2020 Cystic Fibrosis Evidence Analysis Center Evidence-Based Nutrition Practice Guideline

Featuring :: Catherine M McDonald, PhD, MS, RDN, CSP

TRANSCRIPT

Maura: I have a son named Henry. He's 15. He's learning to drive. Into video games and skateboarding and really excellent music. He's taller than me now and believes nothing in the world can stop him, and he's right.

Maura: But when he was a baby, he had serious problems with his lungs. I remember taking Henry to his pediatrician one winter morning after yet another one of his asthma attacks and feeling all the noise leave the room and all the warmth leave my face when his doctor asked what I knew about Cystic Fibrosis. I understood enough to know it was a gut-punch of a diagnosis because, back then, CF kids barely made it to adulthood. His doctor sent him to our local children's hospital for tests, saying, "Look, don't worry yet, I'm just ruling things out."

Maura: So, we go Henry tested. And while we were at the hospital, I met another family whose son was about Henry's age, with similar symptoms. He was smiley and playful, just like my son. And his parents had the same look of masked terror I had. Henry's results were negative, thank goodness. No Cystic Fibrosis. But I don't know how the other boy fared. Even after all these years, I think of him, I think of his parents, all the time. I hope their news was good and that their boy is growing and learning to drive and listening to excellent music, just like mine is. I hope that boy has a shining future ahead of him.

Maura: I'm Maura Bowen, podcasting for Abbott Nutrition Health Institute, and I'm pleased to have Dr Katie McDonald (PhD, MS, RDN, CSP) here with me today. Dr McDonald is a clinical dietitian at Primary Children's Hospital in Salt Lake City, Utah, here in the United States. She has joined us to discuss The Academy of Nutrition & Dietetics' Evidence Analysis Center systematic review of the literature to develop an evidence-based practice guideline for primary nutrition issues in Cystic Fibrosis. Dr McDonald played a key role in this review, and the guideline has been published in the *Journal of the Academy of Nutrition and Dietetics*.

Maura: Hi Dr McDonald. Thank you for joining me today.

Dr McDonald: Such a pleasure Maura. Thank you for your compelling story at the beginning, and I'm so happy for the results that you got.

Maura: Oh, yes, me, too. Thank you! Before we get started, I'll remind everyone that we're still in the middle of a pandemic and practicing safe social distancing, so Dr McDonald and I are both dialing in for this discussion, and the sound quality may sound a little bit different from what you're used to hearing.

Maura: So, Dr McDonald, to start us off, can you tell our listeners a little bit about yourself and your background?

Dr McDonald: Thank you, Maura. As you said, I have been a clinical dietitian at Primary Children's Hospital in Salt Lake City for over 40 years. I spent the first 12 years of that career saying I never wanted to work with cystic fibrosis. The prognosis was just too grim, and I found working with parents of these children was so heartbreaking. However, about thirty years ago improvements in CF care really began to shift from a reactionary model of rescuing from pulmonary exacerbations to a more preventive model. I had an opportunity to work with pulmonologists, people who also worked with CF, and I was really impressed with the pulmonologists' interest in the benefits of good nutrition for children with all kinds of pulmonary disease. When the CF center opened at Primary Children's, I campaigned for that dietitian spot.

Dr McDonald: Both CF Center and Primary Children's have a very strong foundation in quality improvement efforts. It has been such a great opportunity for me to participate in the ongoing process of QI and to see the beneficial outcomes associated with strong guidelines and standardization of care.
Also, I have volunteered for various committee assignments with the Academy of Nutrition and Dietetics and the Evidence Analysis Center over the years. These were great learning experiences. And when the call went out for applications for the AND EAL CF Nutrition guidelines committee, my hand went up. I knew this was going to be a really worthwhile project.

Dr McDonald: Primary Children's CF Center is a top-performing center, one that's been recognized by the CF Foundation. And I am so proud of the work that we do. It's exciting to see the accomplishments and good health of the individuals that we serve. I said how heartbreaking is was to work with the parents of children with CF years ago. Now, those families see a path to a long, fulfilling life for their children. They have hope. It is a tremendous amount of work, blood, sweat, and tears to get there, but the hope is real. That is the best part of my job. And I'm hoping for that little boy that was with your son when he was screened for CF, I hope that his future is bright regardless of a diagnosis of CF—for him and for all of the other children out there.

Maura: That's such a great point, and I so agree with you. And I want to thank you, too, for the helpful background. So, Dr McDonald, let's set the stage a bit. What is Cystic Fibrosis?

