

CF Reference List

(compiled between June 2024 –September 2024)

Adults & Adolescents

Amar S., Badeghiesh A., Baghfalaf H., Dahan MH.

Obstetric and neonatal outcomes among pregnant patients with cystic fibrosis.

Journal of Obstetrics & Gynecology and Reproductive Biology 2024; 300:219-223

Cohen-Cymberknob M., Ariel Dabby M., Gindi Reiss B., Melo Tanner J., Pérez G., Lechtzin N., Polverino E., Perez Miranda J., Gramegna A., Aliberti S., Levine H., Mussaffi H., Blau H., Prais D., Mei-Zahav M., Shteinberg M., Livnat G., Gur M., et al

Maternal and fetal outcomes in multiparous women with Cystic Fibrosis

Respiratory Medicine 2024; 228: e107654

Animal Model

Dastoor P., Muiler C., Garrison A., Egan M., Carlos Dos Reis D., Santos A., Ameen NA.

Localization and function of humanized F508del-CFTR in mouse intestine following activation of serum glucocorticoid kinase 1 and Trikafta.

European Journal of Pharmacology 2024; 978: e978:176771

Antimicrobials

Breen SKJ., Harper M., López-Causapé C., Rogers KE., Tait JR., Smallman TR., Lang Y., Lee WL., Zhou J., Zhang Y., Bulitta JB., Nation RL., Oliver A., Boyce JD., Landersdorfer CB.

Synergistic effects of inhaled aztreonam plus tobramycin on hypermutable cystic fibrosis *Pseudomonas aeruginosa* isolates in a dynamic biofilm mode evaluated by mechanism-based modelling and whole genome sequencing.

International Journal of Antimicrobial Agents 2024; 63(6): e107161

Fowler VG Jr., Das AF., Lipka-Diamond J., Ambler JE., Schuch R., Pomerantz R., Cassino C., Jáuregui-Pereido L., Moran GJ., Rupp ME., Lachiewicz AM., Kuti JL., Wise RA., Kaye KS., Zervos MJ., Nichols WG.

Exebacase in Addition to Standard-of-Care Antibiotics for *Staphylococcus aureus* Bloodstream Infections and Right-Sided Infective Endocarditis: A Phase 3, Superiority-Design, Placebo-Controlled, Randomized Clinical Trial (DISRUPT).

Clinical Infectious Diseases. 2024; 78: 1473-1481.

Jjingo CJ., Bala S., Waack U., Needles M., Bensman TJ., McMaster O., Smith T., Blakely B., Chan IZ., Puthawala K., Dixon C., Kim Y., Lim R., Colangelo P., St Clair C., Nambiar S., Moss RB., Botros R., Bazaz R., Denning DW., Marr KA., et al

Food and Drug Administration Public Workshop Summary-Addressing Challenges in Inhaled Antifungal Drug Development.

Clinical Infectious Diseases. 2024; 78: 1564-1570

Li D., Donnelley M., Parsons D., Habgood MD., Schneider-Futschik EK.

Extent of foetal exposure to maternal elexacaftor/tezacaftor/ivacaftor during pregnancy.

British Journal of Pharmacology 2024; 181: :2413-2428.

Mudgil U., Khullar L., Chadha J., Prerna., Harjai K.

Beyond antibiotics: Emerging antivirulence strategies to combat *Pseudomonas aeruginosa* in cystic fibrosis.

Microbial Pathogenesis 2024; 193: e106730.

Sherwood SJ., Tak C., Bhakta ZN., Packer K., Jacobs H., Liou TG., Young DC.

A comparison of aminoglycoside antibiotic serum concentrations collected by peripheral veins and peripherally inserted central catheters in adults with cystic fibrosis.

Pediatric Pulmonology 2024; 59: 1740-1746

Cardiology

Amoakon JP., Mylavarampu G., Amin RS., Naren AP.

Pulmonary Vascular Dysfunctions in Cystic Fibrosis.

Physiology 2024; 1:39(4):

Cell Biology

Skinner WH., Robinson N., Hardisty GR., Gray RD., Campbell CJ.

SERS Microsensors for the Study of pH Regulation in Cystic Fibrosis Patient-Derived Airway Cultures.

ACS Sensors 2024; 9(5): :2550-2557

CFTR

Akram A., Sakhawat A., Ghani MU., Khan MU., Rehman R., Ali Q., Jin-Liang P., Ali D.

Silibinins and curcumin as promising ligands against mutant cystic fibrosis transmembrane regulator protein.

AMB Express 2024; 14(1): e84

Coatti GC., Vaghela N., Gillurkar P., Leir SH., Harris A.

A promoter-dependent upstream activator augments CFTR expression in diverse epithelial cell types.

Biochimica et Biophysica Acta - Gene Regulatory Mechanisms. 2024; 1867(2): e195031.

