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eLITERATURE REVIEW
eCysticFibrosis Review
Podcast Issue

Presented by the Johns Hopkins University School of Medicine and the Institute for Johns Hopkins Nursing

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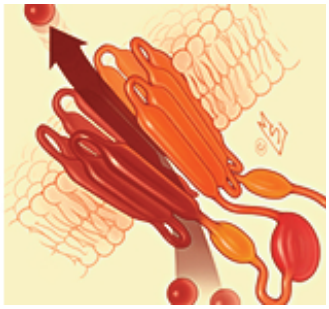

VOLUME 4 — ISSUE 4: TRANSCRIPT

Featured Cases: Strategies for the Improvement of Nutrition Outcomes

Our guest author is Amanda Radmer Leonard, MPH, RD, CDE from The Johns Hopkins Children's Center and The Johns Hopkins Cystic Fibrosis Center.

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

- Describe approaches to the treatment of low vitamin D levels in patients with cystic fibrosis,
- Discuss the variability of fecal elastase result during the first year of life,
- Summarize the importance of tailoring nutritional interventions to specific patient and family situations.

This discussion, offered as a downloadable audio file and companion transcript, covers the important issues related to Nutrition in the format of case-study scenarios for the clinical practice. This program is a follow up to Volume 4, Issue 3 *eCysticFibrosis Review Newsletter* – Strategies for the Improvement of Nutrition Outcomes.

The Johns Hopkins University School of Medicine takes responsibility for the content, quality, and scientific integrity of this CME activity.

MEET THE AUTHOR

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Unlabeled/Unapproved Uses
The author has indicated that there will be no references to unlabeled or unapproved uses of drugs or products.

Guest Faculty Disclosure
The author has indicated that she has served as a consultant for Abbott Pharmaceuticals.

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May 29, 2013

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May 28, 2015

Next Month's Topic
Pseudomonas aeruginosa Eradication

CME/CE INFORMATION

⇩ PROGRAM BEGINS BELOW

ACCREDITATION STATEMENTS

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For United States: Visit this page to confirm that your state will accept the CE Credits gained through this program.

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There are no prerequisites to participate in the activity.

LAUNCH DATE

This program launched on February 28, 2013, and is published monthly; activities expire two years from the date of publication.

STATEMENT OF NEED

Based on a review of the current literature, including national and regional measures, detailed conversations with expert educators at Johns Hopkins, and a survey of potential program participants, this program will address the following core patient care gaps:

Disease-Modifying Therapies

- Clinicians may be unfamiliar with recently introduced disease-modifying therapies and how they are altering the therapeutic landscape for patients with cystic fibrosis.
- Clinicians may be uncertain how to integrate genotyping into therapeutic decisions and how to communicate with patients and families about the relationship between genotype and therapy

Nutrition

- Many clinicians lack strategies to persuade patients to adhere to CF nutritional requirements, resulting in low body weight and nutritional failure in patients with cystic fibrosis.
- Many clinicians remain uncertain how to optimize pancreatic function in patients with cystic fibrosis.

Treating CF Patients with Inhaled Antibiotics

- Clinicians lack knowledge about the use of existing and emerging inhaled ABX to treat chronic pulmonary infections.
- Clinicians need more information to make informed decisions about the use of inhaled ABX in combination.
- Clinicians lack information about best practices for scheduling ABX therapy to suppress chronic airway infections.
- Common clinician assumptions about treating pulmonary exacerbations lack supporting evidence.
- CF clinicians are not aware of and/or are not actively advocating inhaled ABX patient-adherence strategies.

SUCCESSFUL COMPLETION

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Estimated time to complete activity: 30 minutes.

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Guest Author Disclosures

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INTENDED AUDIENCE

This activity has been developed for pulmonologists, pediatric pulmonologists, gastroenterologists, pediatricians, infectious disease specialists, respiratory therapists, dieticians, nutritionists, nurses, and physical therapists.

