National Institute for Health and Care Excellence

Final

Cystic Fibrosis: diagnosis and management

Appendix H

Main appendix document

Excluded 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.

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 All NICE guidance is subject to regular review and may be updated or withdrawn.

Contents

Appendices

Appendix H: Excluded studies

H.1 Diagnosis of cystic fibrosis

Study	Reason for Exclusion
Beck, R., Durie, P. R., Hill, J. G., Levison, H., Malnutrition: a cause of elevated sweat chloride concentration, Acta Paediatrica Scandinavica, 75, 639-44, 1986	Case series.
Bruns, W. T., Connell, T. R., Lacey, J. A., Whisler, K. E., Test strip meconium screening for cystic fibrosis, American Journal of Diseases of Children, 131, 71-3, 1977	Index/ reference test: no symptoms of interest included.
Cain, A. R., Deall, A. M., Noble, T. C., Screening for cystic fibrosis by testing meconium for albumin, Archives of Disease in Childhood, 47, 131-2, 1972	Study design: case report.
Davis, P. B., Cystic fibrosis. Clinical manifestations in older patients, Clinical Notes on Respiratory Diseases, 21, 3-12, 1983	Narrative review
Davis, P. B., Del Rio, S., Muntz, J. A., Dieckman, L., Sweat chloride concentration in adults with pulmonary diseases, American Review of Respiratory Disease, 128, 34-7, 1983	Index/ reference test: no symptoms of interest included.
Desmarquest,P., Feldmann,D., Tamalat,A., Boule,M., Fauroux,B., Tournier,G., Clement,A., Genotype analysis and phenotypic manifestations of children with intermediate sweat chloride test results, Chest, 118, 1591-1597, 2000	Study design: case series
Durieu, I., Bey-Omar, F., Rollet, J., Calemard, L., Boggio, D., Lejeune, H., Gilly, R., Morel, Y., Durand, D. V., Diagnostic criteria for cystic fibrosis in men with congenital absence of the vas deferens, Medicine, 74, 42-7, 1995	Case series.
Egritas, O., Sari, S., Dalgic, B., Poyraz, A., Ensari, A., The diagnosis and outcomes of persistent diarrhea in infants aged 0-24 months - A Turkish cohort study, Turkish Journal of Gastroenterology, 22, 260-269, 2011	Case series
Farahmand, F., Khalili, M., Shahbaznejad, L., Hirbod-Mobarakeh, A., Najafi Sani, M., Khodadad, A., Motamed, F., Rezaei, N., Clinical presentation of cystic fibrosis at the time of diagnosis: a multicenter study in a region without newborn screening, Turkish Journal of Gastroenterology, 24, 541-5, 2013	Only reported data on clinical signs and symptoms in children with CF.
Fitzpatrick, S. B., Rosenstein, B. J., Langbaum, T. S., Diagnosis of cystic fibrosis during adolescence, Journal of Adolescent Health Care, 7, 38-43, 1986	Case series. Only includes people with CF.
Giglio, L., Candusso, M., D'Orazio, C., Mastella, G., Faraguna, D., Failure to thrive: the earliest	Included infants diagnosed by neonatal screening.

Study	Reason for Exclusion
feature of cystic fibrosis in infants diagnosed by neonatal screening, Acta Paediatrica, 86, 1162-5, 1997	
Gilljam, M., Ellis, L., Corey, M., Zielenski, J., Durie, P., Tullis, D. E., Clinical manifestations of cystic fibrosis among patients with diagnosis in adulthood, Chest, 126, 1215-24, 2004	Only reported data on clinical signs and symptoms in adults diagnosed with CF.
Guglani, L., Stabel, D., Weiner, D. J., False-Positive and False-Negative Sweat Tests: Systematic Review of the Evidence, Pediatric, Allergy, Immunology, and Pulmonology, 28, 198-211, 2015	Did not report data on clinical signs or symptoms included in the protocol.
Holsclaw, D. D., Rocmans, C., Shwachman, H., Abdominal complaints and appendiceal changes leading to the diagnosis of cystic fibrosis, Journal of Pediatric Surgery, 9, 867-73, 1974	Case series. Only includes people with CF.
Ibach, E. G., Meconium ileus equivalent in an adult patient, Medical Journal of Australia, 1, 268-9, 1968	Case report.
Jackson, A., Foley, L., Daly, L., Fitzpatrick, P., Harrington, M., Zhou, S., Kelleher, C., Delayed cystic fibrosis presentation in children in the absence of newborn screening, Irish Medical Journal, 103, 113-6, 2010	Only reported data on clinical signs and symptoms in children diagnosed with CF.
Kanavakis, E., Tzetis, M., Antoniadi, T., Pistofidis, G., Milligos, S., Kattamis, C., Cystic fibrosis mutation screening in CBAVD patients and men with obstructive azoospermia or severe oligozoospermia, Molecular Human Reproduction, 4, 333-7, 1998	Reference test (CF transmembrane conductance regulator membrane mutation) not in protocol.
Karakoc, G. B., Yilmaz, M., Altintas, D. U., Kendirli, S. G., Bronchiectasis: still a problem, Pediatric Pulmonology, 32, 175-8, 2001	The study only reported data on causes of brochiectasis, including CF.
Kose, M., Pekcan, S., Kiper, N., Aslan, A. T., Cobanoglu, N., Yalcin, E., Dogru, D., Ozcelik, U., Doll-like face: is it an underestimated clinical presentation of cystic fibrosis?, Pediatric Pulmonology, 43, 634-7, 2008	Symptom not in protocol (doll-like face).
Lim, M. T., Wallis, C., Price, J. F., Carr, S. B., Chavasse, R. J., Shankar, A., Seddon, P., Balfour-Lynn, I. M., Diagnosis of cystic fibrosis in London and South East England before and after the introduction of newborn screening, Archives of Disease in Childhood, 99, 197-202, 2014	Only reported data on people with CF. Case notes and database review.
Maclean, J. E., Solomon, M., Corey, M., Selvadurai, H., Cystic fibrosis newborn screening does not delay the identification of cystic fibrosis in children with negative results, Journal of Cystic Fibrosis, 10, 333-7, 2011	Only reported data on clinical signs and symptoms in children with CF.
Massie, R. J., Curnow, L., Glazner, J., Armstrong, D. S., Francis, I., Lessons learned from 20 years of newborn screening for cystic fibrosis, Medical Journal of Australia, 196, 67-70, 2012	Compares 3 CF newborn strategies. Did not report data on clinical signs and symptoms.

Study	Reason for Exclusion
Massie, R. J., Olsen, M., Glazner, J., Robertson, C. F., Francis, I., Newborn screening for cystic fibrosis in Victoria: 10 years' experience (1989-1998), Medical Journal of Australia, 172, 584-7, 2000	Retrospective review of case notes. Newborn screening only.
McWilliams, T. J., Wilsher, M. L., Kolbe, J., Cystic fibrosis diagnosed in adult patients, New Zealand Medical Journal, 113, 6-8, 2000	Retrospective review of case notes. Only reported data on clinical signs and symptoms in adults with CF.
Mir,T.A., Ashraf,M., Ahmed,K., Chowdhary,J., Rehana,B., Ahmed,J., Clinical profile, diagnostic delay, and genetic make-up of cystic fibrosis in Kashmir, India, Lung India, 28, 97-100, 2011	Reported data on clinical signs and symptoms, but not in relation to whether CF was diagnosed or not.
Murray, J, Cuckle, H, Taylor, G, Littlewood, J, Hewison, J, Screening for cystic fibrosis (Structured abstract), Health Technology Assessment Database, 2016	HTA report. Does not report on signs and symptoms.
Prosser, R., Owen, H., Bull, F., Parry, B., Smerkinich, J., Goodwin, H. A., Dathan, J., Screening for cystic fibrosis by examination of meconium, Archives of Disease in Childhood, 49, 597-601, 1974	Index/ reference test: no symptoms of interest included.
Roberts, G., Stanfield, M., Black, A., Redmond, A., Screening for cystic fibrosis: a four year regional experience, Archives of Disease in Childhood, 63, 1438-43, 1988	Only reported data on clinical signs and symptoms in children diagnosed with CF.
Roussey, M., Le Bihannic, A., Scotet, V., Audrezet, M. P., Blayau, M., Dagorne, M., David, V., Deneuville, E., Ginies, J. L., Laurans, M., Moisan-Petit, V., Rault, G., Vigneron, P., Ferec, C., Neonatal screening of cystic fibrosis: diagnostic problems with CFTR mild mutations, Journal of Inherited Metabolic Disease, 30, 613, 2007	Describes cases of children diagnosed with atypical CF at neonatal screening
Schram, C. A., Atypical cystic fibrosis: identification in the primary care setting, Canadian Family Physician, 58, 1341-5, e699-704, 2012	Narrative review
Shwachman, H., Redmond, A., Khaw, K. T., Studies in cystic fibrosis. Report of 130 patients diagnosed under 3 months of age over a 20-year period, Pediatrics, 46, 335-43, 1970	No diagnostic accuracy of prognostic data reported.
Sousa, M., Servidoni, M. F., Vinagre, A. M., Ramalho, A. S., Bonadia, L. C., Felicio, V., Ribeiro, M. A., Uliyakina, I., Marson, F. A., Kmit, A., Cardoso, S. R., Ribeiro, J. D., Bertuzzo, C. S., Sousa, L., Kunzelmann, K., Ribeiro, A. F., Amaral, M. D., Measurements of CFTR-mediated Cl- secretion in human rectal biopsies constitute a robust biomarker for Cystic Fibrosis diagnosis and prognosis, PLoS ONE [Electronic Resource], 7, e47708, 2012	Compared different screening strategies, but did not report data on clinical signs and symptoms in relation to whether CF was diagnosed or not.
Steinraths, M., Vallance, H. D., Davidson, A. G., Delays in diagnosing cystic fibrosis: can we find ways to diagnose it earlier?, Canadian Family Physician, 54, 877-83, 2008	Retrospective review of hospital medical records and CF clinic charts. Only includes people with CF.

Study	Reason for Exclusion
Tomashefski, J. F., Christoforidis, A. J., Abdullah, A. K., Cystic fibrosis in young adults: an overlooked diagnosis, with emphasis on pulmonary function and radiological patterns, Chest, 57, 28-36, 1970	Study design: case reports
van Dommelen, P., Grote, F. K., Oostdijk, W., de Muinck Keizer-Schrama, S. M., Bouquet, J., Hendriks, J. J., Kouwenberg, J., Verkerk, P. H., van Buuren, S., Wit, J. M., Growth monitoring to detect children with cystic fibrosis, Hormone Research, 72, 218-24, 2009	Evaluates growth monitoring to detect CF in children, but reference standard is not specified.
Wald,E.R., Recurrent and nonresolving pneumonia in children, Seminars in Respiratory Infections, 8, 46-58, 1993	Study design: narrative review/ discussion paper.
Wilcken, B., Towns, S. J., Mellis, C. M., Diagnostic delay in cystic fibrosis: lessons from newborn screening, Archives of Disease in Childhood, 58, 863-6, 1983	Case series.
Wilschanski, M., Famini, H., Strauss-Liviatan, N., Rivlin, J., Blau, H., Bibi, H., Bentur, L., Yahav, Y., Springer, H., Kramer, M. R., Klar, A., L. Lani A, Kerem, B., Kerem, E., Nasal potential difference measurements in patients with atypical cystic fibrosis, European Respiratory Journal, 17, 1208-1215, 2001	Index test (nasal potential difference) not in the protocol.
Wos, H., Sankiewicz-Szkolka, M., Wiecek, S., Kordys-Darmolinska, B., Grzybowska-Chlebowczyk, U., Kniazewska, M., Diagnostic problems in cystic fibrosis - specific characteristics of a group of infants and young children diagnosed positive through neonatal screening, in whom cystic fibrosis had not been diagnosed, Medycyna Wieku Rozwojowego, 19, 25-31, 2015	Included cases of patients who had a positive result of newborn screening for CF, but did not receive a diagnosis of CF because of the lack of symptoms and negative sweat test.

H.2 Information and support

Study	Reason for Exclusion
The experience of families caring for a child with cystic fibrosis: a nursing response	Thesis.
Andersson-Segesten, K., Plos, K., The needs, concerns and coping of mothers of children with cystic fibrosis, Scandinavian Journal of Caring Sciences, 3, 35-41, 1989	Descriptive data, closed ended questionnaire.
Ayers, S., Muller, I., Mahoney, L., Seddon, P., Understanding needle-related distress in children with cystic fibrosis, British Journal of Health Psychology, 16, 329-43, 2011	This study explores the experiences of children with CF/parents on needle-related distress and does not touch upon information or support as such.
Britto, M. T., Slap, G. B., DeVellis, R. F., Hornung, R. W., Atherton, H. D., Knopf, J. M., DeFriese, G. H., Specialists understanding of the health care preferences of chronically ill adolescents, Journal of Adolescent Health, 40, 334-41, 2007	No qualitative data.
Brodeur, Sydney S., Treating families coping with chronic illness: An evaluation of the "Living	Dissertation abstract.

Study	Reason for Exclusion
Well" program, Dissertation Abstracts International: Section B: The Sciences and Engineering, 66, 2811, 2005	
Byczkowski, T. L., Kotagal, U. R., Britto, M. T., Wilmott, R. W., Perceptions of value of routine care among patients with cystic fibrosis and their families, Pediatric Pulmonology, 37, 210-6, 2004	This study reports quantitative data only.
Byczkowski,T.L., Munafo,J.K., Britto,M.T., Family perceptions of the usability and value of chronic disease web-based patient portals, Health Informatics Journal, 20, 151-162, 2014	It is unclear what responses are from parents of children with CF or from parents of children with other chronic conditions.
Bywater, E., Coping with a life-threatening illness: An experiment in parents' groups, British Journal of Social Work, 14, 117-127, 1984	Not a study, no methodolody, no quotations. Describes a CF parents' group programme and the topics discussed in those group meetings.
Cappelli, M., McGrath, P. J., Heick, C. E., MacDonald, N. E., Feldman, W., Rowe, P., Chronic disease and its impact. The adolescent's perspective, Journal of Adolescent Health Care, 10, 283-8, 1989	No qualitative data on information and support.
Chotirmall, S. H., Mann, A. K., Branagan, P., O'Donohoe, C., Lyons, A. M., Flynn, M. G., Gunaratnam, C., O'Neill, S. J., McElvaney, N. G., Male fertility in cystic fibrosis, Irish Medical Journal, 102, 204-6, 2009	No qualitative data.
Cronin, Anne Frances, The influence of attention deficit disorder on mother's perception of family stress: Or, "lady, why can't you control your child?", Dissertation Abstracts International Section A: Humanities and Social Sciences, 56, 4566, 1996	Dissertation abstract.
Dashiff, C., Suzuki-Crumly, J., Kracke, B., Britton, L., Moreland, E., Cystic fibrosis-related diabetes in older adolescents: parental support and self-management, Journal for Specialists in Pediatric Nursing: JSPN, 18, 42-53, 2013	No qualitative data on information and support.
David, V., Feldman, D., Danner-Boucher, I., Rhun, A. L., Guyomarch, B., Ravilly, S., Marchand, C., Identifying the educational needs of lung transplant recipients with cystic fibrosis, Progress in Transplantation, 25, 18-25, 2015	This study only reports quantitative data.
De Wet, B., Cywes, S., The birth of a child with a congenital anomaly. Part III. Response of parents to the diagnosis, South African Medical Journal, 67, 370-373, 1985	This study was excluded due to data saturation. More recent data available from other qualitative studies.
Dellon, Elisabeth P., Shores, Mitchell D., Nelson, Katherine I., Wolfe, Joanne, Noah, Terry L., Hanson, Laura C., Caregiver perspectives on discussions about the use of intensive treatments in cystic fibrosis, Journal of Pain and Symptom Management, 40, 821-828, 2010	The study does not focus on information and support. Richer data available from other qualitative papers.
Devitt, P., Coping strategies of mothers of children with cystic fibrosis, Paediatric Nursing, 14, 12-12, 2002	A commentary of another study. The study that is being commented on will be assessed separately.
Dillard, J. P., Shen, L., Robinson, J. D., Farrell, P. M., Parental information seeking following a positive newborn screening for cystic fibrosis,	Is not a qualitative study even though some videotaping of counselling sessions was done.

Study	Reason for Exclusion
Journal of Health Communication, 15, 880-94, 2010	Does not include qualitative analysis in relation to information or support.
Dodge, J. A., Burton, L., Cull, A., McCrae, W. M., Effectiveness of counselling in cystic fibrosis, Patient Counselling and Health Education, 1, 8-12, 1978	The study methodology poorly described, no quotes.
Dyer, J., Morais, A., Supporting children with cystic fibrosis in school, Professional Nurse, 11, 518-20, 1996	Discussion/review.
Evans, J., An information model for cystic fibrosis patients and carers, Technology and Health Care, 13, 384-386, 2005	Discussion /review.
Finan, C., Nasr, S. Z., Rothwell, E., Tarini, B. A., Primary care providers' experiences notifying parents of cystic fibrosis newborn screening results, Clinical Pediatrics, 54, 67-75, 2015	Not about needs or preferences of people with CF and their families.
Frayman, K. B., Cerritelli, B., Wilson, J., Sawyer, S. M., Reproductive and sexual health in boys with cystic fibrosis: what do parents know and say?, Pediatric Pulmonology, 43, 1107-16, 2008	This study mostly reports quantitative data. Richer qualitative data available from other qualitative studies.
Frost, Julia, Review: Family communication about cystic fibrosis from the mother's perspective: An exploratory study, Journal of Research in Nursing, 12, 635-636, 2007	A review/commentary of another published study.
Gardner, J., What you need to know about cystic fibrosis, Nursing, 37, 52-5, 2007	Discussion/review.
Gjengedal, E., Rustoen, T., Wahl, A. K., Hanesta, B. R., Growing up and living with cystic fibrosis: everyday life and encounters with the health care and social services--a qualitative study, Advances in Nursing Science, 26, 149-59, 2003	This study was excluded due to data saturation. Richer data available from other qualitative studies.
Golden, S. L., Nageswaran, S., Caregiver voices: Coordinating care for children with complex chronic conditions, Clinical Pediatrics, 51, 723-729, 2012	It is unclear which quotes are from caregivers of children with CF or from caregivers of children with other conditions.
Gorlach, Jill, Understanding the Psychosocial Needs of the Teen CF Patient, AARC Times, 39, 18-19, 2015	Review/discussion.
Gotz, I., Labenbacher, I., Eichler, I., Wojnarowski, C., Gotz, M., Health-independent lung transplantation information of parents of children with cystic fibrosis, Transplantation, 64, 742-7, 1997	More comprehensive evidence was included on the topics covered by the qualitative findings of the study.
Hames, A., Beesley, J., Nelson, R., Cystic fibrosis: what do patients know, and what else would they like to know?, Respiratory Medicine, 85, 389-92, 1991	This study only reports quantitative data.
Henley, L. D., Hill, I. D., Global and specific disease-related information needs of cystic fibrosis patients and their families, Pediatrics, 85, 1015-21, 1990	This study reports quantitative data only.
Hinton, S., Watson, S., Chesson, R., Mathers, S., Information needs of young people with cystic fibrosis, Paediatric Nursing, 14, 18-21, 2002	Unclear methodology. Richer data available from other qualitative studies.

Study	Reason for Exclusion
Holton,S., Kirkman,M., Rowe,H., Fisher,J., The Childbearing Concerns and Related Information Needs and Preferences of Women of Reproductive Age with a Chronic, Noncommunicable Health Condition: A Systematic Review, Women's Health Issues, 22, e541-e552, 2012	A review with mixed population groups, including CF. Papers included in this review that relates to CF were picked up by the search of this review and are assessed individually.
Hopkins, Amanda F., Parents' perceptions of the family-school working relationship when children have a chronic condition, Dissertation Abstracts International: Section B: The Sciences and Engineering, 71, 4763, 2011	Dissertation abstract.
Hugo, C. J., van der Merwe, M., South African adolescents with cystic fibrosis: a qualitative exploration of their bio-psychosocial fields, Journal of Child & Adolescent Mental Health, 26, 177-91, 2014	Does not cover the topic of support and information apart from support from friends.
Iles, N., Lowton, K., Young people with cystic fibrosis' concerns for their future: when and how should concerns be addressed, and by whom?, Journal of Interprofessional Care, 22, 436-8, 2008	A short report on a mixed-methods study. The limited data presented does not include relevant qualitative data on information or support.
Jamieson, N., Fitzgerald, D., Singh-Grewal, D., Hanson, C. S., Craig, J. C., Tong, A., Children's experiences of cystic fibrosis: a systematic review of qualitative studies, Pediatrics, 133, e1683-97, 2014	Relevant included studies assessed for inclusion.
Jedlicka-Kohler, I., Gotz, M., Eichler, I., Parents' recollection of the initial communication of the diagnosis of cystic fibrosis, Pediatrics, 97, 204-209, 1996	No qualitative data on information and support.
Korzeniewska, A., Grzelewski, T., Jerzynska, J., Majak, P., Soloniewicz, A., Stelmach, W., Stelmach, I., Sexual and reproductive health knowledge in cystic fibrosis female patients and their parents, Journal of Sexual Medicine, 6, 770-6, 2009	No qualitative data.
Lowton, K., Ballard, K. D., Adult cystic fibrosis patients' experiences of primary care consultations: a qualitative study, British Journal of General Practice, 56, 518-25, 2006	Does not provide data in relation to support or information.
MacDonald, Kath, Irvine, Lindesay, Coulter Smith, Margaret, An exploration of partnership through interactions between young 'expert' patients with cystic fibrosis and healthcare professionals, Journal of Clinical Nursing, 24, 3528-3537, 2015	Does not touch upon support and information provision as such.
Matloff, E. T., Zimmerman, S. J., Framework for a proactive parent support group: the Syracuse cystic fibrosis model, Journal of Pediatric Health Care, 10, 264-71, 1996	Not a study, describes a framework model for a parent support group.
Miroballi, Y., Garber, E., Jia, H., Zhou, J. J., Alba, L., Quittell, L. M., Angst, D., Cabana, M., Saiman, L., Infection control knowledge, attitudes, and practices among cystic fibrosis patients and their families, Pediatric Pulmonology, 47, 144-152, 2012	No qualitative data.

Study	Reason for Exclusion
Nixon, G. M., Glazner, J. A., Martin, J. M., Sawyer, S. M., Urinary incontinence in female adolescents with cystic fibrosis, Pediatrics, 110, e22, 2002	No qualitative data on information and support.
Nolan, T., Desmond, K., Herlich, R., Hardy, S., Knowledge of cystic fibrosis in patients and their parents, Pediatrics, 77, 229-35, 1986	This study reports quantitative data only.
Nuttall, P., Nicholes, P., Cystic fibrosis: adolescent and maternal concerns about hospital and home care, Issues in Comprehensive Pediatric Nursing, 15, 199-213, 1992	This study was excluded based on data saturation, as it mostly reports quantitative data, and there are studies that report more comprehensive data.
Pakhale, S., Baron, J., Armstrong, M., Tasca, G., Gaudet, E., Aaron, S., Cameron, W., Balfour, L., A Cross-Sectional Study of the Psychological Needs of Adults Living with Cystic Fibrosis, PLoS ONE [Electronic Resource], 10, e0127944, 2015	This study reports quantitative data only.
Phillips, S., Bohannon, W. E., Gayton, W. F., Friedman, S. B., Parent interview findings regarding the impact of cystic fibrosis on families, Journal of Developmental & Behavioral Pediatrics, 6, 122-7, 1985	This study mostly reports quantitative data. More recent qualitative studies including parents have already been included.
Popli, K., Bourke, S., Stewart, J., Fertility issues in men with cystic fibrosis: survey of knowledge and opinion of patients, Fertility & Sterility, 91, 1297-8, 2009	More comprehensive evidence was included on the topics covered by the qualitative findings of the study.
Ritchie, J., Stewart, M., Ellerton, M., Thompson, D., Meade, D., Viscount, P. W., Parents' perceptions of the impact of a telephone support group intervention, Journal of Family Nursing, 6, 25-45, 2000	This study included a mixed population, and data for people with CF is not reported separately.
Romero, Sandy Liliana, The development and impact of an online social networking community for adolescents with cystic fibrosis, Dissertation Abstracts International: Section B: The Sciences and Engineering, 74, No Pagination Specified, 2014	Thesis.
Rueegg, C. S., Barben, J., Hafen, G. M., Moeller, A., Jurca, M., Fingerhut, R., Kuehni, C. E., Swiss Cystic Fibrosis Screening, Group, Newborn screening for cystic fibrosis - The parent perspective, Journal of Cystic Fibrosis, 15, 443-51, 2016	Survey reporting quantitative data only.
Salm, N., Yetter, E., Tluczek, A., Informing parents about positive newborn screen results: parents' recommendations, Journal of Child Health Care, 16, 367-81, 2012	This study was excluded due to data saturation. Richer data available from other qualitative studies. The study included a mixed population.
Savage, E, Callery, P, Weight and energy: parents' and children's perspectives on managing cystic fibrosis diet, Archives of Disease in Childhood, 90, 249-52., 2005	The qualitative data reported does not relate to information and support.
Sawicki, G. S., Dill, E. J., Asher, D., Sellers, D. E., Robinson, W. M., Advance care planning in adults with cystic fibrosis, Journal of Palliative Medicine, 11, 1135-41, 2008	No qualitative data.

Study	Reason for Exclusion
Sawicki, G. S., Sellers, D. E., McGuffie, K., Robinson, W., Adults with cystic fibrosis report important and unmet needs for disease information, Journal of Cystic Fibrosis, 6, 411-6, 2007	This study reports quantitative data only.
Sawyer, S. M., Farrant, B., Cerritelli, B., Wilson, J., A survey of sexual and reproductive health in men with cystic fibrosis: new challenges for adolescent and adult services, Thorax, 60, 326-30, 2005	No qualitative data.
Sawyer, S. M., Glazner, J. A., What follows newborn screening? An evaluation of a residential education program for parents of infants with newly diagnosed cystic fibrosis, Pediatrics, 114, 411-6, 2004	Descriptive study based on education program.
Sawyer, S. M., Tully, M. A. M., Dovey, M. E., Colin, A. A., Reproductive health in males with cystic fibrosis: Knowledge, attitudes, and experiences of patients and parents, Pediatric Pulmonology, 25, 226-230, 1998	This study was excluded due to data saturation, as more comprehensive evidence was included on male fertility from other qualitative studies.
Sawyer, S. M., Tully, M. A., Colin, A. A., Reproductive and sexual health in males with cystic fibrosis: a case for health professional education and training, Journal of Adolescent Health, 28, 36-40, 2001	This study reports quantitative data only.
Slatter, A., Francis, S. A., Smith, F., Bush, A., Supporting parents in managing drugs for children with cystic fibrosis, British Journal of Nursing, 13, 1135-9, 2004	Illegible paper, unable to retrieve.
Stewart,M.J., Ritchie,J.A., McGrath,P., Thompson,D., Bruce,B., Mothers of children with chronic conditions: supportive and stressful interactions with partners and professionals regarding caregiving burdens, Canadian Journal of Nursing Research, 26, 61-82, 1994	This study includes a mixed population. It is not always indicated which quotes refer to mother of children with CF. More recent qualitative studies conducted with carers have already been included.
Swisher, A. K., Erickson, M., Perceptions of physical activity in a group of adolescents with cystic fibrosis, Cardiopulmonary Physical Therapy Journal, 19, 107-13, 2008	No relevant data.
Tierney, S., Deaton, C., Webb, K., Jones, A., Dodd, M., McKenna, D., Rowe, R., Isolation, motivation and balance: living with type 1 or cystic fibrosis-related diabetes, Journal of Clinical Nursing, 17, 235-43, 2008	This study explores patient's views on managing diabetes. No relevant data on information and support.
Tluczek, A., Koscik, R. L., Farrell, P. M., Rock, M. J., Psychosocial risk associated with newborn screening for cystic fibrosis: parents' experience while awaiting the sweat-test appointment, Pediatrics, 115, 1692-703, 2005	This study was excluded based on data saturation, as more comprehensive evidence on information and support was included from other qualitative studies.
Ullrich,G., Jansch,H., Schmidt,S., Struber,M., Niedermeyer,J., Consulting the "experts": a pilot study on perceptions of professional support among lung transplant recipients and accompanying relatives, Progress in Transplantation, 15, 115-122, 2005	This study includes mixed population, and it is unclear what comments or suggestions were made by adults with CF or by adults with other health conditions.

Study	Reason for Exclusion
Vyhnalek, M., Vavrova, V., The supporting weekend's meeting of families with CF children, Acta Universitatis Carolinae - Medica, 36, 240-3, 1990	Descriptive quantitative study.
Whyte, D. A., A family nursing approach to the care of a child with a chronic illness, Journal of Advanced Nursing, 17, 317-27, 1992	This study was excluded based on data saturation, as more comprehensive evidence on information and support was included from other qualitative studies.
Widerman, E., Pathways to adult diagnosis of CF: the impact of pre-diagnosis experience on post-diagnosis responses and needs, Patient Education & Counseling, 52, 69-77, 2004	This study was excluded due to data saturation, as more comprehensive evidence was included on adult diagnosis by other qualitative studies.
Zack, J., Jacobs, C. P., Keenan, P. M., Harney, K., Woods, E. R., Colin, A. A., Emans, S. J., Perspectives of patients with cystic fibrosis on preventive counseling and transition to adult care, Pediatric Pulmonology, 36, 376-83, 2003	More comprehensive evidence was included related to preventive counseling. The topic of transition was covered by a separate review.

H.3 Service delivery

H.3.1 Service configuration

Study	Reason for Exclusion
Abbott, J., Hart, A., Measuring and reporting quality of life outcomes in clinical trials in cystic fibrosis: A critical review, Health and Quality of Life Outcomes, 3, 2005	Narrative review.
Aitken, M. L., Caldwell, E., Wilhelm, E., Goss, C. H., Early intervention in pulmonary exacerbation, Pediatric Pulmonology, 46, 2011	Conference abstract of RCT in the United States.
Anonymous,, Services for adults with cystic fibrosis, Journal of Cystic Fibrosis, 1, 103-9, 2002	Meeting report.
Balzano, J., Fresenius, A., Walker, P., Berdella, M., Portenoy, R. K., Bookbinder, M., Glajchen, M., Plachta, A., Langfelder-Schwind, E., Chen, J., Dhingra, L., Web-based symptom screening in cystic fibrosis patients: A feasibility study, Journal of Cystic Fibrosis, 15, 102-8, 2016	No relevant comparison.
Barnes, R., Commissioning of cystic fibrosis services in England, Journal of the Royal Society of Medicine, 99 Suppl 46, 36-45, 2006	Discussion paper.
Bella, S., Murgia, F., Cotognini, C., Alghisi, F., Montemitro, E., Program of home telemonitoring in patients with cystic fibrosis over a period of 2 years: A contribution to the rationalization of care, Clinica Terapeutica, 164, e313-e317, 2013	Costs/economic analysis. No relevant outcomes presented.
Bella, S., Murgia, F., Tozzi, A. E., Cotognini, C., Lucidi, V., Five years of Telemedicine in Cystic Fibrosis Disease, Clinica Terapeutica, 160, 457-60, 2009	FEV1 presented graphically not numerically. Other outcomes not relevant.
Bourke, S. J., Doe, S. J., Gascoigne, A. D., Heslop, K., Fields, M., Reynolds, D., Mannix, K., An integrated model of provision of palliative care to patients with cystic fibrosis, Palliative Medicine, 23, 512-7, 2009	Aim not relevant: a descriptive analysis of the circumstances of death and the palliative care needs of all patients attending one CF centre.

Study	Reason for Exclusion
Bowler, I., Pierrepoint, M., Evans, R., Response to 'Full, shared and hybrid paediatric care for cystic fibrosis (CF) in South and Mid Wales'... Arch Dis Child. 2012 Jan;97(1):17-20, Archives of Disease in Childhood, 97, 88-89, 2012	Comment.
Bradley, J. M., Wallace, E. S., Elborn, J. S., Howard, J. L., McCoy, M. P., An audit of the effect of intravenous antibiotic treatment on spirometric measures of pulmonary function in cystic fibrosis, Irish Journal of Medical Science, 168, 25-8, 1999	Given the availability of prospective studies on the intervention and outcomes covered by this study, this study was excluded because it is retrospective.
Bramwell, E. C., Halpin, D. M. G., Duncan-Skingle, F., Hodson, M. E., Geddes, D. M., Home treatment of patients with cystic fibrosis using the "Intermate": the first year's experience, Journal of Advanced Nursing, 22, 1063-1068, 1995	Given the availability of prospective studies on the intervention and outcomes covered by this study, this study was excluded because it used historical controls.
Bremont, F., Dutau, G., Organization of home care for patients with cystic fibrosis, Pediatric Pulmonology - Supplement, 16, 231, 1997	Discussion paper.
Brown-Ewing, L. J., Finkelstein, S. M., Budd, J. R., Kujawa, S. J., Wielinski, C. L., Warwick, W. J., Nguyen, S., Implementation of a home-based program for early detection of clinical deterioration in cystic fibrosis, Medical Instrumentation, 22, 240-6, 1988	No relevant data.
Byrne, N. M., Hardy, L., Community physiotherapy for children with cystic fibrosis: a family satisfaction survey, Journal of Cystic Fibrosis, 4, 123-7, 2005	Descriptive.
Carruthers, Maureen, Stevens, Anna-Marie, Palliative care and cystic fibrosis -- the benefits of integrated working, European Journal of Palliative Care, 21, 277-282, 2014	Narrative review with case report.
Chait, H., Home care: the cystic fibrosis patient, Resp.Ther, 3, 37-39+68, 1973	Discussion paper / narrative review.
Chowdhury, F., Wildman, M. J., Gunn, E., Bilton, D., Cystic fibrosis deaths in usa and UK: Comparisons of registry and routine data, Thorax, 66, A163-A164, 2011	Abstract only. No relevant comparative interventions presented.
Chuang, S., Doumit, M., McDonald, R., Hennessy, E., Katz, T., Jaffe, A., Annual Review Clinic improves care in children with cystic fibrosis, Journal of Cystic Fibrosis, 13, 186-9, 2014	Not relevant intervention.
Clarke, Blayne T., Home care in respiratory therapy, Canadian Journal of Respiratory Therapy, 52, 51-52, 2016	Commentary.
Cohen, R. I., Jaffrey, F., Fulman, M., Maniscalco, P., Ryan-Frank, C., LaVecchia, G., Application of a microsystem-based project to improve the inpatient care of adults with cystic fibrosis, Annals of the American Thoracic Society, 10, 198-204, 2013	No relevant data.
Cohen-Cymberknoh, M., Shoseyov, D., Kerem, E., Managing cystic fibrosis: strategies that	Narrative review.

Study	Reason for Exclusion
increase life expectancy and improve quality of life, American Journal of Respiratory & Critical Care Medicine, 183, 1463-71, 2011	
Collins, C. E., MacDonald-Wicks, L., Rowe, S., O'Loughlin, E. V., Henry, R. L., Normal growth in cystic fibrosis associated with a specialised centre, Archives of Disease in Childhood, 81, 241-6, 1999	No relevant comparison.
Colombo, C., Littlewood, J., The implementation of standards of care in Europe: state of the art, Journal of Cystic Fibrosis, 10 Suppl 2, S7-15, 2011	Narrative review.
Cote, D., Oruck, J., Thickson, N., A review of a Manitoba Home IV Antibiotic Program, Canadian Journal of Hospital Pharmacy, 42, 137-141, 1989	No relevant data.
Cottrell, J., Burrows, E., Community-based care in cystic fibrosis: role of the cystic fibrosis nurse specialist and implications for patients and families, Disability & Rehabilitation, 20, 254-61, 1998	Narrative review.
Cox, N. S., Alison, J. A., Button, B. M., Wilson, J. W., Holland, A. E., Feasibility and acceptability of an internet-based program to promote physical activity in adults with cystic fibrosis, Respiratory Care, 60, 422-9, 2015	Uncontrolled before-after study; no relevant intervention.
Cox, N. S., Alison, J. A., Rasekaba, T., Holland, A. E., Telehealth in cystic fibrosis: a systematic review, Journal of Telemedicine & Telecare, 18, 72-8, 2012	Systematic review. Individual studies of interest included.
Cox, N. S., McKay, K. O., Follett, J. M., Alison, J. A., Home IV Antibiotic Therapy and Exercise Capacity in Children with CF: A Case Series, Cardiopulmonary Physical Therapy Journal, 22, 16-9, 2011	Case series from Australia.
Cummings, E., Hauser, J., Cameron-Tucker, H., Fitzpatrick, P., Jessup, M., Walters, E. H., Reid, D., Turner, P., Enhancing self-efficacy for self-management in people with cystic fibrosis, Studies in Health Technology & Informatics, 169, 33-7, 2011	Quantitative data analysis subject of forthcoming paper.
David, T. J., Intravenous antibiotics at home in children with cystic fibrosis, Journal of the Royal Society of Medicine, 82, 130-1, 1989	Narrative review.
David, T. J., Home intravenous antibiotic therapy: Practical aspects in children, Journal of the Royal Society of Medicine, Supplement, 90, 26-33, 1997	Recommendations and discussion paper.
Davis, S., Davis, P., Mather, F., Tankersly, P., Waring, W., A randomized trial of home intravenous antibiotic therapy (HIVAT) in cystic fibrosis (CF) : Short-term safety and efficacy [abstract], Pediatric Pulmonology, 5, 1990	Abstract of RCT. Given the availability of full papers on home IV antibiotic therapy, abstracts were excluded from this review.
Davis, S., Davis, P., Mather, F., Waring, W., Home, IVAntibiotic Study Group, A randomized trial of home intravenous antibiotic therapy	No relevant outcomes: psychological scores: self-concept, anxiety, coping.

Study	Reason for Exclusion
(HIVAT) in cystic fibrosis (CF): Short-term psychological effects [abstract], Pediatric Pulmonology, 5, 281-2, 1990	
Dodge, J. A., Patient-centred cystic fibrosis services, Journal of the Royal Society of Medicine, 98 Suppl 45, 2-6, 2005	Narrative review.
Doull, I., Shared care--is it worth it for the patient?, Journal of the Royal Society of Medicine, 105 Suppl 2, S25-9, 2012	Narrative review.
Doull, I., Evans, H., Clarke, J., Creese, K., Hodges, I., Narayan, V., Okuonghae, H., Owen, G., Pitchaikani, P., Full, shared and hybrid paediatric care for cystic fibrosis in South and Mid Wales, Archives of Disease in Childhood, 97, 17-20, 2012	Cross-sectional.
Driessen, Julia, Bonhomme, Andro, Chang, Woody, Nace, David A., Kavalieratos, Dio, Perera, Subashan, Handler, Steven M., Nursing Home Provider Perceptions of Telemedicine for Reducing Potentially Avoidable Hospitalizations, Journal of the American Medical Directors Association, 17, 519-524, 2016	Indirect population.
Dryden, C., O'Berst, E., Corrigan, D., Models of paediatric care for cystic fibrosis: local clinics can deliver equitable care and offer many benefits... Arch Dis Child. 2012 Jan;97(1):17-20, Archives of Disease in Childhood, 97, 88-90, 2012	Narrative commentary on Doull Paper. No relevant data presented.
Elborn, J. S., Bell, S. C., Madge, S. L., Burgel, P. R., Castellani, C., Conway, S., De Rijcke, K., Dembski, B., Drevinek, P., Heijerman, H. G., Innes, J. A., Lindblad, A., Marshall, B., Olesen, H. V., Reimann, A. L., Sole, A., Viviani, L., Wagner, T. O., Welte, T., Blasi, F., Report of the European Respiratory Society/European Cystic Fibrosis Society task force on the care of adults with cystic fibrosis, European Respiratory JournalEur Respir J, 47, 420-8, 2016	Narrative review and consensus statements.
Elliott, R. A., Thornton, J., Webb, A. K., Dodd, M., Tully, M. P., Comparing costs of home- versus hospital-based treatment of infections in adults in a specialist cystic fibrosis center, International Journal of Technology Assessment in Health Care, 21, 506-10, 2005	No relevant outcome (costing study).
Ellis, J. M., Let parents give the care: iv therapy at home in cystic fibrosis, Professional Nurse, 4, 587-92, 1989	No relevant data.
Fernandes, B. N., Jones, A., Simpson, A., Webb, K., Custovic, A., Should adrenaline auto-injectors be prescribed for cystic fibrosis patients having home intravenous antibiotics?- A UK national audit, Thorax, 67, A104, 2012	UK audit, but not relevant to models of care.
Finkelstein, S. M., Budd, J. R., Warwick, W. J., Feasibility and compliance of a home measurement monitoring program for cystic fibrosis, Journal of Chronic Diseases, 39, 195-205, 1986	No relevant comparison.

Study	Reason for Exclusion
Gilbert, J., Robinson, T., Littlewood, J. M., Home intravenous antibiotic treatment in cystic fibrosis, Archives of Disease in Childhood, 63, 512-7, 1988	Poor quality reporting.
Gill, S., Home administration of intravenous antibiotics to children with cystic fibrosis, British Journal of Nursing, 2, 767-70, 1993	Narrative review.
Goss, C. H., West, N. E., Allgood, S., Wilhelm, E., Khan, U., Howe, D., Hamblett, N. M., Aitken, M. L., Ramsey, B. W., Boyle, M. P., Mogayzel Jr, P. J., Gibson, R. L., Orenstein, D., Milla, C., Lechtzin, N., Assessment of the effectiveness of home monitoring trial in CF to identify and treat acute pulmonary exacerbation: The eice study results, Pediatric Pulmonology, 50, 342-343, 2015	Outcomes of RCT are not presented in this abstract.
Greenop, D., Glenn, S., Ledson, M., Walshaw, M., Self-care and cystic fibrosis: a review of research with adults, Health & Social Care in the Community, 18, 653-61, 2010	Narrative review.
Hammond, L. J., Caldwell, S., Campbell, P. W., Cystic fibrosis, intravenous antibiotics, and home therapy, Journal of Pediatric Health Care, 5, 24-30, 1991	Narrative review.
Havermans, T., De Boeck, K., Cystic fibrosis: a balancing act?, Journal of Cystic Fibrosis, 6, 161-2, 2007	Discussion paper.
Horvais, V., Touzet, S., Francois, S., Bourdy, S., Bellon, G., Colin, C., Durieu, I., Cost of home and hospital care for patients with cystic fibrosis followed up in two reference medical centers in France, International Journal of Technology Assessment in Health Care, 22, 525-31, 2006	No relevant data.
Johnson, T., Treating cystic fibrosis patients in the home setting, Infusion, 14, 23-28, 2008	Discussion paper including a description of how information about home care is given to patients at Columbus Children's Hospital.
Jones, A. M., Adults with cystic fibrosis should be treated at a specialist centre, Paediatric Respiratory Reviews, 14, 13-15, 2013	Narrative review.
Kinder, Frances DiAnna, Parents' Perception of Satisfaction With Pediatric Nurse Practitioners' Care And Parental Intent to Adhere To Recommended Health Care Regimen, Pediatric Nursing, 42, 138-144, 2016	No relevant aim; no relevant intervention.
Kirk,S., Beatty,S., Callery,P., Gellatly,J., Milnes,L., Pryjmachuk,S., The effectiveness of self-care support interventions for children and young people with long-term conditions: a systematic review, Child: Care, Health and Development, 39, 305-324, 2013	Included studies are not relevant.
Klettke, U., Magdorf, K., Staab, D., Bisson, S., Paul, K., Wahn, U., [Ambulatory vs. inpatient intravenous antibiotic therapy in mucoviscidosis patients--a controlled study], Pneumologie (Stuttgart, Germany), 53, 31-6, 1999	In German.

Study	Reason for Exclusion
Kuzemko, J. A., Home treatment of pulmonary infections in cystic fibrosis, CHEST, 94, 162S-165S, 1988	Narrative review.
Lavie, M., Vilozni, D., Sokol, G., Somech, R., Szeinberg, A., Efrati, O., Hospital versus home treatment of respiratory exacerbations in cystic fibrosis, Medical Science Monitor, 17, CR698-703, 2011	Setting does not fit inclusion criteria.
Lebecque, P., Leonard, A., De Boeck, K., De Baets, F., Malfroot, A., Casimir, G., Desager, K., Godding, V., Leal, T., Early referral to cystic fibrosis specialist centre impacts on respiratory outcome, Journal of Cystic Fibrosis, 8, 26-30, 2009	No relevant comparison.
Lechtzin, N., Allgood, S., Kahn, U., Hamblett, N. M., Wilhelm, E., West, N. E., Howe, D., Aitken, M. L., Ramsey, B. W., Boyle, M. P., Mogayzel, P., Gibson, R. L., Orenstein, D., Milla, C., Goss, C. H., The effect of home spirometry and symptom monitoring on treatment adherence in CF, Pediatric Pulmonology, 50, 431, 2015	Outcomes of RCT are not presented in this abstract.
Lechtzin, N., West, N., Allgood, S., Wilhelm, E., Khan, U., Mayer-Hamblett, N., Aitken, M. L., Ramsey, B. W., Boyle, M. P., Mogayzel, P. J., Jr., Goss, C. H., Rationale and design of a randomized trial of home electronic symptom and lung function monitoring to detect cystic fibrosis pulmonary exacerbations: the early intervention in cystic fibrosis exacerbation (eICE) trial, Contemporary Clinical Trials, 36, 460-9, 2013	Aim not relevant: to describe the rationale and design of an ongoing trial.
Ledger, S. J., Owen, E., Prasad, S. A., Goldman, A., Willams, J., Aurora, P., A pilot outreach physiotherapy and dietetic quality improvement initiative reduces IV antibiotic requirements in children with moderate-severe cystic fibrosis, Journal of Cystic Fibrosis, 12, 766-72, 2013	No relevant comparison.
Lima, H. A., Cystic fibrosis: management and treatment in the home care setting, Home Health Care Consultant, 5, 6-15, 1998	Narrative review.
Littlewood, J. M., Good care for people with cystic fibrosis, Paediatric Respiratory Reviews, 1, 179-89, 2000	Narrative review.
Loftus, T., Reclaiming their childhood: helping cystic fibrosis patients with high-tech home care, Caring, 7, 22-7, 1988	No relevant data.
Magrabi, F., Lovell, N. H., Henry, R. L., Celler, B. G., Designing home telecare: A case study in monitoring cystic fibrosis, Telemedicine Journal and e-Health, 11, 707-719, 2005	Qualitative review. No relevant outcomes presented.
Mahadeva, R., Webb, K., Westerbeek, R. C., Carroll, N. R., Dodd, M. E., Bilton, D., Lomas, D. A., Dodge, J. A., Clinical outcome in relation to care in centres specialising in cystic fibrosis: Cross sectional study, British Medical Journal, 316, 1771-1775, 1998	Cross-sectional study.

Study	Reason for Exclusion
McClure, M., Home care of patients with cystic fibrosis, Home Health Care Management & Practice, 19, 442-445, 2007	Narrative review.
Merelle, M. E., Schouten, J. P., Gerritsen, J., Dankert-Roelse, J. E., Influence of neonatal screening and centralized treatment on long-term clinical outcome and survival of CF patients, European Respiratory Journal, 18, 306-15, 2001	Poor quality reporting. Baseline characteristics not reported, follow-up not defined.
Mighten, J., Home intravenous therapy training for carers of children and young people, British Journal of Nursing, 16, 272-6, 2007	Intervention not relevant: training package (instead of model of care).
Morrison, L., McIntosh, L., Freeman, A., UK national audit of physiotherapy clinical standards of care in cystic fibrosis, Journal of Cystic Fibrosis, 10, S67, 2011	Abstract only. No relevant data presented.
Muirhead, C. A., Sanford, J. N., McCullar, B. G., Nolt, D., MacDonald, K. D., One Center's Guide to Outpatient Management of Pediatric Cystic Fibrosis Acute Pulmonary Exacerbation, Clinical Medicine Insights.Pediatrics, 10, 57-65, 2016	This paper describes a CF centre's practices. No relevant comparison.
Munzenberger, P. J., Levin, S., Home parenteral antibiotic therapy for patients with cystic fibrosis, Hospital Pharmacy, 28, 20-8, 1993	Narrative review.
Nazer, D., Abdulhamid, I., Thomas, R., Pendleton, S., Home versus hospital intravenous antibiotic therapy for acute pulmonary exacerbations in children with cystic fibrosis, Pediatric Pulmonology, 41, 744-9, 2006	Given the availability of prospective studies on the intervention and outcomes covered by this study, this study was excluded because it is retrospective.
Nightingale, Julia, Shafiq, Irfan, End-of-life care for cystic fibrosis patients - part 2, European Journal of Palliative Care, 22, 136-141, 2015	Narrative review.
Nikander, K., Denyer, J., Dodd, M., Dyche, T., Webb, K., Weller, P., Stableforth, D., The Adaptive Aerosol Delivery system in a telehealth setting: patient acceptance, performance and feasibility, Journal of aerosol medicine & pulmonary drug delivery, 23 Suppl 1, S21-7, 2010	Study design does not fit protocol: no control group.
Oermann, C. M., Sockrider, M. M., Giles, D., Sontag, M. K., Accurso, F. J., Castile, R. G., Comparison of high-frequency chest wall oscillation and oscillating positive expiratory pressure in the home management of cystic fibrosis: a pilot study, Pediatric Pulmonology, 32, 372-7, 2001	No relevant comparison.
Peckham, D., European Cystic Fibrosis Society Standards of Care: Framework for the Cystic Fibrosis Centre, Journal of cystic fibrosis : official journal of the European Cystic Fibrosis Society, 13, S3-22, 2014	Narrative review.
Pedersen, M. G., Jensen-Fangel, S., Olesen, H. V., Tambe, S. D., Petersen, E., Outpatient parenteral antimicrobial therapy (OPAT) in patients with cystic fibrosis, BMC Infectious Diseases, 15, 290, 2015	No relevant comparison.