- Crunkhorn S.**
Discovering CFTR modulators.
Nature Reviews Drug Discovery. 2024; 23(8): 581
- Gao X., Yeh HI., Yang Z., Fan C., Jiang F., Howard RJ., Lindahl E., Kappes JC., Hwang TC.**
Allosteric inhibition of CFTR gating by CFTRinh-172 binding in the pore.
Nature Communications 2024; 15(1): e6668
- Landess L., Prieur MG., Brown AR., Dallon EP.**
Exploring perceptions of and decision-making about CFTR modulators.
Pediatric Pulmonology 2024; 59: 1614-1621
- Loske J., Völler M., Lukassen S., Stahl M., Thürmann L., Seegerbarth A., Röhmel J., Wisniewski S., Messingschläger M., Lorenz S., Klages S., Eils R., Lehmann I., Mall MA., Graeber SY., Trump S.**
Pharmacological Improvement of Cystic Fibrosis Transmembrane Conductance Regulator Function Rescues Airway Epithelial Homeostasis and Host Defense in Children with Cystic Fibrosis. (Comment in *Am J Respir Crit Care Med*. 2024 Jun 1;209(11):1292-1293.)
American Journal of Respiratory and Critical Care Medicine 2024; 209: 1338-1350.
- Luo S., Rollins S., Schmitz-Abe K., Tam A., Li Q., Shi J., Lin J., Wang R., Agrawal PB.**
The solute carrier family 26 member 9 modifies rapidly progressing cystic fibrosis associated with homozygous F508del CFTR mutation.
Clinica Chimica Acta. 2024; 561: e119765.
- Meng X., Ford RC.**
Investigation of F508del CFTR unfolding and a search for stabilizing small molecules
Archives of Biochemistry and Biophysics. 2024; 758: e110050
- No authors listed**
Erratum: Inflammatory Activity of Epithelial Stem Cell Variants from Cystic Fibrosis Lungs Is Not Resolved by CFTR Modulators.
American Journal of Respiratory and Critical Care Medicine 2024; 210(2): e249.
- Reix P., Chassagnon G..**
The younger, the better: lessons learned from real-world studies on CFTR modulators in young children.
European Respiratory Journal. 2024; 64(3): e2401178
- Tanjala AC., Jiang JX., Eckford PDW., Ramjeesingh M., Li C., Huan LJ., Langeveld G., Townsend C., Paone DV., Busch-Petersen J., Pekhletschi R., Tang L., Raju V., Rowe SM., Bear CE.**
Comparison of a novel potentiator of CFTR channel activity to ivacaftor in ameliorating mucostasis caused by cigarette smoke in primary human bronchial airway epithelial cells.
Respiratory Research 2024; 25(1): e269
- van der Sluijs P., Hoelen H., Schmidt A., Braakman I.**
The Folding Pathway of ABC Transporter CFTR: Effective and Robust.
Journal of Molecular Biology. 2024; 436(14): e168591
- Varkki SD., Aaron R., Chapla A., Danda S., Medhi P., Jansi Rani N., Paul GR.**
CFTR mutations and phenotypic correlations in people with cystic fibrosis: a retrospective study from a single centre in south India.
Lancet Regional Health - Southeast Asia 2024; 27: e100434.
- Wu M., Davis JD., Zhao C., Daley T., Oliver KE.**
Racial inequities and rare CFTR variants: Impact on cystic fibrosis diagnosis and treatment.
Journal of Clinical and Translational Endocrinology 2024; 36: e100344
- ## Clinical
- National Health and Medical Research Council Australia**
Long-term outcomes of early exposure to repeated general anaesthesia in children with cystic fibrosis (CF-GAIN): a multicentre, open-label, randomisedcontrolled phase 4 trial.
Lancet Respiratory Medicine 2024; 12: 703-713
- Bush A.**
Learning from cystic fibrosis: How can we start to personalise treatment of Children's Interstitial Lung Disease (chILD)?
Paediatric Respiratory Reviews 2024; 50: 46-53
- Carroll BJ., Ostrenga JS., Fink AK., Antos NJ., Cromwell EA., Ren CL.**
Clinical outcomes at 9-10 years of age in children born with cystic fibrosis transmembrane conductance regulator related metabolic syndrome.
Pediatric Pulmonology. 2024; 59: 1606-1613
- Cuneo A., Smith-Thomas T., Marsac M.**
Opportunities for trauma-informed medical care in cystic fibrosis
Pediatric Pulmonology 2024; 56: 1814-1816
- Fischer DL., Vendruscolo FM., de Oliveira JR., Donadio MVF.**
Association of Neutrophil-Lymphocyte Ratio with Clinical Outcomes in Patients with Cystic Fibrosis.
Indian Journal of Pediatrics 2024; 91: 982
- Guo J., King I., Hill A.**
International disparities in diagnosis and treatment access for cystic fibrosis.
Pediatric Pulmonology 2024; 59: 1622-1630
- Mattison G., Canfell OJ., Smith D., Forrester D., Reid D., Töyräs J., Dobbins C.**
An excellent servant but a terrible master": Understanding the value of wearables for self-management in people with cystic fibrosis and their healthcare providers - A qualitative study.
International Journal of Medical Informatics 2024; 189: e105532

