



VOLUME 6 - ISSUE 8

[LISTEN TO PODCAST](#)[DOWNLOAD PODCAST](#)[PHYSICIAN POST-TEST](#)[NURSE POST-TEST](#)

Nutritional Issues in Cystic Fibrosis

Our guest author is Deepak Agrawal, MD from the the University of Texas Southwestern Medical Center in Dallas.

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

- Evaluate the impact of disease modifying therapies on the nutritional status of patients with cystic fibrosis.
- Describe the impact of behavioral interventions on the nutritional status of children with cystic fibrosis.
- Discuss the effects of parental depression on PERT adherence in their children with cystic fibrosis.

Guest Faculty Disclosure

Dr. Deepak Agrawal reports that he has no relevant relationships with any commercial entities.

Unlabeled/Unapproved Uses

Dr. Deepak Agrawal reports there will be no off-label or unapproved uses of any drugs.

MEET THE AUTHOR



Deepak Agrawal, MD
Adult Gastroenterologist
University Texas Southwestern
Medical Center
Dallas, Texas

Release Date:
July 14, 2016

Expiration Date:
June 13, 2018

[LISTEN TO PODCAST NOW](#)[DOWNLOAD PODCAST](#)[SUBSCRIBE NOW](#)[PHYSICIAN POST-TEST](#)[NURSE POST-TEST](#)

OTHER RESOURCES

[Download the podcast transcript](#)[Go to the companion newsletter](#)[NEWSLETTER ARCHIVE](#)[SHARE WITH A COLLEAGUE](#)

PROGRAM DIRECTORS

Peter J. Mogayzel, Jr., MD, PhD

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 eCysticFibrosis Review Podcast.

Today's program is a follow-up to our newsletter topic "Nutritional Issues in Cystic Fibrosis." Our guest today is one of that issue's authors, Dr. Deepak Agrawal, Adult Gastroenterologist at the University of Texas Southwestern Medical Center in Dallas.

eCysticFibrosis 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 Chiesi USA Inc, Vertex Pharmaceuticals Incorporated, and Gilead Sciences.

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

- Evaluate the impact of disease modifying therapies on the nutritional status of patients with cystic fibrosis.
- Describe the impact of behavioral interventions on the nutritional status of children with cystic fibrosis.
- Discuss the effects of parental depression on PERT adherence in their children with cystic fibrosis.

Dr. Agrawal reports that he has no relevant relationships with any commercial entities, and his presentation today will not discuss any off-label or unapproved uses of any drugs or products.

Dr. Agrawal, welcome to this eCysticFibrosis Review Podcast.

DR. DEEPAK AGRAWAL: Thank you for inviting me.

MR. BOB BUSKER: In the newsletter issue, doctor, you reviewed some of the recent literature describing appropriate PERT dosing, factors that can affect PERT adherence, and the impact of ivacaftor on nutritional outcomes in patients with cystic fibrosis. Our focus today is to show how some of this new information can impact clinical practice. So let me ask you to start us out with a patient presentation, if you would please doctor.

DR. AGRAWAL: This is an adult with cystic fibrosis and G551D mutation who recently started taking ivacaftor. His pulmonary function had improved, and he was taking pancreatic enzymes for his known pancreatic insufficiency at a dose of 1000 units/kg/meal. He was taking a regular diet and was happy, but he noticed that he had gained some weight on this medication. Although he was quite happy about the weight gain, he had a few questions.

MR. BUSKER: Your patient's questions, doctor. Let me ask you to hold those for a moment, because I'd like to ask you a question of my own first: why do you think your patient is gaining weight?

DR. AGRAWAL: Interesting question. Initial studies on ivacaftor in cystic fibrosis looked at improvement in lung function only. But when they looked at the weight data, it showed that the patients had also gained weight. It's not completely clear why people gain weight, but the possible reasons include decreased energy expenditure, meaning patients breathe better and expend less energy and are in a less catabolic state. It's also shown that ivacaftor raises the intestinal pH, and we know that high pH is necessary for digesting food properly. Ivacaftor can do this by regulating gastric acidity or possibly by increasing bicarbonate secretion from the pancreas. Finally, ivacaftor decreases constipation, so overall the patients feel better from a GI standpoint.

MR. BUSKER: Thank you, doctor. So — your patient's questions: what did he ask you about his weight gain on ivacaftor?

DR. AGRAWAL: He asked whether he will become obese if he continues the medication.

