Methods: Eligible participants aged 2–17 years old were asked to complete measures at home for 6-months. KDM-CF provides a score from 0%-100% in 4 subgroups and a total score and was completed by parents of participants aged ≥6 years at visit 1. KDM-CF scores of parents and participants were analysed with the percentage of completed data received at the end of the 6 months.
Results: 148 participants were recruited; 5 withdrew before enrolment. 98 children were aged ≥6; paired scored questionnaires were received for 70. Median (IQR) age was 11(7.8–13.85) years; 41/70(58.6%) were female. Median (IQR)% of requested measures completed over the 6 months was 29.1(10.8–49.7)%. Median (IQR) participant KDM-CF score was 62.9(48.6–73.1)% compared to parental scores of 74.3(67.9–82.9)% ($p < 0.0001$). There was no correlation between participant or parent KDM-CF scores and proportion of measures completed. Those aged 6–10 had significantly higher scores than those aged ≥11 (67.3[52.9–77.9]% v 54.3[40.7–70.7]% $p < 0.05$). The lowest subcategory score for both parents and participants was nutrition.
Conclusion: Knowledge of CF did not impact the percentage of completed home monitoring measures, suggesting this factor may not affect engagement in remote monitoring. The median completion percentage of this subgroup is lower than the CLIMB-CF study cohort (29.1% v 40.1% [ns]) which likely represents the higher percentage of adolescents. In both groups, the nutrition subgroup had the lowest scores which indicates a need for ongoing education.

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Adherence in the use of digital technology for home monitoring in patients with cystic fibrosis

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Objectives: Evidence for using telehealth in Cystic Fibrosis (CF) is growing rapidly. A study evaluating digital technologies in the management of CF adults showed an improvement in patients' quality of life. The behavior of CF patients and their adherence regarding these new approaches need further investigations. The aim was to assess patient's adherence of using the NuvoAir Home platform as part of a virtual model of care.
Methods: The NuvoAir Home platform consists of a smartphone application, Bluetooth spirometer and clinician portal. Patients and/or parents were trained to use the platform and asked to do home spirometry monthly. We calculated and tested the difference of adherence between pediatric and adult CF group using Wilcoxon rank-sum test. To test for significant associations between adherence and other variables, we transformed the adherence variable using inverse-normal transformation and tested in a linear regression within the CF pediatric and adult patients separately.
Results: Thirty-six children CF patients (19 females; mean age 15.2 ± 3.2; 9 homozygous for delta F508; FEV_1 88.0 ± 11.3% predicted) and forty-three adult CF patients (26 females; mean age 31.6 ± 6.8; 16 homozygous for delta F508; FEV1 48.4 ± 16.3% predicted) were recruited from Federico II Hospital, Naples, Italy. All patients had completed six months follow-up. The mean home spirometry performed by pediatric and adult patients was 0.68 ± 0.46 and 0.57 ± 0.28, respectively. Although the mean adherence is a slightly higher in pediatrics than adults, there is no significant difference between the two groups, $p = 0.41$. The adherence is not significantly associated with age, sex, FEV1% predicted or FVC % predicted.
Conclusion: Pediatric and adult CF patients performed similar average amount of spirometry per month. Both groups underuse digital technology for home monitoring, so new strategies should be applied to improve patient adherence.

Physiotherapy

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Effects of the treatment with elexacaftor/tezacaftor/ivacaftor on aerobic fitness of adolescents with cystic fibrosis

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Objective: Treatment with elexacaftor-tezacaftor-ivacaftor (ELE/TEZ/IVA) has proved effective, although little is known about its effects on aerobic fitness. This study aimed to evaluate the effects of treatment with ELE/TEZ/IVA on lung function, aerobic fitness, body mass index (BMI) and physical activity levels of cystic fibrosis (CF) adolescents over a period of 6 months.
Methods: A prospective, ecological, study including patients with at least one F508del mutation, aged 14 to 18 years, was conducted. Demographic, anthropometric, lung function, daily physical activity (PAQ-A questionnaire) and aerobic fitness (cardiopulmonary exercise testing) data were collected immediately before the start of ELE/TEZ/IVA and at 6-months of follow-up. Control data from 1-year before the start of therapy (under the use of TEZ/IVA) was also used for comparisons. Peak oxygen consumption (VO_2peak), ventilatory equivalents of O_2 (V_E/VO_2) and CO_2 (V_E/VCO_2), and ventilatory threshold (VT_1) data were measured. Ethical approval was acquired and data were analyzed by repeated measures ANOVA with the Bonferroni post-hoc test.
Results: Ten patients (70% males), mean age 16.3 ± 1.6 years and mean BMI z-score of –0.43 ± 0.70 were included. A significant increase in the z-score of FEV_1 was seen when comparing the start (–1.7 ± 1.9) and 6 months of therapy (–0.8 ± 1.7). No significant differences were found in both the BMI and PAQ-A score. VO_2peak (39.6 ± 6.2 vs. 34.6 ± 6.7 mL/kg/min) decreased and V_E/VCO_2 at both VT_1 (24.6 ± 4.0 vs. 28.7 ± 2.1) and peak (27.2 ± 5.1 vs. 33.4 ± 4.8) increased when 6 months of treatment was compared to the control period. A significant decrease was also found for the percent of VO_2peak at VT_1 (63.4 ± 14.7 vs. 51.1 ± 8.3) when comparing the start and 6 months of therapy.
Conclusion: Results have shown an increase in lung function after the initiation of ELE/TEZ/IVA therapy that was not followed by BMI or aerobic fitness levels.

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Physical activity and sedentary behavior in cystic fibrosis patients before and after triple modulator therapy: a single-centre retrospective study

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Objectives: Regular physical activity (PA) and reduced sedentary behavior (SB) are associated with health benefits and a greater life expectancy in cystic fibrosis (CF) patients. This retrospective cohort study aimed to investigate whether tezacaftor-ivacaftor-elexacaftor had an impact on daily PA levels and SB in patients with CF.
Methods: 18 CF patients participated in the study. PA, SB and lung function data were measured with accelerometry (for 7 consecutive days) and spirometry respectively. Routine clinical assessments were at baseline (T0), 1 month (T1), 3 months (T3) and 6 months (T6) after receiving treatment. Paired sample t-test was used to show differences, in the lung function and PA data, between T0 and the different measurements (T1, T3 and T6).
Results: Mean Forced expiratory Volume in 1 s (pFEV1) significantly increased in all measurements compared to T0 (68.4%): +9.8% at T1, +13.4%