

ALLM

Assemblée Générale 2021

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Cabinet de Pneumologie du Glacis

Service de Pneumologie HRS – HK /ZK

Nouveautés 2020

- **Rapport ECF – PR / 2018**
- Simeox
- Aiocare
- Symkevi, Kaftrio,
- CF et COVID Luxembourg et Europe
- Problèmes – registre 2019 (année transition) et reprise 2020

2018

ECFS Patient Registry Annual Data Report

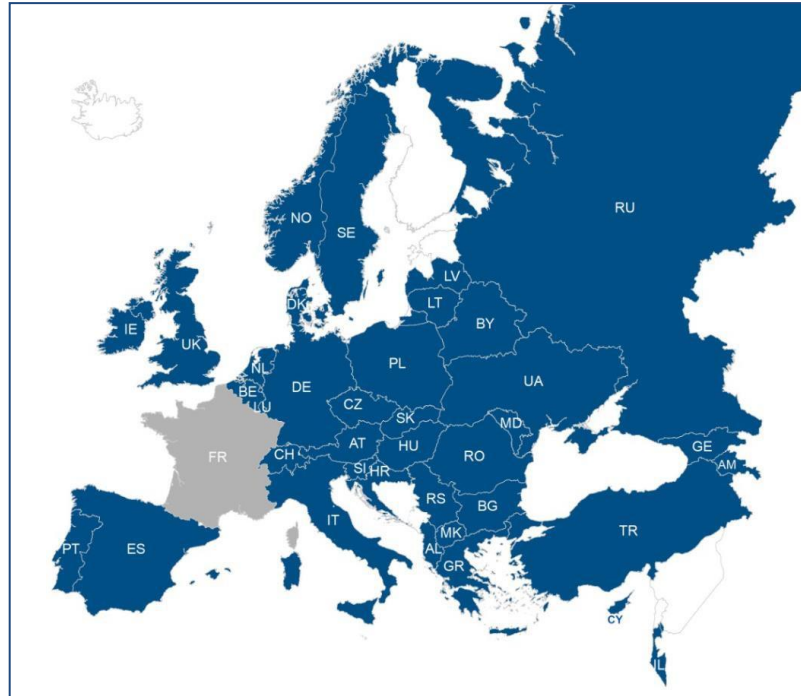


European Cystic Fibrosis Society
Kastanieparken 7
7470 Karup
Denmark
www.ecfs.eu/ecfspr

Data report

1. Demographics

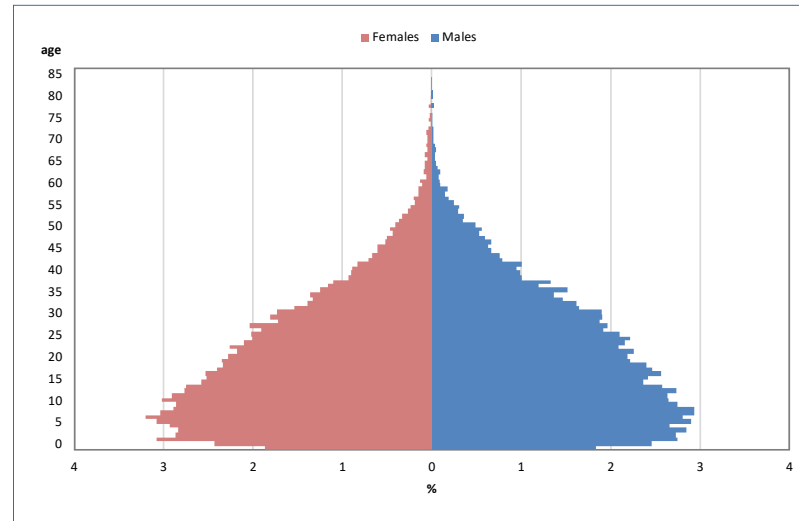
Figure 1.1 Map of countries that contributed to the ECFSPR in year 2018.



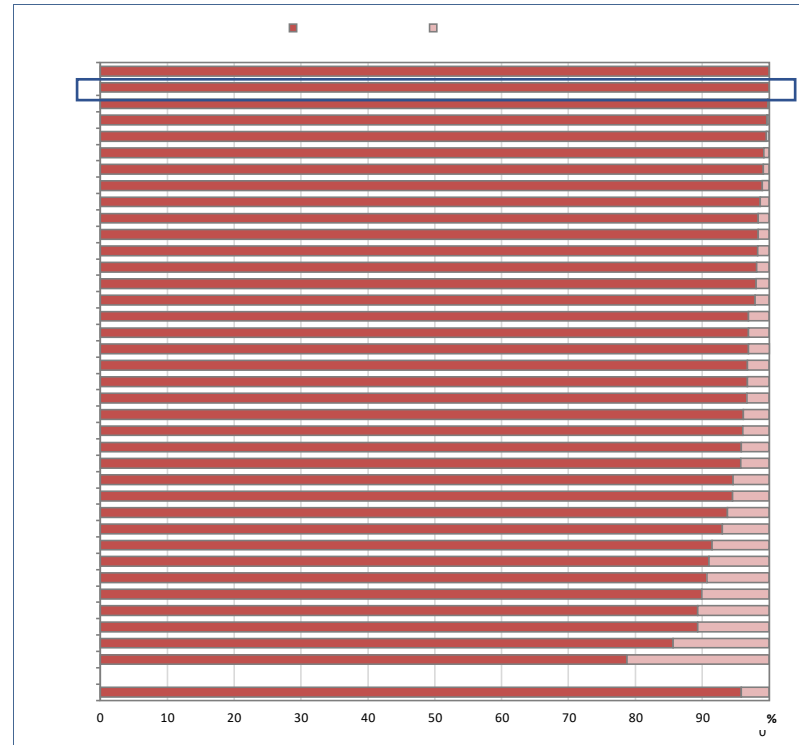
Countries that contributed 2018 data are marked in blue.

