G.3 Service delivery

G.3.1 Service configuration

Review question: Service configuration: What is the effectiveness of different models of care (for example, specialist centre, shared care [delivered by a Network CF Clinic which is part of an agreed designated network with a Specialist CF Centre], community, telehealth and/or home care for people with CF?

Study details	Participants	Interventions	Methods	Outcomes and Results	Comments
Full citation Bosworth, D. G., Nielson, D. W., Effectiveness of home versus hospital care in the routine treatment of cystic fibrosis, Pediatric Pulmonology, 24, 42-7, 1997 Ref Id 330443 Country/ies where the study was carried out USA Study type	Sample size N= 40 patients 19 home group 21 hospital group Subgroup: N=5 patients in the hospital+home group N= 59 courses 27 in home group 32 in hospital group Subgroup: N=12 courses in the hospital+home group 6 home group 6 hospital group Characteristics	Interventions Intervention Patients and families administered home IV antibiotics and chest physiotherapy at home. Prior to receiving home care, patients stayed in the hospital for up to 4 days. Nurses employed by a home care company visited the patients at home at least once a week. Approved companies responded to any	Details Setting Inter mountain Cystic Fibrosis Centre at the University of Utah. Analysis Data was analysed using the t-test, paired sample t-test, and Fisher's exact test as	Results FEV1 at 10-14 days: % change (mean (SEM)): Home (n=27) 13.7 (2.6) (p value=0.11) vs hospital (n=32) 23.3 (4.1) (p value <0.001) Subgroup hospital+home patients (Patients who received both home and hospital IV antibiotic therapy): Home (n=6) 11.2(11.0) (p value 0.12) vs hospital (n=6): 28.6 (2.7) (p value=0.007) Mortality: Not reported	Limitations The quality of this study was assessed using the Newcastle-Ottawa scale assessment tool: Selection: Low risk (Requirements for patients to receive home treatment included the availability of family members to deliver care, financial feasibility, and their demonstrated ability to perform care. This is likely to be representative of the home care population in the UK, except perhaps for financial feasibility)

Study details	Participants	Interventions	Methods	Outcomes and Results	Comments
Comparative cohort study Aim of the study To compare the outcomes of home care with minimal supervision to outcomes of hospital care Study dates Study on patients attending the Intermountain CF Centre over 2 years covered by the study (dates not specified) Source of funding Not mentioned	Confirmed diagnosis of CF Pulmonary exacerbation Age range: Home 7-31 vs hospital 8-29 Age (mean(SEM)): Home 18.8 (1.2) vs Hospital 17.5 (0.9), p value 0.35 Male/female (patients): Home 7/12 vs Hospital 13/8 Male/female (courses): Home 10/17 vs Hospital 20/12 FEV1 (% predicted): Home 40.6 (3.1) vs Hospital 46.0 (3.3), p value 0.24 Percent decrease in FEV1 from best measurement in the year preceding treatment: Home -18.4 (3.6) vs Hospital -21.7 (4.8) Weight (kg): Home 44.6 (2.3) vs Hospital 46.2 (2.0), p value 0.63 Inclusion criteria Care provided bythe Intermountain Cystic Fibrosis Centre at the University of Utah	problems concerning the IV line or antibiotic preparations on a 24 hour basis. Weekly tobramycin serum concentrations were used to adjust the dose Patients were advised to continue physiotherapy at home with the same frequency as in the hospital. Comparison IV antibiotics administered at the hospital. Weekly tobramycin serum concentrations were used to adjust the dose Patients received chest physiotherapy four times a day while in the hospital.	appropriate to each data set. Baseline characteristics were compared with a t-test. Changes in FEV1 between groups were compared with a paired t-test. Time to next exacerbations as a quantitative variable was compared between groups with a t-test. Time to next exacerbations as a categorical variable (did patients start the next course of IV antibiotics more than 12 weeks after completing the previous course: YES/NO)	Patient and carer satisfaction Not reported LCI Not reported Time to next pulmonary exacerbation Quantitative variable (weeks between the end of the treatment course and the start of the next IV antibiotic course): Mean (SEM): home 15.1(3.3) vs hospital 23.1(3.0) (The difference did not achieve statistical significance) Categorical variable (did patients start the next course of IV antibiotics more than 12 weeks after completing the previous course: YES/NO): home (n=27) 13/14 vs hospital (n=32) 28/4, p<0.01 Subgroup hospital+home patients (YES/NO): home (n=6) 1/5 vs hospital (n=6) 6/0, p<0.01 Nutritional status Not reported Quality of life Not reported Frequency of cross-infections (pseudomonas, b.cepacia) Not reported Staff experience	Comparability: Low risk (The groups were matched by age and lung function) Outcome: Unclear for FEV1% pred and time to next exacerbation (Authors specify that patients were evaluation at the CF Clinic or the hospital before treatment but do not mention how evaluation after treatment was carried out). High risk for adherence (self-reported) Overall quality: moderate Other information

Study details	Participants	Interventions	Methods	Outcomes and Results	Comments
	Sputum culture positive for Pseudomonas aeruginosa alone or in combination with S. aureus Patients able to perform spirometry Patients of comparable ages Exclusion criteria Patients with incomplete charts No one in the comparison group of similar age and similar lung function Cases in which the patients stayed in the hospital for more than 4 days and then finished their course of IV antibiotics at home		was compared between groups with a Fisher's exact test.	Not reported Adherence to treatment Adherence (Mean (SEM)) (frequency of chest physiotherapy): home 2.4(1.2) days vs hospital 4.0(0.2) days, p<0.01	
Full citation Donati, M. A., Guenette, G., Auerbach, H., Prospective controlled study of home and hospital therapy of cystic fibrosis pulmonary disease, Journal of Pediatrics, 111, 28-33, 1987 Ref Id	Sample size 64 patents (82 treatments) home group: 26 patients (41 treatments) hospital group: 38 patients (41 treatments) Characteristics	Interventions Intervention Nurses made an initial visit within 24 hours of a patient's enrollment into the home IV program, and daily thereafter. During the visits, intravenous catheters were inserted when needed, clinical status was assessed, and patient/family	Details Setting CF Clinic at the Children's Hospital in Boston, US. Data collection Spirometry was carried out with a 9 L seal spirometer.	Results FEV1 (% predicted) at admission and on discharge (at 18 days): Mean +/- SEM: home (n=31) admission 43.5+/-4.0 discharge 50.2+/-4.2 (p value 0.005) vs hospital (n=32) admission 37.5+/-2.7 vs hospital 49.8+/-3.8 (p value <0.001) Mortality	Limitations The quality of this study was assessed using the Newcastle-Ottawa scale assessment tool: Selection: Low risk (Eligibility criteria for home treatment included >= 1 hour drive form the hospital but this is unlikely to affect differences in outcomes between the home and hospital group)

