviews including 14 RCTs)
  - Publications excluded from review, N=91
    (refer to excluded studies list)
- Excluded, N=1923
  (not relevant population, design, intervention, comparison, outcomes, unable to retrieve)
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

- Titles and abstracts identified, N= 309
  - Full copies retrieved and assessed for eligibility, N= 59
    - Excluded, N=250 (not relevant population, design, intervention, comparison, outcomes, unable to retrieve)
  - Publications included in review, N= 7
  - Publications excluded from review, N= 52 (refer to excluded studies list)

F.13 Distal intestinal obstruction syndrome

Figure 14: Flow diagram of clinical article selection for DIOS review

- Titles and abstracts identified, N= 364
  - Full copies retrieved and assessed for eligibility, N= 12
    - Excluded, N=352 (not relevant population, design, intervention, comparison, outcomes, unable to retrieve)
  - Publications included in review, N= 0
  - Publications excluded from review, N= 12 (refer to excluded studies list)
F.14 Liver disease

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

F.15 Ursodeoxycholic acid

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

Titles and abstracts identified, N=894

Full copies retrieved and assessed for eligibility, N=34
- Publications included in review, N=0
- Publications excluded from review, N=34 (refer to excluded studies list)

Excluded, N=860 (not relevant population, design, intervention, comparison, outcomes, unable to retrieve)

F.17  Bone Mineral density

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

Titles and abstracts identified, N=229

Full copies retrieved and assessed for eligibility, N=16
- Publications included in review, N=6
- Publications excluded from review, N=10 (refer to excluded studies list)

Excluded, N=213 (not relevant population, design, intervention, comparison, outcomes, unable to retrieve)
F.18 Exercise

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

F.19 Psychological 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

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(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.