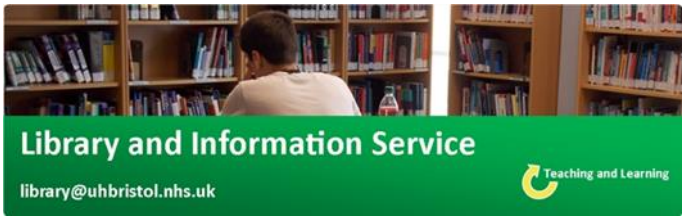
University Hospitals Bristol NHS
NHS Foundation Trust

Cystic Fibrosis

Current Awareness Bulletin

Nov/Dec 2016

Respecting everyone
Embracing change
Recognising success
Working together
Our hospitals.



Training Sessions 2016/17

All sessions are 1 hour

December (12.00)	
Fri 16th	**Literature Searching**
Mon 20th	**Critical Appraisal**
January (13.00)	
Tues 10th	**Literature Searching**
Wed 18th	**Critical Appraisal**
Thur 26th	**Statistics**

Your Outreach Librarian – Jo Hooper

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Contents

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New Additions to NICE, the Cochrane Library and UptoDate

NICE National Institute for Health and Care Excellence

Single versus combination intravenous anti-pseudomonal antibiotic therapy for people with cystic fibrosis

Source: Cochrane Database of Systematic Reviews - 01 December 2016

...infection due to Pseudomonas aeruginosa in cystic fibrosis (CF). Advantages of combination...intravenous antibiotic therapy in cystic fibrosis requires further evaluation. This...antibiotic therapy for treating people with cystic fibrosis.Search methodsWe searched the Cochrane...

Read Summary

Phase III study evaluating lumacaftor/ivacaftor (Orkambi) in patients with cystic fibrosis (CF) meets primary endpoint

07 November 2016 - Publisher: Biospace Inc.

A study evaluating use of Orkambi in children aged 6 to 11 years has met its primary endpoint of absolute change in lung clearance index (LCI2.5) through 24 weeks of treatment, demonstrating a statistically significant improvement in LCI2.5 when compared to

UKMi comment

Microbiological efficacy of early MRSA treatment in cystic fibrosis in a randomised controlled trial

15 November 2016 - Publisher: Thorax

Small study (n=45) found MRSA eradication with a defined protocol for newly acquired MRSA demonstrated microbiological efficacy with a large treatment effect, with 82% of participants being MRSA-negative in treatment arm vs. 26% in observation arm

UKMi comment

Cochrane Library

Pancreatic enzyme replacement therapy for people with cystic fibrosis

Usha Rani Somaraju and Arturo Solis-Moya

Online Publication Date: November 2016

Antifungal therapies for allergic bronchopulmonary aspergillosis in people with cystic fibrosis

Heather E Elphick and Kevin W Southern

Online Publication Date: November 2016

Eradication therapy for *Burkholderia cepacia* complex in people with cystic fibrosis

Kate H Regan and Jayesh Bhatt

Online Publication Date: November 2016

Macrolide antibiotics for non-cystic fibrosis bronchiectasis

Carol Kelly , David J Evans , James D Chalmers , Iain Crossingham , Sally Spencer , Nicola Relph , Lambert M Felix and Stephen J Milan

Online Publication Date: October 2016

UpToDate®

OpenAthens login required. Register here: https://openathens.nice.org.uk/

Cystic fibrosis: Carrier screening

Author: Katharine D Wenstrom, MD

Literature review current through: Nov 2016. | **This topic last updated:** Oct 21, 2016.

This topic will review CF carrier screening and reproductive options for carrier couples

Cystic fibrosis: Nutritional issues

Authors:Robert D Baker, MD, PhD Chris Coburn-Miller, MSRD, CSP Susan S Baker, MD, PhD

Literature review current through: Nov 2016. | **This topic last updated:** Dec 09, 2016.

The evaluation, monitoring, and treatment of nutritional problems will be addressed here.

Cystic fibrosis: Genetics and pathogenesis

Author: **Julie P Katkin, MD**

are updated as new evidence becomes available and our peer review process is complete.

Literature review current through: Nov 2016. | **This topic last updated:** Oct 26, 2016.

The genetics and pathogenesis of cystic fibrosis are discussed here.

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: library@uhbristol.nhs.uk

Journal of Cystic Fibrosis

November 2016, Volume 15, Issue 6

http://www.cysticfibrosisjournal.com/current

Equitable CF care as a basic human right

Kris De Boeck, p703–704 Full-Text HTMLPDF

Improving inhaled antibiotic treatment – Practice defeats the proof

C Kors van der Ent, p705–707 Full-Text HTMLPDF

Nutrition in CF — Two new important guidelines

Susan P Wolfe, p708–70 Full-Text HTMLPDF

News Article

News p710–711PDF

Commentary

Cystic fibrosis biomarker — Commentary

Anthony G. Durmowicz p712–713 Full-Text HTMLPDF

Review

Biomarkers for cystic fibrosis drug development

Marianne S. Muhlebach, p714–723 Full-Text HTMLPDF

Guidelines

Enteral tube feeding for individuals with cystic fibrosis: Cystic Fibrosis Foundation evidence-informed guidelines

Sarah Jane Schwarzenberg, p724–735 Full-Text HTMLPDF

CFTR/Basic Science

Early pulmonary disease manifestations in cystic fibrosis mice

Rebecca J. Darrah, p736–744 Full-Text HTMLPDF

A novel guluronate oligomer improves intestinal transit and survival in cystic fibrosis mice

