



## CF care in the Netherlands

### a short overview

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# Organization of care

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- Multidisciplinary care/ dedicated CF teams
- Guidelines/ Treatment protocol (diagnostics and therapy: national Consensus 2018)
- 7CF Centers (children and Adults)
- Newborn screening since 2011
- (IV) homecare
- E-Health
- NCFS (Dutch CF foundation)
- Dutch CF registry
- CF Research Network
- Dutch CF trial consortium
- International CF Trial consortium (HIT CF Europe, rainbow study)
- CF (nursing) network

# Statistics \*

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## 1600 CF patiënts

- 560 children
- 1040 adults
- 133 > 50 years
- Aprox. 29 babies born with CF every year (registry 2020)
- Average life expectancy 50, changing/ improving due to improved care, therapy and new medication (CF modulators)

\*NCFS, May 2022

**Den Haag: (2020)**  
53 Children  
2 Newborn screening  
197 Adults

**Amsterdam:**  
60 Children  
7/year newborn screening  
140 Adults

**Utrecht:**  
140 Children  
5/6 year newborn screening  
285 Adults

**Rotterdam:**  
143 Children  
6/ year newborn screening  
180 Adults

**Nijmegen:**  
90 Children  
7-8/ year newborn screening  
67 Adults

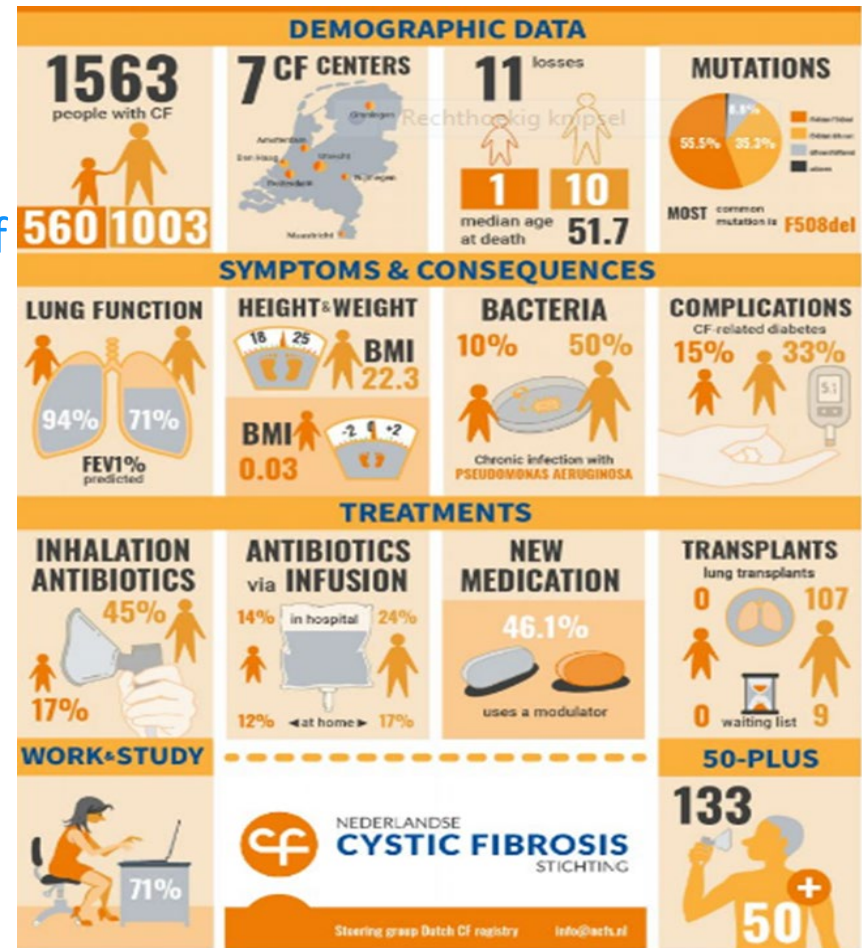
**Maastricht:**  
29 Children  
2/ year newborn screening  
57 Adults

**Groningen:**  
65 Children  
2-3/ year newborn screening  
109 Adults

# Dutch CF registry\*

This report provides information and trends on many aspects of CF diagnostics and treatment, complications of CF and some aspects of living with CF.