Dr McDonald: Cystic Fibrosis is a recessive, genetic disease; it's a progressive disease. It used to be listed as a fatal disease, and now, we talk of it as a chronic disease. It can be life-threatening, it can be life-shortening, but our hope is that this is not the case; that people with CF will lead long, productive lives. There are probably close to two thousand different genetic mutations that can combine to cause Cystic Fibrosis—each parent has to contribute one gene—and so as you can imagine there are many different iterations of the same process. It's all Cystic Fibrosis, but in various shades and degrees. Probably one in 25 Caucasians would be carriers. It's more common in Caucasians, but it is present in all ethnicities, just to a varying extent.

Maura: What are some of the hallmark symptoms of Cystic Fibrosis?

Dr McDonald: The hallmark symptoms of Cystic Fibrosis really stem from a thick, sticky mucus. It's caused at the cellular level, the CFTR Cystic Fibrosis transport protein, that transports sodium and chloride molecules across the cell wall. It's this thick, sticky mucus that causes the problems that we see in CF, including the accumulation of thick, sticky mucus in the lungs; there's an accumulation of this same mucus within the pancreas that decreases the flow of the pancreatic juices and enzymes and causes an autodigestion of the pancreas and therefore the pancreatic function. But any organ within the body that secretes mucus can be affected. And as we learn more about CF, we're seeing more and more effects. But primarily what you'd be looking at would be digestive issues. About 85% of people with Cystic Fibrosis require pancreatic enzyme replacement. The manifestation of pulmonary symptoms has become probably the hallmark of CF. But there are all kinds of problems people with CF have to deal with.

Maura: So as I was preparing for this conversation, I was happy to learn about some of the recent advances in Cystic

Fibrosis research and the role nutrition can play in addressing and managing the disease. Now, I know you've been instrumental to some of this research, and so I'm wondering if you can tell us about some of the accepted evidence on Cystic Fibrosis and nutrition leading up to your most recent investigation?

Dr McDonald: Thank you. The accepted evidence presents a very compelling argument for appropriate nutrition in individuals with CF. From the initial descriptions of Cystic Fibrosis about 80 years ago, nutrition has really been one of the pillars of treatment of this chronic, progressive disease. A turning point occurred about 30 years ago with the increased recognition of the importance of good nutrition not just for the prevention of overt malnutrition. A study compared matched CF groups in Canada and the US. The Canadians followed a more liberal, higher-fat, higher-calorie diet, more enzymes. And they were significantly heavier and taller than their American counterparts. The Canadians were found to have a nine-year survival advantage over the Americans. Since that time, there has been increasing evidence that good nutrition and the best possible lung function are very closely associated. One does not occur without the other. Furthermore, the earlier in life an individual with CF attains normal growth and nutrition parameters, the better they will do long term. And lastly, the new medications that address CF at the cellular level are game changers for the lives of persons with CF. These CFTR modulating drugs have the potential to normalize the lifespan for individuals with CF. We know that the legacy CF diet with high-fat, high-calories, really is going to need to adapt to the new paradigm for living into a healthy old age with CF.

Maura: So, can you tell us: What was the specific aim of your systematic review?

Dr McDonald: We first of all we knew there had been extensive CF nutrition guidelines were already available through the hard work of the Australian/New Zealand CF dietitians' group and a multidisciplinary European CF nutrition consensus group. The CF Foundation has a number of evidence-informed, nutrition-related guidelines, such as enteral nutrition for CF. So, we knew there was a lot of evidence out there already. It was our aim to really complement and add to the existing knowledge base and not duplicate efforts. So, the AND EAL committee really focused on evidence -based, practice guidelines for primary nutrition issues in CF across the lifespan. We chose to emphasize guidelines for medical nutrition therapy for individuals with CF that would be provided by the RDN or international equivalent. The main objective was to provide evidence-based recommendations to RDNs or the international equivalents who deliver Medical Nutrition Therapy (MNT) to improve health and prevent disease progression in CF.

Maura: Can you tell us what prompted this study?

Dr McDonald: Well, the systematic review was commissioned by the Medical Nutrition Therapy Practice Group of the Academy of Nutrition and Dietetics. The CF sub-group of the MNT practice group recognized a need for a closer examination of what was known about nutrition for CF and specifically nutrition for adults with CF. That was a place where the evidence was really slim. The flip side of this was to identify the gaps in knowledge and where further investigation is needed.

Maura: What did you expect to find with this review?