France is in grey, since no 2018 data have been reported due to the COVID-19 pandemic in 2020. In this chapter, 2017 data are used for France.

Figure 1.4 Age at follow-up distribution by sex. Patients alive on 31/12/2018.

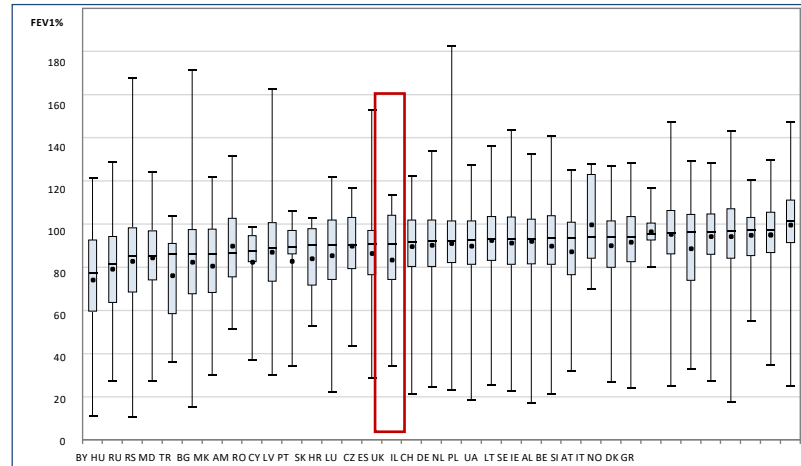


The pyramid shows the percentage of patients of different ages as horizontal bars. The right side of the pyramid (blue) shows, for males, how many patients (as a percentage) are a certain age, the left side (red) shows the same for females. The lower percentage of patients at the bottom of the pyramid is a result of the fact that some patients have not yet been diagnosed (mean age at diagnosis is 4.0 years, see table 2.1).



This graph shows the percentage of mutations that are not identified (unknown in light pink) after DNA analysis, by country and overall. One “allele” means one of the two CFTR genes. The number of non-identified alleles varies greatly from country to country; this is partly due to the different approaches to DNA testing. Overall, more than 4% of mutations remain unidentified after DNA analysis, leaving 6.47% of the patients with at least one mutation unidentified.

Figure 4.1 FEV₁% of predicted: box-plot, by country and overall. Patients aged 6-17 years who have never had a transplant.

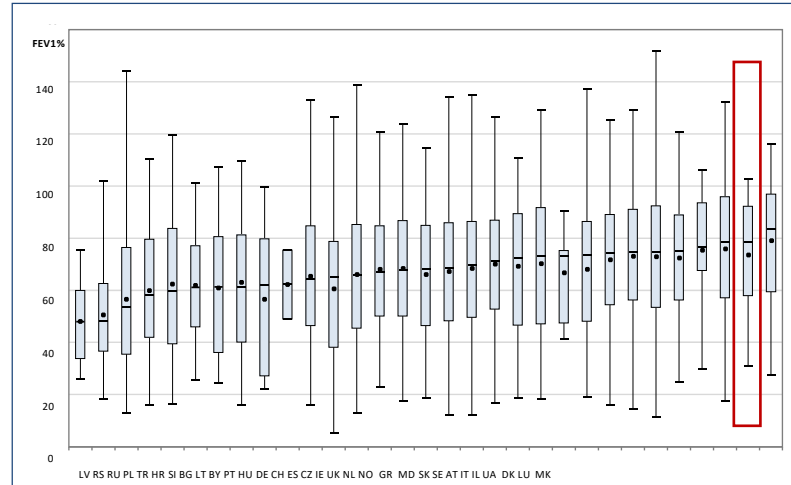


Note: Georgia has <5 patients aged 6-17 years at FEV₁ measurement and is excluded from this graph.

Note: The UK reports best FEV₁ from the annual review, which is the time period between data sets and is not a calendar year. Therefore, in some cases month and day of FEV₁ could be from the previous calendar year.

This box-plot is a graphic representation of the FEV₁ in children, expressed as % of predicted, detailed in table 4.1. For each country, the dash (black line crossing the blue box) is the median, the black dot is the mean and the whiskers (vertical lines with a T-shaped end) are the minimum and the maximum

Figure 4.2 FEV₁% of predicted: box-plot, by country and overall. Patients aged 18 years or older who have never had a transplant.



Note: Belarus and Georgia have 0% coverage for adults and are excluded from the table.

