European Cystic Fibrosis Society Patient Registry

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Data source | Human | Disease registry

Administrative details

Administrative details

Data source ID

1000000014

Data source acronym

ECFSPR - Cystic Fibrosis

Data holder

European Cystic Fibrosis Society (ECFS)

Data source type

Disease registry

Main financial support

Funding by own institution

Care setting

Hospital inpatient care

Hospital outpatient care

Secondary care – specialist level (ambulatory)

Data source qualification

If the data source has successfully undergone a formal qualification process (e.g., from the EMA, ISO or other certifications), this should be described.

Yes

Description of the qualification

September 2018

EMA/CHMP/SAWP/622564/2018

Qualification Opinion on The European Cystic Fibrosis Society Patient Registry (ECFSPR) and CF Pharmaco-epidemiology Studies

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Data source website

www.ecfs.eu/ecfspr

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Data source regions and languages

Data source countries

Albania

Armenia

Austria

Belarus

Bulgaria

Croatia

Cyprus

Czechia

Denmark

Estonia

Finland

France

Georgia

Germany

Greece

Hungary

Iceland

Ireland

Israel

Italy

Latvia

Lithuania

Luxembourg

Moldova, Republic of

Montenegro

Netherlands

North Macedonia

Norway

Poland

Portugal

Romania

Russian Federation

Serbia

Slovakia

Slovenia

Spain

Sweden

Switzerland

Türkiye

Ukraine

United Kingdom

Data source languages

English

Data source establishment

Data source established

01/01/2003

Data source time span

First collection: 01/01/2003

The date when data started to be collected or extracted.

Publications

Data source publications

Effectiveness if lumacaftor/ivacaftor initiation in children with CF aged 2 through 5 years on disease progression: interim results from an ongoing registry-based study

Cystic fibrosis in Europe: improved lung function & longevity: reasons for cautious optimism, but challenges remain

A multinational report on SARS-COV-2 infection outcomes in people with CF and Aspergillus infection or ABPA

Nonsense mutations accelerate lung disease and decrease survival of cystic fibrosis children

Association of Oxygen Therapy with the Natural Disease Progression of Cystic Fibrosis: A Multi-State Model of the European Cystic Fibrosis Society Patient Registry

Risk factors for forced expiratory volume in 1 s decline in European patients with cystic fibrosis: data from the European Cystic Fibrosis Society Patient Registry

Clinical outcomes associated with Achromobacter species infection in people with cystic fibrosis

Cirrhosis and portal hypertension in cystic fibrosis in compound heterozygous people with harboring one F508del CFTR gene mutation

Prevalence, trends and outcomes of long-term inhaled antibiotic treatment in people with cystic fibrosis without chronic Pseudomonas aeruginosa infection – A European cystic fibrosis patient registry data analysis

Disease severity of people with cystic fibrosis carrying residual function mutations: Data from the ECFS Patient Registry

Geographic distribution and phenotype of European people with Cystic Fibrosis carrying A1006E mutation

Factors for severe outcomes following SARS-CoV-2 infection in people with cystic fibrosis in Europe

Incidence of SARS-CoV-2 in people with cystic fibrosis in Europe between February and June 2020

Survival estimates in European cystic fibrosis patients and the impact of socioeconomic factors: a retrospective registry Cohort study

Dornase alfa and rate of lung function decline in European patients with cystic fibrosis: a retrospective registry cohort study

The c.3140-26A>G Variant of the CFTR Gene in homozygous state causes mild cystic fibrosis Overview of longitudinal clinical data of the patient managed in our centre and review of the literature

Cystic Fibrosis Related Diabetes in Europe/ Prevalence, Risk Factors and Outcome

Changing Epidemiology of the respiratory bacteriology of patients with cystic fibrosis from the European Cystic Fibrosis Society Patient Registry

Characteristics of Cystic Fibrosis-related diabetes: Data from two different sources, the European Cystic Fibrosis Society Patient registry and German/Austrian diabetes prospective follow-up registry

Cystic fibrosis mortality in childhood. Data from European Cystic Fibrosis Society Patient Registry

Creating longitudinal datasets and cleaning existing data identifiers in a cystic fibrosis registry using a novel Bayesian probabilistic approach from astronomy

Effect of allergic bronchopulmonary aspergillosis on FEV1 in children and adolescents with cystic fibrosis: a European Cystic Fibrosis Society Patient Registry analysis

Year to year change in FEV1 in patients with cystic fibrosis and different mutation classes

Epidemiology of nontuberculous mycobacteria (NTM) amongst individuals with cystic fibrosis (CF)

International prospective study of distal intestinal obstruction syndrome in cystic fibrosis: Associated factors and outcome

Future trends in Cystic Fibrosis demography in 34 European countries

The relative frequency of CFTR mutation classes in European patients with cystic fibrosis

Multi-Country Estimate of Different Manifestations of Aspergillosis in Cystic Fibrosis

The European Cystic Fibrosis Society Patient Registry: valuable lessons learned on how to sustain a disease registry

Factors associated with FEV1 decline in cystic fibrosis: analysis of the data of the ECFS Patient Registry

Evidence of diminished FEV1 and FVC in 6-year-olds followed in the European cystic fibrosis patient registry, 2007-2009

A new era in the treatment of cystic fibrosis: correction of the underlying CFTR defect

Epidemiology of Cystic Fibrosis Lung Disease progression in adolescents

Reference percentiles for FEV(1) and BMI in European children and adults with cystic fibrosis

Cystic fibrosis across Europe: EuroCareCF analysis of demographic data from 35 countries

Comparative demographics of the European cystic fibrosis population: a cross-sectional database analysis

Publications section on the ECSF Patient Registry website

Data elements collected

The data source contains the following information

Disease information

Does the data source collect information with a focus on a specific disease? This might be a patient registry or other similar initiatives.

