

than previously reported. Based on our limited experience, sweat chloride testing may be useful to identify potential adherence problems and to emphasize the importance of consistent ELX/TEZ/IVA use to achieve the best outcomes.

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GI/NUTRITION

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Body mass index versus mid-upper arm circumference: Can we replace the gold standard?

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Background: Body mass index (BMI) has long been used as a measure and predictor of health in the general population. For people with cystic fibrosis (PwCF), it has been established as the gold standard for nutrition assessment because optimal BMI correlates with better lung function, measured according to forced expiratory volume in 1 second (FEV₁), although as in the general population, BMI use in PwCF has limitations, because it does not distinguish lean body mass from fat mass [1]. The rise in obesity in PwCF prompted our search for a better predictor of nutritional status in the pediatric CF population [2]. Mid-upper arm circumference (MUAC) is an established measure of pediatric nutrition status and predictor of fat mass [3]. In 2021, our care center began including MUAC measurements as part of our nutrition assessment in children with CF. The objective of this study was to compare BMI and MUAC nutrition assessment methods in children with CF.

Methods: An institutional review board–approved, single-center retrospective analysis was performed on 87 patients with an average age of 10.7 ± 4.2. Data were collected on BMI, MUAC, and FEV₁ from April 2021 to March 2022. Correlation was assessed between BMI Z-scores and MUAC Z-scores, between MUAC Z-scores and FEV₁, and between BMI Z-scores and FEV₁. Subsequent-visit BMI, MUAC, and FEV₁ measurements were obtained in 34 patients.

Results: BMI and MUAC Z-scores had a statistically significant positive linear correlation. MUAC Z-scores identified more patients as malnourished than BMI Z-scores. At the baseline visit, MUAC Z-score identified 12 of 75 children (16%) with CF who were identified as well nourished according to BMI Z-score as having mild or moderate malnutrition (Cohen kappa statistic=0.46, 95% CI, 0.27–0.65). FEV₁ did not differ significantly between those who BMI and MUAC identified as well-nourished and those who only MUAC identified as malnourished. Of the three values (BMI, MUAC, FEV₁), FEV₁ at baseline remained the best predictor of FEV₁ at the subsequent visit.

Conclusions: Based on our review, these results lend support to prior evidence that more patients were identified as malnourished according to MUAC than BMI, but because of the small sample size and retrospective nature of this review, a larger, multicenter study on MUAC as a nutrition assessment method in children with CF is necessary to determine correlation between MUAC and lung function.

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Impact of elexacaftor/tezacaftor/ivacaftor on body composition in a small cohort of youth with cystic fibrosis

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Background: The triple combination cystic fibrosis (CF) transmembrane conductance regulator (CFTR) modulator elexacaftor/tezacaftor/ivacaftor (ELX/TEZ/IVA) has been shown to be highly effective in improving pulmonary function, increasing body mass index (BMI), and reducing pulmonary exacerbations [1,2]. With increases in BMI and recent reports of obesity in the CF population, closer evaluation of the impact of ELX/TEZ/IVA on body composition may inform metabolic disease risk.

Methods: This was a secondary analysis of a prospective observational study assessing annual, frequently sampled oral glucose tolerance tests (OGTTs) and dual-energy X-ray absorptiometry (DXA) in people with CF aged 6 and older. Inclusion criteria were confirmed diagnosis of CF treated with ELX/TEZ/IVA and available DXA scan results before and after triple combination therapy. Exclusion criteria were use of medications affecting glucose homeostasis, including insulin; pulmonary exacerbations or admissions in the 8 weeks before the study visit; and pregnancy. Paired t-tests were used to compare normally distributed variables and Wilcoxon signed rank tests for non-normally distributed variables. Measurements of body composition using DXA included body free fat mass adjusted for height (FFMI) and total body fat mass adjusted for height (FMI). OGTTs with sampling at 0, 10, 30, 60, 90, and 120 minutes for glucose, insulin, and c-peptide were obtained. Integrated area under the curve (iAUC) for each was calculated. Homeostatic model assessment of insulin resistance (HOMA2 IR) was calculated as an estimate of insulin sensitivity. Spirometry data from the most-recent clinic visit were obtained through chart review.

Results: Eight participants (median age 22.1, interquartile range (IQR) 16.2–28.2; 87.5% male) were included in the analysis. Mean weight z-score was –0.52 (IQR –1.51 to –0.31), and mean BMI z-score was –0.11 (IQR –0.7–0.87). Median time on ELX/TEZ/IVA was 11 months (IQR 10.8–11.3). All participants were pancreatic insufficient, and 50% were homozygous F508del. Table 1 presents clinical outcomes before and after ELX/TEZ/IVA. Weight increased from 58.6 kg to 68.9 kg ($p=0.01$), whereas BMI z-score did not change. FMI increased ($p=0.04$), but FFMI did not change. Glucose iAUC did not change ($p=0.23$). Insulin secretion measured according to iAUC and C-peptide iAUC increased ($p=0.02$), and HOMA2 IR, as an index of insulin resistance, increased ($p=0.01$).

Conclusions: In a small cohort of people with CF treated with ELX/TEZ/IVA for less than 1 year, weight and FMI increased, whereas FFMI did not change. OGTT-derived estimates of insulin secretion (iAUC) and insulin resistance (HOMA2 IR) increased. Larger studies are needed to evaluate the effects of highly effective triple therapy on body composition, particularly body fat distribution, and its association with insulin resistance in subjects with CF.

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