



VOLUME 2 – ISSUE 6: TRANSCRIPT

Featured Cases: Nutritional Challenges and Complications in Patients with Cystic Fibrosis

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

- Evaluate and treat small bowel overgrowth in CF
- Evaluate and treat distal intestinal obstruction syndrome in CF
- Consider non-enzyme causes of malabsorption in a CF patient with poor growth.

This audio activity has been developed for clinicians caring for patients with issues related to cystic fibrosis. You can also read the **companion newsletter**. Dr. Schwarzenberg will help expand our understanding of the nutritional challenges for patients with CF, with the discussion some typical case scenarios.

Unlabeled/Unapproved Uses

The author has indicated that there will be no reference to unlabeled or unapproved uses of drugs or products in the presentation.

MEET THE AUTHOR



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Guest Faculty Disclosure

Dr. Schwarzenberg discloses that she has no financial relationship with commercial supporters.

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MR. BOB BUSKER: Welcome to this eCystic Fibrosis Review podcast. eCystic FibrosisReview 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 Genentech, Eurand Pharmaceuticals, Vertex Pharmaceuticals and Axcan Pharma, and Gilead Sciences. Today's program is a companion piece to our April, 2010, eCystic Fibrosis Review Newsletter, "Nutritional Challenges and Complications."

Our guest is Dr. Sarah Jane Schwarzenberg from the University of Minnesota Amplatz Children's Hospital. This activity has been developed for physicians, nurses, respiratory therapists, dieticians, and physical therapists caring for patients with issues related to 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 online, please go to our website newsletter archive, www.ecysticfibrosisreview.org, and click in the May, 2010, podcast link.

Learning objectives for this program are that at the conclusion of this audio activity, participants should be better able to evaluate and treat small bowel overgrowth in the cystic fibrosis patient, evaluate and treat distal intestinal obstruction syndrome in the CF patient, and assess non-enzyme causes of malabsorption in a CF patient with poor growth.

I'm **BOB BUSKER**, managing editor of eCysticFibrosis Review. On the line we have with us the newsletter issue's author, Dr. Sarah Jane Schwarzenberg is Associate Professor of Pediatrics and Director of the Division of Gastroenterology, Hepatology and Nutrition at the University of Minnesota Amplatz Children's Hospital in Minneapolis. She has disclosed that she has no relationships with commercial supporters, and that her presentation today will not include discussion of off label uses.

Dr. Schwarzenberg, welcome to this eCystic Fibrosis Review podcast.

DR. SARAH JANE SCHWARZENBERG: I'm very glad to be here.

MR. BUSKER: To help expand our understanding of nutritional issues and complications in cystic fibrosis, we have asked Dr. Schwarzenberg to discuss some typical case scenarios. So if you would, Dr. Schwarzenberg, please present our first case to address the issue of small bowel overgrowth in the CF patient.

DR. SCHWARZENBERG: A 30 year old woman with pancreatic insufficient CF presents in gastroenterology clinic. She is experiencing social embarrassment because of excessive bowel gas which she cannot control. She is also stooling two times daily and expresses no concerns about constipation. She has noted some nausea recently and, in fact, has lost two pounds of weight. At her last visit to her pulmonologist she was told her lung function was stable.

MR. BUSKER: As the clinician works up the differential, what GI problems should be considered?

DR. SCHWARZENBERG: I think in any young woman with cystic fibrosis or without cystic fibrosis who comes in complaining of nausea, we have to consider pregnancy. Occasionally you will find someone in this age group with some lactose intolerance causing excessive bowel gas, but in a cystic fibrosis patient, one of the first things you would think of in this situation would be small bowel overgrowth with associated malabsorption of carbohydrates, leading to the excessive bowel gas, and also leading to the nausea and potentially the weight loss.

MR. BUSKER: Let's translate that into diagnostic tests that can be used to evaluate this patient's GI problems.

DR. SCHWARZENBERG: In patients like this we would always want to do a urine pregnancy test. I think the most important thing to consider at this point is small intestinal bacterial overgrowth. And there are several different ways to evaluate small intestinal bacterial overgrowth, some of them more useful than others.

