



VOLUME 3 – ISSUE 8: TRANSCRIPT

Featured Cases: The Role of Exercise and Physical Activity in Optimizing Outcomes Among Patients with CF

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

- Describe exercise interventions suitable for prepubescent children with cystic fibrosis,
- Differentiate between appropriate exercise interventions for adolescent female and male patients with CF, and,
- Discuss the characteristics of exercise and physical activity interventions for adult patients with CF.

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. Wells will provide an age-appropriate review of exercises that are beneficial in the treatment of cystic fibrosis.

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Dr. Wells has indicated that he has no financial interests or relationships with a commercial entity.

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Next Month's Topic
Interventions to Improve Nutrition for patients with CF

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MR. BOB BUSKER: Welcome to this eCysticFibrosis Review podcast. eCysticFibrosis 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 February 2012 eCysticFibrosis Review newsletter. Our topic is the *Role of Exercise and Physical Activity in Optimizing CF Outcomes*. Our guest today is Dr. Greg Wells, from the Hospital for Sick Children at the University of Toronto.

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.

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

- Describe exercise interventions suitable for prepubescent children with cystic fibrosis,
- Differentiate between appropriate exercise interventions for adolescent female and male patients with CF, and,
- Discuss the characteristics of exercise and physical activity interventions for adult patients with CF.

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 February newsletter author. Dr. Greg Wells is an Associate

Scientist and Assistant Professor of Physiology and Experimental Medicine, Faculty of Kinesiology and Physical Education at the University of Toronto's Hospital for Sick Children.

Dr. Wells has disclosed that he has no financial interest or relationship with a commercial entity whose products or services are relevant to the content of his presentation. He has also indicated that his presentation today will not include references to unlabeled or unapproved uses of any drugs or products.

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

DR. WELLS: Thanks very much, Bob. I'm very happy to be here to do the podcast today.

MR. BUSKER: In your newsletter issue, you reviewed recent research on how physical activity may be able to improve outcomes in patients with cystic fibrosis. I'd like to get into that topic in further depth in today's program, to further explore just what clinicians can do to increase their patients' exercise frequency and capacity. So if you would, Dr. Wells, start us off with a patient description.

DR. WELLS: Our first patient is a young boy with cystic fibrosis, prepubescent, and he's a very good young athlete. It's interesting to look at children with cystic fibrosis because they are very different from adults with cystic fibrosis, and we have to consider children differently from adolescents and adults.

MR. BUSKER: As an exercise physiologist advising clinicians, what do you consider the key factors they need to be aware of when recommending exercise for children before puberty?

DR. WELLS: The first thing for clinicians to consider when they are working with children with CF is what we call peak height velocity or the onset of peak height velocity. That's the growth spurt that begins to initiate a number of changes in the physiology of the young person we're dealing with.

Interestingly, their physiology with regards to exercise changes considerably right in and around that time. Their lungs begin to grow, the heart begins to grow,

the limbs begin to lengthen, and as a result, their ability to exercise changes as well. A number of internal physiological things also change with regard to aerobic metabolism, anaerobic metabolism, and strength and conditioning, all of which can be taken advantage of if we're aware of the timing of the changes in these different elements. So it's very important to consider children before the onset of their growth spurt and then adolescents after the onset of that growth spurt.

Children can be given very specific things to do that can help them as individuals and also as patients with CF.

MR. BUSKER: The concept of stable growth rate — talk to us about that and how it applies here.

DR. WELLS: Children go through two significant growth spurts. The first is from 0 to 2 years of age, and then in and around puberty. In between those times, between the ages of two and approximately let's say 10 for girls, 11 for boys — and that's very individual as to when someone will enter into the second growth spurt obviously — there is a period of stable growth. The wonderful thing about that stable growth period is that boys and girls are very similar physiologically. So the exercise recommendations that you give to children can be consistent from boys to girls. And because they have a very stable growth rate, their ability to learn sports skills is excellent. So you can have children participating in sports, learning, improving, and feeling like they're having a successful experience all the way through that period from 2 right through to 10.

