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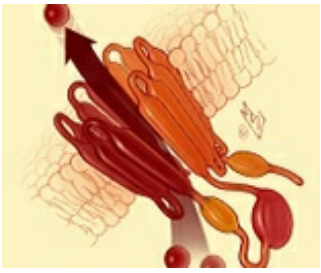

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Nutritional Management for Individuals with CF: Practical Applications

Our guest author is Amanda Leonard, MPH, RD, CDE, from the Johns Hopkins Children's Hospital in Baltimore, MD.

After participating in this activity, the participant will demonstrate the ability to:

- Describe the nutrition-related recommendations in the CF Foundation's Preschool Guidelines;
- Summarize the recommendations in the enteral feeding guidelines and how to apply those in a clinical setting.
- Discuss the impact of ivacaftor on weight in children with cystic fibrosis.

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MEET THE AUTHOR

Amanda Leonard, MPH, RD, CDE
Advanced Nutrition Practitioner
The Johns Hopkins Children's Center
Baltimore, Maryland

Guest Faculty Disclosure
Amanda Leonard has indicated that she has served as an advisor/consultant to Alcresta.

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Director, Cystic Fibrosis Center
Professor of Pediatrics
The Johns Hopkins University
Baltimore, Maryland

Noah Lechtzin, MD
Director, Adult Cystic Fibrosis Program
Associate Professor of Medicine
The Johns Hopkins University
Baltimore, Maryland

Suzanne Sullivan, RN, BSN
Senior Clinical Nurse
Johns Hopkins Hospital
Baltimore, Maryland

Podcast Transcript

BOB BUSKER: Welcome to this *e*CysticFibrosis Review podcast.

I'm Bob Busker, managing editor of the program. We're here today with Amanda Leonard, an advanced nutrition practitioner at the Johns Hopkins Children's Center in Baltimore. We're here to discuss her recent newsletter issue on New Guidelines in Nutritional Management for Patients with Cystic Fibrosis.

***e*CysticFibrosis Review is jointly presented by the Johns Hopkins University School of Medicine and the Institute for Johns Hopkins Nursing. This program is supported by educational grants from Gilead Sciences Inc, Vertex Pharmaceuticals Incorporated, AbbVie Inc, and Chiesi USA Inc.**

Learning objectives for today's audio program include:

- Describe the nutrition-related recommendations in the CF Foundation's Preschool Guidelines.
- Summarize the recommendations in the enteral feeding guidelines and how to apply those in a clinical setting.
- Discuss the impact of ivacaftor on weight in children with cystic fibrosis.

Amanda Leonard has disclosed that she has served as an advisor/consultant to Alcresta. She has indicated that there will be no references to the unlabeled or unapproved use of any drugs or products in today's discussion.

Amanda, thank you for joining us today.

AMANDA LEONARD: Thank you so much for having me, Bob. I'm excited to be here.

BOB BUSKER: **I want to begin our discussion today with a quotation. "As the field of nutrition in cystic fibrosis changes and evolves, new guidelines are needed to ensure that practitioners are provided up-to-date information."**

That's from your newsletter issue, Amanda. You wrote that. And then you went on to review two of the newer guidelines from the Cystic Fibrosis Foundation and explain the evidence behind the recommendations. I'd like to look at the benefits of applying that guidance from a clinical perspective. So please start us out with a patient scenario.

MS. LEONARD: I have a preschool patient we've been following. AJ's a three-year-old female whose CF was diagnosed at birth. At her last clinic visit, her weight was at the 5th percentile, BMI was at the 35th percentile, and her average daily weight gain was about 4 gm/day. Mom reports she's always been a picky eater and has a long history of food refusal. It sometimes takes her up to 45 minutes to finish a meal. The family's tried a variety of supplements at home, but she doesn't seem to like any of them.

She complains about bellyaches five or six times a week, and her enzyme dose seems to be reasonable at about 2200 units of lipase per /kg/meal.

