



VOLUME 5 – ISSUE 8: TRANSCRIPT

# Featured Cases: Optimizing Nutrition in Individuals with Cystic Fibrosis

Our guest author is John Pohl, MD, from the University of Utah, in Salt Lake City, Utah.

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

- Explain how appropriate dosing in pancreatic enzyme replacement therapy is associated with an improved body mass index as well as improved protein and fat absorption in individuals with cystic fibrosis.
- Summarize how health-related quality of life is improved in individuals with CF with a good nutritional status, and why a low BMI is a risk factor for mortality in adolescents with CF.
- Assess the current evidence describing the use of appetite stimulants to improve weight gain in individuals with CF.

This discussion, offered as a downloadable audio file and companion transcript, covers the important issues related to improving nutrition in the format of case-study scenarios for the clinical practice. This program is a follow up to Volume 5, Issue 7 *eCysticFibrosis Review Newsletter* – [Optimizing Nutrition in Individuals with Cystic Fibrosis](#).

### MEET THE AUTHOR



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### Guest Faculty Disclosure

Dr. John Pohl reports he has no relevant relationships with a commercial entity.

### Unlabeled/Unapproved Uses

Dr. John Pohl will not discuss any off-label or unapproved uses of any drugs or products.

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**LAUNCH DATE**

This program launched on November 12, 2014, and is published monthly; activities expire two years from the date of publication.

**INTENDED AUDIENCE**

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

**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.

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**MR. BOB BUSKER:** Welcome to this eCysticFibrosis Review podcast.

Today's program is a follow-up to our newsletter topic, "Optimizing Nutrition in Individuals with Cystic Fibrosis." Our guest today is that issue's author Dr. John Pohl, Professor of Pediatric Gastroenterology at the University of Utah in Salt Lake City.

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 AbbVie, 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:

- Explain how appropriate dosing in pancreatic enzyme replacement therapy is associated with an improved body mass index as well as improved protein and fat absorption in with cystic fibrosis.
- Summarize how health-related quality of life is improved in individuals with CF who have a good nutritional status, and why a low BMI is a risk factor for mortality in adolescents with CF.
- Assess the current evidence describing the use of appetite stimulants to improve weight gain in people with CF.

Dr. Pohl has disclosed that he has no relevant relationships with a commercial entity and that he will not be discussing off-label or unapproved uses of any drugs or products in today's presentation.

**MR. BUSKER:** Dr. Pohl, welcome to this eCysticFibrosis Review Podcast.

**DR. POHL:** Thank you for allowing me to speak today.

**MR. BUSKER:** In your newsletter issue, you described the evidence basis showing that malnutrition is associated with a worse clinical prognosis in patients with cystic fibrosis, including a more rapid decline in pulmonary function. You reviewed new research into pancreatic enzyme replacement therapy (PERT) dosing; the connection between nutritional status and health-related quality of life; prognostic markers of mortality; and the

potential role of appetite stimulants in cystic fibrosis care.

Our objective today is to better understand how some of that new information can be applied in the clinic. So please start us out by describing a patient situation.

**DR. POHL:** A question that is often asked in the cystic fibrosis clinic is how to maximize nutritional outcome. Let's say, for example, the parents of a 3-year-old girl with cystic fibrosis come to your clinic. The child has cystic fibrosis and she is on pancreatic enzyme replacement therapy, or PERT. The family has questions about how to maximize her nutritional outcome. How do we do about doing this?

**MR. BUSKER:** The child is already on pancreatic enzyme replacement therapy, so achieving optimal PERT dosing would seem to be the most obvious solution. What does the guidance say about PERT dosing?

**DR. POHL:** We know there are guidelines for high dosing of pancreatic enzyme replacement therapy to prevent complications like fibrosing colonopathy, but there is definite intercenter variability for PERT in the United States. This is where the [patient registry](#) for the Cystic Fibrosis Foundation is helpful.