Note: Albania, Armenia, Cyprus and Romania have <5 patients aged 18 years or more at FEV₁ measurement and are excluded from this graph.

Note: The UK reports best FEV₁ from the annual review, which is the time period between data sets and is not a calendar year. Therefore, in some cases month and day of FEV₁ could be from the previous calendar year.

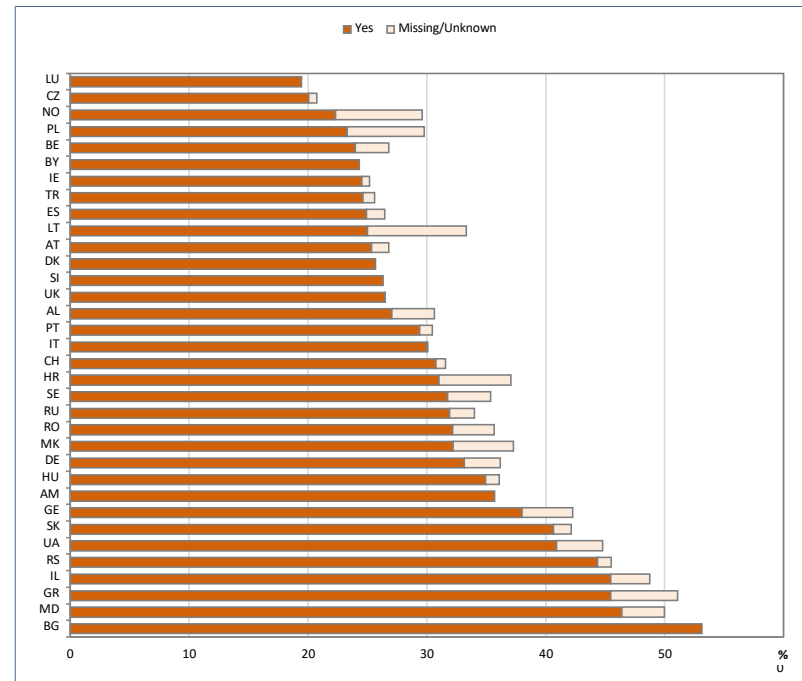
This box-plot is a graphic representation of the FEV₁ in adults, expressed as % of predicted detailed in table 4.2. For each country the dash (black line crossing the blue box) is the median, the black dot is the mean and the whiskers (vertical lines with a T-shaped end) are the minimum and the maximum.

Table 6.9 BMI: descriptive statistics by country. All patients seen in 2018 aged 18 years or older who never had a transplant.								Country	N	N Miss	Mean	Min	25th pctl
(25% of the patients are below this BMI)		Median											
(50% of the patients are below this BMI)		75th pctl											
(75% of the patients are below this BMI)		Max											
Austria	318	1	21.8	14.5	19.5	21.2	23.6	35.5					
Belgium	618	0	22.2	15.8	19.9	21.9	23.7	34.3					
Bulgaria	74	0	20.5	13.8	17.9	19.8	22.5	40.1					
Croatia	36	0	21.5	15.4	19.8	21.5	22.9	28.1					
Czech Republic	242	0	21.5	14.3	19.0	21.1	23.3	33.3					
Denmark	254	0	22.7	16.0	19.9	22.0	24.6	38.2					
Germany	3171	11	21.7	11.8	19.4	21.2	23.4	40.6					
Greece	263	0	22.5	15.0	20.5	22.2	24.1	32.0					
Hungary	148	28	20.6	14.3	18.3	20.0	22.3	39.8					
Ireland	481	102	22.8	15.5	20.4	22.4	24.6	43.4					
Israel	304	1	22.8	14.4	20.3	22.4	25.0	37.9					
Italy	2695	32	22.2	12.9	19.9	21.8	24.0	44.8					
Latvia	<10	0	18.6	16.3	16.5	17.9	19.6	24.5					
Lithuania	15	0	19.9	15.3	18.0	19.6	21.2	24.6					
Luxembourg	18	0	22.9	16.8	21.4	22.4	23.8	35.2					
Rep of Moldova	<10	0	19.9	15.8	17.8	20.1	22.1	23.7					
The Netherlands	778	1	22.5	15.5	20.3	21.9	23.9	45.9					
North Macedonia	33	0	22.1	17.0	20.6	21.8	23.4	26.5					
Norway1	147	3	22.9	15.8	20.0	22.5	24.7	37.7					
Poland	190	0	20.9	14.9	18.3	20.4	22.7	36.0					
Portugal	115	0	22.2	14.5	20.1	21.6	24.1	38.7					
Russian Federation	671	3	19.6	11.8	17.4	19.2	21.3	35.8					
Serbia	45	1	19.7	14.4	17.9	19.5	21.8	25.9					
Slovak Republic	139	0	21.3	14.2	18.8	21.1	23.3	33.7					
Slovenia	31	0	20.6	15.1	18.9	21.1	23.0	24.3					
Spain	871	5	22.5	14.9	20.2	22.0	24.2	41.4					
Sweden	333	2	22.7	15.6	20.2	22.1	24.2	41.7					
Switzerland	445	0	21.7	14.0	19.6	21.5	23.4	37.0					
Turkey	171	3	20.7	13.3	18.9	20.1	22.5	31.7					
Ukraine	27	0	19.4	15.2	17.3	19.2	21.3	26.0					
United Kingdom	5113	18	23.1	13.2	20.4	22.5	25.1	49.4					

