Cystic Fibrosis strength in numbers

UK CF Registry Annual Data Report 2018 - at-a-glance

Our at-a-glance version of the UK CF Registry Annual Data Report 2018 highlights the key information from the full report, available at cysticfibrosis.org.uk/registryreports

Active patients
10,509
Active patients are people who are currently alive and have had an annual review recorded in the last three years.

Median age
20
is the median age of people with cystic fibrosis in the UK.

Mode of presentation
2,751
(27.9%) people were diagnosed by newborn screening. Aside from newborn screening, the most common three presentations were:

- Abnormal stools/fatty stool (steatorrhea)/malabsorption
- Failure to thrive/ malnutrition
- Persistent or acute respiratory infection

Diagnosis
23 days
is the median age that people aged under 16 in 2018 were diagnosed with cystic fibrosis. 8.5% of people in the Registry were diagnosed at age 16 or over.

CF population by devolved nation

Infections

Pseudomonas aeruginosa
41.4%
of people aged 16 and over have chronic Pseudomonas.
The median age of people with chronic Pseudomonas was 24 years in 2008, compared to 29 in 2018. 88.9% of people with chronic Pseudomonas were on inhaled antibiotic therapy in 2018, compared to 76.1% in 2008.

Non-tuberculous mycobacterium (NTM)

has increased from 6% to 7% in the past year.

49%
of people recorded as having NTM were being treated, which is 10% less than in 2017.

Aspergillus is reported in
8.9%
of people.

Allergic bronchopulmonary aspergillosis (ABPA), an immune response to Aspergillus infection, has reduced in prevalence by 10.5% since 2003, to 7.2%.
Median predicted survival age

for people born today, using “2014-2018” data, is 47.3 years old.

The median predicted survival age for females (44.1) is 7 years lower than males (51.0).

Deaths in 2018

Of the 137 people with CF who died in 2018, the median age of death was 32 years old.

Mucus thinners

32.9% of people were on hypertonic saline in 2018, compared to 5% in 2008.

65% of people were on DNase in 2018, compared to 37% in 2008.

IV antibiotics

44.7% of people had at least one course of IV antibiotics in 2018.

37.7% in hospital

24% at home

Burden of treatment

79.4% of people with CF were on at least one form of inhaled therapy.

No inhaled therapy: 2031 (20.6%)

Cystic fibrosis-related diabetes (CFRD)

On CFRD treatment: 33.8%

Screened 53.1%

Existing CFRD diagnosis 26.6%

Not screened 19.4%

Unknown 0.9%

People on CFTR modifiers

Ivacaftor: 612

Lumacaftor/ivacaftor: 372

Tezacaftor/ivacaftor: 30

Transplant

<table>
<thead>
<tr>
<th>Year</th>
<th>Evaluated</th>
<th>Accepted</th>
<th>Double lung</th>
</tr>
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<tbody>
<tr>
<td>2008</td>
<td>126</td>
<td>55</td>
<td>16</td>
</tr>
<tr>
<td>2018</td>
<td>247</td>
<td>104</td>
<td>58</td>
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