CYSTIC FIBROSIS FOUNDATION
ADDING TOMORROWS

February 22nd, 2023

Robert M. Kerr
Director, South Carolina Department of Health and Human Services
P. O. Box 8206
Columbia, SC 29202

Dear Mr. Kerr:

On behalf of patients and families with cystic fibrosis (CF) in South Carolina, we write to you regarding the status of Pancreaze, and Pertzye as non-preferred products on South Carolina Medicaid's preferred drug list. Specifically, **we are writing to request that you continue to allow people with CF who are already stable on a pancreatic enzyme product (PERT) to continue being able to access that therapy.** While PERTs may be interchangeable in some populations, people with CF experience variable responses to these therapies and patients who are stable on a non-preferred product should not be forced to switch – a change that puts CF patients at serious risk for negative health outcomes.

About Cystic Fibrosis & the CF Foundation
Cystic fibrosis is a life-threatening genetic disease that affects nearly 40,000 children and adults in the United States, including 478 people in South Carolina. CF causes the body to produce thick, sticky mucus that clogs the lungs and digestive system, which can lead to life-threatening infections. Cystic fibrosis is both serious and progressive; lung damage caused by infection is irreversible and can have a lasting impact on length and quality of life. As a complex, multi-system condition, CF requires targeted, specialized treatment and medications. While advances in CF care are helping people live longer, healthier lives, half of young adults with CF still die before the age of 35, usually by respiratory failure.

As the world's leader in the search for a cure for CF and an organization dedicated to ensuring access to high quality, specialized CF care, the Cystic Fibrosis Foundation supports the development of CF clinical practice guidelines and accredits 130 care centers and 55 affiliate programs nationally – including 3 care centers that house 4 CF programs in South Carolina.

About Pancreatic Enzyme Replacement Therapy
Pancreatic enzyme replacement therapy (PERT) is a life-sustaining therapy for people with CF as nutritional status is closely linked to pulmonary function and survival. Cystic fibrosis is a multi-system disease that causes the ducts in the pancreas to become clogged with thick, sticky mucus that blocks natural enzymes from reaching food in the small intestine. As a result, approximately 90 percent of CF patients have pancreatic insufficiency, making PERT a vital component of CF care. Decreased pancreatic function leads to malabsorption of calories and nutrients, and therefore, difficulty with growth and weight gain. Patients with pancreatic insufficiency require lifelong PERTs with each meal and snack to maintain adequate nutrition and prevent abdominal distress.

Although the drug substance is the same, the dissolution properties of the PERTs are not identical. The differences in enteric coating, delivery, and size of each FDA-approved product affect a patient's ability to

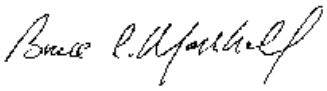
absorb nutrients. The degree of acidification of the GI tract in each CF patient also varies, causing some patients to have a better clinical response to one product over another.

Eliminating any individual enzyme from the formulary disregards the variable clinical responses of CF patients to pancreatic enzyme therapies and jeopardizes the health of patients already on that therapy. Nutritional failure of any type places CF patients at risk for long-term health consequences and a patient's PERT regimen, once stable, should not be modified unless it is clinically indicated. Forcing patients to change their PERT is also especially burdensome for patients who have tried multiple products before identifying one that stabilized their nutritional response.

Given this individualized response, we urge South Carolina Medicaid to allow all patients who are already stable on a PERT to continue using their existing product, while continuing to utilize Creon and Zenpep as preferred first-line product for those who are newly prescribed this type of therapy.

We stand ready to answer any questions about this or other CF treatments and would be happy to schedule a time to meet and further discuss this complicated issue. Please contact Olivia Dieni, Sr. Specialist, Health Systems Innovation and Navigation, at odieni@cff.org or (240) 200-3715. Thank you for all that you do for people with CF.

Sincerely,



Bruce C. Marshall, MD
Executive Vice President
Chief Medical Officer

Mary Dwight
Senior Vice President
Chief Policy and Advocacy Officer

Steven M. Snodgrass, MD
Medical Director, Pediatric Pulmonology
CF Center Director
Prisma Health Children's Hospital–Upstate
Greenville, SC

Heather M. Staples, MD
Pediatric Cystic Fibrosis Center Director
Director CF Therapeutics Development Center
Director of Pediatric Resident Quality Improvement and Research
Prisma Health-Midlands/University of SC
Columbia, SC

Sylvia E. Szentpetery, MD
Director, Pediatric Cystic Fibrosis Program
Assistant Professor of Pediatrics
Division of Pediatric Pulmonology, Allergy & Immunology
Medical University of South Carolina
Charleston, SC

Patrick Flume, MD
The Powers-Huggins Chair for Cystic Fibrosis
Professor of Medicine and Pediatrics
Director, Cystic Fibrosis Center
Medical University of South Carolina
Charleston, SC