The Facts about Cystic Fibrosis

- Cystic fibrosis is the most common fatal genetic disease affecting Canadian children and young adults. There is no cure.
- It is a multi-system disease that affects mainly the lungs and the digestive system. In the lungs, where the effects are most devastating, a build-up of thick mucus causes severe respiratory problems. Mucus and protein also build up in the digestive tract, making it difficult to digest and absorb nutrients from food.
- Cystic fibrosis can cause many symptoms, including: difficulty breathing, life-altering respiratory tract problems, severe, chronic lung infections, impaired growth or weight gain and extreme difficulty absorbing nutrients.
- There are approximately 4,000 Canadians living with cystic fibrosis who receive specialized care at one of the 42 CF clinics across the country.
- Each week in Canada, two children are diagnosed and one person dies from the disease.
- Of the Canadians with cystic fibrosis who died in 2012, half were under 32 years of age.
- In 2012, 57 percent of Canadians newly diagnosed with cystic fibrosis were under the age of six months.
- The median age of survival for Canadians with cystic fibrosis was 49.7 years of age in 2012.

Genetics
- Cystic fibrosis occurs when a child inherits two defective copies of the gene responsible for cystic fibrosis (one from each parent).
- One in 25 Canadians is a cystic fibrosis carrier, with one defective version of the gene responsible for this life-altering disease. Many people are unaware that they are carriers.
- Over 1,900 different mutations in the CFTR gene have been identified. However, over 87.5 percent of Canadian CF patients carry at least one copy of the most common CF-causing mutation, deltaF508.

Adult Population
- Nearly 60 percent of all Canadians with cystic fibrosis are adults.
- Thirty percent of female adults and 19 percent of male adults with cystic fibrosis are classified as underweight.
Treatment and Care

- People with cystic fibrosis consume a large number of enzymes, about 20 pills a day, to help absorb nutrients from food and follow a demanding daily routine of physical and inhalation therapy to keep their lungs free from infection.
- Almost 87 percent of Canadian CF patients must take pancreatic enzymes to digest food and absorb nutrients.
- On average, a person with cystic fibrosis spends the equivalent of four months of full-time work doing life-sustaining treatments every year.
- Cumulatively, Canadians with cystic fibrosis spent over 20,000 days in hospital and attended CF clinics more than 15,000 times in 2012.

Leading Cause of Mortality

- Cystic fibrosis-related deaths are mainly due to lung disease caused by a cycle of chronic lung infections and inflammation.
- Nearly half of all patients with cystic fibrosis are infected with harmful bacteria such as Staphylococcus aureus and/or Pseudomonas aeruginosa in their lungs.

Lung Transplantation

- Lung transplantation is an important treatment option for end-stage lung disease in people with cystic fibrosis.
- Thirty-seven CF patients received a transplant in 2012.

Other Medical Complications

- Sixteen percent of all Canadians with cystic fibrosis have CF-related diabetes (CFRD), and nearly 35 percent of CF patients 35 years of age and older have CFRD.
- Other medical complications in addition to CF-related diabetes include:
  - Allergic bronchopulmonary aspergillosis (ABPA)
  - Arthritis
  - Congenital bilateral absence of the vas deferens (CBAVD)
  - Distal Intestinal Obstruction Syndrome (DIOS)
  - Liver disease
  - Nasal polyps
  - Osteoporosis
  - Pancreatitis

*Facts compiled using data from The Canadian Cystic Fibrosis Registry 2012 Annual Report.*

For more information and to donate, visit [www.cysticfibrosis.ca](http://www.cysticfibrosis.ca).

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