It contains anonymized data, collected, analyzed and published each year on the bases of data from the CF- Centers



\* NCFS, Data 2020, Report from 2021 is not yet published

# Adults with CF

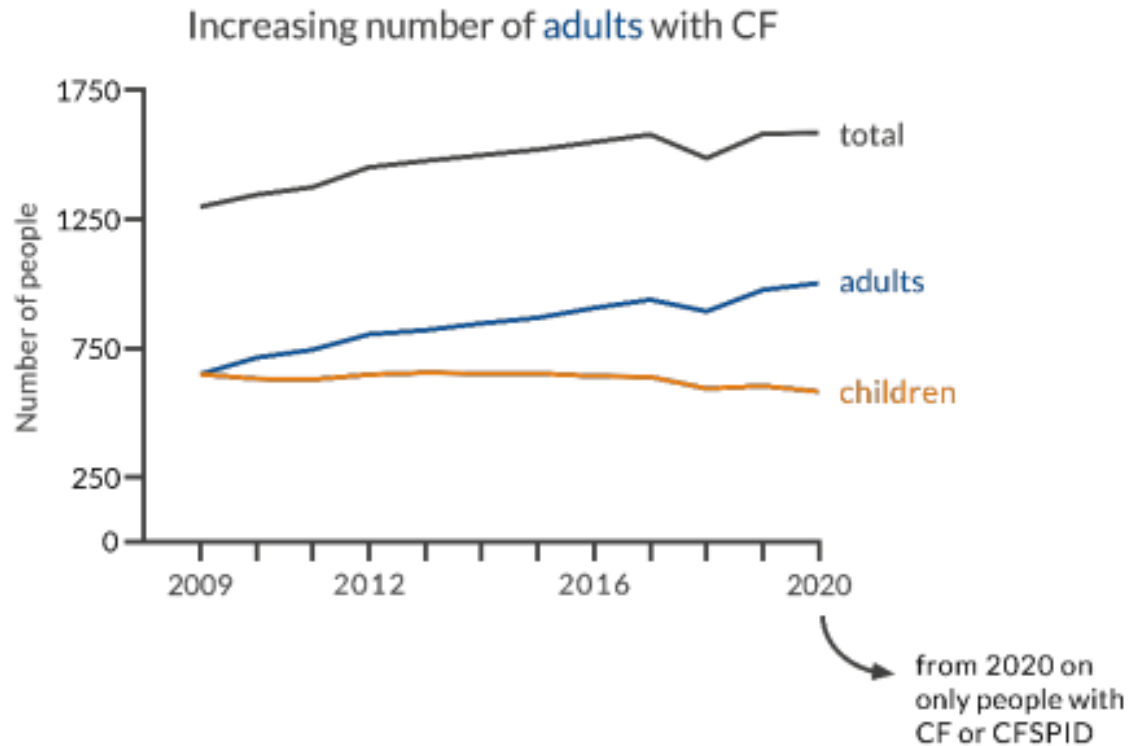
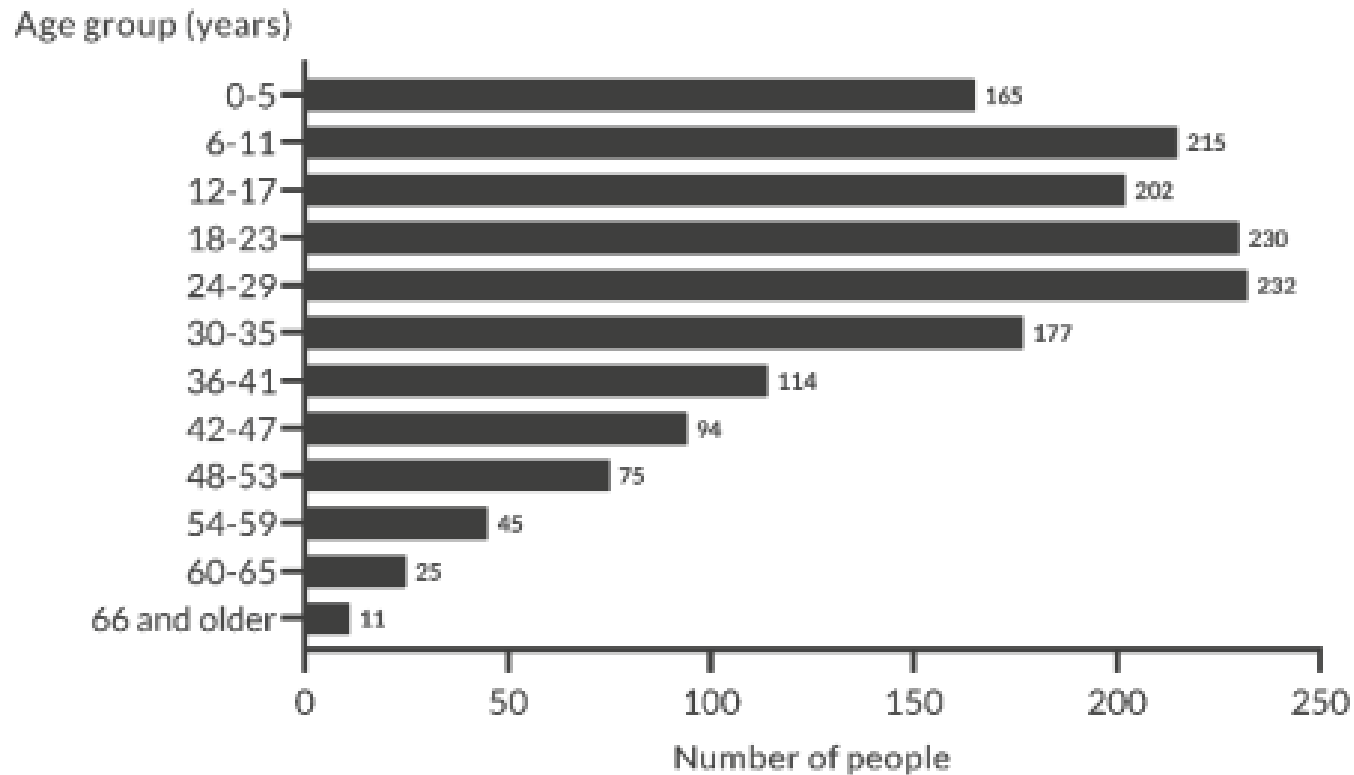


