Background: Physical activity has been shown to have a positive impact on cystic fibrosis (CF) symptoms and slows decline in lung function. Additionally, moderate-intensity activities have been shown to improve sputum expectoration and oxygen saturation in children with CF, improving symptoms [1]. This study assessed whether using fitness trackers and providing a daily step goal would increase physical activity in hospitalized children with CF.

Methods: This pre-post pilot study included participants aged 6 to 21 who were admitted to the hospital for a pulmonary exacerbation of CF between October 2020 and November 2021. Garmin vivosmart 4 wrist-based activity trackers were issued, and baseline data were tracked and analyzed for the first 2 study days. Pre-goal number of steps was defined as average number of steps taken on those 2 days. On study day 3, a step goal and menu of activities designed to increase physical activity were shared with the participants. Steps after goal setting were defined as average number of steps taken on study day 3 and beyond while hospitalized, excluding day of discharge. Data collected from the medical record and the activity tracker web-based profile included daily oxygen requirement, daily step count, sleep duration, overnight pulse oximetry levels, resting heart rate, calories expended, and intensity minutes. The primary outcome of change in daily steps and attainment of step goals was analyzed using descriptive statistical testing, means, and standard deviations. Outpatient data were analyzed for only the first week after discharge because of poor adherence to wearing the device. Although our results show a positive impact, further research is needed to determine the effect such an intervention would have on a larger scale. Future directions of research include determining potential clinical benefit from increased activity during hospitalization and prolonged follow-up to assess long-term benefits of intervention.

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Reference

Cystic fibrosis fitness during inpatient treatment
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Results: Eight participants aged 6 to 18 completed the study. Hospital length of stay ranged from 4 to 14 days. Participants took an average of 1508 ± 1078 steps before goal setting, which increased to an average of 3704 ± 1555 steps after the intervention. Step goals were met 67% of the time (10-year-old girl, 17-year-old girl, 14-year-old boy). During the intervention, patients met their step goal 56% of the time, 60% (10-year-old girl), 50% (17-year-old girl), 75% (14-year-old boy), two met it 0% of the time (18-year-old boy, 17-year-old girl), one met it 44% of the time (6-year-old boy), and the remaining three met it 67% of the time (10-year-old girl, 17-year-old girl, 14-year-old boy).

Conclusions: This intervention shows promise, with daily number of steps doubling from baseline during the intervention period. There was great variability among participants, suggesting that the approach helps some more than others. Enrollment was initially planned for 20 patients, but the SARS-CoV-2 pandemic and changes to CF therapy reduced hospitalizations during the study period. Outpatient data were analyzed for only the first week after discharge because of poor adherence to wearing the device. Although our results show a positive impact, further research is needed to determine the effect such an intervention would have on a larger scale. Future directions of research include determining potential clinical benefit from increased activity during hospitalization and prolonged follow-up to assess long-term benefits of intervention.

Table 1 (abstract 123):
Clinical indicators before and after modulator therapy

<table>
<thead>
<tr>
<th></th>
<th>Prior to Therapy</th>
<th>After Therapy</th>
<th>P-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>First of two sweat chlorides</td>
<td>104.50 (100.00, 112.00)</td>
<td>47.00 (36.00, 54.00)</td>
<td>&lt;0.0001</td>
</tr>
<tr>
<td>ppFEV1 (baseline vs six-months)</td>
<td>27.50 (25.00, 35.00)</td>
<td>42.00 (29.00, 49.00)</td>
<td>&lt;0.0001</td>
</tr>
<tr>
<td>Weight (baseline vs six-months)</td>
<td>50.20 (42.20, 63.40)</td>
<td>58.70 (48.00, 69.10)</td>
<td>0.006</td>
</tr>
<tr>
<td>BMI (baseline vs six-months)</td>
<td>19.15 (16.40, 22.10)</td>
<td>20.60 (18.50, 23.20)</td>
<td>0.0014</td>
</tr>
<tr>
<td>Hospital admissions</td>
<td>4.50 (3.00, 7.00)**</td>
<td>0.50 (0.00, 2.00)**</td>
<td>n/a</td>
</tr>
</tbody>
</table>

*Completed post-drug initiation (earlier than 12 months). **24 months before and after therapy initiation.
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 ± 10.6% (p < 0.001). Mean 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 objective data such as spirometry and weight to be uploaded to a web-based portal.

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

127 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 tezacaftor/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 modulator. 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 in 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 (94th vs 62nd percentile). Of the 28 subjects on HEMT, only four underwent polysomnography after initiation of HEMT. Frequency of OSA after HEMT was higher than before HEMT in our limited sample (50% vs 33.3%). Polysomnographic data were available after therapy for all five subjects on a non-HEMT modulator, 60% of that group was diagnosed with OSA.

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.