Study	Reason for Exclusion
Pinney, M., Home care for CF patients, Nursing, 4, 70-1, 1974	Letters.
Pond, M. N., Newport, M., Joanes, D., Conway, S. P., Home versus hospital intravenous antibiotic therapy in the treatment of young adults with cystic fibrosis, European Respiratory Journal, 7, 1640-4, 1994	Given the availability of prospective studies on the intervention and outcomes covered by this study, this study was excluded because it is retrospective.
Post, P. N., Wittenberg, J., Burgers, J. S., Do specialized centers and specialists produce better outcomes for patients with chronic diseases than primary care generalists? A systematic review, International Journal for Quality in Health Care, 21, 387-96, 2009	Systematic review. Individual studies included and reviewed.
Powers, S. W., Stark, L., Chamberlin, L., Sullivan, S., Filigno, S., Rausch, J., A multi-site, randomized, controlled clinical trial of behavioral and nutrition treatment for preschoolers with cystic fibrosis, Pediatric pulmonology, 48, 2013	No relevant comparison.
Proesmans, M., Heyns, L., Moons, P., Havermans, T., De Boeck, K., Real life evaluation of intravenous antibiotic treatment in a paediatric cystic fibrosis centre: outcome of home therapy is not inferior, Respiratory Medicine, 103, 244-50, 2009	Given the availability of prospective studies on the intervention and outcomes covered by this study, this study was excluded because it is retrospective.
Ramstrom, H., Erwander, I., Mared, L., Kornfalt, R., Seiving, B., Pharmaceutical intervention in the care of cystic fibrosis patients, Journal of Clinical Pharmacy & Therapeutics, 25, 427-34, 2000	Not relevant comparison.
Rim, Matthew H., Smith, Lonnie, Kelly, Michael, Implementation of a patient-focused specialty pharmacy program in an academic healthcare system, American Journal of Health-System Pharmacy, 73, 831-838, 2016	Indirect population; no relevant outcomes.
Rogers, D., Goodchild, M. C., Role of a domiciliary physiotherapist in the treatment of children with cystic fibrosis, Physiotherapy, 82, 396-421, 1996	No relevant data.
Rovedder, P. M. E., Flores, J., Ziegler, B., Casarotto, F., Jaques, P., Barreto, S. S. M., De Tarso Roth Dalcin, P., Exercise programme in patients with cystic fibrosis: A randomized controlled trial, Respiratory Medicine, 108, 1134-1140, 2014	Not relevant comparison.
Sarfaraz, S., Sund, Z., Jarad, N., Real-time, once-daily monitoring of symptoms and FEV1 in cystic fibrosis patients - A feasibility study using a novel device, Clinical Respiratory Journal, 4, 74-82, 2010	No relevant data.
Sawicki, G. S., Goss, C. H., Tackling the increasing complexity of CF care, Pediatric Pulmonology, 50, S74-S79, 2015	Narrative review.
Schneiderman-Walker, J., Pollock, S. L., Corey, M., Wilkes, D. D., Canny, G. J., Pedder, L., Reisman, J. J., A randomized controlled trial of a	No relevant comparison.

Study	Reason for Exclusion
3-year home exercise program in cystic fibrosis, Journal of Pediatrics, 136, 304-10, 2000	
Selekman, J., Cystic fibrosis: what is involved in the home treatment program for these children, adolescents and young adults?, Pediatric Nursing, 3, 32-5, 1977	Discussion paper.
Sequeiros, I. M., Jarad, N. A., Home intravenous antibiotic treatment for acute pulmonary exacerbations in cystic fibrosis - Is it good for the patient?, Annals of Thoracic Medicine, 4, 111-4, 2009	Narrative review.
Sequeiros, I., Jarad, N., Home intravenous antibiotic treatment for acute pulmonary exacerbations in cystic fibrosis - Is it good for the patient?, Annals of Thoracic Medicine, 4, 111-114, 2009	Narrative review.
Shultz, E. K., Finkelstein, S. M., Budd, J. R., Moore, A., Warwick, W. J., A home-based pulmonary function monitor for cystic fibrosis, Medical Instrumentation, 22, 234-9, 1988	Aim not relevant.
Smyth, A. R., Bell, S. C., Bojcin, S., Bryon, M., Duff, A., Flume, P., Kashirskaya, N., Munck, A., Ratjen, F., Schwarzenberg, S. J., Sermet-Gaudelus, I., Southern, K. W., Taccetti, G., Ullrich, G., Wolfe, S., European Cystic Fibrosis, Society, European Cystic Fibrosis Society Standards of Care: Best Practice guidelines, Journal of Cystic Fibrosis, 13 Suppl 1, S23-42, 2014	Narrative review.
Stanford, G., Parrott, H., Bilton, D., Agent, P., Positive pressure--analysing the effect of the addition of non-invasive ventilation (NIV) to home airway clearance techniques (ACT) in adult cystic fibrosis (CF) patients, Physiotherapy Theory & Practice, 31, 270-4, 2015	No relevant comparison.
Stephenson, K., Intravenous therapy. Giving antibiotics at home, Nursing Standard, 3, 24-5, 1989	Description of a home therapy programme.
Strandvik, B., Hjelte, L., Malmborg, A. S., Widen, B., Home intravenous antibiotic treatment of patients with cystic fibrosis, Acta Paediatrica, 81, 340-4, 1992	Given the availability of prospective studies on the intervention and outcomes covered by this study, this study was excluded because it is not stated if it is prospective or retrospective - and seems to be retrospective considering the 1988 publication by Strandvik et al. on the same home care data.
Strandvik, B., Hjelte, L., Widen, B., Home intravenous antibiotic treatment in cystic fibrosis, Scandinavian Journal of Gastroenterology - Supplement, 143, 119-20, 1988	No control group.
Tagliente, I., Trieste, L., Solvoll, T., Murgia, F., Bella, S., Telemonitoring in Cystic Fibrosis: A 4-year Assessment and Simulation for the Next 6 Years, Interactive Journal of Medical Research, 5, e11, 2016	No relevant outcomes.
Termoz, A., Touzet, S., Bourdy, S., Decullier, E., Bouveret, L., Colin, C., Nove-Josserand, R., Reix, P., Cracowski, C., Pin, I., Bellon, G.,	Given the availability of prospective studies on the intervention and outcomes covered by this

Study	Reason for Exclusion
Durieu, I., Effectiveness of home treatment for patients with cystic fibrosis: the intravenous administration of antibiotics to treat respiratory infections, Pediatric Pulmonology, 43, 908-15, 2008	study, this study was excluded because it is retrospective.
Thigpen, Jim, Odle, Brian, Intravenous and Inhaled Antimicrobials at Home in Cystic Fibrosis Patients, Home Health Care Management & Practice, 26, 101-110, 2014	Narrative review.
Thornton, J., Elliott, R. A., Tully, M. P., Dodd, M., Webb, A. K., Clinical and economic choices in the treatment of respiratory infections in cystic fibrosis: comparing hospital and home care, Journal of Cystic Fibrosis, 4, 239-47, 2005	Given the availability of prospective studies on the intervention and outcomes covered by this study, this study was excluded because it is retrospective.
Thornton, J., Elliott, R., Tully, M. P., Dodd, M., Webb, A. K., Long term clinical outcome of home and hospital intravenous antibiotic treatment in adults with cystic fibrosis, Thorax, 59, 242-6, 2004	Given the availability of prospective studies on the intervention and outcomes covered by this study, this study was excluded because it is retrospective.
Van Aalderen, W. M. C., Mannes, G. P. M., Bosma, E. S., Roorda, R. J., Heymans, H. S. A., Home care in cystic fibrosis patients, European Respiratory Journal, 8, 172-175, 1995	Narrative review.
Vinks, A. A. T. M. M., Brimicombe, R. W., Heijerman, H. G. M., Bakker, W., Continuous infusion of ceftazidime in cystic fibrosis patients during home treatment: Clinical outcome, microbiology and pharmacokinetics, Journal of Antimicrobial Chemotherapy, 40, 125-133, 1997	Before-and-after study. Given the availability of comparative cohort studies on home IV antibiotic treatment, before-and-after studies were excluded from the review.
Walker, P., Berdella, M., Fresenius, A., Plachta, A., Wilder, K., Gordon, A., Chen, J., Bookbinder, M., Glajchen, M., Langfelder-Schwind, E., Portenoy, R., Dhingra, L., (192) Feasibility of monthly web-based screening for pain and symptoms in cystic fibrosis patients, Journal of Pain, 17, S23-S24, 2016	Conference abstract, no RCT.
Watson, H., Bilton, D., Truby, H., A randomized controlled trial of a new behavioral home-based nutrition education program, "Eat Well with CF," in adults with cystic fibrosis, Journal of the American Dietetic Association, 108, 847-52, 2008	Not relevant intervention.
Weatherall, Louise, Respiratory nursing: New technology trialled on the ward, Kai Tiaki Nursing New Zealand, 22, 35-35, 2016	Commentary on a forum.
Weber, H. C., Robinson, P. F., Saxby, N., Beggs, S. A., Els, I., Ehrlich, R. I., Do children with cystic fibrosis receiving outreach care have poorer clinical outcomes than those treated at a specialist cystic fibrosis centre?, Australian Journal of Rural HealthAust J Rural Health, 17, 17, 2016	Cross-sectional study.
Winter, R. J., George, R. J., Deacock, S. J., Shee, C. D., Geddes, D. M., Self-administered home intravenous antibiotic therapy in bronchiectasis and adult cystic fibrosis, Lancet, 1, 1338-9, 1984	Indirect population.

Study	Reason for Exclusion
Wolter, J., Bowler, S., Nolan, P., McCormack, J., Intravenous home therapy: a randomised trial in adult cystic fibrosis patients. [abstract], Australian and New Zealand journal of medicine, 25, 1995	Abstract of paper that has been included.
Zinman, R., Corey, M., Coates, A. L., Canny, G. J., Connolly, J., Levison, H., Beaudry, P. H., Nocturnal home oxygen in the treatment of hypoxemic cystic fibrosis patients, Journal of Pediatrics, 114, 368-77, 1989	No relevant intervention.

H.3.2 Multidisciplinary teams

Study	Reason for Exclusion
Anonymous,, Following CF patients throughout continuum, Hospital Case Management, 14, 70, 75, 2006	No relevant data
Anonymous,, Innovative CF program responds to the unique needs of adults, Disease Management Advisor, 12, 28-30, 25, 2006	No relevant data
Antos, N. J., Quintero, D. R., Walsh-Kelly, C. M., Noe, J. E., Schechter, M. S., Improving inpatient cystic fibrosis pulmonary exacerbation care: two success stories, BMJ Quality & Safety, 23 Suppl 1, i33-i41, 2014	No relevant intervention
Barbero, G. J., The science and humanity of cystic fibrosis, Current Problems in Pediatrics, 26, 345-54, 1996	Narrative review / discussion paper
Bilton, D., Cystic fibrosis, Medicine, 36, 273-278, 2008	Narrative review
Bourke, S. J., Doe, S. J., Gascoigne, A. D., Heslop, K., Fields, M., Reynolds, D., Mannix, K., An integrated model of provision of palliative care to patients with cystic fibrosis, Palliative Medicine, 23, 512-7, 2009	Aim not relevant: descriptive analysis of the circumstances of death and the palliative care needs of all patients attending one CF centre
Bowler, I., Pierrepoint, M., Evans, R., Response to 'Full, shared and hybrid paediatric care for cystic fibrosis (CF) in South and Mid Wales', Archives of Disease in Childhood, 97, 88; author reply 89-90, 2012	Comment
Bremont, F., Dutau, G., Organization of home care for patients with cystic fibrosis, Pediatric Pulmonology - Supplement, 16, 231, 1997	Discussion paper
Bryon, M., Wallis, C., Parents as equal partners in the paediatric cystic fibrosis multidisciplinary team, Journal of the Royal Society of Medicine, 104 Suppl 1, S30-5, 2011	Narrative review / discussion paper
Chorro-Mari, V., Christiansen, N., Setting up of a Homecare System for High Cost Nebulisers in a Paediatric Cystic Fibrosis Centre, Archives of Disease in ChildhoodArch Dis Child, 101, e2, 2016	No relevant outcomes
Cohen-Cymberknoh, M., Shoseyov, D., Kerem, E., Managing cystic fibrosis: strategies that increase life expectancy and improve quality of	Narrative review

Study	Reason for Exclusion
life, American Journal of Respiratory & Critical Care Medicine, 183, 1463-71, 2011	
Conway, S., Balfour-Lynn, I. M., De Rijcke, K., Drevinek, P., Foweraker, J., Havermans, T., Heijerman, H., Lannefors, L., Lindblad, A., Macek, M., Madge, S., Moran, M., Morrison, L., Morton, A., Noordhoek, J., Sands, D., Vertommen, A., Peckham, D., European Cystic Fibrosis Society Standards of Care: Framework for the Cystic Fibrosis Centre, Journal of Cystic Fibrosis, 13 Suppl 1, S3-22, 2014	Narrative review
Coulter, Angela, Entwistle, Vikki A., Eccles, Abi, Ryan, Sara, Shepperd, Sasha, Perera, Rafael, Personalised care planning for adults with chronic or long-term health conditions, Cochrane Database of Systematic Reviews, 2015	Aim not relevant: the description of personalised care planning given by the review does not refer to MDTs as an essential component
Crosier, J., Wise, L. C., Coming of age. Cystic fibrosis--once a childhood disease--makes its way to adulthood, Nursing Management, 32, 30-1, 2001	No relevant data
Cuevas, O. A., Pathophysiology and diagnosis, Clinical Pharmacist, 3, 239-244, 2011	Narrative review
Dillman, D. E., Cystic fibrosis: a new challenge for the patient-care team, Respiratory Therapy, 6, 25-7, 1976	Narrative review
Doull, I., Shared care--is it worth it for the patient?, Journal of the Royal Society of Medicine, 105 Suppl 2, S25-9, 2012	Narrative review
Doull, I., Evans, H., Clarke, J., Creese, K., Hodges, I., Narayan, V., Okuonghae, H., Owen, G., Pitchaikani, P., Full, shared and hybrid paediatric care for cystic fibrosis in South and Mid Wales, Archives of Disease in Childhood, 97, 17-20, 2012	No relevant comparison
Dryden, C., O'Berst, E., Corrigan, D., Models of paediatric care for cystic fibrosis: local clinics can deliver equitable care and offer many benefits, Archives of Disease in Childhood, 97, 88-9; author reply 89-90, 2012	Comment
Eickenhorst, H., Cystic fibrosis and the emerging adult: managing the continuum, Continuum (Chicago), 16, 11-5, 1996	No relevant data
Elborn, J. S., Bell, S. C., Madge, S. L., Burgel, P. R., Castellani, C., Conway, S., De Rijcke, K., Dembski, B., Drevinek, P., Heijerman, H. G., Innes, J. A., Lindblad, A., Marshall, B., Olesen, H. V., Reimann, A. L., Sole, A., Viviani, L., Wagner, T. O., Welte, T., Blasi, F., Report of the European Respiratory Society/European Cystic Fibrosis Society task force on the care of adults with cystic fibrosis, European Respiratory JournalEur Respir J, 47, 420-8, 2016	Narrative review and consensus statements
Ellemunter, H., Stahl, K., Smrekar, U., Steinkamp, G., Evaluating patient experience in a cystic fibrosis centre using a disease-specific	No relevant comparison

Study	Reason for Exclusion
patient satisfaction questionnaire, European Journal of Pediatrics, 174, 1451-60, 2015	
Glasper, A., Randall, A., Cystic fibrosis--a cruel inheritance, Nursing - Oxford, 2, 1161-5, 1985	Narrative review
Gledhill, S., A partnership of care: an approach to the management of adults with cystic fibrosis, Australian Health Review, 20, 93-101, 1997	No relevant data
Gozdzik,J., Majka-Sumner,L., Cofta,S., Nowicka,A., Piorunek,T., Batura-Gabryel,H., Challenges in care of adult CF patients--the specialist cystic fibrosis team, Roczniki Akademii Medycznej w Bialymstoku (1995), 50 Suppl 1, 42-45, 2005	Discussion / descriptive paper
Haack, A., Carvalho Garbi Novaes, M. R., Multidisciplinary care in cystic fibrosis: a clinical-nutrition review, Nutricion Hospitalaria, 27, 362-71, 2012	Narrative review
Hodson, M. E., Psychological and social aspects of cystic fibrosis, Practitioner, 224, 301-3, 1980	Narrative review
Iles, N., Lowton, K., Young people with cystic fibrosis' concerns for their future: when and how should concerns be addressed, and by whom?, Journal of Interprofessional Care, 22, 436-8, 2008	No relevant comparison
Jones, A. M., Adults with cystic fibrosis should be treated at a specialist centre, Paediatric Respiratory Reviews, 14, 13-15, 2013	Narrative review
Kaslovsky, R., Sadof, M., How to best deliver care to children with chronic illness: cystic fibrosis as a model, Current Opinion in Pediatrics, 22, 822-8, 2010	Narrative review
Kernen, Y., Durussel, C., Hafen, G. M., Implementation and evaluation of a therapeutic educational multidisciplinary program within the paediatric clinic for cystic fibrosis, Journal of Cystic Fibrosis, 10, S87, 2011	Conference abstract and not an RCT
Long, J. M., Fauset-Jones, J., Dixon, M. J., Worthington-Riley, D., Sharma, V., Patel, L., David, T. J., Annual review hospital visits for patients with cystic fibrosis, Journal of the Royal Society of Medicine, 94 Suppl 40, 12-6, 2001	Aim not relevant: to test the hypothesis that annual review visits generate a larger number of interventions than are generated during routine outpatient clinic visits
Lowton, K., Mathes, L., Wyatt, H., Luce, P., While, A., Carmel, S., Evaluation of transition services for young people with cystic fibrosis in Southeast London, Journal of Interprofessional Care, 19, 408-9, 2005	Protocol for an evaluation of transition services
Madge, S., Bell, S. C., Burgel, P. R., De Rijcke, K., Blasi, F., Elborn, J. S., Limitations to providing adult cystic fibrosis care in Europe: Results of a care centre survey, Journal of Cystic Fibrosis, no pagination, 2016	No relevant comparison
Magtoto, M., Green, V., Simpson, M., Madge, S., How a patient perspective improved IV therapy, Nursing TimesNurs Times, 111, 16-7, 2015	No relevant intervention

Study	Reason for Exclusion
Marciel, K. K., Kimberg, C. I., Riekert, K. A., Swenson, A., Quittner, A. L., Participation of multidisciplinary team members in a behavioral intervention to improve adherence in adolescents with CF, Pediatric Pulmonology, 45, 437, 2010	Aim not relevant: to evaluate the participation of MDT members in in a structured adherence intervention training.
McIntosh, I. D., Health Human Resources Guidelines: Minimum Staffing Standards and Role Descriptions for Canadian Cystic Fibrosis Healthcare Teams, Canadian Respiratory JournalCan Respir J, 2016, 6369704, 2016	Survey on current and optimal staffing levels for each staff discipline included in CF care
Nobili, R. M., Duff, A. J., Ullrich, G., Smrekar, U., Havermans, T., Bryon, M., Borawska-Kowalczyk, U., Malmborg, M. S., Guiding principles on how to manage relevant psychological aspects within a CF team: interdisciplinary approaches, Journal of Cystic Fibrosis, 10 Suppl 2, S45-52, 2011	Narrative review
Petit-Zeman, S., Dramatic licence, Nursing Times, 98, 24-5, 2002	No relevant data
Pettit, R. S., Eaves, S., White, J. K., Victoria, W., Deegan, L. M., Howenstine, M. S., Cystic fibrosis fluid protocol, Pediatric Pulmonology, 49, 398, 2014	Conference abstract and not an RCT
Phelan, P. D., Cystic fibrosis: is it relevant to family practitioners?, Australian Family Physician, 8, 1232-4, 1979	Discussion paper
Piccini, B., Taccetti, G., Neri, A. S., Barni, F., Guasti, M., Casalini, E., Lenzi, L., Toni, S., CFRD clinical care: Experience of Diabetology Unit and Tuscany Regional Centre for Cystic Fibrosis of Meyer Children's Hospital, Pediatric Diabetes, 17, 77, 2016	Conference abstract, no RCT
Puntis, J. W. L., Booth, I. W., The place of a nutritional care team in paediatric practice, Intensive Therapy and Clinical Monitoring, 11, 132-136, 1990	No relevant data
Quittner, A. L. Q., Practical tools to improve adherence, Journal of Aerosol Medicine and Pulmonary Drug Delivery, 24 (3), 5, 2011	No relevant outcomes presented
Quittner, A. L., Alpern, A. N., McLean, K. A., Marciel, K. K., Zhang, J., Riekert, K. A., Clinical supervision improves treatment fidelity to an adherence intervention, Journal of Cystic Fibrosis, 11, S9, 2012	No relevant outcomes presented
Quittner, A. L., Riekert, K. A., Marciel, K. K., Kimberg, C. I., Eakin, M. N., Zhang, J., Adolescent management of CF: GAPS in knowledge and treatment skills in the icare study, Pediatric Pulmonology, 45, 199-200, 2010	No relevant data
Radlovic, N., Cystic fibrosis, Srpski Arhiv Za Celokupno Lekarstvo, 140, 244-9, 2012	Narrative review
Redfern, J., Webb, A. K., Benefits of a dedicated cystic fibrosis pharmacist, Journal of the Royal Society of Medicine, 97 Suppl 44, 2-7, 2004	No relevant outcomes presented

Study	Reason for Exclusion
Rideout,K., Evaluation of a PNP care coordinator model for hospitalized children, adolescents, and young adults with cystic fibrosis, Pediatric Nursing, 33, 29-35, 2007	No relevant intervention
Riekert, K. A., Eakin, M. N., Bilderback, A., Ridge, A. K., Marshall, B. C., Opportunities for cystic fibrosis care teams to support treatment adherence, Journal of Cystic Fibrosis, 14, 142-8, 2015	No relevant comparison
Rogers, C. L., Nutritional management of the adult with cystic fibrosis - Part I, Practical Gastroenterology, 37, 10-24, 2013	Narrative review
Sequeiros, I. M., Jarad, N. A., Outcome of care for home management with intensive input in adult CF patients during pulmonary exacerbations (PExs) - A comparative prospective study with hospital care, Journal of Cystic Fibrosis, 9, S58, 2010	Conference abstract and not an RCT
Sharma, R., Chang, E., See, S., Guleri, A., Improving clinical quality and patient safety in cystic fibrosis paediatric patients. First multidisciplinary clinical audit at a large district hospital of northwest England, Clinical Microbiology and Infection, 17, S64, 2011	Conference abstract and not an RCT
Smith, D. L., Stableforth, D. E., Management of adults with cystic fibrosis, British Journal of Hospital Medicine, 48, 713, 717-23, 1992	Narrative review
Stewart, A., Cystic fibrosis. Two. The primary care team's role, Nursing Times, 80, 34-5, 1984	Discussion paper
Thomas, C. L., O'Rourke, P. K., Wainwright, C. E., Clinical outcomes of Queensland children with cystic fibrosis: a comparison between tertiary centre and outreach services, Medical Journal of Australia, 188, 135-9, 2008	No relevant comparison
Turner, M. A., Unsworth, V., David, T. J., Intravenous long-lines in children with cystic fibrosis: a multidisciplinary approach, Journal of the Royal Society of Medicine, 95 Suppl 41, 11-21, 2002	No relevant intervention
Vandenbranden, S. L., The role of the nurse practitioner in the care of children with chronic respiratory disorders, Pediatric Annals, 39, 800-4, 2010	Discussion paper / narrative review
Webb, A. K., Dudley-Southern, R., Jones, A. M., Development of a modern adult cystic fibrosis centre in Manchester, Journal of the Royal Society of Medicine, 103 Suppl 1, S15-9, 2010	No comparative data presented
Weber, H. C., Robinson, P. F., Saxby, N., Beggs, S. A., Els, I., Ehrlich, R. I., Do children with cystic fibrosis receiving outreach care have poorer clinical outcomes than those treated at a specialist cystic fibrosis centre?, Australian Journal of Rural HealthAust J Rural Health, 17, 17, 2016	This paper compares different models of care rather than different MDT compositions
Whitehead, S., Pearson, C., O'Driscoll, M., Daniels, T., A multidisciplinary approach can	Conference abstract and not an RCT

Study	Reason for Exclusion
improve a low BMI in adults with cystic fibrosis, Journal of Cystic Fibrosis, 14, S50, 2015	
Wilkinson, O. M., Duncan-Skingle, F., Pryor, J. A., Hodson, M. E., A feasibility study of home telemedicine for patients with cystic fibrosis awaiting transplantation, Journal of Telemedicine & Telecare, 14, 182-5, 2008	Aim not relevant: the study does not assess a specific MDT composition, but rather a telemedicine intervention involving an MDT
Zhou, H., Roberts, P., Dhaliwal, S., Della, P., Transitioning adolescent and young adults with chronic disease and/or disabilities from paediatric to adult care services - an integrative review, Journal of Clinical NursingJ Clin Nurs, 25, 3113-3130, 2016	No relevant studies included

H.4 Transition

Study	Reason for Exclusion
Anderson, A., Caraher, M., Johnston, S., Heslop, K., O'Brien, C., Spencer, D., Bourke, S., Transition to adulthood with cystic fibrosis: Current characteristics and challenges, Journal of Cystic Fibrosis, 9, S65, 2010	Quantitative study. Conference abstract.
Anderson, D. L., Flume, P. A., Hardy, K. K., Gray, S., Transition programs in cystic fibrosis centers: perceptions of patients, Pediatric Pulmonology, 33, 327-31, 2002	Descriptive data, closed ended questionnaire.
Anonymous,, Following CF patients throughout continuum, Hospital Case Management, 14, 70, 75, 2006	Newsletter.
Bagnasco, A., Petralia, P., Furnari, S., Ghio, S., Calza, S., Sasso, L., Paediatric nurses' perception of the child-family dyad's autonomy in managing a chronic disease situation: the experience of an Italian paediatric department, Journal of Preventive Medicine & Hygiene, 54, 124-9, 2013	This study includes a mixed population, and it is unclear which quotes are from CF or other conditions.
Baker, K. L., Coe, L. M., Growing up with a chronic condition: transition to young adulthood for the individual with cystic fibrosis, Holistic Nursing Practice, 8, 8-15, 1993	Narrative review/ discussion paper.
Barto, T. L., Simonian, S., Eways, K., Gray, S., Advances in transition: Multi-level factors associated with adult achievement, Journal of Cystic Fibrosis, 11, S139, 2012	Conference abstract.
Bowmer, G., Duff, A. J. A., Sowerby, C., The views of adolescents with CF on their transition and transfer to adult services, Journal of Cystic FibrosisJ Cyst Fibros, 14, S132, 2015	Descriptive data, closed ended questionnaire.
Boyle, M. P., Farukhi, Z., Nosky, M. L., Strategies for improving transition to adult cystic fibrosis care, based on patient and parent views, Pediatric Pulmonology, 32, 428-36, 2001	Descriptive data, closed ended questionnaire.
Bryon, M., Madge, S., Transition from paediatric to adult care: psychological principles, Journal of the Royal Society of Medicine, 94 Suppl 40, 5-7, 2001	Narrative review/ discussion paper.

Study	Reason for Exclusion
Cappelli, M., MacDonald, N. E., McGrath, P. J., Assessment of readiness to transfer to adult care for adolescents with cystic fibrosis, Children's Health Care, 18, 218-24, 1989	Exploratory study with no relevant findings.
Chaudhry, S. R., Keaton, M., Nasr, S. Z., Evaluation of a cystic fibrosis transition program from pediatric to adult care, Pediatric Pulmonology, 48, 658-65, 2013	Descriptive data, closed ended questionnaire.
Claydon, A., Donaghy, B., Lea, S., An audit of the clinical pathway of transition of young people with cystic fibrosis to adult care, Journal of Cystic FibrosisJ Cyst Fibros, 11, S135, 2012	Audit of CF transition clinic. Conference abstract.
Conway, S., Cystic fibrosis in teenagers and young adults, Archives of Disease in Childhood, 75, 99-101, 1996	Commentary on transition.
Corman, Lindsay A., Factors that are associated with readiness to transition from pediatric to adult health care in cystic fibrosis, Dissertation Abstracts International: Section B: The Sciences and Engineering, 75, No Pagination Specified, 2015	Dissertation abstract.
Coyne, I. T., Breen, M., Dempsey, O., While, A., A systematic review of the transition process for young people with cystic fibrosis, Journal of Cystic FibrosisJ Cyst Fibros, 11, S139, 2012	Conference abstract.
Coyne, I. T., While, A. E., Sheehan, A. M., Transitioning of young people (with chronic illnesses) from child to adult healthcare services: Identifying needs from multiple perspectives from three disease groups, Journal of Adolescent HealthJ Adolesc Health, 1), S128, 2015	Conference abstract.
Craig, S. L., Towns, S., Bibby, H., Moving on from paediatric to adult health care: an initial evaluation of a transition program for young people with cystic fibrosis, International Journal of Adolescent Medicine & Health, 19, 333-43, 2007	Descriptive data, closed ended questionnaire.
Crosier, J., Wise, L. C., Coming of age. Cystic fibrosis--once a childhood disease--makes its way to adulthood, Nursing Management, 32, 30-1, 2001	Commentary, discussion
D'Angelo, Sandra L., Adaptation in parents of chronically ill children: The importance of social support, Dissertation Abstracts International, 51, 1490, 1990	Dissertation abstract.
Doug, M., Adi, Y., Williams, J., Paul, M., Kelly, D., Petchey, R., Carter, Y. H., Transition to adult services for children and young people with palliative care needs: a systematic review.[Reprint of Arch Dis Child. 2011 Jan;96(1):78-84; PMID: 19948663], BMJ supportive & palliative careBMJ support, 1, 167-73, 2011	Systematic review. Primary studies checked for inclusion in our review.
Doug, M., Adi, Y., Williams, J., Paul, M., Kelly, D., Petchey, R., Carter, Y. H., Transition to adult	Systematic review of studies.

Study	Reason for Exclusion
services for children and young people with palliative care needs: a systematic review, Archives of Disease in Childhood, 96, 78-84, 2011	
Eickenhorst, H., Cystic fibrosis and the emerging adult: managing the continuum, Continuum (Chicago), 16, 11-5, 1996	Discussion paper.
Flume, P. A., Anderson, D. L., Hardy, K. K., Gray, S., Transition programs in cystic fibrosis centers: perceptions of pediatric and adult program directors, Pediatric Pulmonology, 31, 443-50, 2001	Descriptive data, closed ended questionnaire.
Flume, P. A., Taylor, L. A., Anderson, D. L., Gray, S., Turner, D., Transition programs in cystic fibrosis centers: perceptions of team members, Pediatric Pulmonology, 37, 4-7, 2004	Descriptive data, closed ended questionnaire.
Hallowell, Sara Campbell, Transitioning from Adolescent to Adult Providers: Bridging the Gap in Pediatric Heart Transplant Care Through the Deevelopment of a Transition Program, NaN, 2013	Dissertation abstract.
Henderson, K., Henman, S., Shearing, V., Allen, R., Blatch, J., Johnson, C. J., The development of a website to facilitate transition from paediatric to adult CF services, Journal of Cystic FibrosisJ Cyst Fibros, 14, S9, 2015	Aim not relevant. Conference abstract.
Hewer, S. C., Tyrrell, J., Cystic fibrosis and the transition to adult health services, Archives of Disease in Childhood, 93, 817-21, 2008	Narrative review/ discussion paper.
Iles, N., Lowton, K., Young people with cystic fibrosis' concerns for their future: when and how should concerns be addressed, and by whom?, Journal of Interprofessional Care, 22, 436-8, 2008	No relevant qualitative results reported.
Jamieson, N., Fitzgerald, D., Singh-Grewal, D., Hanson, C. S., Craig, J. C., Tong, A., Children's experiences of cystic fibrosis: a systematic review of qualitative studies, Pediatrics, 133, e1683-97, 2014	Systematic review. Relevant studies checked for inclusion.
Leeman, J., Sandelowski, M., Havill, N. L., Knafl, K., Parent-to-Child Transition in Managing Cystic Fibrosis: A Research Synthesis, Journal of Family Theory & Review, 7, 167-183, 2015	Systematic synthesis. Included studies were checked for inclusion in our review.
Lowton, K., Mathes, L., Wyatt, H., Luce, P., While, A., Carmel, S., Evaluation of transition services for young people with cystic fibrosis in Southeast London, Journal of Interprofessional Care, 19, 408-9, 2005	Commentary, protocol.
Madge, S., Bryon, M., A model for transition from pediatric to adult care in cystic fibrosis, Journal of Pediatric Nursing, 17, 283-8, 2002	Quantitative data, closed ended questionnaire.
Marciel, Kristen K., Self-care independence and readiness to transfer from pediatric to adult health care in cystic fibrosis, Dissertation Abstracts International: Section B: The Sciences and Engineering, 69, 4431, 2009	Dissertation abstract.

Study	Reason for Exclusion
Massey-Chase, R. A., O'Dea, B., Reilly, C. C., Perrin, F., Macedo, P., Elston, C., Understanding young people's anxiety about transitioning from paediatric to adult services-a service improvement project, Journal of Cystic Fibrosis, 14, S133, 2015	Quantitative study, closed ended questionnaire. Conference abstract.
McDonald, G., Transition to adult cystic fibrosis care, RT: The Journal for Respiratory Care Practitioners, 13, 46-48, 2000	Commentary/ discussion paper.
Messick, J., A 21st-century approach to cystic fibrosis: optimizing outcomes across the disease spectrum, Journal of Pediatric Gastroenterology & Nutrition, 51 Suppl 7, S1-7; quiz 3 p following S7, 2010	Commentary. Narrative study.
Morgan, A. L., Culnane, E., Glazner, J., Reardon, N., When i am grown up - Evaluating the effectiveness of a transition program for adolescents with CF transferring to the adult hospital system, Journal of Cystic Fibrosis, 13, S116, 2014	Descriptive data, closed ended questionnaire.
Nasr, S. Z., Campbell, C., Howatt, W., Transition program from pediatric to adult care for cystic fibrosis patients, Journal of Adolescent Health, 13, 682-5, 1992	Descriptive data, closed ended questionnaire.
Okumura, M. J., Ong, T., Dawson, D., Nielson, D., Lewis, N., Richards, M., Brindis, C. D., Kleinhenz, M. E., Improving transition from paediatric to adult cystic fibrosis care: programme implementation and evaluation, BMJ Quality & Safety, 23 Suppl 1, i64-i72, 2014	Quantitative study, closed ended questionnaire.
Oxley, H., Woodhead, S., Biesty, J., Fauset-Jones, J., Trenchard, S., Exploring parents' concerns about transition from paediatric to adult cystic fibrosis (CF) services, Journal of Cystic Fibrosis. Conference: 36th European Cystic Fibrosis Conference. Lisbon Portugal. Conference Start, 12	Quantitative questionnaire survey. Conference abstract.
Reisinho, M. C., Gomes, B., OC37 - Adolescents with cystic fibrosis: their perspective, Nursing Children and Young People, 28, 80, 2016	Conference abstract.
Schlucter, Juliette, Dokken, Deborah, Ahmann, Elizabeth, Family Matters. Transitions from Pediatric to Adult Care: Programs and Resources, Pediatric Nursing, 41, 85-88, 2015	Discussion paper.
Silva Filho, L. V., Castanos, C., Ruiz, H. H., Cystic fibrosis in Latin America-Improving the awareness, Journal of Cystic FibrosisJ Cyst Fibros, 1, 1, 2016	Narrative review/ discussion paper.
Steinkamp, G., Ullrich, G., Muller, C., Fabel, H., von der Hardt, H., Transition of adult patients with cystic fibrosis from paediatric to adult care--the patients' perspective before and after start-up of an adult clinic, European Journal of Medical Research, 6, 85-92, 2001	Purely quantitative study.

Study	Reason for Exclusion
Tierney, S., Kirk, S., Deaton, C., Transferring young people with cystic fibrosis to adult care, Nursing Standard, 30, 41-7, 2015	Conference abstract.
Waldboth, V., Patch, C., Mahrer-Imhof, R., Metcalfe, A., Living a normal life in an extraordinary way: A systematic review investigating experiences of families of young people's transition into adulthood when affected by a genetic and chronic childhood condition, International Journal of Nursing StudiesInt J Nurs Stud, 62, 44-59, 2016	Systematic review. Included studies were checked for inclusion in our review.
Webb, A. K., Jones, A. W., Dodd, M. E., Transition from paediatric to adult care: problems that arise in the adult cystic fibrosis clinic, Journal of the Royal Society of Medicine, 94 Suppl 40, 8-11, 2001	Commentary, recommendation.
Westwood, A., Henley, L., Willcox, P., Transition from paediatric to adult care for persons with cystic fibrosis: patient and parent perspectives, Journal of Paediatrics & Child Health, 35, 442-5, 1999	Quantitative study.
While, A. E., Heery, E., Sheehan, A. M., Coyne, I., Health-related quality of life of young people with long-term illnesses before and after transfer from child to adult healthcare, Child: Care, Health & DevelopmentChild Care Health Dev, 27, 27, 2016	Quantitative survey.
Williams, B., Mukhopadhyay, S., Dowell, J., Coyle, J., From child to adult: an exploration of shifting family roles and responsibilities in managing physiotherapy for cystic fibrosis, Social Science & Medicine, 65, 2135-46, 2007	Aim of the study not relevant to the review (physiotherapy).
Wood, H. M., Phillips, S., The experiences of young people and their parents in the move from paediatric to adult cystic fibrosis services in Bristol: Recommendations for an improved transition pathway, Journal of Cystic FibrosisJ Cyst Fibros, 13, S116, 2014	Conference abstract.
Zack, J., Jacobs, C. P., Keenan, P. M., Harney, K., Woods, E. R., Colin, A. A., Emans, S. J., Perspectives of patients with cystic fibrosis on preventive counseling and transition to adult care, Pediatric Pulmonology, 36, 376-83, 2003	No relevant qualitative findings
Zhou, H., Roberts, P., Dhaliwal, S., Della, P., Transitioning adolescent and young adults with chronic disease and/or disabilities from paediatric to adult care services - an integrative review, Journal of Clinical NursingJ Clin Nurs, 25, 3113-3130, 2016	Integrative review. Included studies were checked for inclusion in our review.

H.5 Complications of cystic fibrosis

Study	Reason for Exclusion
Abman, S. H., Accurso, F. J., Bowman, C. M., Persistent morbidity and mortality of protein calorie malnutrition in young infants with CF,	Sample size <251

Study	Reason for Exclusion
Journal of Pediatric Gastroenterology & Nutrition, 5, 393-6, 1986	
Bahmanyar, S., Ekbom, A., Askling, J., Johannesson, M., Montgomery, S. M., Cystic fibrosis gene mutations and gastrointestinal diseases, Journal of Cystic Fibrosis, 9, 288-291, 2010	Complications not relevant to this review
Bertenshaw, C., Watson, A. R., Lewis, S., Smyth, A., Survey of acute renal failure in patients with cystic fibrosis in the UK, Thorax, 62, 541-545, 2007	Incidence data, no prevalence data
Blackman, S. M., Hsu, S., Vanscoy, L. L., Collaco, J. M., Ritter, S. E., Naughton, K., Cutting, G. R., Genetic modifiers play a substantial role in diabetes complicating cystic fibrosis, Journal of Clinical Endocrinology & Metabolism, 94, 1302-9, 2009	Dates related to prevalence data were not specified
Blackwell, K., Malone, P. S., Denny, A., Connett, G., Maddison, J., The prevalence of stress urinary incontinence in patients with cystic fibrosis: an under-recognized problem, Journal of pediatric urology, 1, 5-9, 2005	Sample size <251
Booth, E., Cystic fibrosis-related diabetes, Journal of Pediatric Nursing, 26, 604-6, 2011	Narrative review
Browne, W. J., Wood, C. J., Desai, M., Weller, P. H., Urinary incontinence in 9-16 year olds with cystic fibrosis compared to other respiratory conditions and a normal group, Journal of Cystic Fibrosis, 8, 50-7, 2009	Sample size <251
Buchdahl, R., Is there a gender gap in cystic fibrosis children?, Journal of the Royal Society of Medicine, Supplement, 100, 35-37, 2007	Narrative review
Burge, A. T; Holland, A. E. , Prevalence and impact of incontinence in adult men with cystic fibrosis, Respirology, 16, 2011 (Study identified by stakeholders at consultation)	Sample size <251
Ciro, D., Padoan, R., Blau, H., Marostica, A., Fuoti, M., Volpi, S., Pilotta, A., Meyerovitch, J., Sher, D., Assael, B. M., Growth retardation and reduced growth hormone secretion in cystic fibrosis. Clinical observations from three CF centers, Journal of Cystic Fibrosis, 12, 165-9, 2013	Unclear whether exclusion criteria were applied before or after identifying the people whose height was under the 3rd percentile. 1 of the 3 centres is from Israel.
Colombo, C., Littlewood, J., The implementation of standards of care in Europe: state of the art, Journal of Cystic Fibrosis, 10 Suppl 2, S7-15, 2011	Narrative review
Cotellessa, M., Minicucci, L., Diana, M. C., Prigione, F., Di Febbraro, L., Gagliardini, R., Manca, A., Battistini, F., Taccetti, G., Magazzu, G., Padoan, R., Pizzamiglio, G., Raia, V., Iapichino, L., Cardella, F., Grinzich, G., Lucidi, V., Tuccio, G., Bignamini, E., Salvatore, D., Lorini, R., Phenotype/genotype correlation and cystic fibrosis related diabetes mellitus (Italian	More recent prevalence data on CFRD available from other studies

Study	Reason for Exclusion
Multicenter Study), Journal of Pediatric Endocrinology, 13, 1087-93, 2000	
Del Ciampo, I. R., Sawamura, R., Fernandes, M. I., Cystic fibrosis: from protein-energy malnutrition to obesity with dyslipidemia, Iranian Journal of Pediatrics, 23, 605-6, 2013	Case report
Dogan, Y., Erkan, T., Yalvac, S., Altay, S., Cokugras, F. C., Aydin, A., Kutlu, T., Nutritional status of patients hospitalized in pediatric clinic, Turkish Journal of Gastroenterology, 16, 212-6, 2005	Number of people with CF included in the study <251
Erkkila, J. C., Warwick, W. J., Bradford, D. S., Spine deformities and cystic fibrosis, Clinical Orthopaedics & Related Research, 146-50, 1978	Sample size < 251
Fainardi, V., Koo, S. D., Padley, S. P., Lam, S. H., Bush, A., Prevalence of scoliosis in cystic fibrosis, Pediatric Pulmonology, 48, 553-5, 2013	Non relevant complication
Farrelly, P. J., Charlesworth, C., Lee, S., Southern, K. W., Baillie, C. T., Gastrointestinal surgery in cystic fibrosis: a 20-year review, Journal of Pediatric Surgery, 49, 280-3, 2014	Relevant sample size (sample size used to calculate the percentage of people treated for DIOS) was <251
Fendler, W., Borowiec, M., Baranowska-Jazwiecka, A., Szadkowska, A., Skala-Zamorowska, E., Deja, G., Jarosz-Chobot, P., Techmanska, I., Bautembach-Minkowska, J., Mysliwiec, M., Zmyslowska, A., Pietrzak, I., Malecki, M. T., Mlynarski, W., Prevalence of monogenic diabetes amongst Polish children after a nationwide genetic screening campaign, Diabetologia, 55, 2631-5, 2012	Indirect population. No prevalence data relating to people with CF only.
FitzSimmons, S. C., The changing epidemiology of cystic fibrosis, Current problems in pediatrics, 24, 171-179, 1994	More recent prevalence data on relevant complications available from other studies
FitzSimmons, S. C., The changing epidemiology of cystic fibrosis, Journal of Pediatrics, 122, 1-9, 1993	More recent prevalence data on relevant complications available from other studies
Frohnert, B. I., Ode, K. L., Moran, A., Nathan, B. M., Laguna, T., Holme, B., Thomas, W., Impaired fasting glucose in cystic fibrosis, Diabetes Care, 33, 2660-4, 2010	More recent prevalence data on CFRD available from other studies
Gumery, L; Lee, J; Whitehouse, J., The prevalence of urinary incontinence in adult cystic fibrosis males [abstract] , Journal of Cystic Fibrosis, 4, 2005 (Study identified by stakeholders at consultation)	Conference abstract. Sample size <251
Hall, W. B., Sparks, A. A., Aris, R. M., Vitamin d deficiency in cystic fibrosis, International Journal of Endocrinology Print, 2010, 218691, 2010	Narrative review
Hamosh, A., FitzSimmons, S. C., Macek, M., Jr., Knowles, M. R., Rosenstein, B. J., Cutting, G. R., Comparison of the clinical manifestations of cystic fibrosis in black and white patients, Journal of Pediatrics, 132, 255-9, 1998	More recent data on prevalence of DIOS available from another study
Havermans, T., Colpaert, K., De Boeck, K., Dupont, L., Abbott, J., Pain in CF: Review of the	Narrative review

Study	Reason for Exclusion
literature, Journal of Cystic Fibrosis, 12, 423-430, 2013	
Hayes, D., Jr., Patel, A. V., Black, S. M., McCoy, K. S., Kirkby, S., Tobias, J. D., Mansour, H. M., Whitson, B. A., Influence of diabetes on survival in patients with cystic fibrosis before and after lung transplantation, Journal of Thoracic & Cardiovascular Surgery, 150, 707-13.e2, 2015	Sample is not representative of most people with CF
Humphries, I. R., Allen, J. R., Waters, D. L., Howman-Giles, R., Gaskin, K. J., Volumetric bone mineral density in children with cystic fibrosis, Applied Radiation & Isotopes, 49, 593-5, 1998	Number of people with CF included in the study <251
Koch, C., Cuppens, H., Rainisio, M., Madessani, U., Harms, H., Hodson, M., Mastella, G., Navarro, J., Strandvik, B., McKenzie, S., Investigators of the, Ercf, European Epidemiologic Registry of Cystic Fibrosis (ERCF): comparison of major disease manifestations between patients with different classes of mutations, Pediatric Pulmonology, 31, 1-12, 2001	More recent prevalence data on relevant complications available from other studies
Koch, C., Rainisio, M., Madessani, U., Harms, H. K., Hodson, M. E., Mastella, G., McKenzie, S. G., Navarro, J., Strandvik, B., Investigators of the European Epidemiologic Registry of Cystic, Fibrosis, Presence of cystic fibrosis-related diabetes mellitus is tightly linked to poor lung function in patients with cystic fibrosis: data from the European Epidemiologic Registry of Cystic Fibrosis, Pediatric Pulmonology, 32, 343-50, 2001	More recent prevalence data on CFRD available from other studies
Konrad, K., Scheuing, N., Badenhoop, K., Borkenstein, M. H., Gohlke, B., Schofl, C., Seufert, J., Thon, A., Holl, R. W., Cystic fibrosis-related diabetes compared with type 1 and type 2 diabetes in adults, Diabetes/Metabolism Research Reviews, 29, 568-75, 2013	No prevalence data on CFRD
Konstan, M. W., Butler, S. M., Wohl, M. E. B., Stoddard, M., Matousek, R., Wagener, J. S., Johnson, C. A., Morgan, W. J., Growth and nutritional indexes in early life predict pulmonary function in cystic fibrosis, Journal of Pediatrics, 142, 624-630, 2003	More recent prevalence data on malnutrition and growth available from other studies
Kumar, N., Balachandran, S., Millner, P. A., Littlewood, J. M., Conway, S. P., Dickson, R. A., Scoliosis in cystic fibrosis: is it idiopathic?, Spine, 29, 1990-5, 2004	Non relevant complication
Lagrange-Puget, M., Durieu, I., Ecochard, R., Abbas-Chorfa, F., Drai, J., Steghens, J. P., Pacheco, Y., Vital-Durand, D., Bellon, G., Longitudinal study of oxidative status in 312 cystic fibrosis patients in stable state and during bronchial exacerbation, Pediatric Pulmonology, 38, 43-9, 2004	Results presented as mean values. No prevalence data on vitamin deficiency.
Lai, H. C., Corey, M., FitzSimmons, S., Kosorok, M. R., Farrell, P. M., Comparison of growth	More recent prevalence data available on malnutrition and growth

Study	Reason for Exclusion
status of patients with cystic fibrosis between the United States and Canada, American Journal of Clinical Nutrition, 69, 531-8, 1999	
Lai, H. C., Kosorok, M. R., Sondel, S. A., Chen, S. T., FitzSimmons, S. C., Green, C. G., Shen, G., Walker, S., Farrell, P. M., Growth status in children with cystic fibrosis based on the National Cystic Fibrosis Patient Registry data: evaluation of various criteria used to identify malnutrition, Journal of Pediatrics, 132, 478-85, 1998	More recent prevalence data available on malnutrition and growth
Lanng,S., Thorsteinsson,B., Lund-Andersen,C., Nerup,J., Schiotz,P.O., Koch,C., Diabetes mellitus in Danish cystic fibrosis patients: prevalence and late diabetic complications, Acta Paediatrica, 83, 72-77, 1994	More recent data available from other studies on CFRD
Lavie, M., Manovitz, T., Vilozni, D., Levy-Mendelovich, S., Sarouk, I., Weintraubv, I., Shoseyov, D., Cohen-Cymberknoh, M., Rivlin, J., Efrati, O., Long-term follow-up of distal intestinal obstruction syndrome in cystic fibrosis, World Journal of Gastroenterology, 21, 318-325, 2015	Context not relevant in relation to prevalence of DIOS, because the climatic conditions may increase the risk of dehydration. Prevalence data on DIOS from a Western European country was available from another study.
Lefaucheur, C., Nochy, D., Amrein, C., Chevalier, P., Guillemain, R., Cherif, M., Jacquot, C., Glotz, D., Hill, G. S., Renal histopathological lesions after lung transplantation in patients with cystic fibrosis, American Journal of Transplantation, 8, 1901-10, 2008	Sample size <251
Li, L., Somerset, S., Dietary intake and nutritional status of micronutrients in adults with cystic fibrosis in relation to current recommendations, Clinical Nutrition, 35, 775-82, 2016	Narrative review
Lind-Ayres, M., Thomas, W., Holme, B., Mauer, M., Caramori, M. L., Moran, A., Microalbuminuria in patients with cystic fibrosis, Diabetes Care, 34, 1526-8, 2011	Prevalence data is not relevant because it is not disaggregated by CFRD status.
Liou, T. G., Jensen, J. L., Allen, S. E., Brayshaw, S. J., Brown, M. A., Chatfield, B., Koenig, J., McDonald, C., Packer, K. A., Peet, K., Radford, P., Reineke, L. M., Otsuka, K., Wagener, J. S., Young, D., Marshall, B. C., Improving performance in the detection and management of cystic fibrosis-related diabetes in the Mountain West Cystic Fibrosis Consortium, BMJ Open Diabetes Research & Care, 4, e000183, 2016	More recent prevalence data on CFRD available from other studies
Logvinoff, M. M., Fon, G. T., Taussig, L. M., Pitt, M. J., Kyphosis and pulmonary function in cystic fibrosis, Clinical Pediatrics, 23, 389-92, 1984	Sample size <251
Marshall, B. C., Butler, S. M., Stoddard, M., Moran, A. M., Liou, T. G., Morgan, W. J., Epidemiology of cystic fibrosis-related diabetes, Journal of Pediatrics, 146, 681-7, 2005	More recent data on prevalence of CFRD available from other studies

