Cystic Fibrosis

Current Awareness Newsletter

April 2015
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New Cochrane Library Systematic Reviews on Cystic Fibrosis

**Standard (head-down tilt) versus modified (without head-down tilt) postural drainage in infants and young children with cystic fibrosis**

Diana A Freitas, Fernando AL Dias, Gabriela SS Chaves, Gardenia MH Ferreira, Cibele TD Ribeiro, Ricardo O Guerra, Karla MPP Mendonça

**Published 10th March 2015**

Postural drainage is used primarily in infants with cystic fibrosis from diagnosis up to the moment when they are mature enough to actively participate in self-administered treatments. However, there is a risk of gastroesophageal reflux associated with this technique. **Objectives:** To compare the effects of standard postural drainage (greater (30° to 45° head-down tilt) and lesser (15° to 20° head-down tilt)) with modified postural drainage (greater (30° head-up tilt) or lesser (15° to 20° head-up tilt)) with regard to gastroesophageal reflux in infants and young children up to six years old with cystic fibrosis in terms of safety and efficacy.

**Standard versus biofilm antimicrobial susceptibility testing to guide antibiotic therapy in cystic fibrosis**

Valerie Waters, Felix Ratjen

**Published 5th March 2015**

The antibiotics used to treat pulmonary infections in people with cystic fibrosis are typically chosen based on the results of antimicrobial susceptibility testing performed on bacteria traditionally grown in a planktonic mode (grown in a liquid). However, there is considerable evidence to suggest that *Pseudomonas aeruginosa* actually grows in a biofilm (or slime layer) in the airways of people with cystic fibrosis with chronic pulmonary infections. Therefore, choosing antibiotics based on biofilm rather than conventional antimicrobial susceptibility testing could potentially improve response to treatment of *Pseudomonas aeruginosa* in people with cystic fibrosis. This is an update of a previously published Cochrane Review. **Objectives:** To compare biofilm antimicrobial susceptibility testing-driven therapy to conventional antimicrobial susceptibility testing-driven therapy in the treatment of *Pseudomonas aeruginosa* infection in people with cystic fibrosis.
New from NICE

IN DEVELOPMENT: Cystic fibrosis: diagnosis and management of cystic fibrosis

Anticipated publication date: Feb 2017

Recent Literature Searches on Cystic Fibrosis

Below is a sample of literature searches carried out by librarians for UH Bristol members of staff on the subject of Cystic Fibrosis. For further details get in touch: bennet.jones@uhbristol.nhs.uk

- CF and instigating non-invasive ventilation
- CF and dry powder antibiotics
Current Awareness Database Articles on Cystic Fibrosis

Below is a selection of articles on cystic fibrosis recently added to the healthcare databases, grouped in the following categories:

- Medical
- Microbiological
- Psychological
- Other

If you would like any of the following articles in full text, or if you would like a more focused search on your own topic, then get in touch: bennet.jones@uhbristol.nhs.uk

Medical

Title: Long-term treatment with oral N-acetylcysteine: Affects lung function but not sputum inflammation in cystic fibrosis subjects. A phase II randomized placebo-controlled trial

Citation: Journal of Cystic Fibrosis, March 2015, vol./is. 14/2(219-227), 1569-1993;1873-5010 (01 Mar 2015)


