At-A-Glance
report 2013

Cystic Fibrosis in Europe – Facts and Figures 2013

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 European public health-planning.
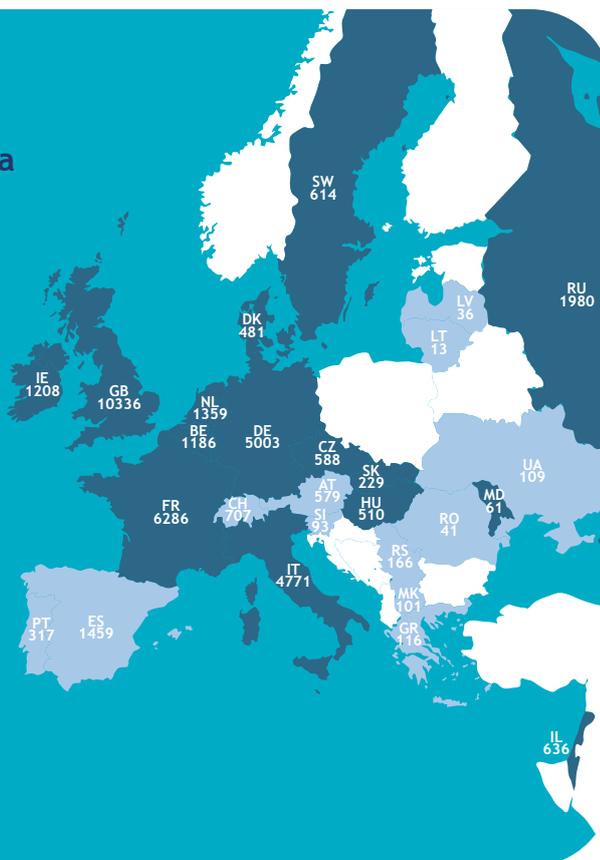
If you want to know more about the ECFSPR visit our website

For more in depth information see the full report here

March 2016

Map of countries
that contributed 2013 data
to the ECFSPR

- Countries that sent the data to the ECFSPR as a national registry
- Countries with individual centres that sent data
- Countries not in the ECFSPR

