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HEALTH SCIENCES LIBRARIES GROUP

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HEAR *(Health Evidence Awareness Report)*

Inside this issue:

HEAR signposts content which is available online. We endeavour to prioritise Open Access content in HEAR to make it easily accessible to all our readers. However, as with all online content, links may change or break over time.

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Welcome! Focus on Cystic Fibrosis

Cystic fibrosis affects the internal organs, especially the lungs and digestive system. It causes them to become clogged with thick, sticky mucus. It is caused by a faulty gene that controls the movement of salt and water in and out of cells in the body. When cystic fibrosis occurs, too much salt and not enough water pass into the cells and turn the body's secretions, which normally act as a lubricant, into a thick mucus. This mucus clogs up many of the body's tubes, ducts and passageways so they cannot work properly. In the lungs, this leads to frequent and severe infections. https://www.hse.ie/eng/health/az/c/cystic-fibrosis/

Cystic fibrosis or CF, is an inherited disease. "Inherited" means the disease is passed from parents to children through genes. People who have CF inherit two faulty genes for the disease—one from each parent. The parents likely don't have the disease themselves. CF mainly affects the lungs, pancreas, liver, intestines, sinuses, and sex organs.

US National Heart, Blood and Lung Institute

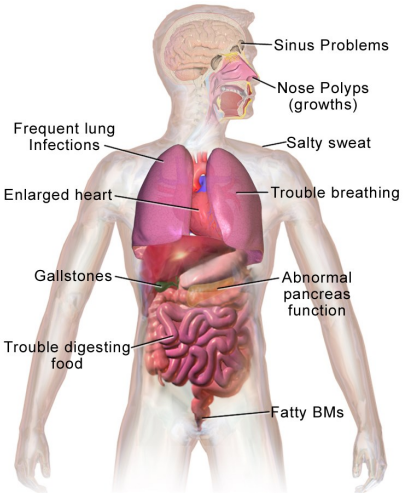


Image copyright: Blausen.com staff (2014). "Medical gallery of Blausen Medical 2014". WikiJournal of Medicine 1 (2). DOI:10.15347/wjm/2014.010. ISSN 2002-4436. CC BY 3.0

There are over 1100 people registered as having cystic fibrosis in Ireland.

Health Service Executive

Ireland has the highest rate of cystic fibrosis worldwide, according to a study in 2014 by Donatello Salvatore et al, which examined international literature from cystic fibrosis registries.

Salvatore, D. (2011). An overview of international literature from cystic fibrosis registries, Part 3. *Journal of Cystic Fibrosis,* 10(2), 71-85.

STORIES FROM THE PRESS

“New report finds better outcomes for people with cystic fibrosis. 04 April 2018”. https://www.irishtimes.com/news/ireland/irish-news/new-report-finds-better-outcomes-for-people-with-cystic-fibrosis-1.3450122

"DWTS judge Julian shares his defiant cystic fibrosis story". 25 March 2018. https://www.rte.ie/entertainment/2018/0324/949797-dwts-judge-julian-shares-his-cystic-fibrosis-story/

"Why Orkambi may not be a 'miracle' drug, but is worth it". Philip Watt, CEO Cystic Fibrosis Ireland. 10 June 2018. https://www.rte.ie/eile/2018/0531/967374-why-orkambi-may-not-be-a-miracle-drug-but-is-worth-it/

“Plans for a new 20-bed cystic fibrosis unit at hospital now at 'advanced stage'“. 20 November 2018. https://www.independent.ie/irish-news/health/plans-for-a-new-20bed-cystic-fibrosis-unit-at-hospital-now-at-advanced-stage-37501005.html

HELPFUL WEBSITES

Cystic Fibrosis Ireland https://www.cfireland.ie/

Health Service Executive https://www.hse.ie/eng/health/az/c/cystic-fibrosis/

NHS UK https://www.nhs.uk/conditions/cystic-fibrosis/

Cystic Fibrosis Trust (UK) https://cms.cysticfibrosis.org.uk/the-work-we-do/about-us

Cystic Fibrosis Foundation (US) https://www.cff.org/About-Us/

Cystic Fibrosis Registry of Ireland https://www.cfri.ie/

RESEARCH WEBSITES

European Cystic Fibrosis Society (EU) https://www.ecfs.eu/society-details/about

National Institute of Diabetes and Digestive and Kidney Diseases https://www.niddk.nih.gov/research-funding/research-programs/cystic-fibrosis-research-translation-centers

National Heart, Lung and Blood Institute (US) https://www.nhlbi.nih.gov/

National Children's Research Centre https://www.nationalchildrensresearchcentre.ie/research-innovation/research-areas/immunity-infection/cystic-fibrosis/

HEALTH BYTES

de Vries, J.V, et al. (2018). Vitamin A and beta (β)-carotene supplementation for cystic fibrosis. Cochrane Database of Systematic Reviews(8). doi:10.1002/14651858.CD006751.pub5

