# The European Clarkson’s syndrome registry

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

**Last updated:** 17/10/2024

**Data source** **Human** **Disease registry** **Other**

## Administrative details

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**Data source ID**

24877

**Data source acronym**

EurêClark

**Data holder**

Institut E3M

**Data source type**

Disease registry

Other

**Data source type, other**

exposure registry

**Care setting**

Hospital inpatient care

Hospital outpatient care

Secondary care – specialist level (ambulatory)

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**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

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**Data source website**

http://institut-e3m.aphp.fr/le-service-de-medecine-interne/

## Contact details

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**Main**

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## Data source regions and languages

**Data source countries**

Austria

Belgium

Estonia

France

Germany

Italy

Spain

Switzerland

United Kingdom

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**Data source languages**

English

French

## Data source establishment

**Data source established**

01/01/1997

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**Data source time span**

**First collection:** 01/01/1997

The date when data started to be collected or extracted.

# Publications

## Data source publications

Intravenous Immunoglobulins Improve Survival in Monoclonal Gammopathy-Associated Systemic Capillary-Leak Syndrome

Myocardial dysfunction is frequent in systemic capillary-leak syndrome (Clarkson disease) severe episodes

The consequences of COVID-19 pandemic on patients with monoclonal gammopathy-associated systemic capillary leak syndrome (Clarkson disease)

Intravenous Immunoglobulins Tapering and Withdrawal in Systemic Capillary Leak Syndrome (Clarkson Disease)

The Clinical Picture of Severe Systemic Capillary-Leak Syndrome Episodes Requiring ICU Admission

# 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

**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)?

No

**Hospital admission and/or discharge**

Yes

**ICU admission**

Is information on intensive care unit admission available?

Yes

**Cause of death**

Captured

**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?

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

Yes

## Procedures

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

Not Captured

## 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?

Not Captured

## 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.

Not Captured

### 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.

No

### Unique identifier for persons

Are patients uniquely identified in the data source?

No

### Diagnostic codes

Captured

### Medicinal product information

Not Captured

### Quality of life measurements

Not Captured

**Lifestyle factors**

Captured

**Lifestyle factors**

Alcohol use

Diet

Tobacco use

**Sociodemographic information**

Captured

**Sociodemographic information collected**

Age

Gender

# 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)

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

<0.1%

## Population

**Population size**

90

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**Active population size**

40

## Population by age group

| Age group | Population size | Active population size |
|---|---|---|
| Paediatric Population (< 18 years) | 10 | 2 |
| Adults (18 to < 46 years) | 10 | 5 |
| Adults (46 to < 65 years) | 50 | 23 |
| Adults (65 to < 75 years) | 10 | 5 |
| Adults (75 to < 85 years) | 10 | 5 |

## Median observation time

**Median time (years) between first and last available records for unique individuals captured in the data source**

15.00

**Median time (years) between first and last available records for unique active individuals (alive and currently registered) capt**

10.00

# Data flows and management

## Access and validation

**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?

Yes

**Description of data collection**

Medical reports

## Event triggering registration

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

Disease diagnosis

Start of treatment

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

Any relevant medical event

# 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

Every 6 months

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### Informed consent for use of data for research

Other

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### Possibility of data validation

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

Yes

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### Data source preservation

Are records preserved in the data source indefinitely?

No

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**Approval for publication**

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

Yes

**Informed consent, other**

There is a committee to evaluate requests for data access

**Data source last refresh**

01/01/2023

## Common Data Model (CDM) mapping

**CDM mapping**

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

No