Abstract: Purpose: To evaluate the effects of oral N-acetylcysteine (NAC), which replenishes systemic glutathione, on decreasing inflammation and improving lung function in CF airways. Methods: A multicenter, randomized, double-blind proof of concept study in which 70 CF subjects received NAC or placebo orally thrice daily for 24 weeks. Endpoints: primary, change in sputum human neutrophil elastase (HNE) activity; secondary, FEV<sub>1</sub> and other clinical lung function measures; and safety, the safety and tolerability of NAC and the potential of NAC to promote pulmonary hypertension in subjects with CF. Results: Lung function (FEV<sub>1</sub> and FEF<sub>25-75%</sub>) remained stable or increased slightly in the NAC group but decreased in the placebo group (p=0.02 and 0.02). Log<sub>10</sub> HNE activity remained equal between cohorts (difference 0.21, 95% CI -0.07 to 0.48, p=0.14). Conclusions: NAC recipients maintained their lung function while placebo recipients declined (24week FEV<sub>1</sub> treatment effect=150mL, p<0.02). However no effect on HNE activity and other selected biomarkers of neutrophilic inflammation were detected. Further studies on mechanism and clinical outcomes are warranted.
Title: Cystic fibrosis and the role of gastrointestinal outcome measures in the new era of therapeutic CFTR modulation

Citation: Journal of Cystic Fibrosis, March 2015, vol./is. 14/2(169-177), 1569-1993;1873-5010 (01 Mar 2015)


Abstract: With the development of new drugs that directly affect CFTR protein function, clinical trials are being designed or initiated for a growing number of patients with cystic fibrosis. The currently available and accepted clinical endpoints, FEV1 and BMI, have limitations. The aim of this report is to draw attention to the need and the ample possibilities for the development and validation of relevant gastrointestinal clinical endpoints for scientific evaluation of CFTR modulation treatment, particularly in young children and infants. The gastrointestinal tract offers very good opportunities to measure CFTR protein function and systematically evaluate CF related clinical outcomes based on the principal clinical gastrointestinal manifestations of CF: intestinal pH, intestinal transit time, intestinal bile salt malabsorption, intestinal inflammation, exocrine pancreatic function and intestinal fat malabsorption. We present a descriptive analysis of a variety of gastrointestinal outcome measures for clinical relevance, reliability, validity, responsiveness to interventions, feasibility in particular in young children and the availability of reference values.

Title: Managing central venous obstruction in cystic fibrosis recipients-lung transplant considerations

Citation: Journal of Cystic Fibrosis, March 2015, vol./is. 14/2(255-261), 1569-1993;1873-5010 (01 Mar 2015)

Author(s): Otani S., Westall G.P., Levvey B.J., Marasco S., Lyon S., Snell G.I.

Abstract: Background: The superior vena cava (SVC) syndrome in cystic fibrosis (CF) patients is rare, but presents unique challenges in the peri-transplant period. We reviewed our experience of SVC syndrome in CF recipients undergoing lung transplantation. Methods: This is a retrospective case series from a single center chart-review. SVC obstruction is defined by clinically significant stenosis or obstruction of the SVC as detected by contrast studies. Results: We identified SVC obstruction in seven post-transplant cases and one pre-transplant case. All eight patients had previous or current history of indwelling central venous catheters. Three recipients experienced operative complications. Five of the seven recipients suffered at least one episode of post-operative SVC obstruction or bleeding despite prophylactic anticoagulation. At a median follow-up of 29 months, six of the seven patients transplanted are well. Conclusions: Strategies are available to minimize the risks of intra/peri-operative acute life-threatening SVC obstruction in CF patients.

Title: Pharmacokinetics and tolerability of oral sildenafil in adults with cystic fibrosis lung disease

Citation: Journal of Cystic Fibrosis, March 2015, vol./is. 14/2(228-236), 1569-1993;1873-5010 (01 Mar 2015)

Abstract: Rationale: Airway inflammation is central to cystic fibrosis (CF) pathophysiology. Preclinical models have shown that phosphodiesterase inhibitors (PDEi) like sildenafil have anti-inflammatory activity. PDEi have not been studied in CF subjects. Objectives: We evaluated the pharmacokinetics, tolerability, and safety of sildenafil in subjects with CF. Sputum biomarkers were used to explore efficacy. Methods: An open-label pilot study of oral sildenafil administration was conducted in adults with mild to moderate CF lung disease. Subjects received oral sildenafil 20 or 40 mg p.o. t.i.d. for 6 weeks. Measurements and main results: Twenty subjects completed the study. Estimated elimination rate constants were statistically different in subjects with CF compared to previously published non-CF subjects. Side effects were generally mild. There were no drug-related serious adverse events. Sputum neutrophil elastase activity decreased. Conclusions: Subjects with CF may eliminate sildenafil at a faster rate than non-CF subjects. Sildenafil administration was safe in subjects with CF and decreased sputum elastase activity. Sildenafil warrants further study as an anti-inflammatory in CF.