MR. BUSKER: **The first thing I want to ask you is: Is this three-year-old at nutritional risk? What does the guidance say?**

MS. LEONARD: That's a great question, Bob. According to the new CFF preschool guidelines, AJ is at nutritional risk. Her BMI is less than the 50th percentile, and her weight for age is less than the 10th percentile. She's also not meeting the average daily weight gain goals of 6 gm/day. She's only been gaining about 4 gm/day. So according to all three criteria, she meets the definition of nutritional risk.

MR. BUSKER: How would you assess this patient?

LEONARD: There's a few different areas that I would look at. First, I would take a close look at the diet history and see what is AJ really taking in. It sounds like it's taking her a long time to eat. How many calories is she eating during those mealtimes?

I'd get more details about the parent-child interaction. Is there frustration or nagging? Is her mealtime stressful?

I would also want to find out a little more about the bellyaches and the stool history. Because five to six times per week for bellyaches is more than we want in any of our patients with CF.

MR. BUSKER: **Based on what you find out from that assessment, what are some of the initial interventions you might**

recommend?

MS. LEONARD: Depending on the answers to some of the questions, we might change around either the dosing of the enzymes or maybe the timing to see if that can help with the bellyaches or look at if there's some other adjunct therapies like adding acid suppression to maybe help with the enzyme efficacy and also see if we might want to have some sort of behavioral intervention or time limit on meals to help with the parent-child interaction and maybe limit some of the stress around mealtimes.

I definitely would want to see AJ back at clinic in eight weeks, or even sooner to see if the interventions helped. If not, we would need to move onto the next step.

MR. BUSKER: **Talk to us about that next step. Would that include referral to a specialist?**

MS. LEONARD: When AJ comes back to clinic, depending on her weight gain, it could be that we would just continue with the current plan and keep a close eye. If she's meeting the average daily weight gain and is closer to meeting BMI in weight-for-age percentile goals, we could just stay the course.

But oftentimes, especially with the preschoolers, a referral to a behavioral psychologist for a little bit more in-depth behavioral intervention could be really helpful. And also with the bellyaches, an assessment by a gastroenterologist to evaluate if there's some other cause, if making little tweaks to the enzymes isn't helping, to find out if there's some other root cause for the bellyaches to help us figure out the whole picture.

MR. BUSKER: **Let's suppose she was referred to a gastroenterologist for assessment. What might the GI be looking for?**

MS. LEONARD: That's a great question, Bob. The gastroenterologist would be looking for other causes besides malabsorption or inadequate enzyme replacement causing her bellyaches. Could be constipation, obstipation, maybe reflux, small bowel overgrowth, celiac disease, motility disorders — lots of other things beyond just pancreatic insufficiency could be causing her bellyaches. We want to make sure those are adequately treated.

MR. BUSKER: Let's assume that the gastroenterology findings are all negative, that there's nothing untoward going on. What might you then suggest for this patient? Would appetite stimulants be appropriate?

MS. LEONARD: AJ's a little on the young side for appetite stimulants. At our institution, we don't use them in patients that young. Our next line of defense, if the behavioral therapy is not as helpful and the gastroenterologist workup is negative, would be to think about an enteral feeding tube. We would probably be talking to the family, and hopefully would've brought it up already as a potential option to give calories when AJ's not able to take in adequate calories by mouth.

MR. BUSKER: Thank you for that case and discussion, Amanda. We'll return, with Amanda Leonard from the Johns Hopkins Children's Center in just a moment.

MR. BUSKER: This is Bob Busker; I'm the managing editor of *e*CysticFibrosis Review.

*e*CysticFibrosis Review is a combination newsletter and podcast program delivered via email to subscribers. Newsletters are published every other month. Each issue reviews the current literature in areas of importance to pulmonologists, gastroenterologists, infectious disease specialists, pediatricians, respiratory therapists, dietitians, nutritionists, nurses, and physical therapists.

