

#### **CYSTIC FIBROSIS** SOPHIE WELLS

## WHAT IS CYSTIC FIBROSIS?

Cystic fibrosis is a condition that is inherited from parents who have CF (cystic fibrosis) or are carriers of the gene. The disease is progressive over the life span of an individual meaning some have significantly shortened life then others. Cystic fibrosis affects the lungs causing persistent infections and limits the individuals ability to breath. CF has long lasting affects on other organs like the digestive system and affects all organ function. The Cystic fibrosis trust help to educate and help those who are suffering with support and donations towards looking at new treatments. This condition is to be affect over 10,000 people within the uk. Internationally over 70,000 individuals or more are suffering from cystic fibrosis.



### HOW DOES CYSTIC FIBROSIS AFFECT THE BODY?

- Cystic fibrosis gene is what controls the movement of salt and water in the cells of the lungs. This is how their is a build up of mucus within the lungs that is able to affect the other organs like this mucus can affect the digestive system.
- This build up of mucus is what causes the difficulty of breathing and increases the risk of the lung infections.
  Over prolonged periods of time the lung capacity decreases and may stop working this is where things like a lung transplant is one of the very last suggestions. The effects on organs such as the digestive system take place when mucus ends up travelling and clogging the pancreas, this then has an effect on how the enzymes are able to digest the food within our stomach which is why many individuals witch CF commonly have feeding tubes or other alternatives.
- Their are many different symptoms that are able to contribute towards if an individual has cystic fibrosis that may be a sign of having cystic fibrosis: recurring chest infections, wheezing, shortness of breath, malnutrition and stunted growth.



### DIAGNOSING & TESTING FOR CYSTIC FIBROSIS

- Throughout the uk, screening to look for cystic fibrosis takes place on all newborns. This is a simple test that is carried out after a short term of babies being born. If the tests come back positive the first time, multiple others are done to ensure and have this confirmed. After this has been diagnosed they will be referred to specialists in order to start treatment and support them.
- Tests are able to be done on older children and adults who some may not have had this cystic fibrosis check. The sweat test is used to measure how much salt their is within your sweat. Those who suffer from cystic fibrosis have a high amount of salt within their sweat, this is a high indicator of having CF. The other form of testing is genetic most commonly done using blood to look closely at your genes if their are any abnormal ones that could cause you to have cystic fibrosis. Genetic testing is also a way of finding out if you are a carrier for cystic fibrosis if this is something that may run through your family.
- The abnormal gene has to be in both parents for a child to have cystic fibrosis, this is most commonly why it is not a high population of those who have it. one individual within 25 are commonly carriers of the cystic fibrosis gene.



# LIVING WITH CYSTIC FIBROSIS

living with cystic fibrosis has many strains on different aspects such as: socialising, nutrition and overall quality of life.

- Nutritionally cystic fibrosis affects the enzymes meaning food is unable to be absorbed or very small amounts. This means many individuals suffering with CF are malnourished and struggle with maintaining a good weight. This eventually becomes so bad CF most commonly have feeding tubes to help them digest food and collect nutrients. Their are two types of tubes that are most commonly given to CF patients: gastrostomy tube or nasogastric. A gastrostomy tube is surgically placed on the abdomen straight into the stomach, this solution to the nutrition is usually long term. A nasogastric tube is inserted through the nose, down the oesophagus and into the stomach, because of the irritation to the nose and throat this is usually a short term solution however it is a much safer one and has less risk of infection whereas the gastrostomy one has higher risk as it is an open wound.
  - Those living with the condition are more commonly susceptible to having more conditions overtime that are related to CF such as diabetes, liver problems, brittle bones and fertility problems. Though most commonly have a wide range of life cystic fibrosis declines over time and worsens usually as an individual gets older or if they catch a serious infection. The usually life expectancy for those with cystic fibrosis is 40 however through developing technology this is rising.

## TREATMENTS

- Cystic fibrosis is one of the many few diseases that has no real cure. The range of treatments that are offered are to improve quality of life, reduce symptoms, prevent further complications and help to elongate life expectancy. Those who have cystic fibrosis are monitored by their doctors and have regular appointments.
- The lung problems caused by CF need to be treat with different medicines to help prevent more lung problems these are both swallowed, inhaled or injected. Antibiotics are used to prevent or treat any chest infections that may occur. Other medicines are used to thin the mucus making it easier to cough up.
- Physical techniques are another to actively open the lungs and reduce mucus such as airway clearances devices send vibration and air pressure to help remove mucus from the airways. oxygen therapy is most commonly used throughout and involves using oxygen within everyday life to help reduce symptoms and increase oxygen flow to the body.
- Severe cases of cystic fibrosis occur the last option is getting a lung transplants. The lungs are so damaged all treatments that were taking place have failed to help and this is where lung failure is most commonly.