Study	Reason for Exclusion
Milla,C.E., Billings,J., Moran,A., Diabetes is associated with dramatically decreased survival in female but not male subjects with cystic fibrosis, Diabetes Care, 28, 2141-2144, 2005	More recent data on prevalence of CFRD available from other studies
Morgan, W. J., Butler, S. M., Johnson, C. A., Colin, A. A., FitzSimmons, S. C., Geller, D. E., Konstan, M. W., Light, M. J., Rabin, H. R., Regelmann, W. E., Schidlow, D. V., Stokes, D. C., Wohl, M. E. B., Kaplowitz, H., Wyatt, M. M., Stryker, S., Epidemiologic study of cystic fibrosis: Design and implementation of a prospective, multicenter, observational study of patients with cystic fibrosis in the U.S. and Canada, Pediatric Pulmonology, 28, 231-241, 1999	More recent prevalence data on relevant complications available from other studies
Morison, S., Dodge, J. A., Cole, T. J., Lewis, P. A., Coles, E. C., Geddes, D., Russell, G., Littlewood, J. M., Scott, M. T., Height and weight in cystic fibrosis: a cross sectional study. UK Cystic Fibrosis Survey Management Committee, Archives of Disease in Childhood, 77, 497-500, 1997	No prevalence data
Munck, A., Alberti, C., Colombo, C., Kashirskaya, N., Ellemunter, H., Fotoulaki, M., Houwen, R., Robberecht, E., Boizeau, P., Wilschanski, M., C. F/Pancreas ESPGHAN Working Group, Dios Study Group, International prospective study of distal intestinal obstruction syndrome in cystic fibrosis: Associated factors and outcome, Journal of Cystic Fibrosis, 15, 531-9, 2016	Incidence data; no prevalence data on DIOS
Nankivell,G., Caldwell,P., Follett,J., Urinary Incontinence in Adolescent Females with Cystic Fibrosis, Paediatric Respiratory Reviews, 11, 95-99, 2010 (Study identified by stakeholders at consultation)	Literature review
Olesen, H. V., Pressler, T., Hjelte, L., Mared, L., Lindblad, A., Knudsen, P. K., Laerum, B. N., Johannesson, M., Scandinavian Cystic Fibrosis Study, Consortium, Gender differences in the Scandinavian cystic fibrosis population, Pediatric Pulmonology, 45, 959-65, 2010	More recent data on prevalence of CFRD available from other studies
Orr, A., McVean, R. J., Webb, A. K., Dodd, M. E., Questionnaire survey of urinary incontinence in women with cystic fibrosis, BMJ, 322, 1521, 2001 (Study identified by stakeholders at consultation)	Sample size <251
Paccou, J., Zeboulon, N., Combescure, C., Gossec, L., Cortet, B., The prevalence of osteoporosis, osteopenia, and fractures among adults with cystic fibrosis: a systematic literature review with meta-analysis, Calcified Tissue International, 86, 1-7, 2010	The systematic review was rated as of low quality using the AMSTAR checklist. The included studies were assessed for inclusion in the current systematic review.
Parasa, R. B., Maffulli, N., Musculoskeletal involvement in cystic fibrosis, Bulletin of the Hospital for Joint Diseases, 58, 37-44, 1999	Narrative review

Study	Reason for Exclusion
Parkins, M. D., Parkins, V. M., Rendall, J. C., Elborn, S., Changing epidemiology and clinical issues arising in an ageing cystic fibrosis population, Therapeutic Advances in Respiratory Disease, 5, 105-19, 2011	Narrative review
Patel, E. M., Swamy, G. K., Heine, R. P., Kuller, J. A., James, A. H., Grotegut, C. A., Medical and obstetric complications among pregnant women with cystic fibrosis, American Journal of Obstetrics & Gynecology, 212, 98.e1-9, 2015	Non relevant population
Pawellek, I., Dokoupil, K., Koletzko, B., Prevalence of malnutrition in paediatric hospital patients, Clinical Nutrition, 27, 72-6, 2008	Number of people with CF <251
Prasad, S. A., Balfour-Lynn, I. M., Carr, S. B., Madge, S. L., A comparison of the prevalence of urinary incontinence in girls with cystic fibrosis, asthma, and healthy controls, Pediatric Pulmonology, 41, 1065-8, 2006 (Study identified by stakeholders at consultation)	Sample size <251
Putman, M. S., Baker, J. F., Uluer, A., Herlyn, K., Lapey, A., Sicilian, L., Tillotson, A. P., Gordon, C. M., Merkel, P. A., Finkelstein, J. S., Trends in bone mineral density in young adults with cystic fibrosis over a 15 year period, Journal of Cystic FibrosisJ Cyst Fibros, 14, 526-32, 2015	Sample size <251
Quon, B. S., Aitken, M. L., Cystic fibrosis: what to expect now in the early adult years, Paediatric Respiratory Reviews, 13, 206-14, 2012	Narrative review
Quon, B. S., Mayer-Hamblett, N., Aitken, M. L., Goss, C. H., Risk of post-lung transplant renal dysfunction in adults with cystic fibrosis, Chest, 142, 185-91, 2012	Incidence data; no prevalence data
Rana, M., Munns, C. F., Selvadurai, H. C., Simonds, S., Cooper, P. J., Woodhead, H. J., Hameed, S., Verge, C. F., Lafferty, A. R., Crock, P. A., Craig, M. E., Increased detection of cystic-fibrosis-related diabetes in Australia, Archives of Disease in Childhood, 96, 823-6, 2011	Incidence only, no prevalence data
Ravilly, S., Robinson, W., Suresh, S., Wohl, M. E., Berde, C. B., Chronic pain in cystic fibrosis, Pediatrics, 98, 741-7, 1996	Sample size < 251
Reisman, J., Corey, M., Canny, G., Levison, H., Diabetes mellitus in patients with cystic fibrosis: effect on survival, Pediatrics, 86, 374-7, 1990	More recent prevalence data on CFRD available from other studies
Rosenecker, J., Eichler, I., Kuhn, L., Harms, H. K., von der Hardt, H., Genetic determination of diabetes mellitus in patients with cystic fibrosis. Multicenter Cystic Fibrosis Study Group, Journal of Pediatrics, 127, 441-3, 1995	More recent data on prevalence of CFRD available from other studies
Salvatore, D., Buzzetti, R., Baldo, E., Furnari, M. L., Lucidi, V., Manunza, D., Marinelli, I., Messore, B., Neri, A. S., Raia, V., Mastella, G., An overview of international literature from cystic fibrosis registries. Part 4: update 2011, Journal of Cystic Fibrosis, 11, 480-93, 2012	Narrative review

Study	Reason for Exclusion
Scheuing, N., Holl, R. W., Dockter, G., Hermann, J. M., Junge, S., Koerner-Rettberg, C., Naehrlich, L., Smaczny, C., Staab, D., Thalhammer, G., van Koningsbruggen-Rietschel, S., Ballmann, M., High variability in oral glucose tolerance among 1,128 patients with cystic fibrosis: a multicenter screening study, PLoS ONE [Electronic Resource], 9, e112578, 2014	Relevant data only presented graphically
Scheuing,N., Holl,R.W., Dockter,G., Fink,K., Junge,S., Naehrlich,L., Smaczny,C., Staab,D., Thalhammer,G., van Koningsbruggen-Rietschel,S., Ballmann,M., Diabetes in cystic fibrosis: multicenter screening results based on current guidelines, PLoS ONE [Electronic Resource], 8, e81545-, 2013	People with a confirmed diagnosis of CFRD or using anti-hyperglycemic treatment were excluded
Schwarzenberg,S.J., Thomas,W., Olsen,T.W., Grover,T., Walk,D., Milla,C., Moran,A., Microvascular complications in cystic fibrosis-related diabetes, Diabetes Care, 30, 1056-1061, 2007	More recent prevalence data on CFRD available from other studies; sample size used to calculate the prevalence of microalbuminuria was too small.
Sharma, A., Morton, A., Peckham, D., Jayne, D., Gastrointestinal surgery in adult patients with cystic fibrosis, Frontline Gastroenterology, 3, 242-247, 2012	No prevalence data on DIOS. Only prevalence data relating to operations for treatment of complications of DIOS.
Sokol, R. J., Butler-Simon, N., Heubi, J. E., Iannaccone, S. T., McClung, H. J., Accurso, F., Hammond, K., Heyman, M., Sinatra, F., Riely, C., et al.,, Vitamin E deficiency neuropathy in children with fat malabsorption. Studies in cystic fibrosis and chronic cholestasis, Annals of the New York Academy of Sciences, 570, 156-69, 1989	Sample size < 251
Solomon, M. P., Wilson, D. C., Corey, M., Kalnins, D., Zielenski, J., Tsui, L. C., Pencharz, P., Durie, P., Sweezey, N. B., Glucose intolerance in children with cystic fibrosis, Journal of Pediatrics, 142, 128-32, 2003	People with CF with CFRD were excluded.
Stankovic Stojanovic, K., Hubert, D., Leroy, S., Dominique, S., Grenet, D., Colombat, M., Clement, A., Fayon, M., Grateau, G., Cystic fibrosis and AA amyloidosis: a survey in the French cystic fibrosis network, Amyloid, 21, 231-7, 2014	Size of CF population not specified precisely ("CF prevalence in France is approximately 6000 patients")
Steinkamp, G., Wiedemann, B., Relationship between nutritional status and lung function in cystic fibrosis: cross sectional and longitudinal analyses from the German CF quality assurance (CFQA) project, Thorax, 57, 596-601, 2002	No relevant indicator. Data too old to be relevant.
Stephens, S. E., Rigden, S. P. A., Cystic fibrosis and renal disease, Paediatric Respiratory Reviews, 3, 135-138, 2002	Narrative review
Stephenson, A., Jamal, S., Dowdell, T., Pearce, D., Corey, M., Tullis, E., Prevalence of vertebral fractures in adults with cystic fibrosis and their relationship to bone mineral density, Chest, 130, 539-44, 2006	Sample size <251

Study	Reason for Exclusion
Sterescu, A. E., Rhodes, B., Jackson, R., Dupuis, A., Hanna, A., Wilson, D. C., Tullis, E., Pencharz, P. B., Natural history of glucose intolerance in patients with cystic fibrosis: ten-year prospective observation program, Journal of Pediatrics, 156, 613-7, 2010	People who were already diagnosed with CFRD before 1996 were excluded
Subhi, R., Ooi, R., Finlayson, F., Kotsimbos, T., Wilson, J., Lee, W. R., Wale, R., Warrier, S., Distal intestinal obstruction syndrome in cystic fibrosis: presentation, outcome and management in a tertiary hospital (2007-2012), ANZ Journal of Surgery, 84, 740-4, 2014	No prevalence data
Usatin, D. J., Perito, E. R., Posselt, A. M., Rosenthal, P., Under Utilization of Pancreas Transplants in Cystic Fibrosis Recipients in the United Network Organ Sharing (UNOS) Data 1987-2014, American Journal of Transplantation, 16, 1620-5, 2016	Sample is not representative of most people with CF
van den Berg, J. M., Kouwenberg, J. M., Heijerman, H. G., Demographics of glucose metabolism in cystic fibrosis, Journal of Cystic Fibrosis, 8, 276-9, 2009	Sample size < 251
van der Doef, H. P., Kokke, F. T., Beek, F. J., Woestenenk, J. W., Froeling, S. P., Houwen, R. H., Constipation in pediatric cystic fibrosis patients: an underestimated medical condition, Journal of Cystic Fibrosis, 9, 59-63, 2010	Sample size < 251; people with a history of DIOS were excluded
VanDevanter, D. R., Kahle, J. S., O'Sullivan, A. K., Sikirica, S., Hodgkins, P. S., Cystic fibrosis in young children: A review of disease manifestation, progression, and response to early treatment, Journal of Cystic Fibrosis, 15, 147-57, 2016	Narrative review
White, D; K, Stiller., The prevalence and severity of symptoms of incontinence in adult cystic fibrosis patients, Physiotherapy Theory & Practice, 16, 35-43, 2000 (Study identified by stakeholders at consultation)	Sample size <251
White, M., Dennis, N., Ramsey, R., Barwick, K., Graham, C., Kane, S., Kepreotes, H., Queit, L., Sweeney, A., Winderlich, J., Wong See, D., Littlewood, R., Prevalence of malnutrition, obesity and nutritional risk of Australian paediatric inpatients: a national one-day snapshot, Journal of Paediatrics & Child Health, 51, 314-20, 2015	Number of people with CF <251
Wiedemann, B., Paul, K. D., Stern, M., Wagner, T. O., Hirche, T. O., German, Cfqa Group, Evaluation of body mass index percentiles for assessment of malnutrition in children with cystic fibrosis, European Journal of Clinical Nutrition, 61, 759-68, 2007	Prevalence data disaggregated by sex. No overall prevalence data.
Wilson, D. C., Kalnins, D., Stewart, C., Hamilton, N., Hanna, A. K., Durie, P. R., Tullis, E., Pencharz, P. B., Challenges in the dietary treatment of cystic fibrosis related diabetes mellitus, Clinical Nutrition, 19, 87-93, 2000	Narrative review

Study	Reason for Exclusion
Ye, X., Kuo, H. T., Sampaio, M. S., Jiang, Y., Bunnapradist, S., Risk factors for development of new-onset diabetes mellitus after transplant in adult lung transplant recipients, Clinical Transplantation, 25, 885-91, 2011	Sample is not representative of most people with CF
Yen, E. H., Quinton, H., Borowitz, D., Better nutritional status in early childhood is associated with improved clinical outcomes and survival in patients with cystic fibrosis, Journal of Pediatrics, 162, 530-535.e1, 2013	More recent prevalence data available on malnutrition and growth
Yung, B., Hodson, M. E., Diabetes in cystic fibrosis, Journal of the Royal Society of Medicine, 92 Suppl 37, 35-40, 1999	Narrative review
Zhang, Z., Lai, H. J., Comparison of the use of body mass index percentiles and percentage of ideal body weight to screen for malnutrition in children with cystic fibrosis, American Journal of Clinical Nutrition, 80, 982-91, 2004	No overall prevalence data. Prevalence data was disaggregated by short, average and tall stature of people with CF.
Zhang, Z., Shoff, S. M., Lai, H. J., Comparing the Use of Centers for Disease Control and Prevention and World Health Organization Growth Charts in Children with Cystic Fibrosis through 2 Years of Age, Journal of Pediatrics, 167, 1089-95, 2015	Prevalence data in the paper was disaggregated by sex; no overall prevalence data was provided.

H.6 Pulmonary monitoring

Study	Reason for Exclusion
Aaron, S. D., Kottachchi, D., Ferris, W. J., Vandemheen, K. L., St Denis, M. L., Plouffe, A., Doucette, S. P., Saginur, R., Chan, F. T., Ramotar, K., Sputum versus bronchoscopy for diagnosis of Pseudomonas aeruginosa biofilms in cystic fibrosis, European Respiratory Journal, 24, 631-7, 2004	Aim and outcomes not relevant to the protocol. The aim of this study was to determine whether bacteria adherent to the airways are genotipically or phenotipically different from the bacteria that are traditionally retreived from sputum cultures. No outcomes of interest are reported.
Agostini, M., Barlocco, G., Bonomi, U., Cappelletti, L. M., Castellani, L., Conforti, M., et al.,, Alternative antibiotics against Pseudomonas infections in cystic fibrosis. In vitro activity, pharmacokinetics, and double-blind randomized clinical trial with azlocillin, piperacillin, cefoperazone, ceftazidime, cefsulodin, cefotaxime and moxalactam. Preliminary results, Drugs under experimental and clinical research, 98, 671-86, 1983	Aim not relevant to the protocol. The study investigates in vitro activity, clinical effectiveness and pharmacokinetics of antibiotics against Pseudomonas infection.
Ahmed, B., Bush, A., Davies, J. C., How to use: bacterial cultures in diagnosing lower respiratory tract infections in cystic fibrosis, Archives of Disease in Childhood Education & Practice, 99, 181-7, 2014	Aim and study design not related to the protocol: review about diagnostic accuracy of bacterial cultures.
Alden, B., Kavanagh, C., Larner, E., Is bronchoalveolar lavage necessary for monitoring and treating respiratory infections in children	Conference abstract. Study design: not a RCT.

Study	Reason for Exclusion
with cystic fibrosis?, Journal of Cystic Fibrosis, 13, S82, 2014	
Almeida, M. B., Bussamra, M. H., Rodrigues, J. C., ABPA diagnosis in cystic fibrosis patients: the clinical utility of IgE specific to recombinant Aspergillus fumigatus allergens, Jornal de Pediatria, 82, 215-20, 2006	Intervention/ comparison: the paper does not include any comparison between monitoring strategies.
Anjorin, A., Schmidt, H., Posselt, H. G., Smaczny, C., Ackermann, H., Deimling, M., Vogl, T. J., Abolmaali, N., Comparative evaluation of chest radiography, low-field MRI, the Shwachman-Kulczycki score and pulmonary function tests in patients with cystic fibrosis, European Radiology, 18, 1153-61, 2008	Aim not related to the protocol. The aim of this study was to investigate whether the parenchymal lung damage in patients suffering from CF can be equivalently quantified by the Chrispin-Norman (CN) scores determined with low-field magnetic MRI and conventional chest X-ray, but only provides data on correlations.
Armstrong, D. S., Grimwood, K., Carlin, J. B., Carzino, R., Gutierrez, J. P., Hull, J., Olinsky, A., Phelan, E. M., Robertson, C. F., Phelan, P. D., Lower airway inflammation in infants and young children with cystic fibrosis, American Journal of Respiratory & Critical Care Medicine, 156, 1197-204, 1997	Intervention/ comparison not relevant to the protocol. The study does not compare monitoring strategies. It examines test results in different population groups and in the same group over time. No outcomes of interest are reported.
Armstrong, D. S., Grimwood, K., Carlin, J. B., Carzino, R., Olinsky, A., Phelan, P. D., Bronchoalveolar lavage or oropharyngeal cultures to identify lower respiratory pathogens in infants with cystic fibrosis, Pediatric Pulmonology, 21, 267-75, 1996	Aim not relevant to the protocol. The study aims to compare oropharyngeal and BAL cultures in infant with CF but only diagnostic accuracy data in reported.
Armstrong,D., Grimwood,K., Carlin,J.B., Carzino,R., Hull,J., Olinsky,A., Phelan,P.D., Severe viral respiratory infections in infants with cystic fibrosis, Pediatric Pulmonology, 26, 371-379, 1998	Aim of the study not relevant to the protocol. This study aims to determine the effect of severe viral respiratory infections upon CF infant lung disease. It does not compare different monitoring strategies.
Aurora, P., Bush, A., Gustafsson, P., Oliver, C., Wallis, C., Price, J., Stroobant, J., Carr, S., Stocks, J., London Cystic Fibrosis, Collaboration, Multiple-breath washout as a marker of lung disease in preschool children with cystic fibrosis, American Journal of Respiratory & Critical Care Medicine, 171, 249-56, 2005	Aim of the study not relevant to the protocol. This study aims to compare multiple breath washout and spirometry in children with CF and healthy children.
Aurora, P., Stanojevic, S., Wade, A., Oliver, C., Kozlowska, W., Lum, S., Bush, A., Price, J., Carr, S. B., Shankar, A., Stocks, J., London Cystic Fibrosis, Collaboration, Lung clearance index at 4 years predicts subsequent lung function in children with cystic fibrosis, American Journal of Respiratory & Critical Care Medicine, 183, 752-8, 2011	Aim of the study not relevant to the protocol.This study aimed to determine whether multiple-breath washout (MBW) results at preschool age can predict subsequent abnormal lung function, but only diagnostic accuracy data in reported.
Bakker, E. M., Borsboom, G. J., van der Wiel-Kooij, E. C., Caudri, D., Rosenfeld, M., Tiddens, H. A., Small airway involvement in cystic fibrosis lung disease: routine spirometry as an early and sensitive marker, Pediatric Pulmonology, 48, 1081-8, 2013	Aim of the study not relevant to the protocol. This study aimed to evaluate the progression of selected spirometry parameters with age in a cohort of CF patients and healthy children aged 6 to 20 years.
Bayramoglu,S., Cimilli,T., Aksoy,S., Yildiz,S., Salihoglu,B., Hatipoglu,S., Celiker,F.B., The role of HRCT versus CXR in children with recurrent	Study design: case series, does not meet protocol criteria for inclusion. Also authors do not report on outcomes of interest for our review.

Study	Reason for Exclusion
pulmonary infections, Clinical Imaging, 29, 317-324, 2005	
Beardsmore, C. S., Respiratory physiological measurements in infants with cystic fibrosis, Pediatric Pulmonology - Supplement, 7, 38-41, 1991	Narrative review, discussion paper.
Blau, H., Linnane, B., Carzino, R., Tannenbaum, E. L., Skoric, B., Robinson, P. J., Robertson, C., Ranganathan, S. C., Induced sputum compared to bronchoalveolar lavage in young, non-expectorating cystic fibrosis children, Journal of Cystic Fibrosis, 13, 106-10, 2014	Aim not related to the protocol. The aim of this study is to compare induced sputum with BAL in young children, non-expectorating CF children in terms of feasibility and safety. No outcomes of interest were reported.
Bortoluzzi, C. F., Volpi, S., D'Orazio, C., Loeve, M., Tiddens, H. A., Assael, B. M., Baseline chest computed tomography scores correlate with future exacerbations in young children with cystic fibrosis, European Respiratory Journal, 42, 2013	Conference abstract. Study design: not a RCT.
Bortoluzzi, C. F., Volpi, S., D'Orazio, C., Loeve, M., Tiddens, H. A., Assael, B. M., Significant lung damage revealed at early chest computed tomography is related to subsequent pseudomonas (PsA) colonization in children with cystic fibrosis, European Respiratory Journal, 42, 2013	Conference abstract. Study design: not a RCT.
Bortoluzzi, C. F., Volpi, S., D'Orazio, C., Tiddens, H. A., Loeve, M., Tridello, G., Assael, B. M., Bronchiectases at early chest computed tomography in children with cystic fibrosis are associated with increased risk of subsequent pulmonary exacerbations and chronic pseudomonas infection, Journal of Cystic Fibrosis, 13, 564-71, 2014	Intervention/ comparison: does not compare strategies and does not measure response following an exacerbation.
Brody, A. S., Sucharew, H., Campbell, J. D., Millard, S. P., Molina, P. L., Klein, J. S., Quan, J., Computed tomography correlates with pulmonary exacerbations in children with cystic fibrosis, American Journal of Respiratory & Critical Care Medicine, 172, 1128-32, 2005	Aim and outcomes not relevant to the protocol. This study aimed to validate HRCT against a true outcome (number of pulmonary exacerbations), and to assess the correlation of CT scores and PFTs with this clinical outcomes. Only correlation data is provided.
Byrne, N. M., O'Brien, C., Spencer, D. A., The efficacy of cough plates in the identification of bacterial pathogens in children with cystic fibrosis, Archives of Disease in Childhood, 98, 768-71, 2013	Outcomes not relevant to the protocol.
Byrnes, C. A., Vidmar, S., Cheney, J. L., Carlin, J. B., Armstrong, D. S., Cooper, P. J., Grimwood, K., Moodie, M., Robertson, C. F., Rosenfeld, M., Tiddens, H. A., Wainwright, C. E., Acfbal Study Investigators, Prospective evaluation of respiratory exacerbations in children with cystic fibrosis from newborn screening to 5 years of age, Thorax, 68, 643-51, 2013	Outcomes are not relevant to the protocol - no relevant comparative data regarding exacerbations are reported.
Castile, R., High resolution CT scans of the chest should be a routine part of CF lung disease assessment: Pro, Pediatric Pulmonology, 45, 130-131, 2010	Opinion paper. Conference abstract.

Study	Reason for Exclusion
Chotirmall, S. H., Branagan, P., Gunaratnam, C., McElvaney, N. G., Aspergillus/allergic bronchopulmonary aspergillosis in an Irish cystic fibrosis population: a diagnostically challenging entity, Respiratory Care, 53, 1035-41, 2008	Aim and outcomes not relevant to the protocol. This study aims to determine the rate of aspergillys colonization and ABPA in a population of patients with CF. No outcomes of interest are reported.
Cleveland, R. H., Stamoulis, C., Sawicki, G., Kelliher, E., Zucker, E. J., Wood, C., Zurakowski, D., Lee, E., Brasfield and Wisconsin scoring systems have equal value as outcome assessment tools of cystic fibrosis lung disease, Pediatric Radiology, 44, 529-34, 2014	Aim not related to the protocol (to assess the reproducibility and reliability of the Brafiel and Wisconsin chest radiograph systems).
Clifton, I. J., Kastelik, J. A., Peckham, D. G., Hale, A., Denton, M., Etherington, C., Conway, S. P., Ten years of viral and non-bacterial serology in adults with cystic fibrosis, Epidemiology & Infection, 136, 128-34, 2008	Aim of the study not related to the protocol.This paper investigates the frequency and impact of viral infections in adults with CF receiving intravenous antibiotics. It does not provide direct comparisons between test or monitoring strategies.
Conway, S. P., Pond, M. N., Watson, A., Etherington, C., Robey, H. L., Goldman, M. H., Intravenous colistin sulphomethate in acute respiratory exacerbations in adult patients with cystic fibrosis, Thorax, 52, 987-93, 1997	Aim not related to the protocol (to assess the safety and efficacy of intravenous colistin in the treatment of acute respiratory exacerbations in adult patients with CF).
Cortese, G., Malfitana, V., Placido, R., Ferrari, A., Grosso, B., De Rose, V., Nespoli, P., Fava, C., Role of chest radiography in the diagnosis of allergic bronchopulmonary aspergillosis in adult patients with cystic fibrosis, Radiologia Medica, 112, 626-36, 2007	Aim not related to the protocol (to verify the usefulness of chest radiography in the diagnosis of allergic bronchopulmonary aspergillosis (ABPA) in adult patients with cystic fibrosis).
Cunningham, S., McColm, J. R., Ho, L. P., Greening, A. P., Marshall, T. G., Measurement of inflammatory markers in the breath condensate of children with cystic fibrosis, European Respiratory Journal, 15, 955-7, 2000	Aim not relevant to the protocol (to assess the value of inflammatory mediators in breath condensate, as a non-invasive, direct assessment of pulmonary inflammation).
Da Silva Filho, L. V. R. F., Ferreira, F. A., Reis, F. J. C., De Britto, M. C. A., Levy, C. E., Clark, O., Ribeiro, J. D., Pseudomonas aeruginosa infection in patients with cystic fibrosis: Scientific evidence regarding clinical impact, diagnosis, and treatment, Jornal Brasileiro de Pneumologia, 39, 495-512, 2013	Systematic review. The research question is not relevant to the protocol.
Dakin, C. J., Pereira, J. K., Henry, R. L., Wang, H., Morton, J. R., Relationship between sputum inflammatory markers, lung function, and lung pathology on high-resolution computed tomography in children with cystic fibrosis, Pediatric Pulmonology, 33, 475-82, 2002	Aim not related to the protocol: to study the relationship between PFTs, sputum cytology, and sputum cytokine interleukin-8 (IL-8) and HRCT in CF patients. Only correlation data is provided.
Dasenbrook, E. C., Merlo, C. A., Diener-West, M., Lechtzin, N., Boyle, M. P., Persistent methicillin-resistant Staphylococcus aureus and rate of FEV1 decline in cystic fibrosis, American Journal of Respiratory & Critical Care Medicine, 178, 814-21, 2008	Aim not related to the protocol (to determine the association between MRSA respiratory infection and FEV(1) decline in children and adults with CF).
Davis, S. D., Fordham, L. A., Brody, A. S., Noah, T. L., Retsch-Bogart, G. Z., Qaqish, B. F., Yankaskas, B. C., Johnson, R. C., Leigh, M. W., Computed tomography reflects lower airway inflammation and tracks changes in early cystic	Intervention/ comparison: the study does not provide a direct comparison between CT and bronchoscopy.

Study	Reason for Exclusion
fibrosis, American Journal of Respiratory and Critical Care Medicine, 175, 943-950, 2007	
Davis,S.D., High resolution CT scans of the chest should be a routine part of CF lung disease assessment: Con, Pediatric Pulmonology, 45, 128-129, 2010	Narrative opinion, review. Conference abstract.
de Almeida, M. B., Zerbinati, R. M., Tateno, A. F., Oliveira, C. M., Romao, R. M., Rodrigues, J. C., Pannuti, C. S., da Silva Filho, L. V., Rhinovirus C and respiratory exacerbations in children with cystic fibrosis, Emerging Infectious Diseases, 16, 996-9, 2010	Intervention/ comparison: the paper does not compare monitoring strategies.
De Baets, F., Schelstraete, P., Van Daele, S., Haerynck, F., Vaneechoutte, M., Achromobacter xylosoxidans in cystic fibrosis: prevalence and clinical relevance, Journal of Cystic Fibrosis, 6, 75-8, 2007	Prevalence of Achromobacter xylosoxidans in people with CF.
de Jong, P. A., Lindblad, A., Rubin, L., Hop, W. C., de Jongste, J. C., Brink, M., Tiddens, H. A., Progression of lung disease on computed tomography and pulmonary function tests in children and adults with cystic fibrosis, Thorax, 61, 80-5, 2006	Outcomes: This study compares the ability of CT scores and PFTs to detect changes in lung disease in children and adults with CF, but does not provide outcomes of interest.
de Jong, P. A., Nakano, Y., Lequin, M. H., Mayo, J. R., Woods, R., Pare, P. D., Tiddens, H. A., Progressive damage on high resolution computed tomography despite stable lung function in cystic fibrosis, European Respiratory Journal, 23, 93-7, 2004	Aim not related to the protocol. The aim of this study was to investigate the ability of HRCT scoring systems and PFT to detect changes in lung disease, but only provides data on correlations.
de Jong, P. A., Ottink, M. D., Robben, S. G., Lequin, M. H., Hop, W. C., Hendriks, J. J., Pare, P. D., Tiddens, H. A., Pulmonary disease assessment in cystic fibrosis: comparison of CT scoring systems and value of bronchial and arterial dimension measurements, Radiology, 231, 434-9, 2004	Aim not related to the protocol. The study aims to compare thin-section CT scores obtained with five scoring systems for assessment of pulmonary disease in children with CF and to determine additional value of bronchial and arterial dimension measurements. Results are reported as correlations. No outcomes of interest are included.
Demirkazik, F. B., Ariyurek, O. M., Ozcelik, U., Gocmen, A., Hassanabad, H. K., Kiper, N., High resolution CT in children with cystic fibrosis: correlation with pulmonary functions and radiographic scores, European Journal of Radiology, 37, 54-9, 2001	Outcomes are not relevant to the protocol. Results are reported as correlations.
Demko, C. A., Stern, R. C., Doershuk, C. F., Stenotrophomonas maltophilia in cystic fibrosis: incidence and prevalence, Pediatric Pulmonology, 25, 304-8, 1998	Aim of the study not relevant to the protocol. The study does not compare two monitoring strategies, but the prognostic significance of S. Maltophilia.
Deschaght, P., Schelstraete, P., Van Simaey, L., Vanderkercken, M., Raman, A., Mahieu, L., Van Daele, S., De Baets, F., Vaneechoutte, M., Is the improvement of CF patients, hospitalized for pulmonary exacerbation, correlated to a decrease in bacterial load?, PLoS ONE [Electronic Resource], 8, e79010, 2013	Aim is not relevant to the protocol. The study correlates bacterial load and patient improvement following an exacerbation.
Dodd, J. D., Barry, S. C., Gallagher, C. G., Respiratory factors do not limit maximal symptom-limited exercise in patients with mild	Aim of the study not related to the protocol.

Study	Reason for Exclusion
cystic fibrosis lung disease, Respiratory Physiology & Neurobiology, 152, 176-85, 2006	
Dorlochter, L., Carlsson, M., Olafsdottir, E. J., Roksund, O. D., Rosendahl, K., Fluge, G., Anti-neutrophil cytoplasmatic antibodies and lung disease in cystic fibrosis, Journal of Cystic Fibrosis, 3, 179-83, 2004	Does not compare two monitoring strategies. It only provides correlation data between BPI-ANCA and lung damage.
Dorlochter, L., Roksund, O., Helgheim, V., Rosendahl, K., Fluge, G., Resting energy expenditure and lung disease in cystic fibrosis, Journal of Cystic Fibrosis, 1, 131-6, 2002	Aim not related to the protocol (to assess the impact of lung parenchyma damage on resting energy expenditure and correlated these findings with forced expiratory volume in 1 s (FEV(1)).
Ellemunter, H., Fuchs, S. I., Unsinn, K. M., Freund, M. C., Waltner-Romen, M., Steinkamp, G., Gappa, M., Sensitivity of Lung Clearance Index and chest computed tomography in early CF lung disease, Respiratory Medicine, 104, 1834-42, 2010	Aim not related to the protocol. This study aims to prospectively investigate the diagnostic accuracy of the LCI in comparison to CT in CF patients with early lung disease and normal FEV(1) (>80% pred.). It only reports on diagnostic accuracy outcomes.
Equi, A. C., Pike, S. E., Davies, J., Bush, A., Use of cough swabs in a cystic fibrosis clinic, Archives of Disease in Childhood, 85, 438-9, 2001	The aim and outcomes of this study are not related to the protocol. This study aims to determine the diagnostic accuracy of cough swabs with spontaneously expectorated sputum samples. Only diagnostic accuracy data is reported.
Falk, B., Nini, A., Zigel, L., Yahav, Y., Aviram, M., Rivlin, J., Bentur, L., Avital, A., Dotan, R., Blau, H., Effect of low altitude at the Dead Sea on exercise capacity and cardiopulmonary response to exercise in cystic fibrosis patients with moderate to severe lung disease, Pediatric Pulmonology, 41, 234-41, 2006	Aim of the study not related to the protocol. The study aims to determine the effect of descent to the Dead Sea on exercise capacity in a group of young, moderately to severely obstructed, non-oxygen dependent CF patients.
Falk, C., Pressler, T., Christiansen, C., Katzenstein, T. L., Virological finding in bronchoalveolar lavage (BAL) among non-transplanted cystic fibrosis patients with clinical exacerbation, Pediatric Pulmonology, 48, 348, 2013	Conference abstract. Study design: not a RCT.
Farrell, P. M., Li, Z., Kosorok, M. R., Laxova, A., Green, C. G., Collins, J., Lai, H. C., Rock, M. J., Splaingard, M. L., Bronchopulmonary disease in children with cystic fibrosis after early or delayed diagnosis, American Journal of Respiratory & Critical Care Medicine, 168, 1100-8, 2003	Aim not related to the protocol (it assess the potential benefits of early diagnosis of CF through neonatal screening in pulmonary prognosis).
Farrell, P. M., Shen, G., Splaingard, M., Colby, C. E., Laxova, A., Kosorok, M. R., Rock, M. J., Mischler, E. H., Acquisition of Pseudomonas aeruginosa in children with cystic fibrosis, Pediatrics, 100, E2, 1997	Aim not related to the protocol (to determine whether patients diagnosed through neonatal screening and treated in early infancy were more likely to become colonized with Pseudomonas aeruginosa compared with those identified by standard diagnostic methods).
Farrell,P.M., Collins,J., Broderick,L.S., Rock,M.J., Li,Z., Kosorok,M.R., Laxova,A., Gershan,W.M., Brody,A.S., Association between mucoid Pseudomonas infection and bronchiectasis in children with cystic fibrosis, Radiology, 252, 534-543, 2009	Aim and outcomes not relevant to the protocol. This study aimed to correlate the severity of bronchiectasis in children with CF with clinical and microbiologic variables in order to clarify risk factors for the development of irreversible lung disease. Only correlation data is provided.
Farrell,P.M., Li,Z., Kosorok,M.R., Laxova,A., Green,C.G., Collins,J., Lai,H.C., Makholm,L.M.,	Aim and outcomes not relevant to the protocol. This study compares quantitative chest

Study	Reason for Exclusion
Rock,M.J., Splaingard,M.L., Longitudinal evaluation of bronchopulmonary disease in children with cystic fibrosis, Pediatric Pulmonology, 36, 230-240, 2003	radiology with spirometry data in the context of respiratory symptoms. Only correlation data is provided. No prognostic outcomes of interest are presented.
Fauroux, B., Delaisi, B., Clement, A., Saizou, C., Moissenet, D., Truffot-Pernot, C., Tournier, G., Vu Thien, H., Mycobacterial lung disease in cystic fibrosis: a prospective study, Pediatric Infectious Disease Journal, 16, 354-8, 1997	Aim of the study is not related to the protocol. The aim of the study was to determine the incidence and clinical picture of mycobacterial infection in CF children.
Fermeiro, J., Reis, P., Castanhinha, S., Pereira, L., Barreto, C., The impact of methicillin-resistant Staphylococcus aureus colonisation on paediatric cystic fibrosis patients' morbidity, Revista Portuguesa de Pneumologia, 16, 527-42, 2010	Intervention: does not compare different strategies.
Finkelstein, S. M., Wielinski, C. L., Kujawa, S. J., Loewenson, R., Warwick, W. J., The impact of home monitoring and daily diary recording on patient status in cystic fibrosis, Pediatric Pulmonology, 12, 3-10, 1992	Aim and monitoring strategy not related to the protocol. This is an implementation evaluation study for an experimental home monitoring system for assessing the progress and planning changes in the care of patients with CF.
Fischer, N., Hentschel, J., Markert, U. R., Keller, P. M., Pletz, M. W., Mainz, J. G., Non-invasive assessment of upper and lower airway infection and inflammation in CF patients, Pediatric Pulmonology, 49, 1065-75, 2014	No outcomes of interest. The study does not report prognostic outcomes, only associations between upper and lower airway samples.
Flight, W. G., Bright-Thomas, R. J., Tilston, P., Mutton, K. J., Guiver, M., Morris, J., Webb, A. K., Jones, A. M., Incidence and clinical impact of respiratory viruses in adults with cystic fibrosis, Thorax, 69, 247-53, 2014	Aim not related to the protocol (to determine the incidence and clinical impact of VRI among adults with CF).
Fontoura, M. S. H., Arajo-Neto, C. A., Andrade, S. C. S., Brim, R. V., Matutino, A. R., Silva, C. C., Santana, M. C., Nobre-Bastos, M., Oliveira, F., Barreto, B. B., Santos, P. M., Noblat, L., Cardoso, M. R. A., Nascimento-Carvalho, C. M., Clinical failure among children with nonsevere community-acquired pneumonia treated with amoxicillin, Expert Opinion on Pharmacotherapy, 11, 1451-1458, 2010	Population: non-CF patients
Fowler, S. J., French, J., Screaton, N. J., Foweraker, J., Condliffe, A., Haworth, C. S., Exley, A. R., Bilton, D., Nontuberculous mycobacteria in bronchiectasis: Prevalence and patient characteristics, European Respiratory Journal, 28, 1204-1210, 2006	Population: non-CF patients (patients attending a tertiary referral bronchiectasis clinic).
Frangolias, D. D., Holloway, C. L., Vedal, S., Wilcox, P. G., Role of exercise and lung function in predicting work status in cystic fibrosis, American journal of respiratory and critical care medicine, 167, 150-157, 2003	Aim not related to the protocol. To determine whether quantification of aerobic fitness improved predictability of disease-related disability. No outcomes of interest are reported.
Fuchs, S. I., Gappa, M., Eder, J., Unsinn, K. M., Steinkamp, G., Ellemunter, H., Tracking Lung Clearance Index and chest CT in mild cystic fibrosis lung disease over a period of three years, Respiratory Medicine, 108, 865-74, 2014	Aim and outcomes. This study aims to investigate the longitudinal course of the LCI in comparison to spirometry and CT and to examine whether LCI results sampled in 2007 are of prognostic value for structural lung abnormalities diagnosed from CT in 2010. The

Study	Reason for Exclusion
	study only reports correlations, and it does not include any outcome of interest.
Galici, V., Cocchi, P., Colombo, C., Cariani, L., Lucidi, V., Fiscarelli, E., Raia, V., Terlizzi, V., Taccetti, G., Early antibiotic treatment for MRSA eradication in cystic fibrosis patients: A randomized multicenter study, Pediatric Pulmonology, 49, 338-9, 2014	Conference abstract of RCT of treatments (not monitoring strategies)
Gangell, C. G., Hall, G. L., Balding, E., Berry, L., Carlin, J., Carzino, R., de Klerk, N., Douglas, T., Ebdon, A., Foo, C., Gangell, C. L., Garratt, L. W., Gibson, A., Harrison, J., Kicic, A., Laing, I., Logie, K., Massie, J., Mott, L., Murray, C., Poreddy, S., Ranganathan, S. C., Robertson, C. F., Robins-Browne, R., Robinson, P., Skoric, B., Sly, P. D., Stick, S. M., Sutanto, E., Early detection of lung function abnormalities in young children with cystic fibrosis, Annals of Respiratory Medicine, 2, 22-27, 2012	Full text unavailable
Gibson, R. L., Emerson, J., McNamara, S., Burns, J. L., Rosenfeld, M., Yunker, A., Hamblett, N., Accurso, F., Dovey, M., Hiatt, P., Konstan, M. W., Moss, R., Retsch-Bogart, G., Wagener, J., Waltz, D., Wilmott, R., Zeitlin, P. L., Ramsey, B., Cystic Fibrosis Therapeutics Development Network Study, Group, Significant microbiological effect of inhaled tobramycin in young children with cystic fibrosis, American Journal of Respiratory & Critical Care Medicine, 167, 841-9, 2003	Aim not related to the protocol. This is an intervention study (RCT) aimed at testing the hypothesis that the administration of TSI to children with CF would be safe and result in a profound decrease in P. aeruginosa (Pa) density from the lower airway of young children with CF. The study does not provide direct comparisons of monitoring strategies.
Grosse-Onnebrink, J., Stehling, F., Olivier, M., Mellies, U., The lung clearance index correlates with markers of pulmonary deterioration in patients with cystic fibrosis, European Respiratory Journal, 42, 2013	Full text unavailable
Gustafsson, P. M., De Jong, P. A., Tiddens, H. A., Lindblad, A., Multiple-breath inert gas washout and spirometry versus structural lung disease in cystic fibrosis, Thorax, 63, 129-34, 2008	Aim not related to the protocol: to determine the agreements between LCI and spirometry, respectively, with structural lung disease as measured by HRCT in children and teenagers with CF. Only results on diagnostic accuracy and correlations between the tests are provided.
Hatziagorou, E., Avramidou, V., Kontouli, K., Georgopoulou, V., Tsanakas, J., Cystic fibrosis lung disease assessed by lung clearance index, European Respiratory Journal, 42, 2013	Conference abstract. Study design: not a RCT.
Hogan, D. A., Willger, S. D., Dolben, E. L., Hampton, T. H., Stanton, B. A., Morrison, H. G., Sogin, M. L., Czum, J., Ashare, A., Analysis of Lung Microbiota in Bronchoalveolar Lavage, Protected Brush and Sputum Samples from Subjects with Mild-To-Moderate Cystic Fibrosis Lung Disease, PLoS ONE [Electronic Resource]PLoS ONE, 11, e0149998, 2016	No outcomes of interest. The study does not report prognostic outcomes. N < 10.
Hoppe, J. E., Towler, E., Wagner, B. D., Accurso, F. J., Sagel, S. D., Zemanick, E. T., Sputum induction improves detection of	Comparison not relevant to the protocol. No prognostic outcomes reported.

Study	Reason for Exclusion
pathogens in children with cystic fibrosis, Pediatric Pulmonology, 50, 638-46, 2015	
Jain, K., Wainwright, C., Smyth, A. R., Bronchoscopy-guided antimicrobial therapy for cystic fibrosis, Cochrane Database of Systematic ReviewsCochrane Database Syst Rev, 1, CD009530, 2016	This is a Cochrane review. It includes only 1 trial, which has already been included in this review.
Jobsis, Q., Raatgeep, H. C., Schellekens, S. L., Kroesbergen, A., Hop, W. C., de Jongste, J. C., Hydrogen peroxide and nitric oxide in exhaled air of children with cystic fibrosis during antibiotic treatment, European Respiratory Journal, 16, 95-100, 2000	Aim and outcomes not relevant to the protocol. This study was to assess whether measurement of hydrogen peroxide (H2O2) and nitric oxide (NO) in exhaled air can serve to monitor the effect of treatment with antibiotics in CF-children with acute infective pulmonary exacerbations. Different monitoring strategies are not compared. Not outcomes of interest are reported.
Johnson, C., Butler, S. M., Konstan, M. W., Morgan, W., Wohl, M. E., Factors influencing outcomes in cystic fibrosis: a center-based analysis, Chest, 123, 20-7, 2003	This study aims to determine whether differences in lung health existed between groups of patients attending different CF care sites and to determine whether these differences are associated with differences in monitoring and intervention. Frequent monitoring received more antibiotics, increased use of appropriate medications. Different monitoring strategies are not compared as such (rather the frequency). The study does not report on outcomes of interest.
Judge, E. P., Dodd, J. D., Masterson, J. B., Gallagher, C. G., Pulmonary abnormalities on high-resolution CT demonstrated more rapid decline than FEV1 in adults with cystic fibrosis, Chest, 130, 1424-1432, 2006	Outcomes are not relevant to the protocol. Results from using different diagnostic techniques are reported but are not compared.
Kraemer, R., Delosea, N., Ballinari, P., Gallati, S., Crameri, R., Effect of allergic bronchopulmonary aspergillosis on lung function in children with cystic fibrosis, American Journal of Respiratory & Critical Care Medicine, 174, 1211-20, 2006	Aim not related to the protocol. This study aims to evaluate onset of A. fumigatus sensitization and development of allergic bronchopulmonary aspergillosis (ABPA), as well as to determine the physiologic factors of lung function influencing these mechanisms in CF.
Lam, J. C., Somayaji, R., Surette, M. G., Rabin, H. R., Parkins, M. D., Reduction in Pseudomonas aeruginosa sputum density during a cystic fibrosis pulmonary exacerbation does not predict clinical response, BMC Infectious Diseases, 15, 145, 2015	Strategy not relevant to the protocol (protocol 3). This study evaluates whether sputum density is an adequate monitoring strategy to predict clinical response in people treated for an exacerbation with P. Aeruginosa.
Laufer, P., Fink, J. N., Bruns, W. T., Unger, G. F., Kalbfleisch, J. H., Greenberger, P. A., Patterson, R., Allergic bronchopulmonary aspergillosis in cystic fibrosis, Journal of Allergy & Clinical Immunology, 73, 44-8, 1984	Aim is not relevant to the protocol. The study examines allergy biomarkers in response to aspergillus fumigatus.
Linnane, B., Vaish, S., Clarke, D., O'Sullivan, N., McNally, P., The findings of a clinical surveillance bronchoalveolar lavage programme in pre-school patients with cystic fibrosis, Pediatric Pulmonology, 50, 327-332, 2015	Aim and outcomes not related to the protocol. This study was carried out to determine the prevalence of pathogens in lower airway samples detected as part of a pilot clinical BAL surveillance programme, in young children a(1-6 years old), and to ascertain if their detection resulted in a change in treatment. However no

Study	Reason for Exclusion
	clinical outcomes are reported. No different surveillance programmes are compared.
Lynch,D.A., Brasch,R.C., Hardy,K.A., Webb,W.R., Pediatric pulmonary disease: assessment with high-resolution ultrafast CT, Radiology, 176, 243-248, 1990	The study includes children with different health conditions, and only n=5 had CF. Although chest X-ray and CT-scan were performed, they are not compared. The study does not report on outcomes of interest either.
Marchant, J. M., Masel, J. P., Dickinson, F. L., Masters, I. B., Chang, A. B., Application of chest high-resolution computer tomography in young children with cystic fibrosis, Pediatric Pulmonology, 31, 24-9, 2001	Aim of the study not relevant to the protocol. This study aims to relate the two available scoring systems to pulmonary function data and to each other and to describe early HRCT changes in younger children. No outcomes of interest are reported.
McEvoy, S., Lavelle, L., Kilcoyne, A., McCarthy, C., deJong, P. A., Loeve, M., Tiddens, H. A., McKone, E., Gallagher, C. G., Dodd, J. D., High-resolution CT of nontuberculous mycobacterium infection in adult CF patients: diagnostic accuracy, European Radiology, 22, 2736-42, 2012	Aim not related to the protocol (to determine the diagnostic accuracy of high-resolution computed tomography for the detection of non-tuberculous mycobacterium infection in adult with CF).
McMahon, C. J., Dodd, J. D., Hill, C., Woodhouse, N., Wild, J. M., Fichele, S., Gallagher, C. G., Skehan, S. J., van Beek, E. J., Masterson, J. B., Hyperpolarized 3helium magnetic resonance ventilation imaging of the lung in cystic fibrosis: comparison with high resolution CT and spirometry, European Radiology, 16, 2483-90, 2006	Outcomes: this paper compares MRI vs HRCT scan spirometry, but it does not report on outcomes of interest, as it only does correlations between both strategies.
McMahon, M. A., Chotirmall, S. H., McCullagh, B., Branagan, P., McElvaney, N. G., Logan, P. M., Radiological abnormalities associated with Aspergillus colonization in a cystic fibrosis population, European Journal of Radiology, 81, e197-e202, 2012	Aim not related to the protocol: To determine if sputum colonization with Aspergillus species in patients with cystic fibrosis (PWCF) correlates with radiological abnormalities and/or a reduction in pulmonary function (FEV1)
Miall, L. S., McGinley, N. T., Brownlee, K. G., Conway, S. P., Methicillin resistant Staphylococcus aureus (MRSA) infection in cystic fibrosis, Archives of Disease in Childhood, 84, 160-2, 2001	Aim not related to the protocol (To determine whether MRSA infection has a deleterious effect on the clinical status of children with CF).
Montella,S., Santamaria,F., Salvatore,M., Pignata,C., Maglione,M., Iacotucci,P., Mollica,C., Assessment of chest high-field magnetic resonance imaging in children and young adults with noncystic fibrosis chronic lung disease: comparison to high-resolution computed tomography and correlation with pulmonary function, Investigative Radiology, 44, 532-538, 2009	Adults with non-CF lung disease.
Morgan, W. J., VanDevanter, D. R., Pasta, D. J., Foreman, A. J., Wagener, J. S., Konstan, M. W., Scientific Advisory, Group, Investigators,, Coordinators of Epidemiologic Study of Cystic, Fibrosis, Forced Expiratory Volume in 1 Second Variability Helps Identify Patients with Cystic Fibrosis at Risk of Greater Loss of Lung Function, Journal of PediatricsJ Pediatr, 169, 116-21.e2, 2016	Insufficient data presented to perform analysis according to the protocol.