Table 6.11 BMI: descriptive statistics by country. All female patients seen in 2018 aged 18 years or older who never had a transplant. Country N

	Mean	Min	25th pctl					N Miss
(25% of the patients are below this BMI)		Median						
(50% of the patients are below this BMI)		75th pctl						
(75% of the patients are below this BMI)		Max						
Austria	153	0	21.1	14.5	19.4	20.7	22.5	35.5
Belgium	283	0	21.8	15.8	19.5	21.5	23.2	34.2
Bulgaria	34	0	19.3	15.2	17.9	19.0	20.4	27.7
Croatia	20	0	21.4	17.7	19.8	21.3	22.9	24.8
Czech Republic	127	0	21.0	14.3	18.9	20.6	23.0	33.0
Denmark	113	0	22.4	16.4	19.6	21.5	23.7	38.2
Germany	1479	5	21.1	11.8	19.0	20.7	22.6	40.6
Greece	123	0	21.7	15.0	19.7	21.7	23.1	31.7
Hungary	74	10	19.8	14.3	17.9	19.2	21.6	39.8
Ireland	194	45	22.1	15.9	19.7	21.6	23.4	43.4
Israel	130	1	22.4	16.4	19.6	21.4	24.4	37.4
Italy	1255	16	21.5	12.9	19.3	21.0	23.1	42.4
Latvia	<10	0	17.0	16.3	16.4	16.5	17.3	18.8
Lithuania	<10	0	19.0	15.3	15.6	19.0	21.0	24.2
Luxembourg	<10	0	23.8	18.2	20.6	22.4	25.4	35.2
The Netherlands	355	1	22.3	15.5	20.1	21.6	23.6	45.9
North Macedonia	12	0	21.5	17.0	20.5	21.6	22.4	24.9
Norway1	66	2	22.1	15.8	19.7	21.2	23.7	33.1
Poland	109	0	20.6	14.9	18.4	20.2	21.7	35.7
Portugal	55	0	22.4	14.5	20.3	21.9	23.3	38.7
Russian Federation	326	2	19.4	13.0	17.3	18.8	21.1	35.8
Serbia	19	1	19.7	14.4	17.7	19.6	22.1	25.2
Slovak Republic	73	0	20.5	14.3	18.2	19.6	22.5	26.0
Slovenia	18	0	19.8	15.1	17.5	20.2	21.3	23.0
Spain	391	2	21.9	15.1	19.4	21.5	23.4	41.4
Sweden	153	1	22.1	15.6	19.9	21.7	23.5	41.7
Switzerland	201	0	21.2	14.3	19.0	20.7	22.3	37.0
Turkey	78	2	20.3	13.3	18.8	19.9	21.9	29.9
Ukraine	<10	0	18.8	16.6	17.3	18.5	20.8	21.4
United Kingdom	2326	11	22.9	13.5	20.1	21.9	24.5	49.4

Figure 5.1 Prevalence of chronic *Pseudomonas aeruginosa* infection in all patients seen in 2018 who have never had a transplant, by country.



Note: We excluded from the graph the countries for which the information on *Pseudomonas aeruginosa* was missing for more than 10% of the patients.

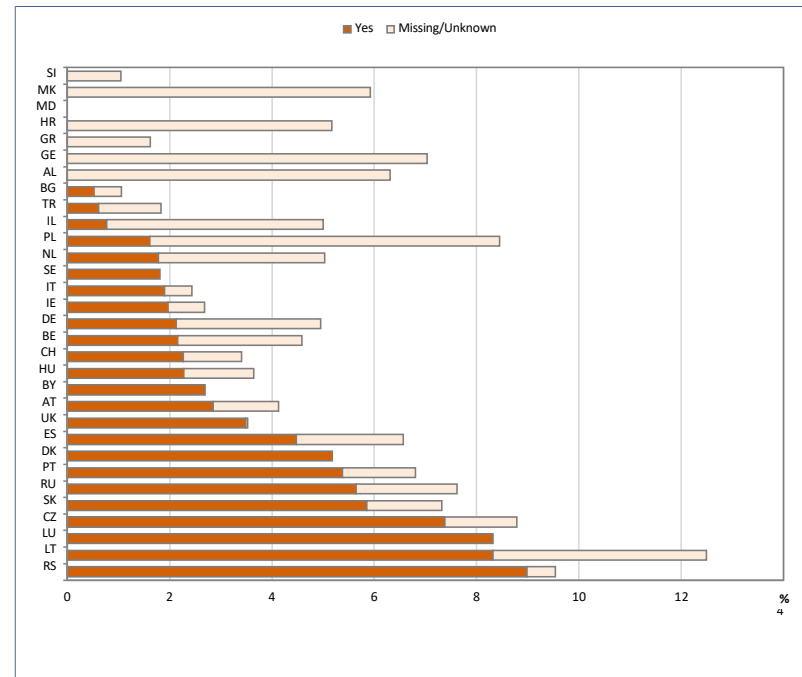
Note: Ireland: chronicity for *Pseudomonas aeruginosa* is defined as: at least 3 or more positive isolates during the last 12 months preceding the last reported culture in 2018.

