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patient); (3) Pa strains isolated from several water points in home 5 and 10 years apart.

Very few studies have been conducted on the microbial communities of the CF domestic environment. For 3 patients, the epidemiological links observed between clinical and environmental strains suggested that the domestic environment could represent a source of contamination. This type of study should be encouraged for a better understanding of the potential cycles of bacterial transmission.

#### P160

##### Detection and management of nontuberculous mycobacteria in cystic fibrosis patients in a tertiary paediatric centre

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**Background:** Detection and management of nontuberculous mycobacteria (NTM) lung disease can be challenging in a paediatric cystic fibrosis (CF) population. Consensus guidelines (Floto et al, Thorax 2016) recommend that all spontaneously expectorating patients have at least 1 sputum sample sent annually for NTM detection, and recommend further investigations and treatment.

**Objective:** To investigate our centre's compliance with guidelines, and explore the challenges of managing NTM lung disease in a paediatric population.

**Methods:** Sputum-productive patients were identified from our large tertiary CF centre in Northwest England. Electronic medical records were reviewed for samples, results, management and outcomes, which were compared to guidelines.

**Results:** 183 patients were identified over a 4-year period, of whom 101 (55.2%) were sputum producing. 91/101 (90%) patients had  $\geq 1$  sample sent for NTM. 10 patients (11%) had  $\geq 1$  positive sample (8 *M. abscessus*, 1 *M. avium*, 2 other/mixed growth).

5 patients (4 male, mean age 12.2 years, mean FEV<sub>1</sub> 78.5%) were treated for NTM lung disease based on clinical condition/repeat samples/CT findings (4 *M. abscessus*, 1 *M. avium*). All patients with *M. abscessus* ultimately tolerated antibiotic treatment according to guidelines (minimum treatment duration 18 months) though initiation and choice of drug were complicated by vomiting in some cases. The patient with *M. avium* tolerated treatment with oral antibiotics for a total duration of 18 months. 1 patient continues on treatment and one transitioned to adult care during treatment. 1 patient completed treatment with 10% improvement in FEV<sub>1</sub> and no further NTM growth. 2 patients continue to grow NTM despite treatment and are considered colonised.

**Conclusion:** NTM remains a significant consideration in paediatric CF patients. Not all positive samples represent true NTM lung disease. International consensus guidelines can be applied to treat this population, but do not always result in eradication.

#### P161

##### The influence of chronic lung infections on the development of bronchiectasis in patients with cystic fibrosis

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**Aim:** Determining the correlation between chronic lung infections and the formation of bronchiectasis in patients with cystic fibrosis (CF).

**Methods:** In this study 80 patients diagnosed with CF were examined, registered in the Cystic Fibrosis Centre, Republic of Moldova. The diagnosis of CF was confirmed by sweat test and DNA molecular research, also by lung imaging examinations to highlight and monitor respiratory syndrome.

**Results:** In the study group, 70.76% of CF patients had complications. The most common complication was bronchiectasis (55.38%). 60% of patients with CF have sacciform bronchiectasis, mainly with localisation in the upper-right lung lobe, but in 1/3 cases they are extended bilaterally. Tubular bronchiectasis is seen in 24.4% of patients. Sacciform and varicose bronchiectasis was determined in children over 15 years of age. In 15% of patients the contralateral lung was not significantly affected, with changes

characterised by thickening of the bronchial walls, mucus plugs, fibrosis sectors, signs of bronchitis, mild hyperinflation. In the majority of patients, bronchiectasis occurred on the background of chronic lung infection with *P. aeruginosa* (73.8%), of which 45.2% associated coinfection with *S. aureus*, 1 patient - *B. cepacia*. Imaging evaluation by HRCT revealed a polymorphic picture of bronchial tree deformities in all patients colonised with *P. aeruginosa*, and in non-*P. aeruginosa* patients - bronchial deformities were identified only in 1/3 cases. Thus, *P. aeruginosa* is the pathogen with the most adverse effects on the respiratory system in children, the frequency of which increases with the advancement of patients ( $r^2 = .81$ ,  $p < 0.01$ ) and is responsible for progressive lung damage.

**Conclusion:** The study demonstrated a significant parallel ( $p < 0.01$ ) between the frequency of assessment of *P. aeruginosa* infection, which is responsible for a more severe evolution of lung pathology by the installation of severe irreversible bronchial deformities.