Megan Vitko, p745–751 Full-Text HTMLPDF

Diagnosis/Screening

A product of immunoreactive trypsinogen and pancreatitis-associated protein as second-tier strategy in cystic fibrosis newborn screening

Sophia Weidler, p752–758 Full-Text HTMLPDFSupplemental Materials

Immunology

IL-22 exacerbates weight loss in a murine model of chronic pulmonary *Pseudomonas aeruginosa* infection

Hannah K. Bayes, p759–768 Full-Text HTMLPDFSupplemental Materials

Microbiology

Extensive cultivation of soil and water samples yields various pathogens in patients with cystic fibrosis but not *Burkholderia multivorans*

Charlotte Peeters, p769–775 Full-Text HTMLPDFSupplemental Materials

Eradication failure of newly acquired *Pseudomonas aeruginosa* isolates in cystic fibrosis

Malena Cohen-Cymberknoh, p776–782 Full-Text HTMLPDF

Epidemiology/Models of Care

Probability of IV antibiotic retreatment within thirty days is associated with duration and location of IV antibiotic treatment for pulmonary exacerbation in cystic fibrosis

D.R. VanDevanter, p783–790 Full-Text HTMLPDF

Cystic fibrosis in Latin America—Improving the awareness

Luiz Vicente Ribeiro p791–793 Full-Text HTMLPDF

Respiratory pathogens mediate the association between lung function and temperature in cystic fibrosis

Joseph M. Collaco, p794–801 Full-Text HTMLPDF Supplemental Materials

Clinical Trials

Continuous alternating inhaled antibiotic therapy in CF: A single center retrospective analysis

C. Van de Kerkhove, p802–808 Full-Text HTMLPDF

Continuous alternating inhaled antibiotics for chronic pseudomonal infection in cystic fibrosis

Patrick A. Flume, p809–815 Full-Text HTMLPDFSupplemental Materials

CT-abnormalities, bacteriology and symptoms of sinonasal disease in children with Cystic Fibrosis

M.C. Berkhout,p816–824 Full-Text HTMLPDF

Gastroenterology/Nutrition

Bioelectrical impedance in young patients with cystic fibrosis: Validation of a specific equation and clinical relevance

A.M. Charatsi, p825–833 Full-Text HTML PDF Supplemental Materials

Ursodeoxycholic acid treatment is associated with improvement of liver stiffness in cystic fibrosis patients

Cathelijne van der Feen,p834–838 Full-Text HTMLPDF

Endocrinology

Variation of glucose tolerance in adult patients with cystic fibrosis: What is the potential contribution of insulin sensitivity?

Valérie Boudreau,p839–845 Full-Text HTMLPDF

Psychosocial

Long-term work participation among cystic fibrosis patients undergoing lung transplantation

Katelyn Krivchenia, p846–849 Full-Text HTMLPDFSupplemental Materials

Online only Papers

Sino nasal inhalation of isotonic versus hypertonic saline (6.0%) in CF patients with chronic rhinosinusitis — Results of a multicenter, prospective, randomized, double-blind, controlled trial

Jochen G. Mainz, e57–e66 Full-Text HTMLPDF

Bone demineralization is improved by ivacaftor in patients with cystic fibrosis carrying the p.Gly551Asp mutation

Isabelle Sermet-Gaudelus, e67–e69 Full-Text HTMLPDF

Coronary artery disease in cystic fibrosis: An emerging concern?

Kate Skolnik, e70–e71 Full-Text HTMLPDF

American Journal of Respiratory and Critical Care Medicine

December 1 2016, Volume 194, Issue 11

http://www.atsjournals.org/toc/ajrccm/current

Abstract | Full Text | PDF (731 KB) | related editorial

Tuberculosis and Mycobacterial Disease

Detection and Quantification of Differentially Culturable Tubercle Bacteria in Sputum from Patients with Tuberculosis

Melissa D. Chengalroyen, pp. 1532–1540

Abstract | Full Text | PDF (733 KB) | related editorial

Beyond The Blue: What Fellows Are Reading in Other Journals

Complications of Central Venous Catheters, Rapid On-Site Specimen Evaluation for Lung Cancer Genotyping, and Endobronchial Valves for Emphysema

Parimalkumar Chaudhari, pp. 1541–1545

First Page | Full Text | PDF (521 KB)

Images in Pulmonary, Critical Care, Sleep Medicine and the Sciences

Is This Heart Normal?

Keren Armoni-Domany, pp. 1546–1547

Citation | Full Text | PDF (600 KB)

Esophagus-like Bronchus in an Adult with Common Variable Immunodeficiency Disease

Jing Zhao, pp. e17–e18

Citation | Full Text | PDF (1408 KB)

Correspondence

Validation of Administrative Definitions of Invasive Mechanical Ventilation across 30 Intensive Care Units

Meeta Prasad Kerlin, pp. 1548–1552

First Page | Full Text | PDF (466 KB)

Omalizumab Is Associated with Reduced Acute Severity of Rhinovirus-triggered Asthma Exacerbation

David B. Kantor, pp. 1552–1555

First Page | Full Text | PDF (454 KB)

Can Dead Space Ventilation Really Be Measured without PaCO2?

