or ICU length of stay. There was an association between pre-transplantation thoracic SMI and pre-transplantation percentage predicted forced expiratory volume in 1 second (FEV1pp) \( p = 0.002 \), with higher SMI associated with higher FEV1pp.

**Conclusions:** We did not identify a significant relationship between pre-transplantation thoracic SMI and post-transplantation outcomes in people with CF. There was an association between thoracic SMI and pre-transplantation pulmonary function, confirming the potential value of sarcopenia as a marker of disease severity.

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**Prescription of elixacaftor/tezacaftor/ivacaftor in lung transplant recipients with cystic fibrosis**


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**Background:** Elexacaftor/tezacaftor/ivacaftor (ELX/TEZ/IVA) received Food and Drug Administration approval in October 2019 for the treatment of cystic fibrosis (CF) in patients aged 12 and older with at least one F508del mutation in the CF transmembrane conductance regulator (CFTR) gene. ELX/TEZ/IVA works by facilitating delivery of functional CFTR protein to the epithelial cell surface and increasing chloride and bicarbonate movement across the cell membrane, helping reverse some of the deleterious effects of CF. The purpose of this study was to develop a protocol for administration and monitoring of ELX/TEZ/IVA at Baylor-St Luke’s Medical Center in qualifying lung transplant recipients with CF.

**Methods:** Inclusion criteria were receipt of a lung transplant and a diagnosis of CF 6 months or longer after lung transplantation; CFTR genotype with at least one F508del allele (genotype documentation required); presence of chronic sinus disease refractory to noninvasive interventions (e.g., irrigation with nasal saline, nasal steroid, intranasal antibiotic washes,) with or without evidence of chronic lung allograft dysfunction and of being underweight with inability to gain weight (Body mass index target in CF is 22 kg/m² for women and 23 kg/m² for men.) Exclusion criteria were Child Pugh Class C liver disease and active use of azole antifungal therapy (considered on case-by-case basis). The Baylor-St Luke’s Medical Center lung transplantation team discussed risks and benefits with eligible patients and arrived at a shared decision as to whether CFTR modulator treatments would be pursued. If pursued, the adult CF team would prescribe full or adjusted ELX/TEZ/IVA dosing with regular monitoring of weight, liver tests, immunosuppression drug levels, blood glucose, glycosylated hemoglobin, and routine spirometry. One-time baseline and 6-month post- ELX/TEZ/IVA assessments of Patient Health Questionnaire-9, General Anxiety Disorder-7, Sino-Nasal Outcome Test, iCAN gastrointestinal tracker, and Cystic Fibrosis Questionnaire Revised (CFQ-R) would also be performed to evaluate for symptomatic and wellbeing improvements.

**Results:** Eight patients were identified for potential enrolment. Two (25%) who enrolled were unable to tolerate the medication because of gastrointestinal side effects and discontinued the medication within days. One (12.5%) was unable to obtain insurance approval for the medication and never initiated therapy. Two (25%) experienced new-onset abdominal pain and nausea but opted to continue therapy because of subjective improvements in sinus symptoms, and one of these patients had improvement in their gastrointestinal symptoms after the dose was reduced. No statistically significant changes were detected in laboratory or symptom assessment scores. None of the patients required immuno-suppression dose adjustment after initiation of ELX/TEZ/IVA.

**Conclusions:** In this small, single-center study examining treatment effects in after lung transplantation in people with CF eligible for triple-combination modulator therapy, ELX/TEZ/IVA did not significantly improve symptom or wellbeing scores or routine laboratory testing, including changes to immunosuppression levels and liver enzyme results. A larger, multicenter study will be needed to better understand and characterize the impact of modulator therapies on lung transplant recipients with CF.

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**Collaboration within the advanced lung disease meeting on transplant referral process at Virginia Commonwealth University adult cystic fibrosis center**

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**Background:** Although medical management options are available for patients with advanced lung disease, lung transplantation is the best option to improve quality of life [1]. Before 2017, Virginia Commonwealth University (VCU) did not have a process for identifying advanced lung disease (ALD) in people with cystic fibrosis (CF). Our CF center joined the Cystic Fibrosis Foundation (CFF) Lung Transplant Learning and Leadership Collaborative (LLC) in 2017. We have elaborated on methods to standardize lung transplant referral guidelines and prevent obstacles to the lung transplant referral process. The goal of continuing to be a part of this initiative is to conduct a formal intervention for improving screening of people with ALD, patient education, and timely transplant referral. For the past few years, we have held meetings with Duke and other transplant centers to build relational coordination, shared knowledge, and effective co-production.

**Methods:** Patients were identified from within the VCU adult CF center and characterized according to the severity of their ALD. Patients were first stratified according to percentage predicted forced expiratory volume in 1 second (FEV1pp). Those with an FEV1pp less than 50% and who were exacerbation free for at least 50% of the time for 1 year were eligible. Between May 2021 and March 2022, there were five meetings, with three patients discussed at each meeting. Since 2017, the intervention has consisted of three elements: implementation of ALD meetings, development of a patient education process, and standardization of a transplant referral process. In 2021, other transplant centers, including Duke, University of Virginia, Inova Health System, and University of Pittsburgh Medical Center (UPMC), were included virtually during the meetings to assist with decision-making and discuss patient updates. An interdisciplinary team discussed people with ALD at the CF center monthly. Once patients were identified, patient education took place, consisting of discussion in a patient room with physician-defined recommendations. Those who wished to pursue transplantation were then referred to a transplant center. Patients who decided against transplantation were still discussed during the ALD meetings to ensure adequate care.

**Results:** Initially in 2017, of 45 patients classified as having ALD, 19 received education, and 23 were referred. In 2021, of 19 patients identified as having ALD, 16 received education, and 16 were referred. During the meetings from May 2021 to March 2022, two patients had an FEV1,pp of less than 30%, four had an FEV1,pp of 30% to 40%, and two had an FEV1,pp of 40% to 50%. Of the eight patients in 2022, two were given education and two were referred to UPMC. Five of the patients are female and three male.

**Conclusions:** Improving and standardizing lung transplant education and the referral process has increased efficiency at the CF center. The CFF LLC has continued to expand, and we are leading other centers to disseminate these guidelines. We will continue holding ALD meetings and identifying new areas of improvement to enhance this process. We will also guide our team to participate in formal training to sustain the education and timely transplant referral efforts. We hope that our experience in standardizing these guidelines may serve as a model for other CF centers nationwide.

**Reference**