KNOWLEDGE IS EMPOWERING:

MAGGIE
Living with CF

CF & PROGRESSION:
UNDERSTANDING THE UNDERLYING IMPACT
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Cystic fibrosis (CF) feels a little different for everyone, but everyone experiences progression of CF over time, whether they feel it or not. Progression can occur when the thick, sticky mucus building up throughout the body causes inflammation and scarring in many different organs, leading to permanent damage. This damage is often present before it can even be detected by tests.

This brochure helps you understand how progression impacts many different parts of the body over time.
Lungs: thick, sticky mucus causes infection and lung damage

Mucus builds up in the lungs and can’t be cleared from the airways easily

Germs build up in the mucus causing infection, inflammation, and scarring

Symptoms may develop such as wheezing, shortness of breath, and persistent, phlegmy cough

Buildup of mucus and germs causes pulmonary exacerbations—periods of time when infection and symptoms get more severe and may require intravenous (IV) antibiotics or a hospital visit
A complete or incomplete blockage can lead to distal intestinal obstruction syndrome (DIOS).

In the pancreas
- Thick juices block ducts within the pancreas
- Blocked ducts make it harder for digestive enzymes to reach the small intestine, where they break down food for digestion
- As a result, the body has trouble absorbing nutrients, which leads to poor growth and slow weight gain

In the liver
- Thicker bile blocks bile ducts and can cause irritation or inflammation in the liver

In the intestines
- A complete or incomplete blockage can lead to distal intestinal obstruction syndrome (DIOS)

CF can also affect other parts of the body, which can lead to problems like sinusitis and infertility. To learn more about these conditions, visit CFSource.com.
KNOWLEDGE IS:
UNCOVERING PROGRESSION

MAUREEN
Living with CF
Beginning early, the buildup of thick, sticky mucus in the lungs results in a cycle of infection, inflammation, and more mucus buildup. Pulmonary exacerbations are often a key part of this cycle. This is especially serious because a pulmonary exacerbation means more than just a hospital stay or an extra round of antibiotics. It may cause permanent lung damage that advances disease progression.

The cycle of pulmonary exacerbations often leads to a condition called bronchiectasis. This is a permanent reshaping of the airways that causes them to become loose and scarred. It can make it harder to clear mucus from the lungs and for the airways to move air in and out as a person breathes.

**Repeate pulmonary exacerbations cause lung damage**

In CF, permanent lung damage may occur even before it can be detected by lung function tests.

Starting from a young age, people with CF may experience a lung function decline of 1 to 3 percentage points each year on average.
A change in lung function can’t always be felt

Part of what makes this decline so serious is that some people don’t feel the change. So it often isn’t until someone has lost a lot of lung function that he or she begins to feel like something is wrong.

Disease progression is universal

CF is different for everyone. But all cases of CF progress over time. Taking a proactive approach to managing CF is the first step toward managing disease.
As someone with CF grows up, the same mucus that caused digestive problems in childhood may have caused scarring in different digestive organs. This scarring can result in the development of other conditions.

Damage caused by thick digestive juices can lead to scarring that makes it more difficult for the pancreas to do its job. As this scarring builds up, many people with CF develop CFRD. This is when the pancreas does not make enough insulin, and the body may not be able to properly use the insulin that is produced.
Although CFRD is serious, the symptoms are often similar to CF or not noticeable. It’s also important to note that CFRD is different from Type 1 and Type 2 diabetes and therefore is treated differently.

Beginning at an early age, you should expect your care teams to check for CFRD regularly.
Loss of function can’t always be felt

The liver

Cirrhosis
Over time, the thick mucus causes scarring and damage to the liver. It sometimes leads to cirrhosis, an advanced stage of liver disease that makes it difficult for blood to travel through the liver. This can result in many complications, including infections, malnutrition, and sometimes liver failure.

Up to 30% of people with CF develop liver disease.
Liver disease is considered to be one of the most serious health risks associated with CF. There are often no symptoms until the scarring and damage have reached an advanced stage.

Care teams regularly monitor liver function in people with CF to check for progression in the liver.

Show off your CF knowledge by taking the CF Fact or Fiction quiz.