Conviser, J. H., et al. (2018). Are children with chronic illnesses requiring dietary therapy at risk for disordered eating or eating disorders? A systematic review. The International journal of eating disorders. doi:10.1002/eat.22831

Nutritional Management of Cystic Fibrosis. *Cystic Fibrosis Trust consensus document 2016* https://www.cysticfibrosis.org.uk/the-work-we-do/clinical-care/consensus-documents#

Hollander, F.M., et al (2017) The optimal approach to nutrition and cystic fibrosis: latest evidence and recommendations. *Current Opinion Pulmonary Medicine*, 23(6):556-561 Abstract available from https://www.ncbi.nlm.nih.gov/pubmed/28991007

IRISH RESEARCH

Kapoor, P., & Murphy, P. (2018). Combination antibiotics against Pseudomonas aeruginosa, representing common and rare cystic fibrosis strains from different Irish clinics. *Heliyon, 4*(3). doi: https://doi.org/10.1016/j.heliyon.2018.e00562

Martin-Loeches, I., et al. (2017). Respiratory research networks in Europe and beyond: aims, achievements and aspirations for the 21st century. *Breathe, 13*(3), 209-215. doi:10.1183/20734735.009217

Reece, E., et al. (2018). Aspergillus fumigatus Inhibits Pseudomonas aeruginosa in Co-culture: Implications of a Mutually Antagonistic Relationship on Virulence and Inflammation in the CF Airway. *Frontiers in Microbiology, 9*. doi:10.3389/fmicb.2018.01205

Cronly, J., et al. (2018). Online versus paper-based screening for depression and anxiety in adults with cystic fibrosis in Ireland: a cross-sectional exploratory study. *BMJ Open*, 8(1). doi:10.1136/bmjopen-2017-019305

McKone, E. F., et al. (2014). Long-term safety and efficacy of ivacaftor in patients with cystic fibrosis who have the gly551asp-cftr mutation: A phase 3, open-label extension study (persist). *Lancet Respiratory Medicine*, 2(11), 902-910. doi:10.1016/s2213-2600(14)70218-8

Twitter accounts

@cf_ireland

@cftrust

@CF_Foundation

@KnowCF

In the US and UK, average life expectancy is 35 to 40 years old.

Cystic Fibrosis Worldwde

An individual must inherit two defective cystic fibrosis genes, one from each parent, to have the disease. Each time two carriers of the disease conceive, there is a 25 percent chance of passing cystic fibrosis to their children ; a 50 percent chance that the child will be a carrier of the cystic fibrosis gene; and a 25 percent chance that the child will be a non-carrier.

World Health Organization

SYSTEMATIC REVIEWS

Ahmed, M. I., & Mukherjee, S. (2018). Treatment for chronic methicillin-sensitive staphylococcus aureus pulmonary infection in people with cystic fibrosis. *Cochrane Database of Systematic Reviews*(7). doi:10.1002/14651858.CD011581.pub3

Dentice, R., & Elkins, M. (2018). Timing of dornase alfa inhalation for cystic fibrosis. *Cochrane Database of Systematic Reviews*(11). doi:10.1002/14651858.CD007923.pub5

Freitas, D. A., et al. (2018). Standard (head-down tilt) versus modified (without head-down tilt) postural drainage in infants and young children with cystic fibrosis. *Cochrane Database of Systematic Reviews*(3). doi:10.1002/14651858.CD010297.pub3

Green, J., et al. (2018). Interventions for treating distal intestinal obstruction syndrome (dios) in cystic fibrosis. *Cochrane Database of Systematic Reviews*(8). doi:10.1002/14651858.CD012798.pub2

Green, J., et al. (2018). Interventions for preventing distal intestinal obstruction syndrome (dios) in cystic fibrosis. *Cochrane Database of Systematic Reviews*(6). doi:10.1002/14651858.CD012619.pub2

Hilton, N., & Solis-Moya, A. (2018). Respiratory muscle training for cystic fibrosis. *Cochrane Database of Systematic Reviews*(5). doi:10.1002/14651858.CD006112.pub4

Jain, K., et al. (2018). Bronchoscopy-guided antimicrobial therapy for cystic fibrosis. *Cochrane Database of Systematic Reviews*(9). doi:10.1002/14651858.CD009530.pub4

Nevitt, S. J., et al (2018). Inhaled mannitol for cystic fibrosis. *Cochrane Database of Systematic Reviews*(2). doi:10.1002/14651858.CD008649.pub3

Oniyangi, O., & Cohall, D. H. (2018). Phytomedicines (medicines derived from plants) for sickle cell disease. *Cochrane Database of Systematic Reviews*(2). doi:10.1002/14651858.CD004448.pub6

Saldanha, I. J., et al. (2018). Immunosuppressive drug therapy for preventing rejection following lung transplantation in cystic fibrosis. *Cochrane Database of Systematic Reviews*(6). doi:10.1002/14651858.CD009421.pub4

Smith, S., et al. (2018). Inhaled antibiotics for pulmonary exacerbations in cystic fibrosis. *Cochrane Database of Systematic Reviews*(10). doi:10.1002/14651858.CD008319.pub3