Study	Reason for Exclusion
Mroueh, S., Spock, A., Allergic bronchopulmonary aspergillosis in patients with cystic fibrosis, Chest, 105, 32-6, 1994	Intervention/ comparison: the study does not include any monitoring strategy comparisons.
Nasr, S. Z., Gordon, D., Sakmar, E., Yu, X., Christodoulou, E., Eckhardt, B. P., Strouse, P. J., High resolution computerized tomography of the chest and pulmonary function testing in evaluating the effect of tobramycin solution for inhalation in cystic fibrosis patients, Pediatric Pulmonology, 41, 1129-37, 2006	This study evaluates the effect of tobramycin solution for inhalation in CF patients. Although they use HRCT scan and spirometry, authors just compare treatment and placebo groups. No outcomes of interest are reported.
Nasr, S. Z., Sakmar, E., Christodoulou, E., Eckhardt, B. P., Streetman, D. S., Strouse, P. J., The use of high resolution computerized tomography (HRCT) of the chest in evaluating the effect of tobramycin solution for inhalation in cystic fibrosis lung disease, Pediatric Pulmonology, 45, 440-9, 2010	Intervention/ comparison: does not compare different monitoring strategies. Evaluates the effectiveness of Tobramycin vs placebo, and then applies same monitoring strategy to all participants.
Oikonomou, A., Manavis, J., Karagianni, P., Tsanakas, J., Wells, A. U., Hansell, D. M., Papadopoulou, F., Efremidis, S. C., Loss of FEV1 in cystic fibrosis: correlation with HRCT features, European Radiology, 12, 2229-35, 2002	Aim not related to the protocol (to determine which high-resolution computed tomography features in patients with CF are most strongly associated with functional impairment as expressed by forced FEV1).
Oikonomou, A., Tsanakas, J., Hatziagorou, E., Kirvassilis, F., Efremidis, S., Prassopoulos, P., High resolution computed tomography of the chest in cystic fibrosis (CF): is simplification of scoring systems feasible?, European Radiology, 18, 538-47, 2008	Aim of the study not relevant to the protocol. This study aims to simplify HRCT scan scoring systems for an easy ad efficient application in clinical practice. Only correlation data is provide.
Olivier, K. N., Weber, D. J., Lee, J. H., Handler, A., Tudor, G., Molina, P. L., Tomashefski, J., Knowles, M. R., Nontuberculous Mycobacteria in Cystic Fibrosis Study, Group, Nontuberculous mycobacteria. II: nested-cohort study of impact on cystic fibrosis lung disease, American Journal of Respiratory & Critical Care Medicine, 167, 835-40, 2003	Outcomes. This study was designed to look systematically at the longitudinal effects of nontuberculous mycobacteria on the clinical course of lung disease, as assessed by changes in spirometry and CT scan. It does not provide any comparison statistics between both tests. It does not report on outcomes of interest.
Owens, C. M., Aurora, P., Stanojevic, S., Bush, A., Wade, A., Oliver, C., Calder, A., Price, J., Carr, S. B., Shankar, A., Stocks, J., London Cystic Fibrosis, Collaboration, Lung Clearance Index and HRCT are complementary markers of lung abnormalities in young children with CF, Thorax, 66, 481-8, 2011	Aim not related to the protocol. The objectives of this study were: (1) To assess whether the lung clearance index (LCI) derived from multiple breath inert-gas washout (MBW) is as effective as HRCT in identifying pulmonary abnormalities; and (2) explore the relationships between abnormalities detected by HRCT and by spirometry, plethysmography and MBW (collectively, LFTs) in young children with CF. Only correlation data is provided.
Robinson, T. E., Goris, M. L., Zhu, H. J., Chen, X., Bhise, P., Sheikh, F., Moss, R. B., Dornase alfa reduces air trapping in children with mild cystic fibrosis lung disease: a quantitative analysis, Chest, 128, 2327-35, 2005	Aim of the study not related to the protocol. This is a RCT that aims to evaluate the therapeutic response to dornase alpha in children with mild CF lung disease. Comparisons between different monitoring strategies are not provided.
Robinson, T. E., Leung, A. N., Chen, X., Moss, R. B., Emond, M. J., Cystic fibrosis HRCT scores correlate strongly with Pseudomonas infection, Pediatric Pulmonology, 44, 1107-17, 2009	Aim and outcomes not relevant to the protocol. This study aimed to compare the association between HRCT scores and P. Aeruginosa status. No outcomes of interest are reported.

Study	Reason for Exclusion
Robinson, T. E., Leung, A. N., Northway, W. H., Blankenberg, F. G., Chan, F. P., Bloch, D. A., Holmes, T. H., Moss, R. B., Composite spirometric-computed tomography outcome measure in early cystic fibrosis lung disease, American Journal of Respiratory & Critical Care Medicine, 168, 588-93, 2003	Aim is not related to the protocol. This study evaluates changes in test scores over one year as part of a RCT of dornase alpha.
Robroeks, C. M., Roozeboom, M. H., de Jong, P. A., Tiddens, H. A., Jobsis, Q., Hendriks, H. J., Yntema, J. B., Brackel, H. L., van Gent, R., Robben, S., Dompeling, E., Structural lung changes, lung function, and non-invasive inflammatory markers in cystic fibrosis, Pediatric Allergy & Immunology, 21, 493-500, 2010	Aim not related to the protocol. The objective of this study was to investigate the relationship between non-invasive inflammatory markers (IM) in exhaled breath condensate (EBC), lung function indices and structural lung changes, visualized by HRCT scans in CF. Only correlation data is provided.
Rosenfeld, M., Gibson, R. L., McNamara, S., Emerson, J., Burns, J. L., Castile, R., Hiatt, P., McCoy, K., Wilson, C. B., Inglis, A., Smith, A., Martin, T. R., Ramsey, B. W., Early pulmonary infection, inflammation, and clinical outcomes in infants with cystic fibrosis, Pediatric Pulmonology, 32, 356-366, 2001	Intervention/ comparison: does not compare techniques.
Rowan, S., Bradley, J., Ennis, M., Horsley, A., Bell, N., Gustafsson, P., Elborn, J. S., Lung clearance index is a reproducible and sensitive measure of airways disease in bronchiectasis, Thorax, 66, A121, 2011	Population includes patients with bronchiectasis (BE) not caused by CF. Conference abstract, not a RCT.
Sagel, S. D., Wagner, B. D., Anthony, M. M., Emmett, P., Zemanick, E. T., Sputum biomarkers of inflammation and lung function decline in children with cystic fibrosis, American Journal of Respiratory & Critical Care Medicine, 186, 857-65, 2012	Intervention/ comparison: this paper examines longitudinal relationships between sputum biomarkers and lung function, but it does not compare different monitoring strategies.
Sanders, D. B., Li, Z., Brody, A. S., Farrell, P. M., Chest computed tomography scores of severity are associated with future lung disease progression in children with cystic fibrosis, American Journal of Respiratory & Critical Care Medicine, 184, 816-21, 2011	Outcomes are not relevant to the protocol - only correlation data is provided.
Sanders,D.B., Li,Z., Brody,A., Collins,J., Broderick,L., Farrell,P.M., Chest CT scores of severity predict future lung disease progression in children with CF, American Journal of Respiratory and Critical Care Medicine, 183, -, 2011	Conference abstract. Study design: not a RCT
Sanders,D.B., Li,Z., Brody,A., Farrell,P.M., Bronchiectasis on chest CT predicts future lung disease severity in children with CF, Pediatric Pulmonology, 46, 330-, 2011	Conference abstract. This is a RCT to evaluate the association of Bronchiectasis and future lung disease. The abstract does not contain enough information to assess it for relevance.
Santamaria, F., Grillo, G., Guidi, G., Rotondo, A., Raia, V., de Ritis, G., Sarnelli, P., Caterino, M., Greco, L., Cystic fibrosis: when should high-resolution computed tomography of the chest Be obtained?, Pediatrics, 101, 908-13, 1998	Aim and outcomes not relevant to the protocol. This study aims to provide indications for HRCT of the chest in patients with CF. Different monitoring strategies are not compared. Only correlation data is presented. No outcomes of interest are reported.
Sheikh, S. I., Long, F. R., Flucke, R., Ryan-Wenger, N. A., Hayes, D., Jr., McCoy, K. S., Changes in Pulmonary Function and Controlled	This study evaluates changes in pulmonary function following an exacerbation in infants, but it does not compare monitoring strategies.

Study	Reason for Exclusion
Ventilation-High Resolution CT of Chest After Antibiotic Therapy in Infants and Young Children with Cystic Fibrosis, Lung, 193, 421-8, 2015	
Sheikh, S. I., Long, F. R., Flucke, R., Ryan-Wenger, N. A., Hayes, D., McCoy, K. S., Changes in Pulmonary Function and Controlled Ventilation-High Resolution CT of Chest After Antibiotic Therapy in Infants and Young Children with Cystic Fibrosis, Lung, 193, 421-428, 2015	Intervention/ comparison: this study does not compare different monitoring strategies.
Sloane, A. J., Lindner, R. A., Prasad, S. S., Sebastian, L. T., Pedersen, S. K., Robinson, M., Bye, P. T., Nielson, D. W., Harry, J. L., Proteomic analysis of sputum from adults and children with cystic fibrosis and from control subjects, American Journal of Respiratory & Critical Care Medicine, 172, 1416-26, 2005	Aim and outcomes not relevant to the protocol. This study aims to identify biomarkers of lung exacerbation for point-of-care monitoring of CF lung disease progression. It does not compare different monitoring strategies. No outcomes of interest are reported.
Sly, P. D., Brennan, S., Gangell, C., de Klerk, N., Murray, C., Mott, L., Stick, S. M., Robinson, P. J., Robertson, C. F., Ranganathan, S. C., Australian Respiratory Early Surveillance Team for Cystic, Fibrosis, Lung disease at diagnosis in infants with cystic fibrosis detected by newborn screening, American Journal of Respiratory & Critical Care Medicine, 180, 146-52, 2009	Descriptive report of surveillance of children with CF using different diagnostic techniques but not comparing these.
Taccone, A., Romano, L., Marzoli, A., Girosi, D., Dell'Acqua, A., Romano, C., High-resolution computed tomography in cystic fibrosis, European Journal of Radiology, 15, 125-129, 1992	Evaluates the sensitivity of high-resolution computed tomography (HRCT) in identifying the pulmonary lesions of CF, but the comparator is not clear. Not outcomes of interest reported.
Taylor, L., Corey, M., Matlow, A., Sweezey, N. B., Ratjen, F., Comparison of throat swabs and nasopharyngeal suction specimens in non-sputum-producing patients with cystic fibrosis, Pediatric Pulmonology, 41, 839-43, 2006	Outcomes: this study does not report outcomes of interest
Tepper, L. A., Ciet, P., Caudri, D., Quittner, A. L., Utens, E. M., Tiddens, H. A., Validating chest MRI to detect and monitor cystic fibrosis lung disease in a pediatric cohort, Pediatric Pulmonology, 51, 34-41, 2016	Aim not relevant to the protocol. This study aimed to validate chest MRI to detect and monitor CF lung disease in children, but only correlation data is provided.
Thia, L. P., Rand, S., Hill, L., Prasad, S. A., Bush, A., Balfour-Lynn, I. M., Pao, C., Ruiz, G., Stocks, J., Wallis, C., Suri, R., Significant bacterial infection missed using cough swabs compared to bronchoalveolar lavage in 1-year old newborn screened CF infants, Journal of Cystic Fibrosis. Conference: 36th European Cystic Fibrosis Conference Lisbon Portugal. Conference Start, 12	Conference abstract. Study design: not a RCT.
Veronez, L., Moreira, M. M., Soares, S. T. P., Pereira, M. C., Ribeiro, M. A. G. O., Ribeiro, J. D., Terzi, R. G. G., Martins, L. C., Paschoal, I. A., Volumetric capnography for the evaluation of pulmonary disease in adult patients with cystic fibrosis and noncystic fibrosis bronchiectasis, Lung, 188, 263-268, 2010	Aim of the study not relevant to the protocol (This study was designed to use volumetric capnography to evaluate the breathing pattern and ventilation inhomogeneities in patients with chronic sputum production and bronchiectasis and to correlate the phase 3 slope of the capnographic curve to spirometric measurements).

Study	Reason for Exclusion
Wielputz, M. O., Heusel, C. P., Herth, F. J., Kauczor, H. U., Radiological diagnosis in lung disease: factoring treatment options into the choice of diagnostic modality, Deutsches Arzteblatt International, 111, 181-7, 2014	Review paper on radiological diagnosis in lung disease. Not particularly focused on CF. It does not provide specific results regarding the monitoring strategy for people with CF.
Willekens, J., De Wachter, E., Ernst, C. W., Basten, I. A., Ilsen, B., Nieboer, K. H., Cornu, P., De Mey, J., Malfroot, A., Assessment of pulmonary disease by low dose ct and spirometry in CF patients with moderate disease: A comparison, Pediatric Pulmonology, 47, 2012	Conference abstract. This is a RCT that aims to compare the findings of spirometry to those of the low-dose CT. Only correlation data is provided. No outcomes of interest are reported in the abstract.
Wilson, C., MacDonald, J., Watter, P., O'Rourke, P., A retrospective audit of hospital records evaluating change in respiratory function during hospitalisation for pulmonary exacerbation in a paediatric population with cystic fibrosis, Physiotherapy, 92, 219-224, 2006	Aim and outcomes not relevant to the protocol. The study aims to determine the rate of change in lung function during hospitalisation in young people with CF using FEV1 and sputum expectorated at admission. The population is stratified by severity of FEV1 and sputum at admission relates to improved FEV1 and FVC during hospitalisation. No relevant outcomes are reported.
Wilson, P., Lambert, C., Carr, S. B., Pao, C., Paranasal sinus pathogens in children with cystic fibrosis: do they relate to lower respiratory tract pathogens and is eradication successful?, Journal of Cystic Fibrosis, 13, 449-54, 2014	Aim not related to the protocol (to assess the association of microflora between the paranasal sinus and the lower airways of children attending a regional paediatric cystic fibrosis centre and to determine the performance of an eradication treatment protocol for positive paranasal sinus samples).
Zhao, Z., Muller-Lisse, U., Frerichs, I., Fischer, R., Moller, K., Regional airway obstruction in cystic fibrosis determined by electrical impedance tomography in comparison with high resolution CT, Physiological Measurement, 34, N107-14, 2013	Full text unavailable

H.7 Airway clearance techniques

Study	Reason for Exclusion
Andrews, J., Sathe, N. A., Krishnaswami, S., McPheeters, M. L., Nonpharmacologic airway clearance techniques in hospitalized patients: a systematic review, Respiratory Care, 58, 2160-86, 2013	Cystic fibrosis not included.
App, E. M., Kieselmann, R., Reinhardt, D., Lindemann, H., Dasgupta, B., King, M., Brand, P., Sputum rheology changes in cystic fibrosis lung disease following two different types of physiotherapy: flutter vs autogenic drainage, Chest, 114, 171-7, 1998	The comparison was not prioritised for inclusion in the protocol (flutter vs AD).
Arens, R., Gozal, D., Omlin, K. J., Vega, J., Boyd, K. P., Keens, T. G., Woo, M. S., Comparison of high frequency chest compression and conventional chest physiotherapy in hospitalized patients with cystic fibrosis, American Journal of Respiratory & Critical Care Medicine, 150, 1154-7, 1994	In Cochrane SR Main (2005). The comparison (percussion postural drainage vs. high frequency oscillation) was not prioritised for inclusion in the protocol.

Study	Reason for Exclusion
Barnes, F., Rowland, M., Watt, P., The high frequency chest compression (HFCC) vest as a complementary intervention in persons with cystic fibrosis who find it difficult to engage in effective exercise or physiotherapy, Irish Journal of Medical Science, 1), S517, 2011	Abstract only.
Bauer, M. L., McDougal, J., Schoumacher, R. A., Comparison of manual and mechanical chest percussion in hospitalized patients with cystic fibrosis, Journal of Pediatrics, 124, 250-4, 1994	In Cochrane SR Main (2005). It compares manual vs mechanical chest percussion. This comparison was not included in the protocol.
Boyd, S., Brooks, D., Agnew-Coughlin, J., Ashwell, J., Evaluation of the literature on the effectiveness of physical therapy modalities in the management of children with cystic fibrosis, Pediatric Physical Therapy, 6, 70-74, 1994	Low quality systematic review (2/11 total score in AMSTAR checklist). Included studies were checked for eligibility.
Bradley, J. M., Moran, F. M., Elborn, J. S., Evidence for physical therapies (airway clearance and physical training) in cystic fibrosis: an overview of five Cochrane systematic reviews, Respiratory Medicine, 100, 191-201, 2006	Provides an overview of Cochrane reviews. References were checked for relevance.
Button, B. M., Heine, R. G., Catto-Smith, A. G., Olinsky, A., Phelan, P. D., Ditchfield, M. R., Story, I., Chest physiotherapy in infants with cystic fibrosis: to tip or not? A five-year study, Pediatric Pulmonology, 35, 208-13, 2003	In Cochrane SR Freitas (2015). The intervention (postural drainage) was not included in the protocol.
Button, B. M., Heine, R., Catto-Smith, A., Olinsky, A., Phelan, P. D., Story, I., A twelve month comparison of standard versus modified chest physiotherapy in twenty infants with cystic fibrosis [abstract], 11th Annual North American Cystic Fibrosis Conference. 299p., 1997	Abstract only.
Button, B. M., Wilson, C., Dentice, R., Cox, N. S., Middleton, A., Tannenbaum, E., Bishop, J., Cobb, R., Burton, K., Wood, M., Moran, F., Black, R., Bowen, S., Day, R., Depiazzi, J., Doiron, K., Doumit, M., Dwyer, T., Elliot, A., Fuller, L., Hall, K., Hutchins, M., Kerr, M., Lee, A. L., Mans, C., O'Connor, L., Steward, R., Potter, A., Rasekaba, T., Scoones, R., Tarrant, B., Ward, N., West, S., White, D., Wilson, L., Wood, J., Holland, A. E. The Thoracic Society of Australia, New Zealand Leaders in Lung, Health, Physiotherapy for cystic fibrosis in Australia and New Zealand, 2016	Clinical practice guideline.
Button, B., Herbert, R., Maher, C., Positive expiratory pressure therapy better maintains pulmonary function than postural drainage and percussion in patients with cystic fibrosis, Australian Journal of Physiotherapy, 44, 285-286, 1998	Abstract only.
Button,B.M., Heine,R.G., Catto-Smith,A.G., Phelan,P.D., Olinsky,A., Chest physiotherapy, gastro-oesophageal reflux, and arousal in infants with cystic fibrosis, Archives of Disease in Childhood, 89, 435-439, 2004	In Cochrane SR Freitas (2015). The intervention (postural drainage) was not included in the protocol.

Study	Reason for Exclusion
Cantin, A. M., Bacon, M., Berthiaume, Y., Mechanical airway clearance using the frequencer electro-acoustical transducer in cystic fibrosis, Clinical & Investigative Medicine - Medecine Clinique et Experimentale, 29, 159-65, 2006	Intervention not relevant to the protocol (frequencer).
Cappelletti, L. M., Cornacchia, M., Braggion, C., Zanolla, L., Mastella, G., Short-term effects of three chest physiotherapy regimens on patients with cystic fibrosis hospitalized for pulmonary exacerbation: a crossover randomized study, Excerpta Medica International Congress Series, 1034, 239-46, 1993	Intervention not relevant to the protocol (postural drainage).
Cardoso, R. M. T., Viana, R. A., Physiotherapy intervention in Cystic Fibrosis: A systematic review, Arquivos de Medicina, 25, 186-195, 2011	Not in English.
Carr, J., Pryor, J. A., Hodson, M. E., Self chest clapping: patients' views and the effects on oxygen saturation, Physiotherapy, 81, 753-757, 1995	Not RCT.
Centre for Reviews and Dissemination, Chest physical therapy management of patients with cystic fibrosis: a meta-analysis (Structured abstract), Database of Abstracts of Reviews of Effects, 2015	Abstract only. Full paper assessed.
Cerny, F. J., Relative effects of bronchial drainage and exercise for in-hospital care of patients with cystic fibrosis, Physical Therapy, 69, 633-9, 1989	In Cochrane SR Main (2005). Comparisons are bronchial drainage and exercise. Study not included as exercise is covered in another evidence review.
Davies, G. A., Banks, A. E., Agent, P., Osman, L. P., Bilton, D., Hodson, M. E., The use of high frequency chest wall oscillation during an acute infective pulmonary exacerbation of cystic fibrosis, Pediatric Pulmonology, 47, 366, 2012	Abstract only.
Dentice, R. L., Elkins, M. R., Bye, P. T., Adults with cystic fibrosis prefer hypertonic saline before or during airway clearance techniques: a randomised crossover trial, Journal of Physiotherapy, 58, 33-40, 2012	Intervention not included in protocol (hypertonic saline).
Dentice, R., Elkins, M. R., Bye, P. T., A randomised trial of the effect of timing of hypertonic saline inhalation in relation to airway clearance physiotherapy in adults with cystic fibrosis, Pediatric Pulmonology, 45, 384, 2010	Abstract only.
Dunn, C. E., Davies, Z., Yacob, A. A., Everson, C. L., Milla, C. E., A comparison of the acute effects of two hand-held percussive devices, the electroflo 5000 and the G5 flimm-fighter, for airway clearance in patients with cystic fibrosis (CF), Pediatric Pulmonology, 49, 372, 2014	Conference abstract.
Dunn, C., Davies, Z., Everson, C., Zirbes, J., Kim, L., Milla, C., Study of acute effects on pulmonary function and sputum production with high frequency chest oscillation (HFCWO) and postural drainage aided by handheld percussion (PD-HP), Pediatric Pulmonology, 48, 359, 2013	Abstract only.

Study	Reason for Exclusion
Dwyer, T. J., Cobb, R., Hall, K., Robbins, L., Kelly, P., Bell, S., Bye, P. T., Randomised controlled two-centre trial of non-invasive ventilation (NIV)-assisted chest physiotherapy (CPT) during an acute exacerbation of cystic fibrosis, Journal of Cystic Fibrosis, 9, S73, 2010	Abstract only.
Dwyer, T. J., Robbins, L., Kelly, P., Piper, A. J., Bell, S. C., Bye, P. T., Non-invasive ventilation used as an adjunct to airway clearance treatments improves lung function during an acute exacerbation of cystic fibrosis: a randomised trial, Journal of Physiotherapy, 61, 142-7, 2015	Intervention not prioritised for inclusion in the protocol (NIV as adjunct to airway clearance treatments).
Eaton, T., Young, P., Zeng, I., Kolbe, J., A randomized evaluation of the acute efficacy, acceptability and tolerability of flutter and active cycle of breathing with and without postural drainage in non-cystic fibrosis bronchiectasis, Chronic Respiratory Disease, 4, 23-30, 2007	Cystic fibrosis not included.
Enright, S., Chatham, K., Ionescu, A. A., Unnithan, V. B., Shale, D. J., Inspiratory muscle training improves lung function and exercise capacity in adults with cystic fibrosis, Chest, 126, 405-11, 2004	Intervention (inspiratory muscle training) not included in protocol.
Fainardi, V., Longo, F., Faverzani, S., Tripodi, M. C., Chetta, A., Pisi, G., Short-term effects of high-frequency chest compression and positive expiratory pressure in patients with cystic fibrosis, Journal of Clinical Medicine Research, 3, 279-84, 2011	This study was included in the Cochrane SR McIlwaine (2015). Data incomplete: "the Fainardi study was a cross-over study from which data from the end of the first randomization arm could not be obtained (quote from Cochrane review p.16).
Falk, M., Kelstrup, M., Andersen, J. B., Kinoshita, T., Falk, P., Stovring, S., Gothgen, I., Improving the ketchup bottle method with positive expiratory pressure, PEP, in cystic fibrosis, European Journal of Respiratory Diseases, 65, 423-32, 1984	Intervention (PEP FET) not included in review.
Fauroux, B., Boule, M., Lofaso, F., Zerah, F., Clement, A., Harf, A., Isabey, D., Chest physiotherapy in cystic fibrosis: improved tolerance with nasal pressure support ventilation, Pediatrics, 103, E32, 1999	This study was included in the Cochrane SR Moran 2013. Inspiratory pressure support ventilation (PSV) not included in protocol.
Fauroux, B., Nicot, F., Essouri, S., Hart, N., Clement, A., Polkey, M. I., Lofaso, F., Setting of noninvasive pressure support in young patients with cystic fibrosis, European Respiratory Journal, 24, 624-30, 2004	Intervention (invasive ventilation) not included in protocol, n = 10.
Flume, P. A., Robinson, K. A., O'Sullivan, B. P., Finder, J. D., Vender, R. L., Willey-Courand, D. B., White, T. B., Marshall, B. C., Clinical Practice Guidelines for Pulmonary Therapies, Committee, Cystic fibrosis pulmonary guidelines: airway clearance therapies, Respiratory Care, 54, 522-37, 2009	Review. Individual studies were checked for relevance.
Freitas, Diana A., Dias, A. L. Fernando, Chaves, S. S. Gabriela, Ferreira, M. H. Gardenia, Ribeiro, T. D. Cibele, Guerra, Ricardo O., Mendonca, M. P. P. Karla, Standard (head-down	Intervention (postural drainage) not included in the protocol.

Study	Reason for Exclusion
tilt) versus modified (without head-down tilt) postural drainage in infants and young children with cystic fibrosis, Cochrane Database of Systematic Reviews, 2015	
Giles, D. R., Wagener, J. S., Accurso, F. J., Butler-Simon, N., Short-term effects of postural drainage with clapping vs autogenic drainage on oxygen saturation and sputum recovery in patients with cystic fibrosis, Chest, 108, 952-4, 1995	Intervention (postural drainage) not included in the protocol.
Gondor, M., Nixon, P. A., Mutich, R., Rebovich, P., Orenstein, D. M., Comparison of Flutter device and chest physical therapy in the treatment of cystic fibrosis pulmonary exacerbation, Pediatric Pulmonology, 28, 255-60, 1999	Comparison (PD percussion or vibration vs. PEP) not included in the protocol.
Gozal, D., Nocturnal ventilatory support in patients with cystic fibrosis: comparison with supplemental oxygen, The European Respiratory Journal 1997 Sep;10(9):1999-2003, 1997	This study was included in the Cochrane SR Moran 2013. It looks at the comparison NIV vs control, but for 1 night only (8hr), no follow-up or repeat tests. N < 10 participants.
Guimaraes, F. S., Lopes, A. J., Moco, V. J., Cavalcanti de Souza, F., Silveira de Menezes, S. L., Eltgol acutelly improves airway clearance and reduces static pulmonary volumes in adult cystic fibrosis patients, Journal of Physical Therapy Science, 26, 813-6, 2014	Intervention (ELTGOL) not included in the protocol.
Hofmeyr, J. L., Webber, B. A., Hodson, M. E., Evaluation of positive expiratory pressure as an adjunct to chest physiotherapy in the treatment of cystic fibrosis, Thorax, 41, 951-4, 1986	This study was included in the Cochrane SR Mckoy 2012. The comparison PEP vs FET was not included in the protocol.
Holland, A. E., Denehy, L., Ntoumenopoulos, G., Naughton, M. T., Wilson, J. W., Non-invasive ventilation assists chest physiotherapy in adults with acute exacerbations of cystic fibrosis, Thorax, 58, 880-4, 2003	This study was included in the Cochrane SR Moran 2013. The comparison ACBT vs ACBT and NIV not included in protocol.
Homnick, D. N., White, F., de Castro, C., Comparison of effects of an intrapulmonary percussive ventilator to standard aerosol and chest physiotherapy in treatment of cystic fibrosis, Pediatric Pulmonology, 20, 50-5, 1995	Comparison (intrapulmonary percussive ventilator) not included in the protocol.
Hough,Judith L., Flenady,Vicki, Johnston,Leanne, Woodgate,Paul G., Chest physiotherapy for reducing respiratory morbidity in infants requiring ventilatory support, Cochrane Database of Systematic Reviews, -, 2008	Cystic fibrosis not included.
Houston, B. W., Mills, N., Solis-Moya, A., Inspiratory muscle training for cystic fibrosis, Cochrane Database of Systematic Reviews, 11, CD006112, 2013	Intervention (inspiratory muscle training) not included in the protocol.
Jarad, N. A., Powell, T., Smith, E., Evaluation of a novel sputum clearance technique--hydro-acoustic therapy (HAT) in adult patients with cystic fibrosis: a feasibility study, Chronic Respiratory Disease, 7, 217-27, 2010	This study was included in the Cochrane SR Warnock 2013. The intervention (hydro-acoustic therapy) was not included in the protocol.

Study	Reason for Exclusion
Kluft, J., Beker, L., Castagnino, M., Gaiser, J., Chaney, H., Fink, R. J., A comparison of bronchial drainage treatments in cystic fibrosis, Pediatric Pulmonology, 22, 271-4, 1996	The comparison (chest physical therapy PD vs. high frequency oscillation) not relevant, as PD was not in the protocol.
Kofler,A.M., Carlesi,A., Cutrera,R., Leone,P., Lucidi,V., Rosati,S., Turchetta,A., Vezzoli,P., BiPAP versus PEP as chest physiotherapy in patients with cystic fibrosis [abstract], Pediatric Pulmonology, 26, 344, 1998-, 1998	This study was included in the Cochrane SR Moran 2013. The comparison PEP vs NIV comparison was not in included in the protocol.
Konstan, M. W., Stern, R. C., Doershuk, C. F., Efficacy of the Flutter device for airway mucus clearance in patients with cystic fibrosis, Journal of Pediatrics, 124, 689-93, 1994	The intervention was not included in the protocol (postural drainage).
Krasovskij, S., Amelina, E., Usacheva, M., Samoylenko, V., Krilova, N., High frequency chest wall oscillation (HFCWO) in the treatment of acute pulmonary exacerbation in adult cystic fibrosis (CF) patients, European Respiratory Journal, 42, 2013	Abstract only.
Kriemler, S., Radtke, T., Christen, G., Kerstan-Huber, M., Hebestreit, H., Short-Term Effect of Different Physical Exercises and Physiotherapy Combinations on Sputum Expectoration, Oxygen Saturation, and Lung Function in Young Patients with Cystic Fibrosis, LungLung, 194, 659-64, 2016	The intervention was not relevant to the protocol (combination of physiotherapy and exercise).
Lagerkvist, A. L., Sten, G. M., Redfors, S. B., Lindblad, A. G., Hjalmarson, O., Immediate changes in blood-gas tensions during chest physiotherapy with positive expiratory pressure and oscillating positive expiratory pressure in patients with cystic fibrosis, Respiratory Care, 51, 1154-61, 2006	In Cochrane SR McIlwaine 2015. The outcome (oxygen tension) not relevant to the protocol.
Lima, C. A., De Andrade, A. D. F. D., Campos, S. L., Brandao, D. C., Fregonezi, G., Mourato, I. P., Aliverti, A., De Britto, M. C. A., Effects of noninvasive ventilation on treadmill 6-min walk distance and regional chest wall volumes in cystic fibrosis: Randomized controlled trial, Respiratory Medicine, 108, 1460-1468, 2014	No comparison for NIV.
Maayan, C., Bar-Yishay, E., Yaacobi, T., Marcus, Y., Katznelson, D., Yahav, Y., Godfrey, S., Immediate effect of various treatments on lung function in infants with cystic fibrosis, Respiration, 55, 144-51, 1989	The comparisons (n-acetyl cysteine, salbutamol) are not relevant to the protocol.
Main,Eleanor, Prasad,Ammani, van der Schans,Cees P., Conventional chest physiotherapy compared to other airway clearance techniques for cystic fibrosis, Cochrane Database of Systematic Reviews, -, 2013	Cochrane review. Does not include comparisons relevant for to the protocol. Individual studies were checked for relevance.
McCarren, B., Alison, J. A., Physiological effects of vibration in subjects with cystic fibrosis, European Respiratory Journal, 27, 1204-9, 2006	Outcomes not included in the protocol.
McIlwaine, P. M., Wong, L. T., Peacock, D., Davidson, A. G., Long-term comparative trial of conventional postural drainage and percussion	In Cochrane SR Main 2005. Study not included as intervention (postural drainage) was not in the protocol.

Study	Reason for Exclusion
versus positive expiratory pressure physiotherapy in the treatment of cystic fibrosis, Journal of Pediatrics, 131, 570-4, 1997	
McKoy, N. A., Saldanha, I. J., Odelola, O. A., Robinson, K. A., Active cycle of breathing technique for cystic fibrosis, Cochrane Database of Systematic Reviews, 12, CD007862, 2012	Cochrane review. Does not include comparisons relevant to the protocol. Individual studies were checked for relevance.
McKoy, N. A., Wilson, L. M., Saldanha, I. J., Odelola, O. A., Robinson, K. A., Active cycle of breathing technique for cystic fibrosis, Cochrane Database of Systematic Reviews, 7, CD007862, 2016	This Cochrane SR is an update to a previous SR (McKoy 2010) and was identified at reruns. No additional studies were included in this review.
Miller, S., Hall, D. O., Clayton, C. B., Nelson, R., Chest physiotherapy in cystic fibrosis: a comparative study of autogenic drainage and the active cycle of breathing techniques with postural drainage, Thorax, 50, 165-9, 1995	In Cochrane SR McKoy 2012 and Robinson 2010. The intervention (combination of ACBT and postural drainage) was not prioritised for inclusion in the protocol.
Milne, S. M., Eales, C. J., A pilot study comparing two physiotherapy techniques in patients with cystic fibrosis, South African Journal of Physiotherapy, 60, 3, 2004	Below sample size requirement (n > 10).
Modi, A. C., Cassedy, A. E., Quittner, A. L., Accurso, F., Sontag, M., Koenig, J. M., Ittenbach, R. F., Trajectories of adherence to airway clearance therapy for patients with cystic fibrosis, Journal of Pediatric Psychology, 35, 1028-37, 2010	In Cochrane SR Morrison 2014. The comparison HFCWO vs postural drainage and percussion was not prioritised for inclusion in the protocol.
Morgan, K., Osterling, K., Gilbert, R., Dechman, G., Effects of Autogenic Drainage on Sputum Recovery and Pulmonary Function in People with Cystic Fibrosis: A Systematic Review, Physiotherapy CanadaPhysiother Can, 67, 319-26, 2015	Moderate quality systematic review (AMSTAR score 6/ 11). All included studies checked for inclusion.
Mortensen, J., Falk, M., Groth, S., Jensen, C., The effects of postural drainage and positive expiratory pressure physiotherapy on tracheobronchial clearance in cystic fibrosis, Chest, 100, 1350-7, 1991	In Cochrane SR Warnock 2013. Study not analysed in Cochrane review and N = 10 or less not included in review.
Newhouse,P.A., White,F., Marks,J.H., Homnick,D.N., The intrapulmonary percussive ventilator and flutter device compared to standard chest physiotherapy in patients with cystic fibrosis, Clinical Pediatrics, 37, 427-432, 1998	Sample size not > 10 (n = 10).
Orlik, T., Evaluation of the efficiency of selected thoracic physiotherapy methods used in the treatment of patients with cystic fibrosis, Medycyna Wieku Rozwojowego, 4, 233-46, 2000	Not in English.
Orlik, T., [Evaluation of autodrainage methods in a selected group of cystic fibrosis patients with home environment factors taken into consideration], Medycyna Wieku Rozwojowego, 4, 247-59, 2000	Not English.
Orlik, T., Sands, D., Long-term evaluation of effectiveness for selected chest physiotherapy methods used in the treatment of cystic fibrosis,	Not in English.

Study	Reason for Exclusion
Medycyna Wieku Rozwojowego, 5, 245-57, 2001	
Osman, L. P., Roughton, M., Hodson, M. E., Pryor, J. A., Short-term comparative study of high frequency chest wall oscillation and European airway clearance techniques in patients with cystic fibrosis, Thorax, 65, 196-200, 2010	In Cochrane SR Morrison 2014. The comparison ACBT manual physiotherapy vs. AD not prioritised for inclusion in the protocol.
Pfleger, A., Theissl, B., Oberwaldner, B., Zach, M. S., Self-administered chest physiotherapy in cystic fibrosis: a comparative study of high-pressure PEP and autogenic drainage, Lung, 170, 323-30, 1992	No follow up period.
Pryor, J. A., Tannenbaum, E., Scott, S. F., Burgess, J., Cramer, D., Gyi, K., Hodson, M. E., Beyond postural drainage and percussion: Airway clearance in people with cystic fibrosis, Journal of Cystic Fibrosis, 9, 187-92, 2010	In Cochrane SR McIlwaine 2015. Data could not be analysed as only p values are provided for the quality of life outcome. Tool for measuring quality of life not included in the protocol.
Pryor, J. A., Webber, B. A., Hodson, M. E., Warner, J. O., The Flutter VRP1 as an adjunct to chest physiotherapy in cystic fibrosis, Respiratory Medicine, 88, 677-81, 1994	The comparison (ACBT vs Flutter) was not prioritised for inclusion in the protocol.
Regelmann, W. E., Elliott, G. R., Warwick, W. J., Clawson, C. C., Reduction of sputum Pseudomonas aeruginosa density by antibiotics improves lung function in cystic fibrosis more than do bronchodilators and chest physiotherapy alone, American Review of Respiratory Disease, 141, 914-21, 1990	The comparison (vs antibiotics) was not included the protocol.
Reisman, J. J., Rivington-Law, B., Corey, M., Marcotte, J., Wannamaker, E., Harcourt, D., Levison, H., Role of conventional physiotherapy in cystic fibrosis, Journal of Pediatrics, 113, 632-6, 1988	The intervention (postural drainage) was not included in the protocol.
Robinson, K. A., McKoy, N., Saldanha, I., Odelola, O. A., Active cycle of breathing technique for cystic fibrosis, Cochrane Database of Systematic Reviews, CD007862, 2010	Cochrane SR. Includes one comparison of interest, but relevant study (Miller 2005) has been excluded from this review as it compares ACBT with postural drainage, which is a comparison not prioritised for inclusion in the protocol.
Rodriguez Hortal, M. C., Nygren-Bonnier, M., Hjelte, L., Non-invasive ventilation as airway clearance technique in cystic fibrosis [with consumer summary], Physiotherapy Research International 2016 Feb 29:Epub ahead of print, 2016	This comparison was not prioritised for inclusion in the protocol (NIV vs PEP).
Rossman, C. M., Waldes, R., Sampson, D., Newhouse, M. T., Effect of chest physiotherapy on the removal of mucus in patients with cystic fibrosis, American Review of Respiratory Disease, 126, 131-5, 1982	In Cochrane SR Warnock 2013. Intervention (postural drainage) not included in the protocol.
Sanchez Riera, H., Dapena Fernandez, F. J., Gomez Dominguez, F., Ortega Ruiz, F., Elias Hernandez, T., Montemayor Rubio, T., Castillo Gomez, J., [Comparative study of the efficacy of 2 respiratory physiotherapy protocols for	Not English.

Study	Reason for Exclusion
patients with cystic fibrosis], Archivos de bronconeumologia, 35, 275-9, 1999	
Sandsund, C. A., Roughton, M., Hodson, M. E., Pryor, J. A., Musculoskeletal techniques for clinically stable adults with cystic fibrosis: a preliminary randomised controlled trial, Physiotherapy, 97, 209-17, 2011	The comparison (musculoskeletal treatment) was not included in the protocol.
Santana-Sosa, E., Gonzalez-Saiz, L., Groeneveld, I. F., Villa-Asensi, J. R., Gomez de Aguero, M. I. B., Fleck, S. J., Lopez-Mojares, L. M., Perez, M., Lucia, A., Benefits of combining inspiratory muscle with 'whole muscle' training in children with cystic fibrosis: a randomised controlled trial [with consumer summary], British Journal of Sports Medicine 2013 Oct;48(20):1513-1517, 2013	The intervention (inspiratory muscle training) was not included in the protocol.
Scherer, T. A., Barandun, J., Martinez, E., Wanner, A., Rubin, E. M., Effect of high-frequency oral airway and chest wall oscillation and conventional chest physical therapy on expectoration in patients with stable cystic fibrosis, Chest, 113, 1019-27, 1998	No relevant numerical evidence.
Sokol, Gil, Vilozni, Daphna, Hakimi, Ran, Lavie, Moran, Sarouk, Ifat, Bar, Bat-El, Dagan, Adi, Ofek, Miryam, Efrati, Ori, The Short-Term Effect of Breathing Tasks Via an Incentive Spirometer on Lung Function Compared With Autogenic Drainage in Subjects With Cystic Fibrosis, Respiratory CareRespir Care, 60, 1819-1825, 2015	Not an RCT.
Sontag, M. K., Quittner, A. L., Modi, A. C., Koenig, J. M., Giles, D., Oermann, C. M., Konstan, M. W., Castile, R., Accurso, F. J., Investigators,, Coordinators of the Airway Secretion Clearance, Trial, Lessons learned from a randomized trial of airway secretion clearance techniques in cystic fibrosis, Pediatric Pulmonology, 45, 291-300, 2010	Incomplete reporting: only p values reported.
Steen, H. J., Redmond, A. O., O'Neill, D., Beattie, F., Evaluation of the PEP mask in cystic fibrosis, Acta Paediatrica Scandinavica, 80, 51-6, 1991	The comparison (PEP vs PEP FET) was not prioritised for inclusion in the protocol.
Swaminathan, Narasimman, Robinson, A. Karen, Ray, Amita, Autogenic drainage for airway clearance in cystic fibrosis, Cochrane Database of Systematic Reviews, 2012	Cochrane systematic review - protocol only.
Thomas, J., Cook, D. J., Brooks, D., Chest physical therapy management of patients with cystic fibrosis. A meta-analysis, American Journal of Respiratory & Critical Care Medicine, 151, 846-50, 1995	Low quality systematic review - 2/11 total score in AMSTAR checklist. Individual studies were checked for relevance.
Tonnesen, P., Kelstrup, M., Self-administered positive end-expiratory pressure (PEEP) employing a face mask as an alternative to conventional chest physiotherapy, Ugeskr-Laeg, 144, 1532-6, 1982	Not English.

Study	Reason for Exclusion
Van Asperen, P. P., Jackson, L., Hennessy, P., Brown, J., Comparison of a positive expiratory pressure (PEP) mask with postural drainage in patients with cystic fibrosis, Australian Paediatric Journal, 23, 283-4, 1987	In Cochrane SR Main 2005. Study not included as the intervention (postural drainage) was not in the protocol.
van der Schans, C. P., van der Mark, T. W., de Vries, G., Piers, D. A., Beekhuis, H., Dankert-Roelse, J. E., Postma, D. S., Koeter, G. H., Effect of positive expiratory pressure breathing in patients with cystic fibrosis, Thorax, 46, 252-6, 1991	In Cochrane SR Warnock 2013 (not analysed). Studies not analysed and below n = 10 excluded from review.
Van Ginderdeuren, F., Verbanck, S., Van Cauwelaert, K., Vanlaethem, S., Schuermans, D., Vincken, W., Malfroot, A., Chest physiotherapy in cystic fibrosis: short-term effects of autogenic drainage preceded by wet inhalation of saline versus autogenic drainage preceded by intrapulmonary percussive ventilation with saline, Respiration, 76, 175-80, 2008	The comparison (same technique with the addition of saline) was not relevant to protocol.
Varekojis, S. M., Douce, F. H., Flucke, R. L., Filbrun, D. A., Tice, J. S., McCoy, K. S., Castile, R. G., A comparison of the therapeutic effectiveness of and preference for postural drainage and percussion, intrapulmonary percussive ventilation, and high-frequency chest wall compression in hospitalized cystic fibrosis patients, Respiratory Care, 48, 24-28, 2004	The interventions were not included in the protocol (intrapulmonary ventilation, postural drainage percussion).
Webber, B, Parker, R, Hofmeyr, J, Hodson, M, Evaluation of self-percussion during postural drainage using the forced expiration technique, Physiother Pract, 1, 42-5., 1985	The comparison was not prioritised for inclusion in the protocol (combination of self-percussion and PD vs PD alone).
Webber, Ba, Hofmeyr, Jl, Morgan, Md, Hodson, Me, Effects of postural drainage, incorporating the forced expiration technique, on pulmonary function in cystic fibrosis, Br J Dis Chest, 80, 353-9., 1986	Not an RCT.
White, D, Stiller, K, Willson, K, The role of thoracic expansion exercises during the active cycle of breathing techniques, Physiotherapy Theory and Practice, 13, 155-62., 1997	Intervention not relevant to the protocol (thoracic expansion exercises during ACBT).

