June 16th, 2023

Linda VanCamp, CPhT Formulary Analyst
Bureau of Medicaid Care Management & Customer Service
Behavioral and Physical Health and Aging Services Administration
P.O. Box 30479
Lansing, Michigan 48909

RE: MDHHS Medicaid Health Plan Common Formulary

Dear Ms.VanCamp:

On behalf of people living with cystic fibrosis (CF) in Michigan, we thank you for the opportunity to comment on Michigan's Medicaid Health Plan (MHP) Common Formulary. Specifically, we are writing to request that you allow people with CF who are already stable on a pancreatic enzyme replacement therapy (PERT) to continue being able to access that therapy. We also request that pancrelipase and at least one dornase alfa-compatible nebulizer devices be added to the MHP Common Formulary. These products are vital components of the CF care regimen and their inclusion on the MHP Common Formulary will ensure that people with CF have access to the care and treatments they need to maintain their health. Additionally, the CF Foundation seeks clarification regarding Medicaid coverage criteria for 3% and 7% inhaled sodium chloride.

**About cystic fibrosis & the Cystic Fibrosis Foundation**
Cystic fibrosis is a life-shortening genetic disease that affects nearly 40,000 children and adults in the United States, including 1,180 in Michigan. Of these, over 15 percent of adults and one in five children with CF in Michigan depend on Medicaid for some or all of their health care coverage. There is no cure for CF today. 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.

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 6 care centers that house eleven CF programs in Michigan.

**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

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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.

Excluding any individual enzyme from the formulary disregards the variable clinical responses of CF patients to pancreatic enzyme therapies and jeopardizes the health of patients who have found stability within a PERT routine. 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 Michigan's Department of Health and Human Services (MDHHS) 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. Furthermore, we ask that the Pancreaze be added to the MHP Common Formulary as a preferred product under the drug class "Digestive Enzyme Mixtures". For children and adults with CF who are unable to use Creon or Zenpep, Pancreaze offers an alternative enzyme therapy that provides critical life-sustaining nutritional support.

**Inhaled sodium chloride**

Mucociliary clearance is an essential component of CF care and CF pulmonary guidelines recommend use of sodium chloride in individuals ages 6 and up.[1] Specifically, treatment with mucolytic products—including inhaled sodium chloride —is shown to help clear mucus from the lungs, resulting in fewer lung infections, improved lung function, and better quality of life for people with CF.[2] Long-term clinical trials also found that people who were treated with inhaled sodium chloride experienced a reduction in antibiotic use for pulmonary exacerbations and subsequently missed fewer days of work and school due to illness.[3] Additionally, the 2016 Clinical Guidelines for Preschoolers with Cystic Fibrosis recommends inhaled sodium chloride be offered to patients based on individual circumstance.[4] Further studies of this age group, released in both 2018 and 2019, conclude that use of inhaled sodium chloride is safe, well-tolerated, and resulted in improved lung clearance for both infants with CF and children aged 3-6 years with CF.[5, 6]

Please see the attached 2013 CF Pulmonary Guidelines and the 2016 Clinical Guidelines for Preschoolers with CF, both of which recommend the use of inhaled sodium chloride as part of the CF care regimen.

We thank Michigan Medicaid for including inhaled sodium chloride .9% on the MHP Common Formulary. However, people with CF often depend on different formulations of inhaled sodium chloride so we are

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[1] Mogayzel, Peter, Jr., Naureckas, Edward, et al. Cystic Fibrosis Pulmonary Guidelines. American Journal of Respiratory and Critical Care Medicine, Vol. 187, 2013.

[2] ibid

[3] Elkins MR, Robinson M, Rose BR, et al. A controlled trial of long-term inhaled hypertonic saline in patients with cystic fibrosis. N Engl J Med 2006;354:229–240.

[4] Lahiri, Thomas, Hampstead, Sarah E., et al. Clinical Practice Guidelines From the Cystic Fibrosis Foundation for Preschoolers With Cystic Fibrosis. Pediatrics, Vol. 137, 2016.

[5] Ratjen F, Davis SD, Stanojevic S, et al. Inhaled hypertonic saline in preschool children with cystic fibrosis (SHIP): a multicentre, randomised, double-blind, placebo-controlled trial. Lancet Respir Med 2019; published online June 6. http://dx.doi.org/10.1016/S2213-2600(19)30187-0.

[6] Stahl, Mirjam, Wielp, Mark O., et al. Preventive Inhalation of Hypertonic Saline in Infants with Cystic Fibrosis (PRESIS). American Journal of Respiratory and Critical Care Medicine. Volume 199. 2018.

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seeking clarification regarding the coverage criteria for inhaled sodium chloride 3% and 7%. Although sodium chloride 3% and 7% are not included on the MHP Common Formulary, both products are included on the Michigan Pharmaceutical Product List (MPPL), seemingly without prior authorization requirements. We are writing to confirm that 3%, 7%, and .9% concentrations of inhaled sodium chloride are covered for people with CF without prior authorization.

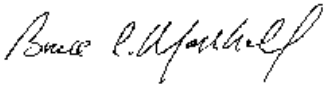
**Dornase alfa-compatible nebulizer and compressor devices**

We thank Michigan Medicaid for including dornase alfa (Pulmozyme®) on the formulary; however we request that you also include at least one compatible nebulizer and compressor device to ensure people are able to access this important therapy. Dornase alfa works to thin the accumulated mucus for people with CF, allowing for improved airway clearance and lung function.[7] This therapy represents an important treatment in CF care as it sustains improvements in lung function and reduces infection rates.[8] Those on dornase alfa experience fewer pulmonary exacerbations, fewer hospitalizations, less shortness of breath, and an overall sense of improved well-being.[9]

Per the FDA label, dornase alfa is administered via specific nebulizer systems that may be connected to an air compressor.[10] It is essential that these devices are accessible for people with CF, and we urge Michigan Medicaid to include either the eRapid® nebulizer system or one of the FDA approved jet nebulizers – such as the Pari® LC plus that was used in the clinical trials – and paired compressors on the formulary.

Thank you for the opportunity to provide feedback on Michigan's MHP Common Formulary. We stand ready to answer any questions about these issues or the CF care regimen. Please contact Beryl Manske, State Policy Specialist, at bmanske@cff.org with any questions or to discuss further. Thank you for all that you do for people with CF.

Sincerely,



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[7] Mogayzel Jr, Peter J., et al. "Cystic fibrosis pulmonary guidelines: chronic medications for maintenance of lung health." *American journal of respiratory and critical care medicine* 187.7 (2013): 680-689.

[8] Cramer, Gena W., et al. "The role of dornase alfa in the treatment of cystic fibrosis." *Annals of Pharmacotherapy* 30.6 (1996): 656-661.

[9] Goa, Karen L., and Harriet Lamb. "Dornase Alfa." *Pharmacoeconomics* 12.3 (1997): 409-422.

[10] https://www.accessdata.fda.gov/drugsatfda_docs/label/2021/103532s5191lbl.pdf

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