Laurent Plantier, pp. 1555–1556

Citation | Full Text | PDF (439 KB)

Reply: Can Dead Space Ventilation Really Be Measured without PaCO2?

Kirk Kee, pp. 1556–1557

Citation | Full Text | PDF (428 KB)

Patient Education/Information Series

Using a Home Ventilator with a Child

pp. P21–P22

Citation | PDF (327 KB)

Thorax

December 2016, Volume 71, Issue 12

http://thorax.bmj.com/content/current

Highlights from this issue

The Triumvirate Thorax 2017;72:1 [Extract][Full text][PDF] FREE

Editorials

What drives neutrophils to the alveoli in ARDS?

Rachel L Zemans, Thorax 2017;72:1 1-3 [Extract][Full text][PDF]

Expanding pulmonary rehabilitation capacity. One size won't fit all

Mike Morgan Thorax 2017;72:1 4-5 [Extract][Full text][PDF]

Optimum low dose CT screening interval for lung cancer: the answer from NELSON?

David R Baldwin, Thorax 2017;72:1 6-7 [Extract][Full text][PDF]

Outcomes from hospitalised acute exacerbations of COPD: a bundle of optimism?

William D-C Man, Thorax 2017;72:1 8-9 [Extract][Full text][PDF]

Macrolides, inflammation and the lung microbiome: untangling the web of causality

Robert P Dickson, Thorax 2017;72:1 10-12 [Extract][Full text][PDF]

Original article: Randomised, double-blind, placebo-controlled trial with azithromycin selects for anti-inflammatory microbial metabolites in the emphysematous lung

Leopoldo N Segal, Thorax 2017;72:1 13-22 [Abstrct][Full text][PDF]

Original article: A validation of the National Early Warning Score to predict outcome in patients with COPD exacerbation

Luke E Hodgson, Thorax 2017;72:1 23-30 [Abstract][Full text][PDF]

Original article: A systematic review of the effectiveness of discharge care bundles for patients with COPD

Maria B Ospina, Thorax 2017;72:1 31-39 [Full text][PDF][Request permissions]

Cystic fibrosis

Original article: Aminoglycoside resistance of Pseudomonas aeruginosa in cystic fibrosis results from convergent evolution in the mexZ gene

Michelle H Prickett, Thorax 2017;72:1 40-47 [Abstract][Full text][PDF]

Original article: Final screening round of the NELSON lung cancer screening trial: the effect of a 2.5-year screening interval

Uraujh Yousaf-Khan, Thorax 2017;72:1 48-56 [Abstract][Full text][PDF]

Rehabilitation

Original article: Home-based rehabilitation for COPD using minimal resources: a randomised, controlled equivalence trial

Anne E Holland, Thorax 2017;72:1 57-65 [Abstract][Full text][PDF]

Respiratory research

Original article: Evidence for chemokine synergy during neutrophil migration in ARDS

Andrew E Williams, Thorax 2017;72:1 66-73 [Abstract][Full text][PDF]

Original article: Latrophilin receptors: novel bronchodilator targets in asthma

A Faiz, Thorax 2017;72:1 74-82 [Abstract][Full text][PDF]

State of the art review

Chest electrical impedance tomography examination, data analysis, terminology, clinical use and recommendations: consensus statement of the TRanslational EIT developmeNt stuDy group

Inéz Frerichs, Thorax 2017;72:1 83-93 [Abstract][Full text][PDF][Online Supplements]

Research letter

The accuracy of pleural ultrasonography in diagnosing complicated parapneumonic pleural effusions

Philip Z Svigals, Thorax 2017;72:1 94-95 [Abstract][Full text][PDF]

Chest clinic

Case based discussion: It ain't necessarily so: a surprising lower airway infection

Rossa Brugha, Thorax 2017;72:1 96-97 Published [Extract][Full text][PDF]

Images in thorax: Sevelamer crystals in the bronchus: a case report

Shroque Zaher, Thorax 2017;72:1 98-99 [Extract][Full text][PDF]

Journal club summaries: What's hot that the other lot got

Ben Soar Thorax 2017;72:1 100 [Extract][Full text][PDF]

Chest

December 2016, Volume 150, Issue 6

http://journal.publications.chestnet.org/issue.aspx

Editorial

FDA Encourages Reporting of Tobacco Product Adverse Experiences

Sandra S. Retzky, Chest. 2016;150(6):1169-1170.

Endobronchial Ultrasonography: A Sublime Procedure and a Guide to the Proper Valuation of Health Care

Kevin L. Kovitz, MD, FCCPChest. 2016;150(6):1171-1173.

The High Road, the Low Road, or Both: Effects of Positive Airway Pressure Route of Administration on Treatment Efficacy for OSA

Nicholas J. Cutrufello, MD and Lee K. Brown, MD, FCCP Chest. 2016;150(6):1174-1176.

Nontuberculous Mycobacterial Disease Therapy: Take It to the Limit One More Time

David E. Griffith, MD, FCCP and Timothy R. Aksamit, MD

Chest. 2016;150(6):1177-1178.

Cost-effectiveness of Fluid Resuscitation of Critically Ill Adults: New Insights From Uncharted Territory

Craig M. Lilly, Chest. 2016;150(6):1179-1180.

Editorials: Point and Counterpoint

POINT: Do the Benefits Outweigh the Risks for Most Patients Under Consideration for Inferior Vena Cava Filters? Yes

Brian Funaki, Chest. 2016;150(6):1181-1182.