The study by Haupt in the 2014 issue of the *Journal of Pediatrics* showed that PERT dosing was significantly higher in top-quartile centers compared to the lowest-quartile centers. One hundred seventy-nine pediatric cystic fibrosis programs were included in this study, divided into quartiles, and the study group included a total of 14,482 patients with cystic fibrosis.

Haupt, et al, looked at multiple factors besides PERT dosing. They looked at patient demographics, pulmonary function testing, and respiratory culture results, and then followed long-term growth and nutrition data. They took mean body mass index (BMI) for each center and divided the results into four quartiles for comparison.

Centers with higher pancreatic enzyme replacement therapy dosing had significantly higher enzyme dosing at 1,755 lipase units per kilogram per meal compared to lower quartile centers, which had a mean enzyme dosing of 1,628 lipase units per kilogram per meal.

It's interesting that multivariate analysis occurred and this difference in pancreatic enzyme replacement therapy — which was significant — remained significant even after adjustment for various covariates.

**MR. BUSKER:** Do high-performing centers, the ones in the top quartile, they have other aspects of care that may have helped increase body mass index?

**DR. POHL:** That's an interesting question. When the authors looked at top-quartile programs, they noted that the patients were younger, they had fewer diagnoses of failure to thrive, and they had fewer diagnoses of malnutrition. Interestingly, the patients in the top-quartile centers had fewer meconium ileus diagnoses, and they also had less history of steatorrhea. The top programs were also noted to have cystic fibrosis diagnosed by newborn screening and had better parameters for lung function testing. For example, in the top-quartile programs the patients had improved forced expiratory volume in one second predicted, also known as FEV1 percent predicted, compared to the lower-quartile programs.

Finally, nutritional supplementation by nasogastric or gastrostomy tube, as well as acid-blockade medication, were more frequent in the top-quartile centers compared to the lower quartile centers.

**MR. BUSKER:** You mentioned the potential risks of too high a dose of PERT. Please explain that.

**DR. POHL:** PERT dosing greater than 10,000 units of lipase per kilogram per day, also dosing greater than 2,500 lipase units per kilogram per meal, appears to be associated with increased risk of various side effects such as fibrosing colonopathy, and it is not recommended that PERT dosing go above that level. We still do not know if there's an optimum PERT dose for absorption with cystic fibrosis, but lower dosing likely affects body mass index in the pediatric age range. And when you look between these centers, low PERT dosing may simply be a proxy marker for a center providing perhaps less aggressive nutrition care in cystic fibrosis.

In summary, care should be spent in your clinic calculating PERT dosing for patients with cystic fibrosis. You want to balance between optimizing nutrition with PERT while reducing the risk of fibrosing colonopathy.

**MR. BUSKER:** The patient's parents asked you to maximize her nutritional outcome. What steps would you take to do that?

**DR. POHL:** For the three year old patient we described in this case I would first calculate PERT dosing, making sure to optimize PERT dosing for adequate nutrition, but at the same time not going over the recommendations for fibrosing colonopathy risk. I also would look at other aspects; for example, does the patient need further nutritional care, such as nasogastric or gastrostomy, or would she benefit from acid suppression.

**MR. BUSKER:** Thank you, doctor. We'll return with Dr. John Pohl from the University of Utah in just a moment.

**MS. MEGAN RAMSEY:** Hello, my name is Meghan Ramsay, nurse practitioner 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](http://www.ecysticfibrosisreview.org).

This podcast is part of eCysticFibrosis Review, a bimonthly, 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.

**MR. BUSKER:** Welcome back to this eCysticFibrosis Review podcast. I'm Bob Busker, managing editor of the program. Our topic is *Optimizing Nutrition in Adolescents with Cystic Fibrosis*. Our guest is Dr. John Pohl, Professor of Pediatric Gastroenterology at the University of Utah in Salt Lake City.



