National Institute for Health and Care Excellence

Final

Cystic Fibrosis: diagnosis and management

Appendix F

Main appendix document
Summary of identified studies
25 October 2017

FINAL

Developed by the National Guideline Alliance, hosted by the Royal College of Obstetricians and Gynaecologist

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.

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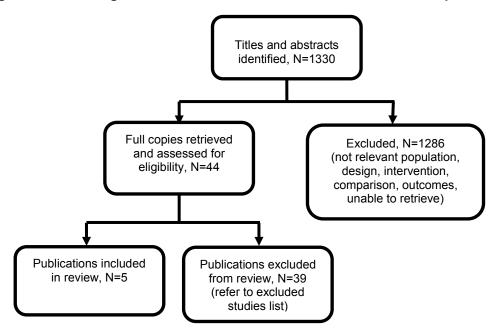
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Appendix F:Summary of identified studies

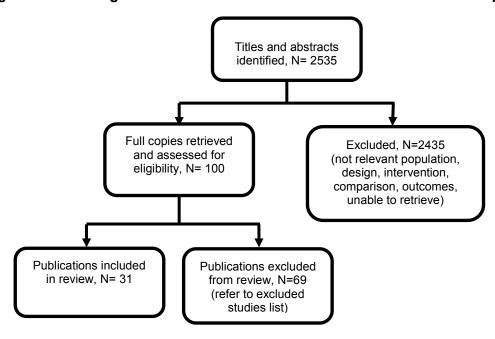
F.1 Diagnosis of cystic fibrosis

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



F.2 Information and support

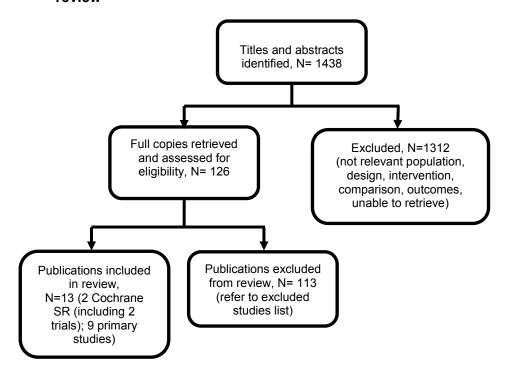
Figure 2: Flow diagram of clinical article selection for information and support review