Dr McDonald: Well, personally, I felt I had a pretty good idea of the research demonstrating the value of good nutrition and CF, specifically in pediatrics where it's the strongest. I expected to find strong evidence on the value of early intervention in nutritional health for CF.

Maura: And what did the data actually show?

Dr McDonald: Well, first of all, I need to outline exactly what we were looking for, because again, as I said, there were already bodies of evidence that had been put together by Australia and New Zealand group and the European Union group. And so, what we were looking for depended on a narrowed focus, and we really decided to kind of laser in on six questions. And those were:

Dr McDonald:

1. How does MNT affect nutrition-related outcomes, such as morbidity and mortality in lung function in CF?
2. Which nutrition screening and assessment methods by the RDN are reliable and valid?
3. What are the longitudinal relationships between nutritional assessment parameters and pulmonary function?
4. What are accurate methods of measuring energy expenditure in CF?
5. What is the relationship between dietary intake and nutrition-related outcomes in CF?
6. What is the impact of CFTR modulating drugs on dietary intake, weight gain and body composition in CF?

Dr McDonald: From these six questions, there grew twenty-eight recommendations, and these recommendations really provide guidance on medical nutrition therapy including frequency and components of nutrition screening, nutrition assessment and dietary intake.

Dr McDonald: I think the strongest piece of this work is that it provides a solid foundation for dietetic practice when working with CF in either an outpatient or inpatient basis.

Dr McDonald: In terms of the article, I feel like one of the most valuable components of this work is an overview nutrition topics table and recommendations for which of the different guideline (Australian/NZ, EU, JAND, CFF) addresses the topic

Maura: Now did anything surprise you about the findings?

Dr McDonald: It didn't surprise me so much, but it pleased me to find that there is no real basis for the CF "legacy" diet, the high fat, high calorie diet with low nutritional value. With the evidence that we were able to establish, we were able to recommend a shift away from this diet with a very low nutritional value toward a more balanced, wholesome diet for individuals with CF. I guess I was surprised that we were able to put that together.

Maura: What learnings from this review can or should clinicians apply to their practice?

Dr McDonald: For RDN who work with CF or for their administrators, these guidelines provide specific recommendations for medical nutrition therapy, numbers of RDN FTEs required in clinical settings, the frequency, method and components of pediatric and adult nutrition screening and assessment, how to deal with CFTR modulators and nutrition assessment, looking at energy requirements, body composition assessment, and nutrition assessment of biochemical values, and then there's just general guidance for food intake, including comorbidities such as CF related diabetes and obesity.

Maura: So you said earlier that you were pleased by your findings, but what makes you feel hopeful?

Dr McDonald: I am hopeful that these recommendations will assist in moving medical nutrition therapy into the next and very exciting phase of CF treatment, one that focuses on individualized nutrition for a long and healthy life with CF. Another hope is that we will be able to collaborate for improved CF nutrition across international borders. I see a very bright future for people with CF, one that will require a lot of work and thought, but still, it's a future and it's bright.

Maura: Can you think of additional research that should be done that could be helpful in this space?

Dr McDonald: That's an excellent question, and you know, I think one of the strongest messages from this work, and that people will take from reading this article, is the great need for additional nutrition-related research, especially in the fields of nutrition for individuals who are receiving these CFTR modulating drugs—to look at the outcomes and to move toward a shift toward a more balanced, health focused diet. I think all of that is very much needed in the research and is something we're really looking forward to seeing in the literature.

Maura: Dr McDonald, thank you so much for your time today. This was so interesting. And we appreciate all you're doing to help build awareness for the role nutrition can play in addressing and managing Cystic Fibrosis. So, thank you.

Dr McDonald: Thank you for this opportunity.

Maura: Now, for our listeners, if you're hoping for more podcast episodes on nutrition topics, rest assured we're continuing to develop a series to help support you. You can find these recordings on anhi.org by clicking "RESOURCES" then "PODCASTS & VIDEOS." And, if you become an anhi.org member today—which you can do by clicking "REGISTER" at the top of our homepage—you'll receive regular nutrition science news updates from our team. And, of course, you can also follow the Abbott Nutrition Health Institute on LinkedIn.

Maura: Finally, our website, anhi.org, has a series of printable resources related to topics like this. And you can find these resources on anhi.org by clicking "RESOURCES" and "PRINTABLE MATERIALS."

Maura: Thanks everyone. Stay healthy and safe.

REFERENCE

Academy of Nutrition and Dietetics: 2020 Cystic Fibrosis Evidence Analysis Center Evidence-Based Nutrition Practice Guideline

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