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https://reachmd.com/programs/cme/cystic-fibrosis-essential-role-pancreatic-enzyme-replacement-therapy-patient-care/8437/

Released: 12/12/2016
Valid until: 12/11/2017
Time needed to complete: 15 minutes

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Cystic Fibrosis: The Essential Role of Pancreatic Enzyme Replacement Therapy in Patient Care

Narrator:
Welcome to CME on ReachMD. This segment, Cystic Fibrosis: The Essential Role of Pancreatic Enzyme Replacement Therapy in Patient Care, is provided by Prova Education. Your host is Dr. John Russell, who welcomes Miss Suzanne Michel, Clinical Assistant Professor of the Medical University of South Carolina in Charleston, South Carolina, and registered dietitian with over 40 years of experience caring for persons who have cystic fibrosis.

Dr. Russell receives consulting fees from Galderma and Speaker Bureau fees from Sanofi Pasteur. Professor Michel receives consulting fees from AbbVie.

This CME activity is supported by an independent educational grant from AbbVie.

Dr. Russell:
Approximately 85 to 90% of patients with cystic fibrosis suffer from pancreatic insufficiency since birth. This significantly complicates the disease and adversely affects both quality of life and life expectancy. Today, we're going to discuss supplemental enzymes and the role in the health of persons who have CF, the barriers to adherence with pancreatic enzyme replacement therapy and how pancreatic enzyme replacement therapy is dosed in patients with CF.

This is CME on ReachMD. I am your host, Dr. John Russell. My guest today is Professor Suzanne Michel from the Medical University of South Carolina.

Professor Michel, welcome to our show.

Professor Michel:
Thank you. It's a pleasure it be here, and I look forward to talking to you about pancreatic enzyme replacement therapy.

Dr. Russell:
Suzanne, to begin your discussion, could you explain why supplemental pancreatic enzymes are so important for our patients who have CF?

Professor Michel:
Certainly. So, exocrine pancreatic insufficiency, which is often referred to as EPI, in CF it’s the result of the thick mucous blocking the pancreatic ducts, so the enzymes that naturally go into the intestine when you and I eat cannot get there for persons who have CF, so they can't digest the food. And since they can't digest the food, they can't absorb the nutrients. So they don't absorb the calories and the nutrients from fat, carbohydrate and protein, also the fat-soluble vitamins A, D, E and K, and minerals are all lost if they don't take enzymes. If you don't take enzymes and you don't grow and gain weight, then you're going to be in poor health if you have CF.

Before we had the current kinds of enzymes we have, it was not unusual for patients to really die of malnutrition, but now with these enteric-coated enzymes, our goal is that they grow, optimally as if they didn't have CF. And there have actually been a number of studies that show optimal growth in the first 4 years of life means better health, better height and fewer infections up to 18 years of age.

Dr. Russell:
Wow.

Professor Michel:
So, it's so important to have optimal growth, and that's where the enzymes come in and play such an important role.

Dr. Russell:
So, Professor, are there weight goals for patients with cystic fibrosis?

Professor Michel:
Well, actually, there are. A few years ago the CF Foundation took a look at their registry data. They keep data on every patient who is seen at a CF center in the United States, and they look to see if there was any correlation between body weight as body mass index, BMI, and pulmonary function, and what they found was as the BMI percentile for children went up, it seemed that there was a better FEV1 percent. So, they aren't saying a better body weight causes better pulmonary function, but there's some type of correlation. So, the recommendation for BMI percentile for children who have CF is to be at the 50th percentile; for adults it's to be at a BMI of 22 for women as a minimum and 23 for men. So, what this shows is if you're at a BMI percentile as a child at the 50th percentile, when you graduate to be an adult, you will be at a BMI of 22 or 23, and so that's what all the CF centers strive for, to meet these BMI goals for patients who have CF.

Dr. Russell:
That's great. So, in talking about these supplemental enzymes, what types are currently available, and are there new forms of enzymes being developed right now?

Professor Michel:
So, the types of enzymes that are available now are all enteric-coated, and what that means is the enzymes come in a gelatin capsule, and the enzymes themselves are beads, and the beads are enteric-coated. The coating protects the enzyme as it moves through the acidic environment of the stomach, and so when the beads enter the intestine, the coating dissolves and the enzymes are then able to work on the food. So, those are the most common-used products.

There is a product that's available that is not enteric-coated, and that is often used by people that will crush it and mix it in a tube-feeding bag, and it sort of predigests the tube-feeding formula. Back in the day we only had the non-enteric-coated enzymes, and patients really suffered from malnutrition. And there was a really famous study. It's called the Boston-Toronto Study, but it's really Toronto versus the rest of the United States, and that showed that if patients got more calories into them and their weight was better, they had a better median age of survival, and that's when the rest of North America looked to Toronto and said, “Well, I guess we better start pushing nutrition because it will improve survival.”

Dr. Russell:
So, how would a clinician calculate the dosage throughout the life cycle for a patient who had CF?

