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Podcast Issue

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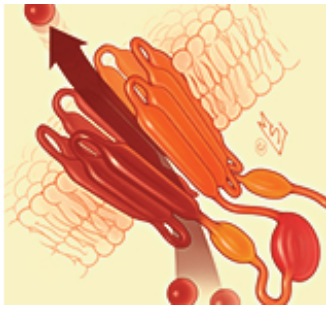

VOLUME 3 — ISSUE 10: TRANSCRIPT

Featured Cases: Interventions to Improve Nutrition in Patients with CF

At the conclusion of this activity, participants will demonstrate the ability to:

- Identify signs and symptoms of gastrointestinal disease that merit further evaluation by a gastroenterologist,
- Discuss important considerations in the nutritional management of patients with CF liver disease and,
- Plan monitoring strategies to ensure optimal outcomes after gastrostomy tube placement.

This audio activity has been developed for clinicians caring for patients with issues related to cystic fibrosis. You can also read the **companion newsletter**. In this edition Dr. Yen will discuss behavioral and nutritional education interventions that provide measurable improvement, as well as the benefits of gastrostomy tube placement for supplemental enteral intake.

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Unlabeled/Unapproved Uses
The author indicates that there will be a reference to unlabeled/unapproved uses of cyproheptadine in the presentation.

MEET THE AUTHORS

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Guest Faculty Disclosures
Dr. Yen discloses that she has an ownership interest in Vertex Pharmaceuticals, Inc., that her husband is an employee of Vertex Pharmaceuticals, Inc., and that she has received an honorarium from Abbott Nutritionals.

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⇩ PROGRAM BEGINS BELOW

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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 an educational grant from Abbott Laboratories, Gilead Sciences Medical Affairs, and Vertex Pharmaceuticals.

Today's program is a companion activity to the April 2012 eCysticFibrosis Review newsletter topic: *Interventions to Improve Nutrition for patients with CF*. Our guest today is Dr. Elizabeth Yen, from the Children's Hospital at Harvard Medical School in Boston.

This activity has been developed for physicians, nurses, respiratory therapists, dietitians, and physical therapists caring for patients with cystic fibrosis. There are no fees or prerequisites for this activity. The Accreditation and Credit Designation Statements can be found at the end of this podcast.

For additional information about accreditation, Hopkins policies, expiration dates, and to take the post-test to receive credit on-line, please go to our website newsletter archive, www.eCysticFibrosisReview.org, and click the May 2012 podcast link.

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

- Identify signs and symptoms of gastrointestinal disease that merit further evaluation by a gastroenterologist,
- Discuss important considerations in the nutritional management of patients with CF liver disease, and,
- Plan monitoring strategies to ensure optimal outcomes after gastrostomy tube placement.

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

I'm **BOB BUSKER**, managing editor of eCysticFibrosis Review. On the line we have with us our April newsletter author Dr. Elizabeth Yen, Assistant in Medicine, Division of Gastroenterology and Nutrition at the Children's Hospital Boston.

DR. ELIZABETH YEN: Well thank you, Bob, I'm very happy to be here and do this podcast with you today.

MR. BUSKER: Dr. Yen has disclosed that she has an ownership interest in Vertex Pharmaceuticals, Inc. and has received an honorarium from Abbott Nutritionals. She has also indicated that her presentation today will include references to the unlabeled or unapproved use of cyproheptadine for the nutritional treatment of patients with cystic fibrosis.

In your newsletter issue, you reviewed new research highlighting the strong positive impact of early behavioral and nutritional education and predictors of long-term response to such interventions. Today we will expand on that information by discussing case scenarios. So please start us out with a presentation.

DR. YEN: Sure. Our first case is of a three year old male patient with cystic fibrosis and pancreatic insufficiency. He presents with poor appetite, chronic vomiting and chronic constipation. He is at the 25th percentile for BMI, but it has been a struggle for the family to maintain this less than ideal growth curve.

His past medical history was additionally notable for being a triplet and born at 35 weeks gestation. There were no sequelae of prematurity, but he has had one hospitalization for a pulmonary exacerbation requiring IV antibiotics.

