# **UK Cystic Fibrosis Registry**

**First published:** 01/02/2024

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Data source

Human

Disease registry

Other

# Administrative details

#### Administrative details

#### **Data source ID**

36763

#### **Data source acronym**

**UK CF Registry** 

#### **Data holder**

Cystic Fibrosis Trust

#### Data source type

Disease registry

Other

#### Data source type, other

Prospective studies database, pharmacovigillance Database

#### Main financial support

Funding by own institution

Funding from industry or contract research

#### **Care setting**

Hospital inpatient care

Hospital outpatient care

Primary care - specialist level (e.g. paediatricians)

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.

No

#### **Data source website**

https://www.cysticfibrosis.org.uk/the-work-we-do/uk-cf-registry

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# Data source regions and languages

#### **Data source countries**

**United Kingdom** 

#### **Data source languages**

English

#### Data source establishment

#### **Data source established**

01/01/1995

#### Data source time span

First collection: 01/01/1995

The date when data started to be collected or extracted.

### **Publications**

# Data source publications

Up-to-date and projected estimates of survival for people with cystic fibrosis using baseline characteristics: A longitudinal study using UK patient registry data

A multinational report to characterise SARS-CoV-2 infection in people with cystic fibrosis

Use of a rare disease patient registry in long-term post-authorisation drug studies: a model for collaboration with industry

Data Resource Profile: The UK Cystic Fibrosis Registry

## **Studies**

# List of studies that have been conducted using the data source

Survey on the collection of data on adverse events related to medicinal products through registries

# 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

ICU	20	lmı	CCI	n

Is information on intensive care unit admission available?

Yes

#### Cause of death

Captured

#### Cause of death vocabulary

Other

#### Cause of death vocabulary, other

Bespoke list of variables

#### **Prescriptions of medicines**

Captured

#### **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?

Yes

#### 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?

Yes

#### 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

Bespoke written Statements of Practice

#### **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.

Yes

#### **Clinical measurements**

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

#### **Genetic data**

Are data related to genotyping, genome sequencing available?

Captured

#### **Genetic data vocabulary**

Other

#### Genetic data vocabulary, other

Use of CFTR2

#### 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

NHS vocabulary

#### **Patient-reported outcomes**

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

Yes

#### **Patient-generated data**

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

#### 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.

No

#### **Unique identifier for persons**

Are patients uniquely identified in the data source?

Yes

#### **Diagnostic codes**

Captured

#### Diagnosis / medical event vocabulary

Other

#### Diagnosis / medical event vocabulary, other

**NHS** variables

#### **Medicinal product information**

Captured

#### Medicinal product information collected

Brand name

Dosage regime

Dose

#### **Medicinal product vocabulary**

Other

# If 'other,' what vocabulary is used? **British National Formulary Quality of life measurements** Captured **Quality of life measurements vocabulary** other Quality of life measurements, other **CFQR** Lifestyle factors Captured **Lifestyle factors** Other Tobacco use Lifestyle factors included other Vaping data **Sociodemographic information** Captured Sociodemographic information collected Age Education level **Ethnicity** Gender Marital status

#### 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 < 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

99% of the population covered. Population age stratification can be found on Figure 1.2 in the UK CF Registry 2022 National report (https://www.cysticfibrosis.org.uk/about-us/uk-cf-registry/reporting-and-resources)

# 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)

It is not recorded how many patients refuse consent to the registry. It is thought not many as the specialist centres ensure all patients are registered.

# **Population**

#### **Population size**

11148

#### **Active population size**

11148

# 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).

https://www.cysticfibrosis.org.uk/about-us/uk-cf-registry

#### **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**

Patient demographic form must be completed. Encounters are entered and an annual review form must be completed once a year.

# Event triggering registration

#### Event triggering registration of a person in the data source

Disease diagnosis

Practice registration

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

Death

**Emigration** 

Practice deregistration

#### Event triggering creation of a record in the data source

NHS patient visit. Patients have 1 annual visit and encounters entered each year.

# 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

Other

#### 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?

No

#### Informed consent, other

There is a formal review process overseen by the UK CF Registry Research Committee to evaluate requests for data access

#### **Data source last refresh**

31/01/2024

# Common Data Model (CDM) mapping

#### **CDM** mapping

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

No