For example, the gold standard in this is jejunal aspiration with quantitation of bacteria, but that's a very difficult test for both the endoscopists and the patient. And often culturing the bacteria is very difficult and you don't get an accurate count. Whereas hydrogen studies are used in some centers, they also suffer from some problems in people with cystic fibrosis. And I think many clinicians would use a trial of antibiotic therapy in a cystic fibrosis patient that they thought had small bowel overgrowth simply because the prevalence appears to be pretty high, perhaps as many as 50 percent of our patients suffer from time to time with small bowel overgrowth, and often you can get a very rapid response and have some reason to believe you have made an accurate diagnosis with a 10-day course of antibiotic therapy.

MR. BUSKER: I would like to step back to something you just said, the breath hydrogen study, why isn't that performed more often?

DR. SCHWARZENBERG: I think in breath hydrogen studies in cystic fibrosis patients, there are a couple of different problems. First of all, many of our patients, particularly as they grow older, have some degree of delayed gastric emptying, or even intestinal dysmotility. And this can create difficulty in interpreting the peaks in the breath hydrogen test and can lead to confusion in making a diagnosis.

Also, some of our patients have enough lung disease that the breath hydrogen test is less accurate. And then many patients with cystic fibrosis are on antibiotic therapy almost all the time, which can also make interpreting the breath hydrogen test very difficult.

So while I think that a lot of centers use it, I've used it from time to time in patients with CF, I think that there is also a place for good clinical judgment in making a decision to treat this disease.

MR. BUSKER: Now this specific patient that you described to us, how was she treated?

DR. SCHWARZENBERG: She received metronidazole for 10 days, and this dramatically improved her bowel gas and her nausea, which allowed her to feel a lot more comfortable in social situations and to regain her lost weight. However, about two months after we treated her, she called the office saying that these symptoms had returned, perhaps not quite as bad as

they had been when we treated her initially, but she certainly was experiencing the significant bowel gas again.

MR. BUSKER: What was your course of action to address this return of symptoms?

DR. SCHWARZENBERG: In this particular patient we chose to repeat her treatment with metronidazole, to see if we could again get the same effect and improve her condition again. It's really pretty common for any patient with small bowel overgrowth, and certainly patients with cystic fibrosis, to have recurrent episodes of small bowel overgrowth, and different clinicians treat this in different ways.

Some clinicians have chosen to give short courses of antibiotic therapy monthly to patients who have recurrent episodes of small bowel overgrowth. So, for example, for the first five days of each month the patient may take metronidazole or some other antibiotic. Some investigators have suggested using an intestinal cleanout every month or two months with polyethylene glycol to try to maintain reduction in numbers of bacteria in the small intestine.

I think that in patients who have recurrent episodes of small bowel overgrowth there may be some value in doing intestinal imaging with an upper GI small bowel follow-through to make sure that you're not dealing with a partial obstruction that might be contributing to small bowel overgrowth. This is particularly important in somebody who has had previous gastrointestinal surgery, and in some cases it may be wise to further investigate the problem.

For example, doing endoscopy with biopsies to see whether you've made the correct diagnosis, is this a patient with celiac disease or some other intestinal infections who is mistakenly being considered to be a patient with small bowel overgrowth.

MR. BUSKER: The weight loss this patient experienced, how does that fit into the overall picture?

DR. SCHWARZENBERG: I think that patients with cystic fibrosis have many reasons for losing weight with different gastrointestinal problems. In patient with small bowel overgrowth there is often abdominal pain or nausea that can reduce their appetite. These patients have to work very hard to maintain their weight when they are feeling healthy, and so even a small reduction in appetite could lead to weight loss.

Small bowel overgrowth, in particular, can lead to malabsorption of nutrients, and even anemia in some cases, and certain vitamin and mineral deficiencies. I think that is one of the reasons why we believe this is such an important entity to understand and treat correctly in cystic fibrosis, because weight management and good nutritional health are so critical to maintaining good lung function in cystic fibrosis.

MR. BUSKER: Well thank you, Dr. Schwarzenberg. I would like us to shift gears now and look at distal intestinal obstruction syndrome. If you would, please, present us with another patient.

DR. SCHWARZENBERG: A six year old boy with pancreatic insufficient cystic fibrosis, presents in the emergency department with a six hour history of vomiting. He was well until three days ago when he developed progressive right lower quadrant abdominal pain. He has not had a stool in two days. He had a recent pulmonary exacerbation, now well controlled, he has been otherwise healthy.