Bimonthly podcasts are also available as downloadable transcripts, providing case-based scenarios to help bring that new information into practice in the clinic.

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MR. BUSKER: Welcome back to this *e*CysticFibrosis Review podcast. I'm Bob Busker, and we're here today with Amanda Leonard from the Johns Hopkins Children's Center. We've been discussing how the New CF Foundation Guidelines in Nutritional Management can be applied in the clinic. So to continue in that vein, Amanda, if you would please bring us another patient scenario.

MS. LEONARD: JT is a 24-year-old male with CF, and he's had a history of poor weight gain most of his life. His current BMI is 20.2 kg/m2, below the goal of 23. He's been on appetite stimulants, and he reports that he eats all day long. He has one to two formed stools per day and no problems with bellyaches, and his enzyme dose is about 2000 units of lipase/kg/day. JT came to clinic today to see what we might be able to offer for him for his desire to improve his weight.

MR. BUSKER: Take us through your steps in assessing this 24-year-old.

MS. LEONARD: According to the new enteral feeding guidelines, we want to make sure to rule out other causes for malnutrition, make sure that JT doesn't have CF-related diabetes or other GI issues that have been undiagnosed. I would also want to assess his intake and see how many calories is he getting when he's eating all day and make sure he's eating high calorie feeds all day, not fruits and vegetables that may fill you up but don't help with the weight gain.

Another thing I would look for is barriers to intake. Are there financial issues? Does he run out of money for food? Is he busy with his job and he doesn't have time to make food or is he trying to eat on the run?

Once we've looked at all of those potential options or potential issues, we would also discuss options that we have with JT for gaining weight, looking at ways to increase his intake, see if he wants to try other supplements, and also bring up the idea of enteral feeding as an option if we can't get his weight to improve with oral intake.

MR. BUSKER: **What other workups might you recommend for this patient?**

MS. LEONARD: I think JT should definitely have an oral glucose tolerance test (OGTT) to make sure he doesn't have CF-related diabetes or just impaired glucose tolerance. If his OGTT is abnormal, we would refer him to an endocrinologist. I also think an assessment by a gastroenterologist to see if there are other causes for inadequate weight gain, if there's constipation or bacterial overgrowth, or other things going on that are causing him to have inadequate gains.

MR. BUSKER: Let's assume the endocrinologist and the gastroenterologist do not find anything relevant. Would it be time to seriously consider enteral tube feeding?

MS. LEONARD: Absolutely. I think that that would be the logical next step

MR. BUSKER: **Wow. Would you present that to the patient and the family? How would you involve them in making this decision?**

MS. LEONARD: I would want to have been talking about enteral feeds as an option from the beginning so JT and his family knew that this is something that might be a good treatment option for them in the future. We want to make sure that we provide information about all of the options, including tubes, and discuss the pros and cons of each so that they can play an active role in the decision-making process.

Another thing that's important is to make sure that we answer questions and also provide them with information, written or electronic or verbal, however they seem to receive information best, so that they can think about it and process it and come up with any questions they may have. Especially with our adult patients, we want to make sure they're involved in the goal-setting. So we will set goals, and if they achieve goals with the tools we're using, then we can continue along that path. But then if it becomes more obvious that we're setting goals for weight gain that are not achieved, we need to try something new, and this is where we would tie back to the enteral feedings as one of the options.

MR. BUSKER: **What kind of barriers have you encountered when you explain to a patient that an enteral tube is the most effective way to go?**

MS. LEONARD: There's a variety of barriers. A common one is they're worried that they won't be able to eat by mouth anymore, and we generally give them their enteral feedings at night so they can eat what they want during the day. Another common concern is that they won't be able to be as active as they were. But with proper care they should be able to participate in all the activities they did before, obviously in consultation with their doctor. I think education to reassure. When you come up with the barriers and they have questions, to make sure that you have an open discussion so they can feel like they're getting the information they need to make the decision that's right for them.

