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| **Impact of elexacaftor/tezacaftor/ivacaftor on muscle strength and exercise capacity in adults with cystic fibrosis: real-world evidence from a large adult CF centre** |
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| **Introduction/Aim:**  Treatment with elexacaftor/tezacaftor/ivacaftor (ETI) improves multiple clinical outcomes in people with cystic fibrosis (pwCF). However, limited information about its effects on muscle strength and exercise capacity exists. This study evaluated the real-world impact of ETI on these outcomes in a group of pwCF following PBS listing in Australia in 2022.  **Methods:**  Prospective cohort design. Isometric Quadriceps strength test (QS) and Modified Shuttle Walk testing (MSWT), using standardised protocols, were assessed at baseline (pre-ETI) with an aim to repeat at one, six- and twelve-month follow-up clinic visits (post-ETI). QS was normalised to body weight (bw%).  **Results:**  At baseline, 109 pwCF consented to participation (mean age 33 ± 12 years; n = 67males [62%]). At baseline, 107 (98%) completed QS and 70(64%) MSWT. This number reduced over time, with QS tests dropping to 45(41%) and MSWT reducing to 24(22%) at the 12-month visit. The primary reason for reduced data capture was an increase in telehealth consultations. There was no significant change to QS (bw %) between baseline and 12 months (110 ± 31[SD] to 113 ± 26[SD], CI -3.4 – 9.9,p = 0.33). MSWT (meters) improved significantly at 12 months (908 ± 220 [SD] to 1004 ± 260 [SD], 95% CI 44 – 149, p < 0.001. Lung function (FEV1%pred) improved significantly overall (65.4 ± 21[SD] to 74.8 ± 20 [SD], 95% CI 8.98 – 16, p < 0.001).  **Conclusion:**  Examination of real-world outcomes demonstrated significant improvements in exercise capacity (MSWT) over 12 months following the commencement of ETI, alongside improvements in lung function. Peripheral muscle strength did not change in this preliminary analysis. |