We've been talking about how some of the new information Dr. Pohl discussed in his newsletter issue can be applied in the clinic. Let's continue by looking at another patient situation.

**DR. POHL:** Our next case is a 15 year old girl with cystic fibrosis who is brought to clinic by her parents. The parents note that she is noncompliant with her pancreatic enzyme replacement therapy medication and her body mass index has decreased from the 30th to the 5th percentile using standard CDC growth charts.

**MR. BUSKER:** Her BMI has decreased rather significantly, and she's been noncompliant with her PERT. What are your thoughts on that?

**DR. POHL:** The practitioner always needs to be careful to make sure that compliance is being followed in complex situations like cystic fibrosis. Obviously, when you are dealing with a teenager you'll potentially have some compliance issues as the patient develops independence. Questions of compliance should be asked to the patient when they get older and to the parents, especially if you are seeing potential risk of malnutrition, such as a decrease in body mass index.

**MR. BUSKER:** It's been known for a long time that PERT can improve fat absorption. But in your newsletter issue you described the effect of PERT on protein absorption as well. Tell us more about that.

**DR. POHL:** When we think about pancreatic enzyme replacement therapy, we need to remember that PERT works by helping with protein as well as fat absorption. It can be somewhat difficult to measure protein absorption; however, Engelen, et al recently came up with a unique method: they gave patients with cystic fibrosis a meal labeled with both nitrogen spirulina protein, which is a protein from blue-green algae, as well as hydrogen-labeled phenylalanine. The spirulina protein is further broken down, while the hydrogen-labeled phenylalanine does not break down at all, and then the ratio is measured. This testing is interesting because it is not affected by phenylalanine metabolism.

Using this method the authors found some interesting results. They compared patients with cystic fibrosis, both adults and children, to a control group. Patients with cystic fibrosis had significantly lower protease function — in other words, protein absorption —

measured at 46.5% compared to control patients. During the study, PERT was given to these patients and protein absorption increased up to 90.3% of control absorption. So it did not get all the way up to control absorption but got very close, with maximum digestion at approximately 80 minutes.

Furthermore, no significant difference was seen in protein digestion between the adult and pediatric patients with cystic fibrosis, and there was no significant difference in protein digestion between patients with cystic fibrosis, regardless of delta-F508 mutation status, regardless of nutritional failure, and regardless of lung function.

**MR. BUSKER:** I want to go back to this adolescent girl and her significant decrease in BMI from the 30th to the 5th percentile. What might her weight loss suggest about an increased risk of mortality?

**DR. POHL:** This certainly could suggest an increased risk of mortality. A recent article by Hulzebos et al looked at adolescent patients with cystic fibrosis. They used a cycle ergometer for cardiopulmonary exercise testing, and using a multivariate analysis of these adolescents with cystic fibrosis, they found that a model of FEV1 percent predicted, peaked ventilatory equivalent ratio, and body mass index was predictive of mortality. Furthermore, when they looked at the study group longitudinally by Kaplan-Meier analysis, they found that patients with two or three of these risk factors had a significantly increased risk of mortality. On the other hand, patients with three risk factors had a significantly greater risk of mortality compared to those patients with only one or two of those risk factors: FEV1 percent predicted, peak ventilatory equivalent ratio, and body mass index.

**MR. BUSKER:** To wrap up this patient we've been discussing, what would you recommend for her?

**DR. POHL:** My recommendations for this patient as well as her family would be to look at her nutrition and make sure we're maintaining good use of pancreatic enzyme replacement therapy, knowing that PERT helps both fat and protein absorption. Finally, knowing there is a multivariate analysis that shows risk of mortality in this teenage population, I would make sure I address her pulmonary function as well as her body mass index, because low body mass index can be a risk for increased mortality in adolescents.

I'm especially worried about her noncompliance because there is a correlation between low body mass index and pulmonary function. I want to preserve her lung function long-term, and if she is not maintaining an adequate body mass index, she is at risk.

