

#### 6.04 The Use of Combination Therapy Drug KAFTRIO in the Treatment of Cystic Fibrosis, and How Pulmonary Function Test Results Can Significantly Change Post Therapy: The Role of the Physiologist.

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Cystic fibrosis (CF) is a multi-organ progressive genetic disease. Approximately 70-80% of CF mutations are Class II, linked to a specific deletion of three base pairs of DNA at position 508 of the gene, known as F508del. KAFTRIO, a combination drug, is utilized in the treatment of these patients. It increases CFTR protein quantity and function at the cell surface and restores cellular transport of sodium and water, reducing mucosal production.

Pulmonary Function Tests are the cornerstone in the clinical management of CF determining the severity of lung disease, capturing clinical course, evaluating therapeutic efficacy, and monitoring disease progression. Forced expiratory volume in 1 second (FEV<sub>1</sub>) being the established marker of CF. Respiratory physiologists play a pivotal role in the care and management plan of a patient by ensuring validity, reliability, and repeatability of PFT's. An emerging pattern of significant improvement was noted in PFT results of returning patients who had been prescribed KAFTRIO. A specific case study is presented in *table 1* showing a marked increase in PFTs.

*Table 1(6.4) – Pulmonary Function Test results, Weight and BMI pre- and post-KAFTRIO.*

<b>Date</b>	<b>FVC</b>	<b>FEV1</b>	<b>PEF</b>	<b>Weight</b>	<b>BMI</b>
29/10/2020 (Pre Kaftrio)	53% 2.66L	32% 1.66L	53	55kg	18.2
26/01/2022 (Post Kaftrio)	74% 3.65L	56% 2.37L	90	77kg	25.4

**Conflict of interest:** none to declare.