Oppelaar MC., Emond Y., Bannier MAGE., Reijers MHE., van der Vaart H., van der Meer R., Altenburg J., Conemans L., Rottier BL., Nuijsink M., van den Wijngaart LS., Merkus PJFM., Heinen M., Roukema J.
Potential, Pitfalls, and Future Directions for Remote Monitoring of Chronic Respiratory Diseases: Multicenter Mixed Methods Study in Routine Cystic Fibrosis Care.
Journal of Medical Internet Research 2024; 26: e54942

Sathe M., Stein A

Remaining barriers to normalcy in cystic fibrosis: Considerations in GI, liver, and nutrition
Pediatric Pulmonology 2024; S1: S5

Smith AD., Schwartzman G., Lyons CE., Flowers H., Albon D., Greer K., Lonabaugh K., Zlotoff BJ

Cutaneous manifestations of cystic fibrosis.

Journal of the American Academy of Dermatology 2024; 91: 490-498

Sun BZ., Sawicki GS.

Advances in Care and Outcomes for Children with Cystic Fibrosis.
Clinics in Chest Medicine 2024; 45: 625-637

Tural DA., Emiralioglu N., Akin S., Alboga D., Ozsezen B., Buyuksahin HN., Guzelkas I., Kasikci M., Sunman B., Gungor I., Yalcin E., Dogru D., Kiper N., Demirel AH., Ozcelik U.

Correction to: Galectin-3 levels in children with cystic fibrosis.

European Journal of Pediatrics 2024; 183: 3633-3634

Diabetes

Alkhateeb AA., Mancl LA., Ramos KJ., Rothen ML., Kotsakis GA., Trencé DL., Chi DL.

The association between cystic fibrosis-related diabetes and periodontitis in adults: A pilot cross-sectional study.
PLoS One. 2024; 19(6): e0305975

Chokkalla AK., Tuley P., Kurteca M., Ona H., Ruiz FE., Devaraj S.

Cystic fibrosis-related diabetes screening at a large pediatric center.
Laboratory Medicine 2024; 55: 580-584

Saegebrecht LS., Röhlig M., Schaub F., Ballmann M., Stachs O., Fischer DC

Glycemic Variability and the Thickness of Retinal Layers in Cystic Fibrosis Patients with and without Cystic Fibrosis Related Diabetes.
Current Eye Research 2024; 49: 759-767.

Endocrinology

Milano RV., Morneau-Gill K., Kamal HY., Barkin JA., Chadwick CB.

Pancreatitis in cystic fibrosis: Presentation, medical and surgical management, and the impact of modulator therapies.
Pediatric Pulmonology 2024; S1: :S53-S60

Ramsey ML., Galante GJ.

Pancreas and pancreatitis: Exocrine pancreatic insufficiency.
Pediatric Pulmonology 2024; S1: S44-S52

Epidemiology

Rafique H., Safdar A., Ghani MU., Akbar A., Awan FI., Naeem Z., Amar A., Awan MF., Wajahat Ullah S., Shaikh RS.

Exploring the diversity of CFTR gene mutations in cystic fibrosis individuals of South Asia.
Journal of Asthma. 2024; 61: 511-519

Gastroenterology

Bertolini A., Nguyen M., Zehra SA., Taleb SA., Bauer-Pisaní T., Palm N., Strazzabosco M., Fiorotto R..

Prominent role of gut dysbiosis in the pathogenesis of cystic fibrosis-related liver disease in mice.
Journal of Hepatology 2024; 81: :429-440.

Lee T., Nissenbaum C.

Improving gastrointestinal health in children and young people with cystic fibrosis.
Archives of Disease in Childhood. 2024; 109: 525-526

Maisonneuve P.

Characteristics and Outcomes of Patients with Cystic Fibrosis and Pancreatic Cancer: A Large Database Analysis.
Journal of Gastrointestinal Cancer. 2024; 55(3): 1467

Raza Z., Islam BN., Hachem CY., Cummings LC.

Evolving data on risk and current screening recommendations for colorectal cancer in cystic fibrosis: Pre- and posttransplant.
Pediatric Pulmonology. 2024; S1: S91-S97

Roca M., Masip E., Colombo C., Boon M., Hulst JM., Garriga M., de Koning BAE., Bulfamante A., de Boeck K., Ribes-Koninkx C., Calvo-Lerma J.

Long-term evaluation of faecal calprotectin levels in a European cohort of children with cystic fibrosis.
Archives of Disease in Childhood 2024; 109: 552-556.

Vélez C., Neuringer I., Schwarzenberg S.

The foregut in cystic fibrosis.

Pediatric Pulmonology 2024; S1: :S61-S69

Wells H., Bough G., Stedman F., Ekerin AR., Hall NJ

Investigations, management and outcome of neonates presenting with distal intestinal obstruction: challenging the need for contrast enemas
Pediatric Surgery International. 2024; 40(1): e154

Gene Therapy

Bulcaen M., Carlon MS.

Gene editing flows to the lungs. (Comment on Science. 2024 Jun 14;384(6701):1196-1202)
Science. 2024; 384: 1175-1176

Porter JJ., Lueck JD.,

A cystic fibrosis gene editing approach that is on target.
Molecular Therapy - Nucleic Acids. 2024; 35(2): e102197.

