UK Cystic Fibrosis Registry

First published: 01/02/2024

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Data source Human Disease registry Other

Administrative details

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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

Yes

ICU admission

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?

Yes

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?

Yes

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

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 < 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-andresources)

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