**MR. BUSKER:** Thank you for that case and discussion, Dr. Pohl. Please bring us one more patient.

**DR. POHL:** Our final case involves a 12 year old boy with cystic fibrosis who comes to clinic with his parents. The parents are concerned about his weight. They know his body mass index is at the 10th percentile. Over time they notice he's become more withdrawn, and he looks different from his friends. They ask if he would benefit from an appetite stimulant.

**MR. BUSKER:** So we're looking at a 12 year old boy, a pubescent preteen. He doesn't fit in, he looks different from his friends, and so he's become withdrawn — that all seems to come under the heading of health-related quality of life. It's easy to assume that it may be worse in patients with cystic fibrosis, but what data actually exist to support that assumption?

**DR. POHL:** A recent study by Shoff et al in Wisconsin, published in the *Journal of Cystic Fibrosis*. They looked at nutritional parameters for health-related quality of life testing obtained by the cystic fibrosis questionnaire, which was applied annually by interview or self-administration for three years during regular clinic visits.

The study had some interesting results. Both height Z scores and body mass index Z scores had a significantly positive association with physical functioning and body image. Physical functioning and body image dimensions improved as height Z scores improved, in other words, as patients became taller, or as their body mass index improved, or as they gained weight. A positive weight Z score association was also seen with physical functioning and body image.

**MR. BUSKER:** Were there differences in health-related quality of life scores between the males and the females who were tested?

**DR. POHL:** There was a difference between girls and boys. Girls showed a positive association between

height Z score and eating disturbances. Also there was a positive association between physical functioning and height Z score and body mass index Z score. Boys, on the other hand, had a positive association between body image and height Z score. The cystic fibrosis questionnaire score was then evaluated to determine a mostly low score. A mostly low score was defined less than 66. A mostly low score was seen between short stature and eating disturbances, as well as between a goal BMI and body image. Improving nutrition over time may prevent a decline in health-related quality of life in patients with cystic fibrosis, again emphasizing the importance of nutrition.

**MR. BUSKER:** What would you tell the parents, who are asking you about appetite stimulants? Have stimulants been shown to be helpful in cystic fibrosis care? What does the evidence say?

**DR. POHL:** The long-term efficacy of appetite stimulants in the care of cystic fibrosis is not known. A recent Cochrane Review by Chinuck et al looked at two appetite stimulants that are commonly used in pediatric and adult care: cyproheptadine hydrochloride and megestrol acetate. They were very restrictive in the studies they reviewed, making sure that they were placebo-controlled, and only three studies existed that studied either cyproheptadine hydrochloride or megestrol acetate. They found that appetite stimulants may be helpful in improving weight gain in the short term, but it is unknown if appetite stimulants are helpful long term.

Both of these appetite stimulants have side effects. Cyproheptadine hydrochloride can cause fatigue and feelings of being tired. Megestrol acetate can be associated with a decrease in morning cortisol levels. Keep in mind that adrenal insufficiency has been described in patients using megestrol acetate, but the side effect has been observed in case reports only.

**MR. BUSKER:** What would you tell this 12 year old boy's parents; what would your recommendations be?

**DR. POHL:** I would tell his parents that he likely has a low health-related quality of life because his body mass index is low, and that may explain why he's become withdrawn and feels that he looks different from his friends. Again, we will discuss with the family the importance of nutrition.

I would also tell the family there is minimal information available about appetite stimulants. While there is information about cyproheptadine hydrochloride and megestrol acetate, I would tell the parents there is only short-term data, and long-term efficacy is not known. We would then discuss other potential therapeutic options such as gastrostomy.

**MR. BUSKER:** Dr. Pohl, thank you for today's cases and discussion. Let me now ask you to look to the future. What do you see happening to help provide better management of patients who are nutritionally compromised?

**DR. POHL:** We are seeing several things for the future. There's increasing evidence that good nutrition is very important in managing patients with cystic fibrosis. Good nutrition early in life likely affects long-term pulmonary function as well as lifespan.

