After 6 months of persistent PNX with 3 more hospitalisations (41 days IV Atb), patient could begin ETI association obtained in a compassionate use setting. At 3 months, CT scan revealed complete PNX resolution with reduced amount of mucoid impact and thickening of bronchial wall in bronchiectasis with partial reventilation of middle lobe.
At 2 years follow-up, FEV_1pp is 45; no further hospitalisations and only 14 days of oral Atb were needed. Listing for lung transplantation list is "suspended."
Conclusion: ETI treatment has been crucial for the resolution of a chronic pneumothorax in this case of severe CF in which all traditional therapeutic approaches failed.

Epidemiology/Registry

P059

Association of *Pseudomonas aeruginosa* infection stage with lung function trajectory in children with cystic fibrosis

M. Rosenfeld[1], A. Faino[2], P. Qu[2], W. Gordon[3], E. Blue[3], M. Collaco[4], Y.-H. Zhou[5], M. Bamshad[3], R. Gibson[1]. *[1]University of Washington, Pediatrics, Seattle, United States; [2]Seattle Childrens Research Institute, Seattle, United States; [3]University of Washington, Seattle, United States; [4]Johns Hopkins University School of Medicine, Baltimore, United States; [5]North Carolina State University, Raleigh, United States*

Background: The effect of chronic *Pseudomonas aeruginosa* (Pa) infection on lung function trajectory is not known. We aimed to compare the association of longitudinal lung function in children aged 6 to 21 with different stages of Pa infection (never, incident, chronic) using 4 definitions of chronic Pa.
Methods: Participants in the Early Pseudomonas Infection Control Observational Study (EPIC-OBS) cohort diagnosed with CF prior to age 3 contributed encounter-based culture and percent-predicted FEV_1 ($ppFEV_1$) data through the U.S. CF Patient Registry from 1992 to 2017. Cubic spline models were used to evaluate the association of Pa stage with longitudinal $ppFEV_1$using 4 chronic Pa definitions (Rosenfeld, et al, J of CF 2021 Aug 12; S1569). All models included age:Pa status interactions and were adjusted for birth year and relevant covariates.

MODEL ESTIMATES AT AGE 16	$ppFEV_1$		$ppFEV_1$ slope (per year)	
	Mean (SE)	Difference from Never Pa Mean (95% CI)	Mean (SE)	Difference from Never Pa Mean (95% CI)
Never Pa	91.3 (0.8)	–	−0.27 (0.16)	–
Incident Pa	89.4 (0.6)	−1.9 (−4.2, 0.5) p = 0.17	−0.74 (0.11)	−0.46 (−0.82, −0.1) p = 0.006
Chronic Pa	85.1 (0.6)	−6.3 (−8.6, −3.9) p < 0.001	−1.23 (0.10)	−0.95 (−1.3, −0.61) p < 0.001

Results: 1,264 subjects born 1992–2006 provided a median 9.5 [IQR 0.25 to 15.75] years of data. 89% developed incident Pa; 39%–58% developed chronic Pa depending on the definition. Estimated mean $ppFEV_1$ was lower after chronic Pa infection at all ages; this effect increased with age. Slope of $ppFEV_1$ decline was greater with chronic Pa after age 12. As an example, the table shows estimated mean $ppFEV_1$ value and slope at age 16 for individuals with reference value of all covariates using chronic Pa definition of 2 Pa+ yrs over 3 yrs. Results were similar across all definitions.
Conclusions: Chronic Pa infection is associated with lower FEV_1pp across all ages and more rapid decline over age 12 in children with CF, regardless of chronic Pa definition. Measures to prevent chronic Pa infection could slow FEV_1 decline and improve long-term outcomes.
Funding: U.S. Cystic Fibrosis Foundation BAMSHA18XX0

P060

Factors associated with pulmonary functions of cystic fibrosis (CF) patients in the National Cystic Fibrosis Patient Registry: a retrospective cohort study

