compared to those reported in Veneto-Trentino Alto Adige (Italy), Israel, Victoria (Australia), and Wisconsin and Colorado (US). The current incidence was compared to that observed in the other European countries that recently reported their experience in NBS for CF.

Results: Over the 43-year period, the incidence of CF was estimated at 1/2811 in Brittany. Poisson regression showed that the incidence decreased significantly over the study period (annual APC: – 1.8%, $p < 0.0001$). It dropped from 1/1983 over the 1975–79 period to 1/3992 over the 2015-17 period, leading to a decline of 50.3% ($p = 0.0019$). Comparison with other areas revealed that: 1°) most areas reported a decline in incidence, but not all; 2°) the most significant declines were observed in areas where a carrier screening program was implemented; 3°) although halved, the incidence of CF in Brittany remained among the highest reported in Europe.

Conclusion: This study highlights how the incidence of CF has evolved in an area where CF is frequent. It reports a clear drop in incidence that results from a complex mixture of factors. Comparison with other areas help to better understand factors influencing the incidence, which depends notably on the health policies implemented on CF.

WS23.2

Epidemiology of European adults with cystic fibrosis

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The European Cystic Fibrosis Society Patient Registry (ECFSPR) collects anonymised demographic and clinical data from consenting people with CF in Europe. The aim of this study is to describe the adult population with CF in Europe.

We considered patients of 18 years or older in the 2017 data, the latest year of follow-up available. Countries with a coverage below 80% were not considered when reporting figures by country. Percentages are computed for categorical variables; quartiles for numerical variables.

For the year 2017, the ECFSPR database contains data of 48,204 patients from 35 countries. 24,491 (51.3%) are adults.

The percentage of adults varies from 6.5% in Albania to 62.8% in Sweden. The proportion of adult females varies from 37.1% in North Macedonia to 59.6% in Slovenia. The highest percentage of patients living with lung transplant is 22.7% in Slovenia.

Considering patients of 18–29 years, the lowest percentage of chronic *Pseudomonas Aeruginosa* is 30.6% in France, the highest is 79.4% in Serbia. For chronic *Burkholderia cepacia complex* species the lowest percentage of infected patients is 2.5% in Italy, the highest 20.6% in Serbia. The lowest percentage of CF related diabetes is 7.3% in Slovak Republic, the highest 34.2% in the Czech Republic. Median of BMI changes from 18.8 in the Russian Federation to 22.4 kg/m^2 in Luxembourg. Considering patients not transplanted, we observe a percentage of patients with FEV1% below 40% that varies from 3.0% in Denmark to 28.5% in the Russian Federation.

We considered only countries with a coverage of 80% or higher, for a realistic reflection of CF and observed variation among those countries. Data show that the percentage of adult population is higher in North Europe, infections are more frequent in Eastern Europe and growth and lung function are worse in Eastern Europe. Further investigation is required to investigate differences in healthcare systems to plan an adequate development of CF care services for adult CF people.

WS23.3

The average rate of lung function decline in adults with cystic fibrosis in the United Kingdom: data from the UK Cystic Fibrosis Registry

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Objectives: The annual rate of change in lung function is an important measure of disease progression. We sought to determine the rate of decline of percent predicted FEV_1 ($ppFEV_1$) for adults attending UK CF centres, accounting for age, sex and pancreatic status.

Methods: Data on $ppFEV_1$ were obtained from the UK Cystic Fibrosis registry from 2015–2017. Patients post-lung transplant were excluded. Multilevel modelling was conducted to calculate the annual rate of change in lung function accounting for sex and age in pancreatic insufficient (PI) and sufficient (PS) adults.

Table 1.
Rate of Decline in ppFEV1 predicted in One Year in Adults with CF

		Pancreatic Insufficient			Pancreatic Sufficient		
		N	Mean annual change in ppFEV1	95% Confidence intervals	N	Mean annual change in ppFEV1	95% Confidence intervals
18–28 years	Females	1124	−1.76	−2.06 to −1.46	171	−1.01	−1.84 to −0.19
	Males	1287	−1.61	−1.91 to −1.31	119	−0.53	−1.39 to 0.33
29–39 years	Females	608	−1.49	−1.85 to −1.13	133	−0.74	−1.47 to −0.02
	Males	851	−1.31	−1.60 to −1.02	125	−0.86	−1.96 to 0.23
40–50 years	Females	217	−1.42	−1.98 to −0.87	94	−0.79	−1.60 to 0.01
	Males	301	−1.15	−1.59 to −0.71	109	−0.13	−1.01 to 0.75
51+ years	Females	81	−1.61	−2.64 to −0.58	92	−0.51	−1.45 to 0.43
	Males	148	−1.06	−1.66 to −0.46	75	0.86	−0.23 to 1.95

Results: Overall, $ppFEV_1$ declined annually by −1.52% (95% CI: −1.66 to −1.38%) and −0.55% (95% CI: −0.86 to −0.23%) in those PI and PS respectively. In PI patients, females had a greater rate decline in ppFEV1. The fastest rate of decline was observed in PI individuals between the ages of 18–28 years (Table 1). The pattern was more inconsistent between sexes and age categories in those PS (Table 1). The largest annual rate of decline in ppFEV1 was seen in 18–28 years old females, with the effect reducing with age. In contrast, men aged 29–39 years had the largest decrease in ppFEV1 and by age 51+ years there was no statistically significant change.

Conclusions: PI individuals had three times the average rate of decline of those PS. Younger adults, especially females tended to have a faster rate of decline and need close monitoring.

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WS23.4

Investigating associations between air pollution and the severity of cystic fibrosis in Great Britain

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Objectives: Our aim was to estimate the effect of air pollution on disease severity among patients with cystic fibrosis (CF) in Great Britain (GB).

Methods: We used a semi-ecological small-area study design. High-resolution (100 × 100 m) modelled surfaces of NO_2 (2009) and $PM_{2.5}$ (2010) concentrations in GB, based on land use regression models, were obtained from the Small-Area Health Statistics Unit, Imperial College London. Exposures were assigned based on postcode of residence. CF data for 2016 were obtained from the UK CF Registry, a centralised database which, for 2016, included records of 9,297 patients from all CF specialist centres across GB. We used two common proxies of disease severity: the percent predicted forced expiratory volume in 1 second ($ppFEV1\%$) and the annual number of days on intravenous antibiotics for pulmonary exacerbation (#IVdays). Linear and Poisson regressions, adjusted for age, gender and deprivation (2015 Index of Multiple Deprivation), were used to explore the association between exposures (NO_2 and $PM_{2.5}$) and outcomes ($ppFEV_1\%$ and #IVdays).

Results: Excluding CF patients < 6 years old (n = 1,493) and those with a missing postcode (n = 64), 7,740 patients were included. Mean $ppFEV_1\%$ and #IVdays were 70.5% (standard deviation (SD): 24%) and 19 (SD: 31) days, respectively. Annual mean concentrations ranged between 2.6 and