Study details	Participants	Interventions	Methods	Outcomes and Results	Comments
363900 Country/ies where the study was carried out USA Study type Controlled Prospective Clinical Trial Aim of the study To compare the efficacy and benefits of home and hospital treatment for patients with exacerbations of pulmonary disease caused by cystic fibrosis. Study dates 1984-1986: data collection 1987: publication date Source of funding Not reported	Age(yr) (Mean+-SEM): Home 23.3+-0.90 vs Hospital 23.3+-100 Inclusion criteria Confirmed diagnosis of CF ≥12 years or older Required IV antibiotic therapy for a pulmonary exacerbation Exclusion criteria Not reported	competence and comfort with the home care regimen were evaluated. Medical backup was provided by the attending physician, and all home care cases were presented at weekly multidisciplinary rounds. Antibiotics were chosen on the basis of results of sputum cultures and sensitivities obtained prior to admission Comparison IV antibiotics administered at the hospital Antibiotics were chosen on the basis of results of sputum cultures and sensitivities obtained prior to admission	All values were obtained at initiation of treatment and on discharge Data analysis The Student t tests for paired and independent samples were applied. In addition, the nonparametric Wilcoxon matched-pairs signed rank and Mann-Whitney U tests were applied. When no discrepancies were found, only those obtained from the Student t test for paired samples are reported.	Not reported Patient and carer satisfaction Not reported LCI Not reported Time to next exacerbation: Intervals between IV antibiotic treatments, months (Mean (SEM)): Home over 18 months before the study 5.9 (1.9) Home After 4.1(1.1), p <= 0.18; hospital over 18 months before the study 6.2 (1.3) hospital after 7.0 (1.0), p<= 0.48 Nutritional status at admission and on discharge (at 18 days): Weight (kg) Mean (SEM) (37 matched pairs): Home Admission 51.2 (1.9) Home Discharge 51.7 (1.9) vs Hospital Admission 50.4 (1.3) Hospital Discharge 52.0 (1.3) (p value comparing home vs hospital at admission NS, p value comparing home vs hospital on discharge NS) Quality of life Not reported Frequency of cross-infections (pseudomonas, b.cepacia) Not reported Staff experience	Comparablity: Low risk (Home and hospital patients were matched according to sex, age, pulmonary function tests and arterial blood gas values) Outcome: Low risk for weight: 37/41 matched pairs had results both on admission and discharge (90% follow up rate) Low risk for FEV1: 31/41 matched pairs in the home group and 32/41 matched pairs in the hospital group had data both on admission and discharge (76% and 78% follow up rate respectively) Overall quality: High Other information Patients meeting the same eligibility criteria for home treatment except distance to the hospital and who were admitted for IV therapy within 4 weeks of a home care patients served as controls. Home and hospital patients were matched according to sex, age, pulmonary function tests, and arterial blood gas values.

Study details	Participants	Interventions	Methods	Outcomes and Results	Comments
				Not reported Adherence to treatment Not reported	
Full citation Esmond, G., Butler, M., McCormack, A. M., Comparison of hospital and home intravenous antibiotic therapy in adults with cystic fibrosis, Journal of Clinical Nursing, 15, 52-60, 2006 Ref Id 330769 Country/ies where the study was carried out UK Study type Quasi-experimental, prospective study. Aim of the study To compare home and hospital treatment for clinical outcome and quality of life in adult cystic fibrosis patients receiving IV antibiotics for acute respiratory exacerbations. Study dates Six-month period. Dates not mentioned. Source of funding	Sample size N= 28 patients (30 courses of treatments) 15 home courses if IV antibiotics 15 hospital courses of IV antibiotics 13 patients received a hospital course of antibiotics only 13 patients received a home course of antibiotics only 2 patients received both a home and hospital course of antibiotics Characteristics Mean(SD) Age: Home (n=15) 26.5 (6.3) vs Hospital (n=15) 22.5 (4.3), p value 0.61; Mean (SD) FEV1 (%predicted) on Day 0: Home (n=15) 33.8 (16.8) vs hospital (n=15) 32.3(16.9), p value 0.66; Mean (SD) BMI on Day 0: Home (n=15) 19.3 (3.0) vs hospital (n=15) 18.9 (2.2), p	Interventions Intervention IV antibiotic treatment administered at home. Patients received a combination of 2 IV antibiotics, which were chosen on the basis of the patient's latest sputum microscopy, culture and sensitivity Mean duration of treatment: 14 days (SD 1.5, range 10-18) The home group was asked to perform their own chest physiotherapy twice a day. Comparison IV antibiotic treatment administered at the hospital. Patients received a combination of 2 IV antibiotics, which were chosen on the basis of the patient's latest sputum microscopy, culture and sensitivity	Details Data collection Quasi-experimental design. The CFQoL questionnaire was used to measure quality of life. Analysis The samples were compared at time of entering the study (day 0) using the Mann-Whitney U-test. Statistical significance of change in FEV1, weight, BMI and quality of life during an antibiotic course was assessed using the	Results FEV1 % predicted at 15 days: Mean change (SD): Home (n=15) 2.0 (5.1) vs hospital (n=15) 5.1(5.6), p value 0.08; Post-treatment (post-Rx) Mean (SD): Home (n=15) 35.8 (19.1); hospital (n=15) 37.4 (19.7) p values (Wilcoxon signed rank test) home post-Rx vs Day 0: 0.16 hospital post-Rx vs Day 0: 0.005 Mortality Not reported Patient and carer satisfaction Not reported LCI Not reported Time to pulmonary exacerbation Not reported Nutritional status at 15 days: Mean change (SD) BMI: Home (n=15) 0.2 (0.3) vs hospital (n=15) 0.4 (0.8), p value 0.22. Post-treatment (Post-Rx) BMI Mean (SD): Home	Limitations The quality of this study was assessed using the Newcastle-Ottawa scale assessment tool: Selection: Unclear (All CF patients over 18 years of age attending the adult cystic fibrosis centre over a six-month period who received IV antibiotics for an acute exacerbation who fulfilled the study criteria were asked to participate in the study once it had been decided if IV antibiotic therapy was going to be administered in hospital or at home. However, authors do not specify if they included in the analysis all the courses of treatment that these patients received over these six months). Comparability: High risk (The study does not control for any factor. The groups were not matched). Outcome: Unclear (Length of follow-up was adequate: one course of antibiotics. However, there was no description of how FEV1 (% predicted) or BMI were

Study details	Participants	Interventions	Methods	Outcomes and Results	Comments
Not mentioned	value= 0.66 (In a different table BMI on Day 0 in hospital group is 19.0(2.3)) Inclusion criteria Confirmed diagnosis of cystic fibrosis Age 18 years and over Acute respiratory exacerbation Exclusion criteria Lung function < 30% predicted On active heart-lung transplant waiting list Pneumothorax Massive haemoptysis (>200 mls blood)	Mean duration of treatment: 15 days (SD 4.7, range 10-25) Chest physiotherapy performed by experienced respiratory physiotherapists twice a day. Input from a specialist CF dietician and availability of a supplements menu.	Wilcoxon signed rank test. The Mann-Whitney U-test was used to compare the change in home and hospital treatment groups.	(n=15) 19.5 (2.9); hospital (n=15) 19.4 (2.5) p values (Wilcoxon signed rank test) home post-treatment BMI post-Rx vs Day 0: 0.05 hospital post-treatments BMI post-Rx vs Day 0: 0.05 Mean (SD) quality of life at 15 days: Physical: Home (n=15): Day 0: 62.0 (14.0); Post-Rx: 72.0 (15.6), P=0.02 vs Hospital (n=15): Day 0: 55.5 (25.4); Post-Rx: 67.7 (21.0), P=0.07 Social: Home (n=15): Day 0: 74.0 (25.6); Post-Rx: 77.0 (22.2), P=0.22 vs Hospital (n=15): Day 0: 61.3 (32.7); Post-Rx: 67.7 (28.7), P=0.06 Treatment: Home (n=15): Day 0: 64.9 (31.2); Post-Rx: 71.1 (16.8), P=0.53 vs Hospital (n=15): Day 0: 62.0 (27.1); Post-Rx:70.2 (18.7) , P=0.21 Symptoms: Home (n=15): Day 0: 49.7 (21.9); Post-Rx: 68.8 (23.2), P=0.03 vs Hospital (n=15): Day 0: 47.0 (22.6); Post-Rx: 70.3 (15.2), P=0.006 Emotional: Home (n=15): Day 0: 66.0 (23.5); Post-Rx: 78.5 (17.6), P=0.01 vs Hospital (n=15): Day 0:	assessed, while QoL was self-reported with the CFQoL questionnaire). Overall quality: low Other information Patients at home All patients who chose home therapy had previously self-administered IV antibiotics at home. Intervention and comparison Patients treated at home were not asked about their adherence with physiotherapy. Patients at home are likely to have more flexibility around eating times and have more types of food available Power calculation The size of the sample was not based on a power calculation, as this was a pilot study. Analysis The article only gives the p values for a comparison of QoL scores on Day 0 vs QoL scores Post-Rx, for both home and hospital, but it does not compare with a statistical test the change in QoL scores during home treatment versus change during hospital treatment.