On reviewing his past medical history you note that he had surgery in infancy for meconium ileus and lost 4 centimeters of bowel. On physical exam, he is an ill appearing boy with dry lips, his abdomen is mildly distended. A 2 centimeter by 3 centimeter mass is palpable in his right lower quadrant.

His rectal exam has no stool present and you elicit no significant tenderness during the rectal exam.

MR. BUSKER: With this patient what would you recommend as the initial approach?

DR. SCHWARZENBERG: One thing that it's important to note is that this child appears dehydrated. And I think that we have to address that before we address almost anything else, because dehydration will contribute to all of his other problems and eventually to concerns about his lung disease.

So we would obtain electrolytes, a BUN, creatinine, urinalysis and a CBC. We would probably do a flat plate and upright of his abdomen, and an ultrasound of his right lower quadrant to evaluate for appendicitis. I think when people are evaluating people with cystic fibrosis, they often focus completely on the cystic fibrosis and cystic fibrosis related

gastrointestinal disease, but a six year old boy could have any of the ordinary problems that six year old boys have that bring them to the emergency room, including gastroenteritis or appendicitis. And it is important not to lose track of the fact that he's still a six year old boy even if he has cystic fibrosis.

MR. BUSKER: And what did these tests find?

DR. SCHWARZENBERG: The flat plate of the abdomen showed an obstructive pattern with air/fluid levels in the small intestine. In addition, our laboratory studies, as well as our clinical exams, demonstrated moderate dehydration, although his electrolyte panel was normal.

MR. BUSKER: Based on these results, what would your diagnosis and next steps be?

DR. SCHWARZENBERG: This young boy meets the criteria for complete distal intestinal obstruction syndrome described by the European Society for Pediatric Gastroenterology in a recent article in *Journal of Pediatric Gastroenterology and Nutrition*. This has been a very interesting piece of progress in our diagnosis and management of children with distal intestinal obstruction syndrome.

In the past we had a somewhat general definition of this problem, but I think with the work of this particular society, we now have some fairly standard definition for complete distal intestinal obstruction syndrome, partial DIOS, or impending DIOS, and constipation, that will allow us to make consistent diagnoses.

So as I said, this young man met the definition for complete DIOS and we began his treatment by starting an IV and rapidly initiating hydration with normal saline, so he received normal saline boluses that totaled 20 cc/kilo. We also made him NPO and we decided we would hold off placing an NG tube, but we were going to place an NG tube if he continued vomiting. I think that six year old boys don't like NG tubes, but I think that recurrent vomiting, particularly in the context of someone with lung disease, can be a source of aspiration pneumonia or sometimes patients can develop a tear in their esophagus, a Mallory Weiss tear, that can lead to very significant bleeding.

So we felt that if we were able to end his vomiting simply by making him NPO and getting him well

hydrated, we wouldn't need to place a tube, but I don't think anybody should be afraid to place an NG tube if they feel they need it.

MR. BUSKER: Once hydration progresses, what would you do next, and please, tell us why?

DR. SCHWARZENBERG: As this child became hydrated, we began to turn towards relieving the obstruction. I think that in complete DIOS it can be somewhat risky to use an oral agent like polyethylene glycol to try to relieve the distal intestinal obstruction. If the child cannot manage to keep down clear liquids, it is very unlikely that he is going to manage to keep down the polyethylene glycol, and again, you put him at a risk of aspiration by trying to force a liter of fluid, let's say, into his intestine, to try to relieve the obstruction.

So in a complete distal intestinal obstruction I would not try to relieve that from above. I would keep the child NPO, keep him well hydrated with IV fluids, and I would have the radiologist come in and do a gastrografin enema to try to relieve the distal intestinal obstruction. Most of the time they're quite successful.

Occasionally, we have patients who require endoscopy reduction of their obstruction. Rarely, we have had to progress to a surgical reduction of the obstruction. I think that one or even two gastrografin enemas are quite safe if the child is well hydrated and you are monitoring him quite closely.

MR. BUSKER: And we will return in a moment with Dr. Sarah Jane Schwarzenberg from the University of Minnesota.

MS. MEGAN RAMSEY: Hello, I'm Megan Ramsey, nurse practitioner and clinical coordinator for adults at 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.