In the family medical history, we have no note of gastrointestinal diseases or cystic fibrosis. He is on some medications currently and these include pancreatic enzyme supplements, lansoprazole, cyproheptadine and respiratory therapies.

His workup thus far has included an upper GI series which did not show evidence of malrotation, but did show some mild reflux.

MR. BUSKER: This patient has many symptoms that interfere with oral intake. What treatment approaches should you consider to improve his caloric intake?

DR. YEN: Indeed, many things are interfering with his oral intake. I think an important note to take right from the beginning is that he's already on a few GI active medications, such as the cyproheptadine and the lansoprazole, and he's still symptomatic. So at this

point I would definitely consider a gastroenterology consultation possibly for additional workup in addition to interventions. Also, the family should have some nutritional education which would already be part of their cystic fibrosis care, but in particular, given that this child has an issue with eating foods in any significant quantity, we would want to make sure that the caloric density of his foods is maximized. So this requires quite a bit of nutritional education of the family.

I also think it would be important to make a careful assessment of mealtime behaviors. Is this child allowed to graze throughout the day so that he can eat whatever he can, which would ultimately interfere with his appetite and possibly result in less total intake, or are mealtimes organized? Additionally, the question of what is the kid doing at mealtimes, is he being goaded to eat or is there begging on the side of the parents, is there bargaining going on, all of these things could actually interfere with the child eating as much as he needs to grow.

MR. BUSKER: Isn't this some of the information you reviewed in the newsletter?

DR. YEN: Yes, it is. Listeners can see these references in the newsletter…

MR. BUSKER: Treatment of constipation improves the vomiting, but poor appetite and inadequate caloric intake continue. What additional steps can you take?

DR. YEN: Well one thing that as a gastroenterologist I think about, is the effect of the appetite stimulant. In this case he's using cyproheptadine and from clinical experience I know that the effect of cyproheptadine can wear off over time. As a result, I tend to recommend to my patients to cycle the cyproheptadine. Some patients prefer to take it only during the weekdays and hold it on the weekends, other patients prefer to take it continuously for three or four weeks and then hold the medication for five days and resume the medication again. This seems to work to improve the effect of the cyproheptadine overall so that it can last a longer period of time.

Other interventions to consider here, of course, are behavioral interventions to improve the mealtime behaviors and this is really not just with the child, but with the entire family or caretakers who are feeding the child. This might be through a clinical psychologist or other resources available through the cystic fibrosis center.

We may also consider increasing the dose of the proton pump inhibitor. We don't know from this case presentation how long this child has been on the lansoprazole, but we do know that if a patient is on this type of medication for several years, he can actually have an increase in the total parietal cell mass in his stomach, and, therefore, the original dose may become less effective over time.

We know from the case presentation that he is still experiencing some reflux, and this may be adversely impacting his appetite. Some patients experience nausea, he can't tell us, he's only three years old, but certainly, I would want to try this intervention before moving forward with anything more invasive.

MR. BUSKER: What additional diagnostic testing should be considered?

DR. YEN: Yes. Well, one thing as a gastroenterologist I often consider is celiac disease. About 1 in 150 Americans have celiac disease, and so it's important not to lose sight of how common this can be and how much of an impact this can have on somebody's nutritional status.

The testing for celiac disease initially is screening blood test, and this includes markers such as tissue transglutaminase antibody and endomysial antibody. These are IgA based tests and so at total IgA level is necessary, as well, in order to be able to interpret the result of that celiac screen.

I would also consider at this point endoscopic evaluation of both the upper and lower GI tract, because the patient is having a lot of symptoms, particularly the vomiting and the constipation, that can be a sign of a more significant disease. Specifically I'm thinking of allergic GI disease, or inflammatory GI disease, that is currently not being treated.

MR. BUSKER: Would you consider food allergies?

DR. YEN: I would. It may be wiser to wait for the results of the endoscopy. It's controversial whether or not testing for food allergies, either through RAS testing of serum or skin prick testing is wise and correlates with improvement in symptoms without there actually being a demonstration of

gastrointestinal injury from an allergic process. So if we do find that there is eosinophilic infiltration at all at any point in the GI tract, then it does make sense to do food allergy testing. However, if there is no evidence of allergy at the microscopic level, then it is still debated. Perhaps a formal allergy evaluation would be the best approach.