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				63.2 (26.1); Post-Rx: 71.5 (22.3), P=0.14 Future: Home (n=15): Day 0: 37.1 (23.5); Post-Rx: 40.9 (17.4), P=0.44 vs Hospital (n=15): Day 0: 42.3 (24.5); Post-Rx: 51.6 (21.3), P=0.04 Relationships: Home (n=15): Day 0: 45.9 (25.7); Post-Rx: 52.8 (22.0), P=0.049 vs Hospital (n=15): Day 0: 56.4 (2216*); Post-Rx: 55.9 (22.5), P=0.93 Body image: Home (n=15): Day 0: 44.0 (31.8); Post-Rx: 46.7 (27.8), P=0.19 vs Hospital (n=15): Day 0: 60.0 (23.0); Post-Rx: 61.8 (22.5), P=0.38 Career: Home (n=15): Day 0: 40.3 (29.4); Post-Rx: 50.3 (20.0), P=0.02 vs Hospital (n=15): Day 0: 51.3 (23.6); Post-Rx: 53.0 (23.4), P=0.65 *Mistake in the paper Frequency of cross-infections (pseudomonas, b.cepacia) Not reported Staff experience Not reported Adherence to treatment Not reported	

Study details	Participants	Interventions	Methods	Outcomes and Results	Comments
Full citation 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 Ref Id 332667 Country/ies where the study was carried out US Study type Retrospective comparative study (follow-up of a RCT) Aim of the study To investigate the effectiveness of an experimental home monitoring system implemented at the University of Minnesota Cystic Fibrosis Center for assessing the progress and planning changes in the care of patients with cystic fibrosis (CF) Study dates	Sample size N= 50 patients 25 in the “intervention” group 25 in the control group Characteristics Age ranges: 6 -43 years (a. 6 -12 years = 24 patients; 13-18 years = 8 patients; older than 18 years =18 patients) Gender: N= 20 F; 30 M Inclusion criteria Patients who returned a minimum of 20% of the diary forms –with at least one of every 6 weeks over the study period Patients who were included in a previous RCT (N=271) Exclusion criteria Not reported	Interventions Intervention: One group of patients and families did daily recording of physical measurements and symptoms, and sent the diary to the data coordinating centre weekly for analysis. Self-measurement and daily recording took place in the absence of any therapeutic intervention. Comparison: No diary recording, no home monitoring	Details Setting This study was settled in the US and it consists of a follow up sturdy (4 years) of an RCT. In this trial patients were randomized by age and gender into either diary (n=173) or non-diary (n=98) groups. Data collection The medical records of the included patients were reviewed retrospectively over a period of 4 years (1983-1987). Pulmonary function and growth measures were	Results FEV1 (% predicted): 1983: intervention=mean [+/-SEM]; control=mean [+/-SEM] 73.6 [6.0]; 72.3 [4.7] 1987 intervention=mean [+/-SEM]; control=mean [+/-SEM] 70.1 [5.2]; 60.8 [4.4] 1983-1987 dif. (over the 4-year period)= intervention=mean [+/-SEM]; control=mean [+/-SEM] 3.5 [2.3] p value=0.33, 95% CI= -2.3,8.1; 11.5 [3.0] p value=<0.01, 95% CI= 5.7,17.3 Intervention-control difference: p value=0.09, 95% CI= -15.5,0.03 Mortality Not reported Patient satisfaction Not reported LCI Not reported Time to next pulmonary exacerbation Not reported Nutritional status Not reported Quality of life Not reported Frequency of cross-infections (pseudomonas, b.cepacia)	Limitations The quality of this study was assessed using the Newcastle-Ottawa scale assessment tool: Selection: low risk of bias (clear sample size strategy, clear representativeness of the analysed cohort) Comparability: unclear risk bias (the authors did not control the analysis for none risk factors –relatively small sample size) Outcome: low risk of bias Overall quality: moderate Other information

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Data collection date: 1983-1987 (4 years-follow-up) Publication date: 1992 Source of funding NIH grant 27355 and 37504			obtained concurrently with clinical score component measures. The pulmonary function measures were performed in the paediatric function laboratory using standard instrumentation under computer control. Analysis Pulmonary changes were evaluated across groups over a period of 4 years. Comparability of the 2 groups at the beginning of the study was established via a t	Not reported Staff experience Not reported Adherence to treatment Not reported	

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			test. Differences for each participant of each group between the onset and endpoint of the study were calculated, and change over time were investigate by means of the t test		
Full citation Goldbeck, L., Fidika, A., Herle, M., Quittner, A. L., Psychological interventions for individuals with cystic fibrosis and their families, Cochrane Database of Systematic Reviews, 6, CD003148, 2014 Ref Id 406192 Country/ies where the study was carried out Wilkinson 2008: UK Study type Goldbeck 2014	Sample size Wilkinson 2008 N randomised: 16 N completed the interventions: 7 4 on telemedicine 3 in the control arm N responses to the telemedicine satisfaction questionnaire: 5 * Of those who did not complete the study 4 patients died, 3 patients received a transplant, 1 withdrew following randomisation and 1	Interventions Wilkinson 2008 Intervention Telemedicine additional to standard care: the participants were provided with an ISDN line to their home and a videoconferencing unit was connected to their home television set. Participants were also given a micro-spirometer, pulse-oximeter and a supply of single use clinical thermometers. Contact was made, on a weekly basis, at a time agreed by the patient and	Details Wilkinson 2008 Setting UK* Analysis RCT. Prospective pilot study. *Information extracted from primary study	Results Wilkinson 2008 FEV1 Not reported Mortality Not reported Patient satisfaction (% of responses) * : Q1 How did you find using the telemedicine equipment provided: "Very easy": 100% Q2: Did you find the link-up helpful in discussing health issues? Yes: 100% Q3 Did you feel a sense of security in seeing someone face to face from the CF team? Yes: 100%	Limitations Goldbeck 2014 AMSTAR score: 9/11 (Likelihood of publication bias not assessed; sources of support only reported for the systematic review, not for included studies) Wilkinson 2008 Random sequence generation (selection bias): Low risk (Participants were randomised by a physiotherapist distributing a pre-prepared sealed envelope, which was made by a third party not involved with recruitment) Allocation concealment (selection bias): Low risk