MR. BUSKER: Let's assume this patient does accept enteral feeding. What kind of formula and what kind of schedule would you recommend? What does the new CF Foundation guidance say?

MS. LEONARD: The guidelines do not recommend a specific kind of formula because there isn't enough data. At our institution we generally start with an intact formula and then assess tolerance. The guidelines recommend nighttime continuous feeds as the starting point for almost all patients. This allows us to adjust the nighttime feeds based on their schedule and allows them to eat during the day.

So what we do at our center is we try and figure out how long the patient's gonna be asleep and how many calories we want to give them during that time, and that lets us figure out how long and how fast to run the feeds. Because obviously you don't want to have the feeds run for 12 hours if someone's only going to sleep for six hours because then they're hooked up to their tube for six hours when they're awake. We really want to try and make it fit into their life.

MR. BUSKER: **Thank you for that case and discussion, Amanda. And let's continue, if you would please, with one more patient scenario.**

MS. LEONARD: Our last patient is KA, who's a seven-year-old female with CF. She has one copy of the G551D mutation. She reports at her clinic visit that she eats well and is on a high-calorie, high-fat diet and has been since birth, when her CF was diagnosed. She's been on ivacaftor for about six months and has gained 4 kg in the past three months. Her BMI at the

visit today is at the 85th percentile, which is up by 35% from her last clinic visit. Her enzyme dose is about 2,125 units/kg/meal, and her mother is concerned about the weight gain and wants to stop the enzymes.

MR. BUSKER: What's your assessment of this child? Is her mother right to be concerned about her weight gain?

MS. LEONARD: KA's rate of weight gain is definitely faster than expected. She was meeting the CF Foundation BMI goals starting ivacaftor, and since being on ivacaftor, obviously her intake or maybe her absorption has improved. For some reason her rate of weight gain has increased. We want to be careful that she doesn't become overweight or obese, because that is not our goal for the CF population.

MR. BUSKER: What about stopping her pancreatic enzymes?

MS. LEONARD: I definitely would not stop the enzymes. There's no good evidence that pancreatic function returns with ivacaftor. I think the absorption might be better, and the lungs are functioning better, so the caloric needs are less. But we definitely don't want to completely stop the pancreatic enzymes. I think that would have potentially negative impact because it might cause an obstruction.

MR. BUSKER: **It's kind of a very new thing for clinicians to have to worry about: obesity in a patient with CF.**

MS. LEONARD: I agree that it's definitely a newer finding that we're worrying about obesity in the CF population. As we get newer treatments and therapies and nutrition is improving, it's definitely something that we need to keep an eye on.

MR. BUSKER: **So in this 7-year-old, what diet changes would you suggest?**

MS. LEONARD: That's a really good question, Bob, and for KA I would suggest following a lower calorie diet. I'd check to see how many snacks she's having and maybe cut back on some of the junk food and switch from whole milk to 2% milk or maybe even to skim milk. Basically move more toward heart healthy diet that's the recommendation for the general population.

MR. BUSKER: **Overall, what would your nutrition goals be for this patient?**

MS. LEONARD: At our center, our goals would be to have her BMI between the 50th and the 85th percentiles, and we would want her to follow a heart healthy diet. I would want to continue to monitor her weight gain and rate of weight gain and try and identify trends so that we can make changes as indicated. So it could be that we need to encourage her to increase her activity in addition to having a healthier diet or a lower diet. We would want to keep track of how she is doing and make sure that she is maintaining a healthy weight.

