At-A-Glance report 2016

Cystic Fibrosis in Europe – Facts and Figures 2016

The European Cystic Fibrosis Society Patient Registry (ECFSPR) is happy to present this report with key information about how cystic fibrosis (CF) affects people with CF and their families throughout Europe.

The ECFSPR collects, measures and compares data of people with CF living in Europe and neighbouring countries who agree to be in the registry. The information is important to better understand CF, encourage new European standards of care and treatment, conduct research, and inform public health-planning.

If you want to know more about the ECFSPR visit our website.

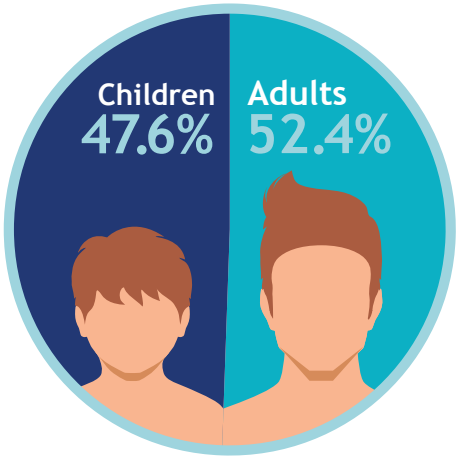
For more in depth information see the full report here.

June 2018

Map of countries
that contributed 2016 data
to the ECFSPR

44,719

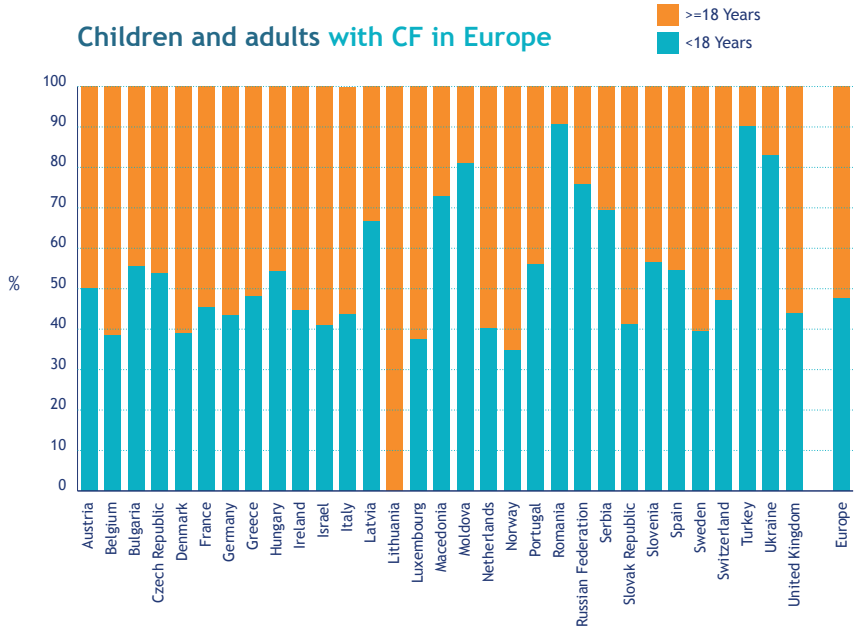
registered patients in 31 countries
number of patients on map



