ABSTRACT

Rationale: The introduction of elexacaftor/tezacaftor/ivacaftor (ETI), a highly effective cystic fibrosis transmembrane conductance regulator (CFTR) modulator therapy, to younger ages and the COVID-19 pandemic have significantly reduced pulmonary exacerbations requiring hospitalization among children with CF.

Objective: To assess demographic and clinical characteristics of children and young adults with CF hospitalized for pulmonary exacerbations before and after pediatric ETI approval.

Methods: A retrospective chart review was conducted at five United States CF Foundation-accredited care centers. Hospitalization data from children and young adults with CF in 2018 and 2022 were analyzed.

Results: Hospitalizations decreased from 471 cases (241 individuals) in 2018 to 163 cases (110 individuals) in 2022. The racial distribution shifted, with more hospitalized patients identifying as Black or non-White in 2022 (28% vs. 14%; p=0.018). A greater proportion of hospitalized children in 2022 had two non-F508del mutations compared with children hospitalized in 2018 (38% vs. 19%) and were less likely to be infected with methicillin-resistant *Staphylococcus aureus* (MRSA). Comparing 2022 to 2018, children on CFTR modulator therapy, including ETI (76%), showed reduced infections with *Pseudomonas aeruginosa* and *Achromobacter xylosoxidans*.

Conclusions: The decline in hospitalizations for pulmonary exacerbations likely reflects the benefits of ETI therapy, as a higher proportion of children and young adults hospitalized in 2022 had two non-F508del mutations and were not eligible for ETI. A greater percentage of those hospitalized in 2022 identified as belonging to minority racial groups, highlighting ongoing health disparities in the ETI era. Additionally, there were notable changes in the microbiological characteristics between 2018 and 2022.