Italy: chronicity for *Pseudomonas aeruginosa* is defined as: at least 3 or more positive cultures during 2018.

United Kingdom: for chronic *Pseudomonas aeruginosa* the definition is: 3 or more positive isolates during the last 12 months.

The horizontal bars represent the percentage of patients with chronic *Pseudomonas aeruginosa* infection (in dark orange) and the percentage of patients where information on *Pseudomonas aeruginosa* infection was missing (in light orange). This is a frequent infection, but prevalence varies considerably between countries.

Figure 5.2 Prevalence of chronic *Burkholderia cepacia complex* species infection in all patients seen in 2018 who have never had a transplant, by country.



Note: We excluded from the graph the countries for which the information on *Burkholderia cepacia complex* species was missing for more than 10% of the patients.

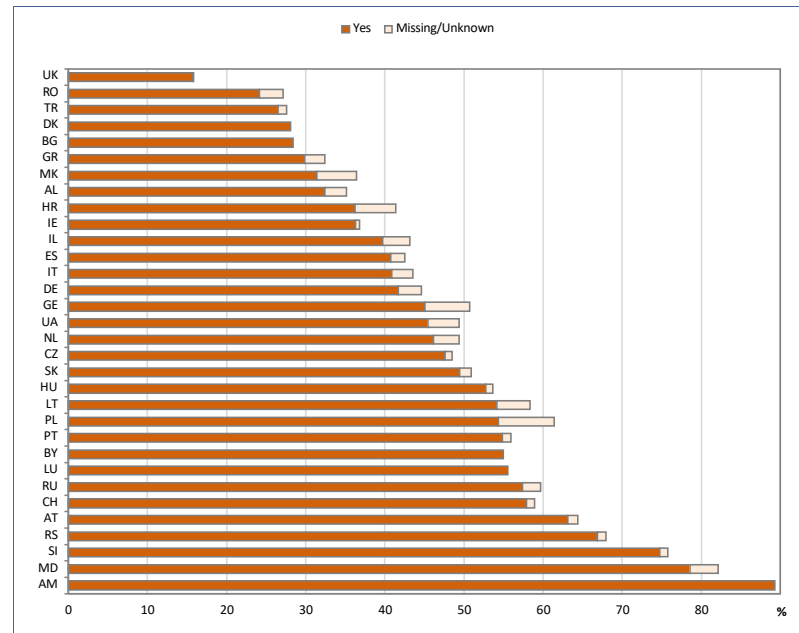
Note: Ireland: chronicity for *Burkholderia cepacia complex* species is defined as: at least 3 or more positive isolates during the last 12 months preceding the last reported culture in 2018.

Italy: chronicity for *Burkholderia cepacia complex* species is defined as: at least 3 or more positive cultures during 2018.

United Kingdom: information on *Burkholderia cepacia complex* species is collected as: *Burkholderia cepacia complex* species grown since last annual review, not necessarily chronic.

The horizontal bars represent the percentage of patients with chronic *Burkholderia* infection (in dark orange) and the percentage of patients where information on *Burkholderia* infection was missing (in light orange). This infection is much less frequent than *Pseudomonas aeruginosa* (note the different scale on the horizontal axis), and there is also some variation.

Figure 5.4 Prevalence of chronic *Staphylococcus aureus* infection in all patients seen in 2018 who have never had a transplant, by country.



Note: We excluded from the graph the countries for which the information on *Staphylococcus aureus* was missing for more than 10% of the patients.

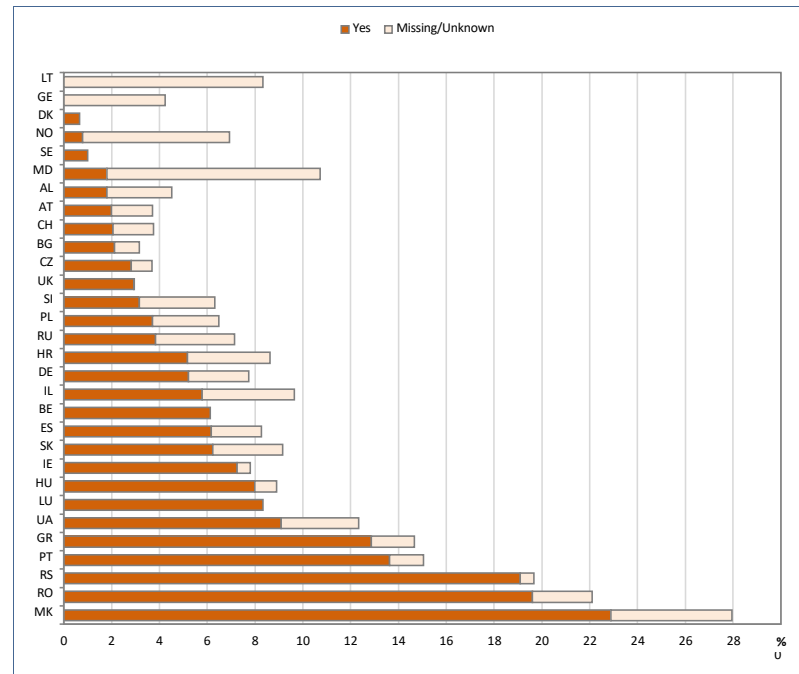
Note: Ireland: chronicity for *Staphylococcus aureus* is defined as: at least 3 or more positive isolates during the last 12 months preceding the last reported culture in 2018.