Figure 1. Number of people with CF, CFSPID\* or CF-related disease in the Dutch CF Registry, for the period 2009–2020.  
\*CFSPID: positive newborn screening, no confirmed CF diagnosis.

# CF patients/ Age

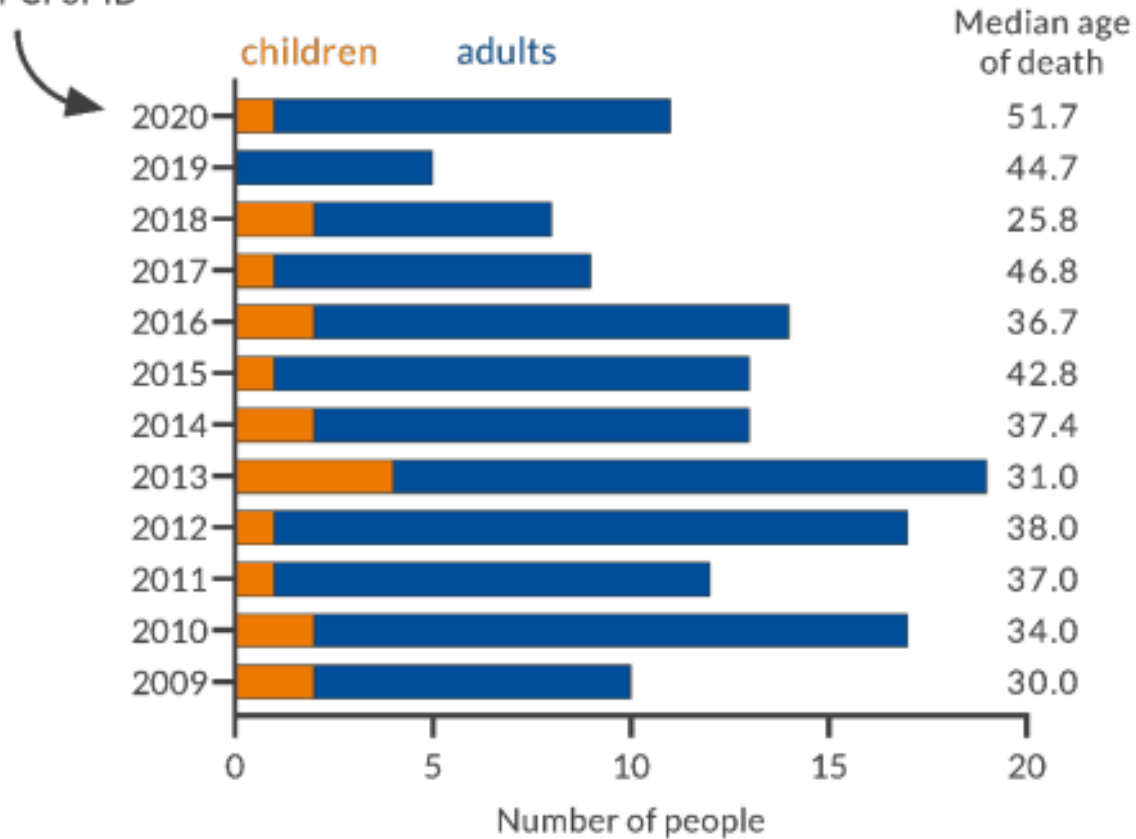
Predominantly young adults with CF in 2020