SW
614

RU
1980

LV
36

LT
13

DK
481

IE
1208

GB
10336

NL
1359

BE
1186

DE
5003

CZ
588

SK
229

UA
109

AT
579

HU
510

FR
6286

CH
707

SI
93

MD
61

RO
41

IT
4771

RS
166

MK
101

GR
116

PT
317

ES
1459

IL
636

38,985
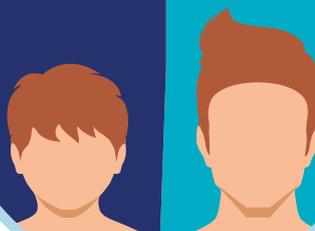## registered patients in 27 countries; number of patients on map.
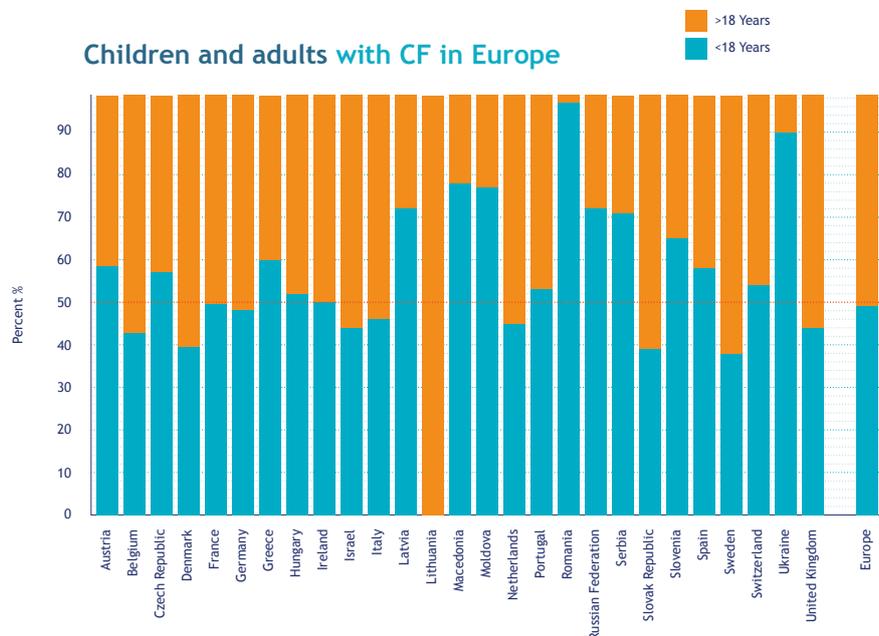
MEDIAN AGE OF
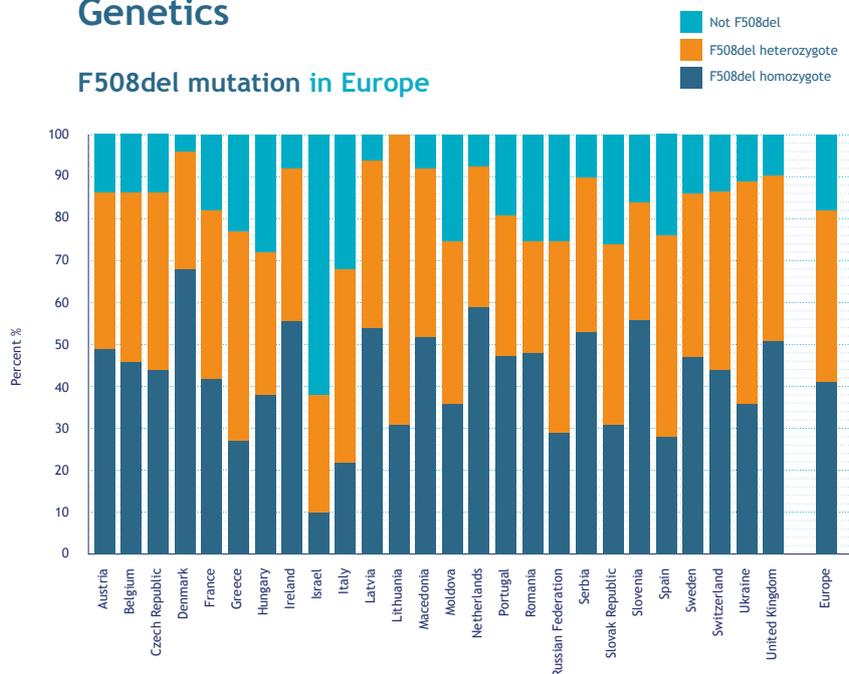DIAGNOSIS
3.6
months

Children
49.1%

Adults
50.9%

Proportion children-adults

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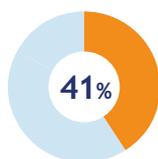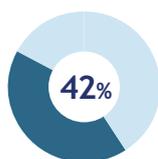· For some countries only a few individual centres sent data to the ECFSPR.

Children and adults with CF in Europe

Legend: >18 Years, <18 Years

Genetics

Legend: Not F508del, F508del heterozygote, F508del homozygote
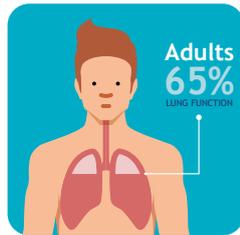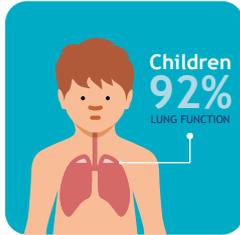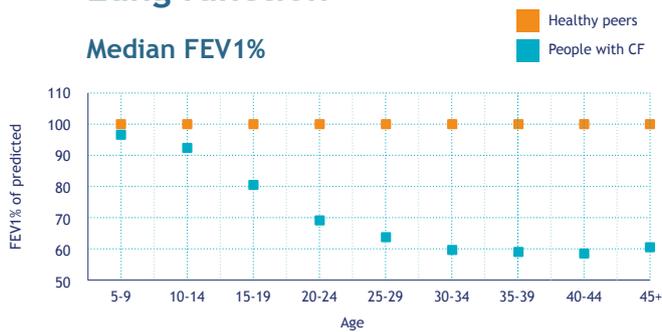
F508del mutation in Europe

- F508 del 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.

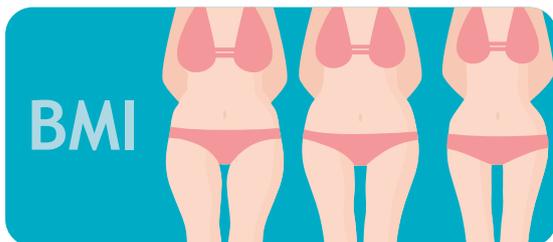- **Heterozygous:** the two mutations are different.