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Cochrane Systematic Review Wilkinson 2008 RCT Aim of the study Goldbeck 2014 To determine whether psychological interventions for people with cystic fibrosis provide significant psychosocial and physical benefits in addition to standard medical care. Wilkinson 2008 To investigate the feasibility of a video link to support patients on the transplantation waiting list and their families. Study dates Goldbeck 2014 Most recent search of the Cystic Fibrosis and Genetic Disorders Group's register: 19 December 2013. Most recent search of the Depression, Anxiety and Neurosis Group's register: 12 November 2013.	was too unwell to continue. Characteristics Wilkinson 2008 Median age of the patients who were randomised: 27 (range 21-41) Inclusion criteria Wilkinson 2008 Patients on the transplantation list At least 16 years of age With a confirmed diagnosis of CF Willing to have an ISDN line installed in their home. Exclusion criteria Wilkinson 2008 Patients were excluded if they could not understand the implications of the study * *Information extracted from primary study	assessor (senior physiotherapist or nurse consultant). The topics which were discussed included: non-invasive ventilation; haemoptysis; physiotherapy and amount of sputum; mobility; difficulties with any clinical procedures; appetite and weight; and any other problems as appropriate. Comparison Standard medical care		Q4 Does this type of service make you feel less isolated from the CF hospital team? Yes: 100% Q5: Did you find it intrusive with the assessor linking up with you in your home? No: 80%: No response: 20% Q6 Do you wish this service to be continued? Yes: 100%: Q7 Have you had any problems with the service? No: 60% Yes: 40% Q8: Do you think this is a good service? Yes: 100% Q9 on preference for the telephone, an extra clinic or telemedicine for clinical review: 1st choice: telephone: 20%, extra clinic: 0%, telemedicine: 80%; 2nd choice: telephone: 60%, extra clinic: 20%, telemedicine: 20%; 3rd choice: telephone: 20%, extra clinic: 80%, telemedicine: 0% LCI: Not reported Time to next pulmonary exacerbations Not reported Nutritional status Not reported	(Participants and investigator could not foresee assignments because the authors reported that they used sequentially numbered sealed envelopes) Blinding (performance bias and detection bias) (all outcomes): Unclear risk (Due to the nature of the intervention the participants and teams providing the intervention could been blinded, but the authors provided no information on blinding of outcome assessment) Incomplete outcome data (attrition bias) (all outcomes): High risk (The authors reported a high number of dropouts. The number of dropoutsfor each group is unclear. Reasons for dropouts were reported: 4 patients died, 3 patients received transplant, 1 withdrew, and 1 was too unwell to continue) Selective reporting (reporting bias): High risk (Means and SDs for all outcome parameters for intervention and control group were not reported in the published article). Other information Wilkinson 2008

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Wilkinson 2008 Data collection date: not reported (Authors only mention this was a "six-month prospective pilot study")* Source of funding Goldbeck 2014 Internal sources: Royal Liverpool Children's NHS Trust, UK. National Institutes of Health, USA. External sources: No sources of support supplied. Wilkinson 2008 Royal Brompton & Harefield Hospital Charitable Fund*				Quality of Life: Cystic Fibrosis Quality of Life Questionnaire (Gee 2000): Telemedicine group: a significant improvement in the subjects' perception of body image (p=0.02) Carer satisfaction: Not reported Frequency of cross-infections Not reported Staff experience Not reported Adherence to treatment Not reported *Information extracted from the primary study	Authors of the Cochrane review contacted the authors of the study for detailed quantitative data on outcome measures, but did not receive a response within the time of updating the review.
Full citation Riethmueller, J., Busch, A., Damm, V., Ziebach, R., Stern, M., Home and hospital antibiotic treatment prove similarly effective in cystic fibrosis, Infection, 30, 387-91, 2002 Ref Id	Sample size N= 36 patients 19 patients in the hospital group 17 patients in the home care group N= 58 courses 28 hospital courses 30 home care courses.	Interventions Intervention IV antibiotic treatment at home 14-day therapy courses Ceftazidime (200 mg/kg body weight/day, 3 infusions per day) combined with tobramycin (10 mg/kg body weight/day, 3 infusions per day)	Details Setting: CF centre at the University Children's Hospital Tuebingen Data collection: Cl inical and laboratory controls	Results FEV1 Mean (SD) FEV1 (%)**: Home (n=29) Pre: 55(28) Post: 63 (29) vs Hospital (n=27) Pre: 66 (29) Post: 72 (30),Hospital vs Home n.s. Mortality Not reported Patient and carer satisfaction Not reported	Limitations The quality of this study was assessed using the Newcastle-Ottawa scale assessment tool: Selection: low risk (Inclusion criteria for participation in the study included good compliance and regular home physiotherapy - it seems reasonable that this might be

Study details	Participants	Interventions	Methods	Outcomes and Results	Comments
331848 Country/ies where the study was carried out Germany Study type Prospective open study Aim of the study To compare home and and hospital IV antibiotic treatment in CF patients with chronic P. aeruginosa infection. Study dates Therapy courses were run between January 1996 and May 1997 Source of funding Financial support by Caremark Germany	Characteristics Patients under the care of the CF centre at the University Children's Hospital Tuebingen Data of patients entering the study *: Mean (SD) Age: Home 16 (5) vs Hospital 15 (4), n.s., Mean (SD) FEV1 (%): Home 55 (28) vs Hospital 66 (29), n.s.; Mean (SD) weight for height (%): Home 86 (9) vs Hospital 94 (10), p <= 0.005. * Please note that authors write "clinical data of patients entering the study", however the data for FEV and weight for height is the same as the data given in the results table which refers to treatments, so it seems that the means were calculated based on treatments rather than patients. Inclusion criteria P. aeruginosa in sputa over a time period of more than 6 months	except single cases of resistance when drugs were chosen according to resistogram Using intermateR containers 100 ml Tobramycin or colistin by inhalation High caloric nutritional intake Patients did their daily training and were supervised once per week by a physiotherapist specialized in CF. Patients were offered a visit by a specialized nurse if intravenous line or other problems occurred. Comparison IV antibiotic treatment in hospital 14-day therapy courses Ceftazidime (200 mg/kg body weight/day, 3 infusions per day) combined with tobramycin (10 mg/kg body weight/day, 3 infusions per day) except single cases of resistance when drugs were chosen according to resistogram	were done on days 1, 3 and 14. Analysis: Student's t-test and paired sample t-test were used for comparison of clinical and laboratory parameters before and after treatment. Analysis of variance was used for group comparison.	LCI Not reported Nutritional status Mean (SD) Weight (kg): Home (n=29) Pre: 38(12) Post: 39.1(13) vs Hospital (n=28) Pre: 36.5 (9) Post: 37.6 (9), Hospital vs Home n.s. Mean (SD) Weight for Height (%): Home (n=29) Pre: 86(9) Post: 89(9) vs Hospital (n=28) Pre: 94 (10) Post: 98(10), Hospital vs Home n.s. Quality of life Not reported Frequency of cross-infections Mean (SD) Pseudomonas counts (log10) (cfu/ml sputum): Home (n=20) Pre: 7.1 (2.1) Post: 3.4 (2.8) vs Hospital (n=16) Pre: 6.4 (2.2) Post: 3.2 (2.9), Hospital vs Home n.s. Staff experience Not reported Adherence to treatment Not reported	required of patients eligible for home care). Comparability: high risk (The study does not control for any factor). Outcome: Low risk for weight, weight for height and FEV1 (Adequate length of follow up and small number of subjects lost to follow up - No. courses in results table: 29 in home group (97% follow up rate) vs 27 or 28 in hospital group (96% or 100% follow up rate)). High risk for Pseudomonas counts (High number lost to follow up and no description of those lost - No. courses in results table: 20 in home group vs 16 in hospital group - this means that follow up rate was 67% in home group and 57% in hospital group) Overall quality: Low Other information This study was planned as a prospective randomized cross-over study, however this could not be realized, because most of the adolescent patients refused hospital treatment. Therefore, the authors decided to open the study.