Spencer, S., et al. (2018). Oral versus inhaled antibiotics for bronchiectasis. *Cochrane Database of Systematic Reviews*(3). doi:10.1002/14651858.CD012579.pub2

GUIDELINES

Castellani, C., et al. (2018). ECFS best practice guidelines: The 2018 revision. Journal of Cystic Fibrosis, 17(2), 153-178. doi:https://doi.org/10.1016/j.jcf.2018.02.006

Ren, C. L., et al. (2018). Cystic fibrosis foundation pulmonary guidelines. Use of cystic fibrosis transmembrane conductance regulator modulator therapy in patients with cystic fibrosis. Annals of the American Thoracic Society, 15(3), 271-280. doi:10.1513/AnnalsATS.201707-539OT

National Institute for Health and Care Excellence. (2017). Cystic fibrosis: Diagnosis and management NICE guideline [ng78]. In. London: National Institute for Health and Care Excellence. https://www.nice.org.uk/guidance/ng78

Royal Brompton Hospital Paediatric Cystic Fibrosis Team. (2017). Care of children with cystic fibrosis 2017. Retrieved from https://www.rbht.nhs.uk/our-services/paediatrics/paediatric-cystic-fibrosis-clinics/care-children-cystic-fibrosis-2017

VIDEOS

What is cystic fibrosis, exactly? Cystic Fibrosis Trust. https://youtu.be/4IGz5p4n8Fg

Useful tips to help with your CF treatment plan. University Hospital Limerick Cystic Fibrosis Centre. https://youtu.be/JhJcBTcKOxA

What Causes Cystic Fibrosis? - Marlyn Woo, MD | UCLAMDCHAT Webinars. https://youtu.be/C_PJdZqXyck

Preschool and Toddler Cystic Fibrosis Exercises. Cincinnati Children's Hospital. https://youtu.be/EUK9zfNhcvw

Living with CF // Exercise. CF Association Ireland. https://youtu.be/JeUR52nNwms

UPCOMING EVENTS

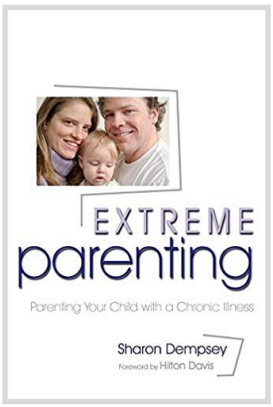
05 December 2018. *AWARE for All - Dublin: Clinical Research Education Day* - a **FREE** educational event that serves as a forum for open dialogue between patients, members of the public, and health and research professionals. AWARE for All will provide information about the clinical research process and activity here in Dublin, as well as food, prizes, and free health screenings. Full details on https://www.eventbrite.com/e/aware-for-all-dublin-2018-registration-44204148799?aff=ebdssbdestsearch

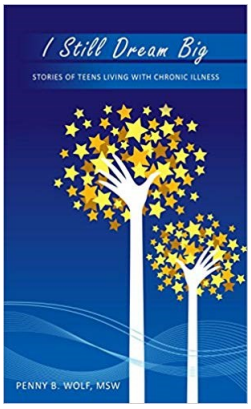
Books

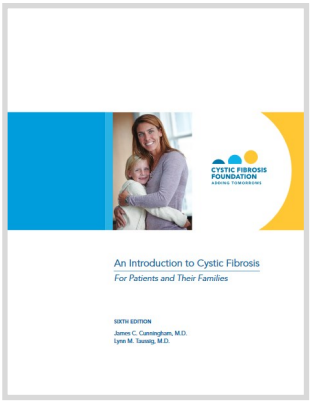
Wolf, P. B. (2009). I still dream big: Stories of teens living with chronic illness. Bloomington, IN: AuthorHouse

Dempsey, S. (2008). Extreme parenting: Parenting your child with a chronic illness. Philadelphia, PA: Jessica Kingsley Publishers

Cunningham, J.C. & Taussig, L.M. (2013). An Introduction to Cystic Fibrosis For Patients and Their Families. 6th edition. Bethseda, MD: Cystic Fibrosis Foundation







HEAR is about sharing information that is useful for both healthcare staff and patients.

Your thoughts, comments and feedback are always welcome. Please contact us at HEAR@hslg.ie

ABOUT US

The purpose of HEAR is to provide specially selected information to patients, the public and to health professionals about key health topics. Each issue is the result of the collaborative effort of librarians from health organisations across Ireland.

We are: Caroline Rowan, St. Michael's Hospital; Anne Madden, St. Vincent's University Hospital; Margaret Morgan, Midland Regional Hospital Mullingar; Joanne Callinan, Milford Care Centre; Bernadette Colley, Temple Street University Hospital; Catherine Dillon, HSE; Gethin White, Dr. Steeven's Hospital; Isabelle Delaunois, University Hospital Limerick; Marie Carrigan, St. Luke's Radiation Oncology Network; Patsy Walsh, University Hospital Limerick.