42% F508del homozygote

41% F508del heterozygote

17% Other mutations

Lung function

Median FEV1%

Legend: ■ Healthy peers ■ People with CF

Chart: Median FEV1%
- Y-axis: FEV1% of predicted (50 to 110)
- X-axis: Age (5-9, 10-14, 15-19, 20-24, 25-29, 30-34, 35-39, 40-44, 45+)

Children **92%** LUNG FUNCTION

Adults **65%** LUNG FUNCTION

- FEV1 is a measure for lung function. It is the maximum amount of air that forcefully can be 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 patients aged 45 and older there is a higher incidence of mutations causing milder forms of CF. This influences the lung function value for the patients in this age group represented.
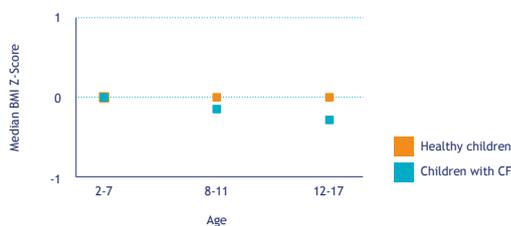
Nutrition

BMI

**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:

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

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

Median BMI Z-score in children

Chart: Median BMI Z-score in children
- Y-axis: Median BMI Z-Score (-1 to 1)
- X-axis: Age (2-7, 8-11, 12-17)
- Legend: ■ Healthy children ■ Children with CF

Median BMI in adults

Chart: Median BMI in adults
- Y-axis: Median BMI (19 to 23)
- X-axis: Age (18-21, 22-27, 28-31, 32-37, 38+)
- Legend: ■ Adults with CF

- A z-score of 0 means that the BMI is the same as the BMI of healthy peers of the same gender and age.

- From the age of 6, children with CF have a lower average BMI than their healthy peers.
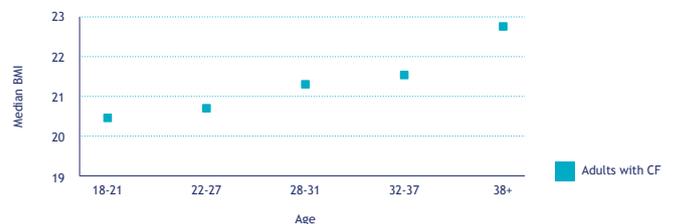
- Healthy peers have values for BMI between 18 and 25.

- As patients get older the BMI values increase.

- In patients aged 38 and older there is a higher incidence of mutations causing milder forms of CF. This influences the BMI for the patients in this age group represented.

Pancreatic enzymes

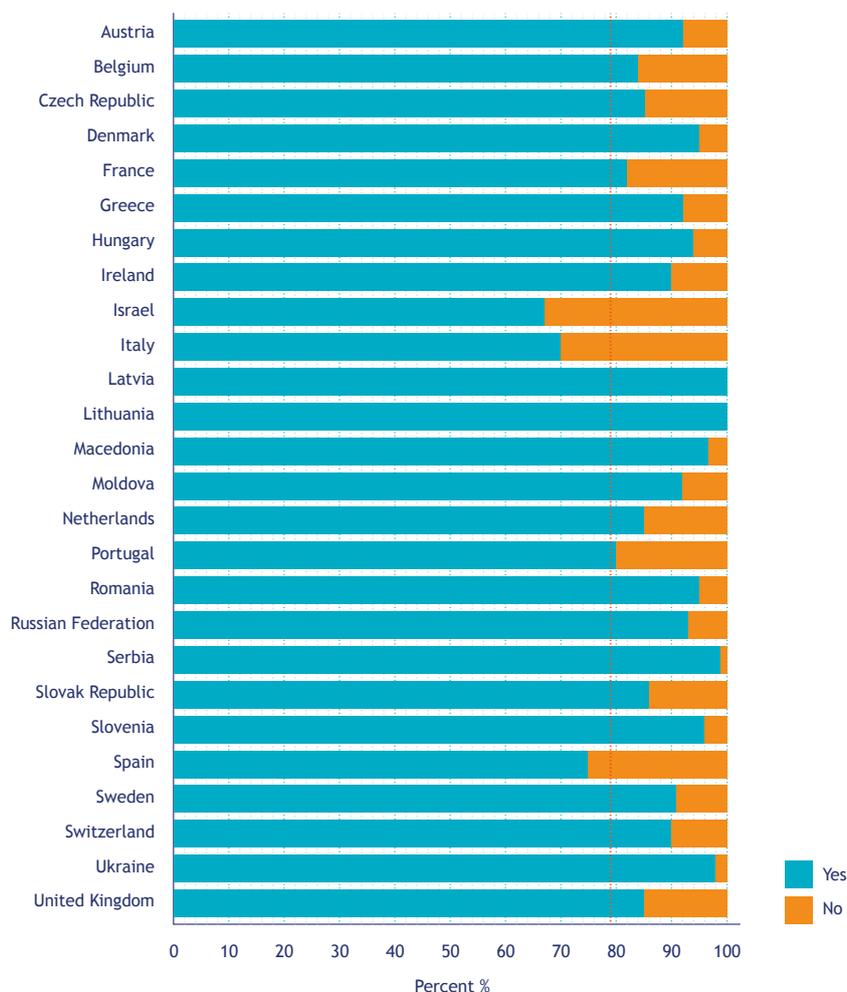**79%**
Pancreatic enzymes
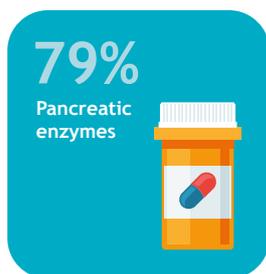
79% of the people with CF are using **pancreatic enzyme** supplementation.

