The New Era of Cystic Fibrosis

Information for Health Professionals

What is CF?

CF is a multi-organ life shortening disease typified by chronic endobronchial infection, progressive obstructive lung disease and malnutrition, secondary to pancreatic insufficiency. CF is the most common autosomal recessive condition in Australia, with 1 in 25 people carrying the faulty gene. Approximately 1 in every 2,500 babies born in WA has CF and there are currently over 3,700 people living with CF in Australia.

There have been significant advancements in CF treatment and outcomes in recent years, with the predicted survival age of a child born after 2017 being 58 years of age.

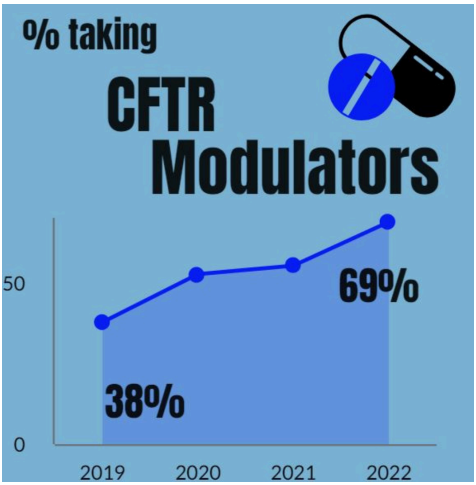
This is an increase in 12 years from previous data. One of the main reasons for this increase is access to modulator medications.

Modulator Medications

Modulator medications aim to improve or restore the function of the cystic fibrosis transmembrane conductance regulator (CFTR) protein.

The CFTR protein helps to maintain the balance of salt and water on many surfaces in the body, such as the surface of the lung. When the protein is not working correctly, the resulting thick, sticky mucus causes many of the symptoms associated with CF, including recurrent lung infections and impaired digestion.

Modulators are not a cure, but they help the body's cells function more normally, resulting in improved lung function, BMI and life expectancy.



Australian CF Data Registry 2022

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Current modulator therapies may not work for everyone. There are many people who do not respond well to these medications, and more who are ineligible or intolerant. CFTR modulators are currently recommended for approximately 90% of people living with CF aged 2 years and above.

Most people on modulator medications will still need to complete daily maintenance therapy to optimise health outcomes, but for many people these new treatments have been life changing and mark a significant change in the CF burden of disease.

Carrier Screening Referral

Medical practitioners are now able to request reproductive carrier testing for CF, spinal muscular atrophy, and fragile X for their patients and reproductive partners under new Medicare Benefits Schedule item numbers.

A typical CF screening test will identify the most common CF genes in Australia, however, there are many rare CF genes that would not be identified. If the patient has a family history of CF, more specific testing relevant to the family gene should be included in the referral.

Find out more about reproductive carrier screening for CF, SFA & FX by scanning the QR code.

Check out our CF information hub for health professionals by scanning the QR code.