#### P162

##### Pneumococcal and influenza vaccination coverage level: data from a cystic fibrosis centre

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**Objectives:** To assess the seasonal influenza and Pneumococcal vaccination coverage level for the 2020–2021 season in a population of CF patients compared to routine vaccination coverage.

**Method:** The study included CF patients over 6 months of age, seen in a specialised CF centre in Northern Greece. The families were asked to fill in a questionnaire and send their vaccination charts by e-mail to assess data on mandatory and recommended vaccines. Telephone interviews were performed as well to assess possible factors associated with nonadherence with vaccination. The study was performed from October to December 2020.

**Results:** The study included 63 patients with CF (54% boys), with a mean age of 14.29 years. Coverage for seasonal flu vaccination was 93.75%, while coverage for polysaccharide pneumococcal vaccine was 73.44%. Coverage for DTPCaHi, conjugate pneumococcal, MMR, and hepatitis B was 100% in 63.5%, 75–99% in 27%, and under 75% in 9.5% of the patients, respectively. Moreover, coverage for varicella and hepatitis A were found 100% for 63.5%, 50–75% for 22.2% and 0% for 14.3% of the study population. The 96.83% of the parents reported that the clinicians informed them regarding their children's routine and specific for CF vaccination recommendations.

**Conclusion:** The vaccination coverage level was high for vaccines more specifically recommended in CF. Nevertheless, there is room for improvement, especially for routine immunisation. Further studies are needed to understand motivators and barriers to vaccination of children with cystic fibrosis.

#### P163

##### Effects of the SARS-CoV-2 pandemic on attendance at the cystic fibrosis centre and the definition of the microbiological status of cystic fibrosis patients in Tuscany

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**Objectives:** The SARS-CoV-2 pandemic and the consequent lockdown periods have profoundly changed the health habits of patients. The fear of the viral infection as well as the reorganisation of staff within the health services due to the pandemic caused a drop in patients' attendance at CF centres. This resulted in a decrease in cultures performed, on which the definition of microbiological status is based. The ECFS definition of patients' microbiological status (mainly the definition of chronic *P. aeruginosa* infection) is based on the modified Leeds criteria (>50% of the samples positive, at least 4 samples collected) or significantly raised anti-Pa antibodies. Following the different lockdown periods imposed in Tuscany in 2020 (March 9th – May 18th) we verified:

- the number of outpatient visits to the Centre

- the number of cultures performed compared with previous years
- compliance with the criteria suggested by disease registries/standards of care for the definition of chronic infection
- the utility of anti-Pa serology in the correct definition of chronic Pain the epidemic context

**Methods:** Retrospective study on data regarding outpatient examinations, number of cultures performed and serology tests.

**Results:** Outpatients' attendance at the Centre (2020)

Jan	Feb	Mar	Apr	May	Jun	Jul	Aug	Sep	Oct	Nov	Dec
206	161	77	55	102	126	140	107	171	163	142	117

% of patients with at least 4 cultures performed yearly.

	2018	2019	2020
Total patients	352	383	370
Mean $\pm$ SD cultures	4.4 (2.28)	3.81 (2.02)	3.19 (1.77)
% patients $\geq$ 4 cultures	64	53	39

In 2020 we observed a significant reduction ( $p < 0.05$ ) in the mean number of cultures per patient and in the percentage of patients who performed at least 4 cultures.

During 2020, 108 (29.2%) patients with fewer than 4 cultures performed Pa serology, 25 (23.1%) tested positive, completing the definition required by the ECFS.

**Conclusions:** The SARS-CoV-2 infection caused a drop in patients' attendance at the centre, compromising the correct definition of Pa chronic infection based on the number of cultures. In the pandemic context, serology could be a useful complementary test to correctly fulfill the ECFS definition.

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#### P164

##### What is the tolerance of antibiotic dry powder inhalers in patients with cystic fibrosis?

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**Background:** Colobreathe dry powder for inhalation (CDPI)<sup>1</sup> and tobramycin inhalation powder (TIP)<sup>2</sup> have both been shown to be safe and effective for the long-term treatment of patients with cystic fibrosis (CF). The aim is to assess the real life tolerance to both dry powder inhalers (DPI) in the cohort at St Bartholomew's Adult CF centre.

**Methods:** Retrospective review of patients that started a DPI between January 2017 and December 2020. Demographic data was collated to review those that continued versus those that discontinued DPI therapy and reasoning.