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HARDWARE & SOFTWARE REQUIREMENTS

PC: Internet Explorer (v6 or greater), or Firefox
MAC: Safari

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

*e*CysticFibrosis Review is presented by the Johns Hopkins University School of Medicine and the Institute for Johns Hopkins Nursing. This program is supported by educational grants from Aptalis Pharma, Inc; Gilead Sciences Inc.; and Vertex Pharmaceuticals.

Today's program is a companion piece to our eCysticFibrosis Review newsletter topic, *Strategies for the Improvement of Nutrition Outcomes*.

Our guest today is that issue's author, Amanda Radmer Leonard from The Johns Hopkins University School of Medicine.

This activity has been developed for pulmonologists, pediatric pulmonologists, gastroenterologists, pediatricians, infectious disease specialists, respiratory therapists, dietitians, nutritionists, pharmacists, nurses and nurse practitioners, physical therapists, and others involved in the care of patients with cystic fibrosis.

The Accreditation and Credit Designation Statements can be found at the end of this podcast. For additional information about accreditation, Hopkins policies, and expiration dates and to take the post-test to receive credit online, please go to our website newsletter archive, www.eCysticFibrosisReview.org, and click the Volume 4, Issue 4 podcast link.

Learning objectives for this audio program are that after participating in this activity, participants will demonstrate the ability to:

- Describe approaches to the treatment of low vitamin D levels in patients with cystic fibrosis,
- Discuss the variability of fecal elastase results during the first year of life, and,
- Summarize the importance of tailoring nutritional interventions to specific patient and family situations.

I'm Bob Busker, managing editor of eCysticFibrosis Review. On the phone we have with us Amanda Radmer Leonard, a pediatric nutrition practitioner at the Johns Hopkins Cystic Fibrosis Center and the Johns Hopkins Children's Center, both part of The Johns Hopkins University School of Medicine.

Ms. Leonard discloses that she has served as a consultant for Abbott Pharmaceuticals and indicated that there will be no reference to unlabeled or unapproved uses of drugs or products in her discussion today.

Amanda Leonard, welcome to this eCystic Fibrosis Review podcast.

MS. AMANDA LEONARD: Thank you, Bob, I'm excited to be here with you today.

MR. BUSKER: In your newsletter issue, you reviewed recent research describing new approaches to increasing the survival of patients with cystic fibrosis, as well as ways to improve their quality of life through better health and decreased comorbid complications. Today I'd like to discuss how some of that research can be applied in clinical practice. So please start us out with a patient description.

MS. LEONARD: Our first case is a five month old male. His CF was diagnosed by newborn screening shortly after birth. His medications include CF specific multivitamin, 1 mL/d, and his mother adds 1/8 teaspoon of salt per day to his feeds. The pediatrician put him on a 24 calorie per ounce infant formula and he takes about 27 ounces per day. He's fed PO ad lib. Relevant lab data includes a fecal elastase at one month of 252 µg/gµg/g, which is consistent with pancreatic sufficiency. His mother reports that he has three to four stools per day with no grease or oil noted. His weight is 8.2 kg, which is at the 29th percentile weight per age; his length is 52 cm, which is the 21st percentile length per age, and his weight per length is at the 56th percentile.

MR. BUSKER: First let me ask you about genetic testing: do you have the mutations for this baby?

MS. LEONARD: Yes, we were able to get mutations for this baby. The mutations came back as F508-del, which is a pancreatic-insufficient mutation, and R347P, which is variable. That means you don't know whether the baby is going to be insufficient or sufficient.

MR. BUSKER: How does this information affect the treatment plan?

MS. LEONARD: It won't affect our treatment plan, but it will affect what we do in the future when there is a pancreatic sufficient mutation that can be dominant. But since the R347P is variable, we don't know where the baby will end up; it could go either way. So it will be important to monitor lab and clinical data.

MR. BUSKER: In your patient description, you noted that the fecal elastase was consistent with pancreatic sufficiency at one month of age. Should the clinician then assume that this baby will always be pancreatic sufficient?

