



A WORD FROM BEST DOCTORS

Cystic fibrosis (CF) is the most common fatal genetic disease affecting Canadian children and young adults. It's estimated one in every 3,600 children born in Canada has CF, a disease that primarily affects the digestive system and lungs.

Although there is presently no cure for cystic fibrosis, improvements in treatments and care have had a dramatic impact on increasing the life expectancy for people born with CF. In Canada, the median age of survival for CF patients has surpassed 50, up from just 32 in 1990. About 4,000 Canadian CF patients are cared for in one of the country's 42 cystic fibrosis clinics—roughly 60 per cent of which are adults, according to *The Globe and Mail*.

Improvements in data and research have helped doctors arrive at a better understanding of CF, leading not only to longer life expectancies for patients, but an improved quality of life for those living with cystic fibrosis.

Treatments focus on alleviating symptoms of the disease, such as enzymes that help with digestion. Antibiotics and vaccines protect CF patients from infection, and in recent years, most provinces have introduced newborn screening for cystic fibrosis, which allows cases to be identified and treated early.

May has been designated Cystic Fibrosis Awareness Month, drawing attention to a disease that, despite the many advances that have been made, continues to claim far too many lives far too early.

UNDERSTANDING CYSTIC FIBROSIS

CF has various effects on the body, beyond its impact on the lungs and digestive system. The severity of symptoms will vary from person to person. However, it's the ongoing infection of the lungs and loss of lung function that eventually leads to death in most people living with CF.

Approximately one in 25 Canadians carries an abnormal version of the gene responsible for cystic fibrosis. Carriers

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BEST DOCTORS IS HERE FOR YOU AND YOUR FAMILY

do not have CF, nor do they exhibit any symptoms of CF. When two carriers have a child, there is a 25 per cent chance the child will be born with cystic fibrosis, a 50 per cent chance the child will be a CF carrier, leaving only a 25 per cent chance the child will neither be born with CF nor be a carrier.

RECOGNIZING THE SIGNS AND SYMPTOMS OF CYSTIC FIBROSIS

Cystic fibrosis produces a variety of symptoms, including:

- Persistent cough with thick mucus
- Wheezing and shortness of breath
- Frequent chest infections (may include pneumonia)
- Bowel disturbances
- Weight loss/failure to gain weight
- Salty tasting sweat
- Infertility (men) and decreased fertility (women)

In adults, cystic fibrosis can cause other health complications, such as cystic fibrosis-related diabetes, liver disease, osteoporosis and nasal polyps (small, sac-like growths of inflamed nasal mucosa – masses in the nose – caused by chronic inflammation in the nasal lining).

DIAGNOSING CYSTIC FIBROSIS

A “sweat test” may be administered to a person suspected of having CF. This is a test that measures the amount of salt content present in the sweat of an individual. A positive test means the sweat collected contains more salt than usual and supports a diagnosis of CF. Other methods used for



diagnosing cystic fibrosis include genetic testing, prenatal screening and newborn screening.

LIVING WITH CYSTIC FIBROSIS

With proper medical care and treatment, people who have cystic fibrosis can, for the most part, lead normal lives. However, CF presents challenges to a person’s lifestyle and health. Time away from school or work is often inevitable due to hospitalizations, or receiving therapy or intravenous antibiotics. Aside from hospital admissions (which vary from person to person), people living with CF require hospital-based check-ups that can take a full day.

People living with cystic fibrosis may also tire more easily for various reasons, must follow daily treatment regimens that vary in length and complexity and must stick to a high-energy diet that can be difficult to maintain.

Still, with proper medical care and by following their treatment regimen, people living with CF can lead relatively normal and productive lives.



Today, there are more adults living with CF than children living with the disease.



Almost 60% of CF patients are diagnosed within their first year of life.



On average, a person with CF spends the equivalent of four months of full-time work doing life-sustaining treatments every year.



Cumulatively, CF patients in Canada spent almost 25,000 days in hospital and attended over 15,500 clinic visits in 2014.



For Canadians born today with CF, the estimated median survival age is 51.8 years.

Source: Cystic Fibrosis Canada

TREATING CYSTIC FIBROSIS

Although there's no cure for cystic fibrosis, various treatment options are available for CF patients. Care primarily consists of performing regular airway clearance, which helps prevent a build-up of thick mucus in the lungs, allowing the lungs to work at their highest capacity. Regular airway clearance may also help minimize chest infections.

There are several different airway clearance techniques—a patient's physiotherapist will work with the patient and caregivers to determine which technique (or combination of techniques) will work best for the individual.

Exercise also forms part of the care plan for people living with cystic fibrosis. Regular physical activity can slow the rate of decline of pulmonary function, and a greater exercise capacity is associated with a lower rate of mortality. A balanced and effective exercise program should consist of cardiovascular, strengthening and stretching exercises.

For some CF patients with an advanced form of the disease, transplantation may be an option. Double-lung, heart-lung and liver transplants are potential treatments for CF patients.

When it comes to day-to-day routines, there are several steps people living with CF can take to help maximize their health and wellness. These include:

- Attending their local CF clinic three to four times a year.
- Taking all prescribed medication.
- Following a diet that helps ensure maximum health, which, for CF patients, means a diet that is high in calories and fat.

- Completing physiotherapy exercises and activities on a regular basis.

People living with CF should also be sure to discuss with their clinic-recommended immunizations to prevent infection (e.g. an annual flu shot), wash their hands frequently to avoid the spread of germs and prevent infection and avoid people who have a respiratory illness. The early recognition and treatment of an impending infection is very important to help maintain overall long-term health.

HOW BEST DOCTORS CAN HELP

Best Doctors can help in a variety of ways when it comes to assisting people living with cystic fibrosis and their families and/or caregivers.

If you or a loved one has received a diagnosis of CF and you want a second opinion, Best Doctors is here to help. Best Doctors can also provide more information about cystic fibrosis following a diagnosis and identify support groups near you.

Given the importance of proper medical care and an appropriate treatment regime, it is important for CF patients to visit a cystic fibrosis clinic regularly, as these clinics provide specialized multidisciplinary care and give patients access to CF adult or pediatric specialist physicians. There are CF clinics in most major cities in Canada, and Best Doctors can help you locate the closest clinic, while also directing you to a clinic that specializes in adults or children.

If you're dealing with a diagnosis of CF or any other medical condition, contact Best Doctors to ensure you have the right diagnosis, the right treatment and the peace of mind you deserve.



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When you're facing an uncertain medical situation, call Best Doctors. We can solve any medical issues – no matter the complexity – to ensure you get the right information, the right diagnosis and the right treatment.

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