D.6 Complications of cystic fibrosis

Item	Details
Key issue in the scope	The complications of cystic fibrosis.
Review question in the scope	What are the complications of cystic fibrosis?
Review question in the protocol	What are the non-lower-respiratory complications of cystic fibrosis in infants, children, young people and adults?
Objective	The objective of this review is to raise awareness of the complications of cystic fibrosis among health care professionals and inform parents and carers of infants and children and young people with cystic fibrosis.
Population and directness	Infants, children, young people and adults with CF, diagnosed clinically and by sweat test or genetic testing. Population size and indirectness: • Only observational studies above sample size of 250 participants will be included (prevalence review). • Studies with indirect populations will not be considered.
Subgroups and sensitivity analyses	The following groups will be assessed separately: • Infants • Children • Young people and adults
Clinical markers	• Malnutrition • Impaired growth • Cystic fibrosis related renal disease • Delayed puberty • Distal intestinal obstruction syndrome (DIOS) • Abdominal pain • Cystic fibrosis related diabetes • Upper airways disease Cystic fibrosis related musculoskeletal disorders • Urinary stress incontinence • Reduced bone mineral density • Cystic fibrosis related liver disease • Infertility • Meconium ileus

Item	Details
Outcomes	Prevalence of complications of cystic fibrosis.
Study design	• Registry data (UK CF Registry) will be prioritised. If no data is available from the registry, observational studies will be eligible for inclusion: • Prospective cohort studies • Retrospective studies • Cross-sectional studies Countries/regions to prioritise: • UK • Western Europe • Australia • North America
Setting	Any healthcare setting where NHS care is delivered (primary, secondary, tertiary or community).
Search strategy	Sources to be searched: Medline, Medline In-Process, Cochrane Central Register of Controlled Trials, Cochrane Database of Systematic Reviews, Cochrane Database of Abstracts of Reviews of Effectiveness, Health Technology Database, Embase Limits (e.g. date, study design): Apply standard exclusions and English language filters. Apply prevalence/incidence filter. Supplementary search techniques: No supplementary search techniques were used. See appendix E.5 for full strategies
Review strategy	Appraisal of methodological quality: • The quality of the evidence will be assessed by using the tool developed and published by Munn et al. 2014 that assesses critical issues of internal and external validity that must be considered when addressing validity of prevalence data. Synthesis of data: • Meta-analysis will not be conducted. Review process: • A list of excluded studies will be provided following weeding. • Evidence tables and an evidence profile will be used to summarise the evidence.
Equalities	• Psychological and behavioural issues are more likely in people with a lower socioeconomic status • Gender- outcomes are worse for women although there is no evidence that this is a consequence of difference in care • Geographical issues – care is given through specialist centres and this may be a problem if a person with CF is living in an isolated location.
Notes/additional information	CF registry.