# Median age

## Median age of death keeps going up

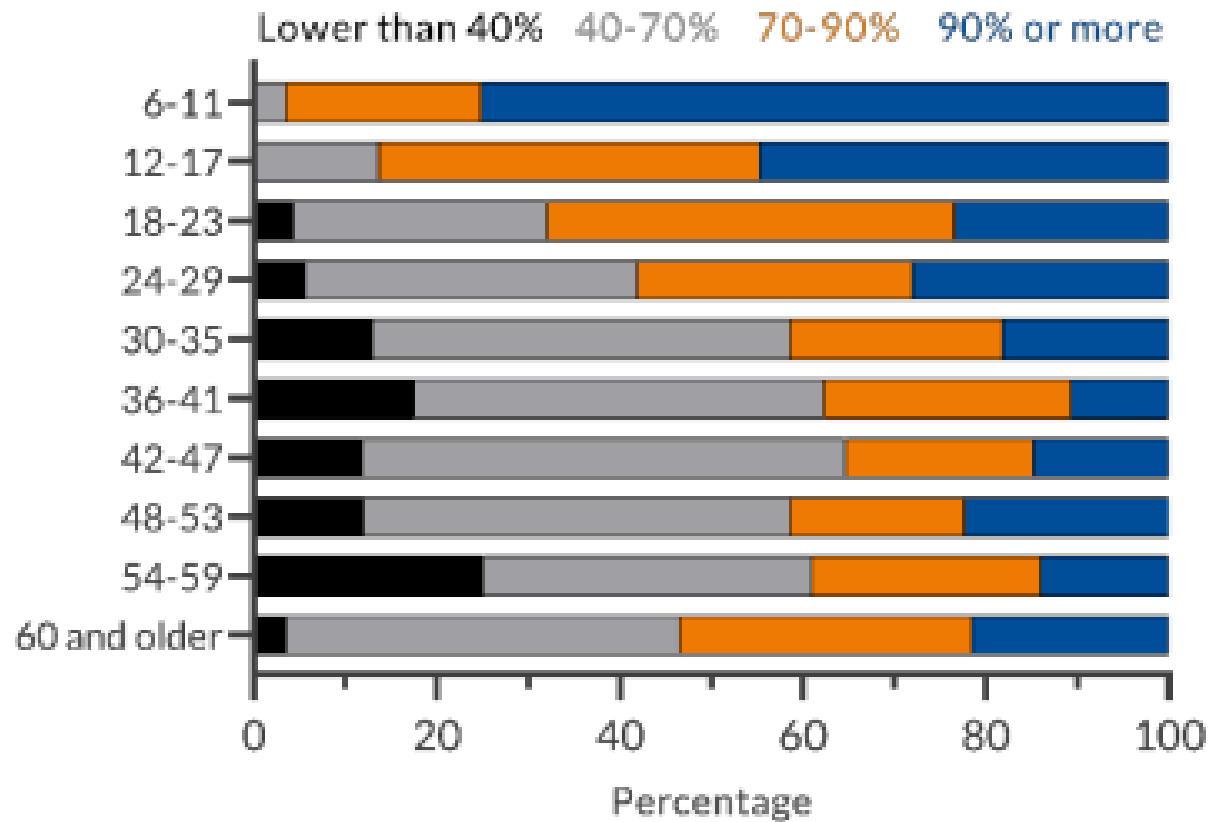
from 2020 on  
only people with  
CF or CFSPID