H.8 Mucoactive agents

Study	Reason for Exclusion
Aitken, M., Bilton, D., Fox, H., Charlton, B., Cf,, C. F. Study Investigators, Bronchitol (inhaled dry powder mannitol) in adult patients with cystic fibrosis [abstract], Journal of Cystic Fibrosis, 11, S68, 2012	Conference abstract
Baran, D., Clinical double-blind trial of oral acetylcysteine and placebo in cystic fibrosis. <ORIGINAL> KLINISCHER DOPPELBLINDVERSUCH MIT ORALEM ACETYLCYSTEIN UND PLACEBO BEI ZYSTISCHER FIBROSE, Die Therapiewoche, 30, 2034-9, 1980	Paper not in English

Study	Reason for Exclusion
Bilton, D., Bellon, G., Charlton, B., Cooper, P., De Boeck, K., Flume, P. A., Fox, H. G., Gallagher, C. G., Geller, D. E., Haarman, E. G., Hebestreit, H. U., Kolbe, J., Lapey, A., Robinson, P., Wu, J., Zuckerman, J. B., Aitken, M. L., Cf,, C. F. Investigators, Pooled analysis of two large randomised phase III inhaled mannitol studies in cystic fibrosis, Journal of Cystic Fibrosis, 12, 367-76, 2013	Pooled results from Aitken 2012 and Bilton 2011
Bollert, F. G., Paton, J. Y., Marshall, T. G., Calvert, J., Greening, A. P., Innes, J. A., Recombinant DNase in cystic fibrosis: a protocol for targeted introduction through n-of-1 trials. Scottish Cystic Fibrosis Group, European Respiratory Journal, 13, 107-13, 1999	No variability/uncertainty reported for outcome of interest
Brivio, A., Ceruti, C., Gambazza, S., Colombo, C., Randomized double-blind monocentric trial on tolerability, acceptability and efficacy of two formulations of inhaled 7% hypertonic saline with and without hyaluronic acid in reducing airways inflammation in patients with cystic fibrosis - preliminary results [abstract], Journal of Cystic Fibrosis : Official Journal of the European Cystic Fibrosis Society, 12, S104, 2013	Conference abstract
Brown, A, Skidmore, B, Hypertonic saline nebules for patients with cystic fibrosis and bronchioectasis: a review of the clinical and cost-effectiveness (Structured abstract), Health Technology Assessment Database, 2016	HTA. References of included studies were assessed to identify relevant studies.
Buonpensiero, P., De Gregorio, F., Sepe, A., Di Pasqua, A., Ferri, P., Siano, M., Terlizzi, V., Raia, V., Hyaluronic acid improves "pleasantness" and tolerability of nebulized hypertonic saline in a cohort of patients with cystic fibrosis, Advances in Therapy, 27, 870-8, 2010	Intervention not of interest
Burness, C. B., Keating, G. M., Mannitol dry powder for inhalation: in patients with cystic fibrosis, Drugs, 72, 1411-21, 2012	Reports results from previously published RCTs
Bush, A., Hodson, M. E., Geddes, D. M., Rosenthal, M., Dinwiddie, R., Wallis, C., Stableforth, D. E., Conway, S. P., Littlewood, J. M., Dornase alfa for cystic fibrosis. Patients should not be denied a safe, effective treatment, BMJ (Clinical research ed.), 310, 1533, 1995	Letter
Bush, A., Hodson, M. E., Geddes, D. M., Rosenthal, M., Dinwiddie, R., Wallis, C., Stableforth, D. E., Conway, S. P., Littlewood, J. M., Collier, J., Dornase alfa for cystic fibrosis [7], British Medical Journal, 310, 1533-1534, 1995	Letter
CADTH, Dornase alfa for patients with cystic fibrosis: a review of the clinical efficacy and cost-effectiveness (Structured abstract), Health Technology Assessment Database, 2016	Abstract of study that had already been assessed for inclusion based on full text
CADTH, Sodium chloride inhalation for the treatment of cystic fibrosis: a review of the clinical evidence, cost-effectiveness and	Abstract of study that had already been assessed for inclusion based on full text

Study	Reason for Exclusion
guidelines (Structured abstract), Health Technology Assessment Database, 2016	
Canadian Agency for Drugs and Technologies in Health, Dornase alfa for patients with cystic fibrosis: a review of the clinical efficacy and cost-effectiveness, 2013	Rapid response report. It includes Jones 2010 Cochrane review (already included) and an additional RCT that is already included (Amin 2011)
Canadian Agency for Drugs and Technology in Health, Sodium chloride inhalation for the treatment of cystic fibrosis: a review of the clnical evidence, cost-effectiveness and guidelines, 2012	CADTH Rapid Response Report. Individual trials have been identified for inclusion.
Cantin, A. M., Berthiaume, Y., Cloutier, D., Martel, M., Prolastin aerosol therapy and sputum taurine in cystic fibrosis, Clinical and Investigative Medicine, 29, 201-207, 2006	Intervention not of interest
Caporalini, R., Giosue, G. L., Neltenexine in lung diseases: An open, randomised, controlled study versus N-acetylcysteine comparison, Minerva pneumologica, 40, 57-62, 2001	No outcome of interest
Carter, E. R., Dornase alfa in young patients with cystic fibrosis, Journal of Pediatrics, 141, 838; author reply 838-9, 2002	Letter
Catalan Agency for Health Information, Assessment and Quality (CAHIAQ) - formerly CAHTA, Manitol for the treatment of cystic fibrosis in adults aged 18 years and above as an add-on therapy to best standard of care (Project record), Health Technology Assessment Database, 2015	Full report not available
Centre for Reviews and Dissemination, Systematic review of N-acetylcysteine in cystic fibrosis (Structured abstract), Database of Abstracts of Reviews of Effects, 2015	Abstract only. Full SR: Duijvestijn 1999 (STAR ID. 360653)
Centre for Reviews and Dissemination, rhDNase therapy for the treatment of cystic fibrosis patients with mild to moderate lung disease (Structured abstract), Database of Abstracts of Reviews of Effects, 2015	Abstract. Full text paper has been assessed for inclusion.
Charlton, B., Lassig, A., C. F. Study Investigators, Inhaled dry powder mannitol (bronchitol) improves FEV1 in cystic fibrosis [abstract], Tsanz, 2006	Conference abstract
Christopher, F., Chase, D., Stein, K., Milne, R., rhDNase therapy for the treatment of cystic fibrosis patients with mild to moderate lung disease, Journal of Clinical Pharmacy & Therapeutics, 24, 415-26, 1999	Low quality systematic review
Cramer, G. W., Bosso, J. A., The role of dornase alfa in the treatment of cystic fibrosis, Annals of Pharmacotherapy, 30, 656-61, 1996	Low quality systematic review
Daviskas, E., Anderson, S. D., Jaques, A., Charlton, B., Inhaled mannitol improves the hydration and surface properties of sputum in patients with cystic fibrosis, Chest, 137, 861-8, 2010	Compares mannitol and placebo, but does not include outcomes relevant to the protocol

Study	Reason for Exclusion
Dentice, R., Elkins, M., Timing of dornase alfa inhalation for cystic fibrosis, Cochrane Database of Systematic Reviews, 7, CD007923, 2016	Non relevant comparison
Deterding, R. R., Lavange, L. M., Engels, J. M., Mathews, D. W., Coquillette, S. J., Brody, A. S., Millard, S. P., Ramsey, B. W., Cystic Fibrosis Therapeutics Development, Network, the Inspire 08-103 Working, Group, Phase 2 randomized safety and efficacy trial of nebulized denufosol tetrasodium in cystic fibrosis, American Journal of Respiratory & Critical Care Medicine, 176, 362-9, 2007	Intervention not of interest
Deterding, R., Retsch-Bogart, G., Milgram, L., Gibson, R., Daines, C., Zeitlin, P. L., Milla, C., Marshall, B., Lavange, L., Engels, J., Mathews, D., Gorden, J., Schaberg, A., Williams, J., Ramsey, B., Cystic Fibrosis Foundation Therapeutics Development, Network, Safety and tolerability of denufosol tetrasodium inhalation solution, a novel P2Y2 receptor agonist: results of a phase 1/phase 2 multicenter study in mild to moderate cystic fibrosis, Pediatric Pulmonology, 39, 339-48, 2005	Intervention not of interest
Duijvestijn, Y. C., Brand, P. L., Systematic review of N-acetylcysteine in cystic fibrosis, Acta Paediatrica, 88, 38-41, 1999	Low quality systematic review
Dwyer, T., Elkins, M., Dentice, R., Forbes, S., McArthur, M., Cooper, P., et al.,, Saline at a lower tonicity in cystic fibrosis (SALTI-CF) trial - a randomised, controlled trial comparing 0.9% v 3% v 6% nebulised saline [abstract], Journal of Cystic Fibrosis, 12, 2013	Conference abstract
Eisenberg, J., [Clinical development of rhDNase in the United States], Archives de pediatrie : organe officiel de la Societe francaise de pediatrie, 2, 674-8, 1995	Conference abstract
Eisenberg, J. D., Aitken, M. L., Dorkin, H. L., Harwood, I. R., Ramsey, B. W., Schidlow, D. V., Wilmott, R. W., Wohl, M. E., Fuchs, H. J., Christiansen, D. H., Smith, A. L., Safety of repeated intermittent courses of aerosolized recombinant human deoxyribonuclease in patients with cystic fibrosis, Journal of Pediatrics, 131, 118-24, 1997	Study not randomised
Eng, P. A., Morton, J., Douglass, J. A., Riedler, J., Wilson, J., Robertson, C. F., Short-term efficacy of ultrasonically nebulized hypertonic saline in cystic fibrosis, Pediatric Pulmonology, 21, 77-83, 1996	No relevant data
Eng, P., Morton, J., Douglas, J., Riedler, J., Wilson, J., Robertson, C. F., Efficacy of short-term ultransonically nebulised hypertonic saline in patients with cystic fibrosis [abstract], European Respiratory Journal. Supplement., 8, 510s, 1995	Conference abstract
Fitzgerald, D. A., Hilton, J., Jepson, B., Smith, L., A crossover, randomized, controlled trial of	Intervention comparison does not fit protocol

Study	Reason for Exclusion
dornase alfa before versus after physiotherapy in cystic fibrosis, Pediatrics, 116, e549-54, 2005	
Frederiksen, B., Pressler, T., Hansen, A., Koch, C., Hoiby, N., Effect of aerosolized rhDNase (Pulmozyme) on pulmonary colonization in patients with cystic fibrosis, Acta Paediatrica, 95, 1070-4, 2006	No outcome of interest
Gotz, M., Kraemer, R., Kerrebijn, K. F., Popow, C., Oral acetylcysteine in cystic fibrosis. A co-operative study, European Journal of Respiratory Diseases - Supplement, 111, 122-6, 1980	Data poorly reported, cannot be pooled.
Grasemann, H., Tullis, E., Ratjen, F., A randomized controlled trial of inhaled L-arginine in patients with cystic fibrosis, Journal of Cystic Fibrosis, 12, 468-74, 2013	Intervention not of interest
Harms, H. K., Matouk, E., Tournier, G., Von Der Hardt, H., Weller, P. H., Romano, L., Heijerman, H. G. M., FitzGerald, M. X., Richard, D., Strandvik, B., Kolbe, J., Kraemer, R., Michalsen, H., Multicenter, open-label study of recombinant human DNase in cystic fibrosis patients with moderate lung disease, Pediatric Pulmonology, 26, 155-161, 1998	Study not randomised. Not control group
Heaf, D. P., Tyson, S., Dinwiddie, R., Matthew, D., A comparison of inhaled therapies in children with cystic fibrosis [abstract], 9th International Cystic Fibrosis Congress, 4, 1984	Conference abstract
Health Technology Assessment, Mannitol dry powder for inhalation for the treatment of cystic fibrosis (Project record), Health Technology Assessment Database, 2016	Project record of study that had already been included
Hodson, M., Multicentric trial of rhDNase in patients with cystic fibrosis and severe pulmonary dysfunction, Archives De Pediatrie, 2, 679-81, 1995	Study not in English
Homola, L., Holeikova, A., Mikolasek, P., Krbkova, L., Efficacy of inhaled amiloride solution versus hypertonic saline, prospective open label single center study in children with cystic fibrosis [abstract], Journal of Cystic Fibrosis, 12, S98, 2013	Conference abstract
Howatt, W. F., DeMuth, G. R., A double-blind study of the use of acetylcysteine in patients with cystic fibrosis, University of Michigan Medical Center Journal, 32, 82-5, 1966	No outcomes of interest
Hubbard, R. C., McElvaney, N. G., Birrer, P., Shak, S., Robinson, W. W., Jolley, C., Wu, M., Chernick, M. S., Crystal, R. G., A preliminary study of aerosolized recombinant human deoxyribonuclease I in the treatment of cystic fibrosis Comment in: N Engl J Med 1992 Aug 20;327(8):571, New England Journal of Medicine, 326, 812-5, 1992	Cross-over trial (n=11 completed both arms). Data is poorly reported and cannot be pooled.

Study	Reason for Exclusion
Jones, A. P., Wallis, C., Dornase alfa for cystic fibrosis, Cochrane Database of Systematic Reviews, CD001127, 2010	More recent update of this review was included (Yang 2016)
Kearney, C. E., Wallis, C. E., Deoxyribonuclease for cystic fibrosis, Cochrane Database of Systematic Reviews, CD001127, 2000	There is an update: see Jones 2010
Mainz, J. G., Schien, C., Schiller, I., Schadlich, K., Koitschev, A., Koitschev, C., Riethmuller, J., Graepler-Mainka, U., Wiedemann, B., Beck, J. F., Sinonasal inhalation of dornase alfa administered by vibrating aerosol to cystic fibrosis patients: a double-blind placebo-controlled cross-over trial, Journal of Cystic Fibrosis, 13, 461-70, 2014	Data is poorly reported and cannot be pooled.
Middleton, A., Robinson, P. D., McKay, K., Jaffe, A., Selvadurai, H., A pilot study of inhaled dry-powder mannitol during cystic fibrosis-related pulmonary exacerbation, European Respiratory Journal, 45, 541-4, 2015	Letter to the editor
Mitchell, E. A., Elliott, R. B., Controlled trial of oral N-acetylcysteine in cystic fibrosis, Australian Paediatric Journal, 18, 40-2, 1982	No outcomes of interest
Mitchell, E. A., Elliott, R. B., Failure of oral N-acetylcysteine to improve the malabsorption of cystic fibrosis, Australian Paediatric Journal, 17, 207-8, 1981	No outcomes of interest
Nasr, S. Z., Kuhns, L. R., Brown, R. W., Hurwitz, M. E., Sanders, G. M., Strouse, P. J., Aerosolized recombinant human DNase in cystic fibrosis patients younger than 5 years of age [abstract], Pediatric Pulmonology, 19, 1999	Conference abstract
Nasr, S. Z., Kuhns, L. R., Brown, R. W., Hurwitz, M. E., Sanders, G. M., Strouse, P. J., Use of computerized tomography and chest x-rays in evaluating efficacy of aerosolized recombinant human DNase in cystic fibrosis patients younger than age 5 years: a preliminary study, Pediatric Pulmonology, 31, 377-82, 2001	CT scan not an outcome of interest
National Horizon Scanning Centre, Mannitol dry powder for inhalation (Bronchitol) for cystic fibrosis (Structured abstract), Health Technology Assessment Database, 2016	Abstract of study that had already been assessed for inclusion based on full text
National Horizon Scanning Centre, Mannitol dry powder for inhalation (Bronchitol) for cystic fibrosis, 2008	National Horizon Scanning Centre. Trials have been identified for inclusion.
Nct,, Sino-nasal Inhalation of Sodium Chloride 6,0% in Patients With Cystic Fibrosis and Chronic Rhinosinusitis A Multicenter, Randomized, Double-blind, Placebo-controlled, Prospective Clinical Trial, Clinicaltrials.gov [www.clinicaltrials.gov]., 2010	Trial results not yet published
Nolan, S. J., Thornton, J., Murray, C. S., Dwyer, T., Inhaled Mannitol (Bronchitol) for Cystic Fibrosis, Paediatric Respiratory ReviewsPaediatr Respir Rev, 18, 52-4, 2016	Summary of systematic review that has been included

Study	Reason for Exclusion
Noone, P. G., Hamblett, N., Accurso, F., Aitken, M. L., Boyle, M., Dovey, M., Gibson, R., Johnson, C., Kellerman, D., Konstan, M. W., Milgram, L., Mundahl, J., Retsch-Bogort, G., Rodman, D., Williams-Warren, J., Wilmott, R. W., Zeitlin, P., Ramsey, B., Cystic Fibrosis Therapeutics Development Research, Group, Safety of aerosolized INS 365 in patients with mild to moderate cystic fibrosis: results of a phase I multi-center study, Pediatric Pulmonology, 32, 122-8, 2001	Intervention not of interest
Paul, K., Ballmann, M., Griese, M., Rietschel, E., Chen, C., Schink, T., Shute, J. K., Doring, G., Ratjen, F., Effect of rhDNase on endobronchial inflammation in CF patients with mild lung disease: results of the multi-center BEAT study [abstract], Pediatric Pulmonology, 24, 310-1, 2002	Conference abstract
Paul, K., Rietschel, E., Ballmann, M., Griese, M., Worlitzsch, D., Shute, J., Chen, C., Schink, T., Doring, G., van Koningsbruggen, S., Wahn, U., Ratjen, F., Bronchoalveolar Lavage for the Evaluation of Antiinflammatory Treatment Study, Group, Effect of treatment with dornase alpha on airway inflammation in patients with cystic fibrosis, American Journal of Respiratory & Critical Care Medicine, 169, 719-25, 2004	No relevant data
Potter, R. W., Hurren, T. J., Nickerson, C., Hatley, R. H., Comparison of the delivery characteristics of dornase alfa from the I-NEB(R) AAD(R) system and the Sidestream(R) et nebulizer [abstract], Pediatric Pulmonology, 43, 2008	Conference abstract
Purins, A, Hiller, JE, Hypertonic saline therapy for cystic fibrosis (Structured abstract), Health Technology Assessment Database, 2016	Abstract of study that had already been assessed for inclusion based on full text
Purins, A, Hiller, JE, Adelaide Health Technology Assessment, Hypertonic saline therapy for cystic fibrosis, Horizon Scanning Prioritising Summary Volume 24, 2009	Horizon Scanning Technology.
Ramsey, B., For the Pulmozyme Study, Group, A summary of the results of the phase III multicenter clinical trial: Aerosol administration of recombinant human DNase reduces the risk of respiratory tract infections and improves pulmonary function in patients with cystic fibrosis [abstract], Pediatric Pulmonology, 9, 152-3, 1993	No relevant data
Ratjen, F., Inhaled hypertonic saline produces small increases in lung function in patients with cystic fibrosis, Journal of Pediatrics, 149, 142, 2006	Journal comment
Ratjen, F., Paul, K., van Koningsbruggen, S., Breitenstein, S., Rietschel, E., Nikolaizik, W., DNA concentrations in BAL fluid of cystic fibrosis patients with early lung disease: influence of	Study not randomised

Study	Reason for Exclusion
treatment with dornase alpha, Pediatric Pulmonology, 39, 1-4, 2005	
Ratjen, F., Posselt, H. G., WonneR,, Stover, B., Bender, Swjw, A double-blind placebo controlled trial with ambroxol and N-Acetylcysteine for mucolytic treatment in cystic fibrosis [abstract], 9th International Cystic Fibrosis Congress, 1984	Conference abstract
Riemsma, R, Al, MJ, Armstrong, N, Misso, K, Allen, A, Manning, N, Tushabe, DA, Severens, JL, Kleijnen, J, Kleijnen Systematic Reviews Ltd, Mannitol dry powder for inhalation for the treatment of cystic fibrosis, 2011	Review of already included technology appraisal
Riethmueller, J., Borth-Bruhns, T., Kumpf, M., Vonthein, R., Wiskirchen, J., Stern, M., Hofbeck, M., Baden, W., Recombinant human deoxyribonuclease shortens ventilation time in young, mechanically ventilated children.[Erratum appears in Pediatr Pulmonol. 2006 Apr;41(4):388], Pediatric Pulmonology, 41, 61-6, 2006	Not CF patients
Robinson, M., Daviskas, E., Eberl, S., Baker, J., Chan, H. K., Anderson, S. D., Bye, P. T., The effect of inhaled mannitol on bronchial mucus clearance in cystic fibrosis patients: a pilot study, European Respiratory Journal, 14, 678-85, 1999	No relevant data
Rodgers, H. C., Knox, A. J., Randomised controlled crossover study of the effect of topically applied benzamil, amiloride or saline on nasal potential difference in cystic fibrosis [abstract], Thorax, 53, A19, 1998	Conference abstract
Ros, M., Casciaro, R., Lucca, F., Alatri, F., Salonini, E., Favilli, F., et al.,, Tolerability and acceptability in patients with cystic fibrosis (CF) of two formulations of 7% hypertonic saline: a prospective multicenter clinical study [abstract], Pediatric Pulmonology, 47, 364, 2012	Conference abstract
Ros, M., Casciaro, R., Lucca, F., Troiani, P., Salonini, E., Favilli, F., Quattrucci, S., Sher, D., Assael, B. M., Hyaluronic acid improves the tolerability of hypertonic saline in the chronic treatment of cystic fibrosis patients: a multicenter, randomized, controlled clinical trial, Journal of aerosol medicine & pulmonary drug delivery, 27, 133-7, 2014	Hyaluronic acid not an intervention of interest
Sawicki, G. S., Chou, W., Raimundo, K., Trzaskoma, B., Konstan, M. W., Randomized trial of efficacy and safety of dornase alfa delivered by eRapid nebulizer in cystic fibrosis patients, Journal of Cystic Fibrosis, 14, 777-83, 2015	Non relevant comparison
Serisier, D. J., Shute, J. K., Hockey, P. M., Higgins, B., Conway, J., Carroll, M. P., Inhaled heparin in cystic fibrosis, European Respiratory Journal, 27, 354-8, 2006	Intervention not of interest
Shak, S., Aerosolized recombinant human DNase I for the treatment of cystic fibrosis, Chest, 107, 65S-70S, 1995	Secondary report of a previously published RCT

Study	Reason for Exclusion
Stafanger, G., Garne, S., Howitz, P., Morkassel, E., Koch, C., The clinical effect and the effect on the ciliary motility of oral N-acetylcysteine in patients with cystic fibrosis and primary ciliary dyskinesia, European Respiratory Journal, 1, 161-7, 1988	Data poorly reported, cannot be pooled.
Stafanger, G., Koch, C., N-acetylcysteine in cystic fibrosis and Pseudomonas aeruginosa infection: clinical score, spirometry and ciliary motility, European Respiratory Journal, 2, 234-7, 1989	Data poorly reported, cannot be pooled.
Stroobant, J., Heaf, D. P., Tyson, S., Matthew, D. J., Effect of inhaled azlocillin, mistabron and combination therapy in children with cystic fibrosis, Pediatric research, 19, 1099	Letter
Subbarao, P., Stanojevic, S., Brown, M., Jensen, R., Rosenfeld, M., Davis, S., Brumback, L., Gustafsson, P., Ratjen, F., Lung clearance index as an outcome measure for clinical trials in young children with cystic fibrosis. A pilot study using inhaled hypertonic saline, American Journal of Respiratory & Critical Care Medicine, 188, 456-60, 2013	No outcome of interest (uses Lung clearance index instead of FEV)
Suri, R, Wallis, C, Bush, A, Thompson, S, Normand, C, Flather, M, Grieve, R, Metcalfe, C, Lees, B, A comparative study of hypertonic saline, daily and alternate-day rhDNase in children with cystic fibrosis (Structured abstract), Health Technology Assessment Database, 2016	Structured abstract of a health technology assessment that was assessed for inclusion in this review using the full text.
Suri, R., Metcalfe, C., Lees, B., Flather, M., Normand, C., Thompson, S., Bush, A., Wallis, C., A cross-over comparative study of hypertonic saline alternate day and daily rhDNase in children with Cystic Fibrosis [abstract], Thorax, 55, 2000	Conference abstract
Suri,R., Wallis,C., Bush,A., Thompson,S., Normand,C., Flather,M., Grieve,R., Metcalfe,C., Lees,B., A comparative study of hypertonic saline, daily and alternate-day rhDNase in children with cystic fibrosis, Health Technology Assessment, 6, -, 2002	No relevant comparison
Taylor, L. M., Kuhn, R. J., Hypertonic saline treatment of cystic fibrosis, Annals of Pharmacotherapy, 41, 481-4, 2007	Low quality systematic review
Teper,A., Jaques,A., Charlton,B., Inhaled mannitol in patients with cystic fibrosis: A randomised open-label dose response trial, Journal of Cystic Fibrosis, 10, 1-8, 2011	Dose-response study. This comparison has not been prioritized for inclusion in the review.
Wark,Peter, McDonald,Vanessa M., Nebulised hypertonic saline for cystic fibrosis, Cochrane Database of Systematic Reviews, -, 2010	There is a recent update for this review
Zhang,Linjie, MendozaSassi,Raul A., Wainwright,Claire, Klassen,Terry P., Nebulised hypertonic saline solution for acute bronchiolitis in infants, Cochrane Database of Systematic Reviews, -, 2013	Not CF patients

H.9 Pulmonary infection

Study	Reason for Exclusion
Aaron, S., Aspergillus (not abpa)-not to treat, Pediatric Pulmonology, 47, 209-10, 2012	This is a conference abstract. The full published RCT (Aaron 2012) is included in the review (protocol 3).
Adeboyeku, D., Jones, A. L., Hodson, M. E., Twice vs three-times daily antibiotics in the treatment of pulmonary exacerbations of cystic fibrosis, Journal of Cystic Fibrosis, 10, 25-30, 2011	Excluded from protocol 2 because the reported comparison regards the frequency of antibiotic administration only.
Adeboyeku, D., Scott, S., Hodson, M. E., Open follow-up study of tobramycin nebuliser solution and colistin in patients with cystic fibrosis, Journal of Cystic Fibrosis, 5, 261-3, 2006	Excluded from protocol 3 as cannot identify treatment difference at specific time points.
Agostini, M., Barlocco, G., Bonomi, U., Alternative antibiotics against Pseudomonas infections in cystic fibrosis. In vitro activity, pharmacokinetics, and double-blind randomized clinical trial with azlocillin, piperacillin, cefoperazone, ceftazidime, cefsulodin, cefotaxime and moxalactam. Preliminary results, Drugs under Experimental and Clinical Research, 98, 671-686, 1983	This study was included in Cochrane SR Hurley 2015, but no data was used for the review.(protocol 2)
Ahmed, Molla I., Mukherjee, Saptarshi, Treatment for chronic Staphylococcus aureus pulmonary infection in people with cystic fibrosis, Cochrane Database of Systematic Reviews, 2015	Protocol only. The review has been included in review 3.
Avgeri, S. G., Matthaiou, D. K., Dimopoulos, G., Grammatikos, A. P., Falagas, M. E., Therapeutic options for Burkholderia cepacia infections beyond co-trimoxazole: a systematic review of the clinical evidence, International Journal of Antimicrobial Agents, 33, 394-404, 2009	Intervention. This SR evaluates co-trimoxazole in the treatment of B. Cepacia and includes people with different illness severity (protocols 2 and 3). The only RCT included in the review had patients suffering an exacerbation, but the intervention was not relevant to protocol 2.
Ballestero, S., Villaverde, R., Escobar, H., Baquero, F., Susceptibility to various antimicrobial agents of Staphylococcus aureus isolates from cystic fibrosis patients, European Journal of Clinical Microbiology & Infectious Diseases, 11, 1193-4, 1992	Study design: non-randomized
Bosso, J. A., Black, P. G., Matsen, J. M., Ciprofloxacin versus tobramycin plus azlocillin in pulmonary exacerbations in adult patients with cystic fibrosis, American Journal of Medicine, 82, 180-4, 1987	Small sample size (N=20. Also the study compares ciprofloxacin vs IV tobramycin and azlocillin for exacerbation, which is not a relevant comparison (not relevant for protocol 2).
Bosso,J.A., Black,P.G., Controlled trial of aztreonam vs. tobramycin and azlocillin for acute pulmonary exacerbations of cystic fibrosis, Pediatric Infectious Disease Journal, 7, 171-176, 1988	Excluded from protocol 2 because the reported comparison is of antibiotics that are not relevant to the protocol.
Bowler, S. D., Effect of long-term treatment with azithromycin on disease parameters in cystic fibrosis, Japanese Journal of Antibiotics, 56 Suppl A, 38, 2003	Not in English
Byrne, S., Maddison, J., Connor, P., Doughty, I., Dodd, M., Jenney, M., Webb, A. K., David, T. J., Clinical evaluation of meropenem versus	Study not randomised.

Study	Reason for Exclusion
ceftazidime for the treatment of Pseudomonas spp. infections in cystic fibrosis patients, Journal of Antimicrobial Chemotherapy, 36 Suppl A, 135-43, 1995	
Cai, Y., Chai, D., Wang, R., Bai, N., Liang, B. B., Liu, Y., Effectiveness and safety of macrolides in cystic fibrosis patients: a meta-analysis and systematic review, Journal of Antimicrobial Chemotherapy, 66, 968-78, 2011	Protocol 3. Comparison not relevant, and clarithromycin is an intervention of interest for chronic PA.
Calabrese, A. T., Pasko, M. T., A review of the clinical efficacy of meropenem, Journal of Infectious Disease Pharmacotherapy, 3, 9-43, 1998	Intervention. This review includes one trial with people with CF and Pseudomonas infection. The treatment (Meroponem) is not relevant for protocol 3.
Carr,R.R., Nahata,M.C., Azithromycin for improving pulmonary function in cystic fibrosis, Annals of Pharmacotherapy, 38, 1520-1524, 2004	Protocol 3. Low quality SR.
Church, D. A., Kanga, J. F., Kuhn, R. J., Rubio, T. T., Spohn, W. A., Stevens, J. C., Painter, B. G., Thurberg, B. E., Haverstock, D. C., Perroncel, R. Y., Echols, R. M., Sequential ciprofloxacin therapy in pediatric cystic fibrosis: comparative study vs. ceftazidime/tobramycin in the treatment of acute pulmonary exacerbations. The Cystic Fibrosis Study Group, Pediatric Infectious Disease Journal, 16, 97-105; discussion 123-6, 1997	This study was included in Cochrane SR Hurley 2015, but no data was used for this review.(protocol 2)
Clancy, J. P., Dupont, L., Konstan, M. W., Billings, J., Fustik, S., Goss, C. H., Lymp, J., Minic, P., Quittner, A. L., Rubenstein, R. C., Young, K. R., Saiman, L., Burns, J. L., Govan, J. R., Ramsey, B., Gupta, R., Arikace Study, Group, Phase II studies of nebulised Arikace in CF patients with Pseudomonas aeruginosa infection, Thorax, 68, 818-25, 2013	Not relevant intervention: amikacin (protocol 3).
Clark, D., Armstrong, J., Young, A., Stirling, R., Wilson, J., Spelman, D., et al.,, An investigator driven, single-centre randomised, double blind comparative study of standard vs high dose intravenous meropenem, in adult cystic fibrosis patients [Abstract], European Respiratory Society Annual Congress, Barcelona, Spain, September, 4796	Excluded from protocol 2 because the reported comparison is of antibiotic dose only.
Clement, A., Tamalet, A., Leroux, E., Ravilly, S., Fauroux, B., Jais, J. P., Long term effects of azithromycin in patients with cystic fibrosis: A double blind, placebo controlled trial, Thorax, 61, 895-902, 2006	Excluded from protocol 3. Population in the P. aeruginosa group <20
Connett, G. J., Pike, K. C., Legg, J. P., Cathie, K., Dewar, A., Foote, K., Harris, A., Faust, S. N., Ciprofloxacin during upper respiratory tract infections to reduce Pseudomonas aeruginosa infection in paediatric cystic fibrosis: A pilot study, Therapeutic Advances in Respiratory Disease, 9, 272-280, 2015	Protocol 2. Not a relevant comparison (placebo)
De Boeck, K., Sauer, K., Vandeputte, S., Meropenem versus ceftazidime plus tobramycin	No numerical data.

Study	Reason for Exclusion
for pulmonary disease in CF patients [abstract], Netherlands journal of medicine, 54, S39, 1999	
Dorkin, H. L., Staab, D., Operschall, E., Alder, J., Criollo, M., Ciprofloxacin DPI: a randomised, placebo-controlled, phase IIb efficacy and safety study on cystic fibrosis, BMJ open respiratory researchBMJ Open Respir Res, 2, e000100, 2015	Not relevant intervention: ciprofloxacin (protocol 3).
Elborn, J. S., Prescott, R. J., Stack, B. H., Goodchild, M. C., Bates, J., Pantin, C., Ali, N., Shale, D. J., Crane, M., Elective versus symptomatic antibiotic treatment in cystic fibrosis patients with chronic Pseudomonas infection of the lungs, Thorax, 55, 355-8, 2000	Protocol 3. Not a relevant comparison.
Elphick, H. E., Jahnke, N., Single versus combination intravenous antibiotic therapy for people with cystic fibrosis, Cochrane Database of Systematic Reviews, 4, CD002007, 2014	Update of this review was assessed for inclusion.
Elphick, H. E., Scott, A., Single versus combination intravenous anti-presudomonal antibiotic therapy for people with cystic fibrosis, Cochrane Database of Systematic Reviews, 2016, no pagination, 2016	Protocol 2 This SR met the inclusion criteria, but it was finally excluded as all relevant trials had already been included in the review. Protocol 3. This SR includes one trial with people with CF and chronic infection with P. Aeruginosa, but the treatment is not relevant to protocol 3.
Equi,A., Balfour-Lynn,I.M., Bush,A., Rosenthal,M., Long term azithromycin in children with cystic fibrosis: A randomised, placebo-controlled crossover trial, Lancet, 360, 978-984, 2002	Protocol 3. Dose of azithromycin classified as immunomodulatory.
Florescu, D. F., Murphy, P. J., Kalil, A. C., Effects of prolonged use of azithromycin in patients with cystic fibrosis: a meta-analysis, Pulmonary Pharmacology & Therapeutics, 22, 467-72, 2009	Protocol 3. Low quality SR.
Fusco, N. M., Toussaint, K. A., Prescott, W. A., Jr., Antibiotic management of methicillin-resistant Staphylococcus aureus--associated acute pulmonary exacerbations in cystic fibrosis, Annals of Pharmacotherapy, 49, 458-68, 2015	Excluded from protocol 2 because this is a narrative review.
Gibson, R. L., Emerson, J., McNamara, S., Burns, J. L., Rosenfeld, M., Yunker, A., Hamblett, N., Accurso, F., Dovey, M., Hiatt, P., Konstan, M. W., Moss, R., Retsch-Bogart, G., Wagener, J., Waltz, D., Wilmott, R., Zeitlin, P. L., Ramsey, B., Cystic Fibrosis Therapeutics Development Network Study, Group, Significant microbiological effect of inhaled tobramycin in young children with cystic fibrosis, American Journal of Respiratory & Critical Care Medicine, 167, 841-9, 2003	Excluded from protocol 2 as the reported comparison to placebo is not relevant.
Gold, R., Carpenter, S., Heurter, H., Corey, M., Levison, H., Randomized trial of ceftazidime versus placebo in the management of acute respiratory exacerbations in patients with cystic fibrosis, Journal of Pediatrics, 111, 907-13, 1987	Protocol 2. N < 30
Goldbeck,Lutz, Fidika,Astrid, Herle,Marion, Quittner,Alexandra L., Psychological	Aim not relevant to any of the protocols.

Study	Reason for Exclusion
interventions for individuals with cystic fibrosis and their families, Cochrane Database of Systematic Reviews, -, 2014	
Harrison, C. J., Marks, M. I., Welch, D. F., Sharma, B. B., Baker, D., Dice, J., A multicenter comparison of related pharmacologic features of cephalexin and dicloxacillin given for two months to young children with cystic fibrosis, Pediatric Pharmacology, 5, 7-16, 1985	Intervention: this study compares Cephalexin and Dicloxacillin. Dicloxacillin is not a treatment of interest in protocol 1.
Horsley, A., Jones, A. M., Antibiotic treatment for Burkholderia cepacia complex in people with cystic fibrosis experiencing a pulmonary exacerbation, Cochrane Database of Systematic Reviews, 10, CD009529, 2012	An update is available for this review.
Huang, N. N., Palmer, J., Braverman, S., Keith, H. H., Schidlow, D., Therapeutic efficacy of ticarcillin and carbenicillin in patients with cystic fibrosis: a double blind study [abstract], 23rd Annual Meeting Cystic Fibrosis Club Abstracts, 14, 1982	This study was included in Cochrane SR Hurley 2015, but no data was used for this review.(protocol 2)
Hubert, D., Le Roux, E., Lavrut, T., Wallaert, B., Scheid, P., Manach, D., Grenet, D., Sermet-Gaudelus, I., Ramel, S., Cracowski, C., Sardet, A., Wizla, N., Deneuville, E., Garraffo, R., Continuous versus intermittent infusions of ceftazidime for treating exacerbation of cystic fibrosis, Antimicrobial Agents & Chemotherapy, 53, 3650-6, 2009	Excluded from protocol 2 because the reported comparison is of antibiotic dose only.
Hurley, M. N., McKeever, T., Lo, D., Rosenfeld, M., Goss, C. H., Fogarty, A., Smyth, A. R., Is flucloxacillin antibiotic prophylaxis associated with differential rates of infection with staphylococcus aureus or pseudomonas aeruginosa in young children?, Pediatric Pulmonology, 47, 319, 2012	Conference abstract, longitudinal study
Jennings, M. T., Boyle, M. P., Weaver, D., Callahan, K. A., Dasenbrook, E. C., Eradication strategy for persistent methicillin-resistant Staphylococcus aureus infection in individuals with cystic fibrosis--the PMEP trial: study protocol for a randomized controlled trial, Trials [Electronic Resource], 15, 223, 2014	This is a protocol of an RCT. (Protocol 3)
Knowles, N. E., Hallberg, T. K., Colombe, J. L., Efficacy of aerosolized antibiotics combined with intravenous treatment of cystic fibrosis pulmonary exacerbation, Pediatric pulmonology, 2, 120-1, 1988	N=10
Kun, P., Landau, L. I., Phelan, P. D., Nebulized gentamicin in children and adolescents with cystic fibrosis, Australian Paediatric Journal, 20, 43-5, 1984	Intervention not in protocol for P aeruginosa
Latzin,P., Fehling,M., Bauernfeind,A., Reinhardt,D., Kappler,M., Griese,M., Efficacy and safety of intravenous meropenem and tobramycin versus ceftazidime and tobramycin in cystic fibrosis, Journal of Cystic Fibrosis, 7, 142-146, 2008	Not a relevant intervention (protocol 3).

Study	Reason for Exclusion
Lenoir, G., Sommelet Olive, D., Derelle, J., Chazalette, J. P., Jehanne, M., Douchain, F., Loeuil, M., Ounis, I., A double-blind comparison of RU-41740 ('Biostim') and placebo in recurrent upper respiratory tract infections in children with cystic fibrosis, Drug Investigation, 3, 76-81, 1991	Treatment. This study aims to evaluate the prophylactic action of Biostim. This is not a treatment of interest for protocol 1.
Littlewood, K. J., Higashi, K., Jansen, J. P., Capkun-Niggli, G., Balp, M. M., Doering, G., Tiddens, H. A., Angyalosi, G., A network meta-analysis of the efficacy of inhaled antibiotics for chronic Pseudomonas infections in cystic fibrosis, Journal of Cystic Fibrosis, 11, 419-26, 2012	This SR compared the efficacy of different inhaled antibiotics for the treatment of patients with CF chronically infected with P. Aeruginosa (protocol 3). It was assessed as low quality according to AMSTAR.
Lo, K. H. David, Hurley, Matthew N., Muhlebach, Marianne S., Smyth, Alan R., Interventions for the eradication of meticillin-resistant Staphylococcus aureus (MRSA) in people with cystic fibrosis, Cochrane Database of Systematic Reviews, 2015	Duplicate. Already included in the review.
Loening-Baucke, V. A., Mischler, E., Myers, M. G., A placebo-controlled trial of cephalexin therapy in the ambulatory management of patients with cystic fibrosis, Journal of Pediatrics, 95, 630-7, 1979	Study design: cross-over (not suitable for protocol 1)
MacLusky,I.B., Gold,R., Corey,M., Levison,H., Long-term effects of inhaled tobramycin in patients with cystic fibrosis colonized with Pseudomonas aeruginosa, Pediatric Pulmonology, 7, 42-48, 1989	Small sample size (N < 30) (protocol 3).
Mahdavinia, M., Grammer, L. C., Management of allergic bronchopulmonary aspergillosis: a review and update, Therapeutic Advances in Respiratory Disease, 6, 173-87, 2012	Population: this review (systematic?) included 2 trials with non-CF patients.
Maiz, L., Giron, R. M., Olveira, C., Quintana, E., Lamas, A., Pastor, D., Canton, R., Mensa, J., Inhaled antibiotics for the treatment of chronic bronchopulmonary Pseudomonas aeruginosa infection in cystic fibrosis: systematic review of randomised controlled trials, Expert Opinion on Pharmacotherapy, 14, 1135-49, 2013	This review evaluated the use of inhaled antibiotics for the treatment of patients with CF chronically infected with P. Aeruginosa (protocol 3). It was assessed as low quality according to AMSTAR.
Marchetti, F., Giglio, L., Candusso, M., Faraguna, D., Assael, B. M., Early antibiotic treatment of pseudomonas aeruginosa colonisation in cystic fibrosis: a critical review of the literature, European Journal of Clinical Pharmacology, 60, 67-74, 2004	Systematic review excluded from protocol 2 after checking for any relevant studies.
Mastella, G., Agostini, M., Barlocco, G., Bonomi, U., Borgo, G., Bozzino, L., Cabrini, G., Cappelletti, L. M., Castellani, L., Conforti, M., et al.,, Alternative antibiotics for the treatment of Pseudomonas infections in cystic fibrosis, Journal of Antimicrobial Chemotherapy, 12 Suppl A, 297-311, 1983	Excluded from protocol 2 because the reported outcomes are not relevant to the protocol.
McCaffery, K., Olver, R. E., Franklin, M., Mukhopadhyay, S., Systematic review of antistaphylococcal antibiotic therapy in cystic fibrosis, Thorax, 54, 380-3, 1999	Excluded as the quality was assessed as low according to AMSTAR checklist (score of 5 out of 11). Included studies were checked for relevance.

Study	Reason for Exclusion
Moreira, A. S., Silva, D., Ferreira, A. R., Delgado, L., Antifungal treatment in allergic bronchopulmonary aspergillosis with and without cystic fibrosis: A systematic review, Clinical and Experimental Allergy, 44, 1210-1227, 2014	Population. This SR includes 4 RCTs with non-CF patients. Not relevant for protocol 3.
Moss, R. B., Critique of trials in allergic bronchopulmonary aspergillosis and fungal allergy, Medical Mycology, 44, 269-272, 2006	Discussion paper. Looks at trials on ABPA (protocol 3).
Moss, R. B., Long-term benefits of inhaled tobramycin in adolescent patients with cystic fibrosis, Chest, 121, 55-63, 2002	Follow up too long to be included in NMA (protocol 3)
Mukhopadhyay, S., Singh, M., Cater, J. I., Ogston, S., Franklin, M., Olver, R. E., Nebulised antipseudomonal antibiotic therapy in cystic fibrosis: a meta-analysis of benefits and risks, Thorax, 51, 364-8, 1996	Ths SR looked at the benefits and risks of nebulised antipseudomonal therapy in CF (protocol 3). It was assessed as low quality according to AMSTAR.
National Horizon Scanning Centre, Tobramycin inhaled powder (TIP) for Pseudomonas aeruginosa infection in cystic fibrosis (Structured abstract), Health Technology Assessment Database, 2015	The studies referenced in this report were checked for inclusion.(protocol 3)
National Horizon Scanning Centre, Colistimethate sodium powder for inhalation (Colobreathe) for pseudomonas aeruginosa lung infection in cystic fibrosis (Structured abstract), Health Technology Assessment Database, 2015	Protocol 3. Lab executive summary. No data.
Nolan,G., Moivor,P., Levison,H., Fleming,P.C., Corey,M., Gold,R., Antibiotic prophylaxis in cystic fibrosis: inhaled cephaloridine as an adjunct to oral cloxacillin, Journal of Pediatrics, 101, 626-630, 1982	Treatment. This paper evaluates the prophylactic effect of inhaled cephaloriding as an adjunct to oral cloxacillin. The second is not a treatment of interest for protocol 1.
Padoan,R., Cambisano,W., Costantini,D., Crossignani,R.M., Danza,M.L., Trezzi,G., Giunta,A., Ceftazidime monotherapy vs. combined therapy in Pseudomonas pulmonary infections in cystic fibrosis, Pediatric Infectious Disease Journal, 6, 648-653, 1987	Excluded from protocol 2 because the reported comparison is of antibiotics that are not relevant to the protocol.
Pedersen, S. S., Pressler, T., Pedersen, M., Immediate and prolonged clinical efficacy of ceftazidime versus ceftazidime plus tobramycin in chronic Pseudomonas aeruginosa infection in cystic fibrosis, Scandinavian Journal of Infectious Diseases, 18, 133-137, 1986	Intervention: this study evaluates ceftazidime in the treatment of chronic P. Aeruginosa. This intervention is not relevant for protocol 3.
Permin, H., Koch, C., Hoiby, N., Christensen, H. O., Moller, A. F., Moller, S., Ceftazidime treatment of chronic Pseudomonas aeruginosa respiratory tract infection in cystic fibrosis, Journal of Antimicrobial Chemotherapy, 12 Suppl A, 313-23, 1983	Intervention: this study evaluates ceftazidime in the treatment of chronic P. Aeruginosa. This intervention is not relevant for protocol 3.
Proesmans, M., Boulanger, L., Vermeulen, F., De Boeck, K., Eradication of recent Pseudomonas aeruginosa infection: TOBI versus Colistineb/ciprofloxacin, Journal of Cystic Fibrosis, 10, S26, 2011	Conference abstract (protocol 2).

Study	Reason for Exclusion
Proesmans, M., Vermeulen, F., Vreys, M., Dupont, L. J., De Boeck, K., Treatment of allergic bronchopulmonary aspergillosis in cystic fibrosis: antifungal therapy with inhaled amphotericin B [abstract], Pediatric Pulmonology, 44, 2009	Study design: this abstract looked at antifungal therapy with amphotericin (protocol 3), but it's not an RCT.
Quittner, A. L., Buu, A., Effects of tobramycin solution for inhalation on global ratings of quality of life in patients with cystic fibrosis and Pseudomonas aeruginosa infection, Pediatric Pulmonology, 33, 269-76, 2002	Retrospective analysis of an RCT.
Regan, K. H., Bhatt, J., Eradication therapy for Burkholderia cepacia complex in people with cystic fibrosis, Cochrane Database of Systematic Reviews, 10, CD009876, 2014	An update is available for this review.
Remmington, T., Jahnke, N., Harkensee, C., Oral anti-pseudomonal antibiotics for cystic fibrosis, Cochrane Database of Systematic Reviews, 10, CD005405, 2013	Update of this review was assessed for inclusion
Retsch-Bogart, G. Z., Burns, J. L., Otto, K. L., Liou, T. G., McCoy, K., Oermann, C., Gibson, R. L., Azli Phase II Study Group, A phase 2 study of aztreonam lysine for inhalation to treat patients with cystic fibrosis and Pseudomonas aeruginosa infection, Pediatric Pulmonology, 43, 47-58, 2008	Later study included (protocol 3)
Saiman, L., Anstead, M., Mayer-Hamblett, N., Lands, L. C., Kloster, M., Hocevar-Trnka, J., Goss, C. H., Rose, L. M., Burns, J. L., Marshall, B. C., Ratjen, F., A. Z. Azithromycin Study Group, Effect of azithromycin on pulmonary function in patients with cystic fibrosis uninfected with Pseudomonas aeruginosa: a randomized controlled trial, JAMA, 303, 1707-15, 2010	Excluded from protocol 3. Evaluates Azitrhomaycin in people not infected with PA.
Schaad, U. B., Wedgwood, J., Ruedeberg, A., Kraemer, R., Hampel, B., Ciprofloxacin as antipseudomonal treatment in patients with cystic fibrosis, Pediatric Infectious Disease Journal, 16, 106-11; discussion 123-6, 1997	Ciprofloxacin received by both groups, abstract (protocol 2).
Southern, K. W., Barker, P. M., Solis-Moya, A., Patel, L., Macrolide antibiotics for cystic fibrosis, Cochrane Database of Systematic Reviews, 11, CD002203, 2012	Protocol 3. Cochrane review. All studies include low-dose Azytrhomycin.
Stead, R. J., Hodson, M. E., Batten, J. C., Inhaled ceftazidime compared with gentamicin and carbenicillin in older patients with cystic fibrosis infected with Pseudomonas aeruginosa, British Journal of Diseases of the Chest, 81, 272-9, 1987	Sample size too small (N<20) for inclusion in NMA; Interventions used not in protocol for P aeruginosa
Steinkamp,G., Schmitt-Grohe,S., Doring,G., Staab,D., Pfrunder,D., Beck,G., Schubert,R., Zielen,S., Once-weekly azithromycin in cystic fibrosis with chronic Pseudomonas aeruginosa infection, Respiratory Medicine, 102, 1643-1653, 2008	This study was excluded as it used azithromysin as immuno (low-does) (protocol 3).

Study	Reason for Exclusion
Stephens, D., Garey, N., Isles, A., Levison, H., Gold, R., Efficacy of inhaled tobramycin in the treatment of pulmonary exacerbations in children with cystic fibrosis, Pediatric Infectious Disease, 2, 209-11, 1983	This study was included in Cochrane SR Hurley 2015, but no data was used for this review (protocol 2).
Szaff, M., Hoiby, N., Antibiotic treatment of Staphylococcus aureus infection in cystic fibrosis, Acta Paediatrica Scandinavica, 71, 821-6, 1982	Study design: non-comparative observational study
Treggiari, M. M., Retsch-Bogart, G., Mayer-Hamblett, N., Khan, U., Kulich, M., Kronmal, R., Williams, J., Hiatt, P., Gibson, R. L., Spencer, T., Orenstein, D., Chatfield, B. A., Froh, D. K., Burns, J. L., Rosenfeld, M., Ramsey, B. W., Early Pseudomonas Infection Control, Investigators, Comparative efficacy and safety of 4 randomized regimens to treat early Pseudomonas aeruginosa infection in children with cystic fibrosis, Archives of Pediatrics & Adolescent Medicine, 165, 847-56, 2011	Excluded from protocol 2 because the reported treatment comparison (cycled antibiotics vs culture based antibiotics)is not relevant to the protocol.
Uttley, L., Harnan, S., Cantrell, A., Taylor, C., Walshaw, M., Brownlee, K., Tappenden, P., Systematic review of the dry powder inhalers colistimethate sodium and tobramycin in cystic fibrosis, European Respiratory Review, 22, 476-86, 2013	This SR is the same as the HTA report (Tappendem 2013), already included in protocol 3.
Wang, C. I., Inderlied, C. B., Armer, C., Roldan, M. A., Osher, A. B., Comparison of the efficacy and safety of oral ciprofloxacin with that of i.v. tobrarmycin plus azlocillin and/or tobramycin plus ticarcillin in patients with cystic fibrosis [abstract], Excerpta Medica, Asia Pacific Congress Series, 74, 1988	Conference abstract.
Wark, P., Pathogenesis of allergic bronchopulmonary aspergillosis and an evidence-based review of azoles in treatment, Respiratory Medicine, 98, 915-23, 2004	Population: this review looked at the management of ABPA (protocol 3), but includes 2 RCTS with non-CF patients.
Wientzen, R., Prestidge, C. B., Kramer, R. I., McCracken, G. H., Nelson, J. D., Acute pulmonary exacerbations in cystic fibrosis. A double-blind trial of tobramycin and placebo therapy, American Journal of Diseases of Children, 134, 1134-8, 1980	Protocol 2. N < 30
Wiesemann, H. G., Steinkamp, G., Ratjen, F., Bauernfeind, A., Przyklenk, B., Doring, G., von der Hardt, H., Placebo-controlled, double-blind, randomized study of aerosolized tobramycin for early treatment of Pseudomonas aeruginosa colonization in cystic fibrosis, Pediatric Pulmonology, 25, 88-92, 1998	This study looks at prophylaxis for P. Aeruginosa (not relevant to protocol 1).
Wolter, J., Seeney, S., Bell, S., Bowler, S., Masel, P., McCormack, J., Effect of long term treatment with azithromycin on disease parameters in cystic fibrosis: a randomised trial, Thorax, 57, 212-6, 2002	Protocol 3. Excluded (low-dose azithromycin)
Wood, D. M., Smyth, A. R., Antibiotic strategies for eradicating Pseudomonas aeruginosa in	Cochrane extracts

Study	Reason for Exclusion
people with cystic fibrosis, Cochrane Database of Systematic Reviews, CD004197, 2006	
Xu, X., Abdalla, T., Sabbatini, G., Roberts, T., Bratcher, P., Jackson, P. L., Blalock, J. E., Clancy, J. P., Gaggar, A., A randomized double-blinded placebo controlled trial of doxycycline as an adjunctive therapy for the treatment of inpatient cystic fibrosis exacerbation, Pediatric Pulmonology, 49, 312, 2014	Conference abstract

H.10 Immunomodulatory agents

Study	Reason for Exclusion
Abdulhamid, I., Beck, F. W. J., Miliard, S., Chen, X., Prasad, A., Effect of zinc supplementation on respiratory tract infections in children with cystic fibrosis, Pediatric Pulmonology, 43, 281-287, 2008	Not relevant intervention (zinc supplemenation).
Balfour-Lynn, I. M., Welch, K., Inhaled corticosteroids for cystic fibrosis, Cochrane Database of Systematic Reviews, 11, CD001915, 2012	An update of this Cochrane SR was included in the review.
Bisgaard,H., Pedersen,S.S., Nielsen,K.G., Skov,M., Laursen,E.M., Kronborg,G., Reimert,C.M., Hoiby,N., Koch,C., Controlled trial of inhaled budesonide in patients with cystic fibrosis and chronic bronchopulmonary Psuedomonas aeruginosa infection, American Journal of Respiratory and Critical Care Medicine, 156, 1190-1196, 1997	Study not relevant for the NMA, as participants were on previous IV treatment. No outcomes of interest for review protocol.
Bowler, S. D., Effect of long-term treatment with azithromycin on disease parameters in cystic fibrosis, Japanese Journal of Antibiotics, 56 Suppl A, 38, 2003	This paper reports the same data as Wolter 2002, already included in the review.
Cheng, K., Ashby, D., Smyth, R., Oral steroids for cystic fibrosis, Cochrane Database of Systematic Reviews, CD000407, 2000	An update of this Cochrane SR was included in the review.
Cheng, K., Ashby, D., Smyth, R. L., Oral steroids for long-term use in cystic fibrosis, Cochrane Database of Systematic Reviews, 6, CD000407, 2013	An update of this Cochrane SR was included in the review.
Conway, S. P., Etherington, C., Peckham, D. G., Whitehead, A., A pilot study of zafirlukast as an anti-inflammatory agent in the treatment of adults with cystic fibrosis, Journal of Cystic Fibrosis, 2, 25-28, 2003	No NMA outcomes of interest. Treatment not included in standard review
Dauletbaev, N., Viel, K., Behr, J., Loitsch, S., Buhl, R., Wagner, T. O., Bargon, J., Effects of short-term inhaled fluticasone on oxidative burst of sputum cells in cystic fibrosis patients, European Respiratory Journal, 14, 1150-5, 1999	Study duration too short for NMA. No outcomes of interest for review protocol.
Dezateux, C., Walters, S., Balfour-Lynn, I., Inhaled corticosteroids for cystic fibrosis, Cochrane Database of Systematic Reviews, CD001915, 2000	An update is available for this review.