As soon as they enter into that second growth spurt, their bodies begin to change and their ability to do sports changes as well. But right through that period we can confidently give boys and girls very consistent, similar recommendations for what they should be doing in terms of exercise.

MR. BUSKER: For children at this age, exercise is really playing. Give us a general overview about the nature of children's play.

DR. WELLS: If you look at a group of children playing in a playground, you'll see that they adopt an interesting pattern. They'll do short sprints and then walk around and recover, or do a burst of climbing and then stop and rest. It's very different

from the way adults might exercise, where we might go for a run, or we might go to the gym and do some work on a treadmill, or we might go into the gym and do an hour of weights for example. Children love to play anaerobically. They love to sprint, they love to do explosive movements and then they will walk around and recover.

The wonderful thing about this is that it has a powerful positive effect on the human physiology of anaerobic metabolism, and also aerobic metabolism inside the muscle. It's the perfect recommendation for children with cystic fibrosis because it targets various different energy systems within the muscle and also the cardiovascular system, which delivers oxygen on the aerobic side of things.

So by being aware of the nature of children's play, we can make effective recommendations for children with cystic fibrosis.

MR. BUSKER: How does the nature of children's play specifically apply to children with CF?

DR. WELLS: Well we talked about the nature of children's play and how it activates both the aerobic and the anaerobic systems. It's critical because children can engage in play all day long, and habitual physical activity and the amount and intensity of that habitual physical activity is critical. Because researchers at the Hospital for Sick Children have identified that the more habitual physical activity that a child is involved in, the slower will be their rate of lung function decline.

It's not just the traditional thinking around exercise or training; it is actually the total volume of movement that a child does over the course of a day that seemed to have a protective effect for long-term lung function in children with CF.

MR. BUSKER: Talk to us about some specific exercise recommendations that can be given to pediatric patients with cystic fibrosis.

DR. WELLS: As clinicians, we should feel that it's safe to recommend physical activity, exercise, and training for children with mild to moderate lung disease. What I would like clinicians to consider is that we want to provide children with the opportunity to do as many different things as possible, to keep it interesting for them, to keep it changing, to continue to develop

them as young athletes who can do all activities. We're talking about walking, running, jumping, throwing, playing, and swimming.

The critical thing that we're looking for here is consistent participation. We want children to be involved in physical activities for at least 60 minutes a day. That might seem like a lot, but this is what we know is important for maintaining health in children.

And interestingly enough, because they're younger and their bodies are adapting a certain way to growth, we've also found that intensity has to be relatively high, as when children play in a playground, there's burst of activity followed by rest. We shouldn't be afraid of recommending intense activities like soccer or basketball to children with cystic fibrosis. The intensity is critical. And we shouldn't be afraid to shy away from anaerobic activities either, anaerobic activities being those where there's 10 to 45 seconds of really intense exercise followed by long periods of two to three minutes of rest.

Those types of activities are perfect for children because they mirror the nature of children's play. They can do a variety of things, all of which have a positive effect on their overall health and lung function.

MR. BUSKER: Very interesting, Dr. Wells. But let's look at it from the other side, at circumstances where exercise would NOT be recommended for these preadolescents.

DR. WELLS: There are two situations where I think we need to be careful when prescribing exercise to all patients with CF. In keeping it simple we'll be able to remember it for everybody.

The first time is during acute pulmonary exacerbations. When a patient is sick, sicker than normal and there's been a precipitous drop in FEV₁ at a specific moment in time, we do not want to be recommending intense activities during that time. At those moments we're looking for just maintenance of just some base level of movement.

The other time that we have to be very careful with patients with CF is during exercise in the heat. The sweat glands in children and patients with CF are different. Children with CF lose more sodium and chloride than healthy people do, so the potential for becoming dehydrated or not maintaining their

electrolyte balance when they're exercising in the heat is great. So those are the two moments where we have to be careful of, acute pulmonary exacerbations and also exercise in the heat.

MR. BUSKER: Thank you, Dr. Wells. Let's move up the age range now and look at exercise recommendations for adolescents.

DR. WELLS: The second case I would like to bring to your attention is a young girl, 15 years of age, with cystic fibrosis. Reasonable FEV₁, above 85 percent, still healthy and participating in lots of activities at her school.

