MSU Health Care

An Update from the Michigan State University Cystic Fibrosis Center.

To say this is an interesting time in the care of those with cystic fibrosis is obviously a massive understatement. In October of 2019 we shared the joy with the CF community of a highly effective modulator therapy becoming available, at some point, to most people with CF. Six short months later we were dealing with the fear and uncertainty of a respiratory pandemic. For the last twelve months everything has felt reactive; reacting to the virus, reacting to clinic closures, reacting to telehealth visits, reacting to research limitations, and reacting to the worry the virus could harm our patients, our families or ourselves. When asked to write this update it was difficult to choose a topic. The trials and tribulations of the past year, and work we have done to react to them, were an easy choice, but I instead want to write about positive things, projects having the potential to improve the lives of those with CF and especially those using more recent data to show how we are using the ongoing research in CF to potentially improve clinical care. I also wanted to pick topics I think people in the CF community might be interested to learn about and I think these two projects are good representations of this.

The first is our ongoing collaboration with basic science researchers to better understand the microbiome of the CF airways. The lung was initially thought to be sterile, so any bacteria found there were harmful. Studies from multiple groups have now shown the lower airways in healthy people are not sterile and there is a complex web of different bacteria known as the lung microbiome. How these bacteria interact with a person's lungs may worsen or protect against disease and the alterations we see in CF are far more complex than we had appreciated previously. Many of these bacteria will not grow on routine clinical cultures and so much more extensive and complicated testing needs to be done to learn about them. We are working to better understand what this bacterial community looks like in the airways of those with CF and how it changes in response to different treatments and with exacerbations. This is important because the bacteria in the airways, and people with CF themselves clinically, do not always respond to antibiotics they way we would predict. For more information on this topic, you can check out a podcast I did with Dr. Robert Quinn of Michigan State University and Dr. Lindsay Caverly of The University of Michigan (https://share.transistor.fm/s/75f13ab9). A better understanding of this area may allow for more effective treatment of airway infections and a better explanation of the effect of antibiotics on the airway infections related to CF. This knowledge will be crucial, especially for those who have mutations that do not allow for the use of highly effective modulators at this time or have more advanced lung disease. The project is still enrolling if people are interested in taking part in the study.

A second project we are working on is early initiation of airway clearance therapies in infants with CF. The time after a diagnosis is obviously and understandably difficult time for families of a child with CF. There is a delicate, and unique to each family, balance between starting therapies and not overwhelming people with information and treatments. The Cystic Fibrosis Foundation guidelines for the treatment of infants with CF published in 2009 suggests considering the initiation of hypertonic saline and Pulmozyme at the age of two, though practically many centers do start this sooner. Much has changed in our understanding of airway clearance treatments in CF since 2009. We have learned that mucus plugging alone can cause inflammation in the airways of those with CF independent of infection. A recent study found initiation of hypertonic saline prior to four months of age was associated with improved weight gain and measures of lung clearance. Children started at a young age maintained normal lung

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clearance though a year of age but initiation of hypertonic saline after a year of age did not return this value to normal. While this data is very new, and we need to learn more, it suggests the sooner the better with hypertonic saline therapy, and potentially Pulmozyme, the better. We have designed a project to work towards this goal and have thus far been very successful and our patients are tolerating it very well. We hope this will help us keep the airways and lung function of children normal until they are old enough to start a modulator which significantly slows decline in lung function but will not recover all of what was lost.

I would like to thank the Bonnell Foundation both for their work on behalf of the CF community and for providing a platform for me to discuss a few of the projects we are working on at our center. This is both an exciting, and scary, time to care for people with CF and it is my privilege to continue to work with the CF community.

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