Professor Michel:
So, you can calculate enzyme dosing in a number of ways. The most common way for cystic fibrosis, which is different from EPI for other reasons, is to do weight-based dosing, and that dosing is done based on patient's weight, and the maximum dose per meal is 2,500 lipase units per kilogram of body weight to a max of 10,000 lipase units per kilogram per day. And at the CF center, there's a dietitian at the CF center who can do these calculations in her sleep, and really in his or her sleep, and that's the major way that we dose enzymes.

The second way you can dose enzymes is based on gram of fat, and that recommendation is less than 4,000 lipase units per gram of fat. And when would you use that? You'd use that, let's say, if somebody is being tube fed and you know exactly how many grams of fat that that patient is getting in that tube feeding. Another way you might use gram dosing is if a patient says to me, “Every afternoon I come home and I drink a can of Ensure.” Well, I know exactly how many grams of fat are in a can of Ensure. You can do fat-based dosing for that. Also, parents have said to me, “You know, after school one day my child drinks a glass of milk one day, and then the next day he might eat three slices of pizza,” and I know that that's a huge difference in fat, so I might teach them how to do gram-based dosing based on differences in fat content of a snack. There are parents who really like to calculate the grams of fat in every meal. I don't encourage that. I think it's a tremendous amount of work, but there are parents who like to do that, and so we teach them how to do that if they're interested in that.

Dr. Russell:
So, it's like carb counting in folks with diabetes.

Professor Michel:
Yes, yes.

Dr. Russell:
So, to follow up on a couple of the points you talked about, one is, how would we give enzymes to a newborn with cystic fibrosis, and how would we give the same enzymes to someone who's being tube fed, as you mentioned?

Professor Michel:
That's a great question, because when you get a newborn, especially, let's say, a newborn who's in the neonatal intensive care unit because they have meconium ileus or another problem, we can actually give them enzyme. And what we do is you open the gelatin capsule -- and there might be very few beads in the lowest dose -- and you put it on the smallest, smallest, smallest amount of apple sauce, and you use a baby spoon, and you put it in the baby's mouth, and you sort of turn the spoon over and wipe it off and then feed them immediately. That's the way I teach parents how to give a newborn enzymes. Also, some parents like to wet their finger with the baby formula or breast milk and then put the beads on their finger and let the baby suck it off. Another way that some parents like to do it, and this is only with babies using a baby bottle and an artificial nipple, would be to put the beads on the outside of the nipple and let the baby suck it off. You don't want to put the enzymes in the bottle for a number of reasons. The baby formula would break down the enteric coating, and also, you'd have to make the nipple opening so big that the baby would choke. So, that's how you give enzymes to newborns, and it works.

I'd like to mention something, though, about dosing enzymes in newborns. It's really a challenge, and that's because newborns up to about 4 months of age are just eating constantly. Especially, breast-fed babies, they could be eating every 2 hours over a 24-hour period. So, how do you keep to that 10,000 lipase units? It's very hard. And there was just a paper that was published about a year ago that addressed it. It was mostly an editorial, what you do about infants, and those of us with a lot of experience in CF know that that baby is growing very rapidly, so we don't tell parents not to feed the baby. We don't tell parents to limit the enzymes. We might go over that 10,000 lipase units, but we know that baby is going to grow, and we're seeing those babies monthly, and they just grow into the dose within that month, and then you have to raise it again. And then by the time they are 4, 5 months old, they are eating less frequently and it's a non-issue.

Dr. Russell:
So, for the tube-feeding patients, is that just that total daily dose of fat, or how do you dose in those patients?

Professor Michel:
Tube feeding has become a big point of discussion among dietitians. What we always used to do was say, “When you start the tube feeding, you take enzyme. If you wake up in the middle of the night, you take enzyme, and if you wake up in the morning when the tube feeding is just finishing, you take enzyme.” And that's what we had been doing for years and years and years, but what's happened recently are two things. People are crushing the enzymes -- and you say you don't want to crush them because it breaks the enteric coating -- but they're crushing them and essentially predigesting the tube feeding by putting it in the tube-feeding bag, shaking it and letting it sit for 15 minutes.

The other development that is new in the past year is an enzyme cartridge, and it's a cartridge that you put in the tube-feeding line, and when you look into the cartridge, there are beads in the cartridge, but these beads are very different. These beads only have lipase, and the lipase is on the outside of the bead, so as the tube feeding runs through the cartridge, it's essentially being predigested. So, that is brand new, people are just using it, and we'll see how people do with that. But you are absolutely right; tube feeding is a challenge.

Dr. Russell:
So, there must be other challenges for adherence for patients who aren't newborns or not on tube feeds, so how do you deal with the various barriers to adherence in our other patients?

Professor Michel:
Well, I can list barriers to adherence. How to deal with them is the one that is the challenge.

Dr. Russell:
The million-dollar question, yes.