**Results:** 33 assessments (17 CDPI and 16 TIP) were performed on 30 patients (female,  $n = 15$ ) with a median age of 23 years (range 17–46). Median forced expiratory volume in 1 second (FEV<sub>1</sub>) percentage predicted was 60% (range 32–104), with those in the CDPI group having a higher median FEV<sub>1</sub> (70% range 32–104) compared to the TIP group (59%, range = 36–93).

A DPI was discontinued in 18 cases (8/17 CDPI and 10/16 TIP), 16 by patients due to intolerance and two by the CF team. There was no difference in FEV<sub>1</sub> percentage predicted between those that discontinued and or continued DPI therapy (CDPI 70% vs. 70% and TIP 59% vs. 60%). However, in the TIP group those that discontinued were generally older, median 29 years (range = 17–44) versus those that continued, 20 years (range = 18–32).

The main reasons for patient discontinuation were cough ( $n = 7$ ) and chest tightness ( $n = 6$ ) and less common were haemoptysis, hoarse voice and poor adherence.

**Conclusion:** More than half of the patients who started a DPI discontinued therapy due to intolerance citing cough and chest tightness as the main reasons for cessation.

There was no difference in age or lung function those that continued or discontinued DPI therapy.

Further evidence to review discontinuation of inhaled therapies, nebulised and dry powder, will help clinical decision-making and selection of inhaled therapies in patients with CF.

#### P165

##### Improvement in antibiotic usage and ppFEV<sub>1</sub> with compassionate use of elxacaftor, tezacaftor and ivacaftor (ETI) for patients with cystic fibrosis

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**Background:** The triple CF transmembrane conductance regulator modulator therapy ETI became available in the UK in 2019 on compassionate grounds. Patients were eligible if they had the following genotypes: homozygous F508del mutation or heterozygous F508del mutation and a minimal function mutation, and one of the following: ppFEV<sub>1</sub> less than 40% for 2 months or referral for lung transplantation. At present, there is limited data on the impact of ETI in CF patients with severe lung disease.

**Objective:** To compare disease burden, using days of antibiotic used for exacerbations and ppFEV<sub>1</sub> as outcome measures, in the 12 months before and after initiation of ETI in CF patients.

**Methods:** We included all patients started on ETI on compassionate grounds at our institution who had been on ETI for at least one year at the time of writing. Electronic records were retrospectively analysed for antibiotic usage for exacerbations per patient, including oral, home IV, and inpatient IV antibiotics, and spirometry readings. We also analysed antibiotic usage for patients who had been on ETI for 6 months. Statistical analysis was performed using a paired t-test.

**Results:** We identified 11 CF patients who had data for 12 months before and after drug initiation. Annual mean total antibiotic days were significantly reduced from 99 (SD 36.4) to 24 (SD 25.2) ( $p < 0.0001$ ). Annual mean intravenous antibiotic days reduced from 66 (SD 44.7) to 12 (SD 24.6), and hospital intravenous antibiotic days reduced from 33 (SD 43.9) to 6 (SD 14.5). Lung function improved in ten patients. Overall mean ppFEV<sub>1</sub> increased from 30% (SD 7) to 45% (SD 15.7) ( $p < 0.0005$ ). In 21 additional patients who had data for 6 months, a similar trend in total antibiotic reduction was observed from 45 (SD 24) days to 13 (SD 18.4) days.

**Conclusions:** Patients prescribed ETI on compassionate grounds demonstrated a significant reduction in total and hospital intravenous antibiotic burden, and an improvement in lung function.

## Gastroenterology/Liver Disease/Endocrinology/ Metabolic Complications/Nutrition

#### P166

##### Prevalence of cystic fibrosis-associated liver disease in Albanian cystic fibrosis patients

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**Introduction:** Cystic fibrosis-associated liver disease (CFLD) affects around 30% of patients, being the third leading cause of death in CF patients.

**Aim:** To estimate the CFLD prevalence in Albanian CF patients.

**Methods:** We analysed all Albanian CF patients' data with CFLD.

CFLD was diagnosed when at least 2 of the following were present:

1. Physical examination with hepatomegaly and/or splenomegaly;
2. Abnormal liver function test;
3. Ultrasonographic evidence of liver involvement, portal hypertension, or biliary abnormalities.

**Results:** We analysed the data collected from 106 CF patients followed up during 2019: clinical assessment, liver biochemical tests, ultrasound examinations.