COUNTERPOINT: Do the Benefits Outweigh the Risks for Most Patients Under Consideration for Inferior Vena Cava Filters? No

Mark L. Lessne, Chest. 2016;150(6):1182-1184.

Rebuttal From Drs Funaki and Haskal

Brian Funaki, MD and Ziv J. Haskal, MD

Chest. 2016;150(6):1184-1185.

Mark L. Lessne,

Chest. 2016;150(6):1185-1186.

Commentary

Macrolides for Clinically Significant Bronchiectasis in Adults: Who Should Receive This Treatment?

Adam T. Hill, MDChest. 2016;150(6):1187-1193.

Original Research: Sleep Disorders

Impact of Acute Changes in CPAP Flow Route in Sleep Apnea Treatment

Rafaela G.S. Andrade, Chest. 2016;150(6):1194-1201.

Effect of CPAP Withdrawal on BP in OSA: Data from Three Randomized Controlled Trials

Esther I. Schwarz, Chest. 2016;150(6):1202-1210 Supplemental materials

Original Research: Chest Infections

Oral Macrolide Therapy Following Short-term Combination Antibiotic Treatment of Mycobacterium massiliense Lung Disease

Won-Jung Koh, Chest. 2016;150(6):1211-1221. Supplemental materials

Lung Function Decline According to Clinical Course in Nontuberculous Mycobacterial Lung Disease

Hye Yun Park, Chest. 2016;150(6):1222-1232.

Antipsychotic Use and Risk of Hospitalization or Death Due to Pneumonia in Persons With and Those Without Alzheimer Disease

Anna-Maija TolppanenChest. 2016;150(6):1233-1241. Supplemental materials

Original Research: Asthma

Association Between Insomnia and Asthma Burden in the Severe Asthma Research Program (SARP) III

Faith S. Luyster, Chest. 2016;150(6):1242-1250.Supplemental materials

Original Research: Critical Care

Culture-Negative Severe Sepsis: Nationwide Trends and Outcomes

Shipra Gupta, Chest. 2016;150(6):1251-1259. Supplemental materials

Risk Factors for In-Hospital Mortality in Smoke Inhalation-Associated Acute Lung Injury: Data From 68 United States Hospitals

Sameer S. Kadri, Chest. 2016;150(6):1260-1268. Supplemental materials

Original Research: COPD

A Subnational Analysis of Mortality and Prevalence of COPD in China From 1990 to 2013: Findings From the Global Burden of Disease Study 2013

Peng Yin, PhD, Chest. 2016;150(6):1269-1280. Supplemental materials

Measuring Airway Remodeling in Patients With Different COPD Staging Using Endobronchial Optical Coherence Tomography

Ming Ding, Chest. 2016;150(6):1281-1290. Supplemental materials

Original Research: Antithrombotic Therapy

OSA Is a Risk Factor for Recurrent VTE

Alberto Alonso-Fernández, Chest. 2016;150(6):1291-1301.Supplemental materials

Direct Comparison of Dabigatran, Rivaroxaban, and Apixaban for Effectiveness and Safety in Nonvalvular Atrial Fibrillation

Peter A. Noseworthy, Chest. 2016;150(6):1302-1312. Supplemental materials

Original Research: Pulmonary Vascular Disease

Echocardiography Combined With Cardiopulmonary Exercise Testing for the Prediction of Outcome in Idiopathic Pulmonary Arterial Hypertension

Roberto Badagliacca, Chest. 2016;150(6):1313-1322.

Original Research: Genetic and Developmental Disorders

Lung Clearance Index in Adults and Children With Cystic Fibrosis

Katherine O'Neill, Chest. 2016;150(6):1323-1332.Supplemental materials

Original Research: Disaster Medicine

Bronchial Reactivity and Lung Function After World Trade Center Exposure

Thomas K. Aldrich,Chest. 2016;150(6):1333-1340.

Evidence-Based Medicine

Chronic Cough Due to Gastroesophageal Reflux in Adults: CHEST Guideline and Expert Panel Report

Peter J. Kahrilas, Chest. 2016;150(6):1341-1360.

Translating Basic Research Into Clinical Practice

Translational Research in Pleural Infection and Beyond

Y. C. Gary Lee, Chest. 2016;150(6):1361-1370.

Recent Advances in Chest Medicine

Therapeutic Approach to Adult Fibrotic Lung Diseases

Ayodeji Adegunsoye, Chest. 2016;150(6):1371-1386.

Topics in Practice Management

Endobronchial Ultrasound: Clinical Uses and Professional Reimbursements

Thomas R. Gildea, MD, FCCP and Katina Nicolacakis, MD, FCCPChest. 2016;150(6):1387-1393.

Contemporary Reviews in Critical Care Medicine

Critical Illness in Patients With Asplenia

Hollis R. O'Neal, Jr.Chest. 2016;150(6):1394-1402.

Pectoriloquy

Informed Consent

Joseph Keefe, EdD Chest. 2016;150(6):1403.

The Ambulance

Helen Reiss Chest. 2016;150(6):1404.

Health Care Proxy

Joseph Duemer Chest. 2016;150(6):1405.

Radiation Therapy

Joseph Duemer Chest. 2016;150(6):1405.

Correspondence

Potential Effects of Hypoxia Preconditioning in Obesity Hypoventilation Syndrome?

Martin Burtscher, MD, PhD

Chest. 2016;150(6):1406.

