Hull Airway Reflex Questionnaire (HARQ) as patient-reported outcome measures (PROMs) to assess the effect of triple CFTR modulators on sino-nasal and laryngoosophageal reflux symptoms. Questionnaires, lung function, and weight were recorded at baseline before starting treatment and after 6 months of treatment.

Results 32 patients (23 male) starting elexacaftor/tezacaftor/ivacaftor were studied. Their baseline characteristics were mean age 34.3 (range 20–65) years, FEV1% predicted of 24.8 (11–40), BMI 21.28 kg/m² (13.2–31.1). All patients continued with treatment throughout the study period. At 6 months there was an improvement in mean FEV1% predicted of 8.63 and BMI 2.6 kg/m². Patient-reported outcomes measures showed significant improvement (table 1): median scores RSI 10, HARQ 19.5 and SNOT 16 (p<0.001 for all outcomes).

Discussion This study shows significant improvement in lung function, weight and sino-nasal and laryngopharyngeal reflux PROMs in patients with advanced CF. The SNOT-20, RSI and HARQ scores showed improvement that exceeded recognised clinically significant changes in these metrics.

Abstract S58 Table 1 Measured values at baseline and after 6 months’ treatment.

<table>
<thead>
<tr>
<th></th>
<th>Baseline</th>
<th>6 months</th>
<th>Difference</th>
</tr>
</thead>
<tbody>
<tr>
<td>ppFEV1</td>
<td>24.8 (7.13)</td>
<td>33.4 (10.33)</td>
<td>8.6 3</td>
</tr>
<tr>
<td>BMI</td>
<td>21.3 (4.13)</td>
<td>23.9 (4.29)</td>
<td>2.6 3</td>
</tr>
<tr>
<td>RSI</td>
<td>15 (10.75–23)</td>
<td>5 (2.25–7)</td>
<td>10 3</td>
</tr>
<tr>
<td>HARQ</td>
<td>26.5 (16–39)</td>
<td>7 (3.75–12.25)</td>
<td>19.5 3</td>
</tr>
<tr>
<td>SNOT-20</td>
<td>36.5 (22–42)</td>
<td>20 (10–31.25)</td>
<td>16.5 3</td>
</tr>
</tbody>
</table>

1 mean (Standard Deviation)
2 median (IQR)
3 p<0.001

Abstract S59 Figure 1 Box plot comparing average adherence of nebulised therapy before and after Kaftrio initiation

47 patients included in analysis. 31 patients (65%) reduced their adherence to nebulised therapies following Kaftrio use. Median nebulised therapy adherence dropped from 65% to 42% (p<0.003, Wilcoxon Signed Rank) pre and post Kaftrio initiation respectively (figure 1). Of the 47 patients, 28 (60%) communicated a decision to change therapy with the CF team, while 19 (40%) did not communicate this change.

Discussion Our data demonstrates a reduction in nebulised therapy adherence after Kaftrio initiation. Decisions to reduce adherence were often patient driven and not disclosed to clinicians.

Our findings underline the importance of including objective measures of adherence to inhaled therapies in the design of CFTR modulator studies.

The lack of CFHH uploads for 34 patients highlights the challenges in monitoring adherence in clinical practice; in our experience, these patients were less adherent to treatment.

We plan to conduct a qualitative study to explore factors influencing patient decisions to stop or continue medication.

Abstract S60 OBSERVATIONAL STUDY OF IVACAFTOR IN PEOPLE WITH CYSTIC FIBROSIS AND SELECTED NON-G551D GATING MUTATIONS: FINAL RESULTS FROM VOCAL

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Introduction and Objectives VOCAL, a Phase 4 observational trial (NCT02445053), assessed real-world effectiveness of ivacaftor (IVA) in people with cystic fibrosis (pwCF) with ≥1 non-G551D gating mutation (G178R, S549N, S549R, G551S, G1244E, S1251N, S1255P or G1349D).

Methods pwCF aged ≥6 years in Italy, the Netherlands and the UK who were IVA-naïve or on IVA for ≤18 months at enrolment were eligible. Data were recorded for 12 months pre-IVA and up to 48 months after enrolment. Continuous outcomes (e.g. percent predicted forced expiratory volume in 1 second [ppFEV1]), body mass index (BMI) were assessed from baseline (the last pre-IVA value recorded) in 6-month intervals up to 48 months post-IVA using a mixed model for...