# Lungfunction by age group

Pulmonary function category by age group



# Lungfunctie

At most centres, at least half of the children has a **healthy pulmonary function**

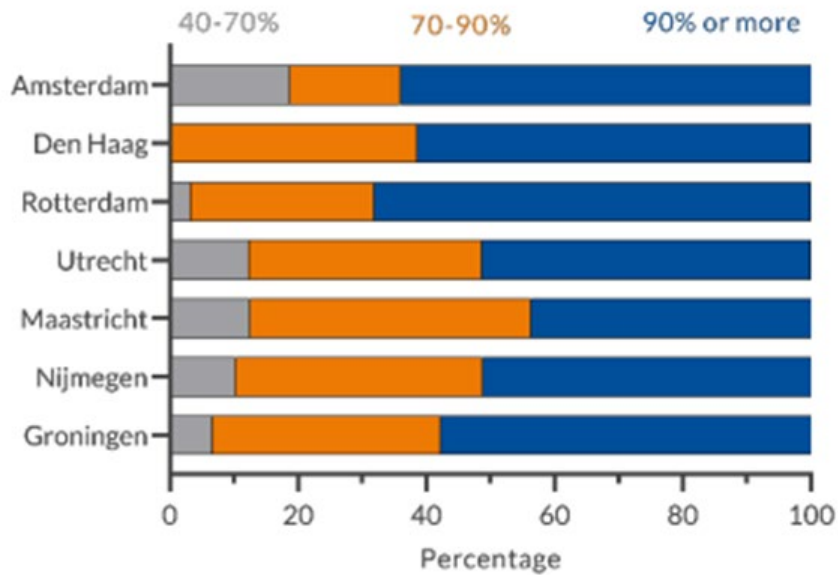


Figure 27. Distribution of pulmonary function in children with CF by CF centre.

At most centres, most adults have a pulmonary function of 40-70%

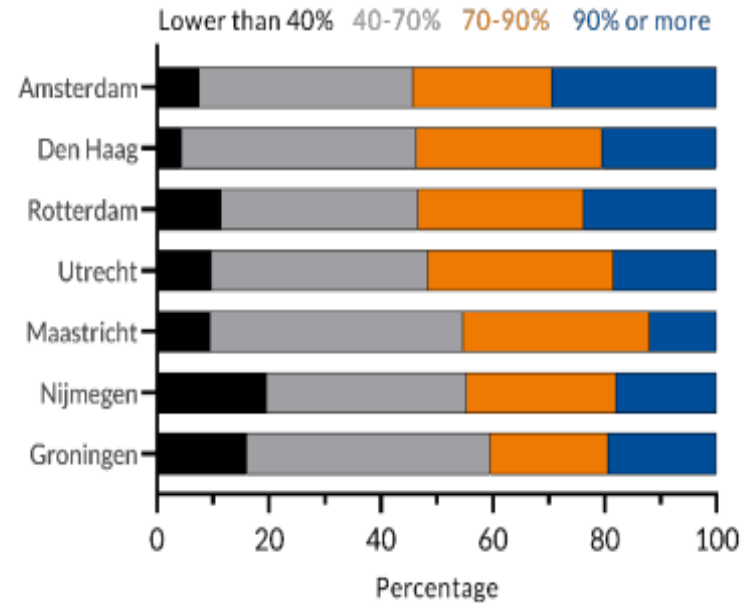


Figure 29. Distribution of pulmonary function in adults with CF by CF centre.

# Mutations

## a. Distribution of mutations in people with CF

Most people with CF in the Netherlands have two F508del mutations (Figure 10). Overall, 90.8% have at least one F508del mutation. A handful of people never had their mutations analysed.