Italy: chronicity for *Staphylococcus aureus* is defined as: at least 3 or more positive cultures during 2018.

United Kingdom: for chronic *Staphylococcus aureus* the definition is: 3 or more positive isolates during the last 12 months.

The horizontal bars represent the percentage of patients with chronic *Staphylococcus aureus* infection (in dark orange) and the percentage of patients where information on *Staphylococcus aureus* was missing (in light orange). This infection is as frequent as chronic *Pseudomonas aeruginosa* infection and a similar degree of variation between the countries can be observed.

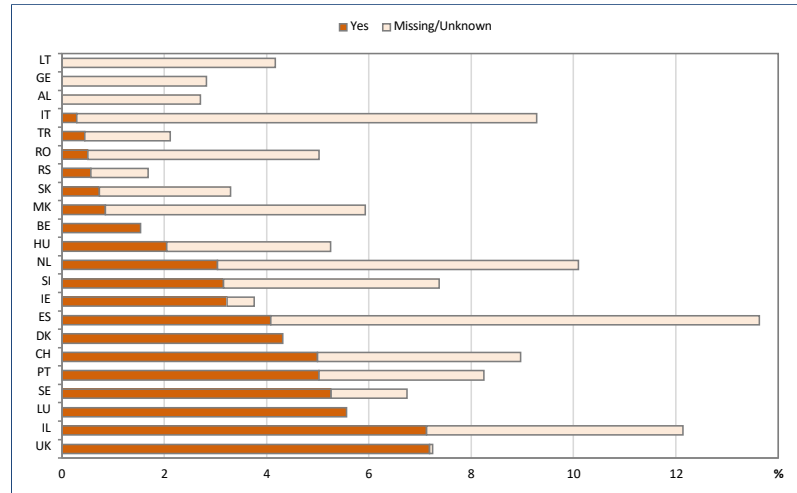
Figure 5.5 Prevalence of methicillin-resistant *Staphylococcus aureus* (MRSA) infection in all patients seen in 2018 who have never had a transplant, by country.



Note: We excluded from the graph the countries for which the information on MRSA was missing for more than 10% of the patients.

The horizontal bars represent the percentage of patients with methicillin-resistant *Staphylococcus aureus* infection (in dark orange) and the percentage of patients where information on methicillin-resistant *Staphylococcus aureus* was missing (in light orange).

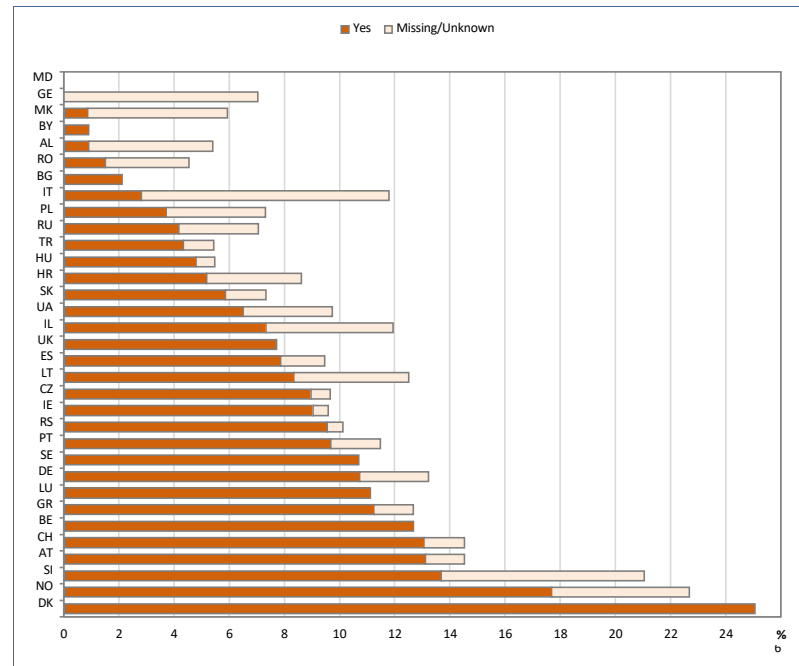
Figure 5.6 Prevalence of non-tuberculous mycobacteria in all patients seen in 2018 who have never had a transplant, by country.



Note: We excluded from the graph the countries for which the information on non-tuberculous mycobacteria was missing for more than 10% of the patients.

The horizontal bars represent the percentage of patients with non-tuberculous mycobacteria infection (in dark orange) and the percentage of patients where information on non-tuberculous mycobacteria infection was missing (in light orange). Detection of non-tuberculous mycobacteria infection depends on sputum production, which is not always possible for all patients, especially younger patients. Generally, infections from these bacteria are not very frequent in any country.

