The European Cystic Fibrosis Society Patient Registry

Map of countries that contributed 2014 data to the ECFSPR

SW 626 · DK 488 · LV 38 · RU 2122 · LT 12 · IE 1218 · UK 10581 · NL 1378 · BE 1186 · CZ 599 · SK 290 · UA 147 · FR 6369 · CH 782 · AT 712 · HU 557 · MD 61 · SI 97 · RS 169 · RO 44 · IT 4981 · PT 295 · ES 1538 · MK 109 · GR 517 · IL 666

Why is a European CF Patient Registry necessary?

Cystic Fibrosis (CF) is a rare disease. To get a good picture of CF in Europe we need as much data as possible. The data will help to understand the disease, encourage new European standards of care, conduct research and inform public health-planning.

The European Cystic Fibrosis Society Patient Registry

Collects, measures and compares data of children and adults with cystic fibrosis living in Europe and neighbouring countries who agree to be in the Registry. The information is used to improve the health and wellbeing of people with CF.
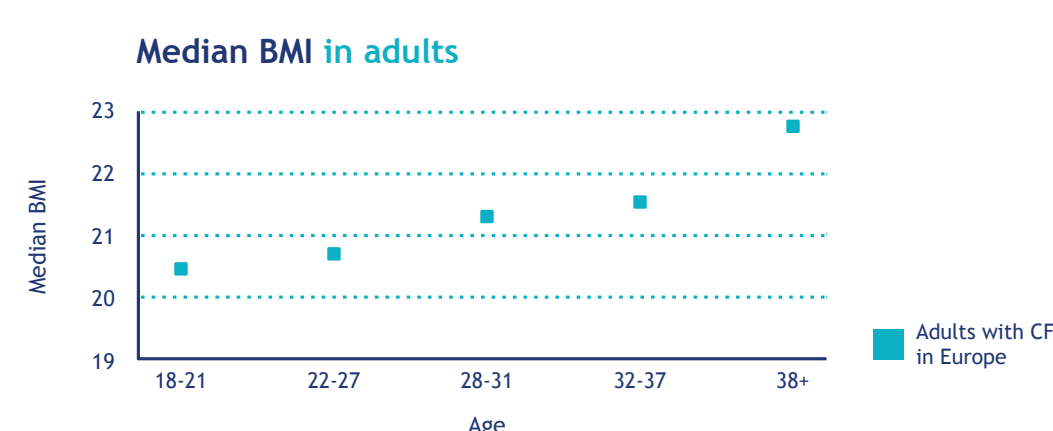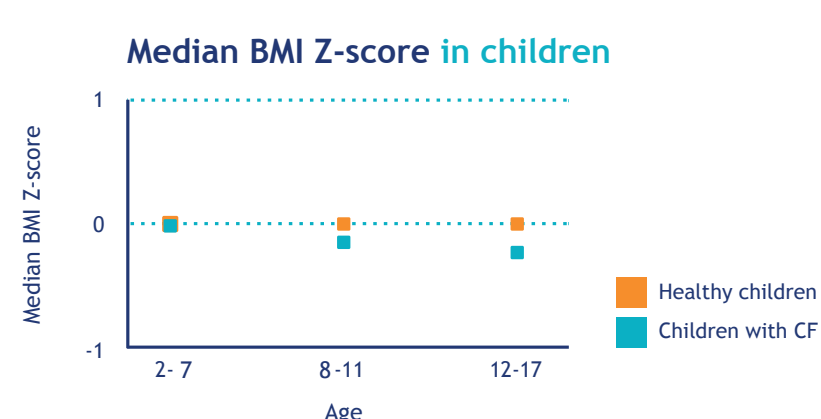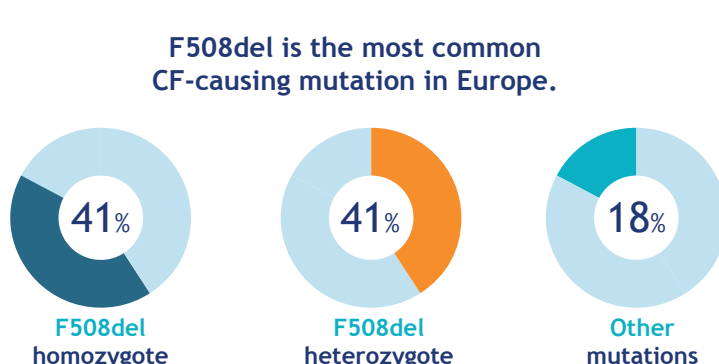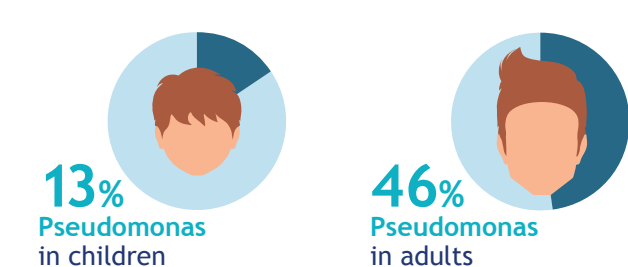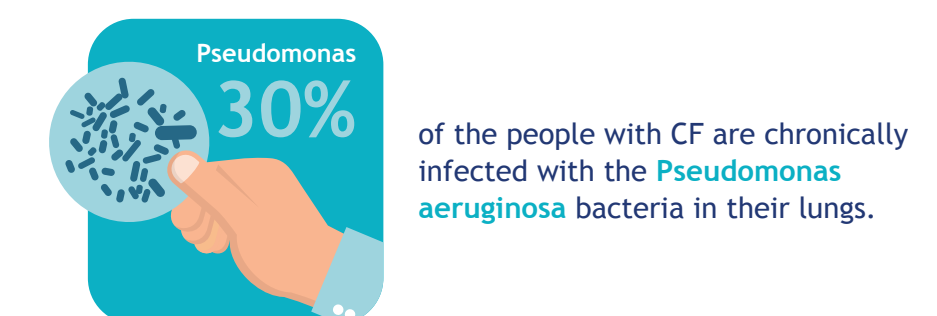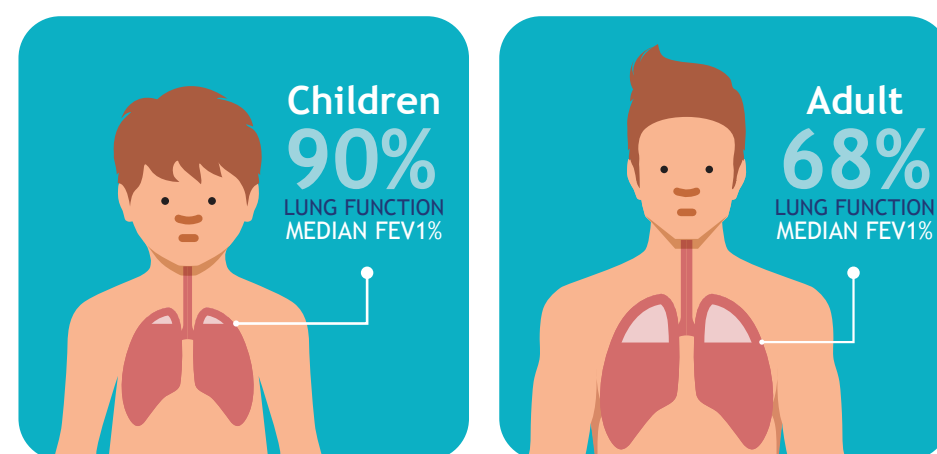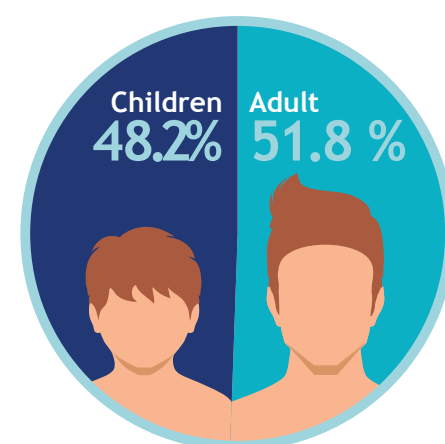
How can I use the Registry's information?