General Review

Mall MA., Burgel PR., Castellani C., Davies JC., Salathe M., Taylor-Cousar JL.

Cystic fibrosis.

Nature Reviews Disease Primers 2024; 10(1): e53

Savant A., Lyman B., Bojanowski C., Upadia J.

Cystic Fibrosis.

in *GeneReviews* 2024; : 1993-2024.

Genetics

Guo R., Zou Y., Guo Y., Gao W.

Compound heterozygous CFTR variants (Q1352H and 5T; TG13) in a Chinese patient with cystic fibrosis.
Diagnostic Pathology. 2024; 19(1): e107

Kumar M., Aaron R., Varkki SD., Danda S., Ranganathan S., Paul GR.

A rare variant c.1802T>C (p. Ile601Thr) associated with severe phenotype among people with cystic fibrosis from south India, and potential genetic admixture in Réunion, France
Pediatric Pulmonology. 2024; 59: 1820-1825

Growth & Development

Gabel ME., Gaudio RE., Shaikhkhailil AK.

Improving growth in infants with CF.

Pediatric Pulmonology. 2024; S1: :S17-S26

Stewart KL., Szczesniak R., Liou TG.

Predicting weight gain in patients with cystic fibrosis on triple combination modulator.

Pediatric Pulmonology 2024; 59: 1724-1730

Immunology & Inflammation

Bardin E., Dietrich C., Attailia M., Ferroni A., Jamet A., Lezmi G., Sermet-Gaudelus I., Leite-de-Moraes M.

Restored Cytokine-Producing Capacities of Mucosal-associated Invariant T Cells in Pediatric Cystic Fibrosis Patients Treated with Elexacaftor/Tezacaftor/Ivacaftor.
American Journal of Respiratory and Critical Care Medicine 2024; 210: :243-245

Capone M., Vanni A., Salvati L., Lamacchia G., Mazzoni A., Maggi L., Cosmi L., Liotta F., Romagnani P., Cirillo L., Buti E., Terlizzi V., Azzari C., Citera F., Barbatì F., Rossolini GM., Bresci S., Borchi B., Cavallo A., Mencarini J., et al

Effect of antimetabolite regimen on cellular and humoral immune response to SARS-CoV-2 vaccination in solid organ transplant recipients.

Immunology Letters 2024; 268: e106886.

Fuchs T., Zlamy M., Zöggeler T., Appelt D., Niedermayr K., Siedl A., Gasser V., Eder J., Ellemunter H.

Detection of cytokines in nasal lavage samples of patients with cystic fibrosis: comparison of two different cytokine detection assays.
BMC Pulmonary Medicine 2024; 24(1): e286

Venegas Garrido C., Mukherjee M., Svenningsen S., Nair P.

Eosinophil-mucus interplay in severe asthma: Implications for treatment with biologicals.

Allergology International 2024; 73: 351-361

Yonker LMv., Kinane TB

Depths of Dysfunctional Epithelial and Immune Crosstalk in Cystic Fibrosis Revealed. - (Comment on Am J Respir Crit Care Med. 2024 Jun 1;209(11):1338-1350)

American Journal of Respiratory and Critical Care Medicine. 2024; 209: 1292-1293.

Liver Disease

Declercq M., Treps L., Geldhof V., Conchinha NV., de Rooij LPMH., Subramanian A., Feyeux M., Cotinat M., Boeckx B., Vinckier S., Dupont L., Vermeulen F., Boon M., Proesmans M., Libbrecht L., Pirenne J., Monbaliu D., Jochmans I., Dewerchin M., et al

Single-cell RNA sequencing of cystic fibrosis liver disease explants reveals endothelial complement activation.

Liver International 2024; 44: 2382-2395

Eldredge JA., Oliver MR., Ooi CY.

Cystic fibrosis liver disease in the new era of cystic fibrosis transmembrane conductance regulator (CFTR) modulators.

Paediatric Respiratory Reviews 2024; : 54-61

Kasper VL., Assis DN.

Pathophysiology of Cystic Fibrosis Liver Disease.

Pediatric Pulmonology 2024; : S98-S106

Palle SK., Leung DH.

Advanced cystic fibrosis liver disease: Endovascular, endoscopic, radiologic, and surgical considerations.

Pediatric Pulmonology 2024; S1: S115-S122

Sankararaman S., Freeman AJ.

Early detection of hepatobiliary involvement in cystic fibrosis: Biomarkers, radiologic methods, and genetic influences.

Pediatric Pulmonology 2024; S1: S107-S114

Microbiology

Adariani AR., Sokhanvari S., Nikbin VS., Hosseinzadeh M., Khoram Z., Solgi H.

First case of New Delhi metallo beta-lactamase-1 (NDM-1)-producing Burkholderia cepacia complex in Iran.

Heliyon 2024; 10(10): e30895

Almeida MM., Bastos LR., Firmida MC., Albano RM., Marques EA., Leão RS.