Response

Juan F. Masa,

Chest. 2016;150(6):1406-1407.

Does Nocturnal Hypoventilation Have a Protective Effect on Cardiovascular Comorbidity in Obesity Hypoventilation Syndrome?

Giorgio Castellana, Chest. 2016;150(6):1407-1408

Response

Juan F. Masa, M Chest. 2016;150(6):1408.

Introducing High-Sensitivity Cardiac Troponin T as a Biomarker of OSA-Related Cardiovascular Morbidity in Obesity Hypoventilation Syndrome

Denis Monneret, Chest. 2016;150(6):1408-1409.

Response

Juan F. Masa, Chest. 2016;150(6):1409-1410.

Cardiovascular Protection From Severe OSA: The Pickwickian Paradox: Is Bigger Really Better?

Yashasvi Chugh, MD and Robert T. Faillace, MD Chest. 2016;150(6):1410-1411.

Response

Juan F. Masa, Chest. 2016;150(6):1411

Cancer and OSA: Beyond Hypoxia

Reviewer Acknowledgment

Selected Reports

Ultrasound Corner

Chest Imaging and Pathology for Clinicians

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**
- **Nutritional**
- **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: library@uhbristol.nhs.uk

Medical

Incident stenotrophomonas infection and lung function decline in patients with cystic fibrosis

Source: Pediatric Pulmonology; Oct 2016; vol. 51 ; p. 313-314

Publication Type(s): Journal: Conference Abstract

Author(s): Barsky E.; Williams K.; Priebe G.P.; Sawicki G.S.

Abstract: Background: Stenotrophomonas maltophilia (SM) is a multi-drug resistant bacterium with increasing prevalence among patients with cystic fibrosis (CF). In 2014, the US CF population prevalence of SM was 13.5%. The impact of incident SM infection on lung function decline and other outcomes is unclear. Objectives: 1) To determine if acquisition of SM is associated with accelerated lung function decline, and 2) To identify clinical differences following incident infection between patients with chronic and intermittent SM infection. **(Abstract Edited)**

A multidisciplinary approach to improving the diagnosis and management of CFRD in an adult cystic fibrosis center

Source: Pediatric Pulmonology; Oct 2016; vol. 51 ; p. 429-430

Publication Type(s): Journal: Conference Abstract

Author(s): Broadhead K.; Kim J.; Bajaj M.; Connealy C.; Wheeler C.; Katz M.; Barto T.

Abstract: Introduction: Cystic fibrosis-related diabetes (CFRD) is the most common comorbid condition in patients with Cystic Fibrosis (CF). According to the 2014 cystic fibrosis Patient Registry Data Report, 35% of people with CF age 18 years or old have CFRD. It is associated with deterioration in pulmonary function, decline in nutrition status (Lanng et al. Eur J Pediatr. 1992;151:847), and increased risk of early death (Rodman et al. Medicine (Baltimore). 1986; 65:389-97). Objective: The

Baylor College of Medicine (BCM) Adult Cystic Fibrosis Center is a large adult center in Houston, Texas. There are 255 active patients in the 2015 registry data set, 99 of which have been diagnosed with either impaired glucose tolerance or CFRD. In light of the significant impact CFRD has on morbidity in adults with CF and the prevalence in our clinic population, it was determined that an organized multidisciplinary approach to diagnosis and management of this cohort is essential to improve clinical outcomes. We hypothesize that the formation of a CFRD multidisciplinary clinic and committee will lead to more timely diagnosis and improved management of CFRD**.(Abstract Edited)**

Microbiological

Comparison of two pancreatic enzyme products for exocrine insufficiency in patients with cystic fibrosis

Source: Journal of Cystic Fibrosis; Sep 2016; vol. 15 (no. 5); p. 675-680

Publication Type(s): Journal: Article

Author(s): Taylor C.J.; Thieroff-Ekerdt R.; Fleming R.; Shiff S.; Magnus L.; Gommoll C.

Abstract: Background Zenpep (APT-1008) is a pancreatic enzyme product for the treatment of exocrine pancreatic insufficiency (EPI) associated with cystic fibrosis (CF). Methods Zenpep and Kreon, both containing 25,000 lipase units, were compared in a randomised, double-blind, crossover, non-inferiority study for CF-associated EPI in patients aged > 12 years. Patients on a standardised diet and stabilised treatment were randomised to two treatment sequences: Zenpep/Kreon or Kreon/Zenpep. The primary efficacy endpoint was the coefficient of fat absorption over 72 h (CFA-72 h). Results 96 patients (mean age 19.2 years, 60.4% males) were randomised with 83 completers of both sequences comprising the efficacy population. Zenpep demonstrated non-inferiority and equivalence to Kreon in fat absorption (LS mean CFA-72 h: Zenpep, 84.1% [SE 1.1] vs. Kreon, 85.3% [SE 1.1]; p = 0.297). Safety and tolerability were similar. Conclusions Zenpep is comparable with Kreon in efficacy and safety for the treatment of adolescents and adults with CF-associated EPI. NCT01641393 Copyright © 2016 The Authors

Epidemiology of nontuberculous mycobacteria (NTM) amongst individuals with cystic fibrosis (CF)

Source: Journal of Cystic Fibrosis; Sep 2016; vol. 15 (no. 5); p. 619-623

Publication Type(s): Journal: Article

Author(s): Viviani L.; Zolin A.; Harrison M.J.; Haworth C.S.; Floto R.A.