Yes

Disease details

Cystic fibrosis

Rare diseases

Are rare diseases captured? In the European Union a rare disease is one that affects no more than 5 people in 10,000.

Yes

Pregnancy and/or neonates

Does the data source collect information on pregnant women and/or neonatal subpopulation (under 28 days of age)?

Yes

Hospital admission and/or discharge

Yes

ICU admission

Is information on intensive care unit admission available?

No

Cause of death

Captured

Cause of death vocabulary

Other

Cause of death vocabulary, other

A prespecified list of causes for mortality

Prescriptions of medicines

Captured

Prescriptions vocabulary

not coded

Dispensing of medicines

Not Captured

Advanced therapy medicinal products (ATMP)

Is information on advanced therapy medicinal products included? A medicinal product for human use that is either a gene therapy medicinal product, a somatic cell therapy product or a tissue engineered products as defined in Regulation (EC) No 1394/2007 [Reg (EC) No 1394/2007 Art 1(1)].

No

Contraception

Is information on the use of any type of contraception (oral, injectable, devices etc.) available?

No

Indication for use

Does the data source capture information on the therapeutic indication for the use of medicinal products?

Not Captured

Medical devices

Is information on medicinal devices (e.g., pens, syringes, inhalers) available?

No

Administration of vaccines

No

Procedures

Does the data source capture information on procedures (e.g., diagnostic tests, therapeutic, surgical interventions)?

Captured

Procedures vocabulary

Other

Procedures vocabulary, other

Disease-specific diagnostic tests

Healthcare provider

Is information on the person providing healthcare (e.g., physician, pharmacist, specialist) available? The healthcare provider refers to individual health professionals or a health facility organisation licensed to provide health care diagnosis and treatment services including medication, surgery and medical devices.

No

Clinical measurements

Is information on clinical measurements (e.g., BMI, blood pressure, height) available?

Yes

Genetic data

Are data related to genotyping, genome sequencing available?

Captured

Genetic data vocabulary

HGVS

Biomarker data

Does the data source capture biomarker information? The term “biomarker” refers to a broad subcategory of medical signs (objective indications of medical state observed from outside the patient), which can be measured accurately and reproducibly. For example, haematological assays, infectious disease markers or metabolomic biomarkers.

Captured

Biomarker data vocabulary

Other

Biomarker vocabulary, other

Disease-specific list of biomarkers

Patient-reported outcomes

Is information on patient-reported outcomes (e.g., quality of life) available?

No

Patient-generated data

Is patient-generated information (e.g., from wearable devices) available?

No

Units of healthcare utilisation

Are units of healthcare utilisation (e.g., number of visits to GP per year, number of hospital days) available or can they be derived? Units of healthcare utilisation refer to the quantification of the use of services for the purpose of preventing or curing health problems.

Yes

Unique identifier for persons

Are patients uniquely identified in the data source?

Yes

Diagnostic codes

Not Captured

Medicinal product information

Not Captured

Quality of life measurements

Not Captured

Lifestyle factors

Not Captured

Sociodemographic information

Not Captured

Quantitative descriptors

Population Qualitative Data

Population age groups

Paediatric Population (< 18 years)

Preterm newborn infants (0 – 27 days)

Term newborn infants (0 – 27 days)

Infants and toddlers (28 days – 23 months)

Children (2 to < 12 years)

Adolescents (12 to < 18 years)

Adults (18 to < 65 years)

Adults (18 to < 46 years)

Adults (46 to < 65 years)

Elderly (≥ 65 years)

Adults (65 to < 75 years)

Adults (75 to < 85 years)

Adults (85 years and over)

Estimated percentage of the population covered by the data source in the catchment area

Over 80%

Description of the population covered by the data source in the catchment area whose data are not collected (e.g., people who are registered only for private care)

Regional subset - Coverage of the population is in most countries higher than 80%; in some countries coverage is below 80%. We are continuously working towards coverage of 80% or higher in all participating countries.

Population

Population size

54000

Active population size

51000

Data flows and management

Access and validation

Governance details

Documents or webpages that describe the overall governance of the data source and processes and procedures for data capture and management, data quality check and validation results (governing data access or utilisation for research purposes).

www.ecfs.eu/projects/efcs-patient-registry/guidelines

Biospecimen access

Are biospecimens available in the data source (e.g., tissue samples)?

No

Access to subject details

Can individual patients/practitioners/practices included in the data source be contacted?

No

Description of data collection

The data from consenting people with Cystic Fibrosis are collected on a centralised web-based platform called as the ECFSTracker.

Event triggering registration

Event triggering registration of a person in the data source

Disease diagnosis

Start of treatment

Event triggering de-registration of a person in the data source

Death

End of treatment

Loss to follow up

Event triggering creation of a record in the data source

The records are collected on the basis of the annual review.

Data source linkage

Linkage

Is the data source described created by the linkage of other data sources (prelinked data source) and/or can the data source be linked to other data source on an ad-hoc basis?

No

Data management specifications that apply for the data source

Data source refresh

Yearly

Informed consent for use of data for research

Required for all studies

Possibility of data validation

Can validity of the data in the data source be verified (e.g., access to original medical charts)?

Yes

Data source preservation

Are records preserved in the data source indefinitely?

Yes

Approval for publication

Is an approval needed for publishing the results of a study using the data source?

Yes

Data source last refresh

28/02/2024

Common Data Model (CDM) mapping

CDM mapping

Has the data source been converted (ETL-ed) to a common data model?

No