Genomic Comparative of Pseudomonas aeruginosa Small Colony Variant, Mucoid and Non-mucoid Phenotypes Obtained from a Patient with Cystic Fibrosis During Respiratory Exacerbations.

Current Microbiology 2024; 81(9): e274

Ciszek-Lenda M., Nowak B., Majka G., Suski M., Walczewska M., Fedor A., Goli?ski E., Górska S., Gamian A., Olszanecki R., Strus M., Marcinkiewicz J.

Saccharomyces cerevisiae beta-glucan improves the response of trained macrophages to severe P. aeruginosa infections.

Inflammation Research 2024; 73: 1283-1297

Crabbé A

Intracellular Pseudomonas aeruginosa: An Overlooked Reservoir in the Lungs of People with Cystic Fibrosis? (Comment on Am J Respir Crit Care Med.

2024; 209:1421-1423)

American Journal of Respiratory and Critical Care Medicine 2024; 209: 1421-1423

Crisan CV., Pettis ML., Goldberg JB.

Antibacterial potential of Stenotrophomonas maltophilia complex cystic fibrosis isolates.

mSphere 2024; 9(7): e0033524

Duncan RP., Moustafa DA., Lewin GR., Diggle FL., Bomberger JM., Whiteley M., Goldberg JB.

Improvement of a mouse infection model to capture Pseudomonas aeruginosa chronic physiology in cystic fibrosis.

Proceedings of the National Academy of Sciences 2024; 121(33): e2406234121

George M., Narayanan S., Tejada-Arranz A., Plack A., Basler M.

Initiation of H1-T6SS dueling between Pseudomonas aeruginosa.

mBio 2024; 15(8): e0035524

Green N., Chan C., Ooi CY.

The gastrointestinal microbiome, small bowel bacterial overgrowth, and microbiome modulators in cystic fibrosis.

Pediatric Pulmonology 2024; S1: S70-S80

Hofstaedter CE., O'Keefe IP., Met CM., Wu L., Vanderwoude J., Shin S., Diggle SP., Riquelme SA., Rasko DA., Doi Y., Harro JM., Kopp BT., Ernst RK.

Pseudomonas aeruginosa Lipid A Structural Variants Induce Altered Immune Responses.

American Journal of Respiratory Cell and Molecular Biology 2024; 71: 207-218

Kruk ME., Mehta S., Murray K., Higgins L., Do K., Johnson JE., Wagner R., Wendt CH., O'Connor JB., Harris JK., Laguna TA., Jagtap PD., Griffin TJ.

An integrated metaproteomics workflow for studying host-microbe dynamics in bronchoalveolar lavage samples applied to cystic fibrosis disease. *mSystems* 2024; 9(7): :e0092923

Lagune M., Kremer L., Herrmann JL..

Mycobacterium abscessus, a complex of three fast-growing subspecies sharing virulence traits with slow-growing mycobacteria.

Clinical Microbiology and Infection 2024; 30: 726-731

Malet K., Faure E., Adam D., Donner J., Liu L., Pilon SJ., Fraser R., Jorth P., Newman DK., Brochiero E', Rousseau S., Nguyen D.. Intracellular Pseudomonas aeruginosa within the Airway Epithelium of Cystic Fibrosis Lung Tissues. (Comment on Am J Respir Crit Care Med. 2024; 209:1421-1423)

American Journal of Respiratory and Critical Care Medicine 2024; 209: 1453-1462

Mangiattera G., Schiavoni V., Cedraro N., Citterio B., Vignaroli C., Gesuita R., Fabrizzi B., Biavasco F., Cirilli N.

Clinical relevance of Pseudomonas aeruginosa viable but non-culturable forms in cystic fibrosis.

European Journal of Clinical Microbiology & Infectious Diseases 2024; 43: 1865-1867.

Maruri-Aransolo A., López-Causapé C., Hernández-García M., García-Castillo M., Caballero-Pérez JD., Oliver A., Cantón R.. In vitro activity of cefiderocol in Pseudomonas aeruginosa isolates from people with cystic fibrosis recovered during three multicentre studies in Spain.

Journal of Antimicrobial Chemotherapy 2024; 79: 1432-1440

Millar BC., Bell J., Rendall JC., Moore JE.

Carriage of *Neisseria meningitidis* (meningococci) in the sputum of people with cystic fibrosis (CF): occupational health risks and mitigating interventions for physiotherapists.
Physiotherapy. 2024; 124: 101-105

Mossop M., Ish-Horowicz J., Hughes D., Dobra R., Cunanan AG., Rosenthal M., Carr SB., Ramadan N., Nolan LM., Davies JC.

Chronicity Counts: The Impact of *Pseudomonas aeruginosa*, *Staphylococcus aureus*, and Coinfection in Cystic Fibrosis.
American Journal of Respiratory and Critical Care Medicine 2024; 210: 240-242

Palucci I., Delogu G.

Alternative therapies against *Mycobacterium abscessus* infections.
Clinical Microbiology and Infection 2024; 30: :732-737

Sheykhsaran E., Abbasi A., Memar MY., Ghotoslou R., Baghi HB., Mazraeh FN., Laghousi D., Sadeghi J.

