Welcome to the COVID-CF project in Europe

CF patient registries throughout Europe have collected data about people with CF who become infected with SARS-Cov-2, causing the illness COVID-19.

Countries that contribute annual data to the ECFS Patient Registry (ECFSPR; www.ecfs.eu/ecfspm) were invited to report COVID-19 case data of people with a confirmed diagnosis of CF. Centres reported data directly to the ECFSPR, and aggregated data was provided by national registries that use their own data-collection system. Here we present centralised, anonymised data, which we hope to update weekly.

It is possible that not all cases have been reported yet. Since the data is preliminary, incomplete, might change over time, and the number of cases is low, the information should not be used to direct clinical decisions. A more substantial statistical analysis will be performed and published at a later date.

Definitions are provided for all the variables measured at the end of this report.

Summary (up to 10 September 2020)

Important update: The ECFSPR database was cleaned in early July to remove patients who did not have a PCR-confirmed diagnosis of infection with SARS-Cov-2. This is because serological antibody tests lack specificity for the SARS-Cov-2 virus compared to other coronaviruses circulating in the population. Aggregate data from national registries may still include patients who have only had an antibody test (without PCR confirmation).

<table>
<thead>
<tr>
<th>People with CF have confirmed COVID-19</th>
<th>Case data available</th>
<th>Needed intensive care</th>
<th>Died</th>
</tr>
</thead>
<tbody>
<tr>
<td>162</td>
<td>144</td>
<td>8</td>
<td>3</td>
</tr>
</tbody>
</table>

- 5 critical cases
- 3 people with CF and COVID-19 died
- The most common treatments were azithromycin and additional antibiotics (oral and intravenous)
Distribution in Europe

The following flow chart presents the number of countries that reported COVID-19 cases in people with CF by 10 September 2020.

38 countries invited to contribute information

38 countries provided information

18 countries reported 162 known cases

18 countries reported zero known cases

Information available for 144 cases

No further information available for 18 cases

COVID-19 in people with CF

Data up to 10 September 2020

Countries in grey did not submit any information

Created with Datawrapper
Summary data for the 144 cases with information

The rest of this report presents data for the 144 reported cases with at least partial data. If case reports were incomplete, the missing information has not been considered.

Demographics

Gender

![Pie chart showing gender distribution: 58 females and 63 males.]

*Data were not available in this format for 23 patients

Age category (years)

![Bar chart showing age distribution: 16 patients aged 0-11, 20 patients aged 12-17, 37 patients aged 18-29, 41 patients aged 30-49, 5 patients aged 50+.

*Data were not available in this format for 25 patients
Cystic fibrosis characteristics
Percent predicted FEV$_1$, by category

**Number of patients (n=104*)**

<table>
<thead>
<tr>
<th>Category</th>
<th>Number</th>
</tr>
</thead>
<tbody>
<tr>
<td>&gt;70%</td>
<td>51</td>
</tr>
<tr>
<td>41-70%</td>
<td>31</td>
</tr>
<tr>
<td>&lt;40%</td>
<td>22</td>
</tr>
</tbody>
</table>

Abbreviation: ppFEV$_1$ = percent predicted forced expiratory volume in one second
*Data were not available in this format for 35 patients. ppFEV$_1$ measurement was not applicable for a further 5 patients.

Other cystic fibrosis characteristics

**Number of patients (n=123*)**

Patients could have between zero and three of these characteristics

<table>
<thead>
<tr>
<th>Condition</th>
<th>Number</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pseudomonas aeruginosa**</td>
<td>41</td>
</tr>
<tr>
<td>CF related diabetes</td>
<td>38</td>
</tr>
<tr>
<td>Transplant</td>
<td>19</td>
</tr>
</tbody>
</table>

*Data were not available in this format for 21 patients
** in the last 12 months (data for Pseudomonas infection were not available in this format for a further 27 patients).
COVID-19 symptoms

Categories of symptoms

Number of patients (n=87*)
Patients could have symptoms in more than one category

- Fever / General: 66
- Pulmonary: 57
- Asymptomatic: 25
- Gastrointestinal: 16
- Other: 4
- Respiratory failure: 4

*Data were not available in this format for 57 patients
Patients could have more than one symptom.