N. Çobanoğlu[17], N. Emiralioglu[1], B. Çakır[2], A. Sertçelik[2], E. Yalçın[1], N. Kiper[1], V. Şen[3], D. Ufuk Altıntaş[4], H. Çokuğraş[5], A.A. Kılınç Sakallı[5], A. Kılıç Başkan[5], E. Hepkaya[5], H. Yazan[6], Ö. Türel[6], A.İ. Yılmaz[7], G. Ünal[7], T. Çağlar[7], E. Damadoglu[8], İ. Irmak[8], E. Demir[9], G. Öztürk[9], A. Bingöl[10], E. Başaran[10], N. Sapan[11], A.T. Aslan[12], P. Asfuroğlu[12], K. Harmancı[13], M. Köse[14], M. Hangül[14], A. Özdemir[15], G. Tuğcu[16], S. Eryılmaz Polat[16], G. Özcan[17], Z.G. Gayretli[18], Ö. Keskin[19], S. Bilgiç[19], H. Yüksel[20], Ş. Özdoğan[21], E. Topal[22], G. Çaltepe[23], D. Can[24], P. Korkmaz Ekren[25], M. Kılıç[26], A. Süleyman[27], T. Şişmanlar Eyüboğlu[12], G. Cinel[16], S. Pekcan[7], E. Çakır[6], U. Özçelik[1], D. Doğru[1]. *[1]Hacettepe University Faculty of Medicine, Pediatric Pulmonology, Ankara, Turkey; [2]Hacettepe University Faculty of Medicine, Public Health/ Epidemiology, Ankara, Turkey; [3]Dicle University Faculty of Medicine, Pediatric Pulmonology, Diyarbakır, Turkey; [4]Cukurova University Faculty of Medicine, Pediatric Allergy and Immunology, Adana, Turkey; [5]Istanbul University-Cerrahpasa, Cerrahpasa Faculty of Medicine, Pediatric Pulmonology, İstanbul, Turkey; [6]Bezmi Alem University Faculty of Medicine, Pediatric Pulmonology, İstanbul, Turkey; [7]Necmettin Erbakan University Faculty of Medicine, Pediatric Pulmonology, Konya, Turkey; [8]Hacettepe University Faculty of Medicine, Adult Pulmonology, Ankara, Turkey; [9]Ege University Faculty of Medicine, Pediatric Pulmonology, İzmir, Turkey; [10]Akdeniz University Faculty of Medicine, Pediatric Pulmonology, Antalya, Turkey; [11]Uludağ University Faculty of Medicine, Pediatric Allergy and Immunology, Bursa, Turkey; [12]Gazi University Faculty of Medicine, Pediatric Pulmonology, Ankara, Turkey; [13]Osmangazi University Faculty of Medicine, Pediatric Allergy and Immunology, Eskisehir, Turkey; [14]Erciyes University Faculty of Medicine, Pediatric Pulmonology, Kayseri, Turkey; [15]Mersin City Hospital, Pediatric Pulmonology, Mersin, Turkey; [16]Ankara City Hospital, Pediatric Pulmonology, Ankara, Turkey; [17]Ankara University Faculty of Medicine, Pediatric Pulmonology, Ankara, Turkey; [18]Karadeniz Technical University, Pediatric Infectious Disease, Trabzon, Turkey; [19]Gaziantep University Faculty of Medicine, Pediatric Allergy and Immunology, Gaziantep, Turkey; [20]Celal Bayar University Faculty of Medicine, Pediatric Pulmonology, Manisa, Turkey; [21]Sarıyer Hamidiye Etfal Hospital, Pediatric Pulmonology, İstanbul, Turkey; [22]İnönü University Faculty of Medicine, Pediatric Allergy and Immunology, Malatya, Turkey; [23]Ondokuz Mayıs University Faculty of Medicine, Pediatric Gastroenterology, Samsun, Turkey; [24]Balıkesir University Faculty of Medicine, Pediatric Pulmonology, Balıkesir, Turkey; [25]Ege University Faculty of Medicine, Adult Pulmonology, İzmir, Turkey; [26]Fırat University Faculty of Medicine, Pediatric Allergy and Immunology, Elazıg, Turkey; [27]İstanbul University, İstanbul Faculty of Medicine, Pediatric Allergy and Immunology, İstanbul, Turkey*