The role of *Staphylococcus aureus* in cystic fibrosis pathogenesis and clinico-microbiological interactions.
Diagnostic Microbiology and Infectious Disease 2024; 109(3): e116294.

Stoodley P., Toelke N., Schwermer C., de Beer D.

Bioenergetics of simultaneous oxygen and nitrate respiration and nitric oxide production in a *Pseudomonas aeruginosa* agar colony biofilm.
Biofilm. 2024; 17: e100181

Tony-Odigie A., Dalpke AH., Boutin S., Yi B.

Airway commensal bacteria in cystic fibrosis inhibit the growth of *P. aeruginosa* via a released metabolite.
Microbiological Research. 2024; 283: e127680

Weimann A., Dinan AM., Ruis C., Bernut A., Pont S., Brown K., Ryan J., Santos L., Ellison L., Ukor E., Pandurangan AP., Krokowski S., Blundell TL., Welch M., Blane B., Judge K., Bousfield R., Brown N., Bryant JM., Kukavica-Ibrulj I., Rampioni G., et al

Evolution and host-specific adaptation of *Pseudomonas aeruginosa*.
Science. 2024; 385(6704): :eadi0908

Yap ZL., Rahman ASMZ., Hogan AM., Levin DB., Cardona ST..

A CRISPR-Cas-associated transposon system for genome editing in *Burkholderia cepacia* complex species.
Applied and Environmental Microbiology 2024; 90(7): e0069924

Nutrition

Bass R., Alvarez JA.

Nutritional status in the era of highly effective CFTR modulators.
Pediatric Pulmonology 2024; S1: S6-S11

Jobanputra AM., Kesavarapu K., Naik S., Ramagopal M., Scharf MT., Jagpal S.

Overnutrition in persons with cystic fibrosis on modulator therapy and the relationship to obstructive sleep apnea.
Pediatric Pulmonology. 2024; S1: S27-S35

Lyons ER., Muther E., Sabharwal S.

Nutrition and behavioral health in cystic fibrosis: Eating and body image.
Pediatric Pulmonology 2024; S1: S36-S43

Shrivastava S., Shaw K., Lee M., Reitich P., Hunter S., Klosterman M., Sathe M.

Association of in-line digestive enzyme cartridge with enteral feeds on improvement in anthropometrics among pediatric patients with cystic fibrosis.
Nutrition in Clinical Practice. 2024; 39: :903-910

Smith C., Lowdon J., Noordhoek J., Wilschanski M.

Evolution of nutritional management in children with cystic fibrosis - a narrative review
Journal of Human Nutrition and Dietetics. 2024; 37: :804-814

Suppakitjanusant P., Wang Y., Sivapiromrat AK., Hu C., Binongo J., Hunt WR., Weinstein S., Jathal I., Alvarez JA., Chassaing B., Ziegler TR., Gewirtz AT., Tangpricha V.

Impact of high-dose cholecalciferol (vitamin D3) and inulin prebiotic on intestinal and airway microbiota in adults with cystic fibrosis: A 2 × 2 randomized, placebo-controlled, double-blind pilot study.
Journal of Clinical and Translational Endocrinology. 2024; 37: e100362

Physiotherapy

Morrison L., McCrea G., Palmer S.

Online activity - A beaming good initiative! Delivering alternative exercise opportunities for people with cystic fibrosis.
Physiotherapy Theory and Practice. 2024; 40: 1609-1615

Sinderholm Sposato N., Bjerså K., Gilljam M., Lannefors L., Fagevik Olsén M.

Effectiveness of manual therapy interventions in cystic fibrosis care: a pilot study.
Journal of Bodywork and Movement Therapies 2024; 39: 323-329

Vidal T., Reyhler G., Sorlat-Maire C., Perceval M., Nove-Josserand R', Durieu I., Reynaud Q

Adherence to chest physiotherapy in adults with cystic fibrosis
Revue des Maladies Respiratoires. 2024; 41: 455-462

Psychosocial

Blais A., Katz SL., Klaassen RJ., Lougheed J., Reisman JJ., Pohl D., Lawrence S., Lai L., Lee S., Gardin L., Wong D., Sell E., Longmuir P. Understanding the physical literacy development of 8- to 12-year-old children living with chronic medical conditions: A comprehensive, mixed methods inquiry.
Child: Care, Health and Development. 2024; 50(5): e13316.

Duck SA., Jansen E., Papantoni A., Sheltry A., Koinis-Mitchell D., D'Sa V., Deoni S., Moran TH., Findling RL., Mogayzel PJ Jr., Carnell S.

Parental perceptions of body weight and appetite in infants and toddlers with cystic fibrosis.
Appetite 2024; 198: e107357

Duff AJA., Lee TWR.

Reply to Piehler et al.: Depression Symptoms in Patients with Cystic Fibrosis Fluctuate at Baseline and Improve with Elexacaftor/Tezacaftor/Ivacaftor Therapy.