- 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.
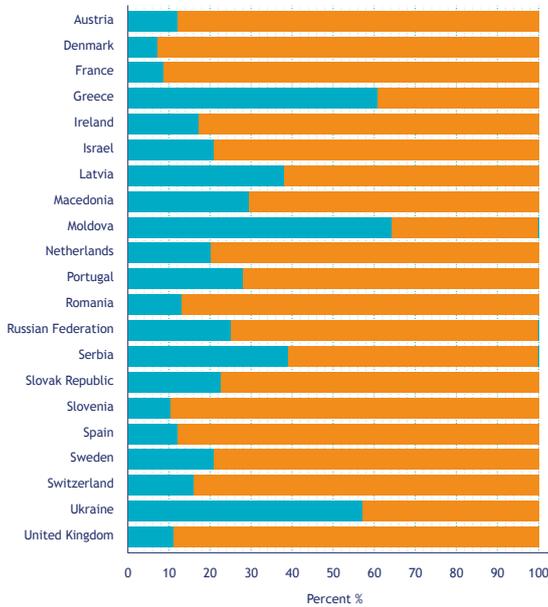
% of patients who use pancreatic enzyme supplements

Percent %

- Yes
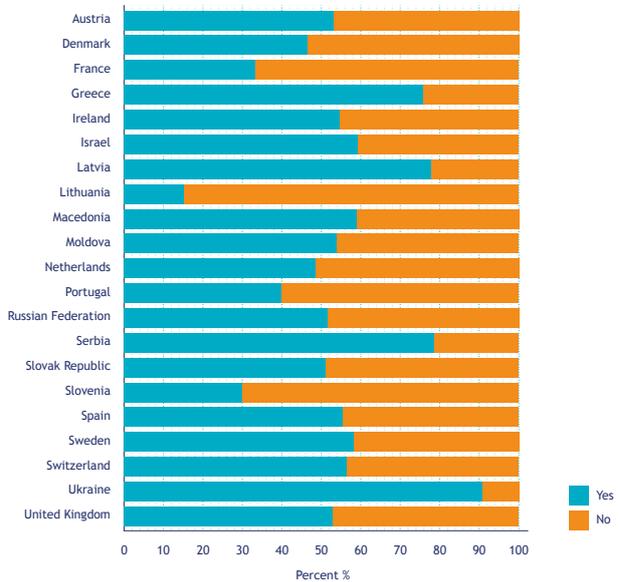- No

Infection

Pseudomonas 33%

33%

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

% chronic Pseudomonas aeruginosa in children*

Austria
Denmark
France
Greece
Ireland
Israel
Latvia
Macedonia
Moldova
Netherlands
Portugal
Romania
Russian Federation
Serbia
Slovak Republic
Slovenia
Spain
Sweden
Switzerland
Ukraine
United Kingdom

0 10 20 30 40 50 60 70 80 90 100

Percent %

■ Yes
■ No

% chronic Pseudomonas aeruginosa in adults*

Austria
Denmark
France
Greece
Ireland
Israel
Latvia
Lithuania
Macedonia
Moldova
Netherlands
Portugal
Russian Federation
Serbia
Slovak Republic
Slovenia
Spain
Sweden
Switzerland
Ukraine
United Kingdom