Proportion children-adults

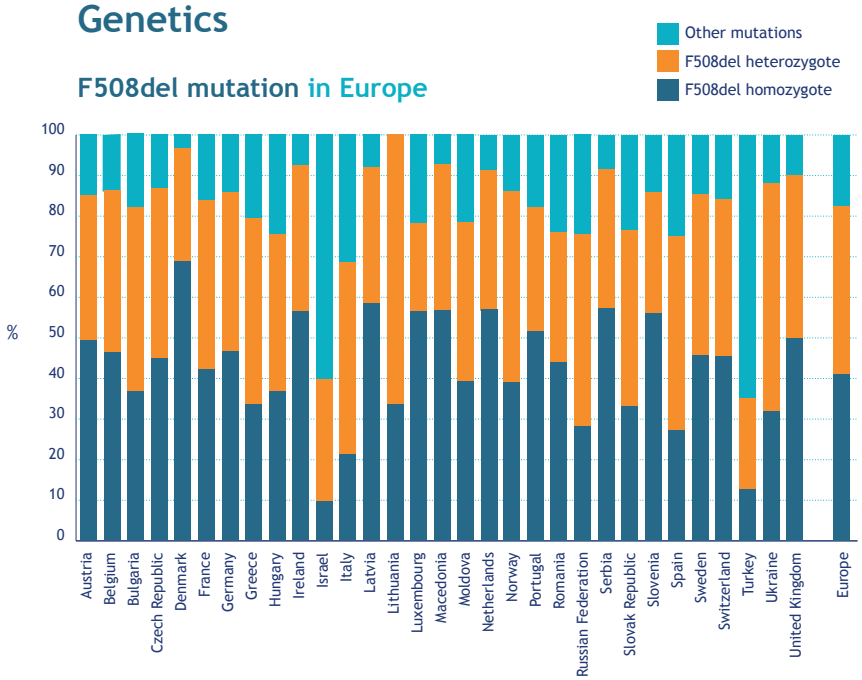
- The proportion children-adults varies between the countries.
- For some countries only a few individual centres sent data to the ECFSPR.

Children and adults with CF in Europe

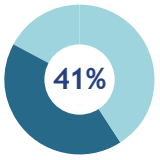


Genetics

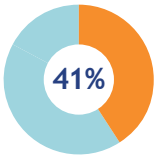
F508del mutation in Europe



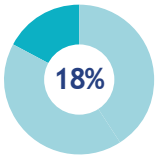
- F508del is the most common CF-causing mutation in Europe.
- People with CF have two CF-causing mutations, one inherited from the mother and one from the father.
- **Homozygous:** both mutations are the same.
- **Heterozygous:** the two mutations are different.



F508del homozygote



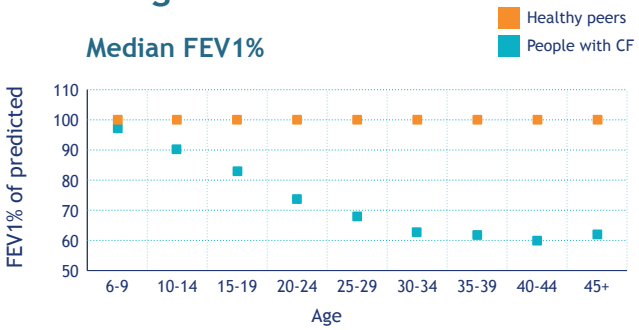
F508del heterozygote

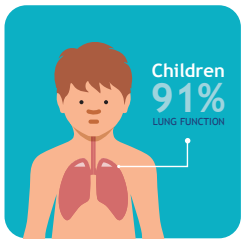


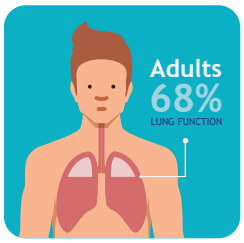
Other mutations

Lung function

Median FEV1%

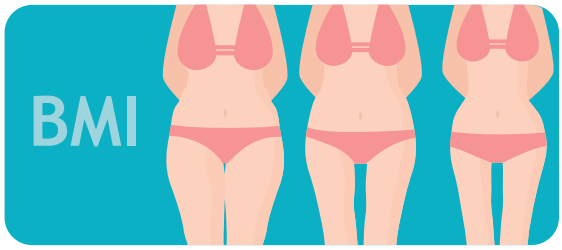






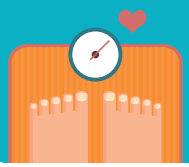
- FEV1 is a measure for lung function. It is the maximum amount of air that can be forcefully exhaled in the first second after taking a deep breath.
- FEV1% is a percentage of the average value for healthy people of the same age, gender and height, which is set at 100%.
- In people with CF aged 45 and older there is a higher incidence of mutations causing milder forms of CF. This influences the lung function value for the people with CF in this age group.

Nutrition



Median means that 50% of the values are above and 50% are below this value.