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	No changes in supportive therapy throughout treatment Good compliance and regular home physiotherapy. All patients had a positive antibody response towards P. aeruginosa measured by ELISA. Exclusion criteria Patients with pulmonary exacerbations Patients with lung transplantation Patients with Burkholderia cepacia infection First time antibiotic treatment Positive CRP (>10 mg/l)	Using conventional infusion pumps Tobramycin or colistin by inhalation High caloric nutritional intake Hospitalized patients had two daily courses of supervised physiotherapy (1 h). Diets were supervised by a dietician specialized in CF care.			
Full citation Thomas, C., Mitchell, P., O'Rourke, P., Wainwright, C., Quality-of-life in children and adolescents with cystic fibrosis managed in both regional outreach and cystic fibrosis center settings in	Sample size N= 162 (Specialist centre or CFC: 91; Shared care or CFOS: 71) CFQ-Teen: 34 (CFC: 24 vs CFOS: 10) CFQ-Child: 83 (CFC: 46 vs CFOS: 37) CFQ-Parent: 80 (CFC: 45 vs CFOS: 35)	Interventions Intervention Cystic Fibrosis Centre (CFC): Children are reviewed at least 3 times a year and have full access to the MDT. (Similar to UK full centre care, although in the UK routine appointments should be	Details Setting The participants were treated by the Royal Children's Hospital CF team in a tertiary CFC or outreach	Results FEV1 Not reported Mortality Not reported Patient and carer satisfaction Not reported LCI Not reported	Limitations The quality of this study was assessed using the Newcastle-Ottawa scale assessment tool: Selection: High risk (There was a significantly higher completion rate in the CFC population (88.4%, 91 of 103) compared with the CFOS population (62.28%, 71 of

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<tr>
<td>Queensland, Journal of Pediatrics, 148, 508-516, 2006
Ref Id
369582
Country/ies where the study was carried out
Australia
Study type
Cross-sectional
Aim of the study
To compare health-related quality of life (HRQOL) in children and adolescents with CF managed by CF Outreach Service (CFOS) with those treated in a CF centre (CFC).
Study dates
Data collection
The study does not mention when questionnaires were sent out.
Clinical data were collected retrospectively, from January 1, 2000 to December 31, 2002.
Source of funding
Not mentioned</td>
<td>Characteristics
Between 2 and 19 years of age

Inclusion criteria
Confirmed CF diagnosis
Exclusion criteria
Not mentioned</td>
<td>at least every 2-3 months)
Comparison
Cystic Fibrosis Outreach Service (CFOS): Children are managed by their local paediatrician or general practitioner and local hospital, and they also attend outreach clinics visited by CFOS. The CFOS varies, although it usually includes a paediatric respiratory physician, physiotherapist, dietician and CF nurse. Regional staff, such as paediatricians, physiotherapists, dieticians, and clinical nurses, are invited to attend the clinics. Outreach clinics occur twice per year except for one site, which has one clinic and two telehealth clinics per year.
(CFOS is similar to shared care in the UK as defined by the UK CF Trust Standards of Care 2011. However, there are differences:
a UK Network CF Clinic should have an MDT</td>
<td>setting (CFOS).
Data collection De mographic details were collected from medical records or available pathology databases.
Two HRQOL surveys were administered : a generic HRQOL measure, PedsQL (TM), and a disease-specific HRQOL measure, the CFQ. Both have been previously validated and tested for reliability. The PedsQL (TM) was administered before the CFQ as per recommende d administratio</td>
<td>Time to pulmonary exacerbation
Not reported
Nutritional status
Not reported
Quality of life
CFQ-Teen, Child and Parent: scale mean (SD) scores
<table>
<tr><td>Scales</td><td>CFC Tee n</td><td>CF OS Tee n</td><td>CFC Chil d</td><td>CF OS Ch d</td></tr>
<tr><td>Physical</td><td>72.6 (23. 7)</td><td>90.4 (13. 1)</td><td>76.0 (21. 9)</td><td>77 (2 1)</td></tr>
<tr><td>Role</td><td>76.2 (21. 4)</td><td>86.6 (21. 9)</td><td>N/A</td><td>N/</td></tr>
<tr><td>Vitality</td><td>56.0 (25. 9)</td><td>74.2 (15. 9)</td><td>N/A</td><td>N/</td></tr>
<tr><td>Emotion al</td><td>77.2 (18. 4)</td><td>82.7 (17. 0)</td><td>76.1 (14. 8)</td><td>74 (1 9)</td></tr>
<tr><td>Social</td><td>76.4 (19. 1)</td><td>94.0 (8.0)</td><td>70.2 (15. 9)</td><td>71 (1 4)</td></tr>
<tr><td>Body</td><td>72.2 (23. 2)</td><td>76.7 (23. 1)</td><td>78.3 (24. 6)</td><td>81 (2 4)</td></tr>
</table>
</td>
<td>114), p<.001. More females than males responded, p= .01. Of the 46 teens, participation was significantly higher for the CFC (24 of 27, 89.0%) compared with those from the CFOS (10 of 19, 53.0%), p=.006)
Comparability: High risk (The study does not mention controlling for any factor)
Outcome: High risk (Not all the p values are given for the statistical tests)
Overall quality: low
Other information</td>
</tr>
</table>

<table>
<tr><th>Study details</th><th>Participants</th><th>Interventions</th><th>Methods</th><th>Outcomes and Results</th><th>Comments</th></tr>
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<td>separate to the specialist centre and a consultant with specialist interest and experience in CF;
MDTs in CFOS include less disciplines than the MDTs in UK CF Specialist CF centres.)</td>
<td>n guidelines. Questionnaires were self-administered for parents and for children aged 12 or over for the CFQ. Questionnaires were administered by interview for children aged 6-11 for CFQ. Interview surveys conducted in the waiting room were completed independently from parents. For the mailed surveys, instructions were included on how to complete the surveys.
Analysis One-way analysis of variance was used to</td>
<td>
<table>
<tr><td>Eating</td><td>80.6 (23.5)</td><td>94.4 (12.0)</td><td>76.1 (26.2)</td><td>76 (2 6)</td></tr>
<tr><td>TB</td><td>56.0 (21.1)</td><td>65.6 (26.4)</td><td>68.4 (25.2)</td><td>63 (2 8)</td></tr>
<tr><td>Health</td><td>57.4 (21.4)</td><td>72.2 (23.6)</td><td>N/A</td><td>N/</td></tr>
<tr><td>Weight</td><td>59.7 (34.0)</td><td>66.7 (31.4)</td><td>N/A</td><td>N/</td></tr>
<tr><td>Respiratory</td><td>68.3 (18.3)</td><td>72.8 (12.7)</td><td>70.8 (20.5)</td><td>66 (2 4)</td></tr>
<tr><td>Digestion</td><td>84.3 (16.4)</td><td>92.2 (10.5)</td><td>76.1 (26.9)</td><td>72 (2 9)</td></tr>
<tr><td>School function</td><td>N/A</td><td>N/A</td><td>N/A</td><td>N/</td></tr>
</table>
"In the CFQ surveys, teens from the CFOS group had a higher HRQOL score for all domains, but this difference was only significant for Social and Vitality scales (p<.05). CFOS children had better HRQOL scores for Physical, Social and Body Image but worse HRQOL scores for Emotional, Treatment Burden,
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Study details	Participants	Interventions	Methods	Outcomes and Results	Comments
			assess differences between CFC and CFOS groups for CFQ HRQOL scores.	Respiratory, and Digestion compared with CFC children, although these differences were not significant. There was no significant difference between any of the scale scores for the CFQ-Parent (proxy). Frequency of cross-infections (pseudomonas, b.cepacia) Not reported Staff experience Not reported Adherence to treatment Not reported	
Full citation 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 Ref Id 333320 Country/ies where the study was carried out Australia Study type	Sample size N= 273 (patients included in the study) Specialist centre: 131 Shared care review 3+ a year: 35 Shared care review 2+ a year: 72 Usual care: 35 6 patients died during the 3-year period (2 from LOC1, 3 from LOC 2, 1 from LOC 4). Analysis of changes in FEV1 was carried out on 150 patients Specialist centre: 74 Shared care review 3+ a year: 21	Interventions Intervention Specialist centre (Called Level of Care 1 (LOC1) in the study) All care is provided by the CFC Admission to the CFC when required Outpatient review at CFC three or more times per year Comparison 1 Shared care review 3+ a year (Called Level of Care 2 (LOC2) in the study)	Details Setting Sites covered by the CF clinic at the Royal Children's Hospital, Brisbane Analysis Pulmonary function rate of change from 1 January 2000 to 31 December 2002 was calculated by simple linear regression	Results FEV1 (% predicted) Mean (95% CI) first to last FEV1 % predicted per year: Specialist centre -1.4 (-2.9 to 0.1) vs Shared care review 3+ a year 0.5 (-4.0 to 5.0) vs Shared care review 2+ a year 1.0 (-2.1 to 4.1) vs Usual care 4.3 (-1.5 to 10.1), p 0.09 Mean (95% CI) slope FEV1% per year: Specialist centre -1.5 (-2.9 to -0.1) vs Shared care review 3+ a year -1.4 (-5.0 to 2.2) vs Shared care review 2+ a year 0.7 (-2.3 to 3.6) vs Usual care 1.8 (-1.0 to 4.7)	Limitations The quality of this study was assessed using the Newcastle-Ottawa scale assessment tool: Selection: High risk (Children under different levels of care live in different geographical areas). Comparability: Unclear (Authors give the following information: “Potential confounding was checked using general linear models and adjustment was made where necessary for comparisons between LOC categories” without specifying what they adjusted for and when.)