Title: 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

Citation: Journal of Cystic Fibrosis, March 2015, vol./is. 14/2(211-218), 1569-1993;1873-5010 (01 Mar 2015)

Author(s): Skov M., Pressler T., Lykkesfeldt J., Poulsen H.E., Jensen P.T., Johansen H.K., Qvist T., Kraemer D., Hoiby N., Ciofu O.

Abstract: Background: Patients with cystic fibrosis (CF) and chronic Pseudomonas aeruginosa lung infection have increased oxidative stress as a result of an imbalance between the production of reactive oxygen species caused by inflammation and their inactivation by the impaired antioxidant systems. Supplementation with anti-oxidants is potentially beneficial for CF patients. Methods: The effect of 4 weeks of oral N-acetylcysteine (NAC) treatment (2400 mg/day divided into two doses) on biochemical parameters of oxidative stress was investigated in an open-label, controlled, randomized trial on 21 patients; 11 patients in the NAC group and 10 in the control group. Biochemical parameters of oxidative burden and plasma levels of antioxidants were assessed at the end of the study and compared to the baseline values in the two groups. Results: A significant increase in the plasma levels of the antioxidant ascorbic acid (p = 0.037) and a significant decrease in the levels of the oxidized form of ascorbic acid (dehydroascorbate) (p = 0.004) compared to baseline were achieved after NAC treatment. No significant differences were observed in the control group. The parameters of oxidative burden did not change significantly compared to baseline in either of the groups. A better lung function was observed in the NAC treated group with a mean (SD) change compared to baseline of FEV1% predicted of 2.11 (4.6), while a decrease was observed in the control group (change - 1.4 (4.6)), though not statistically significant. Conclusion: Treatment with N-acetylcysteine 1200 mg x 2/day for 30 days significantly decreased the level of oxidized vitamin C and increased the level of vitamin C (primary end-points) and a not statistically significant improvement of lung function was observed in this group of patients.
Title: Randomized, single blind, controlled trial of inhaled glutathione vs placebo in patients with cystic fibrosis

Citation: Journal of Cystic Fibrosis, March 2015, vol./is. 14/2(203-210), 1569-1993;1873-5010 (01 Mar 2015)


Abstract: Background: In cystic fibrosis (CF) the defective CF transmembrane conductance regulator protein may be responsible for the impaired transport of glutathione (GSH), the first line defense of the lung against oxidative stress. The aim of this single-blind, randomized, placebo-controlled trial was to evaluate the effect of inhaled GSH in patients with CF. Methods: 54 adult and 51 pediatric patients were randomized to receive inhaled GSH or placebo twice daily for 12 months. Results: Twelve month treatment with inhaled GSH did not achieve our predetermined primary outcome measure of 15% improvement in FEV<sub>1</sub>%. Only in patients with moderate lung disease, 3, 6 and 9 months therapy with GSH resulted in a statistically significant increase of FEV<sub>1</sub>values from the baseline. Moreover GSH therapy improved 6-minute walking test in pediatric population. GSH was well tolerated by all patients. Conclusions: Inhaled GSH has slight positive effects in CF patients with moderate lung disease warranting further study.

Title: Slower rise of exhaled breath temperature in cystic fibrosis

Citation: Indian Pediatrics, 2015, vol./is. 52/2(125-127), 0019-6061;0974-7559 (2015)

Author(s): Bade G., Gupta S., Kabra S.K., Talwar A.