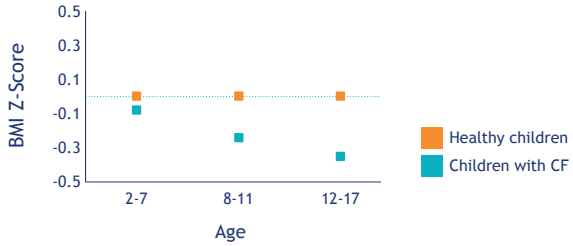
BMI or body mass index is a measure for nutritional status:



$$\frac{\text{weight}}{\text{height}^2}$$

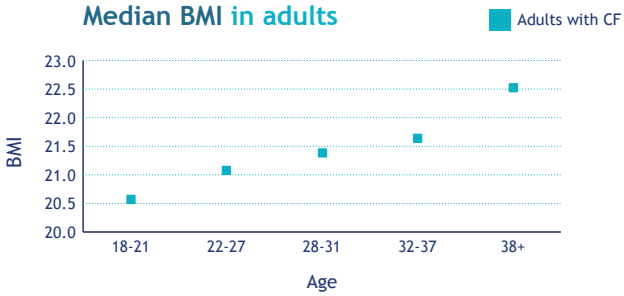
z-score indicates how far a value is from the average (mean) value of the reference population.

Median BMI Z-score in children



- A z-score of 0 means that the BMI is the same as the BMI of healthy children of the same gender and age.
- From the age of 8, children with CF have a lower average BMI than healthy children of a similar age.

