participants had 29 PExs that met criteria for randomization but were not randomized; parent or participant discomfort with randomization was the most common reason (45% of eligible PExs that were not randomized). Among the 33 participants randomized to the “tailored therapy” arm, 23 (70%) were not treated with oral antibiotics within 28 days of randomization. Across both arms, 35% returned to clinical care: 13 (43%) in the “early antibiotic” arm and 9 (27%) of the “tailored therapy” arm. Failure to improve within 7 days and incomplete recovery within 14 days of randomization were the most common reasons. Nineteen participants reported 31 adverse events (AEs). The majority were mild and unrelated to study procedures.

Conclusions: The STOP PExD pilot study met its primary objectives by demonstrating that a single mild PEx in a selected population could resolve without oral antibiotics in 70% of the cases, and by demonstrating the feasibility of enrollment, identification of PEx through weekly texts, randomization of PEx treatment, and adherence to study measures. A definitive clinical trial is necessary to evaluate the short- and long-term safety and efficacy of this approach for repeated PEx events.

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**TRANSPLANTATION**

143 Bronchoalveolar lavage proteome identifies pathways of disease that segregate lung transplant recipients with and without bronchiolitis obliterans syndrome

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**Background:** Lung transplantation (LTx) is a treatment option for many end-stage lung diseases, including cystic fibrosis (CF). Proteomic analysis of bronchoalveolar lavage (BAL) fluid from LTx recipients has not been performed to identify potential biomarkers or pathways associated with bronchiolitis obliterans syndrome (BOS). We hypothesized that determining the differences between patients with and without BOS might shed light on the molecular underpinnings of disease.

**Methods:** We analyzed BAL fluid from 31 adult LTx recipients with (n = 15, 2 female) and without (n = 16, 3 female) BOS. Samples were collected from patients who underwent LTx for idiopathic pulmonary fibrosis, chronic obstructive pulmonary disease, or alpha 1 antitrypsin deficiency. Whole BAL protein was precipitated with acetone, resolubilized, fractionated using sodium dodecyl sulfate gel electrophoresis, in-gel trypsin digested, and subjected to mass spectrometer–based proteomic analysis.

**Results:** Our liquid chromatography mass spectrometry proteomic approach identified 4908 protein isoforms that were present in at least half of either cohort, with 227 isoforms (p < 0.01, false discovery rate (FDR) < 0.1) distinguishing LTx recipients with and without BOS. Pathway analysis of differentially expressed proteins reveal vascular permeability (p = 0.003), proliferation and migration (p = 0.06), protein localization to ciliary membrane (p < 0.001), and prostaglandin E2 immune response (p = 0.049) (Figure 1). A more stringent percolator-decoy analysis identified 9 isoform differences and reveals changes in platelet degranulation (p = 0.001, FDR = 6.09e-6), kallikrein-kinin driven inflammation (p < 0.001, FDR = 3.37e-3), lectin-induced complement pathway (p < 0.001, FDR = 6.09e-6), and IL-6 mediated inflammation (p = 0.004, FDR = 9.96e-3). Disease associations of identified differences included connective tissue diseases (p < 0.001, FDR = 3.26e-3), wounds and injuries (p < 0.001, FDR = 3.26e-3), and autoimmune disease (p < 0.001, FDR = 3.26e-3). Strong signals antigen presentation and immunoglobulin mediated immune response (p < 0.001) and viral transcription (p < 0.001) were also observed in the BOS cohort.

**Conclusions:** The data suggest that antiviral, antimicrobial, complement inhibition, and antigen presentation are features of disease in LTx patients who developed BOS. Therapies that target these pathways may be of benefit. Furthermore, our proteomic approach may offer an additional tool for the diagnosis of BOS in LTx recipients.

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144 Computed tomography body composition and clinical outcomes after lung transplantation in cystic fibrosis

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**Background:** Low muscle mass is common in patients undergoing lung transplantation and may be linked to worse post-transplantation outcomes, but existing studies assessing muscle mass and post-transplantation outcomes include few patients with cystic fibrosis (CF). To address this limitation, we identified a cohort of people with CF and examined the relationship between pre-transplantation muscle mass measured using computed tomography (CT) and post-transplantation outcomes.

**Methods:** Between May 1993 and December 2018, of 152 adults with CF who underwent lung transplantation at our institution, 83 met inclusion criteria and had usable CT scans. Using Cox proportional hazards regression, we evaluated the association between pre-transplantation thoracic skeletal muscle index (SMI) and our primary outcome of death after lung transplantation. Secondary outcomes, including days to post-transplantation extubation and post-transplantation hospital and intensive care unit (ICU) length of stay, were assessed using linear regression. We also examined associations between thoracic SMI and pre-transplantation pulmonary function and 6-minute walk distance.

**Results:** There was no association between pre-transplantation thoracic SMI and death after lung transplantation (HR 1.109; 95% CI, 0.948–1.109), days to post-transplantation extubation, or post-transplantation hospital...
Prescription of elexacaftor/tezacaftor/ivacaftor in lung transplant recipients with cystic fibrosis


Background: Eluxacaftor/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 overweight 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 immuno-suppression 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. Acknowledgements: Our team would like to acknowledge Dra. Amparo Sole Jover and Ines Perez for granting us access to the electronic version of the CFQ-R questionnaire.