Abstract: Objective: To measure exhaled breath temperature in patients with cystic fibrosis. Methods: 17 patients (6-18 years) with cystic fibrosis and 15 age- and gender-matched healthy controls were recruited in this cross sectional study. Exhaled breath temperature was measured in subjects recruited in both the groups with a device X-halo and analyzed as plateau temperature achieved and rate of temperature rise. Results: Patients with cystic fibrosis showed no significant difference in plateau temperature [34.4(32.3-34.6) versus 33.9 (33.0-34.4)°C; P=0.35] while mean (SEM.) rate of temperature rise was significantly less in patients [0.09 (0.01) versus 0.14 (0.02) °C/s; P=0.04] as compared to controls. Conclusion: There was a slower rise of exhaled breath temperature in patients with cystic fibrosis whereas plateau temperature was not significantly different from controls.

Microbiological
Title: Prevalence of Scedosporium species and Lomentospora prolificans in patients with cystic fibrosis in a multicenter trial by use of a selective medium

Citation: Journal of Cystic Fibrosis, March 2015, vol./is. 14/2(237-241), 1569-1993;1873-5010 (01 Mar 2015)


Abstract: Objective: Detection of hyphomycetes of the Scedosporium apiospermum complex and Lomentospora prolificans (Sac.- Lp) is not yet standardized. Prevalence rates in patients with cystic fibrosis (CF) and the resistance pattern of these pathogens in Germany are unknown. Methods: In a one-year prospective study 11 laboratories used a selective medium for isolation of Sac.- Lp, examining >. 11,600 respiratory samples from 2346 patients with CF. Isolates were identified by molecular methods and tested for susceptibility to antifungal drugs. Results: The prevalence of Sac-Lp in patients with CF in Germany varied from 0.0 to 10.5% (mean: 3.1%) among the clinical centres. The benefit of the selective medium SceSel<sup>+</sup> compared to standard media for fungi was documented for >5000 samples. High antifungal resistance was detected in the S. apiospermum complex, and the multiresistance of L. prolificans was confirmed. Conclusion: Microbiology laboratories should be aware of these resistant species in patients with CF and consider using a selective medium.

Title: Randomized controlled trial of biofilm antimicrobial susceptibility testing in cystic fibrosis patients

Citation: Journal of Cystic Fibrosis, March 2015, vol./is. 14/2(262-266), 1569-1993;1873-5010 (01 Mar 2015)


Abstract: This study aimed to determine whether antimicrobial susceptibility testing of Pseudomonas aeruginosa grown as a biofilm, rather than planktonically, improves efficacy of antibiotic treatment for pulmonary exacerbations. This was a multicenter randomized, double-blind controlled trial of 14. days of intravenous antibiotic treatment for pulmonary exacerbations chosen based on conventional vs. biofilm antimicrobial susceptibility results in CF patients with chronic P. aeruginosa infection. There were 74 exacerbations in 39 patients. A total of 46% (12/26) exacerbations in the conventional group compared to 40% (19/48) exacerbations in the biofilm group achieved a >. 3 log drop in P. aeruginosa sputum density (difference - 0.03, 95% CI - 0.5 to 0.4, p. = 0.9). Lung function improvements were similar in both groups. Biofilm antimicrobial susceptibility testing did not lead to improved microbiological or clinical outcomes compared to conventional methods in the treatment of pulmonary exacerbations in CF patients with chronic P. aeruginosa.
Title: Rapid Detection and immune characterization of mycobacterium abscessus infection in cystic fibrosis patients

Citation: PLoS ONE, March 2015, vol./is. 10/3, 1932-6203 (05 Mar 2015)

Author(s): Steindor M., Nkwouano V., Mayatepek E., Mackenzie C.R., Schramm D., Jacobsen M.