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eCysticFibrosis Review, a bimonthly, e-mailed delivered program, available by subscribing. Each issue reviews current literature on focused topics, important to clinicians caring for patients with cystic fibrosis.

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MR. BUSKER: Welcome back to our May, 2010, eCysticFibrosis Review Podcast. I'm Bob Busker, Managing Editor of eCystic Fibrosis Review. Our guest is Dr. Sarah Jane Schwarzenberg, Associate Professor of Pediatrics and Director of the Division of Gastroenterology, Hepatology and Nutrition at the University of Minnesota Amplatz Children's Hospital, and our topic is nutritional challenges and complications in cystic fibrosis. We have been bringing the information presented in our April newsletter into clinical practice via case study scenarios.

Let's continue now with our current patient, a six year old boy with pancreatic insufficiency and CF who has been diagnosed and treated for distal intestinal obstruction syndrome. Dr. Schwarzenberg.

DR. SCHWARZENBERG: The child that we just talked about had resolution of his DIOS after gastrografin enema and good hydration. Two weeks later he returned with mild abdominal distention and nausea. His mother said she had been giving him senna about every other day. He stooled after the senna and felt better for a short period of time, but then failed to stool again and his symptoms developed once again. He's been eating and drinking, but not as well as when he first left the hospital.

MR. BUSKER: Based on what you just presented, what would be your tentative diagnosis, and again, please tell us why?

DR. SCHWARZENBERG: This young boy would meet the criteria for impending distal intestinal obstruction syndrome. He has a short history of abdominal pain and/or distention, he's had a change in frequency or consistency of his bowel movements in the last few weeks, and he has gotten relief of symptoms with laxatives. Senna wouldn't always be my first choice of a treatment for a child with constipation, but I think it proves the principal here that he could at least temporarily receive some relief with a laxative.

And I think the first thing that people might say is why isn't this constipation? Distension is generally not characteristic of constipation, constipation is a non-distending reduction in stool movements. I think we also know that in many cases people who have had a single episode of distal intestinal obstruction syndrome seem to be at risk for a second episode within several weeks of the first episode. Exactly why that is I can't say and I think certainly with the new guidelines available to us it's going to be interesting to see if that pans out in studies. But it has been a clinical observation by several people.

MR. BUSKER: Based on your clinical experience, what factors might be contributing to his recurring DIOS?

DR. SCHWARZENBERG: We know that this is a child who previously had surgery for meconium ileus, and we know that meconium ileus with surgery appears to be a risk factor for later development of distal intestinal obstruction syndrome. This has been demonstrated now in I believe two studies.

We know that this is a child who had poor fluid intake with mild dehydration in the past, and even though he has been eating and drinking since he left the hospital he is obviously not doing as well as he was when he first left the hospital, and so he may gradually be getting dehydrated again.

There are many other factors, however, that we could question him on to see what might be contributing to this second episode. We know that sometimes children skip their pancreatic enzymes and that skipping pancreatic enzymes can lead to undigested food in the small intestine, which can contribute to distal intestinal obstruction syndrome. This child had a recent pulmonary exacerbation, which sometimes can be associated with an episode of DIOS, possibly because of slowing of intestinal motility. And I would also want to ask him about any dietary changes. If this

is a child, perhaps, who has increased his fat intake dramatically recently, and perhaps is not receiving enough enzyme to completely digest his fats. Any of these things could be contributing factors.

MR. BUSKER: Talk to us now about therapeutic options, doctor, what would be appropriate therapy to treat this episode?

DR. SCHWARZENBERG: The nice thing about this episode is that the child has not reached the point where he is completely obstructed. So he is still able to have an intake of fluids. Every child with distal intestinal obstruction syndrome needs to be hydrated well and I think we would have to make a clinical judgment as to whether we could hydrate him orally or whether we needed to place an IV, perhaps in the emergency room, and give him a bolus of normal saline in order to improve his hydration

This is a child that I would probably try to clear out with polyethylene glycol lavage, the same strategy you might use in a child who you were trying to disimpact with severe constipation or clean out for a colonoscopy. At our institution, we've had very good success with placing a small nasogastric tube and giving 10 ccs or 15 ccs per kilo per hour of polyethylene glycol solution through and NG tube.

It's important when you are using volumes like this to use a product that has electrolytes in it. We often use PEG solutions that have no electrolytes orally in children because they are so palatable. But that would be an inappropriate product to use in large volume for a complete lavage because you could risk water intoxication in the child.