MR. BUSKER: **So with that goal in mind, how would you monitor her?**

MS. LEONARD: I would definitely want to see KA at least every three months to see if we needed to make any additional changes. As she's working on decreasing her intake, we would want to see if her weight gain's still faster than we want it to be or has it plateaued, and we also want to make sure she doesn't go too far in the other direction. Sometimes when you suggest that people cut back, they might be a little overzealous and then end up losing weight — which is not our goal. We want her to grow into her weight, because as a seven-year-old we're not looking for any weight loss. We just want to slow down her rate of weight gain. I think close monitoring at follow-up visits at least every three months would be the best way to keep track.

MR. BUSKER: **Thank you for today's cases and discussion, Amanda. Let me change gears on you a little bit here, and ask you to look to the future for us. What changes do you expect to see in the nutritional management of patients with cystic fibrosis?**

MS. LEONARD: That's a great question, Bob. I really think that we're moving more toward mutation-specific treatment recommendations. I think at some point we're going to say this is your mutation, this is the therapy you're on, these are the nutrition recommendations. I think the idea that there will be an overarching certain way to approach CF nutrition for everybody is something that is moving to the back burner, that we're going to say, if you're on this therapy, this is how you should achieve your goals. I think the goals will be similar, but I think the way we get there will be different.

I also think as more data is available to clinicians, we can tailor our recommendations, and hopefully some new guidelines will be developed for some of the new therapies that are coming out. And I also think that at some point we might have people with CF following a heart healthy diet, just like the general population. As the treatments continue to improve, we might be able to make the high calorie, high fat diet a thing of the past.

MR. BUSKER: Thank you for sharing your insights, Amanda. Let's wrap things up now by reviewing today's discussion in light of our leaning objectives. So to begin: the nutrition-related recommendations in the CF Foundation's preschool guidelines.

MS. LEONARD: I think the big nutrition take-home messages from the guidelines were that preschoolers should have weight and height measurements taken at each visit and BMI should be calculated. Your goal BMI is greater than or equal to the 50th percentile, and your weight for age goal is greater than or equal to the 10th percentile. Nutrition risk definitions were given for this age group: BMI less than the 50th, weight for age percentile less than the 10th, a rate of weight gain less

than the 50th percentile for age, or any inappropriate weight loss would put a child in the nutrition risk category. Another helpful guideline from the publication was calorie goal. So your goal of more than 90 to 110 calories/kg/day for preschool-aged children; and protein needs, at least 13 gm/day for 2- to 3-year-olds and 19 gm/day for 4- to 5-year-olds. I think that the guidelines also really stress the need to assess for behavioral issues and refer as needed, and also make sure that GI issues are being identified and treated as needed. I think frequent follow-up and a multidisciplinary approach were two of the cornerstones of the guidelines.

MR. BUSKER: **And our second learning objective: the enteral feeding guideline recommendations and how to apply those in a clinical setting.**

MS. LEONARD: Applying these guidelines in a clinical setting is really important. You want to make sure to have a multidisciplinary approach and the appropriate workup so you assess for other causes of malnutrition before you decide a feeding tube is the right choice. The guidelines didn't give a specific suggestion for a formula, but any formula that's tolerated should be used. There are no specific recommendations for enzymes, because they did not have enough data to support one specific approach. Most feeds should start as continuous overnight feeds and allow for oral intake during the day. You want to make sure that the patient receives education before the tube is placed, during the tube process, and also after discharge to make sure all of their questions are answered and that they're part of the process.

MR. BUSKER: **And our final objective: the impact of ivacaftor on weight in children with CF.**

MS. LEONARD: Ivacaftor has definitely been shown to increase the rate of weight gain in children with CF. It's something that we want to make sure that we monitor so that our children maintain a healthy weight and don't gain weight too quickly. This is a new finding in the CF community that we're worried about obesity, but I think that it's something that could definitely become a problem if it's not monitored appropriately.

MR. BUSKER: **From the Johns Hopkins Children's Center — Amanda Leonard, advanced nutrition practitioner — thank you for participating in this eCystic Fibrosis review podcast.**

MS. LEONARD: Thank you so much for having me, Bob. I really enjoyed it.

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