Study	Reason for Exclusion
Dezateux,C., Crighton,A., Oral non-steroidal anti-inflammatory drug therapy for cystic fibrosis, Cochrane database of systematic reviews (Online), 2000. Date of Publication, -, 2000	An update is available for this review.
Dovey, M., Aitken, M. L., Emerson, J., McNamara, S., Waltz, D. A., Gibson, R. L., Oral corticosteroid therapy in cystic fibrosis patients hospitalized for pulmonary exacerbation: a pilot study, Chest, 132, 1212-8, 2007	Patients not clinically stable.
Equi, A., Bush, A., Balfour-Lynn, I. M., Rosenthal, M., A prospective, double-blind, randomised, placebo controlled crossover trial of azithromycin in paediatric cystic fibrosis, Thorax, 57, iii38, 2002	Conference abstract.
Eubanks, V., Koppersmith, N., Wooldridge, N., Clancy, J. P., Lyrene, R., Arani, R. B., Lee, J., Moldawer, L., Atchison, J., Sorscher, E. J., Makris, C. M., Effects of megestrol acetate on weight gain, body composition, and pulmonary function in patients with cystic fibrosis, Journal of Pediatrics, 140, 439-44, 2002	Small sample size (N=17).
Florescu, D. F., Murphy, P. J., Kalil, A. C., Effects of prolonged use of azithromycin in patients with cystic fibrosis: a meta-analysis, Pulmonary Pharmacology & Therapeutics, 22, 467-72, 2009	Systematic review with MA: all individual studies have already been included in the review.
Jat, Kana R., Walia, Dinesh K., Khairwa, Anju, Anti-IgE therapy for allergic bronchopulmonary aspergillosis in people with cystic fibrosis, Cochrane database of Systematic Reviews, 2015	One study was included in this Cochrane review. The adverse events reported in the included study were not listed in the current review protocol.
Kabra,S.K., Pawaiya,R., Lodha,R., Kapil,A., Kabra,M., Vani,A.S., Agarwal,G., Shastri,S.S., Long-term daily high and low doses of azithromycin in children with cystic fibrosis: A randomized controlled trial, Journal of Cystic Fibrosis, 9, 17-23, 2010	Study compares multiple doses of same treatment.
Konstan, M. W., Hoppel, C. L., Chai, B. L., Davis, P. B., Ibuprofen in children with cystic fibrosis: pharmacokinetics and adverse effects, Journal of Pediatrics, 118, 956-64, 1991	Small sample size (N < 20).
Lands, L. C., Stanojevic, S., Oral non-steroidal anti-inflammatory drug therapy for lung disease in cystic fibrosis, Cochrane Database of Systematic Reviews, 6, CD001505, 2013	An update of this Cochrane SR was included in the review.
Linnane, S. J., Thin, A. G., Keatings, V. M., Moynihan, J. B., McLoughlin, P., FitzGerald, M. X., Glucocorticoid treatment reduces exhaled nitric oxide in cystic fibrosis patients, European Respiratory Journal, 17, 1267-70, 2001	Small sample size (N < 20).
McCormack, J., Bell, S., Senini, S., Walmsley, K., Patel, K., Wainwright, C., Serisier, D., Harris, M., Bowler, S., Daily versus weekly azithromycin in cystic fibrosis patients, European Respiratory Journal, 30, 487-95, 2007	Dose of azithromycin too high for inclusion.

Study	Reason for Exclusion
Moss, R. B., Mayer-Hamblett, N., Wagener, J., Daines, C., Hale, K., Ahrens, R., Gibson, R. L., Anderson, P., Retsch-Bogart, G., Nasr, S. Z., Noth, I., Waltz, D., Zeitlin, P., Ramsey, B., Starko, K., Randomized, double-blind, placebo-controlled, dose-escalating study of aerosolized interferon gamma-1b in patients with mild to moderate cystic fibrosis lung disease, Pediatric Pulmonology, 39, 209-18, 2005	No NMA outcomes of interest. Treatment not included in review protocol.
Nikolaizik,W.H., Schoni,M.H., Pilot study to assess the effect of inhaled corticosteroids on lung function in patients with cystic fibrosis, Journal of Pediatrics, 128, 271-274, 1996	Patients not clinically stable.
Pantin, C. F., Stead, R. J., Hodson, M. E., Batten, J. C., Prednisolone in the treatment of airflow obstruction in adults with cystic fibrosis, Thorax, 41, 34-8, 1986	Non-randomised study (order of treatments is same).
Pukhalsky, A. L., Shmarina, G. V., Kapranov, N. I., Kokarovtseva, S. N., Pukhalskaya, D., Kashirskaja, N. J., Anti-inflammatory and immunomodulating effects of clarithromycin in patients with cystic fibrosis lung disease, Mediators of Inflammation, 13, 111-7, 2004	Non-randomised study (only comparison between before and after treatment).
Ratjen, F., Saiman, L., Mayer-Hamblett, N., Lands, L. C., Kloster, M., Emmett, P., Goss, C. H., Rose, L., Burns, J. L., Marshall, B., Accurso, F. J., Anstead, M., The effect of azithromcyin on inflammatory markers in CF children and adolescents uninfected with pseudomonas aeruginosa, Pediatric Pulmonology, 45, 271, 2010	Conference abstract.
Ratjen, F., Saiman, L., Mayer-Hamblett, N., Lands, L. C., Kloster, M., Thompson, V., Emmett, P., Marshall, B., Accurso, F., Sagel, S., Anstead, M., Effect of azithromycin on systemic markers of inflammation in patients with cystic fibrosis uninfected with Pseudomonas aeruginosa, Chest, 142, 1259-66, 2012	This study is a follow-up of a previous study, Saiman 2010 included in Cochrane SR Southern 2012. No outcomes relevant to the protocol reporte.
Rosenstein, B. J., Eigen, H., Risks of alternate-day prednisone in patients with cystic fibrosis, Pediatrics, 87, 245-6, 1991	Additional data for Eigen 2005. Full study already included in the review (as part of Cochrane SR Cheng 2015).
Saiman, L., Anstead, M., Mayer-Hamblett, N., Lands, L., Kloster, M., Goss, C. H., Rose, L. M., Burns, J. L., Marshall, B. C., Ratjen, F., Open-label, follow-on study to assess continued efficacy & safety of azithromcyin in children & adolescents with CF uninfected with Pseudomonas Aeruginosa, Pediatric Pulmonology, 45, 328, 2010	Conference abstract.
Saiman, L., Mayer-Hamblett, N., Anstead, M., Lands, L. C., Kloster, M., Goss, C. H., Rose, L. M., Burns, J. L., Marshall, B. C., Ratjen, F., A. Z. Macrolide Study Team, Open-label, follow-on study of azithromycin in pediatric patients with CF uninfected with Pseudomonas aeruginosa, Pediatric Pulmonology, 47, 641-8, 2012	Not a randomised study.

Study	Reason for Exclusion
Saiman, L., Mayer-Hamblett, N., Campbell, P., Marshall, B. C., Macrolide Study, Group, Heterogeneity of treatment response to azithromycin in patients with cystic fibrosis, American Journal of Respiratory & Critical Care Medicine, 172, 1008-12, 2005	This study is a follow-up of a previous study, Saiman 2010 included in Cochrane SR Southern 2012. No outcomes relevant to the protocol reported.
Schmitt-Grohe, S., Eickmeier, O., Schubert, R., Bez, C., Zielen, S., Anti-inflammatory effects of montelukast in mild cystic fibrosis, Annals of Allergy, Asthma, & Immunology, 89, 599-605, 2002	No outcomes of interest.
Senini, S., McCormack, J., A randomised, prospective double-blind trial of long-term daily versus weekly azithromycin in cystic fibrosis, Australian New Zealand Clinical Trials Registry, 04, 2010	Comparison not relevant to the protocol.
Sivan, Y., Arce, P., Eigen, H., Nickerson, B. G., Newth, C. J. L., A double-blind, randomized study of sodium cromoglycate versus placebo in patients with cystic fibrosis and bronchial hyperreactivity, Journal of allergy and clinical immunology, 85, 649-654, 1990	Intervention not included in review.
Skov, M., Pressler, T., Lykkesfeldt, J., Poulsen, H. E., Jensen, P. T., Johansen, H. K., Qvist, T., Kraemer, D., Hoiby, N., Ciofu, O., The effect of short-term, high-dose oral N-acetylcysteine treatment on oxidative stress markers in cystic fibrosis patients with chronic P. aeruginosa infection - A pilot study, Journal of Cystic Fibrosis, 14, 211-218, 2015	Intervention not included in review.
Steinkamp,G., Schmitt-Grohe,S., Doring,G., Staab,D., Pfrunder,D., Beck,G., Schubert,R., Zielen,S., Once-weekly azithromycin in cystic fibrosis with chronic Pseudomonas aeruginosa infection, Respiratory Medicine, 102, 1643-1653, 2008	Patients not clinically stable.
Stelmach,I., Korzeniewska,A., Stelmach,W., Majak,P., Grzelewski,T., Jerzynska,J., Effects of montelukast treatment on clinical and inflammatory variables in patients with cystic fibrosis, Annals of Allergy, Asthma and Immunology, 95, 372-380, 2005	Outcomes of interest only reported graphically.
Taylor,C.J., McGaw,J., Rigby,A.S., Threlfall,D., Karmel,J., Pilot safety study of liposomal prostaglandin (PGE1) in respiratory exacerbations in cystic fibrosis, Journal of Cystic Fibrosis, 1, 90-93, 2002	Intervention not included in review.
Tepper,R.S., Eigen,H., Stevens,J., Angelicchio,C., Kisling,J., Ambrosius,W., Heilman,D., Lower respiratory illness in infants and young children with cystic fibrosis: Evaluation of treatment with intravenous hydrocortisone, Pediatric Pulmonology, 24, 48-51, 1997	Treatment (hydrocortisone) not in protocol.
Tirouvanziam, R., Lymp, J., Thompson, V., Chatfield, B. A., Nicholls, D., Clancy, J. P., Vender, R., Egan, M., Quittell, L., Michelson, P.,	Conference abstract.

Study	Reason for Exclusion
Antony, V., Spahr, J., Rubenstein, R. C., Herzenberg, L., Conrad, C. K., A multi-center, phase iib, randomized, placebo-controlled, double-blind study of the effects of N-acetylcysteine (NAC) on redox changes and lung inflammation in cystic fibrosis patients, Pediatric Pulmonology, 46, 280-1, 2011	
van Haren, E. H., Lammers, J. W., Festen, J., Heijerman, H. G., Groot, C. A., van Herwaarden, C. L., The effects of the inhaled corticosteroid budesonide on lung function and bronchial hyperresponsiveness in adult patients with cystic fibrosis, Respiratory Medicine, 89, 209-14, 1995	Incmplete reporting: no variance/uncertainty reported for outcomes of interest.
Xu, X., Abdalla, T., Sabbatini, G., Roberts, T., Bratcher, P., Jackson, P. L., Blalock, J. E., Clancy, J. P., Gaggar, A., A randomized double-blinded placebo controlled trial of doxycycline as an adjunctive therapy for the treatment of inpatient cystic fibrosis exacerbation, Pediatric Pulmonology, 49, 312, 2014	Conference abstract.

H.11 Nutritional interventions

Study	Reason for Exclusion
Alshaikh, B., Schall, J. I., Maqbool, A., Mascarenhas, M., Bennett, M. J., Stallings, V. A., Choline supplementation alters some amino acid concentrations with no change in homocysteine in children with cystic fibrosis and pancreatic insufficiency, Nutrition Research, 36, 418-29, 2016	No relevant intervention
Armand, M., Hamosh, M., Philpott, J. R., Resnik, A. K., Rosenstein, B. J., Hamosh, A., Perman, J. A., Hamosh, P., Gastric function in children with cystic fibrosis: effect of diet on gastric lipase levels and fat digestion, Pediatric research, 55, 457-65, 2004	No relevant outcomes. Cross-over trial.
Baldwin, Christine, Weekes, Elizabeth Christine, Dietary advice with or without oral nutritional supplements for disease-related malnutrition in adults, Cochrane Database of Systematic Reviews, 2012	Relevant included study was included in a Cochrane review specific to CF which has been included
Balzer,B.W., Graham,C.L., Craig,M.E., Selvadurai,H., Donaghue,K.C., Brand-Miller,J.C., Steinbeck,K.S., Low glycaemic index dietary interventions in youth with cystic fibrosis: a systematic review and discussion of the clinical implications, Nutrients, 4, 286-296, 2012	No relevant included studies
Banjar, H., Gastrostomy Tube Feeding of Cystic Fibrosis Patients, Bahrain Medical Bulletin, 26, 15-19, 2004	Retrospective chart review; the comparison between the gastrostomy group and the control group does not focus on change over time.
Bonifant, C. M., Shevill, E., Chang, A. B., Vitamin A supplementation for cystic fibrosis, Cochrane Database of Systematic Reviews, 5, CD006751, 2014	No relevant intervention
Caramia, G., Cocchi, M., Gagliardini, R., Malavolta, M., Mozzon, M., Frega, N. G., Fatty	No relevant intervention

Study	Reason for Exclusion
acids composition of plasma phospholipids and triglycerides in children with cystic fibrosis. The effect of dietary supplementation with an olive and soybean oils mixture, Pediatria Medica e Chirurgica, 25, 42-9, 2003	
Chinuck, R. S., Fortnum, H., Baldwin, D. R., Appetite stimulants in cystic fibrosis: a systematic review, Journal of Human Nutrition & Dietetics, 20, 526-37, 2007	The studies included in this systematic review were assessed for inclusion. The quality of the systematic review was rated as low using the AMSTAR checklist.
Ciofu, O., Lykkesfeldt, J., Antioxidant supplementation for lung disease in cystic fibrosis, Cochrane Database of Systematic Reviews, 8, CD007020, 2014	No relevant intervention
Ellis, L., Kalnins, D., Corey, M., Brennan, J., Pencharz, P., Durie, P., Do infants with cystic fibrosis need a protein hydrolysate formula? A prospective, randomized, comparative study, Journal of Pediatrics, 132, 270-276, 1998	No relevant intervention
Epifanio, M., Marostica, P. C., Mattiello, R., Feix, L., Nejedlo, R., Fischer, G. B., Stein, R. T., A randomized, double-blind, placebo-controlled trial of cyproheptadine for appetite stimulation in cystic fibrosis, Jornal de Pediatria, 88, 155-60, 2012	Portuguese language
Erskine, J. M., Lingard, C. D., Sontag, M. K., Accurso, F. J., Enteral nutrition for patients with cystic fibrosis: comparison of a semi-elemental and nonelemental formula, Journal of Pediatrics, 132, 265-9, 1998	Crossover study design
Ferguson, J. H., Chang, A. B., Vitamin D supplementation for cystic fibrosis, Cochrane Database of Systematic Reviews, 5, CD007298, 2014	No relevant intervention
Forrester, D. L., Knox, A. J., Smyth, A. R., Barr, H. L., Simms, R., Pacey, S. J., Pavord, I. D., Honeybourne, D., Dewar, J., Clayton, A., Fogarty, A. W., Glutamine supplementation in cystic fibrosis: A randomized placebo-controlled trial, Pediatric Pulmonology, 51, 253-7, 2016	No relevant intervention
Francis, D. K., Smith, J., Saljuqi, T., Watling, R. M., Oral protein calorie supplementation for children with chronic disease, Cochrane Database of Systematic Reviews, 5, CD001914, 2015	Included studies on people with CF included in this review were also included in Cochrane reviews specific to people with CF. The latter reviews were assessed for inclusion.
Geukers, V. G., Oudshoorn, J. H., Taminiau, J. A., van der Ent, C. K., Schilte, P., Ruiter, A. F., Ackermans, M. T., Endert, E., Jonkers-Schuitema, C. F., Heymans, H. S., Sauerwein, H. P., Short-term protein intake and stimulation of protein synthesis in stunted children with cystic fibrosis, American Journal of Clinical Nutrition, 81, 605-10, 2005	No relevant intervention. No relevant outcomes.
Goldbeck, L., Fidika, A., Herle, M., Quittner, A. L., Cochrane corner: Psychological interventions for individuals with cystic fibrosis and their families, Thorax, 70, 1089-1091, 2015	This article summarizes a Cochrane systematic review which has been included in the present review

Study	Reason for Exclusion
Grey, V., Mohammed, S. R., Smountas, A. A., Bahlool, R., Lands, L. C., Improved glutathione status in young adult patients with cystic fibrosis supplemented with whey protein.[Erratum appears in J Cyst Fibros. 2004 Mar;3(1):62], Journal of Cystic Fibrosis, 2, 195-8, 2003	No relevant intervention
Groleau, V., Schall, J. I., Dougherty, K. A., Latham, N. E., Maqbool, A., Mascarenhas, M. R., Stallings, V. A., Effect of a dietary intervention on growth and energy expenditure in children with cystic fibrosis, Journal of Cystic Fibrosis, 13, 572-578, 2014	No relevant intervention
Hanssens, L., Thiebaut, I., Lefevre, N., Malfroot, A., Knoop, C., Duchateau, J., Casimir, G., The clinical benefits of long-term supplementation with omega-3 fatty acids in cystic fibrosis patients - A pilot study, Prostaglandins Leukotrienes and Essential Fatty Acids, 108, 45-50, 2016	No relevant intervention
Homnick, D. N., Marks, J. H., Hare, K. L., Bonnema, S. K., Long-term trial of cyproheptadine as an appetite stimulant in cystic fibrosis, Pediatric Pulmonology, 40, 251-6, 2005	Aim not relevant. Follow-up of a study that has been included in the current review.
Homnick, D. N., Marks, J. H., Rubin, B. K., The effect of a first-generation antihistamine on sputum viscoelasticity in cystic fibrosis, Journal of Aerosol Medicine, 20, 45-9, 2007	No relevant outcomes
Hurley, M. N., Forrester, D. L., Smyth, A. R., Antibiotic adjuvant therapy for pulmonary infection in cystic fibrosis, Cochrane Database of Systematic Reviews, 6, CD008037, 2013	No relevant intervention
Jafari, S. A., Mehdizadeh-Hakkak, A., Kianifar, H. R., Hebrani, P., Ahanchian, H., Abbasnejad, E., Effects of probiotics on quality of life in children with cystic fibrosis; A randomized controlled trial, Iranian Journal of Pediatrics, 23, 669-674, 2013	No relevant intervention
Jagannath, Vanitha A., Fedorowicz, Zbys, Thaker, Vidhu, Chang, Anne B., Vitamin K supplementation for cystic fibrosis, Cochrane Database of Systematic Reviews, 2015	No relevant intervention
Janicke, D. M., Mitchell, M. J., Quittner, A. L., Piazza-Waggoner, C., Stark, L. J., The impact of behavioral intervention on family interactions at mealtime in pediatric cystic fibrosis, Children's Health Care, 37, 49-66, 2008	No relevant outcomes
Jelalian, E., Stark, L. J., Reynolds, L., Seifer, R., Nutrition intervention for weight gain in cystic fibrosis: a meta analysis, Journal of Pediatrics, 132, 486-92, 1998	Included studies were assessed for inclusion
Koretz, R. L., Avenell, A., Lipman, T. O., Braunschweig, C. L., Milne, A. C., Does enteral nutrition affect clinical outcome? A systematic review of the randomized trials, American Journal of Gastroenterology, 102, 412-29; quiz 468, 2007	The studies included in the systematic review were assessed for inclusion. The quality of the systematic review was rated as low using the AMSTAR checklist.

Study	Reason for Exclusion
Lepage, G., Yesair, D. W., Ronco, N., Champagne, J., Bureau, N., Chemtob, S., Berube, D., Roy, C. C., Effect of an organized lipid matrix on lipid absorption and clinical outcomes in patients with cystic fibrosis, Journal of Pediatrics, 141, 178-85, 2002	No relevant intervention
Lloyd-Still, J. D., Powers, C. A., Hoffman, D. R., Boyd-Trull, K., Lester, L. A., Benisek, D. C., Arterburn, L. M., Bioavailability and safety of a high dose of docosahexaenoic acid triacylglycerol of algal origin in cystic fibrosis patients: a randomized, controlled study, Nutrition, 22, 36-46, 2006	No relevant intervention
Martin, C. R., Blanco, P. G., Keach, J. C., Petz, J. L., Zaman, M. M., Bhaskar, K. R., Cluette-Brown, J. E., Gautam, S., Sheth, S., Afdhal, N. H., Lindor, K. D., Freedman, S. D., The safety and efficacy of oral docosahexaenoic acid supplementation for the treatment of primary sclerosing cholangitis - a pilot study, Alimentary Pharmacology & Therapeutics, 35, 255-65, 2012	No relevant outcomes
Okebukola, P. O., Kansra, S., Barrett, J., Vitamin E supplementation in people with cystic fibrosis, Cochrane Database of Systematic Reviews, 12, CD009422, 2014	No relevant intervention
Oliver, Colleen, Watson, Helen, Omega-3 fatty acids for cystic fibrosis, Cochrane Database of Systematic Reviews, 2016	No relevant intervention
Opipari-Arrigan, L., Powers, S. W., Quittner, A. L., Stark, L. J., Mealtime problems predict outcome in clinical trial to improve nutrition in children with CF, Pediatric Pulmonology, 45, 78-82, 2010	No relevant data
Oudshoorn, J. H., Klijn, P. H., Hofman, Z., Voorbij, H. A., van der Ent, C. K., Berger, R., Houwen, R. H., Dietary supplementation with multiple micronutrients: no beneficial effects in pediatric cystic fibrosis patients, Journal of Cystic Fibrosis, 6, 35-40, 2007	No relevant intervention
Payne, Cathy, Wiffen, Philip J., Martin, Suzanne, Interventions for fatigue and weight loss in adults with advanced progressive illness, Cochrane Database of Systematic Reviews, 2015	Relevant included systematic reviews have been assessed for inclusion. The update of a relevant review has been included.
Powers, S. W., Jones, J. S., Ferguson, K. S., Piazza-Waggoner, C., Daines, C., Acton, J. D., Randomized clinical trial of behavioral and nutrition treatment to improve energy intake and growth in toddlers and preschoolers with cystic fibrosis, Pediatrics, 116, 1442-50, 2005	No relevant data
Powers, S. W., Piazza-Waggoner, C., Jones, J. S., Ferguson, K. S., Daines, C., Acton, J. D., Examining clinical trial results with single-subject analysis: an example involving behavioral and nutrition treatment for young children with cystic fibrosis, Journal of Pediatric Psychology, 31, 574-81, 2006	No relevant data

Study	Reason for Exclusion
Rust, P., Eichler, I., Renner, S., Elmadfa, I., Long-term oral beta-carotene supplementation in patients with cystic fibrosis - Effects on antioxidative status and pulmonary function, Annals of Nutrition and Metabolism, 44, 30-37, 2000	No relevant intervention
Schall, J. I., Mascarenhas, M. R., Maqbool, A., Dougherty, K. A., Elci, O., Wang, D. J., Altes, T. A., Hommel, K. A., Shaw, W., Moore, J., Stallings, V. A., Choline supplementation with a structured lipid in children with cystic fibrosis: A randomized placebo-controlled trial, Journal of Pediatric Gastroenterology and Nutrition, 62, 618-626, 2016	No relevant outcomes
Schwarzenberg, S. J., Hempstead, S. E., McDonald, C. M., Powers, S. W., Wooldridge, J., Blair, S., Freedman, S., Harrington, E., Murphy, P. J., Palmer, L., Schrader, A. E., Shiel, K., Sullivan, J., Wallentine, M., Marshall, B. C., Leonard, A. R., Enteral tube feeding for individuals with cystic fibrosis: Cystic Fibrosis Foundation evidence-informed guidelines, Journal of Cystic FibrosisJ Cyst Fibros, 15, 724-735, 2016	Guidelines and systematic review; quality of the systematic review was rated as low using the AMSTAR checklist; included studies were checked to identify relevant ones
Sharma, G., Lodha, R., Shastri, S., Saini, S., Kapil, A., Singla, M., Mukherjee, A., Jat, K. R., Kabra, M., Kabra, S. K., Zinc supplementation for one year among children with cystic fibrosis does not decrease pulmonary infection, Respiratory Care, 61, 78-84, 2016	No relevant intervention
Shoff,S.M., Ahn,H.Y., Davis,L., Lai,H., Temporal associations among energy intake, plasma linoleic acid, and growth improvement in response to treatment initiation after diagnosis of cystic fibrosis, Pediatrics, 117, 391-400, 2006	No relevant data
Skypala,, Ashworth,, Hodson,, Leonard,, Knox,, Hiller,, Wolfe,, Littlewood,, Morton,, Conway,, Patchell,, Weller,, McCarthy,, Redmond,, Dodge,, Oral nutritional supplements promote significant weight gain in cystic fibrosis patients, Journal of Human Nutrition and Dietetics, 11, 95-104, 1998	Subjects acted as their own control; no randomization was used
Smith, C., Winn, A., Seddon, P., Ranganathan, S., A fat lot of good: balance and trends in fat intake in children with cystic fibrosis, Journal of Cystic Fibrosis, 11, 154-7, 2012	No relevant data
Smyth, R., Jahnke, N., Cochrane systematic reviews in cystic fibrosis, Journal of the Royal Society of Medicine, Supplement, 99, 6-12, 2006	Narrative review
Stallings, V. A., Stark, L. J., Robinson, K. A., Feranchak, A. P., Quinton, H., Clinical Practice Guidelines on, Growth, Nutrition, Subcommittee, Ad Hoc Working, Group, Evidence-based practice recommendations for nutrition-related management of children and adults with cystic fibrosis and pancreatic insufficiency: results of a	Recommendations for management based on a systematic review. The studies that were included in the systematic review are referenced and were assessed for inclusion.

Study	Reason for Exclusion
systematic review, Journal of the American Dietetic Association, 108, 832-9, 2008	
Stapleton, D. R., Gurrin, L. C., Zubrick, S. R., Silburn, S. R., Sherriff, J. L., Sly, P. D., What do children with cystic fibrosis and their parents know about nutrition and pancreatic enzymes?, Journal of the American Dietetic Association, 100, 1494-500, 2000	No relevant outcomes
Stapleton, D., Tunnecliffe, L., McGuiness, D., Sherriff, J., Sly, P., - Development of a nutrition and behaviour intervention program: go and grow with CF, 1998	No relevant outcomes
Stapleton, Denise R., Gurrin, Lyle C., Zubrick, Stephen R., Silburn, Sven R., Sherriff, Jill L., Sly, Peter D., The effect of 'Go and Grow with CF' on nutrition and pancreatic enzyme knowledge of children with cystic fibrosis, Australian Journal of Nutrition and Dietetics, 2001	No relevant outcomes
Stark, L. J., Opipari, L. C., Spieth, L. E., Jelalian, E., Quittner, A. L., Higgins, L., Mackner, L., Byars, K., Lapey, A., Stallings, V. A., Duggan, C., Contribution of Behavior Therapy to Dietary Treatment in Cystic Fibrosis: A Randomized Controlled Study with 2-Year Follow-up, Behavior Therapy, 34, 237-258, 2003	No relevant data (No standard deviation of the mean provided)
Stark, L. J., Opipari-Arrigan, L., Quittner, A. L., Bean, J., Powers, S. W., The effects of an intensive behavior and nutrition intervention compared to standard of care on weight outcomes in CF, Pediatric Pulmonology, 46, 31-5, 2011	No relevant comparison
Steinkamp, G., Demmelmair, H., Ruhl-Bagheri, I., von der Hardt, H., Koletzko, B., Energy supplements rich in linoleic acid improve body weight and essential fatty acid status of cystic fibrosis patients, Journal of Pediatric Gastroenterology & Nutrition, 31, 418-23, 2000	Quasi-randomised design; groups not comparable at the start of the study
Stephenson, A. L., Mannik, L. A., Walsh, S., Brotherwood, M., Robert, R., Darling, P. B., Nisenbaum, R., Moerman, J., Stanojevic, S., Longitudinal trends in nutritional status and the relation between lung function and BMI in cystic fibrosis: a population-based cohort study, American Journal of Clinical Nutrition, 97, 872-7, 2013	No relevant data
Stratton, R. J., Summary of a systematic review on oral nutritional supplement use in the community, Proceedings of the Nutrition Society, 59, 469-76, 2000	Summary of systematic review. Relevant included studies assessed for inclusion.
Van Biervliet, S., Devos, M., Delhaye, T., Van Biervliet, J. P., Robberecht, E., Christophe, A., Oral DHA supplementation in DeltaF508 homozygous cystic fibrosis patients, Prostaglandins Leukotrienes & Essential Fatty Acids, 78, 109-15, 2008	No relevant intervention

Study	Reason for Exclusion
Van Biervliet, S., Vande Velde, S., Van Biervliet, J. P., Robberecht, E., The effect of zinc supplements in cystic fibrosis patients, Annals of Nutrition & Metabolism, 52, 152-6, 2008	No relevant intervention
Woestenenk, J. W., Castelijns, S. J., van der Ent, C. K., Houwen, R. H., Nutritional intervention in patients with Cystic Fibrosis: a systematic review, Journal of Cystic Fibrosis, 12, 102-15, 2013	The included studies were assessed for inclusion. The quality of the systematic review was rated as low using the AMSTAR checklist.

H.12 Exocrine pancreatic insuffiency

Study	Reason for Exclusion
Borowitz, D., Goss, C. H., Stevens, C., Hayes, D., Newman, L., O'Rourke, A., Konstan, M. W., Wagener, J., Moss, R., Hendeles, L., Orenstein, D., Ahrens, R., Oermann, C. M., Aitken, M. L., Mahl, T. C., Young, K. R., Jr., Dunitz, J., Murray, F. T., Safety and preliminary clinical activity of a novel pancreatic enzyme preparation in pancreatic insufficient cystic fibrosis patients, Pancreas, 32, 258-63, 2006	Insufficient reporting of data
Bowler, I. M., Wolfe, S. P., Owens, H. M., Sheldon, T. A., Littlewood, J. M., Walters, M. P., A double blind lipase for lipase comparison of a high lipase and standard pancreatic enzyme preparation in cystic fibrosis, Archives of Disease in Childhood, 68, 227-30, 1993	Use of Nutrizym 22 in this age group (4.9-14.1 years)is not relevant to the protocol
Boyle,B.J., Long,W.B., Balistreri,W.F., Widzer,S.J., Huang,N., Effect of cimetidine and pancreatic enzymes on serum and fecal bile acids and fat absorption in cystic fibrosis, Gastroenterology, 78, 950-953, 1980	Cohort study design with n = 8 participants.
Braggion, C., Mastella, G., [Clinical trial of several preparation types of the pancreatic extract Pancrex V in the treatment of cystic fibrosis], Minerva pediatrica, 37, 891-6, 1985	Italian
Cameron,D.J., Pitcher-Wilmott,R., Milla,P.J., More,J., Ghale,G.K., Matthew,D.J., Harries,J.T., The effect of cimetidine on meal-stimulated gastric function and exogenous pancreatic enzymes in cystic fibrosis, Human Nutrition - Clinical Nutrition, 36, 475-481, 1982	Population too small to be relevant to the protocol
Carroccio, A., Pardo, F., Montalto, G., Iapichino, L., Soresi, M., Averna, M. R., Iacono, G., Notarbartolo, A., Use of famotidine in severe exocrine pancreatic insufficiency with persistent maldigestion on enzymatic replacement therapy. A long-term study in cystic fibrosis, Digestive Diseases & Sciences, 37, 1441-6, 1992	Insufficient reporting of data
Chalmers, D. M., Brown, R. C., Miller, M. G., Clarke, P. C., Kelleher, J., Littlewood, J. M., Losowsky, M. S., The influence of long-term cimetidine as an adjuvant to pancreatic	No PERT intervention - Placebo vs Cimetidine.

Study	Reason for Exclusion
enzyme therapy in cystic fibrosis, Acta Paediatrica Scandinavica, 74, 114-7, 1985	
Chazalette, J. P., Dain, M. P., Castaigne, J. P., Efficacy of pancrease: crossover comparative study versus eurobiol in cystic fibrosis, Journal of Pediatric Gastroenterology & Nutrition, 7 Suppl 1, S46-8, 1988	Randomisation was not performed
Cleghorn, G. J., Shepherd, R. W., Holt, T. L., The use of a synthetic prostaglandin E1 analogue (misoprostol) as an adjunct to pancreatic enzyme replacement in cystic fibrosis, Scandinavian Journal of Gastroenterology, Supplement, 23, 142-147, 1987	Not and RCT. Also Misoprostal is not relevant to the protocol.
Dudley, F., McNeil, S., Smith, M., Malabsorption in cystic fibrosis: failure to respond to cimetidine, Australian and New Zealand journal of medicine, 11, 223-4, 1981	Abstract
Duhamel, J. F., Vidailhet, M., Le Luyer, B., Douchain, F., Jehanne, M., Clavel, R., Guillot, M., [Multicenter comparative study of a new formulation of pancreatin in gastro-resistant microgranules for the treatment of exocrine pancreatic insufficiency in children with mucoviscidosis], Annales de pediatrie, 35, 69-74, 1988	French
Dutta, S. K., Hubbard, V. S., Appler, M., Critical examination of therapeutic efficacy of a pH-sensitive enteric-coated pancreatic enzyme preparation in treatment of exocrine pancreatic insufficiency secondary to cystic fibrosis, Digestive Diseases & Sciences, 33, 1237-44, 1988	Randomisation was not performed
Elliott, R. B., Escobar, L. C., Lees, H. R., Akroyd, R. M. D., Reilly, H. C., A comparison of two pancreatin microsphere preparations in cystic fibrosis, New Zealand Medical Journal, 105, 107-108, 1992	Numerical data are not presented in results
Francisco, M. P., Wagner, M. H., Sherman, J. M., Theriaque, D., Bowser, E., Novak, D. A., Ranitidine and omeprazole as adjuvant therapy to pancrelipase to improve fat absorption in patients with cystic fibrosis, Journal of Pediatric Gastroenterology & Nutrition, 35, 79-83, 2002	Outcomes not stated in protocol.
Giuliano, C. A., Dehoorne-Smith, M. L., Kale-Pradhan, P. B., Pancreatic enzyme products: digesting the changes, Annals of Pharmacotherapy, 45, 658-66, 2011	Narrative review
Goodchild, M. C., Sagaro, E., Brown, G. A., Cruchley, P. M., Jukes, H. R., Anderson, C. M., Comparative trial of pancrex V forte and nutrizym in treatment of malabsorption in cystic fibrosis, British Medical Journal, 3, 712-4, 1974	Use of Nutrizym in this age group (5-15 years) is not relevant to the protocol
Heijerman, H. G. M., Lamers, C. B. H. W., Dijkman, J. H., Bakker, W., Ranitidine compared with the dimethylprostaglandin E2	Comparison is not relevant (adjuvant ranitidine vs adjuvant enprostil)

Study	Reason for Exclusion
analogue enprostil as adjunct to pancreatic enzyme replacement in adult cystic fibrosis, Scandinavian Journal of Gastroenterology, Supplement, 25, 26-31, 1990	
Hill, C. M., Rolles, C. J., Keegan, P., Chand, R. A., Pancreatic enzyme supplementation in cystic fibrosis [letter], Archives of Disease in Childhood, 68, 150, 1993	Insufficient reporting of data
Kalnins, D., Ellis, L., Corey, M., Pencharz, P. B., Stewart, C., Tullis, E., Durie, P. R., Enteric-coated pancreatic enzyme with bicarbonate is equal to standard enteric-coated enzyme in treating malabsorption in cystic fibrosis, Journal of Pediatric Gastroenterology & Nutrition, 42, 256-61, 2006	enteric-coated buffered pancreatic enzyme (EC buffered PE) containing 1.5 mEq of bicarbonate per capsule with a conventional enteric-coated enzyme (EC-PE) capsule
Morrison, G., Morrison, J. M., Redmond, A. O., Byers, C. A., McCracken, K. J., Dodge, J. A., Guilford, S. A., Bowden, M. W., Comparison between a standard pancreatic supplement and a high enzyme preparation in cystic fibrosis, Alimentary Pharmacology & Therapeutics, 6, 549-55, 1992	Randomisation was not performed
Morrison, G., Morrison, J., Redmond, A., Byers, C., McCracken, K., Dodge, J. A., Pancreatic enzyme supplements in cystic fibrosis, Lancet, 338, 1596-7, 1991	Abstract
Munck, A., Duhamel, J. F., Lamireau, T., Le Luyer, B., Le Tallec, C., Bellon, G., Roussey, M., Foucaud, P., Ginies, J. L., Houzel, A., Marguet, C., Guillot, M., David, V., Kapel, N., Dyard, F., Henniges, F., Pancreatic enzyme replacement therapy for young cystic fibrosis patients, Journal of Cystic Fibrosis, 8, 14-8, 2009	Comparison not relevant to the protocol (Minimicrospheres vs microspheres)
Ng, May Sze, Franchini, Angelo J., Drug therapies for reducing gastric acidity in people with cystic fibrosis, Cochrane Database of Systematic Reviews, 2014	Relevant studies already identified from searches
Patchell, C. J., Desai, M., Weller, P. H., Macdonald, A., Smyth, R. L., Bush, A., Gilbody, J. S., Duff, S. A., Creon 10,000 Minimicrospheres vs. Creon 8,000 microspheres--an open randomised crossover preference study, Journal of Cystic Fibrosis, 1, 287-91, 2002	Comparison not relevant to the protocol (Minimicrospheres vs microspheres)
Petersen, W., Heilmann, C., Garne, S., Pancreatic enzyme supplementation as acid-resistant microspheres versus enteric-coated granules in cystic fibrosis. A double placebo-controlled cross-over study, Acta Paediatrica Scandinavica, 76, 66-9, 1987	Insufficient data reported
Pollack, P., Sander-Struckmeier, S., Beckmann, K., Janssen-Van Solingen, G., No effect of drugs that reduce gastric acid production on the efficacy of creon in randomized trials of patients with exocrine	Conference abstract

Study	Reason for Exclusion
pancreatic insufficiency, Pancreas, 40 (8), 1348, 2011	
Robinson, P. J., Olinsky, A., Smith, A. L., Chitravanshi, S. B., High compared with standard dose lipase pancreatic supplement, Archives of Disease in Childhood, 64, 143-5, 1989	Insufficient data reported (graph)
Robinson, P., Sly, P. D., Placebo-controlled trial of misoprostol in cystic fibrosis, Journal of Pediatric Gastroenterology & Nutrition, 11, 37-40, 1990	Adjuvant acid suppressant used is not relevant to protocol (Misoprostol)
Robinson,P.J., Sly,P.D., Smith,A.L., Effect of misoprostol on fat malabsorption in cystic fibrosis, Archives of Disease in Childhood, 63, 1081-1082, 1988	Adjuvant acid suppressant used is not relevant to protocol (Misoprostol)
Shafiq, Nusrat, Rana, Surinder, Bhasin, Deepak, Pandhi, Promila, Srivastava, Puja, Sehmby, Surjit S., Kumar, Raj, Malhotra, Samir, Pancreatic enzymes for chronic pancreatitis, Cochrane Database of Systematic Reviews, 2010	No relevant studies included (participants did not have cystic fibrosis)
Shah, A., Dinwiddie, R., Madge, S., Prescott, P., Hudson, G., High dose Nutrizym 22 in cystic fibrosis, European Journal of Pediatrics, 152, 763-4, 1993	Use of Nutrizym 22 in this age group (7-16 years)is not relevant to the protocol
Somaraju, Rani Usha, SolisMoya, Arturo, Pancreatic enzyme replacement therapy for people with cystic fibrosis, Cochrane Database of Systematic Reviews, 2014	Relevant studies already identified from searches. The review only included RCTs with a treatment duration of 28 days or more
Stead, R. J., Skypala, I., Hodson, M. E., Treatment of steatorrhoea in cystic fibrosis: a comparison of enteric-coated microspheres of pancreatin versus non-enteric-coated pancreatin and adjuvant cimetidine, Alimentary Pharmacology & Therapeutics, 2, 471-82, 1988	Comparison is not relevant according to the protocol (EC PERT vs Non-EC PERT Acid suppression)
Taylor, J. R., Gardner, T. B., Waljee, A. K., Dimagno, M. J., Schoenfeld, P. S., Systematic review: efficacy and safety of pancreatic enzyme supplements for exocrine pancreatic insufficiency, Alimentary Pharmacology & Therapeutics, 31, 57-72, 2010	Relevant studies already identified from searches
Toskes, P. P., Secci, A., Thieroff-Ekerdt, R., A randomized, double-blind, dose-response control, crossover study of two doses of eur-1008 (Zenpep) in Chronic Pancreatitis (CP) patients with Exocrine Pancreatic Insufficiency (EPI), Gastroenterology, 1), S394-S395, 2010	Conference abstract
Van de Vijver, E., Desager, K., Mulberg, A. E., Staelens, S., Verkade, H. J., Bodewes, F. A., Malfroot, A., Hauser, B., Sinaasappel, M., Van Biervliet, S., Behm, M., Pelckmans, P., Callens, D., Veereman-Wauters, G., Treatment of infants and toddlers with cystic fibrosis-related pancreatic insufficiency and fat malabsorption with pancrelipase MT, Journal	Pancrelipase microtablets (2-mm, enteric coated) were provided orally. Sixteen subjects, 6 to 30 months of age, were provided 500 U lipase/kg/meal for 5 days (baseline period). Subsequently, subjects were randomly assigned to 1 of 4 treatment groups (each n = 4), receiving 500, 1000, 1500, or 2000 U (Ph. EUR) of lipase/kg/meal, respectively, for 5 days (experimental period).

Study	Reason for Exclusion
of Pediatric Gastroenterology & Nutrition, 53, 61-4, 2011	
Vyas, H., Matthew, D. J., Milla, P. J., A comparison of enteric coated microspheres with enteric coated tablet pancreatic enzyme preparations in cystic fibrosis. A controlled study, European Journal of Pediatrics, 149, 241-3, 1990	Comparison not relevant to the protocol (EC microspheres vs EC tablets)
Waljee, A. K., Dimagno, M. J., Wu, B. U., Schoenfeld, P. S., Conwell, D. L., Systematic review: pancreatic enzyme treatment of malabsorption associated with chronic pancreatitis, Alimentary Pharmacology & Therapeutics, 29, 235-46, 2009	Relevant studies already identified from searches

H.13 Distal intestinal obstruction syndrome

Study	Reason for Exclusion
Khoshoo, V., Udall Jr, J. N., Meconium ileus equivalent in children and adults, American Journal of Gastroenterology, 89, 153-157, 1994	Narrative review.
Koletzko,S., Corey,M., Ellis,L., Spino,M., Stringer,D.A., Durie,P.R., Effects of cisapride in patients with cystic fibrosis and distal intestinal obstruction syndrome, Journal of Pediatrics, 117, 815-822, 1990	This RCT assesses the effects of cisapride vs placebo. Cisapride is not a treatment of interest.
Lavie, M., Manovitz, T., Vilozni, D., Levy-Mendelovich, S., Sarouk, I., Weintraubv, I., Shoseyov, D., Cohen-Cymberknoh, M., Rivlin, J., Efrati, O., Long-term follow-up of distal intestinal obstruction syndrome in cystic fibrosis, World Journal of Gastroenterology, 21, 318-325, 2015	Comparison not relevant for the review: compares DIOS and non-DIOS groups. Descriptive results of treatments in the DIOS group are provided but without comparison.
Lillibridge, C. B., Docter, J. M., Eidelman, S., Oral administration of n-acetyl cysteine in the prophylaxis of "meconium ileus equivalent", Journal of Pediatrics, 71, 887-9, 1967	Case report (1 case)
Mabogunje, O. A., Wang, C. I., Mahour, H., Improved survival of neonates with meconium ileus, Archives of Surgery, 117, 37-40, 1982	Retrospective study. Provides descriptive data only. The main aim of the study is to evaluate if treatments have improved over time, which is not the aim of our review.
McPartlin, J. F., Dickson, J. A., Swain, V. A., The use of gastrografin in the relief of residual and late bowel obstruction in cystic fibrosis, British Journal of Surgery, 60, 707-10, 1973	Case reports only (3 cases)
Munck, A., Alberti, C., Boizeau, P., Colombo, C., Kashirskaya, N., Ellemunter, H., Fotoulaki, M., Houwen, R., Robberecht, E., Wilschanski, M., Dios Study G, International prospective study of distal intestinal obstruction syndrome, Pediatric Pulmonology, 49, 411, 2014	Conference abstract. No data relevant to the protocol is available in the abstract.
O'Halloran, S. M., Gilbert, J., McKendrick, O. M., Carty, H. M., Heaf, D. P., Gastrografin in acute meconium ileus equivalent, Archives of Disease in Childhood, 61, 1128-30, 1986	No comparison of interest: evaluates Gastrografin but it is a non-comparative study.
Sawyer, S. M., Taylor, R., McMahon, R., Robertson, C. F., Meconium ileus in cystic fibrosis	Retrospective review of hospital data of patients with MI and admitted to hospital.

Study	Reason for Exclusion
- a 20-year review of morbidity, mortality and management, Pediatric Surgery International, 9, 180-184, 1994	
Wagget, J., Bishop, H. C., Koop, C. E., Experience with gastrografin enema in the treatment of meconium ileus, Journal of Pediatric Surgery, 5, 649-54, 1970	Case reports only (8 cases)
Wagget, J., Johnson, D. G., Borns, P., Bishop, H. C., The nonoperative treatment of meconium ileus by Gastrografin enema, Journal of Pediatrics, 77, 407-11, 1970	Case reports only(4 cases)
Winfield, R. D., Beierle, E. A., Pediatric surgical issues in meconium disease and cystic fibrosis, Surgical Clinics of North America, 86, 317-27, viii-ix, 2006	Narrative review. Does not provide data on different treatments.

H.14 Liver disease

Study	Reason for Exclusion
Addy, C., Bateman, K., Bell, N. J., Diagnoses of cystic fibrosis liver disease in an adult cystic fibrosis centre, Journal of Cystic Fibrosis. Conference: 36th European Cystic Fibrosis Conference Lisbon Portugal. Conference Start, 12	Conference abstract.
Aqul, A. A., Harney, S., Raza, R., Sawicki, G. S., Mitchell, P. D., Jonas, M. M., Fawaz, R., Correlation of liver stiffness, as determined by transient elastography, with the presence and severity of clinical liver disease in children and young adults with cystic fibrosis, Hepatology, 1), 819A, 2013	Conference abstract.
Aqul, A., Jonas, M. M., Harney, S., Raza, R., Sawicki, G. S., Mitchell, P. D., Fawaz, R., Correlation of Transient Elastography With Severity of Cystic Fibrosis Related Liver Disease, Journal of Pediatric Gastroenterology and Nutrition, no pagination, 2016	This study aims to assess the correlation between liver stiffness measurement and severity of liver disease. Insufficient data presented to perform analysis according to the protocol.
Behrens, C. B., Langholz, J. H., Eiler, J., Jenewein, R., Naehrlich, L., Fuchs, K., Harth, S., Krombach, G. A., Alzen, G. F., A pilot study of the characterization of hepatic tissue strain in children with cystic-fibrosis-associated liver disease (CFLD) by acoustic radiation force impulse imaging, Pediatric Radiology, 43, 552-7, 2013	ARFI intervention is not relevant to the protocol.
Bodewes, F. A. J. A., Van Der Doef, H. P. J., Houwen, R. H. J., Verkade, H. J., Elevated serum gamma-glutamyl transpeptidase predicts the future development of cirrhotic cystic fibrosis liver disease, Journal of Pediatric Gastroenterology and Nutrition, 50, E145-E146, 2010	Conference abstract.
Bodewes, F. A., van der Doef, H. P., Houwen, R. H., Verkade, H. J., Increase of Serum gamma-Glutamyltransferase Associated With Development of Cirrhotic Cystic Fibrosis Liver	Insufficient data presented to perform analysis according to the protocol.

Study	Reason for Exclusion
Disease, Journal of Pediatric Gastroenterology & NutritionJ Pediatr Gastroenterol Nutr, 61, 113-8, 2015	
Canas, T., Macia, A., Munoz-Codoceo, R. A., Fontanilla, T., Gonzalez-Rios, P., Miralles, M., Gomez-Mardones, G., Hepatic and Splenic Acoustic Radiation Force Impulse Shear Wave Velocity Elastography in Children with Liver Disease Associated with Cystic Fibrosis, BioMed Research International, 2015, 517369, 2015	Not relevant intervention (hepatic and splenic acoustic radiation force impulse shear wave velocity elastography).
Ciuca, I., Pop, L., Popa, Z., Deleanu, A., Turcu, S., Guta-Almajan, B., Tamas, L., Popa, I., Liver elasticity in CF associated liver disease, Journal of Cystic Fibrosis, 11, S112, 2012	Conference abstract.
Costa, P. C., Barreto, C. C., Pereira, L., Lobo, M. L., Costa, M. A., Lopes, A. I., Cystic fibrosis-related liver disease: a single-center experience, Pediatric Reports, 3, e21, 2011	Descriptive study that does not present any data relevant to the protocol.
De Ledinghen, V., Clouzeau, H., Foucher, J., Vergniol, J., Chermak, F., Gaboreau, M., Lamireau, T., Longitudinal assessment of patients with cystic fibrosis with liver stiffness measurement (Fibroscan), Journal of Hepatology, 56, S543-S544, 2012	Conference abstract.
Durieu, I., Pellet, O., Simonot, L., Durupt, S., Bellon, G., Durand, D. V., Minh, V. A., Sclerosing cholangitis in adults with cystic fibrosis: a magnetic resonance cholangiographic prospective study, Journal of Hepatology, 30, 1052-6, 1999	Study does not present any data relevant to the protocol.
Feigelson, J., Anagnostopoulos, C., Poquet, M., Pecau, Y., Munck, A., Navarro, J., Liver cirrhosis in cystic fibrosis - Therapeutic implications and long term follow up, Archives of Disease in Childhood, 68, 653-657, 1993	Study does not present any data relevant to the protocol.
Fiel, S. B., Friedman, A. C., Caroline, D. F., Radecki, P. D., Faerber, E., Grumbach, K., Magnetic resonance imaging in young adults with cystic fibrosis, Chest, 91, 181-4, 1987	Study does not present any data relevant to the protocol.
Foster, J. A., Ramsden, W. H., Conway, S. P., Taylor, J. M., Etherington, C., The role of IDA scintigraphy in the follow-up of liver disease in patients with cystic fibrosis, Nuclear Medicine Communications, 23, 673-81, 2002	Scintigraphy is not relevant to the protocol.
Friedrich-Rust, M., Schlueter, N., Smaczny, C., Eickmeier, O., Rosewich, M., Feifel, K., Herrmann, E., Poynard, T., Gleiber, W., Lais, C., Zielen, S., Wagner, T. O., Zeuzem, S., Bojunga, J., Non-invasive measurement of liver and pancreas fibrosis in patients with cystic fibrosis, Journal of Cystic Fibrosis, 12, 431-9, 2013	Insufficient data presented to perform analysis according to the protocol. ARFI intervention is not relevant to the protocol.
Fustik, S., Liver cirrhosis and portal hypertension in cystic fibrosis, Srpski Arhiv Za Celokupno Lekarstvo, 141, 764-9, 2013	Insufficient data presented to perform analysis according to the protocol.
Gominon, A. L., Frison, E., Vergniol, J., Clouzeau, H., Bui, S., Fayon, M., De Ledinghen, V., Lamireau, T., Evolutivity of cystic fibrosis-	Conference abstract.