F.3 Service delivery

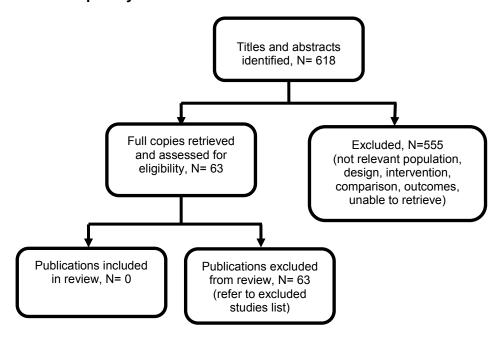
F.3.1 Service configuration

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



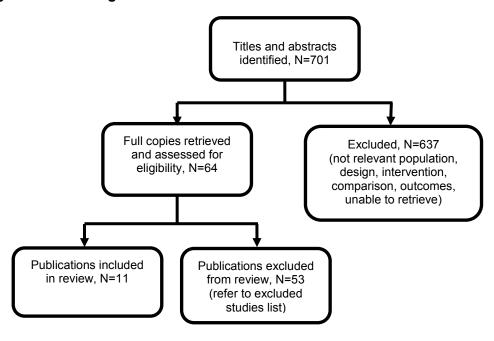
F.3.2 Multidisciplinary teams

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



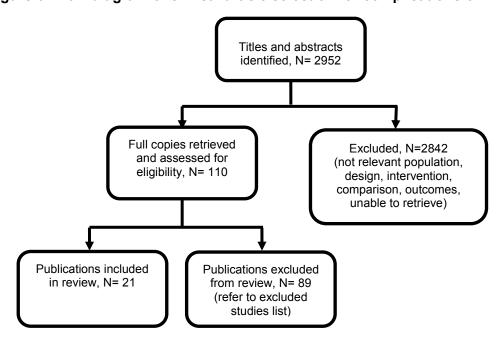
F.4 Transition

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



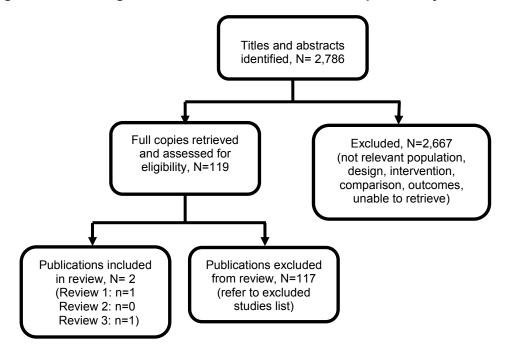
F.5 Complications of CF

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



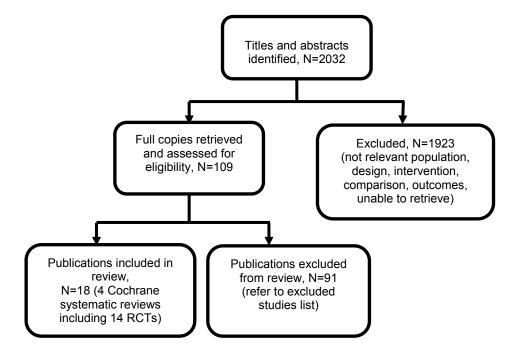
F.6 Pulmonary monitoring

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



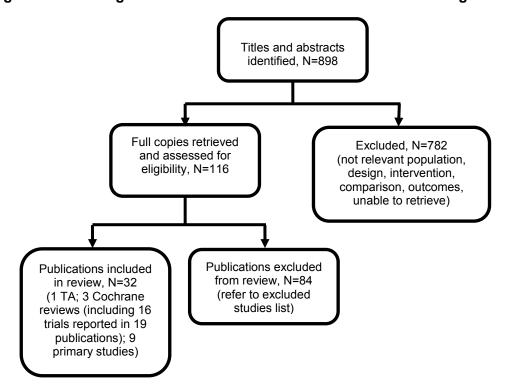
F.7 Airway clearance

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