MS. LEONARD: No, that's not necessarily the case. With a variable mutation it's possible for a baby to convert from being pancreatic sufficient to becoming pancreatic insufficient. It's important to monitor the baby's growth and his stools for signs and symptoms of malabsorption, and also think about some lab data, as well.

MR. BUSKER: So regarding the lab data: when would you recheck fecal elastase?

MS. LEONARD: I would check fecal elastase again at a about one year of age or maybe sooner if there are any signs of pancreatic insufficiency such as increase in the number of stools, noticing grease or oil in the diaper, or if the baby seems to be having problems growing. At this point the weight for length is good at 56%, so the baby seems to be doing fine. But if his weight for length seemed to drop off or his mother noticed he was having other GI issues, we would definitely want to recheck it sooner.

The workup of Sullivan and colleagues discussed in the newsletter issue showed us that the fecal elastase can change over the first year of life. So if you just look at one dataset, you're not getting the full picture. They recommend for any baby whose initial value is greater than 50µg/g , that you recheck it at one year because the babies can become pancreatic insufficient over time. One infant in their study went from pancreatic insufficient fecal elastase levels to pancreatic sufficient at the one year check.

MR. BUSKER: Would you recommend starting pancreatic enzyme replacement therapy — PERT — in this baby?

MS. LEONARD: I wouldn't recommend starting PERT at this time since the clinical evidence points to the fact that the baby is pancreatic sufficient, so we don't want to just empirically start enzymes unless they're indicated by the stool history and the lab data. The baby is growing well now and there is some chance that he may end up being pancreatic sufficient. We don't know for sure.

With the variable mutations, it's hard to tell early on what the pancreatic status will be later on in life. And although many babies present with pancreatic insufficiency early on, some don't develop insufficiency until later in life. So close clinical management and monitoring is the best choice for this child.

I think it's important when you're assessing pancreatic status in a baby or in children that you look not just at one data point or one lab value, or just the clinical picture; you can look at it all together, you can take the labs, the genetics and how the baby's doing clinically, and assess what your next steps should be.

MR. BUSKER: Thank you for that case and discussion, Amanda. Please bring us another patient.

MS. LEONARD: Our next patient is a 15 year old female whose CF was diagnosed at 4 months of age. She's an active teenager whose weight is 50.5 kilos, which is the 42nd percentile weight for age, her height is 160 centimeters, which is the 38th percentile height for age, and her BMI is at the 46th percentile. Her pulmonary function is pretty good, with an FEV1 at 89% predicted. Medications include CF specific vitamin, one soft gel twice a day, enzyme replacement therapy with lipase at a dose of about 2,100 units/kg/meal, and she follows a high calorie diet with two to three snacks at home. Relevant lab data includes a 25-hydroxyvitamin D of 20 ng/mL measured at her last clinic visit.

MR. BUSKER: Just to confirm — that vitamin D level in her lab data is low. Is that correct?

MS. LEONARD: Yes, her vitamin D level of 20 ng/mL is low. The CF Foundation recommends a goal of at least 30 ng/mL in people with cystic fibrosis.

MR. BUSKER: What would you do to address the low vitamin D level?

MS. LEONARD: I'd look at a few things. One of the first would be to make sure that she's taking her CF

specific vitamins. She's a busy teenager, and sometimes they can forget to take doses, so we wouldn't want to add another medication if she's not already taking the initial amount that we think she is. And another way to make sure if she is taking her vitamins, that they're absorbed better, is for her to take them at a meal when she's taking enzymes. Since vitamin D is a fat soluble vitamin, it is absorbed better if taken with PERT at a mealtime. If she's taking her enzymes twice a day and she's taking them with a meal when she's taking her enzymes and her levels are still low, I would consider adding cholecalciferol or vitamin D3, 2000 IU twice a day.

MR. BUSKER: We know that medication adherence is a problem for all patients, even more so for teenagers. They're busy, they forget, they don't want to, they don't feel its important. What, specifically, would you do to address better adherence in this 15 year old girl?