Objectives: Decline in pulmonary functions is a predictor of disease progression in patients with CF. This study aimed to determine the annual variability of percent predicted Forced Expiratory Volume in 1 second ($ppFEV_1$), and to investigate the risk factors related with the change in $ppFEV_1$, using national Registry data.
Methods: A retrospective cohort study of all CF patients over 6 years old with pulmonary functions data over at least 2 years of follow-up were extracted from the national CF Registry for years 2017 through 2019. Patients were classified according to disease severity and age groups. Multivariate analysis was used to predict the annual change in $ppFEV_1$ and to investigate the associated risk factors.
Results: Of the 574 children, the median age was 12 (IQR = 9–16) years. Overall mean annual $ppFEV_1$ decline was −0.97% (95%CI = −0.02 to −1.92%) and −4.2% (95%CI = −8.5 to 0.1%) in patients in the age group 20–24 years and −2.77% (95%CI = −6.9 to 1.4%) over 25 years old. The mean age at diagnosis was older ($p = 0.003$) and height for age, weight for age and body mass index z score ($p < 0.001$) were lower in the severe group. The mean change of $ppFEV_1$ was significantly higher in the group with $ppFEV_1 \geq 70$ compared with the other ($ppFEV_1 < 40$ and $ppFEV_1$: 40–69) 2 groups ($p = 0.004$). Chronic *P. aeruginosa* colonisation was a siginificant risk factor (OR:1.79; 95%CI = 1.26–2.54); however, genetic analysis, age at diagnosis, body weight, height, body mass index z score, and CF-related complications were not statistically significant risk factors for FEV_1 decline.
Conclusions: National Registry data analysis revealed the important effect of growth parameters on pulmonary functions. Close follow-up is critical

for patients over 20 years to slow lung disease progression, for those with normal initial ppFEV$_1$ levels, in particular. The association between chronic *P. aeruginosa* colonisation and lung disease progression calls attention to early interventions for *P. aeruginosa*.

P061

Low FEV$_1$ performance in the 30–34 years of age cohort in Norway: utilising Registry data in the search for causes

A. Os[1], H. Alderslyst[1], B. Akselsen Grøterud[1], P.L. Finstad[1], E. Bakkeheim[1]. [1]*Oslo University Hospital, Norwegian Resource Sentre for Cystic Fibrosis, Oslo, Norway*

Objectives: Norwegian CF Registry data has shown a low FEV$_1$ performance for the 30–34 year age-cohort in the years 2016–18. However, in 2019 the FEV$_1$ performance for this group improved significantly. We analysed data from the Norwegian CF Registry in the search for causes of the poor FEV$_1$ results and the sudden improvement.
Methods: Data from 2018 and 2019 in the Norwegian CF Registry regarding the age-span 25–39 years were extracted. Lung-TX patients were excluded. The following Registry parameters were both sufficiently reported and deemed interesting for further analysis: gender, PERT, CFTR modulator treatment, F508-homozygosity, diabetes, hemoptysis and CF liver disease. Further, the patient composition was assessed to determine the proportion of common and unique patients in the 30–34 year cohort for 2018 and 2019.
Results: Median FEV$_1$ in %-predicted for the age-cohorts 25–29, 30–34 and 35–39 years were 76, 48 and 73 for 2018 and 77, 69 and 66 for 2019. The number of patients was 27, 15 and 21, and 29, 14 and 19, respectively. Regarding the 30–34 year cohort and the specific parameters listed above; there were no significant changes between 2018 and 2019. In 9 patients that were common for 2018 and 2019 a median FEV$_1$change of −3.8% points was found. Six patients were unique to 2018 (4 moved to the next age-cohort, 1 died and 1 was lost to follow-up) and had 4% points higher, whereas 5 patients were unique to 2019 (2 from the age-cohort below and 3 new to the Registry) and had a 43%-point higher median FEV$_1$ value as compared to the 2018 overall median, respectively.
Conclusion: No specific Registry parameter was identified as a plausible reason for the low FEV$_1$ and the improvement seen in 2019. However, the low number of patients combined with missing data prevented analysis for several parameters. Patient mobility in and out of the age-cohort between 2018 and 2019 was substantial and the main contributor to the improvement in FEV$_1$ preformance.

P062

An international survey: understanding the health and perspectives of people with cystic fibrosis (CF) not benefitting from CFTR modulators

E. Kramer-Golkinkoff[1], A. Camacho[1], L. Kramer[1], J. Taylor-Cousar[2]. [1]*Emily's Entourage, Merion Station, United States;* [2]*National Jewish Health, Internal Medicine and Pediatrics, DENVER, United States*

