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Efficacy of cyproheptadine for appetite stimulation in children with cystic fibrosis

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Background: Cystic fibrosis (CF) is an autosomal-recessive disease that results in progressive lung disease and may lead to poor nutritional status secondary to pancreatic insufficiency (PI). Because of impaired digestion and malabsorption, weight loss and malnutrition are complications of CF, and have been linked to declining lung function and increased infection. This study aims to evaluate the impact of cyproheptadine on nutritional status and lung health in children with CF and PI.
Methods: A retrospective chart review was conducted on patients aged 1 to 21 with CF and PI initiated on cyproheptadine for appetite stimulation between 2013 and 2019. Fifty-two patients with poor nutritional status were analyzed, comparing changes in body mass index (BMI) z-scores or weight for length based upon age, lung function (forced expiratory volume in 1 second [FEV_1]), and pulmonary exacerbations for 1 year before and 1 year after starting cyproheptadine or until discontinuation.
Results: Patients initiated on cyproheptadine realized an improvement in BMI z-scores of 0.363 ($p = 0.002$) and weight-for-length z-scores of 0.96 ($p < 0.001$) after 12 months. Reduction in the number of exacerbations was demonstrated, with 1.41 fewer exacerbations per calendar year ($p < 0.001$) 12 months after cyproheptadine initiation. There were no significant changes in FEV1.
Conclusions: The results of this study indicate a clinical benefit with cyproheptadine for appetite stimulation based on use for at least 12 months. Cyproheptadine was found to be an appropriate therapy to improve nutritional status, as demonstrated by significant increase in BMI and weight-for-length z-scores, and was correlated with fewer exacerbations. These findings suggest that cyproheptadine is an effective treatment addition for providing alternative nutritional support in people with CF and PI as young as 1 year old.

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Caregiver perceptions of a lifestyle education tool and subsequent behavior changes with elexacaftor/tezacaftor/ivacaftor initiation

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Background: Elexacaftor/tezacaftor/ivacaftor (ELX/TEZ/IVA) is a cystic fibrosis (CF) transmembrane conductance regulator (CFTR) modulator currently approved for persons with CF aged 6 and older with at least one copy of the F508del mutation or another eligible mutation. ELX/TEZ/IVA should be taken every 12 hours with fat-containing (10–20 g) foods and pancreatic enzyme therapy for optimal absorption. In clinical trials, ELX/TEZ/IVA increased body mass index (BMI) by 1 kg/m^2. Some adolescents started on ELX/TEZ/IVA at our center had rapid weight gain leading to overweight or obese status. We developed a proactive educational tool to increase caregiver knowledge about nutrition, physical and mental wellbeing, and anticipated outcomes with ELX/TEZ/IVA use in children aged 6 to 11, with the goal of providing education and avoiding rapid weight gain.
Methods: As part of a quality improvement project, multidisciplinary clinical care team members and family partners created an educational tool (Figure 1) to educate patients and families on the impact of ELX/TEZ/IVA therapy on BMI and potential lifestyle changes. Several months after the ELX/TEZ/IVA educational session, a survey was sent via the patient portal in our hospital's electronic medical record to caregivers of children aged 6 to 11 started on ELX/TEZ/IVA. This survey assessed caregiver perceptions of the educational tool and subsequent behavioral changes at home after ELX/TEZ/IVA initiation.

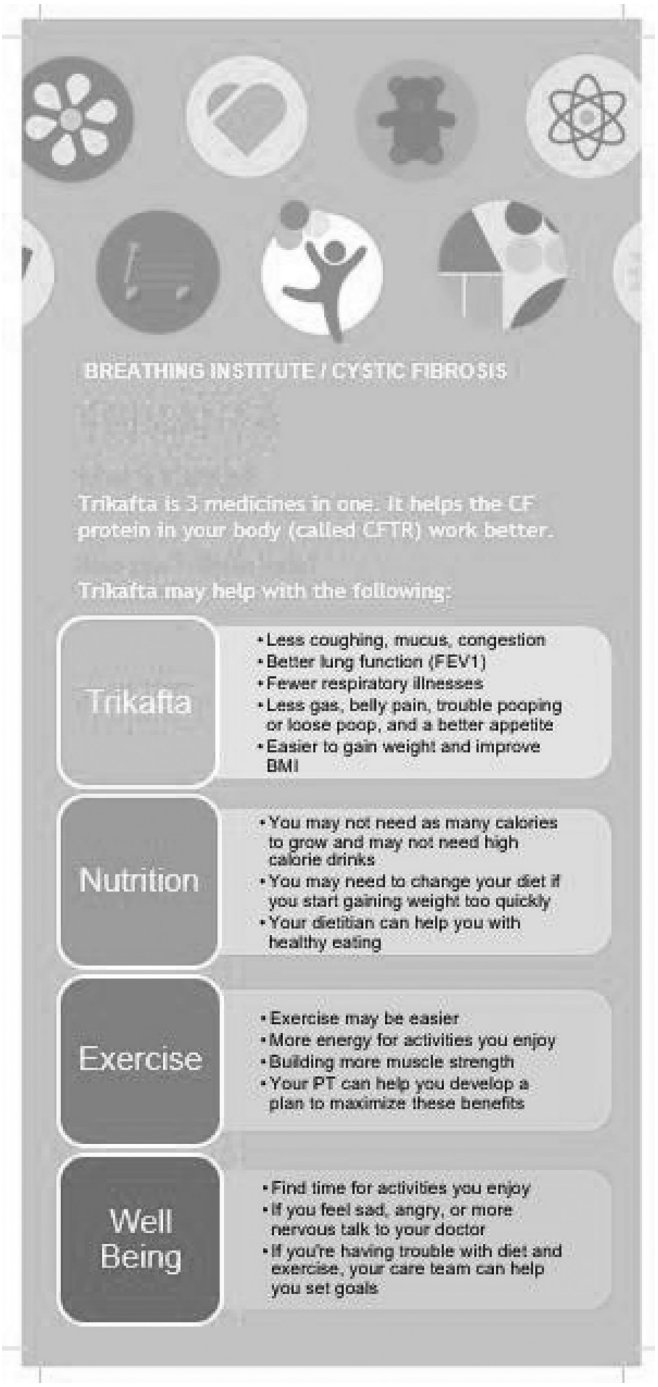

Figure 1. Elexacaftor/tezacaftor/ivacaftor (Trikafta) education tool

Results: We received 37 survey responses. Twenty-eight (76%) remembered receiving the educational tool and found it helpful, seven (19%) did not remember receiving it, and two (5%) did not find it helpful. Two-thirds requested that future educational materials be provided electronically. Caregivers reported potential side effects ($n = 26$) as the biggest concern with ELX/TEZ/IVA initiation, and some were concerned about insurance coverage ($n = 13$) or lack of improvement with treatment ($n = 11$). Nutritional and lifestyle changes that were made after ELX/TEZ/IVA