Professor Michel:
So, first of all, cost can be an issue, but cost shouldn't be an issue anymore because there are programs out there. Some are sponsored by the enzyme companies, some are sponsored by private organizations that can pick up the copay, so cost should not be an issue. Time can be a challenge, especially if you think about you're out, you're at a friend's house with your toddler, and you have to stop the toddler every time the toddler wants to eat something to give him enzyme. Time is an issue at school, at the schools that do not allow

kids to carry their enzyme. So, what do you have for lunch now at public schools or private schools? It really doesn't matter which. You have about 20 minutes for lunch.

Dr. Russell:
Yes.

Professor Michel:
So, they have to go to the nurse first, they have to get their enzyme, and then they have to go down and eat lunch, so time can be an issue. With that, also embarrassment.

Dr. Russell:
Sure. Kids want to feel like every other kid, right?

Professor Michel:
They don't want to have to be taking these pills every time they eat, and with the stigma of drugs in school, they can be teased for taking drugs. I mentioned grazing. Grazing is a huge problem. And what I mean by that is the person who just eats all day long. People forget to bring their enzymes. I'm always amazed when people travel 2, 3 hours to the CF center and they don't have their enzymes with them, and we have to get them enzymes, but, “Wait, you're going away for the whole day. Don't you have to take your enzymes with you?” And then a big problem is the desire to lose weight. If you don't take your enzymes, you could lose weight, and that takes the help of the dietitian, the social worker, maybe a psychologist, because if you don't take your enzymes, not only don't you lose weight, you can end up with all kinds of GI complications and malnutrition.

Dr. Russell:
So, speaking of GI complications, say I have an adult patient, their dose is maxed out, but they still have GI symptoms. Are there other things I should be thinking about on my differential? How should we handle that?

Professor Michel:
There are a lot of things to think about. And I have to say that when you talk about differential, in CF for us it's easy. A patient comes in; a newborn is diagnosed through newborn screening. We can do what's called a fecal elastase, and the patient is either pancreatic sufficient or pancreatic insufficient. But in a typical GI office, the patient comes in, and you have a huge differential that you have to work through.

Dr. Russell:
Sure.

Professor Michel:
And so it's so much harder. But in CF, just because you have cystic fibrosis doesn't mean you can't have other problems. Now, what can contribute to GI problems that are nonmedical, and then I'll give you some ideas about the medical ones. It could be outdated enzymes. You know, you have a stockpile of enzymes and they are just outdated. Enzymes are very sensitive to extremes of temperature. You put your bottle of enzymes in the glove compartment, and it was 90 degrees out. That denatures the enzyme, so they aren't any good. Excessive juice intake can cause diarrhea.

Dr. Russell:
Sure.

Professor Michel:
So, that I would look at, especially in children. We say so often, “Take your enzymes with meals and snacks,” and we might forget to say, “and beverages,” so the patient is not taking enzyme with a milkshake, a glass of whole milk; that can cause it. A child chewing the beads and cracking the coating, and as I said, that child grazing, and then that patient who has these huge differences in meals, so breakfast might be a bowl of cereal and lunch might be, as we say here in Philadelphia, a giant cheese steak. So there's a huge fat difference. So as a dietitian, we have to try and figure that out. Medical reasons are a challenge, also. So, the patient could be lactose intolerant; the patient could have celiac disease; a patient could have liver disease, small bowel bacterial overgrowth, Giardia. I mean, these are some of the medical reasons that can cause GI symptoms that are separate from enzymes.

Dr. Russell:
Right.

Professor Michel:
So, there are so many things you have to think of when a CF patient comes in and they are maxed out on their enzymes. You know they're adherent. It could be another issue.

Dr. Russell:
It might not be a short visit.

Professor Michel:
It might not be a short visit. And some of the longer visits involve some of the social issues, especially that teen would wants to lose weight or that chaotic household where there's no routine to anything or that child who has multiple caretakers, in the morning with the parents and then goes to grammy's house during the day and then goes somewhere else in the evening, and no one knows what they are doing in terms of enzymes.

Dr. Russell:
And that's why there are Centers of Excellence for cystic fibrosis to kind of pull all this together.

Professor Michel:
To pull all that together, because there is a complete team. And as a dietitian, there are so many times when I need to get a social worker in there with me to help me out, or if it's a behavioral issue, maybe the psychologist to help with some behavior modification with a young patient.

Dr. Russell:
So, before we close, is there anything you'd like to discuss that we haven't talked about, or is there anything you'd like to revisit before we wrap up today?

Professor Michel:
Well, you had some wonderful questions, and you've really covered so many of the important aspects of enzymes and CF. I just want to say, again, the importance of enzymes, because without optimal growth, you are not going to get optimal health, and you can't get optimal growth without taking enzymes correctly and eating that high-fat diet that you need to eat to meet your calorie needs. So, I think it's so important that those working with people who have CF make sure that they are getting what they need in terms of enzyme dosing.

Dr. Russell:
Well, thank you so much for being with us, Professor Michel. Thanks for joining us. I'm your host, Dr. John Russell, for ReachMD. Please visit us at ReachMD.com where you can be part of the knowledge. Thanks for listening.

Narrator:
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