0 10 20 30 40 50 60 70 80 90 100

Percent %

■ Yes
■ No

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

Inhalation Treatment

% of patients on hypertonic saline*

Countries (top to bottom): Austria, Czech Republic, France, Greece, Ireland, Israel, Latvia, Lithuania, Macedonia, Moldova, Netherlands, Portugal, Romania, Russian Federation, Serbia, Slovak Republic, Slovenia, Spain, Sweden, Switzerland, Ukraine, United Kingdom

Legend: Yes / No
X-axis: Percent %

Hypertonic saline is prescribed to help to clear thick mucus from the lungs.
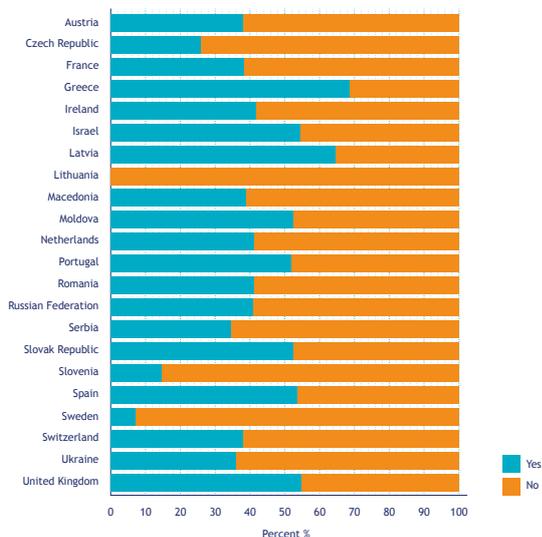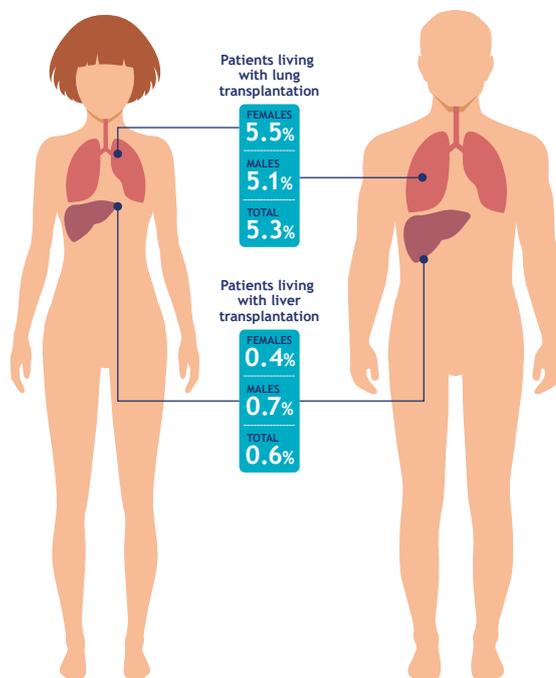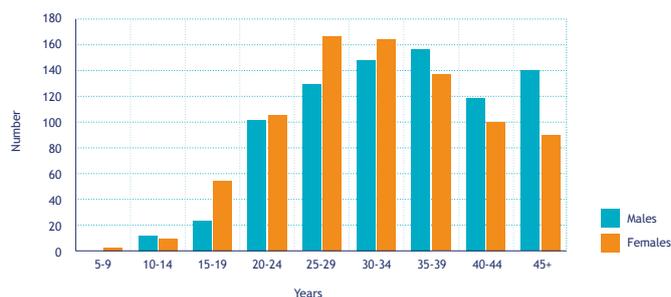
* In this graph the missing values are not included nor are data from countries with a high % of missing values.

% of patients on inhaled antibiotics*

Countries (top to bottom): Austria, Czech Republic, France, Greece, Ireland, Israel, Latvia, Lithuania, Macedonia, Moldova, Netherlands, Portugal, Romania, Russian Federation, Serbia, Slovak Republic, Slovenia, Spain, Sweden, Switzerland, Ukraine, United Kingdom

Legend: Yes / No
X-axis: Percent %

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

Transplantation

Number of people with CF living with a lung transplant

X-axis: Years (5-9, 10-14, 15-19, 20-24, 25-29, 30-34, 35-39, 40-44, 45+)
Y-axis: Number
Legend: Males / Females

Patients living with lung transplantation
FEMALES 5.5%
MALES 5.1%
TOTAL 5.3%

Patients living with liver transplantation
FEMALES 0.4%
MALES 0.7%
TOTAL 0.6%