Objective data such as spirometry and weight to be uploaded to a web-based portal.

**Methods:** The study recruited 18 patients about to start on triple therapy. Patients were brought into the clinic, and baseline data including height, weight, lung function, sputum volume, sputum culture, and routine blood tests were collected. Initial data on sputum volume and quality of life were recorded on an app (PatientMpower) on the patient’s smartphone. Weighing scales and a spirometer were connected to the smartphone app via Bluetooth. At 3-month intervals, a virtual consultation took place with our study coordinator. Weight and lung function were measured and downloaded to the app. Lung function was assessed using the Mir Spirobank Smart Handheld spirometer. Lung function was also measured in the lab at baseline and between 6 and 9 months. Weight was recorded at the patient’s home using a Bodytrace scale, and the results were transmitted to the PatientMpower app. Patients recorded sputum volume on the app at baseline and 3-month intervals. The Cystic Fibrosis Respiratory Symptom Diary (CFRSD) was recorded on the app, and the Cystic Fibrosis Questionnaire Revised (CFQ-R) was completed.

**Results:** Eighteen patients completed the study (4 female, 14 male) and were included in the analysis. Fifteen were homozygous for F508del, and three were heterozygous and therefore naive to CFTR modulators. The absolute change in percentage predicted forced expiratory volume in 1 second was 16.2 ± 16.0% (p < 0.001). Weight also increased by up to 4.2 ± 3.4 kg (p < 0.001). Mean reduction in sputum volume per day was 22.9 ± 15.8 mL. At baseline, more than half of patients had sputum production in excess of 30 mL per day, but after initiation of CFTR triple therapy, sputum production fell to much lower volumes for at least half and remained at negligible levels for 12 months. Mean improvement in CFQ-R was 17.6 ± 12.8 points. CFRSD score also improved significantly, falling by 20.3 ± 11.9 points. (A change of 4 points is considered to be clinically significant).

**Conclusions:** The excellent results seen in our study mirror those of phase III clinical trials. The reduction in sputum volume, albeit subjective, was impressive and has not been reported quantitatively before to our knowledge. The smartphone app was particularly useful in providing accurate data remotely and would allow for more frequent and convenient opportunities to capture clinical data from patients without having to come to the study center.

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**Change in pulmonary function after introduction of elexacaftor/tezacaftor/ivacaftor: Results from the national cystic fibrosis cohort in Denmark**

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**Background:** Triple modulator treatment has improved lung function in people with cystic fibrosis (CF) after just few months of treatment. Initially introduced through trials and compassionate use programs in May 2018, Denmark was among the first countries to attain market authorization in September 2020, and elexacaftor/tezacaftor/ivacaftor (ELX/TEZ/IVA) was introduced in the following months. Denmark has the greatest prevalence of the F508del mutation in Europe (95% homo- or heterozygote), with most people with CF being eligible for treatment with triple modulators. Routine CF care involves close monitoring of spirometry data obtained at every consultation. This provides high-resolution data for monitoring disease progression over short time spans, as well as pre-modulator observation time for longitudinal evaluation. Our objective was to evaluate changes in percentage predicted forced expiratory volume in 1 second (FEV1/pp) for 12 months after implementation of ELX/TEZ/IVA in the Danish population.

**Methods:** This was a national observational cohort study at the two Danish CF centers in a real-world set-up to evaluate the impact of ELX/TEZ/IVA on lung function. Study participants included all people with CF in Denmark, including Greenland and the Faroe Islands, eligible for CF transmembrane conductance regulator (CFTR) triple modulator treatment. Inclusion criteria are aged 12 and older, at least one F508del mutation, no solid organ transplantation, and a CF diagnosis and F508del status verified by sweat test and gene sequencing. The primary endpoint is FEV1/pp. Subgroup analyses include prior modulator treatment, chronic lung infection, and birth cohort. FEV1 was measured at study visits at treatment initiation and 1-, 3-, 6-, 9-, and 12-month follow-up, in addition to routine monthly contacts. Spirometry was performed using a Carefusen Jaeger Vynus Spiro spirometer. FEV1/pp was calculated using the Global Lung Initiative reference equations. For statistical analysis, patients contributed individual observation time, and impact of treatment was assessed using a longitudinal mixed-effects model.

**Results:** As of September 2022, 300 patients (n = 145 male) have been treated for 12 months, of whom 194 (65%) switched from modulator treatment. At treatment initiation, median age was 25 (interquartile range 18–35), mean body mass index was 21.4 ± 3.6 kg/m2, mean FEV1/pp was 74.9 ± 25.3%. 221 (74%) were homozygous for F508del. At the time of abstract submission, 1780 FEV1 measurements after initiation of triple modulator treatment had been collected (6.0 ± 2.3 per subject). In addition, 10,258 FEV1 measurements (34.0 ± 15.8 per subject) are available for analysis of observation time before treatment with ELX/TEZ/IVA. All eligible patients for triple modulator therapy have completed baseline data collection.

**Conclusions:** The final data at 1-year follow-up will have been collected and analyzed for the conference.

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**Exploring the impact of highly effective modulator therapy on sleep in children with cystic fibrosis**

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**Background:** Cystic fibrosis (CF) is an autosomal-recessive chronic respiratory disorder that affects approximately one in 3200 live births in Caucasians. Obstructive sleep apnea (OSA) is increasingly being identified as a comorbid condition of consequence in individuals with chronic lung disease, including those with CF. Widespread use of highly effective modulator therapy (HEMT), including ivacaftor and elexacaftor/tezacaftor/ivacaftor, has led to dramatic improvements in nutritional status, lung function, and quality of life. The objective of this study was to explore the effects of HEMT on sleep in children with CF.

**Methods:** This was a retrospective analysis of polysomnographic data and medical records from 2009 to 2022 in children with CF followed at the Children’s Health Pediatric CF Center in Dallas, Texas.

**Results:** Polysomnographic data were available for 49 children with CF. Mean age of the subjects at the time of polysomnography was 7.0 ± 4.4; 87.8% were white, 8.2% were Black, and 28.6% were Hispanic; and 79.6% had at least one copy of the delF508 mutation. Twenty-eight (57.1%) were on HEMT at the time of the study, and five (10.2%) were on a different combination. A diagnosis of OSA was present in 36.7% of subjects with CF. Sleep duration was shorter in those with OSA (378.3 minutes) than those without (428.2 minutes). Sleep onset latency was longer (41.9 vs 32.4 minutes) and sleep efficiency was poorer (80% vs 86%) in those with OSA. The OSA group had a lower body mass index (BMI) percentile than the non-OSA group (54th vs 62nd percentile). Of the 28 subjects on HEMT, only four (14.3%) were in the OSA group (54th vs 62nd percentile). Of the 28 subjects on HEMT, only four (14.3%) were in the OSA group (54th vs 62nd percentile).

**Conclusions:** With HEMT affording improvements in lung health, nutritional status, and overall life expectancy, a new comorbid condition, OSA, is quickly emerging in people with CF. In our limited sample, we found an increase in OSA frequency after use of modulators. A larger study is needed to elucidate the impact of HEMT on frequency and severity of OSA.