The ECFSPR publishes: **Annual data reports** with demographic and clinical outcomes across Europe and **At-a-glance reports** with key information about CF in Europe.
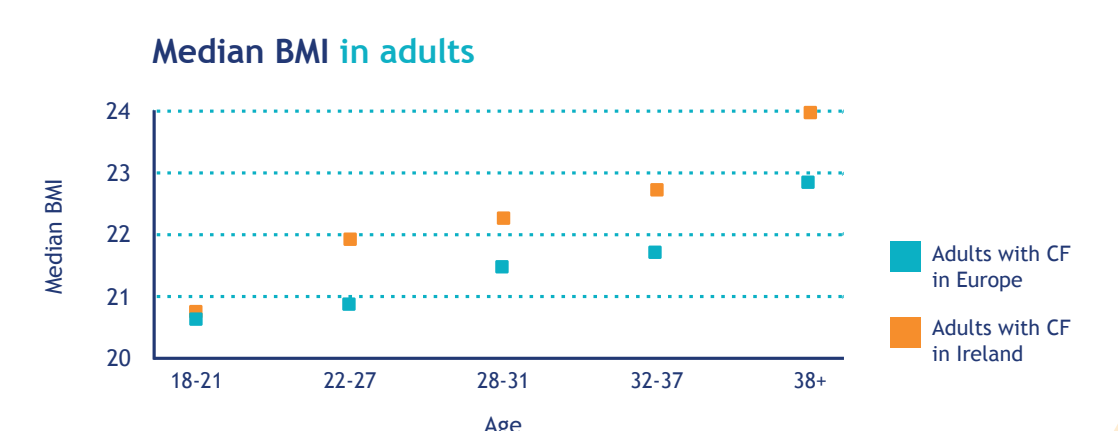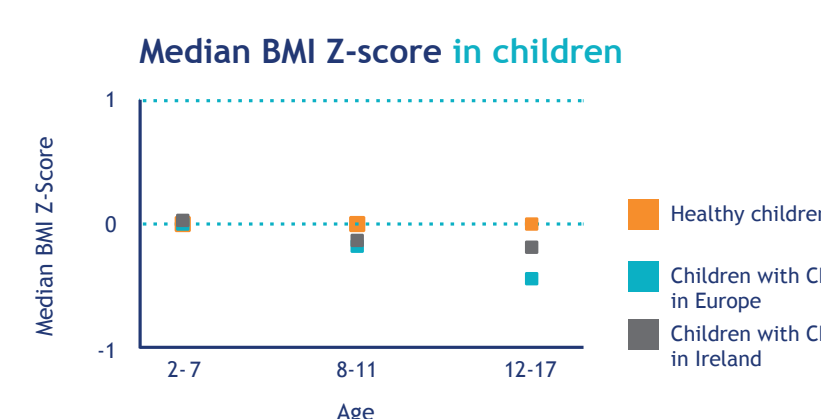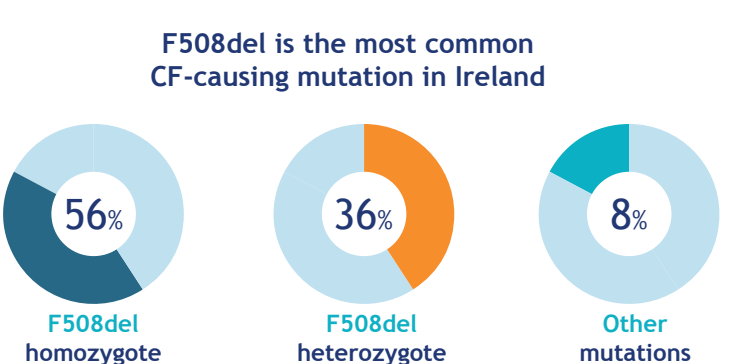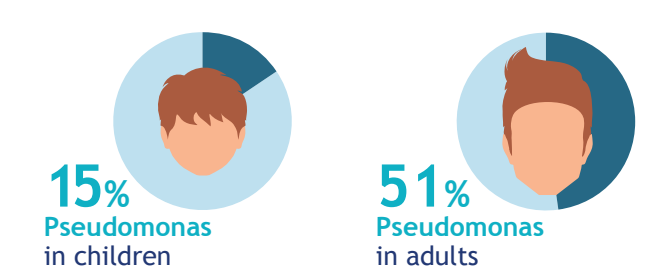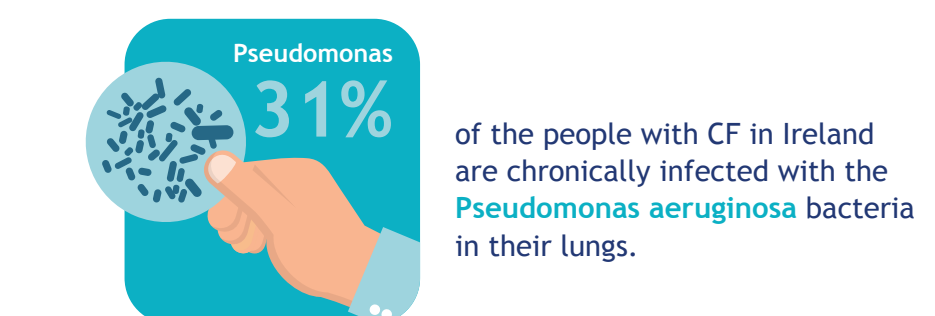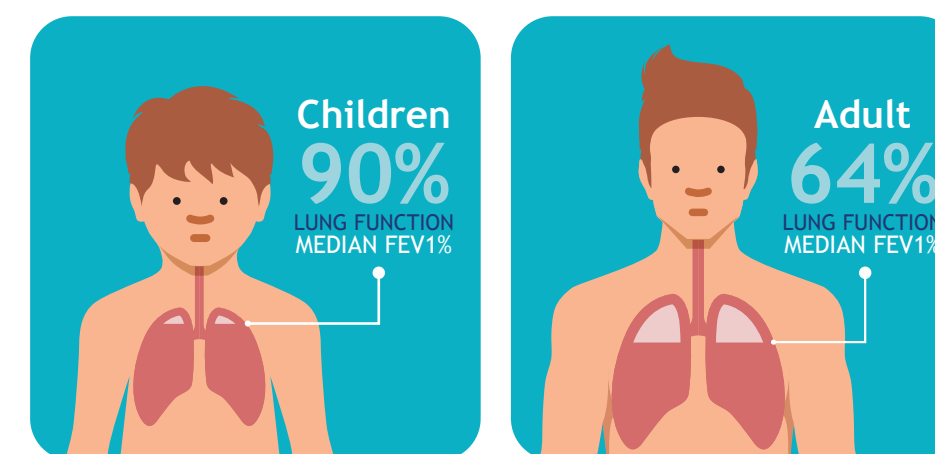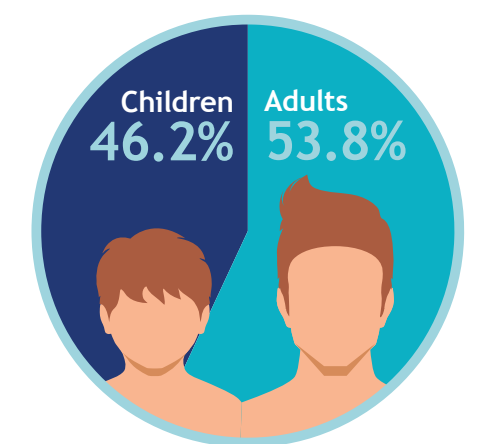
These reports are valuable tools for:

- local patient organisation to discuss improvements in CF care in your country with the national healthcare organisation and other relevant parties.
- CF centre(s) to compare with other countries and identify fields for improvement.
- you to discuss outcomes and results with your doctor.

CF in Europe

35,582 registered patients in 26 countries; number of patients on map above.

Children 48.2% · Adult 51.8%

Children **90%** LUNG FUNCTION MEDIAN FEV1% · Adult **68%** LUNG FUNCTION MEDIAN FEV1%

MEDIAN AGE OF DIAGNOSIS **3.6** months

Pseudomonas **30%** of the people with CF are chronically infected with the **Pseudomonas aeruginosa** bacteria in their lungs.

13% Pseudomonas in children · **46%** Pseudomonas in adults

85% Pancreatic enzymes of the people with CF are using **pancreatic enzyme** supplementation.

F508del is the most common CF-causing mutation in Europe.

41% F508del homozygote · **41%** F508del heterozygote · **18%** Other mutations

Median BMI Z-score in children

Median BMI Z-score · Age · 2-7 · 8-11 · 12-17 · Healthy children · Children with CF

Median BMI in adults

Median BMI · Age · 18-21 · 22-27 · 28-31 · 32-37 · 38+ · Adults with CF in Europe

CF in Ireland

1,218 registered patients in Ireland

Children 46.2% · Adults 53.8%

Children **90%** LUNG FUNCTION MEDIAN FEV1% · Adult **64%** LUNG FUNCTION MEDIAN FEV1%

MEDIAN AGE OF DIAGNOSIS **3.6** months

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

15% Pseudomonas in children · **51%** Pseudomonas in adults

93% Pancreatic enzymes of the people with CF in Ireland are using **pancreatic enzyme** supplementation.

F508del is the most common CF-causing mutation in Ireland

56% F508del homozygote · **36%** F508del heterozygote · **8%** Other mutations

Median BMI Z-score in children

Median BMI Z-Score · Age · 2-7 · 8-11 · 12-17 · Healthy children · Children with CF in Europe · Children with CF in Ireland

Median BMI in adults

Median BMI · Age · 18-21 · 22-27 · 28-31 · 32-37 · 38+ · Adults with CF in Europe · Adults with CF in Ireland

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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:

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

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

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%.

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.

Which data is collected?

- year/month of birth, gender
- genotype, symptoms at diagnosis
- lung function, weight, height, types of infections, treatment, some complications

The data is non-identifiable and stored in a secured database. We use well defined guidelines to handle the data, overseen by a committee of experts.

How can I become part of the ECFSPR?

Ireland has a national registry, and data is collected on a national level from all CF centres in the country. Each year a data-set of the registered CF patients in your country is sent to the ECFSPR to include in the database.

Check with your local CF centre. Your data should already be included, provided you signed a form to give your consent.

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