MR. BUSKER: That may be kind of a unique question, considering that low weight issues are one of the hallmarks of cystic fibrosis.

DR. AGRAWAL: You are right; for once it's a good problem to have. I feel people with cystic fibrosis are quite cautious about their weight and their diets. In the Borowitz article reviewed in the newsletter, children continued gaining weight, and it was appropriate weight gain, but adults stopped gaining after some time. I would tell him he does not have to fear becoming

obese, but I will still review his diet to make sure it's appropriate and is not obesogenic.

MR. BUSKER: What other questions did he ask?

DR. AGRAWAL: He also asked me about the number of medications he was taking. He was taking a lot of enzymes and a proton pump inhibitor and asked if he could reduce the number of pills he takes.

MR. BUSKER: Just about every patient with CF would love to reduce their pill burden. And a lot of their pills are pancreatic enzymes. So the question is, is it possible that this patient could safely reduce his PERT dose by switching to a different formulation? But before you answer that, doctor, let me ask you to give us a quick summary of the different PERT formulations.

DR. AGRAWAL: There are two main types of pancreatic enzymes, coated and uncoated. Coated preparations protect the enzymes from the gastric acid and they're realized in the duodenum. Uncoated enzymes are affected by gastric acid and are best taken along with a proton pump inhibitor. Most of the enzymes available on the market are coated enzymes; only one uncoated enzyme is available.

The other difference in the formulation is that some capsules have microspheres and others have micro tablets. I should also add that one enzyme has added bicarbonate, the rationale being that it can decrease the pH in the duodenum and thus aid in digestion and absorption of food.

MR. BUSKER: How do you determine which is the best PERT formulation for a particular patient, and how you do determine what the dose should be?

DR. AGRAWAL: There is not much difference in performance of these different formulations. An individual patient may respond differently, so if somebody is not having the desired results despite maximum doses of enzyme replacement therapy, I may try a different formulation.

The optimal dose of pancreatic enzymes remains to be defined. Most patients are started on 500 or 1000 units/kg/meal, and then the dose is adjusted according to their response. The maximum dose of pancreatic enzymes can be 2000 to 2500 units/kg/meal.

MR. BUSKER: Thank you for that clarification. Let's go back to your patient's question. Could his success on ivacaftor therapy result in reducing his enzyme pill burden?

DR. AGRAWAL: It may be possible to reduce the dose slightly, so he can decrease the number of pills. By the way, he probably cannot stop pancreatic enzymes completely. Ivacaftor can stimulate the pancreas to secrete more pancreatic enzyme, but it is not a substitute for pancreatic enzymes.

MR. BUSKER: His proton pump inhibitor — is that a medication he might be able to stop taking?

DR. AGRAWAL: It depends on why he is taking the proton pump inhibitor. Is it because of a peptic condition or because he has heartburn, or was it an attempt to improve absorption through neutralization of acid? If he is taking coated enzymes, he may not need a proton pump inhibitor. So if he doesn't have any symptoms I will try discontinuing the proton pump inhibitors.

MR. BUSKER: Thank you, doctor. And we'll return, with Dr. Deepak Agrawal, in just a moment.

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

eCysticFibrosis 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.

Subscription to eCysticFibrosis Review is provided without charge or prerequisite.

Continuing education credit for each issue and each podcast is provided by the Johns Hopkins University School of Medicine and the Institute for Johns Hopkins Nursing. For more information on this educational activity, to subscribe to and receive eCysticFibrosis Review without charge, and to access back issues, please go to our website: www.ecysticfibrosisreview.org.

I'd also like to tell you about the "CF Family Day Meeting Builder." This is a one-stop shop to help you create patient and caregiver education and Family Day meetings. To find out more, please visit www.CFFamilyDay.org.

One more thing I'd like to tell you about is new the GetSMART app. GetSMART (Safe Means of Administering the Right Therapy Extended Release and Long Acting Opioids) is available for CME/CE/MOC credit at no cost.

Visit dkbmed.com/smart to download the GetSMART app for Apple iOS, Android, or desktop today.

That's dkbmed.com/smart.

MR. BUSKER: Welcome back to this eCysticFibrosis Review podcast on "Nutritional Issues in Cystic Fibrosis." I'm Bob Busker, managing editor of the program. We've been talking with Dr. Deepak Agrawal, from the University of Texas Southwestern Medical Center in Dallas, about how the information in his recent newsletter issue can impact clinical practice. So let's continue with another patient scenario — if you would please, doctor.