MS. LEONARD: In teenage patients, one of the things I find most helpful is to learn what their schedule is and what their goals are. If they don't feel the medication is helping them, they may not put it at the top of their list if they only have time to do a certain number of things. So I want to make sure that teens understand why the vitamins are important, vitamin D is important for bone health, and as we saw in Grossman's work in the newsletter commentary, it's also linked to inflammation and potentially is helpful in improving relevant clinical outcomes.

Once you can get the teen to buy into the idea that vitamins are important, you can troubleshoot ways to help them remember. You can see if there's a certain time of day, such as with breakfast and dinner. Maybe they can get a pill case that they can leave out so that they see it, sometimes we'll have teens set reminders on their cell phone since everyone seems to be very connected these days, that's a good little reminder to say, oh, yeah, it's time for me to take my vitamins.

Sometimes in our clinic we are able to consult with behavioral psychology to help teens figure out ways to improve their adherence and also to help them realize how important these issues are.

MR. BUSKER: I'd like you to address a specific point there might be some confusion about, and that's when should vitamin D levels be drawn? Does it matter?

MS. LEONARD: Yes it does. The time of year can affect the results of your vitamin D levels. The CF Foundation recommends checking levels at the end of winter when vitamin D stores are likely to be at their lowest. Since vitamin D can be made by the body in sunlight, in the summertime and the fall levels tend to be higher. We want to make sure that we're capturing the lowest point. However, it's not always feasible to get everybody's levels drawn at the end of winter , especially in a busy clinic setting, so you have to make sure that you include some clinical judgments along with the lab data when you're making recommendations.

MR. BUSKER: With this patient we've been discussing, when would you recheck her levels?

MS. LEONARD: I would check it at the next clinic visit, probably in the next two to three months. It's important to keep track and we don't want to lose sight of a low level and want to make sure that our treatment had the desired effect.

MR. BUSKER: What would you do if her levels were still low at her next clinic visit?

MS. LEONARD: If her levels continue to be low at the next clinic visit, I would recommend an additional 2000 IU of cholecalciferol or vitamin D3 every day, and I would want to make sure that she has been taking the extra cholecalciferol that I had recommended at the first low level and that she was still taking her CF specific vitamins.

MR. BUSKER: Please summarize assessing vitamin D levels.

MS. LEONARD: It's important to make sure that when you're assessing vitamin levels, especially in teenagers, you're checking for adherence and you're not just throwing more medications at someone. You want to make sure the teens can fit the treatment into their life and they're taking what you think they're taking so when you give them more medicine it is helpful. Keeping vitamin levels up can be very tricky.

MR. BUSKER: We'll return with Ms. Amanda Radmer Leonard in a just moment.

MS. MEGAN RAMSEY: Hello my name is Meghan Ramsay, Nurse Practioner and Adult Clinical Coordinator for the Johns Hopkins Cystic Fibrosis

Program at The Johns Hopkins University School of Medicine.

I am one of the program directors of eCysticFibrosis Review. These podcast programs will be provided on a regular basis to enable you to receive additional current concise peer reviewed information through podcasting, a medium that is gaining wide acceptance throughout the medical community. In fact today there are over 5,000 medical podcasts.

To receive credit for this educational activity and to review Hopkins' policies please go to our website at www.ecysticfibrosisreview.org. This podcast is part of eCysticFibrosis Review a bi-monthly email delivered program available by subscribing. Each issue reviews a current literature on focus topics important to clinicians caring for patients with Cystic Fibrosis.

Continuing education credit for each newsletter and each podcast is provided by the Johns Hopkins University School of Medicine for physicians and by the Institute for Johns Hopkins Nursing for nurses. Subscription to eCysticFibrosis Review is provided without charge and nearly a thousand of our colleagues have already become subscribers. The topic focus literature reviews help them keep up-to-date on issues critical to maintaining the quality of care for their patients.

For more information to register to receive eCysticFibrosis Review without charge and to access back issues please go to www.ecysticfibrosisreview.org.