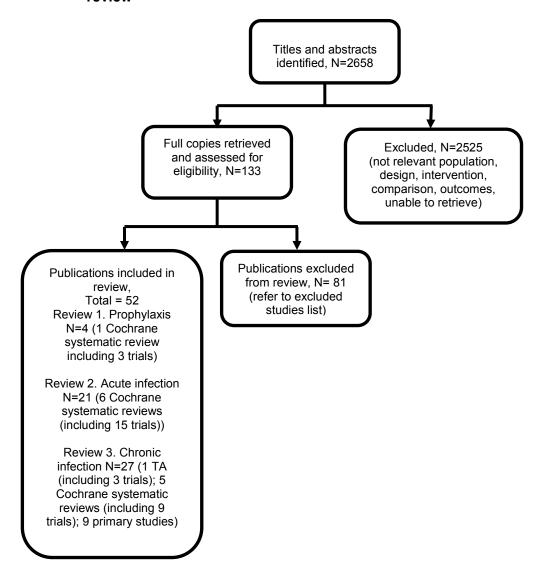
F.8 Mucoactive agents

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



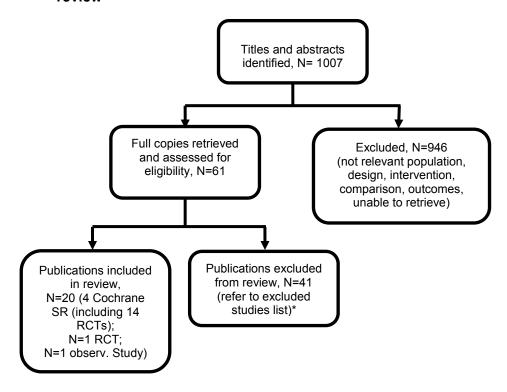
F.9 Pulmonary infection

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



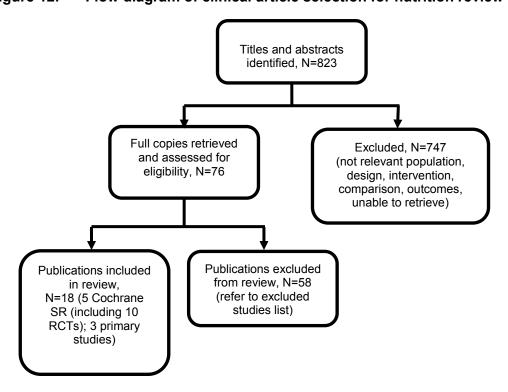
F.10 Immunomodulatory agents

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



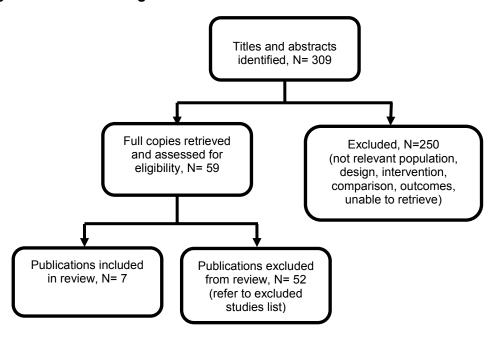
F.11 Nutritional interventions

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



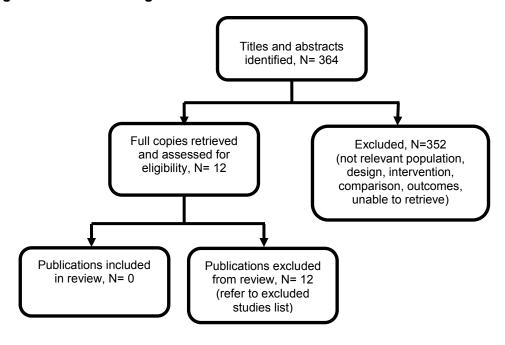
F.12 Exocrine pancreatic insufficiency

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



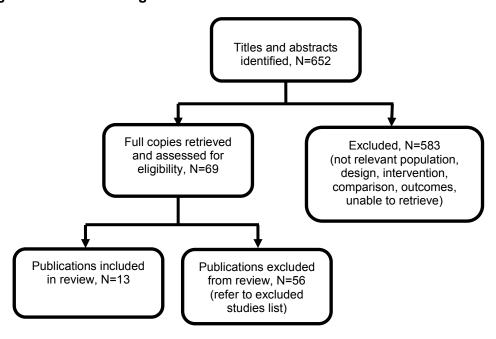
F.13 Distal intestinal obstruction syndrome