Abstract: Cystic fibrosis patients are highly susceptible to infections with non-tuberculous mycobacteria. Especially Mycobacterium abscessus infections are common but reliable diagnosis is hampered by non-specific clinical symptoms and insensitive mycobacterial culture. In the present study we established novel methods for rapid detection and immune characterization of Mycobacterium abscessus infection in cystic fibrosis patients. We performed Mycobacterium abscessus specific DNA-strip- And quantitative PCR-based analyses of noncultured sputum samples to detect and characterize Mycobacterium abscessus infections. Concomitantly in vitro T-cell reactivation with purified protein derivatives (PPDs) from different mycobacterial species was used to determine Mycobacterium abscessus specific T-cell cytokine expression of infected cystic fibrosis patients. Four of 35 cystic fibrosis patients (11.4%) were Mycobacterium abscessus culture positive and showed concordant DNAstrip- Test results. Quantitative PCR revealed marked differences of mycobacterial burden between cystic fibrosis patients and during disease course. Tandem-repeat analysis classified distinct Mycobacterium abscessus strains of infected cystic fibrosis patients and excluded patient-to-patient transmission. Mycobacterium abscessus specific T-cells were detected in the blood of cystic fibrosis patients with confirmed chronic infection and a subgroup of patients without evidence of Mycobacterium abscessus infection. Comparison of cytokine expression and phenotypic markers revealed increased proportions of CD40L positive T-cells that lack Interleukin-2 expression as a marker for chronic Mycobacterium abscessus infections in cystic fibrosis patients. Direct sputum examination enabled rapid diagnosis and quantification of Mycobacterium abscessus in cystic fibrosis patients. T-cell in vitro reactivation and cytokine expression analyses may contribute to diagnosis of chronic Mycobacterium abscessus infection.

Title: High-frequency audiometry reveals high prevalence of aminoglycoside ototoxicity in children with cystic fibrosis

Citation: Journal of Cystic Fibrosis, March 2015, vol./is. 14/2(248-254), 1569-1993;1873-5010 (01 Mar 2015)

Author(s): Al-Malky G., Dawson S.J., Sirimanna T., Bagkeris E., Suri R.

Abstract: Background: Intravenous aminoglycoside (IV AG) antibiotics, widely used in patients with cystic fibrosis (CF), are known to have ototoxic complications. Despite this, audiological monitoring is not commonly performed and if performed, uses only standard pure-tone audiometry (PTA). The aim of this study was to investigate ototoxicity in CF children, to determine the most appropriate audiological tests and to identify possible risk factors. Methods: Auditory assessment was performed in CF children using standard pure tone audiometry (PTA), extended high-frequency (EHF) audiometry and distortion-product otoacoustic emissions (DPOAE). Results: 70 CF children, mean (SD) age 10.7 (3.5) years, were recruited. Of the 63 children who received IV AG, 15 (24%) children had ototoxicity detected by EHF audiometry and DPOAE. Standard PTA only detected ototoxicity in
13 children. Eleven of these children had received at least 10 courses of IV AG courses. A 25 to 85 dB. HL hearing loss (mean +/- SD: 57.5 +/- 25.7 dB. HL) across all EHF frequencies and a significant drop in DPOAE amplitudes at frequencies 4 to 8 kHz were detected. However, standard PTA detected a significant hearing loss (> 20 dB. HL) only at 8 kHz in 5 of these 15 children and none in 2 subjects who had significantly elevated EHF thresholds. The number of courses of IV AG received, age and lower lung function were shown to be risk factors for ototoxicity. Conclusions: CF children who had received at least 10 courses of IV AG had a higher risk of ototoxicity. EHF audiometry identified 2 more children with ototoxicity than standard PTA and depending on facilities available, should be the test of choice for detecting ototoxicity in children with CF receiving IV AG.