DR. AGRAWAL: The parents of a four year-old child with pancreatic insufficiency come to me with concerns about the child's weight. The child had been growing normally up until about nine months back and was on the 50th percentile for body mass index, when her weight began to decline; she's now at the 25th body mass index percentile. Her eating habits have become worse, and she just doesn't want to eat much.

MR. BUSKER: So this 4 year old has developed an eating problem that's confirmed by the decline in her weight and BMI, and her parents are asking your advice on what to do about it. What kind of information do you need from them before you can make a recommendation?

DR. AGRAWAL: History is a big part of the evaluation. I would first want to make sure that there is no medical cause, no pulmonary issues, the lungs are doing fine, there's no infection, constipation, and similar medical issues. The second thing I would want to know is, is she taking an adequate dose of pancreatic enzymes and is she taking the enzymes at the right time; that is, with or during meals.

If all the things look good, I would like to know more about the child's mealtime behavior. What does she like to eat, how long does she spend at the table, what is the parents' reaction to her eating habits.

MR. BUSKER: Can you give us some examples of the kind of responses you might get to these questions? And what do those responses tell you?

DR. AGRAWAL: This is very interesting. Quite often parents can tell me about the kids but less about their own reaction. They often can't recall what prompts they give their kids. Probably the most common way to persuade kids to eat appears to be telling them to eat, or eat quickly; many patients often take a long time to finish meals.

MR. BUSKER: Once you get that kind of information, what would be the next steps?

DR. AGRAWAL: The first step would be to create awareness and let them observe the child's behavior and their own reaction. Second is education about the existing literature — things like what prompts have been shown to be helpful. For example, rather than just telling the child to eat, the children can be told to heap up the food on the plate and fill up the spoon. That's a specific clue that can be given to the child.

Literature also shows that children eat most within the first few minutes, and longer mealtimes do not necessarily translate into increased caloric intake.

The third thing would be actual treatment and counseling.

MR. BUSKER: When you tell the parents that you're recommending behavioral counseling to address their child's eating problems, what kind of reactions do you get from them?

DR. AGRAWAL: A few puzzled looks. Very rarely I've had parents get a little upset about it because they feel we are telling them they are not taking care of the children, but the majority of the time they want to know more about it. Most of them are unaware that there is such a thing as behavioral counseling, and once we tell them this is going to help them feed the child better and will decrease their stress level, they want to know more about it.

MR. BUSKER: What specifics do you tell them about treatment and counseling — about how it works, what it entails, potential success — what kind of things do they need to know?

DR. AGRAWAL: Behavior intervention involves teaching parents about caloric goals, introducing a specific mealtime, which is to be only 20 minutes, when to praise the child, when to ignore their bad behavior, how to reward the child, dealing with tantrums, and then finally reinforcing all the skills they have learned. Every child's situation is slightly different.

An important part of behavioral intervention is individualizing the counseling. The protocol used in the recent randomized control trial was weekly sessions for two months and then monthly sessions for next four months. The sessions were given by postdoctoral psychology fellows. Importantly, the sessions can be given in person or by telephone, which can greatly

increase the reach of this program because not everybody can make it to the clinic every time for an in-person visit.

After the behavior intervention, the positive changes last for a long time, and in the study they lasted during the one-year follow-up. This was reviewed in the newsletter in the Powers article.

MR. BUSKER: Thank you. We've got time to look at one more patient.

DR. AGRAWAL: Imagine a five year old boy with cystic fibrosis whose growth curve has plateaued despite multiple efforts. He seems to be on optimal dosages of pancreatic enzymes, and his pulmonary symptoms are under control. The mother says she is doing everything she can. She has two older children, and in between school and other activities she spends as much time as possible with her son with cystic fibrosis. She feels tired and doesn't sleep well at night. She has also lost some weight.

MR. BUSKER: So at least on paper, this child should be doing well. Optimal PERT dosage, pulmonary symptoms under control. But he's not growing, and the mother's upset. How do you respond to this kind of situation?

DR. AGRAWAL: In this case the child is not growing as well as expected. The first step would be to make sure he is taking pancreatic enzymes appropriately: with or during meals.

Apart from the child, I'm also concerned about the mother's well-being. I get a sense that the mother is overwhelmed and may have depression. In a study involving 154 cystic fibrosis centers in nine countries, one-third of parents taking care of CF children reported depressive symptoms, and almost half had anxiety. Studies have shown that depression in parents is related to treatment adherence in children. So I would be a little worried whether depression in the parent in this case has something to do with the child's not gaining weight.