MR. BUSKER: How do you approach placement of a gastrostomy tube?

DR. YEN: We could consider placement of a gastrostomy tube if all other interventions are not resulting in increased caloric intake. One thing we have to be cautious about is this chronic vomiting. We want to make sure we have a diagnosis and treatment for this vomiting before we put in a gastrostomy tube. In some cases, after placement of a gastrostomy tube you get leveling of reflux and vomiting.

MR. BUSKER: Let's assume you, in this patient you do consider that, how would you approach the placement of a gastrostomy tube?

DR. YEN: This may be institution dependent, but in our institution, we recommend gastroenterology consultation for education of the patient and the families of the procedure. We show them what the tube looks like on a doll and how we would care for it.

The tube can be placed either by a gastroenterologist or a surgeon, and there are two different placement approaches. The gastroenterologist would do an endoscopic placement of a PEG tube, that's a percutaneous endoscopically placed gastrostomy. This tube has a disadvantage in that it's a long tube that's always long, for anywhere from three to six months after placement before it can get exchanged for a skin level device.

On the other hand, the surgical approach might be a longer intraoperative procedure but the end result is a skin level device right from the get-go. And many patients with CF, they prefer going directly to the skin-level device, it really is up to the family and the patient.

We also find it important to make sure that the parents and the patients have contact with other patients with G tubes. Obviously we try not to pair them up with other CF patients with G tubes, just for infectious precaution, but there are many patient families in the G tube community that are willing to share their experiences with patients who are considering placement of a G tube. Also online now, the CF community can talk a lot about how successful these things have been in each case.

MR. BUSKER: How would you explain to the family what the goal and the expectations for a G tube placement would be?

DR. YEN: I would start with explaining that placement of the G tube does not mean that the patient isn't going to eat by mouth. It's important that we only use the G tube primarily at night while the patient is sleeping as a way to add calories to the 24 hour intake.

During the day the G tube will be clamped and he or she will eat regular amounts of food, as much as they can tolerate. The overnight feeds will gradually be ramped up to a percent of total core at goal. So let's say we want eventually to get to the overnight feeds be 40 percent of the total calories for the day, we'll probably start at 15 percent and gradually ramp it up so that we don't trigger any GI symptoms, such as pain, intolerance, nausea, et cetera. We want this to be a pleasant experience. Over time, we would expect weight gain and improvement in BMI.

MR. BUSKER: Shall we transition to case No. 2?

DR. YEN: Certainly. The next patient is a 15 year old female with CF and pancreatic insufficiency. She presents with symptoms of abdominal bloating and pain after eating. She has cut down her total intake because of the symptoms over the past four months, and her BMI has gone down from 45 percent to 30th percentile. She's otherwise well and has no respiratory symptoms.

Her past medical history is notable for mild lung disease. She suffers from chronic sinusitis and had a surgical drainage procedure one year ago. She takes antibiotics to clear her sinusitis on average every three to four months. She is on her standard respiratory therapies and takes omeprazole twice a day. She is also on a weekly bowel regimen, taking stool softeners and a stimulant laxative combination to prevent constipation problems. She does this every weekend. She also takes that soluble vitamin.

Her family medical history is important. There's Hashimoto's thyroiditis in the mother and maternal aunt. There is also a history of osteoporosis in the maternal grandmother and great grandmother.

Her dietary history is notable for dairy consumption with almost every meal. She is compliant with taking her pancreatic enzymes. On exam, her abdomen is slightly distended, but soft and it's not tender. Here the differential diagnosis includes lactose intolerance, peptic ulcer disease, celiac disease, small intestinal bacterial overgrowth and chronic constipation.

MR. BUSKER: Please tell us some pertinent factors about our second case.

DR. YEN: Well the first thing I notice is that her symptoms seem to be triggered by food. So this makes me think about a few things, is there excessive secretion of acid production in the stomach that's aggravating some injury such as an ulcer? I wonder is she having some alterations in her digestive process of absorption of nutrients that are causing her to have this abdominal bloating and discomfort, and then I also wonder whether there may be some motility problems where the food is just not traveling down the pipe as well as it should, and, therefore, contributing to some of the bloating and gas.