Study details	Participants	Interventions	Methods	Outcomes and Results	Comments
Retrospective study. Aim of the study To evaluate and compare the clinical outcomes of children with cystic fibrosis managed primarily at a tertiary cystic fibrosis centre (CFC) with those treated at regional centres by local health care professionals and the cystic fibrosis outreach service (CFOS). Study dates Clinical data between 1 January 2000 and 31 December 2002 Source of funding Not mentioned	Shared care review 2+ a year: 37 Usual care: 18 Characteristics Characteristics of 273 children included in the study (authors give the following characteristics without specifying what year they refer to): Median age: 9 years (range 0-20, IQR: 5-13) Boys and girls 0-4 years: 64. Specialist centre: 28 vs Shared care review 3+ a year: 9 vs Shared care review 2+ a year: 21 vs Usual care: 6 Boys and girls 5-9 years: 76. Specialist centre: 38 vs Shared care review 3+ a year: 8 vs Shared care review 2+ a year: 23 vs Usual care: 7 Males >= 10 years: 71. Specialist centre: 35 vs Shared care review 3+ a year: 8 vs Shared care review 2+ a year: 15 vs Usual care: 13 Females >= 10 years: 62.	Children living in regional centres and attending CFOS who also attend CFC regularly Admission to CFC or local hospital with local hospital care provided by local paediatrician Outpatient review by CFC or CFOS three or more times per year Comparison 2 Shared care review 2+ a year (Called Level of Care 3 (LOC3) in the study) Care is predominantly provided by the local paediatrician with consultation with CFC Admission to local hospital with care provided by local paediatrician Outpatient review by CFOS at least twice a year Comparison 3 Usual care (Called Level of Care 4 (LOC4) in the study)	using two methods: using all FEV1 % predicted measurements available for each child against time (slope FEV1 %) and using only the first and last FEV1 % predicted measurements available for each child against time (first to last FEV1 %). Associations between categorical variables were tested using the chi-squared test of association. Differences in patients' characteristics were assessed by one-way analysis of variance for	Mortality Not reported Patient and carer satisfaction Not reported LCI Not reported Time to next pulmonary exacerbation Not reported Nutritional status Not reported Quality of life Not reported Frequency of cross-infections Not reported Staff experience Not reported Adherence to treatment Not reported	Outcome: High risk (Data on FEV1 were only available for 150 children (55% of the 273 children included in the study) and there was no description of those lost). Overall quality: low Other information

Study details	Participants	Interventions	Methods	Outcomes and Results	Comments
	Specialist centre: 30 vs Shared care review 3+ a year: 10 vs Shared care review 2+ a year: 13 vs Usual care: 9 p value for sex and age group: 0.59 Characteristics of 150 patients included in the analysis on the change from first to last FEV1: Mean (95% CI) maximum FEV1 % predicted measurement over two years: Specialist centre: 86.9 (82.1 to 91.7) vs Shared care review 3+ a year: 84.9 (75.0 to 94.8) vs Shared care review 2+ a year: 86.0 (80.7 to 91.2) vs Usual care: 84.2 (75.9 to 92.4), p 0.94 Inclusion criteria Children with confirmed diagnosis of CF born between 19 October 1982 and 19 February 2002 Children with clinical data available between 1 January 2000 and 31 December 2002. Exclusion criteria	Involvement by CFC or CFOS once a year or no CFC/CFOS involvement Includes children seen by respiratory physicians but with no CFC or CFOS multidisciplinary health care involvement Alternatively, care provided by local paediatrician or general practitioner or unknown	pulmonary function and anthropometric measurements. Potential confounding was checked using general linear models and adjustment was made where necessary for comparisons between LOC categories.		

Study details	Participants	Interventions	Methods	Outcomes and Results	Comments
	Not mentioned				
Full citation Van Koolwijk, L. M. E., Uiterwaal, C. S. P. M., Van der Laag, J., Hoekstra, J. H., Gulmans, V. A. M., Van der Ent, C. K., Treatment of children with cystic fibrosis: Central, local or both?, Acta Paediatrica, International Journal of Paediatrics, 91, 972-977, 2002 Ref Id 406560 Country/ies where the study was carried out The Netherlands Study type Longitudinal, prospective Aim of the study To study the effects of the different levels of involvement of centralized care on the clinical conditions of children with CF. Study dates Patients attending the Cystic Fibrosis Centre Utrecht between	Sample size N= 105 Central care group: n=41 Shared care group: n=41 Local care group: n=23 Characteristics Males (%): central care 43.9 vs shared care 53.7 vs local care 52.2, n.s. Age (Mean (SEM): central 10.8 (0.5) vs shared 10.7 (0.5) vs local 9.4 (0.5), n.s. Age range: 5-17 Height (cm) (Mean (SEM): central 141.5 (2.7) vs shared 140.5 (3.0) vs local 134.9 (3.2), n.s. Weight (kg) (Mean (SEM): central 33.6 (1.8) vs shared 33.1 (1.9) vs local 29.9 (1.7), n.s. BMI, kg/m2 (Mean (SEM): central 16.2 (0.3) vs shared 16.2 (0.3) vs local 16.2 (0.3), n.s. FEV1 % pred (Mean (SEM): central 87.5	Interventions Intervention Specialist centre (Called centralized care in the study) Patients receive their treatment completely in the Centre Regular visits at minimum intervals of 3 months Comparison 1 Shared care (also called shared care in the study) Includes a half-yearly visit to the Centre (annual check-up and an MDT outpatient clinic visit) combined with regular visits to the local paediatrician The local paediatrician comes to the centre during the annual check-up and participates in the multidisciplinary consultation Regular visits at minimum intervals of 3 months Comparison 2	Details Data collection: A nnual data on FEV1, height and weight were obtained from the database of the CF Centre Utrecht. Spirometry was performed according to standards established by the American Thoracic Society. Pseudomon as colonization was studied throughout the whole study period at all outpatient visits Analysis: Me an differences	Results FEV1 (%predicted) FEV1 % pred (Annual change, Mean (SEM)): Central -2.9 (0.7) vs shared -2.4 (1.1) vs local -5.6 (1.5), none of the differences between groups was statistically significant Differences at the end of follow-up (3 years) between the three groups (I don't think we need this for our review): FEV1 % pred (Unadjusted mean difference (95% CI)): Shared-Central -7.1 (-17.6; 3.3) vs Local-Central 3.1 (-9.2;15.4) vs Local-Shared 10.2 (-2.1;22.5) FEV1 % pred (Mean difference adjusted for age, gender and corresponding baseline level (95% CI)) at 3 years: Shared-central 1.5 (-3.8, 6.7) vs Local-Central 2.2 (-1.2, 5.6) vs Local-Shared 12.5 (-11.5, 36.4) Mortality Not reported Patient and carer satisfaction Not reported LCI Not reported	Limitations The quality of this study was assessed using the Newcastle-Ottawa scale assessment tool: Selection: Low risk (Different groups were drawn from the same community: although referral to any care model is predominantly based on the distance between the patient's home and the centre, on personal habituation of the referring paediatrician, and on the severity and complexity of the disease, authors excluded patients with well known factors for progressive disease because these patients often require specialized care) Comparability: High risk (The authors did not control for any factor when comparing annual changes across groups) Outcome: Unclear (Authors do not mention how they measured FEV1 or BMI. Authors mention that only patients from whom at least 2 years of follow up data were available were included in the study, but do not say how many were excluded for this reason.)