MR. BUSKER: Depression and anxiety in parents caring for children with CF — those percentages you just mentioned are pretty high. Do you think clinicians are aware of the prevalence of these mental health issues?

DR. AGRAWAL: This is a very important question, I feel. Only recently we have recognized that depression and anxiety are a significant problem in parents who are taking care of children with cystic fibrosis. In fact, now the international guideline on mental health and cystic fibrosis recommends annual screening of parents and caregivers for symptoms of depression and anxiety. Many trials are also ongoing to address mood disorders. But the key part here is that the physician should be able to recognize mood disorders, and parents should recognize that it is a common problem. It is okay to feel overwhelmed and seek care when they have these kinds of symptoms. So we should be supportive and sympathetic and treat when needed.

MR. BUSKER: In the situation you described for us, the mother has symptoms of anxiety and depression. The child is not gaining weight as expected. Connect the dots for us, please.

DR. AGRAWAL: This is a very interesting observation, which was mentioned in the newsletter by Barker and Quittner.¹ They hypothesized that children of parents who have depression were less likely to comply with pancreatic enzyme replacement therapy, and they showed that in the study. Interestingly, these children were compliant with the pancreatic enzyme treatment at school, which clearly showed that the problem was at home.

It's not that the parents are deliberately not giving these enzymes, it's just that they are overwhelmed, they are burnt out, and pancreatic enzyme treatment does not seem to be an important part of CF treatment, at least not in an immediately obvious way. In fact, studies have reported that parents with depression actually took better care of airway clearance in children with cystic fibrosis, so they clearly are trying hard, but they tend to miss out on pancreatic enzyme replacement treatment.

MR. BUSKER: There is a logic in that, I think, because the pulmonary symptoms present a much more obvious need, whereas the digestive symptoms tend to be less immediately obvious.

DR. AGRAWAL: Oh, I agree.

MR. BUSKER: Dr. Agrawal, I want to thank you for today's cases and discussion. Before we wrap things up, I'd like to get your opinion on something: what kinds of improvements in our understanding of the use of pancreatic enzymes you'd find most beneficial.

DR. AGRAWAL: I think we need better and more consistent ways of assessing efficacy of different pancreatic enzyme formulations. We need head-to-head comparisons of different formulations, including coated and uncoated enzymes. We need to understand how to dose these pancreatic enzymes, especially when patients are taking medications like ivacaftor, which have the potential to increase bicarbonate secretion and even secretion of pancreatic enzymes.

The importance of behavioral mealtimes should become a standard teaching for all parents; we all can benefit a lot by

improving the interaction with our children during mealtimes.

Finally, recognizing depression and anxiety as a common feature amongst parents taking care of cystic fibrosis patients will help us recognize and treat these disorders. All of these can result in better treatment adherence in children with cystic fibrosis.

MR. BUSKER: Thank you for sharing your insights, doctor. Let's wrap up our discussion by reviewing what we've talked about today in light of our learning objectives. So to begin: the impact of disease modifying therapies on the nutritional status.

DR. AGRAWAL: Children who receive ivacaftor can continue to gain weight for 48 weeks after treatment. Adults can also gain weight but do not become obese. Patients who are taking ivacaftor and taking pancreatic enzyme replacement treatment may be able to reduce the amount of pancreatic enzymes they are taking, but ivacaftor is not a replacement for pancreatic enzymes.

MR. BUSKER: And our second learning objective: the impact of behavioral interventions on the nutritional status of children with cystic fibrosis.

DR. AGRAWAL: Behavioral intervention, which is usually given by psychologists and lasts about six months, is a very useful intervention in increasing nutrition status. This intervention can be given in person or by phone, and the effects last much longer than the six month intervention period.

MR. BUSKER: And finally: the effects of parental depression on their children's adherence to pancreatic enzymes.

DR. AGRAWAL: Taking care of children with cystic fibrosis can be overwhelming, and depression among parents is common. So we should screen for mood disorders, not just depression but also for anxiety, and treat as needed. This can improve the nutrition outcomes in children with cystic fibrosis by improving adherence to treatment such as pancreatic enzymes.

MR. BUSKER: Dr. Deepak Agrawal, from the University of Texas Southwestern Medical Center, thank you for participating in this eCysticFibrosis Review Podcast.