Abstract: Background Infection by nontuberculous mycobacteria (NTM) in patients with cystic fibrosis (CF) is often associated with significant morbidity. Limited, conflicting results are published regarding risk factors for pulmonary NTM disease. We analysed factors potentially associated with

NTM in a large population of European patients with CF. Methods We investigated associations between presence of NTM and various factors for patients registered in the European Cystic Fibrosis Society Patient Registry. Results 374 (2.75%) of 13,593 patients studied had at least one positive NTM culture within the study year. Age- and FEV<ovid:inf>1</ovid:inf>-adjusted odds of NTM infection was more than 2.5 times higher (95%CI: 1.79; 3.60) in patients infected by Stenotrophomonas maltophilia than in patients not infected (p < 0.0001), 2.36 times higher (95%CI: 1.80;3.08) in patients with ABPA than without (p < 0.0001), 1.79 times higher (95%CI: 1.34; 2.38) in patients who use bronchodilators than in patients who don't (p < 0.0001), 1.49 times higher (95%CI: 1.18; 1.89) in patients who use inhaled antibiotics than in patients who don't (p = 0.001), and 1.30 times higher (95%CI: 1.02; 1.66) in patients who use rhDNase than in patients who don't (p = 0.032). Conclusions NTM-positive cultures in individuals with CF are associated with distinct clinical variables. Improved data collection identifying risk factors for NTM infection will allow more focused screening strategies, and influence therapeutic choices and infection control measures in high-risk patients.

Antimicrobial susceptibility of microorganisms isolated from sputum culture of patients with cystic fibrosis: Methicillin-resistant Staphylococcus aureus as a serious concern

Source: Microbial Pathogenesis; Nov 2016; vol. 100 ; p. 201-204

Publication Type(s): Journal: Article

Author(s): Mazloomi Nobandegani N.; Nabavizadeh Rafsanjani R.; Mamishi S.; Mahmoudi S.; Pourakbari B.; Hosseinpour Sadeghi R.; Najafi Sani M.; Farahmand F.; Motamed F.

Abstract: Introduction Infection is a major cause of morbidity and mortality in patients with cystic fibrosis (CF). Antimicrobial resistance of the bacterial spp. particularly methicillin resistance in Staphylococcus aureus has caused a lot of attention. The aim of this study was to describe the prevalence of S. aureus, Pseudomonas aeruginosa and Burkholderia cepacia-complex as well as their antimicrobial susceptibility patterns in CF patients in an Iranian referral pediatrics Hospital. **(Abstract Edited)**

The relationship between implementation of the 2013 CF foundation infection control guideline on the incidence and prevalence of pathogenic bacteria in respiratory cultures from patients with cystic fibrosis

Source: Pediatric Pulmonology; Oct 2016; vol. 51 ; p. 345

Publication Type(s): Journal: Conference Abstract

Author(s): Kim C.; Welter J.; DelaRiva-Velasco E.; Dozor A.; Nolan S.; Paul L.

Abstract: Introduction: The views of the cystic fibrosis (CF) community on optimal infection control practices have evolved over the years. In the past, it was thought that patients with CF rarely acquired organisms from other CF patients. More recent data suggests patient-to-patient transmission is possible. There is increasing evidence that implementation of strict infection control practices in CF clinics and inpatient units can decrease the spread of pathogenic bacteria. The 2013 update to the Infection Prevention and Control Guideline for CF reflects this emerging viewpoint on organism transmission in CF. This guideline was instituted at our CF center in August 2014. Objective: To determine the effect of implementation of the new infection control recommendations on incidence and prevalence of pathogenic organisms in patients at one CF center. **(Abstract Edited)**

Psychology

The sleep/wake cycle: Fatigue, sleep, insomnia and their effects on cystic fibrosis health; tackling sleep issues in the adult with CF

Source: Pediatric Pulmonology; Oct 2016; vol. 51 ; p. 136-138

Publication Type(s): Journal: Conference Abstract

Author(s): Franco R.

Abstract: Adults with CF report poor quality sleep more frequently and in some but not all objective studies there are more frequent awakenings and disturbed sleep architecture. The sleep issues result in measurable impact on quality of life and perhaps mood. Source of sleep complaints may be mood, chronic pain, digestive issues, obstructed breathing (lower airways and upper airways), sleep disorders, as well as typical triggers such as sleeping environment, work stress and even parental duties. Linking CF to sleep issues requires first an inventory of how cystic fibrosis impacts quality of life (QoL). (**Abstract Edited)**

CF care notebooks: Encouraging pediatric patients with cystic fibrosis to be active participants in their care

Source: Pediatric Pulmonology; Oct 2016; vol. 51 ; p. 475

Publication Type(s): Journal: Conference Abstract

Author(s): Gore W.; Leyva C.; Morse K..

Abstract: Background and Significance: Cystic fibrosis is no longer a disease confined to the pediatric population. As the life expectancy of those with CF continues to rise, it is important to prepare pediatric patients to live as independent adults. Transition and transfer are widely discussed within the CF community: "The timing of transfer to adult CF care often coincides with a dynamic period in

adolescents and young adulthood when lung function may be declining and treatment burden and complications of a multisystem disease are increasing." (Tuchman E, et al. Pediatrics. 2009;125:566-73). **(Abstract Edited)**

Nutrition

Improving nutrition in adult cystic fibrosis patients: A quality improvement project

Source: Pediatric Pulmonology; Oct 2016; vol. 51 ; p. 393-394

Publication Type(s): Journal: Conference Abstract

Author(s): Stamm J.A.; Shoff C.T.; Brosius H.; Savage J.; Ritter B.; McCurley D.; Roush L.; Michel S.

