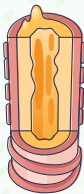




Early Intervention in Cystic Fibrosis: Limiting Multisystem Disease Progression

The publication of this infographic was funded by Vertex Pharmaceuticals. Medical writing assistance was provided by Bethany Cooper, Ownership Health Limited, UK. EMJ Respir. 2026;14[1]:90-91. <https://doi.org/10.33590/emjrespir/X7J3YIF3>

The burden of CF



CF is a progressive autosomal recessive disease caused by variants in the *CFTR* gene,¹⁻³ characterised by the accumulation of sticky mucus in multiple organs, of which the lungs, GI tract and pancreatic ducts are the most commonly affected.⁴



CF is an inherited disorder^{1,5,6} associated with high symptom burden and reduced quality of life.^{7,8}



Structural lung disease is present within the first year of life, contributing to associated morbidity and mortality rates.⁹⁻¹² 85% of infants with CF have exocrine pancreatic insufficiency.¹³

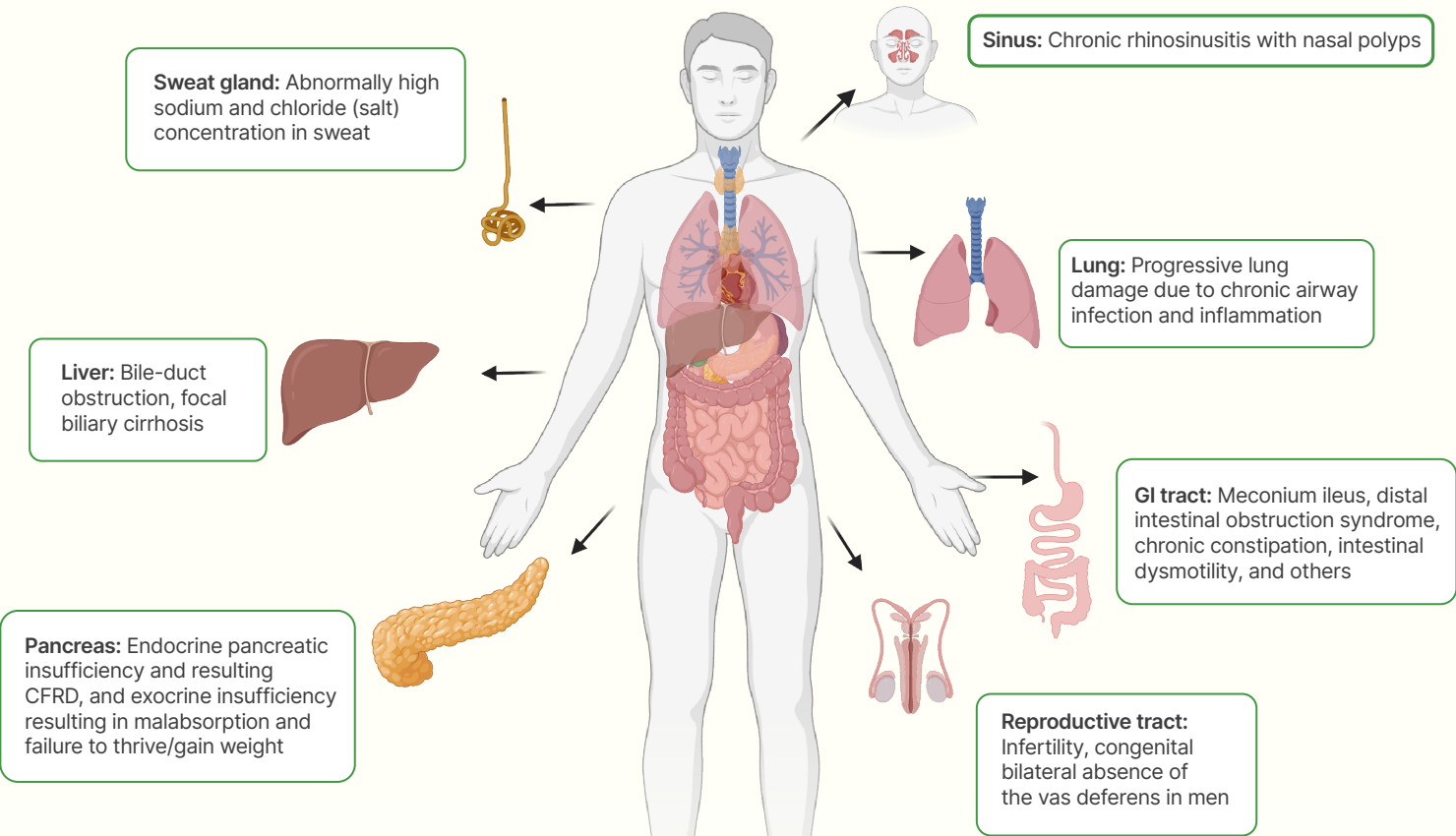


In countries with CF screening programmes such as the USA, CF is typically diagnosed shortly after birth, enabling early specialist care and timely intervention.¹⁴