Median BMI in adults

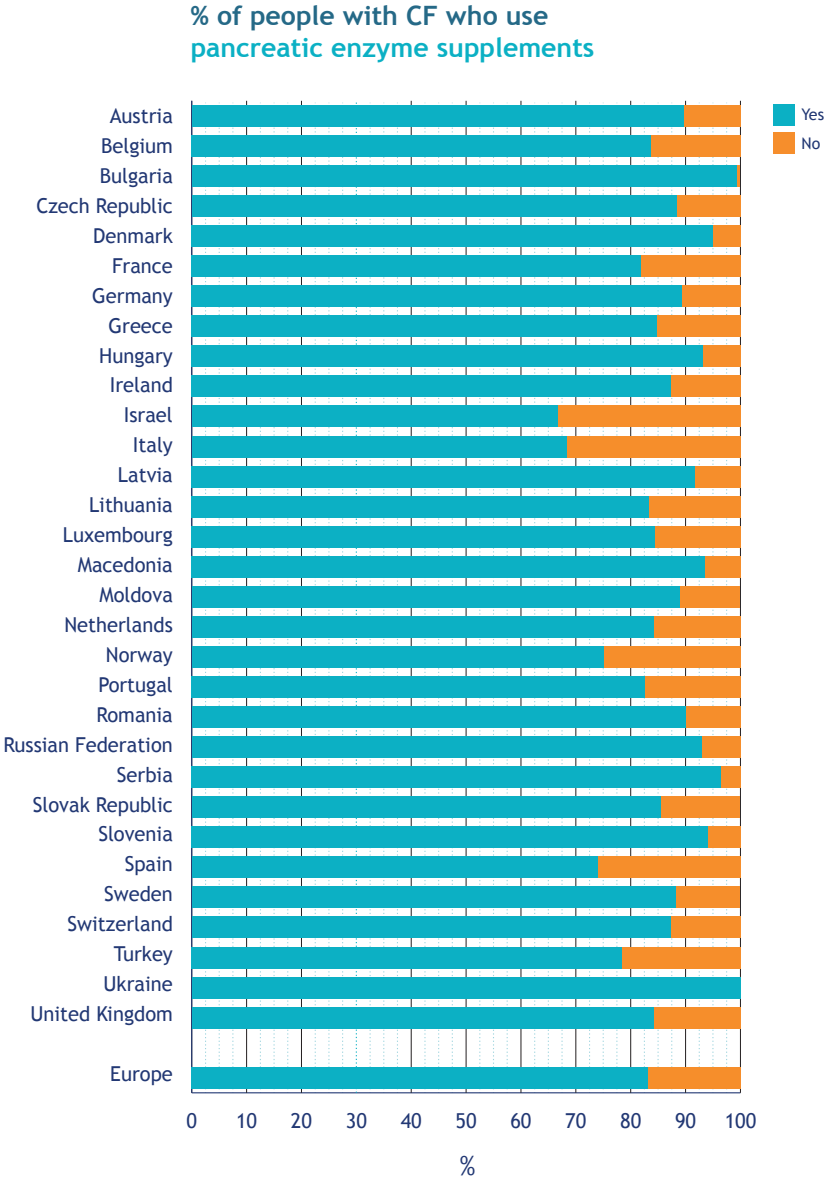


- Healthy peers have values for BMI between 18 and 25.
- As people with CF get older BMI increases.
- In adults with CF aged 38 and older there is a higher incidence of mutations causing milder forms of CF. This influences the BMI for the adults in this age group.

Pancreatic enzymes

83% of the people with CF use pancreatic enzyme supplements.*

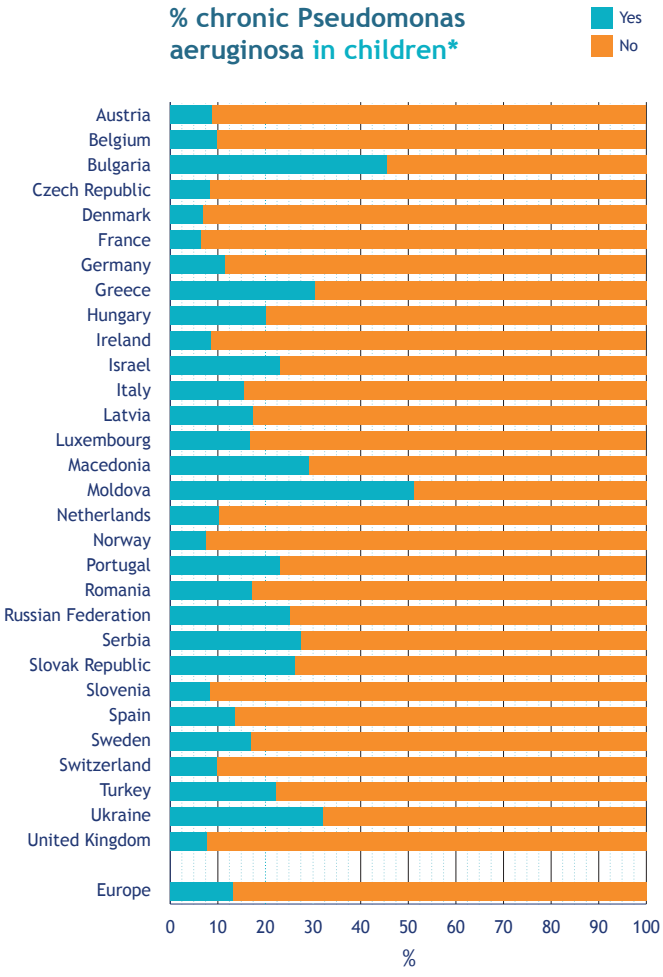
- Enzymes released from the pancreas are vital for the digestion and absorption of fat and vitamins from food. In many people with CF the release of these enzymes is blocked and they must take supplements.