Abstract: Introduction: Prior research has shown that malnutrition is associated with reduced lung function and increased mortality in adult cystic fibrosis (CF) patients. Maintaining optimal nutrition is an essential part of CF care. Only 50% of our adult CF patients are at their goal BMI and therefore we undertook a quality improvement (QI) project to improve the nutrition status of our adult CF population. (**Abstract Edited)**

Efficacy of percutaneous endoscopic gastrostomy tube feedings in patients with cystic fibrosis

Source: Pediatric Pulmonology; Oct 2016; vol. 51 ; p. 432

Publication Type(s): Journal: Conference Abstract

Author(s): Khalaf R.; Green D.M.; Amankwah E.; Carr V.; Goldenberg N.; Martinez D.; Wilsey M.

Abstract: Background: Cystic fibrosis (CF) is the most common autosomal recessive disease in Caucasians. In a recent consensus statement, the Cystic Fibrosis Foundation emphasized that adequate nutrition is a vital element in preserving lung function and survival in patients with CF. Malnutrition is a common problem in patients with CF as a result of their inability to meet nutritional demands. Improving the nutritional status of patients with CF has been shown to have a positive effect on pulmonary function, respiratory status, body composition and survival. Few studies have investigated the outcomes and efficacy of nutritional supplementation via percutaneous endoscopic gastrostomy (PEG) tube feedings. (**Abstract Edited)**

Evaluating knowledge and attitudes about nutrition and weight among adults with cystic fibrosis

Source: Pediatric Pulmonology; Oct 2016; vol. 51 ; p. 409

Publication Type(s): Journal: Conference Abstract

Author(s): Wilson A.; Poch K.; Brayshaw S.; Trout C.; Fischler K.; McVey-Tyson A.; Rasmussen L.M.

Abstract: Background: It is widely agreed upon that nutritional status is an important, independent determinant of lung function and survival in individuals living with CF. The Cystic Fibrosis Foundation

(CFF) established the goal body mass index (BMI) for men to be a minimum of 23kg/m<ovid:sup>2</ovid:sup> and for women to be a minimum of 22kg/m<ovid:sup>2</ovid:sup>. However, nutritional knowledge and understanding of basic dietary concepts is low and there is a lack of awareness about the link between diet and disease. These problems can be attributed to poor education, inadequate adult-centered educational materials, or misconceptions by the care team. (**Abstract Edited)**

Lean body mass deficits and glucose tolerance in cystic fibrosis

Source: Pediatric Pulmonology; Oct 2016; vol. 51 ; p. 446

Publication Type(s): Journal: Conference Abstract

Author(s): Sheikh S.; Kelly A.; Rubenstein R.C.

Abstract: Objectives: Cystic fibrosis-related diabetes (CFRD) is associated with worse cystic fibrosis (CF) outcomes including lower body mass index (BMI). Lean body mass (LBM) deficits arising from hypoinsulinemia are suspected to at least partially mediate worsening pulmonary function, and we recently demonstrated that LBM is even more strongly associated with pulmonary function than BMI. However, changes in body composition in CFRD have received limited attention. Hypothesis: Worsening glucose tolerance as reflected by oral glucose tolerance test (OGTT) is associated with LBM deficits. **(Abstract Edited)**

Clinical impact of impaired glucose tolerance in children and adolescents with cystic fibrosis

Source: Pediatric Pulmonology; Oct 2016; vol. 51 ; p. 443

Publication Type(s): Journal: Conference Abstract

Author(s): Finocchiaro J.A.; Reinoso A.; Massa S.; Pietropaolo G.; Fernandez A.; Balbi V.; Diez G.; Fasano M.V.

Abstract: Introduction: Cystic fibrosis-related diabetes (CFRD) is a frequent comorbidity that produces impacts on pulmonary function (PF) and nutritional status (NS) which can be detected 3 to 6 years previous to diagnosis. Impaired glucose tolerance (IGT) defined as a fasting glycemia (FG) <126 mg/dL and 140-199 mg/dL 2 hours after administration of glucose on an oral glucose tolerance test (OGTT), correlates with lower PF and poorer NS, although it is not clear whether these can be tracked retrospectively. Other glucose tolerance alterations such as indeterminate (INDET) category (normal FG and 2 hour, with values over 200 mg/dL during the test) can be detected by a complete OGTT. We hypothesized that patients with IGT and INDET have worse PF and NS and that these are present previously to the detection of the alteration. Objective: To assess glucose tolerance in a

group of children and adolescents with cystic fibrosis (CF) followed at a CF center and the retrospective association with PF and NS data. (**Abstract Edited)**

Enteral tube feeding for individuals with cystic fibrosis: Evidence-informed guidelines-clinical pathway

Source: Pediatric Pulmonology; Oct 2016; vol. 51 ; p. 182-183

Publication Type(s): Journal: Conference Abstract

Author(s): McDonald C.M.