I also think that the family history is very important here. There is thyroiditis and osteoporosis, and both of these are associated with celiac disease. So I will make sure to consider that in my differential.

And then finally, I also think that her frequent antibiotic exposures are having an impact on her GI tract. All of these antibiotics can alter the bacteria that normally grow in the GI tract, and as a result she can have some bacterial, what we call dysbiosis, so not the bacteria that are necessarily promoting healthy intestinal function, and she may be a candidate for probiotics.

MR. BUSKER: What diagnostic testing should you consider?

DR. YEN: Well once again, here I would consider looking at her celiac serologies, that tissue transglutaminase, IgA antibody and the endomysial IgA antibody. I also would consider a lactose-hydrogen breath test because she is a big dairy consumer, and it's important that we not cut out the dairy in her diet because it probably is giving her a lot of calories that are important for her. So I would go directly to a breath test to get a yes or no answer as to whether or not she is lactose intolerant.

I would also consider an abdominal x-ray. She has a history of constipation and even though she's doing these weekly clean-outs, it might not be enough. She might be significantly backed up and this could be contributing to her abdominal bloating and discomfort with eating.

MR. BUSKER: One of the celiac tests is positive. What are the next steps for this patient?

DR. YEN: The celiac serology testing is a screening test. While it does have very high sensitivity and specificity, it's still not 100 percent. And in a patient with cystic fibrosis, we know that there are sometimes inflammatory lesions in the small intestine which could theoretically give you a false positive read on a celiac serology. Therefore, it is important to proceed with an endoscopy for small intestinal biopsies to look for the telltale signs of celiac disease on microscopy. And that includes blunting or atrophy of the intestinal villi, increased intra-epithelial lymphocytes, and elongation of the intestinal crypt.

Once the diagnosis is confirmed, then she would need to move on to nutritional education on a gluten-free diet. Celiac is treated with a strictly gluten-free diet. With the gluten-free diet, the injury from celiac disease is completely eliminated. This is important because the injury in celiac disease interferes with absorption of important nutrients. Once on a gluten-free diet she will need close follow-up of her symptoms and monitoring of her dietary intake to make sure that she is gaining weight and doing well.

MR. BUSKER: How does celiac disease affect the nutritional status of this CF patient?

DR. YEN: Well this gluten-free diet certainly poses greater challenges for her to meet caloric requirements. It's going to be difficult for her to eat out with her friends and family, she will need to be very cautious to insure that there is not cross-contamination of gluten in the food that she eats. So it might require a very significant lifestyle change for her.

Apart from the dietary restrictions posing challenges, she'll also have a higher risk of low bone mineral density due to the impact of celiac disease on vitamin D and calcium metabolism. Vitamin D is absorbed intestinally, as we know it is already impaired in cystic fibrosis because of the pancreatic insufficiency, but

celiac disease can further impair this absorption. It can also impair calcium absorption.

Other important nutrients that can be impacted in celiac disease include iron absorption and other micro nutrients. Iron is exclusively absorbed in the duodenum and celiac disease has the strongest impact on the duodenum.

MR. BUSKER: We'll return in a moment with Dr. Elizabeth Yen from the Children's Hospital Boston.

MR. BUSKER: Welcome back to our May 2012 eCysticFibrosis Review podcast. I'm Bob Busker, managing editor of the program. Our guest is Dr. Elizabeth Yen from Children's Hospital Boston and Harvard University. Our topic is *Interventions to Improve Nutrition for patients with CF.*

We've been discussing how the information in our April newsletter issue can be applied in the exam room. So let's continue with another age group and case scenario, Dr. Yen?

DR. YEN: The next patient is a 13 year old male with CF, pancreatic insufficiency, liver disease, and a BMI at the 50th percentile for age. His lung disease is mild. He presents for routine follow-up and has no active respiratory or GI symptoms.

His past medical history is notable for diagnosis of liver disease during early childhood. His last ultrasound a year ago showed and enlarged spleen, and a normal sized liver with a hyperechoic, heterogeneous echotexture.

His current medications are pancreatic enzyme supplements, a proton pump inhibitor, ursodiol, respiratory therapy and fat soluble vitamins. The family history is unremarkable. He is very active in soccer and skiing.

