when their menstrual cycle started and were sent RedCap surveys to assess respiratory symptoms at different menstrual cycle phases over a 6-month period. Time points correlated with the different phases of the menstrual cycle: day 1 (start of follicular phase, menstrual period), day 13 (ovulation phase), and day 21 (luteal phase). Ten symptoms were evaluated to calculate a Respiratory Symptom Menstrual Cycle (RSMC) score. Self-reported symptoms were compared with their subjective baseline; improved from baseline (−1); at baseline (0); and mildly (1), moderately (2), and severely increased from baseline (3). Total symptom scores ranged from −10 to 30. We determined changes in RSMC score between points in their cycles and constructed a mixed-effects linear regression model to explore the association between menstrual cycle day (13 vs 21 vs 1) and total symptom score (dependent variable, RSMC score; independent variable, menstrual cycle day; random effect, patient).

**Results:** Despite multiple contact attempts, only seven of 13 WwCF who provided consent completed at least three surveys. We analyzed data from these 7 patients. Participants were aged 20 to 45 and had a mean body mass index of 22.6 kg/m² and mean percentage predicted forced expiratory volume in 1 second of 71.5%, all had chronic *Pseudomonas* infection, 86% had at least one copy of F508del, and 86% were on highly effective modulator therapy (HEMT). The most common reported symptom changes were cough, sputum, and overall sputum quality, each reported in 57% of patients. There was a large variation in overall RSMC scores across participants, with no clear worsening for the cohort at any point in the cycle. There was no statistical association between day and RSMC score when each day was compared with day 1, day 13, or day 22 (day 13 vs day 1, regression coefficient 0.00, 95% CI, −1.77 to 1.77, p > 0.09; day 21 vs day 1, regression coefficient −0.41, 95% CI, −2.15 to 1.32, p = 0.64) (Figure 1).

**Conclusions:** Our results show that there is large variation in RSMC scores between WwCF, with no association between RSMC score and different time points of the menstrual cycle. Poor survey response, resulting in a small sample size, limited our study. Most patients had been started on HEMT, which decreased their overall respiratory symptoms. Online surveys may not be the best way to engage participants; tracker phone applications may be a better option for several cycles. Larger studies are needed to evaluate the impact of sex hormones on respiratory symptoms and other physiologic parameters in WwCF to understand better the gender gap.

**References**


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**Figure 1.** Respiratory symptom menstrual cycle total score stratified according to patient at days 1, 13, and 22 of their menstrual cycle.
Conclusions: These seven patients with N1303K and a second nonresponsive mutation demonstrated significant clinical improvement after treatment with ELX/TEZ/IVA. A controlled clinical trial is needed to confirm these results and allow people with the N1303K mutation to register and be reimbursed.

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Effect of hypertonic saline therapy on lung clearance index in preschool children with cystic fibrosis

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Background: Hypertonic saline (HS) may improve mucociliary clearance in the context of long-term maintenance therapy in people with cystic fibrosis (CF). We aimed to assess the effect of inhaled HS on the lung clearance index (LCI2.5) and percentage predicted forced expiratory volume in 1 second (FEV1pp) in children aged 4 to 6 with CF.

Methods: The study was conducted at the Division of Pediatric Pulmonology, Marmara University, between May and October 2021. Participants, who had a confirmed diagnosis of CF between the ages of 48 and 72 months, were divided into two groups as 7% HS users and 0.9% isotonic saline (IS) users. LCI2.5 was measured according to nitrogen multiple-breath washout, FEV1pp were measured, and the Pediatric Quality of Life Inventory (PedsQL) was administered at baseline and week 24.

Results: There were 16 children each in the HS and IS groups. All were on dornase alpha treatment. Cough was reported as an adverse event in nine children in the HS group. Mean LCI2.5 was 9.4 ± 3.1 at baseline and 8.0 ± 1.9 at week 24 in the HS group (p = 0.01) and 8.2 ± 1.6 at baseline and 7.8 ± 2.1 at week 24 in the IS group (p = 0.42). There was no significant change in FEV1pp between baseline at week 24 in either group (p > 0.05). Scores on the physical domain of the PedsQL in the IS group increased between baseline and week 24 (p = 0.04).

Conclusions: In this group of preschool-aged children with CF, treatment with HS for 24 weeks was associated with significantly greater improvement in LCI2.5 than treatment with IS.

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Electronic home monitoring of children with cystic fibrosis to detect and treat acute pulmonary exacerbations and its effect on 1-year FEV1 loss

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Background: People with cystic fibrosis (CF) have recurrent respiratory tract infections and acute pulmonary exacerbations (PEx), which cause loss of lung function and thus decrease life expectancy. One sign of PEx is decline in forced expiratory volume in 1 second (FEV1). Early detection of impairment in pulmonary function tests (PFTs) allows early diagnosis and treatment of PEx. We aimed to investigate the effect of electronic home spirometry in people with CF on number of PEx and 1-year change in percentage predicted FEV1pp.

Methods: This was a randomized, 1-year, prospective study. Children with CF aged 6 to 18 who could perform spirometry and had a smartphone were included. Eligible children were randomized into two groups: home spirometry (HSG) and usual care (UCG). HSG participants performed two PFTs per week, and a registered CF nurse evaluated the results for decline in FEV1; simultaneously. Number of PEx, days in hospital for PEx, and days on oral antibiotics for PEx were evaluated from patients’ records and compared with data from the previous year. A health-related quality of life questionnaire for children with CF who was administered and lung clearance index (LCI) was measured at the beginning and end of the study.

Results: Each group included 30 patients; 22 in the HSG and 24 in the UCG completed the study. Median age was 13.5 (interquartile range (IQR) 11.7–14.9) in the HSG and 12.7 (IQR 10.6–15.5) in the UCG. Median 1-year change in FEV1pp was 0.95 (IQR −2.61–6.73) in the HSG and −0.41 (IQR −3.68–3.09) in the UCG (p = 0.27). The results are summarized in Table 1.

Conclusions: Electronic home monitoring of children with CF using spirometry and early treatment of PEx may result in slower decline in lung function, which would be likely to have a beneficial effect on QOL. Full results, including QOL analysis, will be available before the conference.

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Long-term clinical impact of a virtual model of care in cystic fibrosis

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Background: Health professionals have explored and tested the use of digital technologies in managing chronic respiratory diseases remotely during the COVID-19 pandemic. Further investigation into whether these new approaches to care delivery provide an opportunity to improve cystic fibrosis (CF) management is needed. The aim of this study was to assess the long-term clinical impact of use of e-health as part of a virtual model of care in CF.