DR. AGRAWAL: It was a pleasure, and I thank you for the opportunity.

MR. BUSKER: To receive CME credit for this activity, please take the post-test at www.ecysticfibrosisreview.org/test.

This podcast is presented in conjunction with eCysticFibrosis Review newsletter, a peer-reviewed CME/CE credit publication that is emailed monthly to clinicians treating patients with cystic fibrosis.

This activity has been developed for the CF care team, including 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.

There are no fees or prerequisites for this activity.

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.

The Johns Hopkins University School of Medicine is accredited by the ACCME to provide continuing medical education for physicians. The Johns Hopkins University School of Medicine designates this enduring material for a maximum of *0.5 AMA PRA Category 1 credit*.™ Physicians should claim only the credit commensurate with the extent of their participation in this activity.

The Institute for Johns Hopkins Nursing is accredited as a provider of continuing nursing education by the American Nurses Credentialing Center's Commission on Accreditation. For nurses, this 0.5 contact hour Educational Activity is provided by the Institute for Johns Hopkins Nursing. Each podcast carries a maximum of 0.5 contact hour.

This educational resource is provided without charge, but registration is required. To register to receive eCysticFibrosis Review by email, please go to our website: www.ecysticfibrosisreview.org.

The opinions and recommendations expressed by faculty and other experts whose input is included in this program are their own. This enduring material is produced for educational purposes only.

Use of the names of the Johns Hopkins University School of Medicine and the Institute for Johns Hopkins Nursing implies review of educational format, design, and approach.

Please review the complete prescribing information for specific drugs, combinations of drugs, or use of medical equipment, including indication, contraindications, warnings, and adverse effects, before administering therapy to patients.

eCysticFibrosis Review is supported by educational grants from, Gilead Sciences Inc., Vertex Pharmaceuticals, Incorporated, and Chiesi USA, Inc. This program is copyright with all rights reserved by the Johns Hopkins University School of Medicine and the Institute for Johns Hopkins Nursing.

References

1. Barker DH, Quittner AL. [Parental depression and pancreatic enzymes adherence in children with cystic fibrosis.](#) *Pediatrics*. 2016 Feb;137(2):1-9.

CME/CE INFORMATION

ACCREDITATION STATEMENT

Physicians

This activity has been planned and implemented in accordance with the accreditation requirements and policies of the Accreditation Council for Continuing Medical Education (ACCME) through the joint providership of the Johns Hopkins University School of Medicine and The Institute for Johns Hopkins Nursing. The Johns Hopkins University School of Medicine is accredited by the ACCME to provide continuing medical education for physicians.

Nurses

The Institute for Johns Hopkins Nursing is accredited as a provider of continuing nursing education by the American Nurses Credentialing Center's Commission on Accreditation.

The Institute for Johns Hopkins Nursing and the American Nurses Credentialing Center do not endorse the use of any commercial products discussed or displayed in conjunction with this educational activity.

CREDIT DESIGNATION STATEMENT

Physicians

Podcast: The Johns Hopkins University School of Medicine designates this enduring material for a maximum of 0.5 *AMA PRA Category 1 Credit(s)*[™]. Physicians should claim only the credit commensurate with the extent of their participation in the activity.

Nurses

Podcast: This 0.5 contact hour Educational Activity is provided by the Institute for Johns Hopkins Nursing. Each Podcast carries a maximum of 0.5 contact hour, or a total of 3 contact hours for the 6 podcasts in this program.

Respiratory Therapists For United States: [Visit this page](#) to confirm that your state will accept the CE Credits gained through this program.

POLICY ON SPEAKER AND PROVIDER DISCLOSURE

It is the policy of the Johns Hopkins University School of Medicine and the Institute for Johns Hopkins Nursing that the speaker and provider globally disclose conflicts of interest. The Johns Hopkins University School of Medicine OCME has established policies in place that will identify and resolve all conflicts of interest prior to this educational activity. Detailed disclosure will be made in the instructional materials.

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

[INTERNET CME/CE POLICY](#)

[INTENDED AUDIENCE](#)

[DISCLAIMER STATEMENT](#)

[CONFIDENTIALITY DISCLAIMER FOR CME ACTIVITY PARTICIPANTS](#)

[STATEMENT OF RESPONSIBILITY](#)

[HARDWARE & SOFTWARE REQUIREMENTS](#)

[STATEMENT OF NEED](#)

[COMPLETE CME INFORMATION](#)