one percent of the patients had pancreatic exocrine insufficiency, with $p < 0.01$ as a predictor variable for survival. Forty-four percent of patients had suboptimal values for FEV_1, and 26% patients had suboptimal values for FVC. Major effect on survival within 7 years were detected in patients with positive *P. aeuroginosa* and those with undernutrition. Sixty-one percent did not have government coverage of treatment, which had a clear effect on survival (Figure 1).

Conclusions: To our knowledge, this is the first report of survival in people with CF in Latin America. Median survival was 43.6 years in 2017 and 46.2 years in 2019 in the United States and 47 years in the United Kingdom in 2019, a difference of more than 2 decades from survival reported in our study. The predictor variables affecting survival are consistent those previously reported in the literature but not with diagnosis before 2 years old. There are two possible reasons for this: CF diagnosis based on newborn screening started in 2015, so it might be that the effect on survival was not yet evident, and patients with early diagnosis did not have full treatment coverage. Many interventions have been performed since the beginning of the cohort. We want to highlight the treatment coverage for the government from 2015 and the intervention of the nutrition team from 2006. The nutrition team was integrated full time into the CF clinic—counting fat grams, dynamically managing pancreatic enzymes, increasing energy density, providing recipes for a homemade elemental diet, and managing food portions, so we expect to see a greater effect on survival in the coming years. Treatment with CFTR modulators is not available in Mexico, so the survival results of this study are strongly associated with the multidisciplinary approach and continuous education of patients and parents, which has made an important difference in the quality of life of patients.

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Impact of early growth trajectories and cystic fibrosis transmembrane conductance regulator modulator therapy on puberty in cystic fibrosis

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Background: The advent of cystic fibrosis (CF) transmembrane conductance regulator (CFTR) modulators has led to a rapidly changing landscape in CF outcomes. It remains unclear to what degree puberty is affected in children with CF in this new era. Historically, children with CF experienced a 2- to 4-year delay in pubertal onset, possibly as a manifestation of poor nutrition or CFTR dysfunction [1,2]. CFTR modulators have the potential to affect pubertal timing by improving nutritional status and normalizing CFTR function in the hypothalamic-pituitary-gonadal axis [3]. Pubertal progression in CF has not been explored in the era of CFTR modulators, and the impact of early growth on puberty in CF is unknown.

Methods: Using national data from the U.S. Cystic Fibrosis Foundation Patient Registry (CFFPR), this study aims to analyze associations between early growth trajectories and puberty using peak height velocity (PHV) as a proxy measure. PHV (adolescent growth spurt) is the maximum rate of height increase for an individual. Age at PHV (APHV) is frequently used to approximate the timing of puberty. PHV and APHV will be derived using a shape-invariant mixed model to approximate growth. Individuals aged 18 to 21 between 2010 and 2019 with confirmed CF according to sweat test or genotype analysis were included. We will classify early growth trajectories (aged 0–6) based on weight for length (WFL) or body mass index (BMI) into mutually exclusive categories representing growth patterns that clinicians may seek to encourage (WFL-BMI always >50th percentile, increasing, or stable) or avoid (WFL-BMI decreasing). We will analyze the impact of early growth on APHV (primary endpoint), as well as actual PHV and final adult height (secondary endpoints). We will perform unadjusted analyses using t-tests or analysis of variance. We will perform adjusted analyses using multivariable linear regression, including various potential confounders and effect modifiers. We will conduct stratified analyses based on sex and modulator use. We will also split analyses according to birth cohort to determine whether pubertal patterns have changed over time. Lastly, we will compare PHV, APHV, and adult height with published North American longitudinal standards to compare pubertal outcomes of individuals with CF with those of the general population.

Results: We secured CFFPR data in late March 2022. Analyses are underway and are expected to be complete in 2 months. The dataset captures 12,615 people with CF: 52.5% male, 91.9% white, 5.2% Black, 0.7% Asian, and 2.2% of another race. Median age at CF diagnosis was 0.5 years. Sixty-three percent are treated with CFTR modulator therapy, although mean age at which therapy was prescribed is 21.7 years (after completion of puberty). Approximately 14% (894 male, 830 female) were started on CFTR modulator therapy before age 18. Mean adult height is at the 36th percentile.

Conclusions: Conclusions are pending analytic results.

Acknowledgements: This work was supported by the Cystic Fibrosis Foundation [grant PATIL20B0].

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Lipid trends on highly effective modulators in cystic fibrosis

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Background: There are few reports of cardiovascular disease (CVD) in people with cystic fibrosis (PwCF), but with advances such as highly effective modulator therapy (HEMT), PwCF are living longer, with increasing exposure to CF-related diabetes (CFRD), and are increasingly overweight or obese. All these factors increase the risk of CVD. Prior studies of serum lipids in PwCF before the advent of HEMT typically demonstrated low total cholesterol (TC), low-density lipoprotein (LDL), and high-density lipoprotein (HDL). It is not known whether HEMT may change serum lipid profiles in PwCF, perhaps because of altered absorption or metabolism.

Methods: We addressed this knowledge gap using the Carolina Data Warehouse to identify PwCF aged 18 and older at the University of North Carolina (UNC) Cystic Fibrosis Center with available lipid panel data within 5 years before starting ivacaftor or elexacaftor/tezacaftor/ivacaftor and at any timepoint thereafter. Retrospective chart review was performed after obtaining institutional review board approval. Exclusion criteria included solid organ transplantation or type 1 diabetes. The primary endpoint was change in TC; secondary endpoints included changes in HDL, TC/HDL ratio, change in BMI, and a new diagnosis of hypertension. Unadjusted mean changes in TC were assessed using paired t-tests. Available samples were not consistently collected in the fasting state.

Results: We identified 42 patients meeting study criteria; 27 (64%) were female, 24 (57%) had CFRD, 12 (29%) had CF liver disease, and 36 (86%) were pancreatic insufficient. Mean age at HEMT initiation was 40 (range 16–73). On average, there was a 3-year interval between the pre- and post-HEMT lipid panel and 17 months between HEMT initiation and the post-HEMT lipid panel. We noted significant increases in TC, LDL, and TC/HDL ratio after HEMT (Table 1). There was no significant change in HDL. BMI rose significantly. At baseline, nine patients had diagnoses of hypertension; three additional patients were diagnosed after starting HEMT.