MR. BUSKER: And your follow-up plan?

DR. SCHWARZENBERG: Once we have this young man relieved of his impending distal intestinal obstruction syndrome, it would be important to put him on therapy to try to prevent another episode from occurring. I think we would want to have him taking a PEG solution daily for one to two months. And here you could go back and use the more palatable solution that has no electrolytes in it, because the dose will be pretty small.

We would really want to try to improve his fluid intake during the day, and with a six year old, I would probably tell his mom to buy him a really snazzy water

bottle of some kind that he could carry to school. You would want to be sure the school would allow him to do that, and set a goal for him of water that he should drink during the day, to try to really push and improve his hydration.

If we continued to see this young man bank in clinic with recurrent episodes of DIOS, that would be a time when we might think about doing some imaging studies. We have tried to move to MR enterography for these studies. In the past we would have done an upper GI small bowel follow through looking for a narrowing, or even sometimes a nociception in the terminal ileum at the site of his previous surgery. MR enterography will give us some of the same information without having to use radiation.

Sometimes our radiologists will supplement that with an ultrasound of the right lower quadrant if they feel as though they are seeing something on MR enterography and want to better delineate it.

I think we would also want to reinforce the importance of using his pancreatic enzymes appropriately and spreading his fat intake through the day, instead of allowing him to perhaps eat some low fat meals and then binge on a lot of fat at a particular meal. It's kind of hard to cover that completely with enzymes. Those are the kinds of things that might prevent this from happening again.

MR. BUSKER: Let's switch topics now to address our third objective and focus on malabsorption problems in CF patients. Dr. Schwarzenberg, describe a patient scenario for it us if you would.

DR. SCHWARZENBERG: A 14 year old girl with pancreatic insufficient cystic fibrosis was sent to me for evaluation and management of growth failure. She had good growth along the 25th percentile for weight, and the 50th percentile for height, until about 8 months ago when she stopped gaining weight. She has not gained weight for about six months.

Her appetite is good. She takes 2000 lipase units per kilo per meal, pancreatic enzymes, and she takes about half that with snacks and rarely misses a dose of enzyme.

She has three to four loose bowel movements each day and she perceives that she has more problems with bowel gas than do her peers. Her pulmonary status has been fairly stable, she has not yet had menarche.

On physical exam, she is a slender girl. On her abdominal exam she has no tenderness, masses, or distension of her abdomen. She is Tanner stage 3 for breasts and Tanner stage 4 for pubic hair.

MR. BUSKER: And your assessment of this child?

DR. SCHWARZENBERG: I think that the case suggests a child who is malnourished. She has delayed development and her growth has stalled over the last several months. It's possible that the three to four stools a day would represent malabsorption. And I think in any child with cystic fibrosis, we have to be concerned when growth is delayed and when we begin to see evidence in a child with good pancreatic enzyme intake of malabsorption.

MR. BUSKER: Based on those thoughts, what would your first steps be in her evaluation?

DR. SCHWARZENBERG: It is always important to get perspective analysis of what a patient is really eating. People tend to forget what they have eaten, even in the last 24 hours, when they are sitting in your clinic and you're trying to get an understanding of what they are eating.

So we often ask patients to perform a six-day diet diary to assess their intake. They take home a pad of paper and they fill it out with what they're eating as they're eating it, sometimes their parents have to help them, but it gives us a little better feel for how much someone is really taking in. There are limitations to this method but it can be effective in helping us understand how much people are eating.

I think that we have to consider cystic fibrosis related diabetes in any young CF patient who is losing weight, and if this young woman has not had a recent oral glucose tolerance test, she would get an oral glucose tolerance test in our clinic at this time to try to assess for diabetes.

We would certainly want to look at her micro nutrient status. Again, if she had not recently had vitamin A, E and D levels performed we would perform them in the clinic. And I think those would be laboratory studies that would be appropriate for this young woman at this time.

MR. BUSKER: Are there other GI considerations you might consider?

DR. SCHWARZENBERG: I think you would want to consider the possibility that this woman could have celiac disease, she could have a gastroenteritis that is somewhat persistent, like *Giardia*, for example, that could be contributing to her diarrhea. These are fairly common diseases that affect many people in the population. So a reasonable evaluation of chronic diarrhea would be appropriate in this young woman as we try to assess and improve her nutritional status.