American Journal of Respiratory and Critical Care Medicine. 2024; 210: 367

Gamel B., Albon D., Bandla S., Davison DW., Flath J., Sabadosa KA., Seid M., Silva L., Ong T.; Cystic Fibrosis Learning Network Group.

Interventions to improve system-level coproduction in the Cystic Fibrosis Learning Network.
BMJ Open Quality. 2024; 13(30): e002860

Ladune R., Hayotte M., Vuillemin A', d'Arripe-Longueville F.

Development of a Web App to Enhance Physical Activity in People With Cystic Fibrosis: Co-Design and Acceptability Evaluation by Patients and Health Professionals.
JMIR Formative Research. 2024; 8: e54322

Liu CM., Han EJ., Fischer JL., Mace JC., Mattos JL., Markarian K., Alt JA., Bodner TE., Chowdhury NI., Eshaghian PH., Getz AE., Hwang PH., Khanwalkar A., Kimple AJ., Lee JT., Li DA., Norris M., Nayak JV., Owens C., Patel ZM., Poch K., et al

Patient perspectives on chronic rhinosinusitis in cystic fibrosis: Symptom prioritization in the era of highly effective modulator therapy.
International Forum of Allergy & Rhinology. 2024; 14: 1282-1293

Piehler L., Thalemann R., Lehmann C., Stahl M., Mall MA., Graeber SY.

Depression Symptoms in Patients with Cystic Fibrosis Fluctuate at Baseline and Improve with Elexacaftor/Tezacaftor/Ivacaftor Therapy.
American Journal of Respiratory and Critical Care Medicine 2024; 210: 365-367

Sermet-Gaudelus I., Benaboud S., Bui S., Bihouée T., Gautier S.; MODUL-CF study group.

Behavioural and sleep issues after initiation of elexacaftor-tezacaftor-ivacaftor in preschool-age children with cystic fibrosis.
Lancet. 2024; 404: 117-120

Shadi D., Jabraeili M., Hassankhani H., Alhani F., Bostanabad MA.

Development and validation of a supportive programme for family caregivers of children suffering from cystic fibrosis: protocol for a sequential exploratory mixed-methods study.

BMJ Open. 2024; 14(6): e081560

Silvestri L., Issanchou D., Schuft L., Ferez S.

How workplaces produce or reduce disability along the career paths of young people with cystic fibrosis
Health 2024; 28: 507-525

Pulmonology

Bech M., Skov M., Andersen ISB., von Buchwald C., Aanæs K.

The criteria for chronic rhinosinusitis in children with cystic fibrosis are rarely fulfilled after initiation of CFTR modulator treatment.
APMIS 2024; 132: 625-631

Beswick DM., Liu CM., Overdevest JB., Zemke A., Khatriwada A., Gudis DA., Miller JE., Kimple A., Tervo JP., DiMango E., Goralski JL., Keating C., Senior B., Stapleton AL., Eshaghian PH., Mace JC., Markarian K., Alt JA., Bodner TE., et al
Predictors of Sinonasal Improvement After Highly Effective Modulator Therapy in Adults with Cystic Fibrosis.
Laryngoscope. 2024; 134: 3965-3973

Harris ES., McIntire HJ., Mazur M., Schulz-Hildebrandt H., Leung HM., Tearney GJ., Krick S., Rowe SM., Barnes JW.

Reduced sialylation of airway mucin impairs mucus transport by altering the biophysical properties of mucin.
Scientific Reports 2024; 14(1): e16568.

Mazio C., Scognamiglio LS., Casale C., Panzetta V., Urciuolo F., Galletta LJV., Imparato G., Netti PA.

A functional 3D full-thickness model for comprehending the interaction between airway epithelium and connective tissue in cystic fibrosis.
Biomaterials 2024; 308: e122546

Naseem R., Howe N., Williams CJ., Pretorius S., Green K

What diagnostic tests are available for respiratory infections or pulmonary exacerbations in cystic fibrosis: A scoping literature review.
Respiratory Investigation 2024; 62: 817-831

Sousa AM., Pereira MO.

Challenges with drug efficacy prediction of in vitro models of biofilms infecting cystic fibrosis airway.
Expert Opinion on Drug Discovery. 2024; 19: 635-638.

Radiology

Bugenhagen SM., Grant JCE., Rosenbluth DB., Bhalla S.

Update on the Role of Chest Imaging in Cystic Fibrosis.
Radiographics 2024; 44(9): e240008.

David M., Benlala I., Bui S., Benkert T., Berger P., Laurent F., Macey J., Dournes G..
Longitudinal Evaluation of Bronchial Changes in Cystic Fibrosis Patients Undergoing Elexacaftor/Tezacaftor/Ivacaftor Therapy Using Lung MRI With Ultrashort Echo-Times.
Journal of Magnetic Resonance Imaging 2024; 60: 116-124.