Most people with CF have one or two F508del mutations

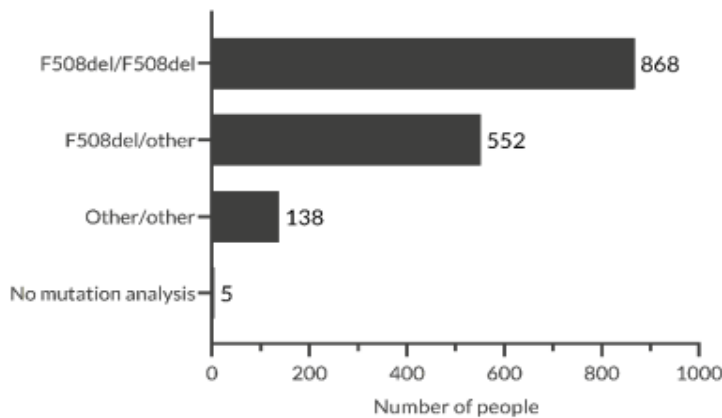


Table 2. The fifteen most common mutations in people with CF and CFSPID in the Netherlands in 2020. Together, 1,585 people have 3,170 mutations.

Mutation	Number of people	Percentage (%)
<i>F508del</i>	2,306	72.7
<i>A455E</i>	123	3.9
<i>R117H</i>	59	1.9
<i>G542X</i>	57	1.8
<i>1717-1G-&gt;A</i>	43	1.4
<i>3272-26A-&gt;G</i>	39	1.2
<i>S1251N</i>	38	1.2
<i>N1303K</i>	36	1.1
<i>R553X</i>	31	1.0
<i>R1162X</i>	30	0.9
<i>2789+5G-&gt;A</i>	24	0.8
<i>3849+10kbC-&gt;T</i>	18	0.6
<i>E60X</i>	18	0.6
<i>711+1G-&gt;T</i>	16	0.5
<i>W1282X</i>	13	0.4
<i>Unknown</i>	23	0.7
<i>Mutations not analysed</i>	10	0.3
<i>Other</i>	286	9.0
<b>Total number of mutations</b>	<b>3,170</b>	<b>100.0</b>

# Standard of care/ update

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## 1. CF modulators available in the Netherlands

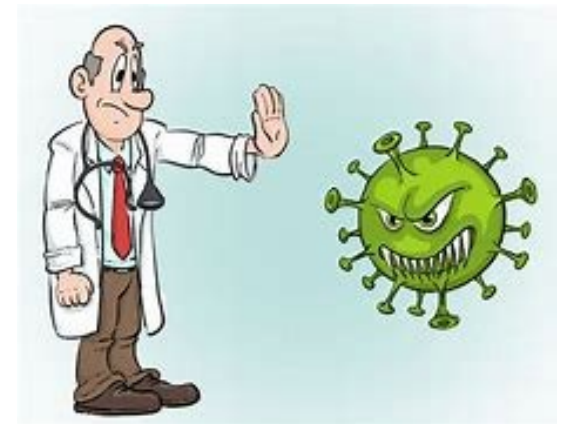
- Kalydeco 2015
- Orkambi 2018
- Symkevi 2020
- Kaftrio 2022

## 2. Coronavirus Sars-CoV-2

## 3. E-health

# Coronavirus Sars-CoV-2 and CF

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The CF Registry reported

- 51 people with CF tested positive for Coronavirus in 2020.  
This is probably an underestimation
- In 2021 10% have tested positive  
(Based on data from the CF centers (65%))
- In the first months of 2022 this increased to 25%  
(40% of the centers provided data)

Data from Wilhelmina Childrens Hospital

- 44 children reported positive (1x2020, others 2021/22 till now)
- All had mild symptoms, 1 hospital admission (CF exacerbation and Covid positive)

# Coronavirus Sars-CoV-2 and CF

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## Impact on care:

- Less face to face appointments, some appointments were canceled and rescheduled, some tests were postponed (if possible) and most of the treatments were done at home
- (iv homecare, as little as possible hospitalizations)

## Furthermore:

- Less adults received IV treatment
- Less Influenza in patients with CF
- CF centers continued most of the standard care

SO: Standard of care ok?

\* Info NCFS

# E health

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Already broadly used before Covid 19  
Next to or instead of a visit to CF  
Center

- Longfunction test at home e-  
portaal/ apps
- Video-consulting
- Telephone contacts
- Beveiligde mail
- Apps
- E-portal in patientfile > e-consulting  
(mails)
- online questionnaires (in patient  
file)
- Teams meetings/ online  
information meetings (new  
medication)

# Round up

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