Conclusions: These seven patients with N1303K and a second nonrespon-
sive mutation demonstrated significant clinical improvement after treat-
ment 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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134 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) aged 7 and older. 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 (FEV1,pp) 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, FEV1,pp 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 9.2 ± 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 FEV1,pp 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 improve-
ment in LCI2.5 than treatment with IS.

135 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 FEV1,pp.

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 (QOL) questionnaire for children with CF 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 FEV1,pp 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.

136 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.