Mustafina M., Silantyev A., Krasovskiy S., Chernyak A., Naumenko Z., Suvorov A., Gognieva D., Abdullaev M., Bektimirova A., Bykova A., Dergacheva V., Betelin V., Kopylov P.
Exhaled breath analysis in adult patients with cystic fibrosis by real-time proton mass spectrometry.
Clinica Chimica Acta 2024; 560: e119733

Navas-Moreno V., Sebastian-Valles F., Rodríguez-Laval V., Knott-Torcal C., Marazuela M., de la Blanca NS., Arranz Martín JA., Girón RM., Sampedro-Núñez MA.
Impact of CFTR modulator therapy on body composition as assessed by thoracic computed tomography: A follow-up study.
Nutrition 2024; 123: e112425.

Therapy

Arenhoevel J., Kuppe A., Addante A., Wei LF., Boback N., Butnarasu C., Zhong Y., Wong C., Graeber SY., Duerr J., Gradzielski M., Lauster D., Mall MA., Haag R.
Thiolated polyglycerol sulfate as potential mucolytic for muco-obstructive lung diseases.
Biomaterials Science 2024; 12(17): 4376-4385.

Chun SW., Somers ME., Burgener EB.
Highly effective cystic fibrosis transmembrane conductance (regulator) modulator therapy: shifting the curve for most while leaving some further behind.
Current Opinion in Pediatrics 2024; 36: 290-295

Hansen CME., Breukelman AJ., van den Bemt PMLA., Zwitserloot AM., van Dijk L., van Boven JFM.
Medication adherence to CFTR modulators in patients with cystic fibrosis: a systematic review.
European Respiratory Review 2024; 33(173): e240060

Hong E., Zampoli M., Beringer PM.
Pharmacokinetic Enhancement of Elexacaftor/Tezacaftor/Ivacaftor for Cystic Fibrosis: A Cost Reduction Strategy to Address Global Disparities in Access.
Clinical Pharmacology & Therapeutics 2024; 115: 1204-1207.

Jarosz-Griffiths HH., Gillgrass L., Caley LR., Spoletini G., Clifton IJ., Etherington C., Savic S., McDermott MF., Peckham D.
Anti-inflammatory effects of elexacaftor/tezacaftor/ivacaftor in adults with cystic fibrosis heterozygous for F508del.
PLoS One 2024; 19: e0304555

Mazio C., Scognamiglio LS., Casale C., Panzetta V., Urciuolo F., Galletta LJV., Imparato G., Netti PA., Cohen A., Mass A., Reiter J., Zangen DH., Cohen-Cymberknob M.
Long-term therapy with CFTR modulators consistently improves glucose metabolism in adolescents and adults with cystic fibrosis
Respiratory Medicine. 2024; 228: e107664.

McKinzie CJ., Kam CW., Jones MC., Gifford LB., Loughlin CE., Noah TL., Shenoy VK., Dallon EP.
Elevated creatine phosphokinase and rhabdomyolysis associated with elexacaftor/tezacaftor/ivacaftor use in cystic fibrosis.
Pediatric Pulmonology 2024; 59: 1795-1797

Pudukodu H., Powell MZ., Ceppe A., Donaldson SH., Goralski JL., Sowa NA.
Analysis of Depression and Anxiety Scores Following Initiation of Elexacaftor/Tezacaftor/Ivacaftor in Adults With Cystic Fibrosis.
Clinical Respiratory Journal . 2024; 18(9): e70007

Sandler RD., Lai L., Dawson S., Cameron S., Lynam A., Sperrin M., Hoo ZH., Wildman MJ.
Development of data processing algorithm to calculate adherence for adults with cystic fibrosis using inhaled therapy - a multi-center observational study within the CFHealthHub learning health system.
Expert Review of Pharmacoeconomics & Outcomes Research 2024; 24: 759-771

Stahl M., Dohna M., Graeber SY., Sommerburg O., Renz DM., Pallenberg ST., Voskrebenev A., Schütz K., Hansen G., Doellinger F., Steinke E., Thee S., Röhmel J., Barth S., Rückes-Nilges C., Berges J., Hämmерling S., Wielputz MO., Naehrlich L., et al
Impact of elexacaftor/tezacaftor/ivacaftor therapy on lung clearance index and magnetic resonance imaging in children with cystic fibrosis and one or two F508del alleles.
European Respiratory Journal 2024; 64(3): e2400004

Stastna N., Hrabovska L., Homolka P., Homola L., Svoboda M., Brat K., Fila L.
The long-term effect of elexacaftor/tezacaftor/ivacaftor on cardiorespiratory fitness in adolescent patients with cystic fibrosis: a pilot observational study.
BMC Pulmonary Medicine. 2024; 24(1): e260.

Turuvekere Vittala Murthy N., Vlasova K., Renner J., Jozic A., Sahay G.
A new era of targeting cystic fibrosis with non-viral delivery of genomic medicines.
Advanced Drug Delivery Reviews 2024; 209: e115305

Woynarowski M., Sapiejka E., Jó?wiak M., Milczewska J., Zybert K., Krusi?ski A., Siwiec J., Wierzbicka-Ruciska A.
Macroenzymes as a reason for aminotransferases flare in cystic fibrosis patients on CFTR modulators therapy - Report of three cases. *Heliyon* 2024; 10(10): e31189.