Study	Reason for Exclusion
related liver disease assessed by elastometry, Hepatology, 63, 324A-325A, 2016	
Graham, N., Manhire, A. R., Stead, R. J., Lees, W. R., Hodson, M. E., Batten, J. C., Cystic fibrosis: ultrasonographic findings in the pancreas and hepatobiliary system correlated with clinical data and pathology, Clinical Radiology, 36, 199-203, 1985	Insufficient data presented to perform analysis according to the protocol.
Hillaire, S., Regnard, N. E., De Miranda, S., Grenet, D., Stern, M., Paradis, V., Vilgrain, V., Picard, C., Mellot, F., Non invasive liver elastography (LSM) and computed tomography (CT) for evaluation of liver disease in 50 cystic fibrosis adult patients, Hepatology, 56, 831A, 2012	Conference abstract.
Hung, J. C., Howie, A. F., Beckett, G. J., Sood, M., Hambleton, G., Super, M., The use of human glutathione S-transferase A1 in the detection of cystic fibrosis liver disease, Journal of Paediatrics & Child Health, 34, 335-8, 1998	hGST A1 enzyme is not of relevant to the protocol.
Jones, G. H., Richardson, P., Ledson, M., Walshaw, M., Greenwood, J., Transient elastography measurements in adults with cystic fibrosis liver disease, Journal of Cystic Fibrosis, 13, S94, 2014	Conference abstract.
Jones, G. H., Richardson, P., Ledson, M., Walshaw, M., Greenwood, J., Transient elastography can be used to guide monitoring of cystic fibrosis related liver disease in adult patients, Journal of Cystic Fibrosis, 13, S94, 2014	Conference abstract.
Katzenstein, T. L., Andersen, E. S., Pressler, T., Jensen, F., Transient elastography and abdominal ultra-sound investigation of adult CF patients monitored at the Copenhagen CF centre, Journal of Cystic Fibrosis. Conference: 36th European Cystic Fibrosis Conference Lisbon Portugal. Conference Start, 12	Conference abstract.
Koh, C., Chernick, M., Zhao, X., Eccleston, J., Sakiani, S., Hoofnagle, J. H., Heller, T., Long term follow-up of adult cystic fibrosis patients: Noninvasive markers predict a second wave of liver disease, Hepatology, 52, 905A, 2010	Conference abstract.
Kumari-Subaiya, S., Gorvoy, J., Phillips, G., Portal vein measurement by ultrasonography in patients with long-standing cystic fibrosis: Preliminary observations, Journal of Pediatric Gastroenterology and Nutrition, 6, 71-78, 1987	Insufficient data presented to perform analysis according to the protocol.
Kumari-Subaiya, S., Gorvoy, J., Phillips, G., Ross, P., Riddelsberger, M. M., Portal vein measurement by ultrasonography in patients with long-standing cystic fibrosis: preliminary observations, Journal of Pediatric Gastroenterology & Nutrition, 6, 71-8, 1987	Insufficient data presented to perform analysis according to the protocol.
Lenaerts, C., Lapierre, C., Patriquin, H., Bureau, N., Lepage, G., Harel, F., Marcotte, J., Roy, C. C., Surveillance for cystic fibrosis-associated	Insufficient data presented to perform analysis according to the protocol.

Study	Reason for Exclusion
hepatobiliary disease: early ultrasound changes and predisposing factors, Journal of Pediatrics, 143, 343-50, 2003	
Leung, D. H., Ye, W., Molleston, J. P., Weymann, A., Ling, S., Paranjape, S. M., Romero, R., Schwarzenberg, S. J., Palermo, J., Alonso, E. M., Murray, K. F., Marshall, B. C., Sherker, A. H., Siegel, M. J., Krishnamurthy, R., Harned, R., Karmazyn, B., Magee, J. C., Narkewicz, M. R., Cystic Fibrosis Liver Disease, Network, Baseline Ultrasound and Clinical Correlates in Children with Cystic Fibrosis, Journal of PediatricsJ Pediatr, 167, 862-868.e2, 2015	Aim not relevant to the protocol. This study aims to determine which demographic and clinical features are associated with abnormal US results.
Lindblad, A., Hultcrantz, R., Strandvik, B., Bile-duct destruction and collagen deposition: a prominent ultrastructural feature of the liver in cystic fibrosis, Hepatology, 16, 372-81, 1992	Insufficient data presented to perform analysis according to the protocol.
Ling, S. C., Wilkinson, J. D., Hollman, A. S., McColl, J., Evans, T. J., Paton, J. Y., The evolution of liver disease in cystic fibrosis, Archives of Disease in Childhood, 81, 129-32, 1999	Insufficient data presented to perform analysis according to the protocol.
Ling, S. C., Ye, W., Leung, D. A., Weymann, A., Karnsakul, W., Freeman, A. J., Magee, J. C., Narkewicz, M. R., Simple biomarkers correlate with liver ultrasound scan abnormalities in children with cystic fibrosis, Hepatology, 63, 275A, 2016	Conference abstract.
Malbrunot-Wagner, A. C., Bridoux, L., Nousbaum, J. B., Riou, C., Dirou, A., Ginies, J. L., Maurage, C., Cagnard, B., Pelatan, C., Dabadie, A., Transient elastography and portal hypertension in pediatric patients with cystic fibrosis Transient elastography and cystic fibrosis, Journal of Cystic Fibrosis, 10, 338-42, 2011	Insufficient data presented to perform analysis according to the protocol.
Manco, M., Zupone, C. L., Alghisi, F., D'Andrea, M. L., Lucidi, V., Monti, L., Pilot study on the use of acoustic radiation force impulse imaging in the staging of cystic fibrosis associated liver disease, Journal of Cystic Fibrosis, 11, 427-32, 2012	Insufficient data presented to perform analysis according to the protocol. ARFI intervention is not relevant to the protocol.
McHugo, J. M., McKeown, C., Brown, M. T., Weller, P., Shah, K. J., Ultrasound findings in children with cystic fibrosis, British Journal of Radiology, 60, 137-41, 1987	Insufficient data presented to perform analysis according to the protocol.
Menten, R., Leonard, A., Clapuyt, P., Vincke, P., Nicolae, A. C., Lebecque, P., Transient elastography in patients with cystic fibrosis, Pediatric Radiology, 40, 1231-5, 2010	Insufficient data presented to perform analysis according to the protocol.
Monti, L., Manco, M., Lo Zupone, C., Latini, A., D'Andrea, M. L., Alghisi, F., Lucidi, V., Toma, P., Bonomo, L., Acoustic radiation force impulse (ARFI) imaging with Virtual Touch Tissue Quantification in liver disease associated with	Insufficient data presented to perform analysis according to the protocol. ARFI intervention is not relevant to the protocol.

Study	Reason for Exclusion
cystic fibrosis in children, Radiologia Medica, 117, 1408-18, 2012	
Namazova-Baranova, L., Surkov, A., Tomilova, A., Simonova, O., Torshkhoeva, R., Diagnostic capabilities of liver ultrasound elastography in the care of children with cystic fibrosis, Journal of Cystic Fibrosis, 11, S62, 2012	Conference abstract.
Nash, K. L., Allison, M. E., McKeon, D., Lomas, D. J., Haworth, C. S., Bilton, D., Alexander, G. J., A single centre experience of liver disease in adults with cystic fibrosis 1995-2006, Journal of Cystic Fibrosis, 7, 252-7, 2008	Study does not present any data relevant to the protocol.
Palermo, J., Kotyk, J. J., Siegel, M. J., Magnetic resonance elastography for assessment of cystic fibrosis liver disease, Gastroenterology, 1), S686, 2011	Conference abstract.
Potter, C. J., Fishbein, M., Hammond, S., McCoy, K., Qualman, S., Can the histologic changes of cystic fibrosis-associated hepatobiliary disease be predicted by clinical criteria?, Journal of Pediatric Gastroenterology & Nutrition, 25, 32-6, 1997	Insufficient data presented to perform analysis according to the protocol.
Quattrucci, S., Angelico, M., Stancati, M., Bertasi, S., Cantusci, D., De Sanctis, A., Antonelli, M., Hepatobiliary involvement in adolescents and adults with cystic fibrosis, Acta Universitatis Carolinae - Medica, 36, 180-2, 1990	Study does not present any data relevant to the protocol.
Rattenbury, J. M., Taylor, C. J., Heath, P. K., Howie, A. F., Beckett, G. J., Serum glutathione S-transferase B1 activity as an index of liver function in cystic fibrosis, Journal of Clinical Pathology, 48, 771-4, 1995	Study does not present any data relevant to the protocol. GST B1 enzyme is not of relevant to the protocol.
Robberecht, E., Van Langenhove, K., Declercq, D., Peeters, H., Van Vlierberghe, H., Transient elastography (TE; Fibroscanj) as systematic screening test forincipient cystic fibrosis liver disease (CFLD), Journal of Pediatric Gastroenterology and Nutrition, 52, E139-E140, 2011	Conference abstract.
Rowland, M., Gallagher, C. G., O'Laoide, R., Canny, G., Broderick, A., Hayes, R., Greally, P., Slattery, D., Daly, L., Durie, P., Bourke, B., Outcome in cystic fibrosis liver disease, American Journal of Gastroenterology, 106, 104-9, 2011	Descriptive study that does not present any data relevant to the protocol.
Schoenau, E., Boeswald, W., Wanner, R., Herzog, K. H., Boewing, B., Boehles, H. J., Stehr, K., High-molecular-mass ("biliary") isoenzyme of alkaline phosphatase and the diagnosis of liver dysfunction in cystic fibrosis, Clinical Chemistry, 35, 1888-90, 1989	Study does not present any data relevant to the protocol.
Shapira, R., Hadzic, N., Francavilla, R., Koukulis, G., Price, J. F., Mieli-Vergani, G., Retrospective review of cystic fibrosis presenting as infantile liver disease, Archives of Disease in Childhood, 81, 125-8, 1999	Study does not present any data relevant to the protocol.

Study	Reason for Exclusion
Staufer, K., Halilbasic, E., Trauner, M., Kazemi-Shirazi, L., Cystic fibrosis related liver disease--another black box in hepatology, International Journal of Molecular Sciences, 15, 13529-49, 2014	Narrative review.
Stonebraker, J. R., Ooi, C. Y., Pace, R. G., Corvol, H., Knowles, M. R., Durie, P. R., Ling, S. C., Features of Severe Liver Disease With Portal Hypertension in Patients With Cystic Fibrosis, Clinical Gastroenterology & HepatologyClin Gastroenterol Hepatol, 14, 1207-1215.e3, 2016	Aim not relevant to the protocol. This study evaluates the phenotype associated with severe liver disease.
Swann, R. E., Mustafa, Z., Ross, E., Priest, M., Validation of fibroscan as a screening tool for cystic fibrosis associated liver disease in an adult population, Gut, 61, A410, 2012	Conference abstract.
Valletta, E. A., Loreti, S., Cipolli, M., Cazzola, G., Zanolla, L., Portal hypertension and esophageal varices in cystic fibrosis. Unreliability of echo-Doppler flowmetry, Scandinavian Journal of Gastroenterology, 28, 1042-6, 1993	Study does not present any data relevant to the protocol.
Van Biervliet, S., Verdievel, H., Vande Velde, S., De Bruyne, R., De Looze, D., Verhelst, X., Geerts, A., Robberecht, E., Van Vlierberghe, H., Longitudinal Transient Elastography Measurements Used in Follow-up for Patients with Cystic Fibrosis, Ultrasound in Medicine & Biology, 42, 848-54, 2016	Insufficient data presented to perform analysis according to the protocol.
Van Der Feen, C., Van Der Doef, H., Mundt, M., Arends, J., Van Der Ent, C. K., Houwen, R., Liver elastography to detect early stage cystic fibrosis related liver disease in children, Journal of Cystic Fibrosis, 9, S80, 2010	Conference abstract.
Walsh, M. J., Lewindon, P. J., Shepherd, R. W., Greer, R. M., Williamson, R., Pereira, T. N., Frawley, K., Bell, S., Smith, J. L., Ramm, G. A., Detection and follow-up of hepatic fibrosis in cystic fibrosis: A role for diagnostic liver biopsy and serum markers in evaluating outcomes of Cystic Fibrosis liver disease, Journal of Gastroenterology and Hepatology, 24, A225, 2009	Conference abstract.
Williams, S. G., Evanson, J. E., Barrett, N., Hodson, M. E., Boultbee, J. E., Westaby, D., An ultrasound scoring system for the diagnosis of liver disease in cystic fibrosis, Journal of Hepatology, 22, 513-21, 1995	Study does not present any data relevant to the protocol.
Williams, S. M., Goodman, R., Thomson, A., McHugh, K., Lindsell, D. R., Ultrasound evaluation of liver disease in cystic fibrosis as part of an annual assessment clinic: a 9-year review, Clinical Radiology, 57, 365-70, 2002	Insufficient data presented to perform analysis according to the protocol.
Wilschanski, M., Menachem, M., Kerem, E., Simanovski, N., Armoni, S., Kelly, D., Shteyer, E., Evaluation of a new ultrasound scoring system for CF liver disease, Journal of Cystic Fibrosis, 9, S80, 2010	Conference abstract.

H.15 Ursodeoxycholic acid

Study	Reason for Exclusion
Bittner, P., Posselt, H. G., Sailer, T., Ott, H., Magdorf, K., Wahn, U., Arleth, S., Bertele-Harms, R. M., Wolf, A., Krawinkel, M., Lindemann, H., The effect of treatment with ursodeoxycholic acid in cystic fibrosis and hepatopathy: results of a placebo-controlled study, Bile acids as therapeutic agents. From basic science to clinical practice. Falk symposium, 58, 1991	Data for placebo arm not reported.
Bittnere, P., Sailer, T., Ott, H., Posselt, H. G., Magdorf, K., Krawinkel, M., Therapy of the accompanying hepatopathy in mucovischidosis with ursodesoxychloric acid (UDC). Results of a placebo-controlled study, Monatsschrift fur Kinderheilkunde, 138, 1990	Article in German.
Cheng, K., Ashby, D., Smyth, R., Ursodeoxycholic acid in cystic fibrosis-related liver disease: a systematic review, Journal of the Royal Society of Medicine, 90 Suppl 31, 6-12, 1997	Sys review - studies included in Cochrane 2014 review.
Colombo, C., Allocca, M., Quattrucci, S., Traverso, G., Farina, S., Lucidi, V., Santini, B., Casciaro, R., Carnovale, E., Cipolli, M., De Rose, V., Gagliardini, R., Miano, A., Travan, L., Viviani, L., Battezzati, P., Ursodeoxycholic acid for liver disease associated to cystic fibrosis: long-term follow-up of patients enrolled in the italian multicenter trial [abstract], Pediatric pulmonology, 40, 343, 2005	Non-comparative cohort design (abstract).
Colombo, C., Crosignani, A., Assaisso, M., Battezzati, P. M., Podda, M., Giunta, A., Zimmer-Nechemias, L., Setchell, K. D., Ursodeoxycholic acid therapy in cystic fibrosis-associated liver disease: a dose-response study, Hepatology, 16, 924-30, 1992	No comparison with placebo/control.
Colombo, C., Crosignani, A., Battezzati, P. M., Castellani, M. R., Comi, S., Melzi, M. L., Giunta, A., Delayed intestinal visualization at hepatobiliary scintigraphy is associated with response to long-term treatment with ursodeoxycholic acid in patients with cystic fibrosis-associated liver disease, Journal of Hepatology, 31, 672-7, 1999	Non-comparative cohort design.
Costa, P. C., Barreto, C. C., Pereira, L., Lobo, M. L., Costa, M. A., Lopes, A. I., Cystic fibrosis-related liver disease: a single-center experience, Pediatric Reports, 3, e21, 2011	Non-comparative cohort design.
Debray, D., Kelly, D., Houwen, R., Strandvik, B., Colombo, C., Best practice guidance for the diagnosis and management of cystic fibrosis-associated liver disease, Journal of Cystic Fibrosis, 10 Suppl 2, S29-36, 2011	Guidelance for best practice. Taken into consideration for linking evidence to recommendations (LETR).
Desmond, C. P., Wilson, J., Bailey, M., Clark, D., Roberts, S. K., The benign course of liver disease in adults with cystic fibrosis and the	Non-comparative cohort design.

Study	Reason for Exclusion
effect of ursodeoxycholic acid, Liver International, 27, 1402-8, 2007	
Galabert, C., Montet, J. C., Lengrand, D., Lecuire, A., Sotta, C., Figarella, C., Chazalette, J. P., Effects of ursodeoxycholic acid on liver function in patients with cystic fibrosis and chronic cholestasis, Journal of Pediatrics, 121, 138-41, 1992	Non-comparative cohort design.
Kappler, M., Espach, C., Schweiger-Kabesch, A., Lang, T., Hartl, D., Hector, A., Glasmacher, C., Griese, M., Ursodeoxycholic acid therapy in cystic fibrosis liver disease - A retrospective long-term follow-up case-control study, Alimentary Pharmacology and Therapeutics, 36, 266-273, 2012	Case control design.
Lenaerts, C., Lapierre, C., Patriquin, H., Bureau, N., Lepage, G., Harel, F., Marcotte, J., Roy, C. C., Surveillance for cystic fibrosis-associated hepatobiliary disease: early ultrasound changes and predisposing factors, Journal of Pediatrics, 143, 343-50, 2003	Non-comparative cohort design.
Lepage, G., Paradis, K., Lacaille, F., Senechal, L., Ronco, N., Champagne, J., Lenaerts, C., Roy, C. C., Rasquin-Weber, A., Ursodeoxycholic acid improves the hepatic metabolism of essential fatty acids and retinol in children with cystic fibrosis, Journal of Pediatrics, 130, 52-8, 1997	Poor outcome reporting. Better quality data for hepatobiliary enzymes was already included in the review.
Nousia-Arvanitakis, S., Fotoulaki, M., Economou, H., Xefteri, M., Galli-Tsinopoulou, A., Long-term prospective study of the effect of ursodeoxycholic acid on cystic fibrosis-related liver disease, Journal of Clinical Gastroenterology, 32, 324-8, 2001	Non-comparative cohort design.
O'Brien, S. M., Campbell, G. R., Burke, A. F., Maguire, O. C., Rowlands, B. J., FitzGerald, M. X., Hegarty, J. E., Serum bile acids and ursodeoxycholic acid treatment in cystic fibrosis-related liver disease, European Journal of Gastroenterology and Hepatology, 8, 477-483, 1996	Cohort study design in which the control group had no liver disease. Therefore, groups are not comparable. This is a further study of O'Brien 1992 which was an RCT of n = 12 participants with CF and liver disease and was included in the review.
O'Connor, P. J., Southern, K. W., Bowler, I. M., Irving, H. C., Robinson, P. J., Littlewood, J. M., The role of hepatobiliary scintigraphy in cystic fibrosis, Hepatology, 23, 281-7, 1996	No UDCA intervention.
Poropat, Goran, Giljaca, Vanja, Stimac, Davor, Gluud, Christian, Bile acids for liver-transplanted patients, Cochrane Database of Systematic Reviews, 2010	No CF participants included.
Rubin, R. A., Kowalski, T. E., Khandelwal, M., Malet, P. F., Ursodiol for hepatobiliary disorders, Annals of Internal Medicine, 121, 207-218, 1994	A review on the use of UDCA (study design not appropriate).
Siano, M., De Gregorio, F., Boggia, B., Sepe, A., Ferri, P., Buonpensiero, P., Di Pasqua, A., Raia, V., Ursodeoxycholic acid treatment in patients with cystic fibrosis at risk for liver disease, Digestive & Liver Disease, 42, 428-31, 2010	Cohort study, both groups received UDCA.

Study	Reason for Exclusion
van de Meeberg, P. C., Houwen, R. H., Sinaasappel, M., Heijerman, H. G., Bijleveld, C. M., Vanberge-Henegouwen, G. P., Low-dose versus high-dose ursodeoxycholic acid in cystic fibrosis-related cholestatic liver disease. Results of a randomized study with 1-year follow-up, Scandinavian Journal of Gastroenterology, 32, 369-73, 1997	Both groups received UDCA (low vs high) rather than UDCA vs placebo/control/no treatment.

H.16 Cystic fibrosis related diabetes

Study	Reason for Exclusion
Assael,B.M., Kronfeld,K., Honer,M., Holl,R.W., Staden,U., Classen,M., Schuster,A., Mellies,U., Posselt,H.G., Ballmann,M., Wiebel,M., Rietschel,E., Stern,M., Teschler,H., Smaczny,C., Kohnlein,T., Wienhausen-Wilke,V., Claass,A., Biedermann,T., Nahrlich,L., Dockter,G., Koster,H., Hebestreit,A., Balke,E.H., Heuer,H.E., Kamin,W., Kuster,P., Szczepanski,R., Keller,K.M., Generlich,H., Bresser,H.G., Kopp,M., Herting,E., Kreger,K., Feikert,H.J., Hautz,J., Schilling,B., Meyer,E., Mall,M., Wiebicke,W., Tegtmeye,F.K., Assael,B., Hubert,D., Mosnier-Pudar,H., Lenoir,G., Robert,J.J., Kessler,L., Weiss,L., Nove-Josserand,R., Vantygehm,M.C., Munck,A., Wizla,N., Leroy,S., Loeuille,G.A., Thalhammer,D., Huttegger,I., Eichler,I., Gotz,M., Open randomised prospective comparative multi-centre intervention study of patients with Cystic fibrosis and early diagnosed diabetes mellitus, BMC Pediatrics, 14, -, 2014	This study compares 2 treatments following diagnosis of CFRD.
Balzer, B. W. R., Simmons, L. R., Moriarty, C. P., Bye, P. T. P., Steinbeck, K. S., Continuous glucose monitoring as a useful decision-making tool for adults with cystic fibrosis, Internet Journal of Pulmonary Medicine, 16, 2014	Case series.
Bilbao, L., Clemente, M., Costas, L., Gartner, S., Losada, M., Armengol, E., Yeste, D., Carrascosa, A., Evaluation of continuous glucose monitoring in cystic fibrosis patients, Hormone Research in Paediatrics, 84, 195, 2015	The study does not report prognostic outcomes. Conference abstract.
Bizzarri, C., Lucidi, V., Ciampalini, P., Bella, S., Russo, B., Cappa, M., Clinical effects of early treatment with insulin glargine in patients with cystic fibrosis and impaired glucose tolerance, Journal of Endocrinological Investigation, 29, RC1-4, 2006	No relevant screening tool (OGTT) (review question 2).
Bizzarri, C., Montemitro, E., Pedicelli, S., Ciccone, S., Majo, F., Cappa, M., Lucidi, V., Glucose tolerance affects pubertal growth and final height of children with cystic fibrosis, Pediatric Pulmonology, 50, 144-9, 2015	No relevant screening tool (OGTT) (review question 2).
Boudreau, V., Reynaud, Q., Dubois, C. L., Coriati, A., Desjardins, K., Durieu, I., Rabasa-Lhoret, R., Screening for Cystic Fibrosis-Related	Discussion paper

Study	Reason for Exclusion
Diabetes: Matching Pathophysiology and Addressing Current Challenges, Canadian Journal of DiabetesCan, 40, 466-470, 2016	
Brennan, A. L., Beynon, J., Clinical updates in cystic fibrosis-related diabetes, Seminars in Respiratory & Critical Care MedicineSemin, 36, 236-50, 2015	Narrative review/ discussion paper. Relevant references checked for inclusion.
Brodsky, J., Dougherty, S., Makani, R., Rubenstein, R. C., Kelly, A., Elevation of 1-hour plasma glucose during oral glucose tolerance testing is associated with worse pulmonary function in cystic fibrosis, Diabetes Care, 34, 292-5, 2011	The study does not report prognostic outcomes.
Cawood, T. J., McKenna, M. J., Gallagher, C. G., Smith, D., Chung, W. Y., Gibney, J., O'Shea, D., Cystic fibrosis-related diabetes in adults, Irish Medical Journal, 99, 83-6, 2006	No relevant screening tool (OGTT) (review question 2).
Chan, C. L., Pyle, L., Vigers, T., Zemanick, E. T., Zeitler, P. S., Sagel, S. D., Nadeau, K., Continuous glucose monitoring reveals glucose abnormalities in CF youth despite normal oral glucose tolerance testing, Pediatric PulmonologyPediatr Pulmonol, 50, 419, 2015	The study does not report prognostic outcomes. Conference abstract.
Coriati, A., Ziai, S., Azar, M., Berthiaume, Y., Rabasa-Lhoret, R., Characterization of patients with cystic fibrosis presenting an indeterminate glucose tolerance (INDET), Journal of Cystic Fibrosis, 15, 127-32, 2016	The study does not report prognostic outcomes.
Dobson, L., Sheldon, C. D., Hattersley, A. T., Conventional measures underestimate glycaemia in cystic fibrosis patients, Diabetic Medicine, 21, 691-6, 2004	The study does not report prognostic outcomes.
Dyce, P., Daniels, J., Dunne, J., Govin, B., Malone, V., Nazareth, D., Walshaw, M., Continuous glucose monitoring (CGM) for cystic fibrosis related diabetes (CFRD): Sweet success or bitter disappointment?, Pediatric PulmonologyPediatr Pulmonol, 47, 420, 2012	Conference abstract. Not an RCT.
Dyce, P., Malone, V., Nazareth, D., Walshaw, M., The diagnostic utility of continuous glucose monitoring (CGM) in cystic fibrosis related diabetes (CFRD), Journal of Cystic Fibrosis. Conference: 36th European Cystic Fibrosis Conference. Lisbon Portugal. Conference Start, 12	Conference abstract. Not an RCT.
Ebdon, A. M., Nolan, S., Dick, K., Alexander, S., Bridges, N., Carr, S. B., Continuous glucose monitoring is a useful tool for diagnosis of cystic fibrosis related diabetes, Journal of Cystic Fibrosis, 13, S13, 2014	No prognostic outcomes reported. Conference abstract.
Franzese, A., Valerio, G., Buono, P., Spagnuolo, M. I., Sepe, A., Mozzillo, E., De Simone, I., Raia, V., Continuous glucose monitoring system in the screening of early glucose derangements in children and adolescents with cystic fibrosis, Journal of Pediatric Endocrinology, 21, 109-16, 2008	The study does not report prognostic outcomes.

Study	Reason for Exclusion
Hameed, S., Jaffe, A., Verge, C. F., Advances in the detection and management of cystic fibrosis related diabetes, Current Opinion in Pediatrics, 27, 525-533, 2015	No relevant screening tool (OGTT) (review question 2).
Hameed, S., Morton, J. R., Jaffe, A., Field, P. I., Belessis, Y., Yoong, T., Katz, T., Verge, C. F., Early glucose abnormalities in cystic fibrosis are preceded by poor weight gain, Diabetes Care, 33, 221-6, 2010	The study does not report prognostic outcomes.
Jefferies, C., Solomon, M., Perlman, K., Sweezey, N., Daneman, D., Continuous glucose monitoring in adolescents with cystic fibrosis, Journal of Pediatrics, 147, 396-8, 2005	Case series.
Jonas, A. M., Collaco, J. M., Blackman, S. M., Impaired fasting glucose is followed by more rapid decline in lung function in cystic fibrosis, Pediatric PulmonologyPediatr Pulmonol, 48, 418, 2013	The study does not report prognostic outcomes. Conference abstract.
Leclercq, A., Gauthier, B., Rosner, V., Weiss, L., Moreau, F., Constantinescu, A. A., Kessler, R., Kessler, L., Early assessment of glucose abnormalities during continuous glucose monitoring associated with lung function impairment in cystic fibrosis patients, Journal of Cystic Fibrosis, 13, 478-84, 2014	The study does not report prognostic outcomes.
Leclercq, A., Kessler, L., Rosner, V., Weiss, L., Gauthier, B., Kessler, R., Lung function and early abnormality of glucose tolerance (GT) in cystic fibrosis (CF) patients, European Respiratory Journal. Conference: European Respiratory Society Annual Congress, 42, 2013	Conference abstract. See full paper study in list of excluded studies (Leclercq 2014).
McLachlan, C., Beach, R., Laing, R., Frazer, G., Lunt, H., Cawood, T. J., Could glucose self-monitoring become the preferred tool for screening and monitoring glucose control in cystic fibrosis?, Practical Diabetes International, 28, 173-176, 2011	Intervention not relevant to the protocol (glucose self-monitoring).
Milla, C. E., Warwick, W. J., Moran, A., Trends in pulmonary function in patients with cystic fibrosis correlate with the degree of glucose intolerance at baseline, American Journal of Respiratory & Critical Care Medicine, 162, 891-5, 2000	No relevant screening tool (OGTT) (review question 2).
Mohan, K., Israel, K. L., Miller, H., Grainger, R., Ledson, M. J., Walshaw, M. J., Long-term effect of insulin treatment in cystic fibrosis-related diabetes, Respiration, 76, 181-6, 2008	No relevant screening tool (treatment based on WHO and US CF consensus criteria) (review question 2).
Mohan, K., Miller, H., Burhan, H., Ledson, M. J., Walshaw, M. J., Management of cystic fibrosis related diabetes: a survey of UK cystic fibrosis centers, Pediatric Pulmonology, 43, 642-7, 2008	Survey of UK cystic fibrosis centres.
Moran, A., Becker, D., Casella, S. J., Gottlieb, P. A., Kirkman, M. S., Marshall, B. C., Slovis, B., Alexander, P., Beall, R. J., Brunzell, C., Campbell, Iii P. W., Chin, M., Cohen, R. C., Brooks, J. F., George, C., Hazle, L., Katz, M., McKeon, C., Onady, G., Robinson, K. A.,	Narrative review.

Study	Reason for Exclusion
Rodgers, T., Sabadosa, K. A., Schindler, T., Stecenko, A., Wood, M. E., Young, D., Epidemiology, pathophysiology, and prognostic implications of cystic fibrosis-related diabetes: A technical review, Diabetes Care, 33, 2677-2683, 2010	
Moreau, F., Weiller, M. A., Rosner, V., Weiss, L., Hasselmann, M., Pinget, M., Kessler, R., Kessler, L., Continuous glucose monitoring in cystic fibrosis patients according to the glucose tolerance, Hormone & Metabolic Research, 40, 502-6, 2008	The study does not report prognostic outcomes.
Ode, K. L., Frohnert, B., Laguna, T., Phillips, J., Holme, B., Regelmann, W., Thomas, W., Moran, A., Oral glucose tolerance testing in children with cystic fibrosis, Pediatric Diabetes, 11, 487-92, 2010	No relevant screening tool (OGTT) (review question 2).
O'Riordan,S.M., CFRD in childhood and the use of continuous glucose monitoring, Pediatric Pulmonology, 44, 204-205, 2009	Discussion piece. Conference abstract.
Rayner,R.J., Williams,V., Archer,M., Salt,G., Jones,S., Early recognition of CF related diabetes in children by the use of continuous glucose monitoring systems, Journal of Cystic Fibrosis, 11, S52-, 2012	Conference abstract. Not an RCT.
Sheikh, S., Putt, M. E., Forde, K. A., Rubenstein, R. C., Kelly, A., Elevation of one hour plasma glucose during oral glucose tolerance testing, Pediatric Pulmonology, 50, 963-9, 2015	No relevant screening tool (OGTT) (review question 2).
Walshaw, M., Routine OGTT screening for CFRD - no thanks, Journal of the Royal Society of Medicine, 102 Suppl 1, 40-4, 2009	Discussion paper.
Widger, J., Ranganathan, S., Robinson, P. J., Progression of structural lung disease on CT scans in children with cystic fibrosis related diabetes, Journal of Cystic Fibrosis, 12, 216-21, 2013	No relevant screening tool (OGTT) (review question 2).

H.17 Bone mineral density

Study	Reason for Exclusion
Aris, R. M., Renner, J. B., Winders, A. D., Buell, H. E., Riggs, D. B., Lester, G. E., Ontjes, D. A., Increased rate of fractures and severe kyphosis: sequelae of living into adulthood with cystic fibrosis, Annals of Internal Medicine, 128, 186-93, 1998	The study does not report on the predictive ability of measuring bone mineral density.
Bianchi, M. L., Leonard, M. B., Bechtold, S., Hogler, W., Mughal, M. Z., Schonau, E., Sylvester, F. A., Vogiatzi, M., van den Heuvel-Eibrink, M. M., Ward, L., Bone health in children and adolescents with chronic diseases that may affect the skeleton: The 2013 ISCD pediatric official positions, Journal of Clinical DensitometryJ Clin Densitom, 17, 281-294, 2014	Narrative review. Relevant references have been identified. Good for background reading.
Donovan, D. S., Jr., Papadopoulos, A., Staron, R. B., Addesso, V., Schulman, L., McGregor, C.,	The study does not report on the predictive ability of measuring bone mineral density.

Study	Reason for Exclusion
Cosman, F., Lindsay, R. L., Shane, E., Bone mass and vitamin D deficiency in adults with advanced cystic fibrosis lung disease, American Journal of Respiratory & Critical Care Medicine, 157, 1892-9, 1998	
Ellis, K. J., Shypailo, R. J., Hardin, D. S., Perez, M. D., Motil, K. J., Wong, W. W., Abrams, S. A., Z score prediction model for assessment of bone mineral content in pediatric diseases, Journal of Bone & Mineral Research, 16, 1658-64, 2001	The study does not report on the predictive ability of measuring bone mineral density.
Lakey, W. C., Spratt, S., Vinson, E. N., Gesty-Palmer, D., Weber, T., Palmer, S., Osteoporosis in lung transplant candidates compared to matched healthy controls, Clinical Transplantation, 25, 426-435, 2011	The study does not report on the predictive ability of measuring bone mineral density.
Robertson, J., Macdonald, K., Prevalence of bone loss in a population with cystic fibrosis, British Journal of Nursing, 19, 636-9, 2010	The study does not report on the predictive ability of measuring bone mineral density.
Rossini, M., Del Marco, A., Dal Santo, F., Gatti, D., Braggion, C., James, G., Adami, S., Prevalence and correlates of vertebral fractures in adults with cystic fibrosis, Bone, 35, 771-6, 2004	The study does not report on the predictive ability of measuring bone mineral density.
Rossini, M., Viapiana, O., Del Marco, A., de Terlizzi, F., Gatti, D., Adami, S., Quantitative ultrasound in adults with cystic fibrosis: correlation with bone mineral density and risk of vertebral fractures, Calcified Tissue International, 80, 44-9, 2007	Cross-sectional study design.
Stephenson, A., Jamal, S., Dowdell, T., Pearce, D., Corey, M., Tullis, E., Prevalence of vertebral fractures in adults with cystic fibrosis and their relationship to bone mineral density, Chest, 130, 539-44, 2006	The study does not report on the predictive ability of measuring bone mineral density.(cross-sectional study)
Ujhelyi, R., Treszl, A., Vasarhelyi, B., Holics, K., Toth, M., Arato, A., Tulassay, T., Tulassay, Z., Szathmari, M., Bone mineral density and bone acquisition in children and young adults with cystic fibrosis: a follow-up study, Journal of Pediatric Gastroenterology & Nutrition, 38, 401-6, 2004	The study does not report on the predictive ability of measuring bone mineral density.

H.18 Exercise

Study	Reason for Exclusion
Asher, M. I., Pardy, R. L., Coates, A. L., Thomas, E., Macklem, P. T., The effects of inspiratory muscle training in patients with cystic fibrosis, American Review of Respiratory Disease, 126, 855-9, 1982	No relevant data
Baldwin, D. R., Hill, A. L., Peckham, D. G., Knox, A. J., Effect of addition of exercise to chest physiotherapy on sputum expectoration and lung function in adults with cystic fibrosis, Respiratory Medicine, 88, 49-53, 1994	No relevant intervention (No exercise programme; exercise is performed on one single day). Cross-over study with 8 participants.

Study	Reason for Exclusion
Boyd, Stephanie, Brooks, Dina, Agnew-Coughlin, Jennifer, Ashwell, Jo, Evaluation of the Literature on the Effectiveness of Physical Therapy Modalities in the Management of Children With Cystic Fibrosis, Pediatric Physical Therapy, 6, 70-76, 1994	Narrative review
Bradley, J., Moran, F., Physical training for cystic fibrosis, Cochrane Database of Systematic Reviews, CD002768, 2008	The update of this Cochrane review was included (Radtke 2015)
Burtin, C., Van Remoortel, H., Vrijsen, B., Langer, D., Colpaert, K., Gosselink, R., Decramer, M., Dupont, L., Troosters, T., Impact of exacerbations of cystic fibrosis on muscle strength, Respiratory Research, 14, 46, 2013	No relevant data
Button, B. M., Wilson, C., Dentice, R., Cox, N. S., Middleton, A., Tannenbaum, E., Bishop, J., Cobb, R., Burton, K., Wood, M., Moran, F., Black, R., Bowen, S., Day, R., Depiazzi, J., Doiron, K., Doumit, M., Dwyer, T., Elliot, A., Fuller, L., Hall, K., Hutchins, M., Kerr, M., Lee, A. L., Mans, C., O'Connor, L., Steward, R., Potter, A., Rasekaba, T., Scoones, R., Tarrant, B., Ward, N., West, S., White, D., Wilson, L., Wood, J., Holland, A. E., Physiotherapy for cystic fibrosis in Australia and New Zealand: A clinical practice guideline, Respirology, 21, 656-67, 2016	Guideline and systematic review. The quality of the systematic review was rated low using the AMSTAR checklist. The references of included studies were checked to identify relevant trials
Cardoso, R. M. T., Viana, R. A., Physiotherapy intervention in Cystic Fibrosis: A systematic review, Arquivos de Medicina, 25, 186-195, 2011	Portuguese language
Centre for, Reviews, Dissemination,, Evaluation of the literature on the effectiveness of physical therapy modalities in the management of children with cystic fibrosis (Structured abstract), Database of Abstracts of Reviews of Effects, 2015	Structured abstract of a systematic review that has been assessed for inclusion.
Centre for, Reviews, Dissemination,, Effects of inspiratory muscle training in cystic fibrosis: a systematic review (Structured abstract), Database of Abstracts of Reviews of Effects, 2015	Structured abstract of a systematic review that has been assessed for inclusion.
Centre for, Reviews, Dissemination,, The evidence regarding exercise training in the management of cystic fibrosis: a systematic review (Structured abstract), Database of Abstracts of Reviews of Effects, 2015	Structured abstract of a systematic review that has been assessed for inclusion
Centre for, Reviews, Dissemination,, The effects of physical exercise on cystic fibrosis: a review of the literature (Provisional abstract), Database of Abstracts of Reviews of Effects, 2015	Structured abstract of a systematic review that has been assessed for inclusion.
Cerny, F. J., Relative effects of bronchial drainage and exercise for in-hospital care of patients with cystic fibrosis, Physical Therapy, 69, 633-9, 1989	No relevant outcomes (change in weight in the study was assessed with a follow-up shorter than one month)
Collaco, Joseph M., Blackman, Scott M., Raraigh, Karen S., Morrow, Christopher B.,	This is a cohort study on habitual physical activity. Although there were no RCT on habitual

Study	Reason for Exclusion
Cutting, Garry R., Paranjape, Shruti M., Self-reported exercise and longitudinal outcomes in cystic fibrosis: a retrospective cohort study, BMC Pulmonary Medicine, 14, 1-8, 2014	physical activity, cohort studies relating to this intervention were not assessed for inclusion because the Committee thought that the information from RCTs on long-term exercise programmes was sufficient to formulate recommendations.
Cox, N. S., Alison, J. A., Holland, A. E., Interventions to promote physical activity in people with cystic fibrosis, Paediatric Respiratory Reviews, 15, 237-239, 2014	This paper summarizes a Cochrane review which has been assessed for inclusion.
Cox, N. S., Alison, J. A., Holland, A. E., Interventions for promoting physical activity in people with cystic fibrosis, Cochrane Database of Systematic Reviews, 12, CD009448, 2013	Included studies are included in Cochrane review on physical exercise training for CF by Radtke et al., published in 2015.
De Jong, W., Van Aalderen, W. M. C., Kraan, J., Koeter, G. H., Van Der Schans, C. P., Inspiratory muscle training in patients with cystic fibrosis, Respiratory Medicine, 95, 31-36, 2001	The intervention in this study was IMT at 40% of PImax.
Debska, G., Mazurek, H., Factors related to changes in the quality of life among Polish adolescents and adults with cystic fibrosis over a 1-year period, Patient preference & adherence, 9, 1763-70, 2015	No relevant data
del Corral, T., Percegona, J., Seborga, M., Rabinovich, R. A., Vilaro, J., Physiological response during activity programs using Wii-based video games in patients with cystic fibrosis (CF), Journal of Cystic Fibrosis, 13, 706-711, 2014	No relevant intervention (no exercise programme)
Dwyer, T. J., Alison, J. A., McKeough, Z. J., Daviskas, E., Bye, P. T. P., Effects of exercise on respiratory flow and sputum properties in patients with cystic fibrosis, Chest, 139, 870-877, 2011	No relevant intervention (not an exercise programme; exercise is only performed for 20 minutes)
Flores, J. S., Rovedder, P. M., Ziegler, B., Pinotti, A. F., Barreto, S. S., Dalcin Pde, T., Clinical Outcomes and Prognostic Factors in a Cohort of Adults With Cystic Fibrosis: A 7-Year Follow-Up Study, Respiratory Care, 61, 192-9, 2016	No relevant intervention
Goodill, S. W., Dance/movement therapy for adults with cystic fibrosis: pilot data on mood and adherence, Alternative Therapies in Health & Medicine, 11, 76-7, 2005	No relevant intervention (The authors mention that dance/movement therapy is a creative arts psychotherapy)
Gruber, W., Orenstein, D. M., Braumann, K. M., Do responses to exercise training in cystic fibrosis depend on initial fitness level?, European Respiratory Journal, 38, 1336-42, 2011	This study was a non-randomized trial. Non-randomized trials were only included if they reported data on the critical outcome time to next exacerbation, or on the intervention high intensity interval training (both were not covered by RCTs). This study did not cover the aforementioned outcome/intervention.
Hebestreit, H., Kriemler, S., Radtke, T., Exercise for all cystic fibrosis patients: Is the evidence strengthening?, Current Opinion in Pulmonary Medicine, 21, 591-595, 2015	Narrative review

Study	Reason for Exclusion
Hebestreit, H., Schmid, K., Kieser, S., Junge, S., Ballmann, M., Roth, K., Hebestreit, A., Schenk, T., Schindler, C., Posselt, H. G., Kriemler, S., Quality of life is associated with physical activity and fitness in cystic fibrosis, BMC Pulmonary Medicine, 14, 26, 2014	No relevant outcomes. The data that are analyzed are from two studies that are included in this review
Hind, K., Truscott, J. G., Conway, S. P., Exercise during childhood and adolescence: a prophylaxis against cystic fibrosis-related low bone mineral density? Exercise for bone health in children with cystic fibrosis, Journal of Cystic Fibrosis, 7, 270-6, 2008	Narrative review
Hristara-Papadopoulou, A., Tsanakas, J., Results of active cycle of breathing techniques and conventional physiotherapy in mucociliary clearance in children with cystic fibrosis, Hippokratia, 11, 202-204, 2007	No relevant intervention
Irons, Yoon J., Petocz, Peter, Kenny, Theadora Dianna, Chang, Anne B., Singing as an adjunct therapy for children and adults with cystic fibrosis, Cochrane Database of Systematic Reviews, 2015	No relevant intervention
Kriemler, S., Radtke, T., Christen, G., Kerstan-Huber, M., Hebestreit, H., Short-Term Effect of Different Physical Exercises and Physiotherapy Combinations on Sputum Expectoration, Oxygen Saturation, and Lung Function in Young Patients with Cystic Fibrosis, LungLung, 194, 659-64, 2016	Interventions are exercise sessions lasting less than a day rather than exercise programmes
Kuys, S. S., Hall, K., Peasey, M., Wood, M., Cobb, R., Bell, S. C., Gaming console exercise and cycle or treadmill exercise provide similar cardiovascular demand in adults with cystic fibrosis: a randomised cross-over trial, Journal of Physiotherapy, 57, 35-40, 2011	No relevant intervention (no exercise programme; each exercise intervention comprised 15 minutes of exercise)
Lannefors, L., Wollmer, P., Mucus clearance with three chest physiotherapy regimes in cystic fibrosis: a comparison between postural drainage, PEP and physical exercise, European Respiratory Journal, 5, 748-53, 1992	No relevant intervention (no exercise programme; one of the interventions is exercise performed on one single day); Cross-over study with 9 participants.
Macfarlane, P. I., Heaf, D., Changes in airflow obstruction and oxygen saturation in response to exercise and bronchodilators in cystic fibrosis, Pediatric Pulmonology, 8, 4-11, 1990	No relevant intervention (no exercise programme, 6-min run)
Nixon, P. A., Orenstein, D. M., Curtis, S. E., Ross, E. A., Oxygen supplementation during exercise in cystic fibrosis, The American Review of Respiratory Disease 1990 Oct;142(4):807-811, 1990	No relevant intervention (the intervention is oxygen supplementation during exercise test)
O'Keefe, K., Orr, R., Huang, P., Selvadurai, H., Cooper, P., Munns, C. F., Singh, M. A., The effect of whole body vibration exposure on muscle function in children with cystic fibrosis: a pilot efficacy trial, Journal of Clinical Medicine Research, 5, 205-16, 2013	Non-randomised controlled cross-over trial; 7 participants were recruited
Orenstein, D. M., Franklin, B. A., Doershuk, C. F., Hellerstein, H. K., Germann, K. J., Horowitz,	This study was a non-randomized trial. Non-randomized trials were only included if they

Study	Reason for Exclusion
J. G., Stern, R. C., Exercise conditioning and cardiopulmonary fitness in cystic fibrosis. The effects of a three-month supervised running program, Chest, 80, 392-8, 1981	reported data on the critical outcome time to next exacerbation, or on the intervention high intensity interval training (both were not covered by RCTs). This study did not cover the aforementioned outcome/intervention.
Papaioannou, M., A randomized controlled trial of a 3-year home exercise program in cystic fibrosis, Pediatric Physical Therapy, 13, 94-5, 2001	This paper summarizes the paper by Schneiderman-Walker et al. published in 2000, which is included in this review
Paranjape, Shruti M., Barnes, Laura A., Carson, Kathryn A., von Berg, Karen, Loosen, Holly, Mogayzel Jr, Peter J., Exercise improves lung function and habitual activity in children with cystic fibrosis, Journal of Cystic Fibrosis, 11, 18-23, 2012	Study design does not fit protocol; the paper compares before and after values for one group only; moreover, the study includes other non relevant comparisons
Perez, M., Groeneveld, I. F., Santana-Sosa, E., Fiuza-Luces, C., Gonzalez-Saiz, L., Villa-Asensi, J. R., Lopez-Mojares, L. M., Rubio, M., Lucia, A., Aerobic fitness is associated with lower risk of hospitalization in children with cystic fibrosis, Pediatr Pulmonol, 49, 641-9, 2014	No relevant intervention (no exercise programme)
Radtke, T., Benden, C., Kriemler, S., Physical Activity and Exercise Training in Lung Transplant Recipients with Cystic Fibrosis: 'What We Know, What We Don't Know and Where to Go', LungLung, 194, 177-8, 2016	Discussion paper/narrative review
Reid, W. D., Geddes, E. L., O'Brien, K., Brooks, D., Crowe, J., Effects of inspiratory muscle training in cystic fibrosis: a systematic review, Clinical Rehabilitation, 22, 1003-13, 2008	Included studies are included in the Cochrane systematic review from 2013, which is included in this review
Reix, P., Aubert, F., Werck-Gallois, M. C., Toutain, A., Mazzocchi, C., Moreux, N., Bellon, G., Rabilloud, M., Kassai, B., Exercise with incorporated expiratory manoeuvres was as effective as breathing techniques for airway clearance in children with cystic fibrosis: a randomised crossover trial, Journal of Physiotherapy, 58, 241-7, 2012	No relevant intervention (no exercise programme; the exercise intervention consisted of three periods of exercise each lasting 5 minutes)
Salonini, E., Gambazza, S., Meneghelli, I., Tridello, G., Sanguanini, M., Cazzarolli, C., Zanini, A., Assael, B. M., Active video game playing in children and adolescents with cystic fibrosis: Exercise or just fun?, Respiratory Care, 60, 1172-1179, 2015	No relevant intervention (no exercise programme; each exercise intervention lasted 20 min)
Sandsund, C. A., Roughton, M., Hodson, M. E., Pryor, J. A., Musculoskeletal techniques for clinically stable adults with cystic fibrosis: a preliminary randomised controlled trial, Physiotherapy, 97, 209-17, 2011	No relevant intervention
Savi, D., Di Paolo, M., Simmonds, N., Onorati, P., Internullo, M., Quattrucci, S., Winston, B., Laveneziana, P., Palange, P., Relationship between daily physical activity and aerobic fitness in adults with cystic fibrosis, BMC Pulmonary Medicine, 15, 59, 2015	Case-control study
Savi, D., Simmonds, N., Di Paolo, M., Quattrucci, S., Palange, P., Banya, W.,	Daily physical activity is an outcome, not an intervention

Study	Reason for Exclusion
Hopkinson, N. S., Bilton, D., Relationship between pulmonary exacerbations and daily physical activity in adults with cystic fibrosis, BMC Pulmonary Medicine, 15, 151, 2015	
Sawyer, E. H., Clanton, T. L., Improved pulmonary function and exercise tolerance with inspiratory muscle conditioning in children with cystic fibrosis, Chest, 104, 1490-7, 1993	The intervention in this study was IMT at 60% of PImax.
Schaar, B., Platen, P., Literaturanalyse: sportliche aktivitaten bei zystischer fibrose (The effects of physical exercise on cystic fibrosis: a review of the literature) [German], Atemwegs- und Lungenkrankheiten 2006 Sep;32(9):361-365, 2006	German
Schmidt, Anne Mette, Jacobsen, Ulla, Bregnballe, Vibeke, Olesen, Hanne Vebert, Ingemann-Hansen, Thorsten, Thastum, Mikael, Schietz, Peter Oluf, Exercise and quality of life in patients with cystic fibrosis: A 12-week intervention study, Physiother Theory PractPhysiotherapy theory and practice, 27, 548-556, 2011	Uncontrolled before-after study
Schneiderman, J. E., Wilkes, D. L., Atenafu, E. G., Nguyen, T., Wells, G. D., Alarie, N., Tullis, E., Lands, L. C., Coates, A. L., Corey, M., Ratjen, F., Longitudinal relationship between physical activity and lung health in patients with cystic fibrosis, European Respiratory Journal, 43, 817-23, 2014	This is a cohort study. Although there were no RCT on habitual physical activity, cohort studies relating to habitual physical activity were not assessed for inclusion because the Committee thought that the information from RCTs on long-term exercise programmes was sufficient to formulate recommendations.
Shoemaker, M. J., Hurt, H., Arndt, L., The evidence regarding exercise training in the management of cystic fibrosis: a systematic review, Cardiopulmonary Physical Therapy Journal, 19, 75-83, 2008	Included studies were assessed for potential inclusion. The quality of the systematic review was low according to the AMSTAR checklist.
Thomas, J., Cook, D. J., Brooks, D., Chest physical therapy management of patients with cystic fibrosis. A meta-analysis, American Journal of Respiratory & Critical Care Medicine, 151, 846-50, 1995	Relevant included studies assessed for inclusion
Thompson, K., Cystic Fibrosis-Update on Exercise, Physician & SportsmedicinePhys Sportsmed, 18, 103-6, 1990	Discussion paper
van de Weert-van Leeuwen, P. B., Hulzebos, H. J., Werkman, M. S., Michel, S., Vijftigschild, L. A., van Meegen, M. A., van der Ent, C. K., Beekman, J. M., Arets, H. G., Chronic inflammation and infection associate with a lower exercise training response in cystic fibrosis adolescents, Respir MedRespiratory medicine, 108, 445-52, 2014	This study was not an RCT. Non-RCTs were only included if they reported data on the critical outcome time to next exacerbation, or on the intervention high intensity interval training (both were not covered by RCTs). This study did not cover the aforementioned outcome/intervention.
van Doorn, N., Exercise programs for children with cystic fibrosis: a systematic review of randomized controlled trials, Disability & Rehabilitation, 32, 41-9, 2010	Included studies were assessed for potential inclusion. The quality of the systematic review was low according to the AMSTAR checklist.
Vivodtzev, I., Decorte, N., Wuyam, B., Gonnet, N., Durieu, I., Levy, P., Cracowski, J. L., Cracowski, C., Benefits of neuromuscular	No relevant intervention

Study	Reason for Exclusion
electrical stimulation prior to endurance training in patients with cystic fibrosis and severe pulmonary dysfunction, Chest, 143, 485-93, 2013	
Wheatley, C. M., Baker, S. E., Morgan, M. A., Martinez, M. G., Morgan, W. J., Wong, E. C., Karpen, S. R., Snyder, E. M., Effects of exercise intensity compared to albuterol in individuals with cystic fibrosis, Respiratory Medicine, 109, 463-74, 2015	No relevant intervention (No exercise programme; exercise is performed during one single visit for three separate 15 min exercise bouts)
Williams, C. A., Physical activity and health of adults with cystic fibrosis, RespirologyRespirology, 21, 404-5, 2016	Discussion paper
Williams, C. A., Benden, C., Stevens, D., Radtke, T., Exercise training in children and adolescents with cystic fibrosis: theory into practice, International Journal of Pediatrics, 2010	Narrative review
Wlodarczyk, O. M., Barinow-Wojewodzki, A., The impact of resistance respiratory muscle training with a SpiroTiger() device on lung function, exercise performance, and health-related quality of life in respiratory diseases, Kardiochirurgia I Torakochirurgia PolskaKardiochir, 12, 386-90, 2015	No relevant studies included

H.19 Psychological assessment

Study	Reason for Exclusion
Alpern, A. N., Hoffman, M. F., Riekert, K. A., Quittner, A. L., Using the CFQ-R to screen for eating and body image disturbances in adolescents with CF, Pediatric Pulmonology, 48, 423-424, 2013	Abstract only. No specific psychometric properties or diagnostic accuracy data reported.
Burrows, J. A., Bunting, J. P., Masel, P. J., Bell, S. C., Nebulised dornase alpha: adherence in adults with cystic fibrosis, Journal of Cystic Fibrosis, 1, 255-9, 2002	No specific psychometric properties or diagnostic accuracy data reported.
Dalcin, P. D. T. R., Rampon, G., Pasin, L. R., Ramon, G. M., Abrahao, C. L. D. O., De Oliveira, V. Z., Adherence to treatment in patients with cystic fibrosis. [Portuguese, English], Jornal Brasileiro de Pneumologia, 33, 663-670, 2007	Paper in Portuguese.
Duff, A. J. A., Abbott, J., Cowperthwaite, C., Sumner, C., Hurley, M. A., Quittner, A., Depression and anxiety in adolescents and adults with cystic fibrosis in the UK: A cross-sectional study, Journal of Cystic Fibrosis, 13, 745-753, 2014	Evaluates one screening tool. No reference standard.
Duff, A. J. A., Bowmer, G., Waldron, R., Cammidge, S., Peckham, D., Latchford, G., Administering the PHQ8 and GAD7 in routine UK CF care: In situ utilisation in a paediatric and an adult centre, Journal of Cystic Fibrosis, 14, S124, 2015	Abstract only. No specific psychometric properties or diagnostic accuracy data reported.
Duff, A. J., Abbott, J., Cowperthwaite, C., Sumner, C., Anxiety and depression in CF in the UK (TIDES-UK), Journal of Cystic Fibrosis, 11, S138, 2012	Abstract only. No specific psychometric properties or diagnostic accuracy data reported.