MR. BUSKER: Welcome back to this eCysticFibrosis Review podcast. I'm Bob Busker, managing editor of the program. Our guest is Ms. Amanda Radmer Leonard, a pediatric nutrition practitioner at the Johns Hopkins Children's Center and the Johns Hopkins Cystic Fibrosis Center. Our topic is, *Strategies for the Improvement of Nutrition Outcomes*.

We've been looking at how some of the new information Ms. Leonard described in her newsletter issue can be applied in clinical practice. Please present with another case.

MS. LEONARD: Our last patient is a three year old female whose CF was diagnosed at birth. Medical history is remarkable for meconium ileus with resection. Anthropometrics include weight of 14 kg, which is the 42nd percentile for weight for age; a height of 97cm, which is the 62nd percentile of height per age; and a BMI at the 26th percentile.

Medications include CF specific vitamins, one chewable per day, 2000 IU of cholecalciferol, and an enzyme dose that's about 2400 IU of lipase/kg/meal.

The mother reports that the stool history is unremarkable, with one to two formed stools per day and rare bellyaches, and the diet history includes a high calorie diet with two to three snacks. This little girl also drinks two cans of a high calorie supplement per day.

The family has been struggling with trying to get her to gain weight, and they've tried appetite stimulants with limited success. The family discontinued this therapy.

When weight is discussed in clinic, the mother reports that their whole family is thin when they're young and she will catch up eventually, and also reports that she eats all the time.

MR. BUSKER: From what you just described, it sounds like the family doesn't realize the importance of nutrition issues. How would you approach them about that?

MS. LEONARD: That's an interesting point, Bob. It's really important for the family to know that nutrition plays an important role in cystic fibrosis. We've learned from the study by Yen and colleagues that was reviewed in the newsletter, that early nutrition has an impact on later clinical outcomes. So sharing that information with the family is important.

Sometimes I show the CF Foundation graph that shows the relationship or the association between body mass index and pulmonary function to help the family know that nutrition can help make your lungs better.

I think that also telling the family that the goal that the CF Foundation suggests is a BMI at or above the 50th percentile, and we have found that in patients who can achieve this, there is some association with having better lung function, as well.

Having the entire team involved, not just the dietitian, is also helpful. If the dietitian is saying one thing and

the family is not hearing it reflected by the other team members, by the physician or the nurse, or even in our clinic sometimes the physical therapist or respiratory therapist will help push the idea that good nutrition is important message so the family knows that we're all on the same page.

MR. BUSKER: How would you evaluate this three year old girl?

MS. LEONARD: I would want to look at a few things. I would look at her intake and mealtime behaviors, make sure her diet is high fat and high calorie as the mother is reporting. I would also want to make sure that she's drinking the high calorie supplement every day, because that will provide additional calories. I also want to make sure that her enzyme dose is okay.

So assessing the intake is checking the calories that are in, and checking the enzyme dose can help us with the potential losses if she's malabsorbing. From the stool history and her enzyme dose at 2400 units of lipase/kg/meal, it seems to be in a pretty good range.

Another thing I would think about would be appetite stimulants, but this family has already tried them and did not feel they were helpful. So I don't think I would suggest them again.

MR. BUSKER: What other things would you consider here?

MS. LEONARD: If the girl really is drinking her high calorie supplement and is eating three meals and two or three snacks a day and is maximizing what she is able to take in, I might consider a referral to a gastroenterologist. Because if she's absorbing and is taking in high calories but still can't eat enough to keep up with her energy needs, something like a gastrostomy might help her to grow.

We use gastrostomy feeds in kids with CF as an additive, not as a replacement for food. We want them to eat a high calorie diet during the day, but they just need that extra boost at night, and it does help improve their growth.

There weren't any other symptoms that pointed to malabsorption or other GI issues, but a referral to a gastroenterologist will allow for those issues to be ruled out before we proceed with the placement of a gastrostomy tube.

MR. BUSKER: If the GI findings support it, and the family chooses to go with a gastrostomy, how and when would this patient be fed?

