

Cystic Fibrosis Research News

Title:

Nutritional Considerations for a New Era: A CF Foundation Position Paper

Lay Title:

Nutritional Considerations for a New Era: A CF Foundation Position Paper

Authors:

Amanda Leonard¹, Julianna Bailey², Amanda Bruce³, Shijing Jia⁴, Adam Stein⁵, Judith Fulton⁶, Meagan Helmick⁷, Marina Litvin⁸, Alpa Patel⁹, Kate E. Powers¹⁰, Elizabeth Reid¹¹, Senthilkumar Sankararaman¹², Cristen Clemm¹³, Kim Reno¹³, Sarah E. Hempstead¹³, and Emily DiMango¹⁴

Affiliations:

1. The Johns Hopkins Children's Center, Baltimore, Maryland, USA
2. The University of Alabama at Birmingham, Birmingham, Alabama, USA
3. University of Kansas Medical Center, Kansas City, Kansas USA
4. University of Michigan Medical School, Ann Arbor, Michigan, USA
5. Northwestern Medicine, Feinberg School of Medicine, Chicago, Illinois, USA
6. Children's Hospital Colorado, Aurora, Colorado, USA
7. Community Advisor to the Cystic Fibrosis Foundation, Bethesda, Maryland, USA
8. Division of Endocrinology, Metabolism and Lipid Research. Washington University School of Medicine. St. Louis, MO, USA
9. Nationwide Children's Hospital, Columbus Ohio, USA
10. Albany Medical Center, Albany, New York, USA
11. Children's Hospital of Philadelphia, Philadelphia, Pennsylvania, USA
12. UH Rainbow Babies & Children's Hospital, Cleveland, Ohio, USA
13. Cystic Fibrosis Foundation, Bethesda, Maryland USA
14. Columbia University Irving Medical Center, New York, New York, USA

What was your research question?

With increasing survival in PwCF, the impact of increasing overweight and obesity on CF outcomes, such as pulmonary status, diabetes and cardiometabolic risk, warrants further investigation and updated recommendations for nutrition management.

Why is this important?

Improved care and use of highly effective modulator therapy has led to a high rate of overweight and obesity in PwCF. The traditional high fat, high calorie CF diet may have

Cystic Fibrosis Research News

negative nutritional consequences as PwCF age; following population-based guidelines may be more appropriate. Individuals with CF may have poor diet quality, food insecurity, distorted body image, and an increased incidence of eating disorders. Further study is needed to better define optimal treatments in PwCF in many areas including diet makeup, vitamin monitoring/supplementation, pancreatic function, sodium needs, treatment of overweight and obesity.

What did you do?

A multidisciplinary workgroup, including physicians from various disciplines, dietitians, a psychologist, social worker, PwCF, and representatives from CFF was formed to consider important nutritional topics in the current era for both children and adults with CF, emphasizing changes in nutritional status following introduction of HEMT. Surveys were distributed by email utilizing SurveyMonkey® to determine topics important to PwCF, their families, and healthcare providers (HCP). While there is insufficient evidence to develop a formal guideline, the need for guidance led the North American Cystic Fibrosis Foundation (CFF) to organize a multidisciplinary workgroup to develop this position paper.

What did you find?

The relationship between pulmonary and cardiovascular outcomes and nutritional status needs to be better defined as the CF population ages. As PwCF develop overweight and obesity, insulin resistance may play a role in cystic fibrosis related diabetes (CFRD); treatment of CFRD may be changing with the introduction of HEMT. Emerging data suggests that HEMT may improve/ reverse pancreatic insufficiency in some young children with CF. Exercise is important for cardiovascular health and maintaining a healthy weight in pwCF. Current guidelines for weight loss in the general population offer some guidance, but caution is required in extrapolating these recommendations to PwCF.

What does this mean and reasons for caution?

The main take home from our work is that we need more research in the area of nutrition and HEMT. We know that HEMT impacts nutrition but are not certain if it impacts everyone in the same way. As more information becomes available, it will be possible to have more concrete guidelines. Until that time nutritional approaches should be individualized based on the available data for each pwCF.



Cystic Fibrosis Research News

What's next?

The next step is to continue to study the impact of HEMT on nutrition in pwCF so that we can develop appropriate guidelines.

Original manuscript citation in PubMed

<https://pubmed.ncbi.nlm.nih.gov/37230807/>

Cystic Fibrosis Research News

cfresearchnews@gmail.com