Abstract: Optimal nutritional status for individuals with cystic fibrosis (CF) is associated with improved pulmonary function. Enteral tube feeding can benefit nutritional status in some individuals with cystic fibrosis (CF). In 2015, the Cystic Fibrosis Foundation convened an expert panel to develop enteral tube feeding guidelines based on a systematic review of the evidence and expert opinion.1 The guidelines describe essential steps to standardize care, improve outcomes, and reduce cost in the care of individuals with CF who are determined to be candidates for enteral tube placement. Criteria for recommending enteral tube feeding, assessment of confounding causes of poor nutrition in CF, preparation for placement of the enteral feeding tube, management after placement, and education about enteral tube feeding for the individual and caregivers are addressed by the enteral tube feeding guidelines. Preoperative assessment and care. Prior to the placement of a gastrostomy, individuals with CF should be evaluated by a multidisciplinary team for any treatable conditions (including medical, psychosocial, and/or financial) contributing to nutritional decline. **(Abstract Edited)**

Other

Use of pancreatic enzyme management smartphone app in adult cystic fibrosis patients

Source: Pediatric Pulmonology; Oct 2016; vol. 51 ; p. 427

Publication Type(s): Journal: Conference Abstract

Author(s): Breeding Z.R.; Stephen M.J.

Prevalence and impact of pelvic floor dysfunction in an adult cystic fibrosis population: a questionnaire survey

Source: International Urogynecology Journal and Pelvic Floor Dysfunction; Oct 2016 ; p. 1-14

Publication Type(s): Journal: Article In Press

Author(s): Chambers R.; Reihill A.; Hough J.; Lucht A.

A targeted intervention improves infection control compliance on an inpatient cystic fibrosis unit

Source: Pediatric Pulmonology; Oct 2016; vol. 51 ; p. 394

Publication Type(s): Journal: Conference Abstract

Author(s): McNeal M.K.; Johnson S.; Adams V.; Burger S.; Polineni D.; Mermis J.

Investigating the improvement in physical and pulmonary endurance as measured by the modified shuttle walk test and FEV1 in pediatric patients admitted to the hospital for acute cystic fibrosis exacerbation

Source: Pediatric Pulmonology; Oct 2016; vol. 51 ; p. 373

Publication Type(s): Journal: Conference Abstract

Author(s): Griego B.; Tataria S.; Kirchner K.

Physiotherapy in end-stage cystic fibrosis-the role of home care

Source: Pediatric Pulmonology; Oct 2016; vol. 51 ; p. 408

Publication Type(s): Journal: Conference Abstract

Author(s): Parkinson H.F.; Channon K.; Elston C.; Reilly C.C.

The physiotherapy management of the critically ill patient with cystic fibrosis including DIOS, pneumothorax, hemoptysis and acute respiratory failure

Source: Pediatric Pulmonology; Oct 2016; vol. 51 ; p. 150-152

Publication Type(s): Journal: Conference Abstract

Author(s): Agent P.

Exercise: Relative Risk

The relative risk is the ratio of probability of an event (a specified outcome) occurring in one group (i.e. those exposed to a particular intervention) compared to those in another group (i.e. those not exposed – a control group).

The relative risk can be interpreted using the following chart. First, you must determine whether the event (the outcome measure) is adverse or beneficial.

Relative Risk	Adverse outcome (e.g. death)	Beneficial outcome (e.g. recovery of limb function)
<1	Intervention better than control	Intervention worse than control
1	Intervention no better or worse than control	Intervention no better or worse than control
>1	Intervention worse than control	Intervention better than control

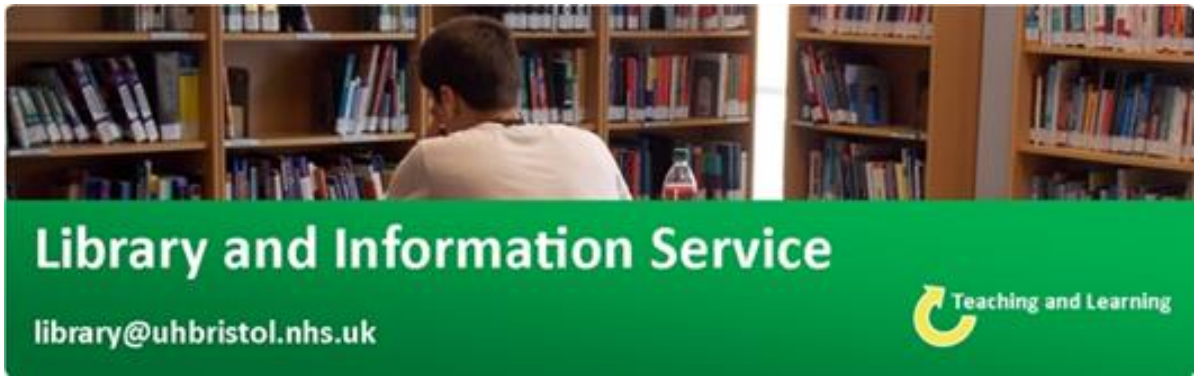
Have a go at interpreting the relative risks for these three studies using the chart above. Is the intervention better or worse than the control?

	Intervention	Population	Outcome measure (think: adverse or beneficial?)	Relative Risk
Study 1	Drug X	Adults at risk of a heart attack	Heart attack	1.2
Study 2	Therapy programme Y	Smokers	Smoking cessation	0.8
Study 3	Probiotic Z	Children on antibiotics	Diarrhoea	0.3

Find out more about relative risk in one of our Statistics training sessions.

For more details, email library@uhbristol.nhs.uk.

Answers: Study 1: worse; Study 2: worse; Study 3: better



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