Study details	Participants	Interventions	Methods	Outcomes and Results	Comments
January 1997 and January 2001. Source of funding Not mentioned	(3.6) vs shared 81.7 (4.4) vs local 98.9 (3.9), p value 0.030 P. aeruginosa colonization (%): 53.7 vs 58.5 vs 39.1, n.s. Inclusion criteria Not mentioned Exclusion criteria Colonization with Burkholderia cepacia Allergic bronchopulmonary aspergillosis (ABPA) Diabetes mellitus Inability to perform lung function tests (because of young age or mental/physical handicap) Pulmonary exacerbation at time of the tests Lung transplantation during the time of follow-up Patients that did not have at least two years of follow-up data available	Usual care (Called "Local care" in the study) Patients visit the centre only once a year at the annual check-up, but remain fully treated at their local hospitals. The local paediatrician comes to the centre during the annual check-up and participates in the multidisciplinary consultation Regular visits at minimum intervals of 3 months	in baseline characteristics of the patients were assessed by one-way ANOVA or by the chi-squared test. Data on FEV1 % predicted, BMI collected 3 year before were subtracted from the most recent data on these parameters to derive the changes during the time of analysis. By dividing these changes by follow-up time, authors calculated the annual changes. The annual changes were	Time to pulmonary exacerbation Not reported Nutritional status BMI kg/m2 (Annual change, Mean (SEM)): Central care 0.42 (0.08) vs shared 0.54 (0.14) vs local 0.51 (0.15), none of the differences between groups was statistically significant Differences at the end of follow-up (3 years) between the three groups (I don't think we need this for our review): BMI kg/m2 (Unadjusted mean difference (95% CI)) at 3 years: Shared-Central 0.04 (-0.93;1.02) vs Local-Central -0.14 (-1.29; 1.01) vs local minus shared -0.19 (-1.34;0.97) BMI kg/m2 (Mean difference adjusted for age, gender and corresponding baseline level (95% CI)) at 3 years: Shared-Central -0.08 (-0.71;0.54) vs Local-Central 0.08(-0.30;0.46) vs Local-Shared 0.22(-0.53;0.96) Quality of life Not reported Frequency of cross-infections (pseudomonas, b.cepacia)	Overall quality: Low Other information

Study details	Participants	Interventions	Methods	Outcomes and Results	Comments
			compared between the groups using linear regression, with annual changes as dependent variables and a group indicator as independent variable. Similar models were also used to adjust differences in levels found at the end of follow-up for differences in levels at baseline. Effect measures are presented as linear regression coefficients indicating mean group differences.	Not reported Staff experience Not reported Adherence to treatment Not reported	
Full citation	Sample size	Interventions	Details	Results FEV1	Limitations

Study details	Participants	Interventions	Methods	Outcomes and Results	Comments
Walters, S., Britton, J., Hodson, M. E., Hospital care for adults with cystic fibrosis: an overview and comparison between special cystic fibrosis clinics and general clinics using a patient questionnaire, Thorax, 49, 300-6, 1994 Ref Id 363517 Country/ies where the study was carried out UK Study type Cross-sectional study Aim of the study To assess the current pattern of medical service received by adults with cystic fibrosis and to compare the type of care between special cystic fibrosis and general clinics Study dates Data collection date: Not explicitly stated (1990) Publication date: 1994 Source of funding	N= 886 people with CF (59% of the total number of people with cystic fibrosis over 15 years of age in the UK at the time of the study). Characteristics N= 886 people with CF members of the Association of Cystic Fibrosis Adults N= 494 [62%] (All special cystic fibrosis clinics) N= 252 [33.8%] (All general clinics) Inclusion criteria Patients attending large special cystic fibrosis clinics and general clinics at local hospitals. Exclusion criteria Not reported	Intervention: Large special cystic fibrosis clinics Comparison: Non general clinics at local hospitals	Setting This study was placed In the UK, and it was basedf on a survey of the Association of Cystic Fibrosis Adults (ACFA) extends to approximatel y 68% of the UK population with cystic fibrosis aged over 16 years, and to over 80% of those over 25 years of age. Data collection Questionnair es were sent to all 1052 members of the Association of Cystic Fibrosis Adults. 746 patients (494 patients	Not reported LCI Not reported Time to next pulmonary exacerbation Not reported Mortality Not reported Nutritional status Not reported Quality of life Not reporte patient and carer satisfaction: General clinics vs fibrosis general clinics [95% CI of mean difference between CF clinics and general clinics]; Hospital accommodation: 3.64 VS 3.74 [-0.26 to 0.06]; N = 636 Hospital food: 2.73 VS 2.76 [-0.22 to 0.16] ; N = 631 Consultant's knowledge of CF: 4.33 VS 4.74 [-0.53 to -0.29*]; N = 690 Consultant's understanding of your problems:3.93 VS 4.31 [-0.54 to – 0.23]; N = 687 Junior doctors' understanding of CF: 3.13 VS 3.65 [-0.68 to -0.36*]; N = 645	The quality of this study was assessed using the Newcastle-Ottawa scale assessment tool: Selection: low risk of bias (clear representativeness of the analysed cohort) Comparability: unclear risk bias (the authors did not control the analysis for none risk factors –relatively small sample size) Outcome: low risk of bias Overall quality: moderate Other information

Study details	Participants	Interventions	Methods	Outcomes and Results	Comments
Cystic Fibrosis Research Trust			were attending a cystic fibrosis clinic and 252 a general clinic). Analysis Data were analysed using, where appropriate, x2, Mantel-Haenszel, analysis of variance, and confidence intervals for single proportions and the difference between proportions. Not all respondents answered all questions, and analysis for each question is confined only to those who made valid responses.	Nurses' understanding of CF: 3.27 VS 3.93 [-0.81 to -0.51*]; N = 662 Physiotherapy advice you receive: 3.97 VS 4.27 [-0.47 to -0.13*]; N = 642 Dietary advice you receive: 3.23 VS 3.86 [-0.83 to -0-36*]; N = 694 Social work advice you receive: 2.24 VS 2.89 [-1.00 to - 0.30*]; N = 369 Overall rating of hospital care: 3.76 VS 4.20 [-0.58 to -0.29*]; N = 686*p<0.05 frequency of cross-infections (pseudomonas, b.cepacia) Not reported staff experience Not reported adherence to treatment Not reported	