Study	Reason for Exclusion
Duff, A. J., Cowperthwaite, C. J., Abbott, J., Sumner, C. L., Anxiety and depression in CF: Preliminary data from the TIDES-UK study, Journal of Cystic Fibrosis, 9, S96, 2010	Abstract only. No specific psychometric properties or diagnostic accuracy data reported.
Elmasry, S., Mok, S. S., Braithwaite, M., Sofianopoulos, S., Clark, D., Finlayson, F., Williams, E., Poole, S., Dooley, M., Liew, D., Wilson, J., Adherence behaviour of adult cystic fibrosis (CF) patients to prescribed azithromycin, Journal of Cystic Fibrosis, 9, S24, 2010	Abstract only. No specific psychometric properties or diagnostic accuracy data reported.
Farach, L., Quittner, A. L., Chau, H., Yang, S., Rutkowski, A., Use of the modified medication possession ratio to determine adherence in cystic fibrosis patients, Pediatric Pulmonology, 46, 410-411, 2011	Conference abstract. No specific psychometric properties or diagnostic accuracy data reported.
Fidika, A., Mai, S., Herle, M., Goldbeck, L., Fear of Progression Questionnaire for caregivers of youth with cystic fibrosis (FoP-Q/C), Journal of Cystic Fibrosis, 13, S115, 2014	Includes parents
Filigno, S. S., Weiland, J. L., McPhail, G. L., Miller, J., Moore, S., Backstrom, J. M., Measurement of psychosocial risk using an empirically-based tool in CF care, Pediatric Pulmonology, 49, 430, 2014	Assessment tool not in protocol (Psychosocial Assessment Tool, PAT).
Flavin, S., Adult cystic fibrosis depression screening with the PHQ9 as a routine intervention in evaluation of quality of life and self care, Pediatric Pulmonology, 49, 432, 2014	Abstract only. No specific psychometric properties or diagnostic accuracy data reported.
Fries, L., Savant, A. P., Berger, A., Nufer, J., Roach, C. M., Smith, B. A., Cogswell, A., Borowitz, D., Routine depression screening: Dissemination of a standardized depression screening and intervention process, Pediatric Pulmonology, 50, 422, 2015	Abstract only. No specific psychometric properties or diagnostic accuracy data reported.
Goetz, D. M., Frederick, C. A., Roach, C. M., Cogswell, A., Smith, B. A., The relationship between depression and patient outcomes in children and adults with CF, Pediatric Pulmonology, 50, 442-443, 2015	Abstract only. No specific psychometric properties or diagnostic accuracy data reported.
Goldberg, S., Janus, M., Washington, J., Simmons, R. J., MacLusky, I., Fowler, R. S., Prediction of preschool behavioral problems in healthy and pediatric samples, Journal of Developmental & Behavioral Pediatrics, 18, 304-13, 1997	Assessment tool not in protocol (Child Behaviour Checklist).
Hawthorne, N., Hawwa, A. F., Shields, M. D., Reid, A. J., McElnay, J. C., Non-adherence to therapy in children with cystic fibrosis, Pediatric Pulmonology, 46, 408, 2011	Conference abstract. No specific psychometric properties or diagnostic accuracy data reported.
Helms, S. W., Prinstein, M. J., Dellon, E. P., Body image and disordered eating among adolescents with cystic fibrosis, Pediatric Pulmonology, 48, 424, 2013	Abstract only. No specific psychometric properties or diagnostic accuracy data reported.
Heroux, C., Cloutier, K., Gervais, P., Daigneault, P., Pharmacy records: The best way to identify low adherence in a pediatric cystic fibrosis population?, Pediatric Pulmonology, 50, 430-431, 2015	Conference abstract. No specific psychometric properties or diagnostic accuracy data reported.

Study	Reason for Exclusion
Jacque, C. A., Stress reactions in children with cystic fibrosis, Scandinavian Journal of Gastroenterology, Supplement, 23, 47-51, 1987	Case report and narrative review.
Latchford, G., Duff, A. J., Screening for depression in a single CF centre, Journal of Cystic Fibrosis, 12, 794-6, 2013	Compares two screening tools. No reference standard.
Latchford, G., Duff, A. J. A., Oracz, K., Bradley, N., Assessing anxiety, depression and suicidal ideation in a single CF centre; Experiences using the PHQ and HADS, Journal of Cystic Fibrosis, 9, S96, 2010	Abstract only. No specific psychometric properties or diagnostic accuracy data reported.
Logan, G., Oken, T., Simons, L., Garcia, G., Depression, anxiety, and suicide screening in cystic fibrosis: Patient accessibility to mental health treatment, Pediatric Pulmonology, 47, 441, 2012	Abstract only. No specific psychometric properties or diagnostic accuracy data reported.
McLean, K. A., Madan, A., Monzon, A., Quittner, A. L., Trajectories of depression and anxiety in adolescents with CF, Pediatric Pulmonology, 49, 2014	Abstract only. No specific psychometric properties or diagnostic accuracy data reported.
Modi, A. C., Driscoll, K. A., Montag-Leifling, K., Acton, J. D., Screening for symptoms of depression and anxiety in adolescents and young adults with cystic fibrosis, Pediatric Pulmonology, 46, 153-9, 2011	Reports prevalence according to one assessment tool. No reference standard used.
Modi, A. C., Lim, C. S., Yu, N., Geller, D., Wagner, M. H., Quittner, A. L., A multi-method assessment of treatment adherence for children with cystic fibrosis, Journal of Cystic Fibrosis, 5, 177-85, 2006	Report adherence rates. No specific psychometric properties or diagnostic accuracy data reported.
Olveira, G., Giron, R. M., Olveira, C., Escobedo, M. C., Espildora, F., Juan, D. S., Gaspar, I., Depression and anxiety in patients with cystic fibrosis in Spain, Journal of Cystic Fibrosis, 9, S96, 2010	Abstract only. No specific psychometric properties or diagnostic accuracy data reported.
Pakhale, S., Armstrong, M., Holly, C., Edjoc, R., Gaudet, E., Aaron, S., Tasca, G., Cameron, W., Balfour, L., Assessment of stigma in patients with cystic fibrosis, BMC Pulmonary Medicine, 14, 76, 2014	Scale not in protocol (stigma scale).
Quittner, A. L., Abbott, J., Georgiopoulos, A. M., Goldbeck, L., Smith, B., Hempstead, S. E., Marshall, B., Sabadosa, K. A., Elborn, S., International Committee on Mental, Health, Epos Trial Study Group, International Committee on Mental Health in Cystic Fibrosis: Cystic Fibrosis Foundation and European Cystic Fibrosis Society consensus statements for screening and treating depression and anxiety, Thorax, 71, 26-34, 2016	Narrative review and expert consensus.
Quittner, A. L., Modi, A. C., Lemanek, K. L., Ievers-Landis, C. E., Rapoff, M. A., Evidence-based assessment of adherence to medical treatments in pediatric psychology, Journal of Pediatric Psychology, 33, 916-936, 2008	Narrative review. Individual relevant studies check for inclusion.
Quittner, A. L., Saez-Flores, E., Barton, J. D., The psychological burden of cystic fibrosis, Current Opinion in Pulmonary Medicine, 22, 187-91, 2016	Narrative review.
Randlesome, K., Bryon, M., Evangeli, M., Developing a measure of eating attitudes and	Assessment tool not in protocol (CFEAB).

Study	Reason for Exclusion
behaviours in cystic fibrosis, Journal of Cystic Fibrosis, 12, 15-21, 2013	
Rodgers, B., Ferholt, J., Cooper, C. L., A screening tool to detect psychosocial adjustment of children with cystic fibrosis, Nursing Research, 23, 420-5, 1974	Assessment tool not in protocol (semi-structure interview, open-ended parent questionnaire and a self-administered teacher questionnaire)
Rohovyk, N., Bober, L., Makukh, H., Rohovyk, M., Evaluation of parents' compliance in the treatment of cystic fibrosis children, Journal of Cystic Fibrosis, 13, S117, 2014	Conference abstract. No specific psychometric properties or diagnostic accuracy data reported.
Sands, D., Sapiejka, E., Mazurek, H., Gaszczyk, G., Use of an electronic monitoring system to generate objective information on patients' adherence to taking treatments of a novel inhaled tobramycin solution (VANTOBRA), Journal of Cystic Fibrosis. Conference: 36th European Cystic Fibrosis Conference Lisbon Portugal. Conference Start, 12	Conference abstract. No specific psychometric properties or diagnostic accuracy data reported.
Sapina-Vivo, R., Ledson, M., Walshaw, M., Assessing medication adherence in adult CF patients: Involvement of the community, Journal of Cystic Fibrosis, 11, S135, 2012	Conference abstract. No specific psychometric properties or diagnostic accuracy data reported.
Sheehan, J., Massie, J., Hay, M., Jaffe, A., Glazner, J., Armstrong, D., Hiscock, H., The natural history and predictors of persistent problem behaviours in cystic fibrosis: a multicentre, prospective study, Archives of Disease in Childhood, 97, 625-31, 2012	Prevalence and predictors of behavioural problems.
Simonton, S., Sherman, A. C., Campbell, D., Reddy, R. M., Kumar, S., O'Brien, C. E., Anderson, P. J., Adherence to airway clearance therapy among adults with cystic fibrosis, Pediatric Pulmonology, 46, 406, 2011	Conference abstract. No specific psychometric properties or diagnostic accuracy data reported.
Siracusa, C. M., Ryan, J., Burns, L., Wang, Y., Zhang, N., Clancy, J. P., Drotar, D., Electronic monitoring reveals highly variable adherence patterns in patients prescribed ivacaftor, Journal of Cystic Fibrosis, 14, 621-626, 2015	Conference abstract. No specific psychometric properties or diagnostic accuracy data reported.
Smith, B. A., Goldbeck, L., Georgiopoulos, A. M., Screening and assessment; psychological interventions; pharmacological interventions, Pediatric Pulmonology, 49, 171-173, 2014	Good clinical practice summary. Abstract only.
Smith, B. A., Modi, A. C., Quittner, A. L., Wood, B. L., Depressive symptoms in children with cystic fibrosis and parents and its effects on adherence to airway clearance, Pediatric Pulmonology, 45, 756-63, 2010	No specific psychometric properties or diagnostic accuracy data reported.
Smith, L. J., Staples, P., Buchanan, S., Lougheed, D., Adult depression screening in cystic fibrosis, Pediatric Pulmonology, 49, 443, 2014	Abstract only. No specific psychometric properties or diagnostic accuracy data reported.
Stawski, M., Auerbach, J. G., Barasch, M., Lerner, Y., Zimin, R., Miller, M. S., Behavioral problems of adolescents with chronic physical illness: a comparison of parent-report and self-report measures, European Child & Adolescent Psychiatry, 4, 14-20, 1995	Assessment tool not in protocol (Child Behaviour Checklist and Youth self-report).
Thompson, R. J., Jr., Hodges, K., Hamlett, K. W., A matched comparison of adjustment in children with	Prevalence data only.

Study	Reason for Exclusion
cystic fibrosis and psychiatrically referred and nonreferred children, Journal of Pediatric Psychology, 15, 745-59, 1990	
Thompson, Robert J., Gustafson, Kathryn E., Hamlett, Kim W., Spock, Alexander, Psychological adjustment of children with cystic fibrosis: The role of child cognitive processes and maternal adjustment, Journal of Pediatric Psychology, 17, 741-755, 1992	No specific psychometric properties or diagnostic accuracy data reported.
Tluczek, A., Henriques, J. B., Brown, R. L., Support for the reliability and validity of a six-item state anxiety scale derived from the State-Trait Anxiety Inventory, Journal of Nursing Measurement, 17, 19-28, 2009	Mixed population.
Walker, L. S., Ford, M. B., Donald, W. D., Cystic fibrosis and family stress: effects of age and severity of illness, Pediatrics, 79, 239-46, 1987	Includes parents/ caregivers.
Wenninger, Kerstin, Weiss, Christa, Wahn, Ulrich, Staab, Doris, Body Image in Cystic Fibrosis - Development of a Brief Diagnostic Scale, Journal of Behavioral Medicine, 26, 81-94, 2003	Assessment tool not in protocol (Body Image Scale).
Westell, S., Dang, K., Thomas, A., Tullis, E., Improving the quality of cystic fibrosis mental health through the introduction of validated screening tools, Pediatric Pulmonology, 49, 394-395, 2014	Abstract only. No specific psychometric properties or diagnostic accuracy data reported.
Wheat Butt, E., Wentz, D., Bitsko, M., Schmidt, J., Improving psychosocial care for patients: Implementing an annual screening program for anxiety and depression, Pediatric Pulmonology, 49, 438-439, 2014	Abstract only. No specific psychometric properties or diagnostic accuracy data reported.
Williams, E., Edgeworth, D., Fantidis, M., Finlayson, F., Button, B. M., Clark, D., Tierney, A. C., Keating, D., Kotsimbos, T., Wilson, J., Patient reported adherence to ivacaftor, Journal of Cystic Fibrosis, 14, S46, 2015	Conference abstract. No specific psychometric properties or diagnostic accuracy data reported.

H.20 Cross-infection control

Study	Reason for Exclusion
Amalfitano, G., Tonolli, E., Favari, F., Perazzoli, C., Cazzola, G., Mastella, G., Piacentini, I., Field inversion gel electrophoresis on Pseudomonas cepacia strains isolated from cystic fibrosis patients, European Journal of Epidemiology, 12, 149-53, 1996	No relevant outcomes.
Antony, B., Cherian, E. V., Boloor, R., Shenoy, K. V., A sporadic outbreak of Burkholderia cepacia complex bacteremia in pediatric intensive care unit of a tertiary care hospital in coastal Karnataka, South India, Indian Journal of Pathology & MicrobiologyIndian J Pathol Microbiol, 59, 197-9, 2016	Indirect population.
Ashish, A., Shaw, M., Winstanley, C., Humphreys, L., Walshaw, M. J., Halting the spread of epidemic pseudomonas aeruginosa in an adult cystic fibrosis centre: a prospective cohort study, JRSM Short Reports, 4, 1, 2013	No relevant comparison.

Study	Reason for Exclusion
Bosshammer, J., Fiedler, B., Gudowius, P., von der Hardt, H., Romling, U., Tummler, B., Comparative hygienic surveillance of contamination with pseudomonads in a cystic fibrosis ward over a 4-year period, Journal of Hospital Infection, 31, 261-74, 1995	No relevant outcomes.
Botzenhart, K., Wolz, C., Doring, G., Cross-colonization and routes of infection assessed with a DNA probe, Antibiotics & Chemotherapy, 44, 8-12, 1991	No relevant intervention.
Bowmer, G., Latchford, G., Duff, A., Denton, M., Dye, L., Lawton, C., Lee, T., Adherence to infection prevention and control guidelines: A vignette-based study of decision-making and risk-taking in young adults with cystic fibrosis, Journal of Cystic Fibrosis, 22, 22, 2016	Survey focuses on hypothetical situations rather than on real life situations.
Bryant, J. M., Grogono, D. M., Greaves, D., Foweraker, J., Roddick, I., Inns, T., Reacher, M., Haworth, C. S., Curran, M. D., Harris, S. R., Peacock, S. J., Parkhill, J., Floto, R. A., Whole-genome sequencing to identify transmission of Mycobacterium abscessus between patients with cystic fibrosis: a retrospective cohort study, Lancet, 381, 1551-60, 2013	Aim not relevant.
Corkill, J. E., Sisson, P. R., Smyth, A., Deveney, J., Freeman, R., Shears, P., Heaf, D., Hart, C. A., Application of pyrolysis mass spectroscopy and SDS-PAGE in the study of the epidemiology of Pseudomonas cepacia in cystic fibrosis, Journal of Medical Microbiology, 41, 106-11, 1994	No relevant outcomes.
De Smet, B., Veng, C., Kruy, L., Kham, C., van Griensven, J., Peeters, C., Ieng, S., Phe, T., Vlieghe, E., Vandamme, P., Jacobs, J., Outbreak of Burkholderia cepacia bloodstream infections traced to the use of Ringer lactate solution as multiple-dose vial for catheter flushing, Phnom Penh, Cambodia, Clinical Microbiology & Infection, 19, 832-7, 2013	No relevant setting (no Western country).
Dedeckova, K., Fila, L., Skalicka, V., Bartosova, J., Kucerova, T., Vavrova, V., Zemkova, D., Kalferstova, L., Melter, O., Cinek, O., Drevinek, P., PCR detection of Burkholderia cepacia complex as one of key factors to handle a long-term outbreak, Journal of Cystic Fibrosis, 11, 440-5, 2012	No relevant comparison.
Doe, S. J., McSorley, A., Isalska, B., Kearns, A. M., Bright-Thomas, R., Brennan, A. L., Webb, A. K., Jones, A. M., Patient segregation and aggressive antibiotic eradication therapy can control methicillin-resistant Staphylococcus aureus at large cystic fibrosis centres, Journal of Cystic Fibrosis, 9, 104-9, 2010	Study design does not fit protocol.
Doring, G., Staphylococcus aureus in cystic fibrosis, Pediatric Pulmonology, 24, 235-236, 1997	Discussion paper.

Study	Reason for Exclusion
Driessche, K. V., Hens, N., Tilley, P., Quon, B. S., Chilvers, M. A., de Groot, R., Cotton, M. F., Marais, B. J., Speert, D. P., Zlosnik, J. E., Surgical masks reduce airborne spread of Pseudomonas aeruginosa in colonized patients with cystic fibrosis, American Journal of Respiratory & Critical Care Medicine, 192, 897-9, 2015	No relevant outcomes.
Duff, A. J. A., Psychological consequences of segregation resulting from chronic Burkholderia cepacia infection in adults with CF, Thorax, 57, 756-758, 2002	Discussion paper / narrative review.
Elborn, J. S., Hodson, M., Bertram, C., Implementation of European standards of care for cystic fibrosis--control and treatment of infection, Journal of Cystic Fibrosis, 8, 211-7, 2009	No relevant outcomes: focus on adherence of centres instead of adherence of staff.
Farrell, P. M., Shen, G., Splaingard, M., Colby, C. E., Laxova, A., Kosorok, M. R., Rock, M. J., Mischler, E. H., Acquisition of Pseudomonas aeruginosa in children with cystic fibrosis, Pediatrics, 100, E2, 1997	Better quality evidence (Hayes 2010 study) has been included on cohort segregation in the outpatient setting and the incidence of PA. With regards to prevalence of PA, the analysis is cross-sectional.
Festini, F., Buzzetti, R., Bassi, C., Braggion, C., Salvatore, D., Taccetti, G., Mastella, G., Isolation measures for prevention of infection with respiratory pathogens in cystic fibrosis: a systematic review, Journal of Hospital Infection, 64, 1-6, 2006	Quality of the systematic review is low according to the AMSTAR checklist. Included studies assessed for potential inclusion.
Fothergill, J. L., Walshaw, M. J., Winstanley, C., Transmissible strains of Pseudomonas aeruginosa in cystic fibrosis lung infections, European Respiratory Journal, 40, 227-38, 2012	Narrative review.
Garber, E., Desai, M., Zhou, J., Alba, L., Angst, D., Cabana, M., Saiman, L., C. F. Infection Control Study Consortium, Barriers to adherence to cystic fibrosis infection control guidelines, Pediatric Pulmonology, 43, 900-7, 2008	No relevant intervention.
Geddes, D., Segregation is not good for patients with cystic fibrosis, Journal of the Royal Society of Medicine, 101 Suppl 1, S36-8, 2008	Discussion paper.
Govan, J. R., Infection control in cystic fibrosis: methicillin-resistant Staphylococcus aureus, Pseudomonas aeruginosa and the Burkholderia cepacia complex, Journal of the Royal Society of Medicine, 93 Suppl 38, 40-5, 2000	Narrative review.
Govan, J. R., Brown, P. H., Maddison, J., Doherty, C. J., Nelson, J. W., Dodd, M., Greening, A. P., Webb, A. K., Evidence for transmission of Pseudomonas cepacia by social contact in cystic fibrosis, Lancet, 342, 15-9, 1993	Relevant intervention is only mentioned in the discussion, without mentioning when it was introduced.
Hoiby, N., Isolation and treatment of cystic fibrosis patients with lung infections caused by Pseudomonas (Burkholderia) cepacia and multiresistant Pseudomonas aeruginosa,	Narrative review.

Study	Reason for Exclusion
Netherlands Journal of Medicine, 46, 280-7, 1995	
Holland, A. E., Button, B. M., International Physiotherapy Group for Cystic Fibrosis, Australian Chapter, Physiotherapy for cystic fibrosis in Australia: knowledge and acceptance of the Consensus Statement recommendations, Respirology, 18, 652-6, 2013	Unclear whether relevant interventions were being implemented at the centres/clinics where respondents worked.
Johansen, H. K., Kovesi, T. A., Koch, C., Corey, M., Hoiby, N., Levison, H., Pseudomonas aeruginosa and Burkholderia cepacia infection in cystic fibrosis patients treated in Toronto and Copenhagen, Pediatric Pulmonology, 26, 89-96, 1998	Cross-sectional study design.
Johansen, H. K., Moskowitz, S. M., Ciofu, O., Pressler, T., Hoiby, N., Spread of colistin resistant non-mucoid Pseudomonas aeruginosa among chronically infected Danish cystic fibrosis patients, Journal of Cystic Fibrosis, 7, 391-7, 2008	No relevant outcomes.
Jorgensen, I. M., Johansen, H. K., Frederiksen, B., Pressler, T., Hansen, A., Vandamme, P., Hoiby, N., Koch, C., Epidemic Spread of Pandoraea apista, a New Pathogen Causing Severe Lung Disease in Cystic Fibrosis Patients, Pediatric Pulmonology, 36, 439-446, 2003	Study design does not fit protocol (study dates and date of isolation regime are unclear).
Kalferstova, L., Vilimovska Dedeckova, K., Antuskova, M., Melter, O., Drevinek, P., How and why to monitor Pseudomonas aeruginosa infections in the long term at a cystic fibrosis centre, Journal of Hospital InfectionJ Hosp Infect, 92, 54-60, 2016	No relevant intervention.
Kapnadak, S. G., Hisert, K. B., Pottinger, P. S., Limaye, A. P., Aitken, M. L., Infection control strategies that successfully controlled an outbreak of Mycobacterium abscessus at a cystic fibrosis center, American Journal of Infection Control, 44, 154-9, 2016	Study design does not fit protocol (pre-intervention incidence/prevalence not reported).
Koch, C., Frederiksen, B., Hoiby, N., Patient Cohorting and Infection Control, Seminars in Respiratory and Critical Care Medicine, 24, 703-715, 2003	Narrative review.
Kosorok, M. R., Jalaluddin, M., Farrell, P. M., Shen, G., Colby, C. E., Laxova, A., Rock, M. J., Splaingard, M., Comprehensive analysis of risk factors for acquisition of Pseudomonas aeruginosa in young children with cystic fibrosis, Pediatric Pulmonology, 26, 81-8, 1998	No relevant findings.
LiPuma, J. J., Mortensen, J. E., Dasen, S. E., Edlind, T. D., Schidlow, D. V., Burns, J. L., Stull, T. L., Ribotype analysis of Pseudomonas cepacia from cystic fibrosis treatment centers, Journal of Pediatrics, 113, 859-62, 1988	No relevant intervention.
Luna, R. A., Millecker, L. A., Webb, C. R., Mason, S. K., Whaley, E. M., Starke, J. R., Hiatt, P. W., Versalovic, J., Molecular epidemiological surveillance of multidrug-resistant Pseudomonas	No relevant data.

Study	Reason for Exclusion
aeruginosa isolates in a pediatric population of patients with cystic fibrosis and determination of risk factors for infection with the Houston-1 strain, Journal of Clinical Microbiology, 51, 1237-40, 2013	
Madge, S., Bell, S. C., Burgel, P. R., De Rijcke, K., Blasi, F., Elborn, J. S., Limitations to providing adult cystic fibrosis care in Europe: Results of a care centre survey, Journal of Cystic Fibrosis, no pagination, 2016	No relevant outcomes: focus on infection control policies of centres rather than on adherence of staff.
Marchant, J. L., Little, S. A., Bush, A., Flow volume curves in children with cystic fibrosis--a possible risk of cross-infection?, Respiratory Medicine, 88, 235-6, 1994	Aim not relevant.
Marshall, B. C., Liou, T. G., Elusiveness of ideal approach to Pseudomonas aeruginosa infection complicating cystic fibrosis, Lancet, 356, 613-4, 2000	Discussion paper.
Masterson, T. L., Wildman, B. G., Newberry, B. H., Omlor, G. J., Impact of age and gender on adherence to infection control guidelines and medical regimens in cystic fibrosis, Pediatric Pulmonology, 46, 295-301, 2011	No relevant aim.
Masterson, T., Wildman, B. G., Newberry, B., Omlor, G., Bryson, E., Kukay, A., Compliance in cystic fibrosis: an examination of infection control guidelines, Pediatric Pulmonology, 43, 435-42, 2008	No relevant aim.
McCallum, S. J., Corkill, J., Gallagher, M., Ledson, M. J., Hart, C. A., Walshaw, M. J., Superinfection with a transmissible strain of Pseudomonas aeruginosa in adults with cystic fibrosis chronically colonised by P aeruginosa, Lancet, 358, 558-60, 2001	No evaluation of relevant intervention.
McCarthy,C.A., Hall,C.B., Respiratory syncytial virus: Concerns and control, Pediatrics in Review, 24, 301-308, 2003	Narrative review.
Miroballi, Y., Garber, E., Jia, H., Zhou, J. J., Alba, L., Quittell, L. M., Angst, D., Cabana, M., Saiman, L., C. F. Infection Control Study Consortium, Infection control knowledge, attitudes, and practices among cystic fibrosis patients and their families, Pediatric Pulmonology, 47, 144-52, 2012	No relevant data.
Morris, D., Fallon, L., Heaf, L., Burrows, E., Wallace, H., McNamara, P., Winstanley, C., Southern, K. W., A REDUCING PREVALENCE OF THE LIVERPOOL EPIDEMIC STRAIN OF PSEUDOMONAS AERUGINOSA IN CHILDREN ATTENDING THE INDEX PAEDIATRIC CLINIC: 324, Pediatric Pulmonology, 44 Supplement, 325, 2009	Conference abstract no RCT.
Muhdi, K., Edenborough, F. P., Gumery, L., O'Hickey, S., Smith, E. G., Smith, D. L., Stableforth, D. E., Outcome for patients colonised with Burkholderia cepacia in a	Insufficient reporting relating to relevant outcomes.

Study	Reason for Exclusion
Birmingham adult cystic fibrosis clinic and the end of an epidemic, Thorax, 51, 374-7, 1996	
O'Connell, N., Improving the management of cystic fibrosis, Hospital Pharmacist, 9, 114, 2002	Report on symposium and narrative review.
O'Malley, C. A., Infection control in cystic fibrosis: cohorting, cross-contamination, and the respiratory therapist, Respiratory Care, 54, 641-57, 2009	Narrative review.
Pant, C., Deshpande, A., Gilroy, R., Olyaee, M., Donskey, C. J., Rising Incidence of Clostridium difficile Related Discharges among Hospitalized Children in the United States, Infection Control & Hospital EpidemiologyInfect Control Hosp Epidemiol, 37, 104-6, 2016	No relevant intervention
Pant, C., Sferra, T. J., Deshpande, A., Olyaee, M., Gilroy, R., Anderson, M. P., Donskey, C., Clostridium difficile infection in hospitalized patients with cystic fibrosis, Infection Control & Hospital Epidemiology, 35, 1547-8, 2014	Aim not relevant.
Parkins, M. D., Floto, R. A., Emerging bacterial pathogens and changing concepts of bacterial pathogenesis in cystic fibrosis, Journal of Cystic Fibrosis, 14, 293-304, 2015	Narrative review.
Paul, M. L., Pegler, M. A., Benn, R. A., Molecular epidemiology of Burkholderia cepacia in two Australian cystic fibrosis centres, Journal of Hospital Infection, 38, 19-26, 1998	No relevant outcomes.
Pedersen, S. S., Jensen, T., Hoiby, N., Koch, C., Flensborg, E. W., Management of Pseudomonas aeruginosa lung infection in Danish cystic fibrosis patients, Acta Paediatrica Scandinavica, 76, 955-61, 1987	Relevant data are only reported graphically.
Pedersen, S. S., Jensen, T., Pressler, T., Hoiby, N., Rosendal, K., Does centralized treatment of cystic fibrosis increase the risk of Pseudomonas aeruginosa infection?, Acta Paediatrica Scandinavica, 75, 840-5, 1986	No relevant intervention.
Pedersen, S. S., Koch, C., Hoiby, N., Rosendal, K., An epidemic spread of multiresistant Pseudomonas aeruginosa in a cystic fibrosis centre, Journal of Antimicrobial Chemotherapy, 17, 505-16, 1986	Relevant data are only reported graphically.
Prior, A. R., Gunaratnam, C., Humphreys, H., Ralstonia species - do these bacteria matter in cystic fibrosis?, Paediatric Respiratory Reviews, 21, 21, 2016	Narrative review.
Ramsay, K. A., Butler, C. A., Paynter, S., Ware, R. S., Kidd, T. J., Wainwright, C. E., Bell, S. C., Factors influencing acquisition of Burkholderia cepacia complex organisms in patients with cystic fibrosis, Journal of Clinical Microbiology, 51, 3975-80, 2013	Study design does not fit protocol.
Ramsey, A. H., Skonieczny, P., Coolidge, D. T., Kurzynski, T. A., Proctor, M. E., Davis, J. P., Burkholdria cepacia lower respiratory tract infection associated with exposure to a	No relevant intervention.

Study	Reason for Exclusion
respiratory therapist, Infection Control and Hospital Epidemiology, 22, 423-426, 2001	
Razvi, S., Quittell, L., Sewall, A., Quinton, H., Marshall, B., Saiman, L., Respiratory microbiology of patients with cystic fibrosis in the United States, 1995 to 2005, Chest, 136, 1554-60, 2009	Aim not relevant.
Ryley, H. C., Doull, I. J. M., Burkhoideria cepacia complex infection in patients with cystic fibrosis: Laboratory investigations, epidemiology and clinical management, Reviews in Medical Microbiology, 14, 15-24, 2003	Narrative review.
Saiman, L., Siegel, J. D., LiPuma, J. J., Brown, R. F., Bryson, E. A., Chambers, M. J., Downer, V. S., Fliege, J., Hazle, L. A., Jain, M., Marshall, B. C., O'Malley, C., Pattee, S. R., Potter-Bynoe, G., Reid, S., Robinson, K. A., Sabadosa, K. A., Schmidt, H. J., Tullis, E., Webber, J., Weber, D. J., Cystic Fibrous, Foundation, Society for Healthcare Epidemiology of, America, Infection prevention and control guideline for cystic fibrosis: 2013 update, Infection Control & Hospital Epidemiology, 35 Suppl 1, S1-S67, 2014	Recommendations and systematic review. Quality of the systematic review is low according to the AMSTAR checklist. Included studies were assessed for potential inclusion.
Saiman, L., Siegel, J., Cystic Fibrosis Foundation Consensus Conference on Infection Control, Participants, Infection control recommendations for patients with cystic fibrosis: Microbiology, important pathogens, and infection control practices to prevent patient-to-patient transmission, American Journal of Infection Control, 31, S1-62, 2003	Consensus document including a narrative review of the evidence.
Salsgiver, E. L., Fink, A. K., Knapp, E. A., LiPuma, J. J., Olivier, K. N., Marshall, B. C., Saiman, L., Changing Epidemiology of the Respiratory Bacteriology of Patients With Cystic Fibrosis, ChestChest, 149, 390-400, 2016	No evaluation of relevant intervention.
Sanders, D. B., Emerson, J., Ren, C. L., Schechter, M. S., Gibson, R. L., Morgan, W., Rosenfeld, M., Epic Study Group, Early Childhood Risk Factors for Decreased FEV1 at Age Six to Seven Years in Young Children with Cystic Fibrosis, Annals of the American Thoracic SocietyAnn Am Thorac Soc, 12, 1170-6, 2015	No relevant intervention.
Schaffer, K., Epidemiology of infection and current guidelines for infection prevention in cystic fibrosis patients, Journal of Hospital Infection, 89, 309-13, 2015	Narrative review.
Shepherd, S. L., Goodrich, E. J., Desch, J., Quinton, P. M., Counterpoint: Does the risk of cross infection warrant exclusion of adults with cystic fibrosis from cystic fibrosis foundation events? No, Chest, 145, 680-683, 2014	Discussion paper.
Siracusa, C. M., Weiland, J. L., Acton, J. D., Chima, A. K., Chini, B. A., Hoberman, A. J., Wetzel, J. D., Amin, R. S., McPhail, G. L., The impact of transforming healthcare delivery on	No relevant outcomes.

Study	Reason for Exclusion
cystic fibrosis outcomes: a decade of quality improvement at Cincinnati Children's Hospital, BMJ Quality & Safety, 23 Suppl 1, i56-i63, 2014	
Somayaji, R., Waddell, B., Workentine, M. L., Surette, M. G., Brager, N. P., Rabin, H. R., Parkins, M. D., Infection control knowledge, beliefs and behaviours amongst cystic fibrosis patients with epidemic Pseudomonas aeruginosa, BMC Pulmonary Medicine, 15, 138, 2015	No relevant findings.
Spicuzza, L., Sciuto, C., Vitaliti, G., Di Dio, G., Leonardi, S., La Rosa, M., Emerging pathogens in cystic fibrosis: ten years of follow-up in a cohort of patients, European Journal of Clinical Microbiology & Infectious Diseases, 28, 191-5, 2009	No relevant intervention.
Steinkamp, G., Stahl, K., Ellemunter, H., Heuer, E., Van Koningsbruggen-Rietschel, S., Busche, M., Bremer, W., Schwarz, C., Cystic fibrosis (CF) care through the patients' eyes - A nationwide survey on experience and satisfaction with services using a disease-specific questionnaire, Respiratory Medicine, 109, 79-87, 2015	No relevant data.
Steinkamp, G., Stahl, K., Ellemunter, H., Heuer, E., van Koningsbruggen-Rietschel, S., Busche, M., Bremer, W., Schwarz, C., Patient Experience Working, Group, Cystic fibrosis (CF) care through the patients' eyes - a nationwide survey on experience and satisfaction with services using a disease-specific questionnaire, Respiratory MedicineRespir Med, 109, 79-87, 2015	No relevant intervention.
Steinkamp, G., Ullrich, G., Different opinions of physicians on the importance of measures to prevent acquisition of Pseudomonas aeruginosa from the environment, Journal of Cystic Fibrosis, 2, 199-205, 2003	Unclear whether relevant interventions were being implemented at the centres where respondents worked.
Tummler, B., Koopmann, U., Grothues, D., Weissbrodt, H., Steinkamp, G., von der Hardt, H., Nosocomial acquisition of Pseudomonas aeruginosa by cystic fibrosis patients, Journal of Clinical Microbiology, 29, 1265-7, 1991	No relevant outcomes.
Vonberg, R. P., Gastmeier, P., Isolation of infectious cystic fibrosis patients: results of a systematic review, Infection Control & Hospital Epidemiology, 26, 401-9, 2005	Quality of the systematic review is low according to the AMSTAR checklist. Included studies assessed for potential inclusion.
Waters, V., Ratjen, F., Multidrug-resistant organisms in cystic fibrosis: management and infection-control issues, Expert Review of Antiinfective Therapy, 4, 807-19, 2006	Narrative review.
Wiehlmann, L., Cramer, N., Ulrich, J., Hedtfeld, S., Weissbrodt, H., Tummler, B., Effective prevention of Pseudomonas aeruginosa cross-infection at a cystic fibrosis centre - results of a 10-year prospective study, Ijmm International	Study design does not fit protocol.

Study	Reason for Exclusion
Journal of Medical Microbiology, 302, 69-77, 2012	
Wood, M. E., Stockwell, R., Johnson, G., Ramsay, K., Sherrard, L., Jabbour, N., Knibbs, L., Kidd, T., Wainwright, C. E., Morawska, L., Bell, S. C., Face masks and cough etiquette reduce cough-generated bioaerosols containing pseudomonas aeruginosa in patients with cystic fibrosis, Pediatric Pulmonology. Conference: 30th Annual North American Cystic Fibrosis Conference. United States. Conference Start, 51, 2016	No relevant comparison (comparing N95 mask to surgical mask) in relation to relevant outcome (patient comfort); conference abstract of crossover trial.
Zhou, J., Garber, E., Saiman, L., Survey of infection control policies for patients with cystic fibrosis in the United States, American Journal of Infection Control, 36, 220-2, 2008	No relevant outcomes: focus on adherence of centres instead of adherence of staff.
Zimakoff, J., Hoiby, N., Rosendal, K., Guilbert, J. P., Epidemiology of Pseudomonas aeruginosa infection and the role of contamination of the environment in a cystic fibrosis clinic, Journal of Hospital Infection, 4, 31-40, 1983	No relevant outcomes.
Zlosnik, J. E., Zhou, G., Brant, R., Henry, D. A., Hird, T. J., Mahenthiralingam, E., Chilvers, M. A., Wilcox, P., Speert, D. P., Burkholderia species infections in patients with cystic fibrosis in British Columbia, Canada. 30 years' experience, Annals of the American Thoracic Society, 12, 70-8, 2015	No relevant comparison.
Zuckerman, J. B., Clock, S. A., Prato, B. S., McDevitt, J. J., Zhou, J. J., Leclair, L. W., Lucas, F. L., Saiman, L., Air contamination with bacteria in cystic fibrosis clinics: implications for prevention strategies, American Journal of Respiratory & Critical Care Medicine, 191, 598-601, 2015	No relevant outcomes.

H.21 Health economics excluded studies

Study	Reason for Exclusion
Angelis, A., Kanavos, P., Lopez-Bastida, J., Linertova, R., Nicod, E., Serrano-Aguilar, P., Burqol-Rd Research Network, Social and economic costs and health-related quality of life in non-institutionalised patients with cystic fibrosis in the United Kingdom, BMC Health Services ResearchBMC Health Serv Res, 15, 428, 2015	Irrelevant. Cost of illness study that takes a societal perspective.
Brown, A, Skidmore, B, Hypertonic saline nebules for patients with cystic fibrosis and bronchioectasis: a review of the clinical and cost-effectiveness (Structured abstract), Health Technology Assessment Database, 2015	Review of the literature, no further studies identified.
Brown, TER, Glennie, JL, Carleton, BC, A pharmacoeconomic evaluation of DNase in cystic fibrosis, Pharmacotherapy, 16, 134-5, 1996	Conference paper with insufficient detail. Pre-dates inclusion criteria for conference abstracts and higher quality full-papers identifed in this area.

Study	Reason for Exclusion
CADTH, Sodium chloride inhalation for the treatment of cystic fibrosis: a review of the clinical evidence, cost-effectiveness and guidelines (Structured abstract), Health Technology Assessment Database, 2016	Review of the literature, no further studies identified.
CADTH, Inhaled tobramycin versus intravenous tobramycin for patients with cystic fibrosis: a review of the clinical effectiveness, cost effectiveness, and guidelines (Structured abstract), Health Technology Assessment Database, 2016	Review of the literature, no further studies identified.
CADTH, Dornase alfa for patients with cystic fibrosis: a review of the clinical efficacy and cost-effectiveness (Structured abstract), Health Technology Assessment Database, 2016	Review of the literature, no further studies identified.
Chevreul, K., Berg Brigham, K., Michel, M., Rault, G., Burqol-Rd Research Network, Costs and health-related quality of life of patients with cystic fibrosis and their carers in France, Journal of Cystic FibrosisJ Cyst Fibros, 14, 384-91, 2015	Irrelevant. Non-UK cost of illness study.
Dewitt, E. M., Grussemeyer, C. A., Friedman, J. Y., Dinan, M. A., Lin, L., Schulman, K. A., Reed, S. D., Resource use, costs, and utility estimates for patients with cystic fibrosis with mild impairment in lung function: analysis of data collected alongside a 48-week multicenter clinical trial, Value in Health, 15, 277-83, 2012	Irrelevant. Cost of illness study that includes indirect costs from missed school and/or work days.
Goa, K. L., Lamb, H., Dornase alfa. A review of pharmacoeconomic and quality-of-life aspects of its use in cystic fibrosis, Pharmacoeconomics, 12, 409-22, 1997	Review of the literature, no further studies identified.
Gu, Y., Garcia-Perez, S., Massie, J., van Gool, K., Cost of care for cystic fibrosis: an investigation of cost determinants using national registry data, European Journal of Health Economics, 16, 709-717, 2015	Irrelevant. Non-UK cost of illness study.
Kane, R. E., Jennison, K., Wood, C., Black, P. G., Herbst, J. J., Cost savings and economic considerations using home intravenous antibiotic therapy for cystic fibrosis patients, Pediatric Pulmonology, 4, 84-9, 1988	Irrelevant. US study that does not reflect UK health care system today.All patients underwent private home-care IV therapy.
LeLorier, J., Perreault, S., Birnbaum, H., Greenberg, P., Sheehy, O., Savings in direct medical costs from the use of tobramycin solution for inhalation in patients with cystic fibrosis, Clinical Therapeutics, 22, 140-51, 2000	US data extrapolated to a Canadian setting reducing the directness of their analysis to the UK. Not a full cost-effectiveness analysis as only medical savings are calculated. The cost of tobramycin is also excluded.
National Horizon Scanning Centre, Tobramycin inhaled powder (TIP) for Pseudomonas aeruginosa infection in cystic fibrosis (Structured abstract), Health Technology Assessment Database, 2015	Not a cost-effectiveness analysis. Report the estimated cost and cost impact (BNF acquisition cost) of colistin IV (colomycin), colistin nebuliser (promixin) and tobramycin nebuliser (TOBI and Bramitob) based on 28 day dose.
National Horizon Scanning Centre, Mannitol dry powder for inhalation (Bronchitol) for cystic fibrosis (Structured abstract), Health Technology Assessment Database, 2015	Not a cost-effectiveness analysis. Report the cost impact "the cost of mannitol dry powder is currently unknown".

Study	Reason for Exclusion
National Horizon Scanning Centre, Colistimethate sodium powder for inhalation (Colobreathe) for pseudomonas aeruginosa lung infection in cystic fibrosis (Structured abstract), Health Technology Assessment Database, 2015	Not a cost-effectiveness analysis. Report the estimated cost and cost impact (BNF acquisition cost) of colistin IV (colomycin), colistin nebuliser (promixin) and tobin nebuliser based on 28 day dose.
NIHR, HSC, Lumacaftor and ivacaftor combination therapy for cystic fibrosis - first line (Structured abstract), Health Technology Assessment Database, 2015	Not a cost-effectiveness analysis and not a comparator of interest in the final protocols.
Oster, G., Huse, D. M., Lacey, M. J., Regan, M. M., Fuchs, H. J., Effects of recombinant human DNase therapy on healthcare use and costs in patients with cystic fibrosis, Annals of Pharmacotherapy, 29, 459-64, 1995	Cost-benefit analysis reporting the costs of RTI-related inpatient and outpatient care for daily rhDNASe, alternate day rhDNase and placebo. The cost of rhDNase therapy was not included, as it was not being marketed at the time the assessment was undertaken (1995). As US practice and the use of antibiotics is likely to vary considerably from that in the UK, any savings resulting from the decreased use of antibiotics cannot be generalizable to the UK. Overall it was considered reasonable to exclude this study because higher quality UK evidence on the cost effectiveness of rhDNase has been included for this review question.
Purins, A, Hiller, JE, Hypertonic saline therapy for cystic fibrosis (Structured abstract), Health Technology Assessment Database, 2015	Irrelevant. Not a cost-effectiveness analysis - report the expected cost impact.
Robson, M., Abbott, J., Webb, K., Dodd, M., Walsworth-Bell, J., A cost description of an adult cystic fibrosis unit and cost analyses of different categories of patients, Thorax, 47, 684-9, 1992	Irrelevant. Not a cost-effectiveness analysis - UK delivery of care costing study.
Suri,R., Wallis,C., Bush,A., Thompson,S., Normand,C., Flather,M., Grieve,R., Metcalfe,C., Lees,B., A comparative study of hypertonic saline, daily and alternate-day rhDNase in children with cystic fibrosis, Health Technology Assessment, 6, -, 2002	Duplicate. Methods and results also reported by: Suri R, Grieve R, Normand C, Metcalfe C, Thompson S, Wallis C, Bush A. â□œEffects of hypertonic saline, alternate day and daily rhDNase on healthcare use, costs and outcomes in children with cystic fibrosis.â□□ Thorax. 2002: Oct;57(10):841-6.
Weiner, J. R., Toy, E. L., Sacco, P., Duh, M. S., Costs, quality of life and treatment compliance associated with antibiotic therapies in patients with cystic fibrosis: a review of the literature, Expert Opinion on Pharmacotherapy, 9, 751-66, 2008	Review of the literature, no further studies identified.
Whiting, P., Al, M., Burgers, L., Westwood, M., Ryder, S., Hoogendoorn, M., Armstrong, N., Allen, A., Severens, H., Kleijnen, J., Ivacaftor for the treatment of patients with cystic fibrosis and the G551D mutation: a systematic review and cost-effectiveness analysis, Health Technology Assessment (Winchester, England), 18, 1-106, 2014	Irrelevant. Not a population or comparator of interest in the final protocols.