Infection

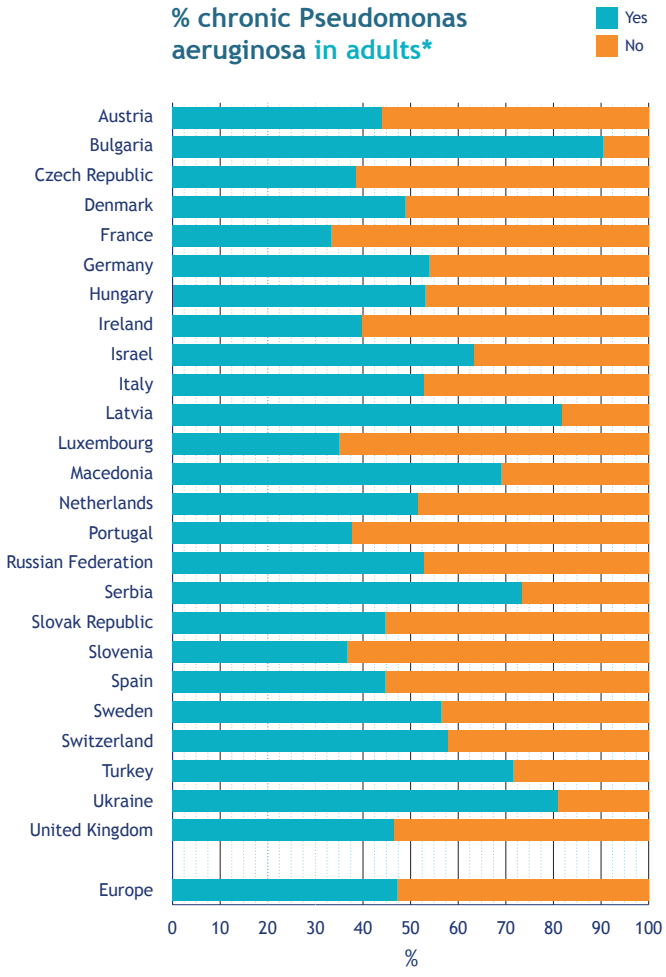
31% of the people with CF are chronically infected with the **Pseudomonas aeruginosa** bacteria in their lungs.*

% chronic Pseudomonas aeruginosa in children*



* Missing values are not included nor are data from countries with a high % of missing values.

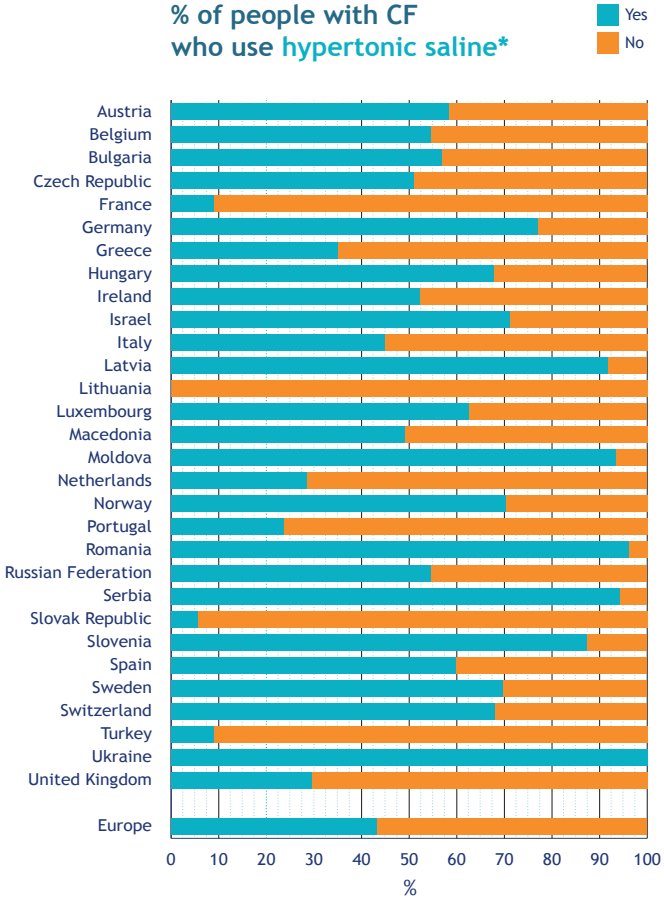
% chronic Pseudomonas aeruginosa in adults*



Note: Moldova and Romania have less than 5 patients of 18 years or older, and are therefore excluded from the graph.