**Psychological**

**Title:** Integration of palliative care into the routine care of cystic fibrosis patients.

**Citation:** Palliative Medicine, March 2015, vol./is. 29/3(282-283), 0269-2163;1477-030X (Mar 2015)

**Author(s):** Karlekar, Mohana, Doherty, Kathleen E, Guyer, Dana, Slovis, Bonnie

**Abstract:** This study conducted a retrospective chart review of patients referred to specialist palliative care (SPC) providers by cystic fibrosis (CF) teams in the outpatient adult CF clinic between 1 January and 30 June 2012. A total of 30 different patients were identified. Patients' electronic medical records were reviewed for the presence of various symptoms including pain, anxiety, shortness of breath, depression, forced expiratory volume in one second (FEV1), willingness to discuss advance care directives, and evidence of completed advance care directives. Patient confidentiality was maintained throughout this study. Our chart review indicates that a significant number of CF patients not at the EOL have a high prevalence of distressing symptoms that are potentially very treatable. The literature suggests that patients with CF at the EOL suffer from significant symptoms that negatively impact quality of life. Less is known about the symptom burden of CF patients in relatively good health. Our data suggest that patients with earlier stage disease suffer from significant symptoms. These symptoms may not be adequately addressed routinely in clinic, as CF providers are tasked to address a multitude of different issues, and may not feel like they have the clinical expertise to assess and manage complex symptoms. (PsycINFO Database Record (c) 2015 APA, all rights reserved)

**Other**

**Title:** Cost-effectiveness of newborn screening for cystic fibrosis determined with real-life data

**Citation:** Journal of Cystic Fibrosis, March 2015, vol./is. 14/2(194-202), 1569-1993;1873-5010 (01 Mar 2015)


Language: English

Abstract: Background: Previous cost-effectiveness studies using data from the literature showed that newborn screening for cystic fibrosis (NBSCF) is a good economic option with positive health effects and longer survival. Methods: We used primary data to compare cost-effectiveness of four screening strategies for NBSCF, i.e. immunoreactive trypsinogen-testing followed by pancreatitis-associated protein-testing (IRT-PAP), IRT-DNA, IRT-DNA-sequencing, and IRT-PAP-DNA-sequencing, each compared to no-screening. A previously developed decision analysis model for NBSCF was fed with model parameters mainly based on a study evaluating two novel screening strategies among 145,499 newborns in The Netherlands. Results: The four screening strategies had cost-effectiveness ratios varying from 23,600 to 29,200 per life-year gained. IRT-PAP had the most favourable cost-effectiveness ratio. Additional life-years can be gained by IRT-DNA but against higher costs. When treatment costs reduce with 5% due to early diagnosis, screening will lead to financial savings. Conclusion: NBSCF is as an economically justifiable public health initiative. Of the four strategies tested IRT-PAP is the most economic and this finding should be included in any decision making model, when considering implementation of newborn screening for CF.

Title: Evaluation of mold exposure in cystic fibrosis patients' dwellings and allergic bronchopulmonary risk

Citation: Journal of Cystic Fibrosis, March 2015, vol./is. 14/2(242-247), 1569-1993;1873-5010 (01 Mar 2015)

Author(s): Rocchi S., Richaud-Thiriez B., Barrera C., Grenouillet F., Dalphin J.-C., Millon L., Reboux G.

Abstract: Very few studies have been conducted on cystic fibrosis (CF) patients' exposure to the indoor environment and, to our knowledge, there are no studies dealing with the link between specific fungal environmental exposure at home and fungal colonization resulting in allergic bronchopulmonary aspergillosis (ABPA). Fungal exposure of CF adult patients with ABPA (n = 4) with fungal sensitization (n = 7) and with no ABPA (n = 5) was assessed in 16 homes by dust sampling with electrostatic dust fall collectors (EDCs). Aspergillus fumigatus was specifically quantified by real-time quantitative polymerase chain reactions (qPCRs), and A. fumigatus DNA concentrations were significantly higher in homes of ABPA patients (p < 0.001). Results indicate that indoor fungal contamination could be a factor favoring ABPA and suggest that environmental surveys could help in preventing fungal risk in CF patients.