So what does the future hold? I think we're going to see more information about how to appropriately dose PERT in patients with cystic fibrosis based on the study that we reviewed. We want to find a dosing regimen that allows maximal intestinal absorption while at the same time preventing side effects such as fibrosing colonopathy.

Additionally, the research that's recently been done on protein absorption is intriguing. In patients with cystic fibrosis, with and without PERT use, we still do not fully understand how medications such as PERT work but expect more understanding of their absorption pathways in patients with cystic fibrosis to be more fully elucidated over time.

We will continue to strive for an increased understanding of the risk factors for morbidity and mortality of patients with cystic fibrosis. Although the articles we discussed today looked at children with cystic fibrosis, there are also obvious adult correlations.

We talked about how poor nutrition can decrease health-related quality of life in patients with cystic fibrosis, and we need to do more work to determine what patients are at highest risk based on their age, gender, or pulmonary status. Other aspects to consider are exposure to the newborn screening, earlier diagnosis of cystic fibrosis, and perhaps we can do modeling to allow us to look at nutritional factors such as BMI to address underlying pulmonary status

to identify patients who are high risk early in their medical care. And then we can allocate resources of care appropriately. In this regard the Cystic Fibrosis Foundation patient database is very beneficial.

Finally, we really do not know if appetite stimulants are a helpful adjunct to nutrition care of patients with cystic fibrosis. They appear to work in the short term, but their long term efficacy is really unknown. And only two appetite stimulants, cyproheptadine hydrochloride and megestrol acetate, have been studied in people with cystic fibrosis, and only in short-term trials, so we do not know how effective other appetite stimulant medications are.

**MR. BUSKER:** Thank you for sharing your insights. Let's wrap things up by reviewing today's discussion in light of our learning objectives. So to begin: explaining how appropriate PERT dosing is associated with improved BMI as well as both protein and fat absorption.

**DR. POHL:** We discussed that higher but appropriate PERT dosing is associated with an improvement of body mass index in children and adolescents with cystic fibrosis. We also discussed that PERT dosing has been shown to improve protein absorption in people with cystic fibrosis when compared to healthy controls.

We want to make sure we have an adequate PERT dose without dosing too high and placing the patient at risk of fibrosing colonopathy, which is a serious complication of overuse of PERT.

**MR. BUSKER:** And our second objective: how health-related quality of life is improved in people with CF who have good nutritional status; and conversely, why poor nutrition with a low BMI is a risk factor for mortality, particularly in adolescents with CF.

**DR. POHL:** We looked at two studies today and noted that health related quality of life is lower in adolescents with cystic fibrosis, if they have poor nutritional parameters compared to peers with cystic fibrosis who have improved parameters. This is important for early identification of patients at risk. We also discussed that a low body mass index, combined with an impaired pulmonary status, increases the risk of mortality. This is determined using a multivariate analysis of adolescents with cystic fibrosis. These risk factors should be

considered when evaluating nutritional status of a patient with cystic fibrosis.

**MR. BUSKER:** And finally: the current evidence about the use of appetite stimulants to improve weight gain.

**DR. POHL:** Appetite stimulants have been studied only in the short term; long-term nutritional effects on body mass index in patients with cystic fibrosis are unknown, and more research is needed. Appetite stimulants are a short-term therapy, and if longer-term nutritional therapy is needed we may have to consider other options such as working with a feeding therapist for a younger child, or consider nasogastric or gastrostomy placement.

**MR. BUSKER:** Dr. John Pohl from the University of Utah, thank you for participating in this eCystic Fibrosis Review Podcast.

**DR. POHL:** Thank you for inviting me. It's been a real pleasure talking to you today.

**MR. BUSKER:** To receive CME credit for this activity, please take the post-test at [www.ecysticfibrosisreview.org/test](http://www.ecysticfibrosisreview.org/test).

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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.

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