On exam, his abdomen is mildly distended, the liver edge is not palpable, but the spleen tip extends to the level of the umbilicus and the spleen is firm. There is no evidence of ascites on exam. His extremities are muscular but with little overlying fat.

MR. BUSKER: What are some important points in this case?

DR. YEN: We have no active symptoms, but this patient has liver disease and splenomegaly. These are signs of portal hypertension and they will require some careful monitoring.

He also is a very active young man, so his caloric requirements are going to be elevated.

MR. BUSKER: What considerations should be taken when assessing this patient's nutritional status?

DR. YEN: Interestingly, his BMI was at the 50th percentile, but the question is how much of this is fluid weight that's in his abdomen, specifically in his spleen. The BMI may be misleading in this case, therefore, it's important for a nutritionist or dietician to measure mid-arm anthropometrics, these include the triceps skin fold thickness, and the mid-arm circumference. Other measures include bioelectrical impedance measurement, which can give us a sense of the percent body fat.

Also, because of his liver disease, he's at higher risk for the fat soluble vitamin deficiencies. We know that there is already a risk for this in CF, but in the case of liver disease, bile salt synthesis is further decreased and can further interfere with absorption of these vital nutrients.

MR. BUSKER: What increased risks do the physical findings indicate?

DR. YEN: His physical findings, again indicate portal hypertension. So we want to make sure that this patient is followed by a hepatologist. He's at risk for gastrointestinal bleeding from portal hypertension, which can present as esophageal varicies and gastric varicies. He also is at risk nutritionally from early satiety due to mass effect in the abdomen from that very large spleen.

Additionally, that large spleen is particularly at risk for abdominal trauma. He's very active in soccer and skiing, so he'll need to wear some form of shield device to protect that spleen.

MR. BUSKER: What considerations should be taken in regard to this patient's nutritional management?

DR. YEN: Even though is BMI is at the 50th percentile, as I mentioned earlier, this may be misleading. I would definitely focus on dietary

supplements to improve his fat stores. The physical exam did show that he had good muscle tone but decreased fat stores and we want to make sure that he has enough fat stores.

In liver disease, we also have to worry about loss of liver function where the glucose tolerance is lower and the glycogen stores are lower. So this can actually lead to an accelerated starvation state, therefore, we want to make sure that he has excess calories onboard.

At this point, I might also start educating him on nasogastric tube feeds, as well as potentially gastrostomy tube feeds. I don't want this to be brought in at the very last minute, I want him to be aware of the possibility of needing these interventions early on. He certainly doesn't seem to be needing it now, but it's best to have him aware early.

Additionally, I might consider medications to augment his appetite. In an earlier case we mentioned cyproheptadine as an appetite stimulant, that's something that I might bring up here.

MR. BUSKER: Shall we go to our next patient scenario?

DR. YEN: This last patient is a 14 year old female with CF and pancreatic insufficiency. She has mild liver disease, a history of poor weight gain, and has a gastrostomy tube placed for supplemental enteral feeds. She complains of abdominal pain, nausea and vomiting with her overnight enteral feeds. She has found that the enteral feeds are tolerable at a maximum rate of 40 milliliters an hour. Currently, her supplemental feeds are only providing about 20 percent of her total recommended daily calories.

Her past medical history is notable for mild lung disease. Two years ago her BMI fell over the course of one year from 35th percentile to 15th percentile. Nutritional education, appetite stimulants, optimizing of pancreatic enzymes and treatment of mild respiratory symptoms did not result in improvements in her weight gain. At that point a gastrostomy tube was recommended. She initially experienced improvements in her BMI to the 40th percentile. This was the best she had ever been at.

Her current medications include pancreatic enzymes, a proton pump inhibitor, ursodiol, respiratory therapies and fat soluble vitamins. Overnight her enteral supplement is a formula composed of whole protein with a caloric density of 1.5 kilocalories/milliliter, and 10 percent NCT oil. During the daytime she averages 80 to 90 percent of her caloric goal by mouth.

The family history is notable for depression in the mother that is currently well treated. The patient, herself, has missed a lot of school lately because of complaints of abdominal pain in the morning. On exam, her weight is down 1 kilogram from the previous visit just two months ago, and her BMI is at the 25th percentile. She has a flat affect. Her abdomen is soft, nontender, nondistended, with a well healed gastrostomy. A stool mass is palpable in the left lower quadrant.