MS. LEONARD: We usually give about 50% of the estimated calorie needs overnight through the tube, and we adjust it based on the patient's growth and the family's schedule and preferences. The idea is that she would eat as much as she wanted during the day and then at night we would give her more. A lot of families say that this decreases some stress in encouraging the children to eat — or maybe the children would call it nagging — saying "come on, eat some more, eat some more," when the parents know that she will be getting a substantial amount of calories overnight.

One of the nice things about overnight feeds is that they can be adjusted to the family's schedule. If she's going to bed later or they have an activity, they can start the feeds later or earlier, depending on what they need. It's really important for the health care provider or the dietitian to work with the family to make sure they're fitting this therapy into their schedule.

MR. BUSKER: In this patient — or in any patient with a gastrostomy — what's the best way to ensure they get their enzymes?

MS. LEONARD: That's not an easy question to answer. Ideally, we would give enzymes all throughout the feeding every few hours because they're only active for a few hours, but that would not be helpful in allowing the patient or her parents to sleep.

So at our center we give a meal dose of enzymes at the beginning and the end of feeds. Other centers use other approaches , but this is what we've chosen to do at our center, and we've had good success.

I think that when you're working with a family and talking about getting a gastrostomy, it's important to make sure to involve the family in the process so they believe this will help their child. It's also important to tailor the feeds to meet their schedule and their needs.

MR. BUSKER: Thank you for those case presentations and discussions. Let me change subjects now and ask you to look into the future for us. What's on the horizon for advances in understanding and improving cystic fibrosis nutrition?

MS. LEONARD: I think the field of CF nutrition is emerging lately and I think there's a lot of exciting things on the horizon. I would see as we move forward that CF nutrition will be more tailored to promoting health and not just to preventing deficiencies. I think once we get a better handle on managing some of the vitamin and mineral deficiencies, we'll find that maybe giving different doses or different kinds of foods will improve patients' health.

I think also that we're going to be moving toward a tailored nutrition approach in that we might be able to get modifier genes and know from the genetics what sorts of foods or what sorts of therapies will be helpful for these patients.

MR. BUSKER: Thank you. Let's wrap things up by reviewing what we've talked about today in light of our learning objectives. What are the approaches to the treatment of low vitamin D levels in patients with cystic fibrosis?

MS. LEONARD: I think recognizing the importance of vitamin D that should be done in clinical practice, and continued monitoring will help keep patients healthy. And if you notice a low vitamin D level, knowing what medicines your patients are already taking, and if they're not taking the vitamins that you think they are, you want to make sure to address that rather than just throwing more medications on top of something that maybe they already find to be overwhelming.

MR. BUSKER: What about the variability of fecal elastase results during the first year of life?

MS. LEONARD: I think it's important to know that just getting one fecal elastase value during the first year of life doesn't provide good picture of that patient's pancreatic status going forward. So getting an initial reading and then another reading at a year, especially if the initial reading was greater than 50 μg/g, is important.

MR. BUSKER: And finally, the importance of tailoring nutritional interventions to the patient and family situation.

MS. LEONARD: Practicing patient- and family-centered care when you're looking at the nutritional approach for a family is very important, because you want to make sure you're not recommending or prescribing things that won't work for their particular situation. So I think taking the whole picture into account and making sure that they have a voice in the planning is very important.

MR. BUSKER: Ms. Amanda Radmer Leonard from the Johns Hopkins Children's Center, thank you for participating in this eCystic Fibrosis Review podcast.

MS. LEONARD: Thanks, Bob, I really enjoyed it.

MR. BUSKER: This podcast is presented in conjunction with eCysticFibrosis Review, a peer-reviewed CME and CNE-accredited literature review emailed monthly to clinicians treating patients with cystic fibrosis. This activity has been planned and implemented in accordance with the Essential Areas and policies of the Accreditation Council for Continuing Medical Education through the joint sponsorship of The Johns Hopkins University School of Medicine and The Institute for Johns Hopkins Nursing.

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Thank you for listening.

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