MR. BUSKER: What are some of the things you might do to improve her nutrition?

DR. SCHWARZENBERG: There are several things that you could try in a person with cystic fibrosis to see if you can improve their weight gain. We would have a dietary consultation to see if we could increase the calories in the food she is eating now. And, of course, at any CF center there are dietitians who have a great deal of expertise in counseling families and patients about good nutritional care in CF.

This is a patient where I might give her a trial of metronidazole to treat small bowel overgrowth to see if that is contributing to her diarrhea, her excess bowel gas, and perhaps even to her weight loss.

I also might give a trial of proton pump inhibitor to see if by reducing acid in her stomach, I could perhaps alter the pH of her small intestine.

In the small intestine, the bicarb coming out of the pancreas in a normal pancreas is what keeps our intestine at a reasonably neutral pH, and allows our bile salts to remain in solution. In cystic fibrosis there is some evidence that because patients don't produce good levels of bicarbonate or any bicarbonate from the pancreas, that the acid coming out of the stomach tends to create a more acidic environment in the intestine. This allows the bile salts to fall out of solution and can reduce the formation of mixed micelles in the intestine.

Many investigators and clinicians have suggested that putting patients with cystic fibrosis on acid suppression might improve absorption of lipid that is being adequately digested but cannot be adequately absorbed because of the poor production of mixed micelles. So I think that is always worth a try in someone who is losing weight in cystic fibrosis.

MR. BUSKER: So let's say you take those therapeutic steps. Now if she returns in three months with no improvement in weight gain, what would your next course of action be?

DR. SCHWARZENBERG: Poor weight gain in cystic fibrosis, particularly if it continues on for month after month, is a risk factor for decline in pulmonary function. And I think that we are very aggressive about trying to maintain good weight gain in our patient population.

We would really move forward in a patient who was not regaining their weight and achieving a normal weight gain again to a therapy that would insure weight gain. This is a patient that I might give a trial of Periactin as an appetite stimulant. I think that at the same time I would be preparing this young girl and her family for the possibility that a gastrostomy tube may be necessary in order to improve her weight gain. I think that this gives the family a time to be considering the gastrostomy at the same time that they are giving a trial to an appetite stimulant. If she didn't see a fairly rapid improvement in weight with the appetite stimulant, we might give her a temporary NG tube, perhaps for five days, to see if she tolerated supplemental NG feedings at night, which would give us some confidence that if we place a gastrostomy tube that she would be able to tolerate gastrostomy tube feedings at night.

MR. BUSKER: One final question, Dr. Schwarzenberg, and this is about future directions in addressing nutritional challenges in CF, tell us what you would like to see?

DR. SCHWARZENBERG: I think nutrition in cystic fibrosis is a key issue in maintaining health and good lung function in this population of patients. And while I'm not sure what the future holds, I would like to see the establishment of better protocols for diagnosis and management of common cystic fibrosis conditions such as small bowel overgrowth and distal intestinal obstruction syndrome.

I think we have a great start with DIOS. I think that we have a tool that would allow us to establish criteria for treatment and protocols for treatment in the future.

I also think we're moving into an era where we are refining lipid absorption beyond just the use of

pancreatic enzymes. People are investigating non-pancreatic enzyme associated malabsorption of lipids with novel techniques and I think this will allow us to introduce new therapies that will allow cystic fibrosis patients to absorb more of the nutrients that they are already eating and hopefully reduce the need for gastrostomy tube use in the future.

Finally, I am very excited about the work that has been done in the CF mouse model, and that I hope will be extended to the CF pig model, looking at the pathophysiology of gastrointestinal disease in CF that again may give us new options for diagnosis and treatment.

MR. BUSKER: Dr. Sarah Jane Schwarzenberg from the United States Minnesota Amplatz Children's Hospital in Minneapolis, thank you for participating in this eCysticFibrosis Review Podcast.

DR. SCHWARZENBERG: You are very welcome.

MR. BUSKER: This podcast is presented in conjunction with eCysticFibrosis Review, a peer reviewed CME and CNE accredited literature review e-mailed 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, with a joint sponsorship of the Johns Hopkins University School of Medicine, and the Institute for Johns Hopkins Nursing.

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