Table 1 (abstract 46):
Lipid profile measurements (means) before and after patients were started on highly effective modulator therapy and mean difference. Results of paired t-tests are displayed.

Table 1. Measurements before and after modulator initiation				
	Pre-HEMT[e], mean	**Post-HEMT**, mean	**Difference**, mean (95%CI[f])	**p value**
TC[a] (mg/dL) n = 41	153.1	169.9	16.8 (5.55, 30.0)	0.0043
HDL[b] (mg/dL) n = 39	54	53.2	-0.821(-6.24, 4.61)	0.76
LDL[c] (mg/dL) n = 36	73.4	91.2	17.9 (9.27, 26.5)	0.0002
TC/HDL n = 40	3.07	3.47	0.395 (0.0728, 0.716)	0.016
BMI[d] (kg/m^2) n = 41	25.1	26.1	1.00 (0.460, 1.54)	0.00062

a. TC = total cholesterol, b. HDL = high density lipoprotein, c. LDL = low density lipoprotein d. BMI = body mass index, e. HEMT = highly effective modulator therapy, f. CI = confidence interval

Conclusions: Our results are limited by their retrospective nature, variable time between lipid assessments and exposure to HEMT, but in this small sample, the increases in TC, TC/HDL ratio, and LDL may be clinically meaningful because higher values were associated with greater CVD risk. HDL, a cardioprotective lipid, did not change. If confirmed in larger, more tightly controlled studies, this could mean that long-term exposure to HEMT increases CVD risk. This knowledge may influence health screening practices. When combined with other CVD risk factors, lipid-lowering therapy may be considered for some PwCF on HEMT.

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47

Longitudinal assessment of health-related quality of life and clinical outcomes in children and adolescents with cystic fibrosis in Australia

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Background: The Australian Cystic Fibrosis Data Registry (ACFDR) collects data from more than 3,500 people with cystic fibrosis (CF). The registry does not capture health-related quality of life (HRQOL) measures. HRQOL is an important outcome, influencing decisions regarding the efficacy and value of health interventions in clinical settings. The aim of this study was to assess and compare child-reported HRQOL and evaluate relationship with clinical outcomes by analyzing 10 years of patient-reported outcome measures collected at one ACFDR contributing center.

Methods: Children and adolescents aged 6 to 18 described their HRQOL using the Cystic Fibrosis Questionnaire-Revised (CFQ-R) between 2010 and 2020. Eight HRQOL domains were assessed: respiratory, physical, emotional, body, digestive, treatment burden, emotion, social. Demographic and diagnostic data, as well as information on body mass index (BMI) and forced expiratory volume in 1 second (FEV_1), of study participants were extracted from the ACFDR. Descriptive analysis was used to describe the study population. Spearman rank correlation was calculated for CFQ-R domains, BMI, and FEV_1. $P < 0.05$ was considered to be significant.

Results: Forty-seven children and adolescents completed at least one CFQ-R between 2010 and 2020: 23 (48.9%) female; 40 (85%) aged 6 to 13, and 22 (46.8%) F508del homozygous. A Spearman rho correlation test demonstrated a strong relationship between FEV_1 and CFQ-R body domain ($r = 0.328$, $p = 0.004$) in girls. A significant negative relationship between FEV_1 and CFQ-R treatment body domain ($r = -0.262$, $p = 0.005$) was observed in boys. A strong positive relationship was also observed between BMI and CFQ-R physical domain ($r = 0.243$, $p = 0.009$ in boys only) and the CFQ-R eating domain ($r = 0.372$, $p < 0.001$ in boys; $r = 0.342$, $p = 0.003$ in girls). Additional analyses and associations between HRQOL scores stratified according to age group and sex will be presented at the conference.

Conclusions: The initial findings of our study are consistent with those in the literature, suggesting that the CFQ-R is a meaningful clinical endpoint to evaluate clinical interventions in children. By incorporating the CFQ-R into the ACFDR, this study will provide a patient perspective for clinicians and policy makers, enhancing the treatment and management of children and adolescents with CF.

48

Longitudinal outcomes of adults and children with cystic fibrosis during the COVID-19 pandemic

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Background: The onset of the COVID-19 pandemic was associated with restricted community movement, including limited access to health care facilities, resulting in a change in clinical service delivery to people with cystic fibrosis (CF). The aim of this study was to determine clinical outcomes of Australian adults and children with CF in the 12 months after the onset of the COVID-19 pandemic.

Methods: This longitudinal cohort study used prospectively entered national registry data. Primary outcomes were 12-month change in percentage predicted forced expiratory volume in one second (FEV_1pp), body mass index (BMI) in adults, and BMI z-scores in children. A piecewise linear mixed-effects model was used to determine trends in outcomes in the 24 months before and 12 months after the onset of the pandemic. Comparative analysis of hospitalization and service delivery data before and after the onset of the pandemic was conducted.

Results: Data were available for 3,662 individuals (median age 19.6, range 0–82). Overall registry data completeness was 95%. When trends in outcomes before and after pandemic onset were compared; FEV_1pp went from a mean annual change of –0.13% (95% CI, –0.39 to 0.13) to a mean improvement of 1.73% (95% CI, 1.29–2.17). Annual mean annual change in