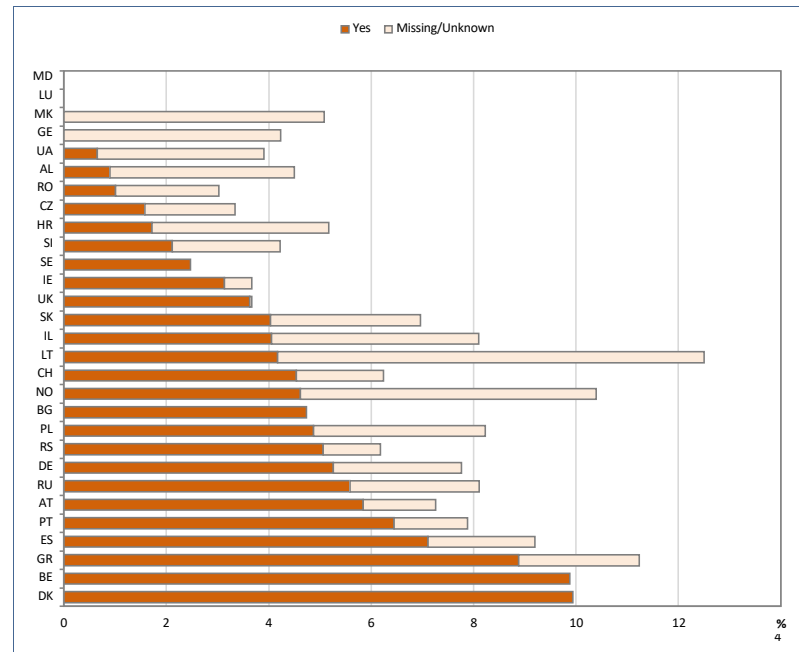
Figure 5.7 Prevalence of *Stenotrophomonas maltophilia* infection in all patients seen in 2018 who have never had a transplant, by country.



Note: We excluded from the graph the countries for which the information on *Stenotrophomonas maltophilia* was missing for more than 10% of the patients.

The horizontal bars represent the percentage of patients with *Stenotrophomonas maltophilia* infection (in dark orange) and the percentage of patients where information on *Stenotrophomonas maltophilia* was missing (light orange). The frequency varies considerably between countries.

Figure 5.8 Prevalence of *Achromobacter* species in all patients seen in 2018 who have never had a transplant, by country.



Note: We excluded from the graph the countries for which the information *Achromobacter species* was missing for more than 10% of the patients.

The horizontal bars represent the percentage of patients with *Achromobacter species* infection (in dark orange) and the percentage of patients where information on *Achromobacter species* infection was missing (in light orange).

Nouveautés 2020

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- **Simeox**
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- Symkevi, Kaftrio
- CF et COVID Luxembourg et Europe
- Problèmes – registre 2019 (année transition) et reprise 2020

Simeox



Nouveautés 2020

- Rapport ECF – PR / 2018
- Simeox
- **Aiocare – spirométrie à domicile + questionnaire spécifique**
- Symkevi, Kaftrio
- CF et COVID Luxembourg et Europe
- Problèmes – registre 2019 (année transition) et reprise 2020

Aiocare - spirométrie



Transfert listing

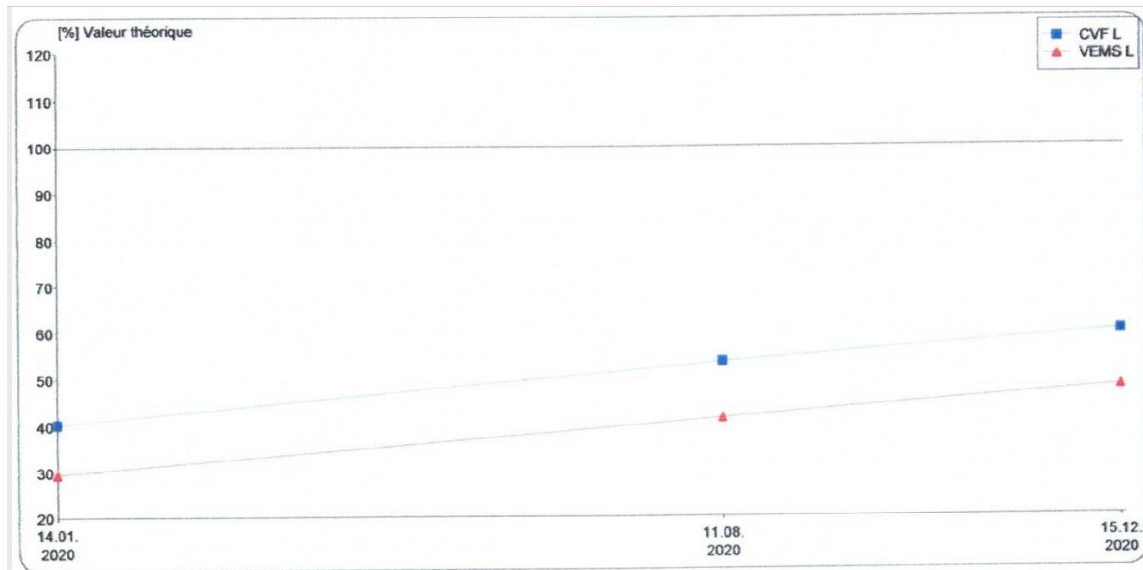
- Demander aux patients de contacter l'ALLM

