**Conclusions:** The current study showed that, in our country, every year, a large proportion of new CF diagnoses is made in adult subjects; compared to the previous 2004 study these patients have a higher median age at diagnosis, less prevalence of *Ps. aeruginosa* chronic infection, a good nutritional status and a good median $FEV_1$p.p. Prevalence of pancreatic insufficiency is confirmed low, compared to the national data (67%); this might suggest a mild condition of the disease among this specific patient group. These data might be due to a better knowledge of CF disease by adult physicians.

Further studies are needed to clarify the weight of a true diagnostic delay or a late manifestation of CF symptoms.

**Reference:**

1. Epidemiologia e Prevenzione, 2019, in press.

## 1.1.10
## Air pollution exposure is associated with lung function decrease in adults with cystic fibrosis

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**Background**: Several studies have found an association between outdoor air pollution and cystic fibrosis (CF) exacerbation and/or lung function deterioration, but these are limited to children populations. Now that life expectancy of cystic fibrosis patients has significantly increased, it becomes relevant to investigate these relationships in adults with CF. The objective of this study was to assess the effect of outdoor air pollution on lung function in adults with CF.

**Methods**: The relationship between the daily mean concentration of major air pollutants (particulate matter, nitrogen dioxide, ozone) and lung function was studied retrospectively in a cohort of 283 patients aged over 18 years followed-up between January 1, 2012 and December 31, 2014 in the Parisian region. Daily average concentrations of air pollutants were modeled and evaluated using the chemistry-transport model CHIMERE near the patient's residence. A Generalized Additive Model (GAM) following a Quasi-Poisson distribution with a Distributed Lag Non-Linear Model (dlnm) associated with every 10µg/m$^3$ increase in air pollutant concentration was used to examine the relationship of exposure to air pollutants to having a decreased lung function. (Having a decreased lung function is characterized by clinically significant deficits in Forced vital capacity (FVC). A low FVC as a FVC below 80 percent of the predicted value). We adjusted the model to the effect of day of the week, temperature, calendar time, relative humidity.

**Results**: 121 of the patients were women (42.8%) and 162 men (57.2%). With lag 0 as reference corresponding to the day of the visit, a decreased lung function was significant for $PM_{10}$ from the 7th day before it ($RR_{lag\ 7}$: 1.04, 95% CI, 1.001–1.07) and for $NO_2$ from the 2$^{nd}$ day ($RR_{lag\ 2}$: 1.08, 95% CI, 1.005–1.17).

**Conclusion**: For the first time, our data provide evidence of a significant association between exposure to outdoor air pollutants and lung function in adult patients with CF according to various lags.

## 1.1.11
## Epidemiology of European adults with Cystic Fibrosis

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**Background**: The European Cystic Fibrosis Society Patient Registry (ECFSPR) collects anonymised demographic and clinical data from consenting people with CF in Europe. A common set of variables and definitions is used to collect the information. Countries with a national registry extract data from their own database and import data into the ECFSPR software, countries with individual centres enter data directly in the system.

**Aim**: To describe the adult population with CF in Europe.

**Methods**: We considered people with CF of 18 years or older in the European data of 2017, the latest year of follow-up available. Countries with a coverage below 80% were not considered when reporting figures by country (1). Percentages are computed for categorical variables; quartiles are computed for numerical variables. We did not consider a country if there was 10% of missing information for a variable and we did not report the percentage if computed on less than 5 patients. We did not compute quartiles when the number of patients is less than 10 in an age group. FEV1% of predicted was computed using Global Lung Function Initiative equations (2).

**Results**: For the year 2017, the ECFSPR database contains data of 48,204 people with CF from 35 countries. 24,491 (51.3%) are adults, the median age at follow-up is 29.7 years and the maximum age is 88.4 years.

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.

Considering patients of 18–29 years, the lowest percentage of chronic *Pseudomonas Aeruginosa* is 30.6% in France and the highest is 79.4% in Serbia. For chronic *Burkholderia cepacia complex* species the lowest percentage of infected patients is 2.5%, reported in Italy, and the highest 20.6% in Serbia. The lowest percentage of CF related diabetes is 7.3% in Slovak Republic and the highest 34.2% in Czech Republic. The highest percentage of patients living with lung transplant is 12.5% in Portugal. Median of BMI changes from 18.8 in the Russian Federation to 22.4 kg/m$^2$ in Luxembourg. Considering the patients 18–29 years who have never been transplanted, we observe a percentage of patients with FEV1% below 40% that varies from 3.0% in Denmark to 28.5% in Russian Federation.

**Conclusions**: We considered only countries with a coverage of 80% or higher, for a realistic reflection of CF in those countries, 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 Countries and growth and lung function are worse in eastern European countries. Differences in healthcare systems may be a possible explanation. Further investigation is required to investigate this aspect to plan an adequate development of CF care services for adult CF people.

**References**:

1. Zolin A et al., ECFSPR Annual Report 2017, 2019.
2. Quanjer PH et al., Multi-ethnic reference values for spirometry for the 3–95-yr age range: the global lung function 2012 equations. *Eur Respir J* 2012; 40: 1324–1343.