Categories of symptoms:
- Pulmonary symptoms: increased cough, dyspnoea, chest tightness, wheezing, sputum production, haemoptysis
- Fever / General symptoms: fever, fatigue, headache, arthralgia/myalgia
- Other: none of the above
- Asymptomatic: none of the symptoms reported
- Gastrointestinal symptoms: diarrhoea, vomiting/nausea, abdominal pain
- Respiratory failure: respiratory failure
Individual symptoms

Number of patients (n=83*)
Patients could have more than one symptom

- Fever: 54
- Increased cough: 39
- Fatigue: 28
- Increased sputum: 26
- Headache: 24
- Increased dyspnoea: 22
- Pulmonary exacerbation: 17
- Pharyngitis: 16
- Muscle/joint pain: 16
- Acute rhinitis: 15
- Chest tightness: 11
- Vomiting/Nausea: 9
- Diarrhoea: 9
- Loss of taste: 8
- Loss of smell: 7
- Abdominal pain: 5
- Wheezing: 4
- Haemoptysis: 4
- Arterial Hypotension: 4
- Respiratory failure: 3
- Conjunctivitis: 2

*Data were not available in this format for 61 patients
COVID-19 severity

Number of patients (n=104*)

<table>
<thead>
<tr>
<th>Severity</th>
<th>Count</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mild</td>
<td>93</td>
</tr>
<tr>
<td>Severe</td>
<td>6</td>
</tr>
<tr>
<td>Critical</td>
<td>5</td>
</tr>
</tbody>
</table>

*Data were not available in this format for 40 patients

Mild: Patients without pneumonia or cases of mild pneumonia

Severe: Patients who suffered from shortness of breath, respiratory frequency ≥ 30/minute, blood oxygen saturation ≤93%, PaO2/FiO2 ratio <300, and/or lung infiltrates >50% within 24–48 hours

Critical: Patients who suffered respiratory failure, septic shock, and/or multiple organ dysfunction or failure.

COVID-19 treatment

Place of care

73 patients were hospitalised

8 hospitalised patients needed ICU care
Pharmacological treatment

Number of patients (n=94*)
Patients could receive more than one treatment

<table>
<thead>
<tr>
<th>Treatment</th>
<th>Number of Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Azithromycin</td>
<td>28</td>
</tr>
<tr>
<td>Additional oral antibiotics</td>
<td>28</td>
</tr>
<tr>
<td>Additional IV antibiotics</td>
<td>23</td>
</tr>
<tr>
<td>(Hydroxy) chloroquine</td>
<td>15</td>
</tr>
<tr>
<td>Systemic steroids</td>
<td>10</td>
</tr>
<tr>
<td>Antiviral</td>
<td>7</td>
</tr>
<tr>
<td>Antifungal</td>
<td>5</td>
</tr>
<tr>
<td>Tocilizumab</td>
<td>3</td>
</tr>
<tr>
<td>Vitamin C</td>
<td></td>
</tr>
<tr>
<td>Anakinra</td>
<td></td>
</tr>
</tbody>
</table>

*Data were not available in this format for 50 patients

Oxygen and respiratory support

<table>
<thead>
<tr>
<th>Treatment</th>
<th>Number of Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Oxygen therapy needed</td>
<td>29</td>
</tr>
<tr>
<td>Respiratory support needed*</td>
<td>7</td>
</tr>
<tr>
<td>Non-invasive ventilation (BIPAP, CPAP)</td>
<td>2</td>
</tr>
<tr>
<td>High flow nasal canula oxygen therapy</td>
<td>4</td>
</tr>
<tr>
<td>Invasive Ventilation</td>
<td>3</td>
</tr>
<tr>
<td>ECMO</td>
<td>1</td>
</tr>
</tbody>
</table>

Abbreviations: BIPAP = bi-level positive airway pressure, CPAP = continuous positive airway pressure, ECMO = extra corporeal membrane oxygenation

* In certain cases, patients can have more than one type of respiratory support (e.g. ECMO and invasive ventilation). Therefore, the numbers of patients who needed each type of respiratory support may exceed the total number of patients who needed respiratory support.
COVID-19 complications

Number of patients (n=83*)
Patients could have more than one complication

- CF pulmonary exacerbation: 15
- Bacterial pneumonia: 6
- Sepsis: 4
- ARDS: 3
- Other: 3
- Renal insufficiency: 3
- Multi-organ failure: 2
- Pleural effusion: 1

*Data were not available in this format for 61 patients
ARDS = acute respiratory distress syndrome

COVID-19 outcomes

Number of patients (n=144)

- Recovered*: 115
- Not answered: 13
- Unknown: 7
- Illness ongoing: 6
- Died: 3

*or discharged alive from hospital
Footnotes and references

For data submitted directly to ECFSPR, the following references were used for computation of ppFEV1.

- Percent predicted FEV₁ was calculated using the calculator http://gligastransfer.org.au/calcs/spiro.html based on the last 3 FEV₁ (pre-bronchodilator) measurements before infection with SARS-Cov-2.

Icons are from:

- Home by Kahalap from the Noun Project
- Hospital by Made from the Noun Project
- Medical treatment by visual world from the Noun Project