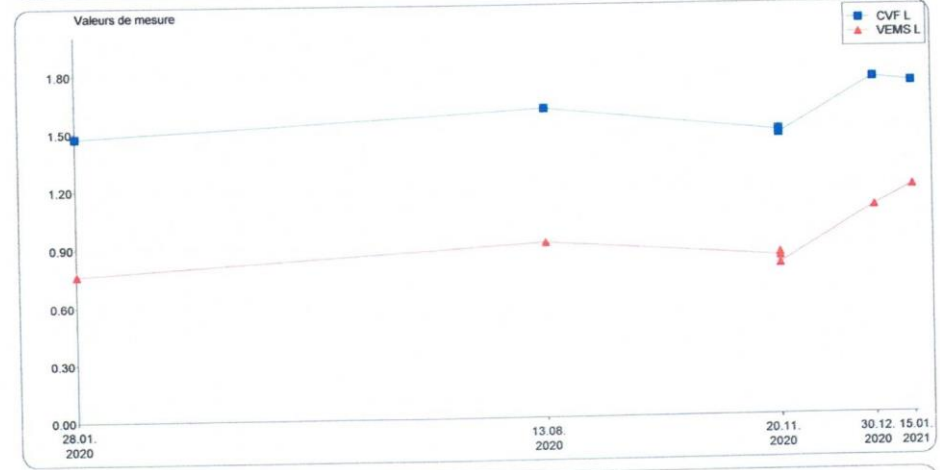
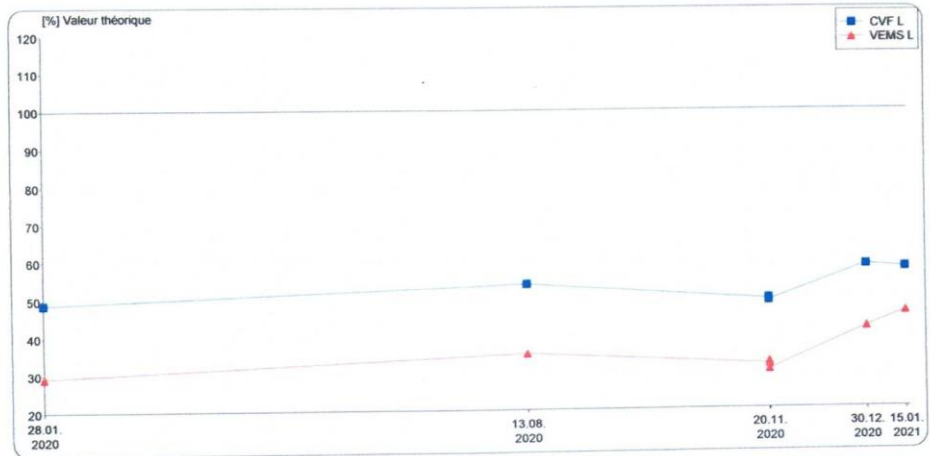
Nouveautés 2020

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KAFTRIO

- Echec compassionate use program
- Premier pays Benelux
- 6 + 2 patients
- Tolérance
- Prise de poids
- Exemple de courbe d'évolution des premières patientes





Nouveautés 2020

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Incidence of COVID-19 in people with cystic fibrosis in Europe between February and June 2020

Target journal is Journal of Cystic Fibrosis

- SARS-CoV-2 incidence was significantly higher in pwCF compared to the general population in age categories up to 49 years of age. Lung-transplanted pwCF have more severe illness than non-transplanted pwCF. Further work is required to elucidate independent risk factors, and to monitor the second wave of SARS-CoV-2 in Europe.
- Overall, the case fatality rate observed in this study was 4.07% (CI: 1.33 – 9.23) for the CF population. This is notably but not significantly lower than the case fatality rate calculated for the general population of the same group of countries (7.46%) (p=0.172).
- For the period up to 30 June 2020, there were 123 cases of PCR-confirmed SARS-CoV-2 in pwCF for a total at-risk population of 48211 pwCF. This yields an incidence of 2.55 cases per 1000 pwCF (95% CI: 2.12-3.04). This is significantly lower than the incidence of 3.10 cases per 1000 people in the general population of the entire 38-country ECFSPR area in the same period (based on ECDC data [13])
- When incidence was considered by age bracket, SARS-CoV-2 incidence was significantly higher in pwCF versus the general population for those aged <15 years, 15-24 years, and 25-49 years (p<0.001)
- **In conclusion, SARS-CoV-2 incidence was significantly higher in pwCF compared to the general population in age categories up to 49 years of age. Lung-transplanted pwCF have more severe illness than non-transplanted pwCF. Further work is required to elucidate independent risk factors, and to monitor the second wave of SARS-CoV-2 in Europe.**

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