MR. BUSKER: What are the important features of this case?

DR. YEN: Well this young woman has weight loss and that's very concerning, especially since she has a gastrostomy tube in place. That makes me concerned that either it's not being used properly or perhaps there is something else interfering with her intake. She also has GI symptoms that are not being addressed.

MR. BUSKER: What diagnostic testing might you consider for her weight loss and GI symptoms?

DR. YEN: On exam I also felt a stool mass, so I'm going to start off with an abdominal x-ray and see what the stool burden looks like. If there is evidence of constipation on the x-ray I may consider a significant bowel cleanout followed by maintenance medications for her constipation.

Again, I'm going to consider celiac serologies because I always consider it when there are GI symptoms that are not explainable by other findings. And finally I'm going to consider a gastroenterology consultation if this constipation cleanout doesn't improve her symptoms.

MR. BUSKER: What changes in her nutritional management can be trialed while workup for GI symptoms is completed?

DR. YEN: Well, it should be noted that her pain and nausea and vomiting is occurring in the morning after her overnight feeds. So I might target that formula and change it to something that is more easily

digested. A polypeptide formula with higher MCT percent is indicated.

I would also consider an appetite stimulant medication since she is not on one right now. She had trialed one in the past and it wasn't effective, but I would trial it again. And I would make sure that her proton pump inhibitor is optimized with twice daily dosing to minimize reflux impact on nausea and abdominal pain.

MR. BUSKER: What psychosocial factors in this case might impact success with enteral supplementation?

DR. YEN: Yes. Well this patient has some depressive symptoms. She is missing school, she has a flat affect, and she also has a family history of depression. So we definitely have to consider the impact of depression on her oral intake and her tolerance of her enteral feeds.

Also, this missing of school may be related to some difficulty with her peers. Are they accepting of the fact that she has a G tube. Perhaps some self-esteem issues are also contributing to the school absenteeism. It was mentioned in the review that poor body satisfaction is associated with inadequate caloric intake in CF patients, particularly in young women with CF. So this is something that we certainly have to address and perhaps refer to a psychologist or a psychiatrist.

MR. BUSKER: Would you please summarize our program today?

DR. YEN: Well, CF patients are living longer and so we think we will be looking at nutritional status in older CF patients and how to optimize this. Also, there has been some interesting research in the adult critical care literature on the use of anti-inflammatory nutritional supplements that I think would be interesting to look at in the CF population.

We continue to consider whether or not it's useful to use omega-3 fatty acids in high doses for CF. And I think also, as we consider the nutritional status of older patients, we'll have to continue to assess our use of gastrostomy tubes and see if we can come up with guidelines on when to really consider their use and how to best — sorry, and how to optimize its use.

MR. BUSKER: Dr. Yen, I just want to summarize what we talked about today, what did we learn about identifying the signs and symptoms of GI disease that can merit further evaluation by a gastroenterologist?

DR. YEN: Well we've seen that symptoms such as vomiting, anorexia, bloating, abdominal pain, can all be presenting signs of various GI diseases that require specific diagnosis and treatment of, such as celiac disease, small intestinal bacterial overgrowth, lactose intolerance and peptic ulcer disease.

MR. BUSKER: The most important considerations in the nutritional management of patients with CF liver disease?

DR. YEN: We learned that CF liver disease has higher nutrient requirements because of the lack of function of the liver and that there is a higher risk for fat soluble vitamin deficiencies due to the low bile salt pool.

MR. BUSKER: Monitoring strategies to insure optimal outcomes after G tube placement?

DR. YEN: Well we saw in the last case that even after placement of a gastrostomy tube you can have some weight loss. So it's important to keep track of its use, make sure that the patient is tolerating the feeds, and that we are meeting the nutritional goals.

MR. BUSKER: I want to thank you, Dr. Elizabeth Yen from the Division of Gastroenterology and Nutrition, at Children's Hospital Boston and the Faculty in Pediatrics at the Harvard Medical School, participating in this eCystic Fibrosis Review podcast.

DR. YEN: Well thank you, it was quite a pleasure.

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.

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