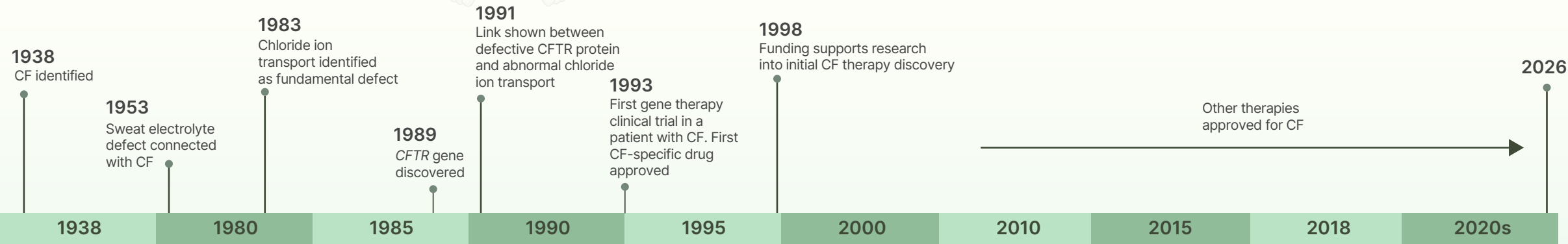


While CF has traditionally been thought of as a lung disease, it affects various other organ systems, including the pancreas, sweat glands, liver, and GI tract, extending into adulthood.^{1-3,15}

CF is a multisystem disease that impacts multiple organs^{1-3,16}



CF research milestones¹⁷⁻²⁰



Patient outlook then

In the 1980s, most people with CF did not reach adulthood.^{2,21}



1980s: A life expectancy of <20 years

Significant improvements in care have changed the outlook of CF.²⁵

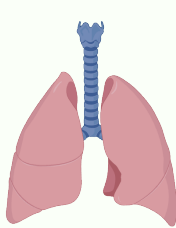
Patient outlook now

The median age of survival now is above 50 years in Canada, Australia, New Zealand, France, and the USA^{2,21-23} with latest estimates reaching 65 years and beyond.²⁴



2026: Living into their 50s and beyond

Early intervention can change the trajectory of CF⁶



May reduce progression of irreversible damage



May support healthy growth and development



May reduce cumulative disease burden

- Early treatment of CF is crucial to improving long-term health outcomes and potentially improving the disease trajectory.²⁶⁻³⁰
- Early treatment initiation may delay, reduce, or limit progression of organ damage, reduce cumulative disease burden, and potentially support healthy growth.²⁶⁻³⁰
- Despite advances in CF therapy and care, unmet needs still remain for some patients living with CF.

Why intervene early?

KEY MESSAGES

CF is a systemic disease that requires a whole-body, multidisciplinary approach.¹⁻³
CF begins before symptoms become apparent. Disease-related changes in the lungs, pancreas, and GI tract can start early in life, making timely diagnosis and intervention critical.^{6,9,13}
Significant improvements in care have changed the outlook of CF; however unmet needs still remain.

Abbreviations

CF: cystic fibrosis; CFRD: cystic fibrosis-related diabetes; CFTR: cystic fibrosis transmembrane conductance regulator; GI: gastrointestinal.

References

- Davies JC et al. BMJ. 2007;335(7632):1255-9.
- Grasemann H, Ratjen F. N Engl J Med. 2023;389:1693-707.
- Carbone A et al. Curr Issues Mol Biol. 2024;46(9):10396-410.
- Morrison CB et al. Pediatr Pulmonol. 2019;54(Suppl 3):S84-96.
- Allen L et al. Nat Commun. 2023;14:693.
- Coverston AM, Ferkol TW. Front Pediatr. 2020;8:608821.
- Smirnova N et al. J Cyst Fibros. 2024;23(5):831-5.
- Sawicki GS et al. J Pain Symptom Manage. 2008;35(4):372-80.
- Hoo AF et al. Thorax. 2012;67(10):874-81.
- VanDevanter DR et al. J Cyst Fibros. 2016;15(2):147-57.
- Sly PD et al. Am J Resp Crit Care. 2009;180(2):146-52.
- Sun X et al. Sci Transl Med. 2019;11(485):eaau7531.
- Slae M, Wilschanski M. Frontline Gastroenterol. 2020;12(7):622-8.
- McGarry ME et al. Int J Neonatal Screen. 2025;11(2):24.
- O'Sullivan BP, Freedman SD. Lancet. 2009;373(9678):1891-904.
- Singh VK, Schwarzenberg SJ. J Cyst Fibros. 2017;16(Suppl 2):S70-8.
- Cystic Fibrosis Foundation. Available at: <https://www.cff.org/about-us/our-history>. Last accessed: 30 September 2026.
- Davis PB. Am J Respir Crit Care Med. 2006;173(5):475-82.
- Dalesmans W et al. Nature. 1991;354:526-8.
- National Human Genome Research Institute. 2013. Available at: <https://www.genome.gov/Genetic-Disorders/Cystic-Fibrosis>. Last accessed: 30 September 2026.
- Dodge JA et al. Eur Res J. 2007;29(3):522-6.
- Coriati A et al. Thorax. 2023;78(3):242-8.
- Stephenson AL et al. Ann Intern Med. 2017;166(8):537-46.
- Cystic Fibrosis Foundation. 2024. Available at: <https://www.cff.org/sites/default/files/2026-06/2025-Patient-Registry-Highlights-Report.pdf>. Last accessed: 30 September 2026.
- Diener BL et al. Healthcare (Basel). 2025;13(23):3022.
- Goralski JL et al. Am J Respir Crit Care Med. 2023;208(1):59-67.
- Kurgansky KE et al. Chest. 2026;S0012-3692(26)00614-8.
- Salvatore D, Pepe A. J Clin Med. 2025;14(11):3978.
- Rosenfeld M et al. Lancet. 2018;6(7):545-53.
- Shteinberg M et al. Eur Respir Rev. 2020;29(155):190112.