Title: Higher mobility scores in patients with cystic fibrosis are associated with better lung function

Citation: Pulmonary Medicine, 2015, vol./is. 2015/, 2090-1836;2090-1844 (2015)

Author(s): Thobani A., Alvarez J.A., Blair S., Jackson K., Gottlieb E.R., Walker S., Tangpricha V.
Abstract: Objective. The purpose of this study was to determine whether mobility and physical activity were associated with lung function in adults with cystic fibrosis (CF). Design. This was a prospective cohort observational study in an urban, academic, specialized care center. Participants were ambulatory, nonhospitalized adults with CF. Main Outcome Measures. Mobility was assessed monthly by the Life-Space Assessment (LSA) questionnaire and quarterly by pedometer. Lung function was assessed by spirometry. Results. Twenty-seven subjects participated. Subjects recorded mean pedometer steps of 20,213 +/- 11,331 over three days and FEV<sub>1</sub>% predicted of 77.48% +/- 22.60% over one year. The LSA score at enrollment was correlated with initial pedometer steps (r = 0.42 and P = 0.03), and mean LSA score over one year was correlated with mean number of steps (r = 0.51 and P = 0.007). LSA mobility and pedometer scores were correlated with FEV<sub>1</sub>% predicted at enrollment and throughout the study. Conclusions. Mobility and physical activity measured by LSA questionnaire and pedometer are positively associated with lung function in adults with CF. This study confirms the importance of mobility and physical activity and supports the utility of a simple office-based questionnaire as a measure of mobility in adults with CF.

Title: A contemporary survival analysis of individuals with cystic fibrosis: A cohort study

Citation: European Respiratory Journal, March 2015, vol./is. 45/3(670-679), 0903-1936;1399-3003 (01 Mar 2015)

Author(s): Stephenson A.L., Tom M., Berthiaume Y., Singer L.G., Aaron S.D., Whitmore G.A., Stanojevic S.

Abstract: Previously established predictors of survival may no longer apply in the current era of cystic fibrosis (CF) care. Our objective was to identify risk factors associated with survival in a contemporary CF population. We used the Canadian CF Registry, a population-based cohort, to calculate median age of survival and summarise patient characteristics from 1990 to 2012. Clinical, demographic and geographical factors, and survival were estimated for a contemporary cohort (2000-2012) using Cox proportional hazards models. There were 5787 individuals in the registry between 1990 and 2012. Median survival age increased from 31.9 years (95% CI 28.3-35.2 years) in 1990 to 49.7 years (95% CI 46.1-52.2 years) in the most current 5-year window ending in 2012. Median forced expiratory volume in 1 s improved (p=0.04) and fewer subjects were malnourished (p<0.001) over time. Malnourished patients (hazard ratio (HR) 2.1, 95% CI 1.6-2.8), those with multiple exacerbations (HR 4.5, 95% CI 3.2-6.4) and women with CF-related diabetes (HR 1.8, 95% CI 1.2-2.7) were at increased risk of death. Life expectancy in Canadians with CF is increasing. Modifiable risk factors such as malnutrition and pulmonary exacerbations are associated with an increased risk of death. The sex gap in CF survival may be explained by an increased hazard for death in women with CF-related diabetes.
Journal Tables of Contents

The most recent issues of the following journals:

- Journal of Cystic Fibrosis
- American Journal of Respiratory and Critical Care Medicine
- Thorax
- Chest

Click on the links for abstracts. If you would like any of these papers in full text then get in touch: bennet.jones@uhbristol.nhs.uk

Journal of Cystic Fibrosis

Vol. 14, iss. 2, March 2015

American Journal of Respiratory and Critical Care Medicine

Vol. 191, iss. 6, 15th March 2015

Thorax

Vol. 70, iss. 4, April 2015

Chest

Vol. 147, iss. 3, March 2015
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