

6.06 Effect of Elexacaftor-Tezacaftor-Ivacaftor on a West of Ireland cohort of patients with Cystic Fibrosis (CF) and comparison of “smart” monitoring of patients compared to traditional lab and clinic based measurements

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CF is a rare autosomal recessive multisystem disorder secondary to cystic fibrosis transmembrane protein (CFTR) gene dysfunction. ETI has been shown to improve a range of outcomes in CF patients internationally. We performed a prospective, non-randomized, non-controlled study to assess if ETI had similar effect in our real life West of Ireland cohort over a 9 month period. Data was collected in the traditional method but also remotely using a novel monitoring mobile phone app in conjunction with smart hand-held spirometers and weighing scales. We aimed to assess the accuracy of this form of remote monitoring compared to traditional laboratory monitoring. 18 patients commencing ETI therapy were recruited to the study. Characteristics are outlined in table 1. Pre commencing ETI the mean predicted FEV1 in our cohort was 59.6% (± 18.8). This increased to 73.7% (± 23.2) ($p < 0.0001$) with therapy. ETI therapy also resulted in significant improvements in body mass index, sputum volume and in quality of life and symptom scores (table 1). Total combined days spent in hospital among the cohort fell from 434 days the year prior to starting ETI treatment to 39 days the following year. Our study confirmed the hand-held spirometer correlated strongly with those measured in the pulmonary function lab.

Conflict of Interest: None to declare

Table 1(6.6). Demographics and clinical characteristics pre-and-post treatment

Characteristics	Start of study	End of Study
Male sex: no. (%)	14 (78%)	
Age		
Mean: year (SD)	28.7 (± 8)	
Genotype		
Homozygous Phe508del- no. (%)	15 (83%)	
Heterozygote Phe508del- no (%)	3 (17%)	
Clinical characteristics		
Height- cm (SD)	170.6 (± 6.4)	
Weight- Kg (SD)	67.8 ± 12.3)	72.8 (± 11.8)
Body Mass Index- Kg/m ² (SD)	23.3 (± 3.9)	25.1 (± 3.9)
Percent Predicted FEV1- % (SD)	59.6% (± 18.8)	73.7% (± 23.2)
Sweat Chloride- mmol/L (IQR)	84 (66-101)	46 (32-58)
Sputum volume: ml/day (IQR)	28.8 (12-37.5)	1 (0-10)
CFRSD- no. (IQR)	34 (23-41)	14 (0-23)
CFQ-R- no. (IQR)	78 (66-83)	94. (92.3-100)