Study details	Participants	Interventions	Methods	Outcomes and Results	Comments
Full citation 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 Ref Id 367007 Country/ies where the study was carried out See Cochrane SR Goldbeck 2014 Study type See Cochrane SR Goldbeck 2014 Aim of the study See Cochrane SR Goldbeck 2014 Study dates See Cochrane SR Goldbeck 2014 Source of funding See Cochrane SR Goldbeck 2014	Sample size See Cochrane SR Goldbeck 2014 Characteristics See Cochrane SR Goldbeck 2014 Inclusion criteria See Cochrane SR Goldbeck 2014 Exclusion criteria See Cochrane SR Goldbeck 2014	Interventions See Cochrane SR Goldbeck 2014	Details See Cochrane SR Goldbeck 2014	Results See Cochrane SR Goldbeck 2014	Limitations See Cochrane SR Goldbeck 2014 Other information See Cochrane SR Goldbeck 2014
Full citation Wolter, J. M., Bowler, S. D., Nolan, P. J.,	Sample size See Cochrane SR Balaguer 2015	Interventions See Cochrane SR Balaguer 2015	Details See Cochrane	Results See Cochrane SR Balaguer 2015	Limitations See Cochrane SR Balaguer 2015

Study details	Participants	Interventions	Methods	Outcomes and Results	Comments
McCormack, J. G., Home intravenous therapy in cystic fibrosis: a prospective randomized trial examining clinical, quality of life and cost aspects, European Respiratory Journal, 10, 896-900, 1997 Ref Id 363511 Country/ies where the study was carried out See Cochrane SR Balaguer 2015 Study type See Cochrane SR Balaguer 2015 Aim of the study See Cochrane SR Balaguer 2015 Study dates See Cochrane SR Balaguer 2015 Source of funding See Cochrane SR Balaguer 2015	Characteristics See Cochrane SR Balaguer 2015 Inclusion criteria See Cochrane SR Balaguer 2015 Exclusion criteria See Cochrane SR Balaguer 2015		SR Balaguer 2015		Other information See Cochrane SR Balaguer 2015
Full citation Balaguer, Albert, Gonzalez de Dios, Javier, Home versus hospital intravenous antibiotic therapy for cystic fibrosis, Cochrane Database	Sample size Wolter 1997 17 participants 31 admissions 13 admissions: home therapy group *	Interventions Wolter 1997 Intervention Home therapy: Patients spent 2 - 4 days in hospital before	Details Wolter 1997 Participants were initially randomized in blocks of four by sealed	Results Wolter 1997 FEV1 (% predicted) mean (SD*): Day 0: Home 39 (17) vs hospital 44 (20) *	Limitations Balaguer 2015 Amstar score: 9/11 (The authors did not mention that publication bias could not be assessed because there were fewer than 10 included studies; Source of support or

Study details	Participants	Interventions	Methods	Outcomes and Results	Comments
of Systematic Reviews, 2015 Ref Id 425672 Country/ies where the study was carried out Wolter 1997: Australia* Study type Balaguer 2015 Cochrane systematic review Wolter 1997 RCT and cross-over open study Aim of the study Balaguer 2015 To determine whether home intravenous antibiotic therapy in cystic fibrosis is as effective as inpatient intravenous antibiotic therapy and if it is preferred by individuals or families or both. Wolter 1997 To determine if home IV antibiotic therapy in adult patients with CF is a feasible, effective and less costly	18 admissions: control group * *Information extracted from individual paper Characteristics Wolter 1997 Agre range (median): 19-41 (22)* All patients had colonization of their sputum with P. aeruginosa* *Information extracted from individual paper Inclusion criteria Wolter 1997 Consenting adolescents and adults* With an infective exacerbation of cystic fibrosis Attending two Brisbane hospitals* *Information extracted from individual paper Exclusion criteria Wolter 1997 Unstable disease* Dwelling outside Brisbane* A history of noncompliance*	discharge and were taught to prepare and administer their own IV antibiotics; Participants were discharged with medication and equipment for the duration of the proposed course of treatment; Home visits were conducted. All participants received the same antibiotic therapy with ceftazidime 2 g 12 hourly and tobramycin 4 to 6 mg/kg daily as a single bolus for a minimum of 10 days. Comparison Control group: Whole treatment was administered in the hospital. All participants received the same antibiotic therapy with ceftazidime 2 g 12 hourly and tobramycin 4 to 6 mg/kg daily as a single bolus for a minimum of 10 days.	envelopes, to home or hospital therapy. Participants experiencing recurrent episodes automatically alternated tr eatment arms after initial randomizatio n.	Day 10: Home 45 (22) vs hospital 50 (21) Day 21 (post-treatment): Home 43 (19) vs hospital 51 (21) p value comparing magnitudes of overall changes in the home vs hospital arm: 0.27 Mortality Not reported Patient and carer satisfaction Not reported LCI Not reported Time to next pulmonary exacerbation Not reported Nutritional status Weight (kg)* mean (SD*): Day 0: Home 53.7 (8.6) vs hospital 52.5 (7.5) Day 10: Home 54.1 (8.9) vs hospital 53.4 (7.6) Day 21 (post-treatment): Home 53.9 (8.7) vs hospital 53.2 (7.6) p value comparing magnitudes of overall changes in the home vs hospital arm: 0.10 Quality of life Not reported Frequency of cross-infections	funding was mentioned for the systematic review, but not for the included study) Wolter 1997 Random sequence generation (selection bias): Low risk (Randomized in blocks of four) Allocation concealment (selection bias): Low risk (Randomization used sealed envelopes) Blinding (performance bias and detection bias), all outcomes: High risk (Participants and clinicians could not be blinded due to the nature of the treatment. No information given on whether outcome assessors were blinded) Incomplete outcome data (attrition bias), all outcomes: Low risk (Reasons for exclusions given) Selective reporting (reporting bias): Low risk (Authors were unable to detect any selective reporting) Other information The unit of analysis is the admission. 9 participants had 1 admission, 5 had 2 admissions, 1 had 3 admissions, 1 had 4 admissions, and 1 had 5 admissions.

Study details	Participants	Interventions	Methods	Outcomes and Results	Comments
alternative to hospitalization, and to assess the impact of home therapy on quality of life. * *Information extracted from individual paper Study dates Balaguer 2015 Search date: The evidence is current to 23 November 2015. Wolter 1997 Not mentioned* *Information extracted from individual paper Source of funding Balaguer 2015 Internal sources: Universitat Internacional de Catalunya. Barcelona, Spain. External sources: National Institute for Health Research, UK. This systematic review was supported by the National Institute for Health Research, via Cochrane Infrastructure funding	Inability to learn treatment techniques, including home physiotherapy* Personal request* Patients with lung transplants* Patients on their first admission* * Information extracted from individual paper			Not reported Staff experience Not reported Adherence to treatment Not reported * Information extracted from individual paper	It is not known whether admissions were different episodes or recurrences. All episodes, initial or recurrent, were analysed together. The statistical analysis considered recurrent episodes as independent events. Data on first randomized episodes are not currently available.

Study details	Participants	Interventions	Methods	Outcomes and Results	Comments
to the Cochrane Cystic Fibrosis and Genetic Disorders Group. Wolter 1997 "The authors wish to acknowledge Glaxo Australia, Brisbane Teaching Hospitals Scholarship 1993, John P. Kelly Research Foundation, D. Battistutta from Medical Biostatistics Pty. Ltd" * *Information extracted from primary study					