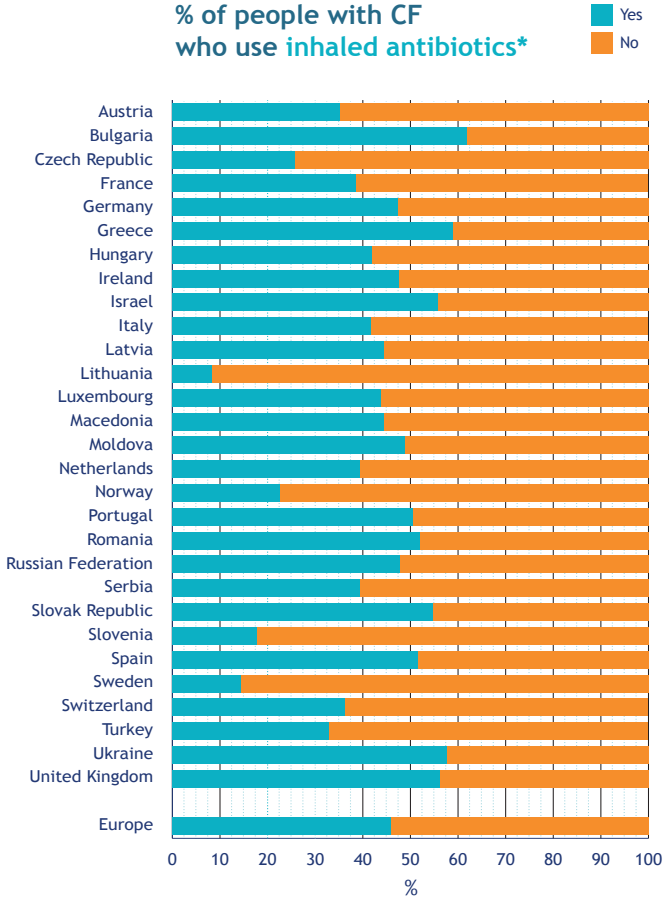
Inhalation Treatment

% of people with CF who use hypertonic saline*



Hypertonic saline is prescribed to clear thick mucus from the lungs.

% of people with CF who use inhaled antibiotics*

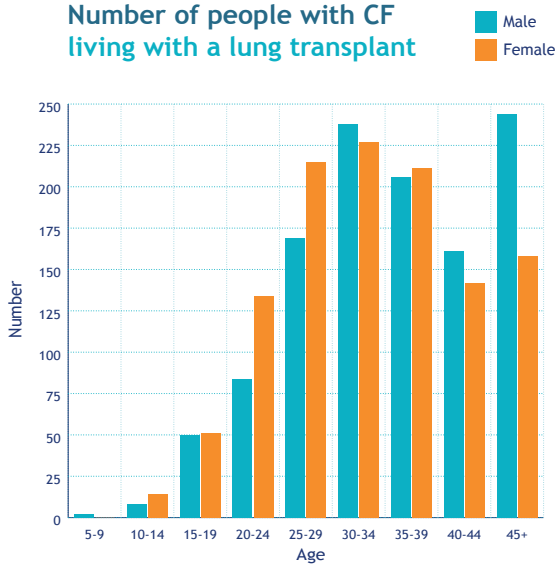


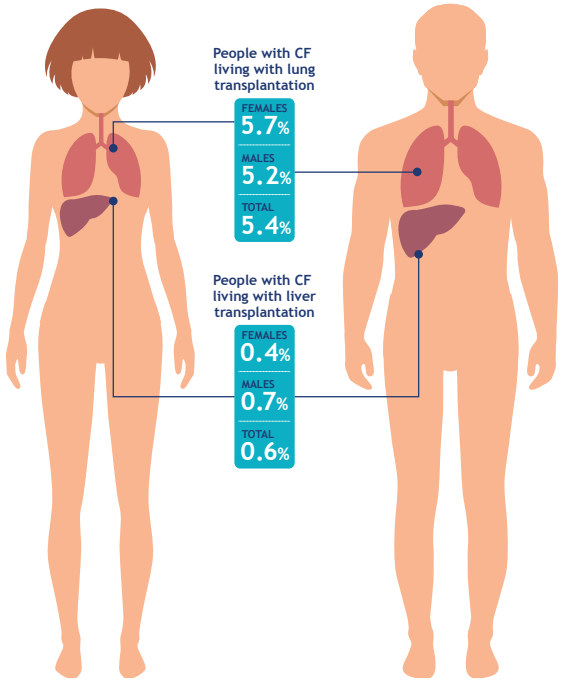
Inhaled antibiotics are prescribed to treat CF specific bacterial infection, such as chronic Pseudomonas aeruginosa.

* In these graphs the missing values are not included nor are data from countries with a high % of missing values.

Transplantation

Number of people with CF living with a lung transplant






ecfs

Europe

GILEAD

VERTEX