Objectives: While the advent of cystic fibrosis transmembrane conductance regulator (CFTR) modulator use has improved daily life and long-term prognosis of CF for many with approved CFTR mutations, approximately 10% of people with CF (pwCF) have only symptomatic treatments available. We sought to understand the demographics, clinical characteristics, care perceptions, treatment needs, mental health impacts of lack of access, and clinical research participation interest of those unable to access or tolerate modulator therapy.
Methods: Between 10 June and 1 July 2021, Emily's Entourage developed and distributed a 38-question anonymous survey targeted at pwCF not benefitting from approved modulators; distribution occurred via social media and email to pwCF and advocacy groups in and outside the US.
Results: There were 431 survey respondents representing pwCF on five continents. Most survey respondents were parents of pwCF (43.4%) or adults with CF (42.1%). The majority of pwCF had moderate lung disease (50.3%). Ineligibility based on CFTR mutation (64.1%) was the most frequently reported reason that pwCF were not on modulators. PwCF reported that the most impacted aspects of life were mental (66.7%) and physical (40.7%) health. Financial concerns and feelings of isolation were commonly reported. Witnessing improvements for peers with access to modulators was simultaneously uplifting and disheartening for many. The majority of pwCF said they would be interested in participating in future clinical research (77.6%), although some living outside of the US cited lack of opportunity to participate in clinical trials as a barrier.
Conclusions: PwCF not able to benefit from CFTR modulators have a high burden of disease impacting their physical and mental health. While most are happy for those who are benefiting from modulators, they are eager for the opportunity to experience similar improvements for themselves, and willing to participate in clinical trials of new therapies.

P063

Physiotherapy data for the UK cystic fibrosis Registry - review and re-launch

L. Morrison[1], M. Yip[2], O. Tomlinson[3], C. Brown[4]. [1]*Queen Elizabeth University Hospital, West of Scotland Adult CF Unit, Glasgow, United Kingdom;* [2]*Cystic Fibrosis Trust, UK CF Registry, London, United Kingdom;* [3]*University of Exeter Medical School, Exeter, United Kingdom;* [4]*West Midlands Adult Cystic Fibrosis Centre, Birmingham, United Kingdom*

Objectives: Data collection within the UK CF registry is valuable for research, however physiotherapy-related data was considered insufficient in evaluating benefits of interventions and standardising specialist services.
Methods: An electronic survey was sent to 47 ACPCF committee members, representing the wider UK physiotherapy community. The 10-point survey examined current data captured, clinical relevance, specificity and what data was considered lacking.
Results: 34 responses were received. If members belonged to the same centre, a single response was collated, reflecting a total response rate of 72%. 85% valued information on nebulisers, specifically device type and availability of adherence data. 100% wanted information on non-invasive ventilation (NIV) use, specifically type of device (56%), settings and humidification (47%) and use of NIV for acute exacerbation, as an airway clearance (ACT) adjunct or for long-term use. Exercise test outcome measures were considered essential, such as heart rate (82%), oxygen saturations (91%), VO_{2peak} (71%) and distance or level completed (74%); allowing annual comparisons both locally and nationally. The selection of ACT in combination was challenging. The opportunity to highlight different techniques used throughout the day or week was considered advantageous. 58% use the Manchester postural screening tool widely recommended by the ACPCF. More information regarding type of postural anomaly, if fixed or correctable, and requirement for treatment and follow-up was considered necessary.
Conclusion: Collaboration with the CF Trust registry over 2 iterations has allowed improvements in data collection and updated software, whereby clinically meaningful physiotherapy data, with modifications in exercise testing, postural screening, NIV and clearer information on primary and secondary ACT, are achievable. We will support the rollout through the national ACPCF with training and further audit and the inclusion of sinus management in future updates.

P064

Iron deficiency in cystic fibrosis: a prospective study in a modern adult cohort

H. Lobbes[1], S. Durupt[2], B. Pereira[1], S. Mainbourg[2], R. Nove-Josserand[2], I. Durieu[2], Q. Reynaud[2]. [1]*CHU de Clermont Ferrand, Médecine interne, Clermont Ferrand, France;* [2]*Hospices Civils de Lyon, Groupe Hospitalier Sud, adult CF center, Pierre bénite, France*

Objectives: Iron homeostasis is a crucial issue in cystic fibrosis (CF) because of its close relationship with *Pseudomonas aeruginosa* colonisation. Iron deficiency (ID) diagnosis in CF is challenging because of frequent systemic inflammation. We aimed to determine the prevalence and risk factors of ID in adult patients with CF.
Methods: A monocentric prospective study was performed in a referral centre for adult CF patients. Clinical and biological parameters were collected during the annual medical check-up. Apparent exacerbation was an exclusion criterion. ID was defined by transferrin saturation ≤16% or ferritin ≤20 (women) or 30 (men) µg/L or ≤100 µg/L in the case of significant systemic inflammation.