Visit CFSource.com to take the quiz now.
For people with CF, 2 different factors contribute to low bone density:

- **Bones don’t grow strong**
  For some people, bone development is affected because they may have trouble absorbing enough vitamin D for healthy bone growth. As a result, bones are weak and can fracture more easily

- **Bones lose density**
  Some people may lose bone density more easily. The inflammation caused by frequent lung infections, as well as the steroids used to treat these infections, can cause bone tissue to break down

Both of these factors can contribute to osteoporosis, a disease that can lead to stooped posture, loss of height, and bones that break easily.

**Care teams begin screening people with CF for bone health at an early age.**
About 70% of adults living with CF will develop osteoporosis, compared to 55% of adults who do not have CF.
KNOWLEDGE IS: ADVANCING CF MANAGEMENT
ADVANCES IN CF MANAGEMENT

CF management has come a long way in a short period of time
It is always important to stay up to date on the latest advances and maintain your health as best as possible. Advancements in diagnosis and disease management have increased the life expectancy of people living with CF.

• In 1955, the median predicted survival for someone with CF was 5 years
• In 1985, the median predicted survival for someone with CF was 25 years
• In 2015, the median predicted survival for someone with CF was 42 years

Median predicted survival numbers are based on data relating to all people in the CFF Patient Registry. Median predicted survival means that in this year, half the people with CF were expected to live longer and half were expected to live less than this age.

HIGHLIGHTS IN CF HISTORY
Advances in science, medicine, and the care of people with CF continue to be made

Understanding of CF
• In 1989, the cystic fibrosis transmembrane conductance regulator (CFTR) gene was identified as the gene responsible for the defective CFTR proteins that cause CF
Developments in CF management

1959
Sweat chloride testing becomes a standard procedure for diagnosing CF.

1978
Coated pancreatic enzymes that help with nutrient absorption are introduced.

1997
Inhaled antibiotics targeting lung bacteria arrived.

1993
Mucolytics (mucus-thinning medicines) are introduced.

1988
A vest is introduced to make chest physical therapy at home easier.

2012
Medicines that modulate activity of the CFTR protein are introduced.

2015
Nearly 60% of new CF diagnoses are detected by newborn screening.

To learn more about available treatments to help manage symptoms and treat CF, talk to your care team.
One of the most common tests used to measure CF progression is a **pulmonary function test**. People with CF may know it better as **FEV<sub>1</sub>**, or forced expiratory volume in 1 second. The FEV<sub>1</sub> test is usually done after age 6.

But FEV<sub>1</sub> does not always find the beginnings of permanent lung damage and disease progression. There are other tests that can show progression in the lungs as well. You can learn more about the tests shown below at [CFSource.com](http://CFSource.com).

**Computed tomographic (CT) scans**

**Magnetic resonance imaging (MRI)**

**Lung clearance index (LCI)**
There are also other tests that allow care teams to determine the impact of CF on the body. You can learn more about the tests shown below at CFSource.com.

- Sweat chloride levels
- Palpation
- Fecal elastase-1
- Immunoreactive trypsinogen (IRT)
- Blood sugar testing
- Dual energy x-ray absorptiometry
- Bacterial culturing
- Transaminase levels
- Palpation
- Sinus endoscopy
Whether you have CF or are a parent or caregiver for someone with CF, it’s common to have questions about what progression looks and feels like, how it may affect you or your child, and how to measure and track it. Here are a few questions you may want to ask at your next care center visit.

**What are some signs of disease progression?**

**How can someone with CF find out if they’re experiencing progression?**

**What are the signs of cystic fibrosis–related diabetes (CFRD)?**

**What kinds of tests are used to measure progression?**

**What are the signs of distal intestinal obstruction syndrome (DIOS)?**
Have more questions? Add your own here:
LEARN MORE ABOUT CF PROGRESSION, and get links to helpful online resources, such as videos, by visiting CFSource.com

THE CFDAILY iPHONE® APP

Help for your disease management routine

CFDaily is a free iPhone® app for people with CF. This app is designed especially for people with CF and their caregivers to help manage their treatment routines. Visit CFDailyApp.com to learn more.

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