

SHIELDS NETWORK OUTCOMES*

Cystic Fibrosis

Cystic fibrosis (CF) is a genetic progressive disorder caused by mutations in the chloride ion channel cystic fibrosis transmembrane conductance regulator (CFTR), which helps to maintain the balance of salt and water on many surfaces in the body. Many pharmacological treatments on the market address pulmonary manifestations of CF. However, oral CFTR protein modulators transformed the management of CF in the last decade.¹ They are highly effective in most CF patients with eligible genotypes. CFTR modulators decrease pulmonary exacerbations, improve pulmonary function (ppFEV1), nutritional status, and respiratory health-related quality of life.^{2, 3}

Pulmonary Exacerbations Hospital and ER Utilization 30% PATIENTS 21% ≤**2**5% 20% ⁄⁄₀ ЦО 10% Adult Pediatric % Pediatric Adult Of patients require ER visits or <18 v/o inpatient hospital stays for CF Benchmark⁴ Shields Network Of patients report feeling very well **Patient Reported** or well about how their specialty **Treatment Efficacy** medication is helping them with CF Of patients reported no missed days No Absenteeism of planned activity, school, or work Of patients receive and take Adherence** medications on time

What We Measure

Why It Matters

Pulmonary exacerbations are closely related to IV antibiotic use and hospitalizations and are a significant measure of disease severity. Acute pulmonary exacerbations are associated with rapid lung function decline, permanent loss of lung function, diminished quality of life, reduced survival, and increased healthcare costs.⁹ Decreasing the number of pulmonary exacerbations is a target in the care of CF patients. Measuring absenteeism and adherence is also crucial in identifying gaps in care, ensuring timely interventions, and improving overall health outcomes.

shields Health Solutions Network includes data from a collective of member health systems that partner with Shields o elevate an integrated specialty pharmacy model.

- Il metrics are reflective of data collected in 2024
- Adherence is measured by the proportion of days covered (PDC) Pediatric: < 18 years old Adult: > 18 years old
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