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



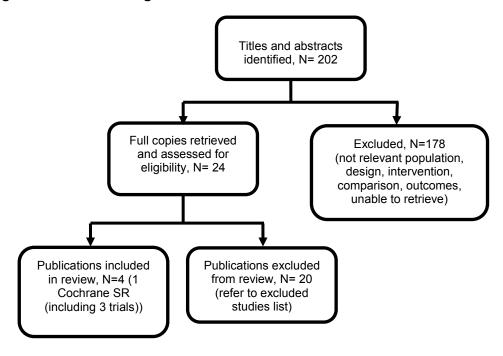
F.14 Liver disease

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



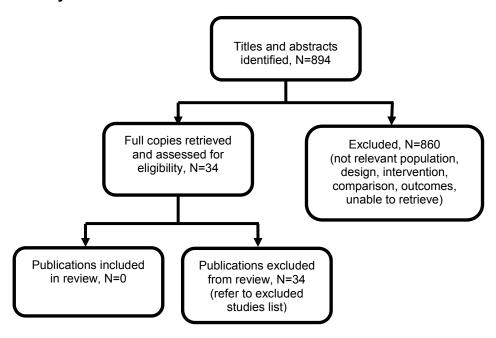
F.15 Ursodeoxycholic acid

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



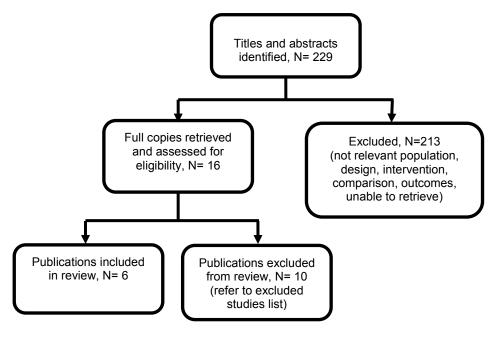
F.16 Cystic fibrosis related diabetes

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



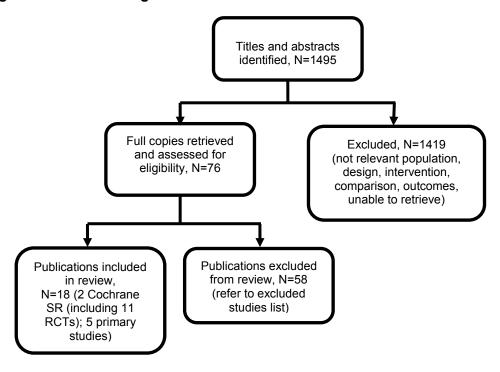
F.17 Bone Mineral density

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



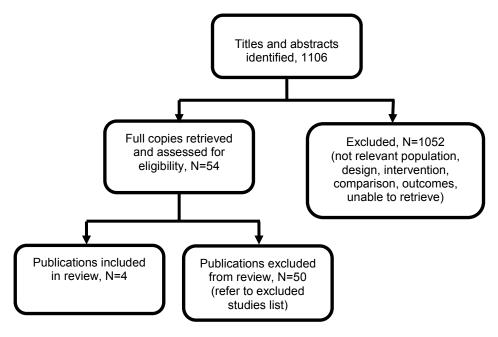
F.18 Exercise

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



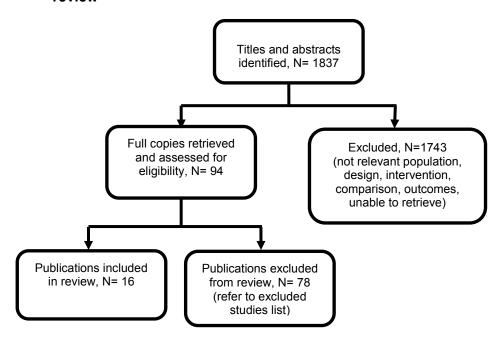
F.19 Psychological assessment

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



F.21 Health economics

Figure 22: Flow diagram of selection for economic evaluations

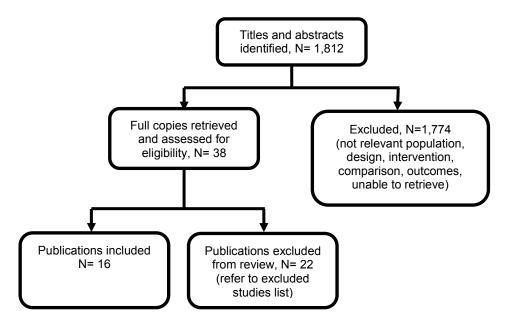


Table 1: Number of included studies by area

Area	Include
Airway clearance	1
Monitoring pulmonary disease	2

Area	Include
Monitoring liver disease	0
DIOS	0
PERT	0
Nutrition	0
Mucoactive or mucolytic agents	6
Antimicrobials	4
Service configuration	3
Cross-infection	0
Immunomodulatory agents	0
UDCA	0
Psychological and behavioural assessment	0
Exercise	0
BMD	0
CFRD	0
Clinical manifestations	0
Information and support	0
Complications of CF	0
Transition	0
Total	16

⁽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.