MR. BUSKER: You've described an adolescent female. Aside from the obvious ones, are there major differences between adolescent females and males?

DR. WELLS: There are differences that we have to consider for male and female adolescents. Two reasons are most important. First of all, related to cystic fibrosis, we know that adolescent girls with cystic fibrosis are at increased risk of a precipitous decline in FEV₁. There's a critical period for adolescent girls in terms of maintenance of their health as patients with CF.

The other thing we have to consider is that boys and girls during their adolescent years begin to differentiate in terms of their internal physiology with regards to strength, anaerobic function, and aerobic function. For example, girls begin to plateau in terms of their VO₂ peak, their cardiopulmonary exercise testing results, in and around age 14, where boys continue to increase right through age 18. So it is important to consider boys and girls separately once they reach adolescence.

MR. BUSKER: Aerobic training for adolescent girls with CF — that would be a recommendation, wouldn't it?

DR. WELLS: It is a very important recommendation for girls with CF. Mainly because aerobic capacity begins to plateau around age 14 for girls. There are several benefits of aerobic training for children with CF. Because aerobic training or aerobic exercise like running or swimming, even sports like soccer, they push the physiology of the oxygen transport system. And that involves lungs, heart, blood, as well as muscle, so it has a very powerful effect on the entire oxygen transport pathway.

There also have been demonstrated powerful effects on increasing mucus clearance in children who participate in aerobic activities. In fact, you can increase daily mucus clearance by up to 30 percent. There is something that we need to be careful of, especially with adolescent girls with CF. We know that maintenance of body composition is critical, and we do want to maintain a certain percentage of body fat in patients with CF. When we do aerobic training, those percentages begin to decrease. So it's critical to support exercise recommendations for adolescents with CF with an increased consumption of calories across proteins, carbohydrates, and fats to be able to support that effectively.

MR. BUSKER: Doctor Wells, let me ask you to go a little deeper into the importance of nutrition for these adolescent female patients.

DR. WELLS: Exercise demands energy. The way that we get energy into our bodies to fuel exercise is obviously through nutrition. And of course maintaining proper body composition in patients with CF is critical to long-term health. We want to maintain lean body mass, and we also want to maintain the body fat percentage.

So I would like young athletes or young children, adolescents who are participating in an activity, especially children with CF, to have increased amounts of complex carbohydrates like whole wheat pasta or whole wheat bread after exercise to help refuel their muscles. I want them to be having protein to help their muscles and different body parts to rebuild themselves. But I also want them consuming good fats like avocado, coconut, or olive oil because those sorts of foods help the nervous system repair itself and provide a base level of energy to help support the increased levels of physical activity, as well as to maintain proper body composition in patients with CF.

MR. BUSKER: Thank you. Talk to us now about some of the actual exercises you would recommend for aerobic exercise training.

DR. WELLS: Aerobic exercise training is really interesting. Even though it's aerobic and we often think of that as sort of long runs or long bike rides, there are a number of different things that we can do. We can do aerobic-base type activities that are something like going for a hike, that's something like walking, that's a general level of activity in our lives,

walking to and from school, the light easy activities over an extended period of time. It has very powerful effects on the oxygen transport system and can be very helpful for children with CF.

We can then take it to a slightly more intense level where we do something called interval training, where someone might do a block of exercise, two to three minutes in length, and then a shorter period of rest in between. That might be something like running during a track practice where you do a couple of laps of the track and then walk one, or where we would go to swimming and do some laps, take a rest, do some laps take a rest. That would be considered interval training, slightly higher intensity and a little bit more rest integrated into those.

Examples of some activities of sports that can place an emphasis on aerobic capacity are swimming, soccer, cross country running, even things like yoga can be beneficial, because there is an elevated level of activity for an extended period of time. All of those can be very powerful and beneficial for children with CF, especially adolescent females.

MR. BUSKER: Let me ask you about other activities, particularly anaerobic activities? Give us your recommendations for those in these adolescent girls.

DR. WELLS: Anaerobic activities can be beneficial for girls with CF as well. Let's take an example of something like soccer or basketball or volleyball, very popular sports to be involved in. When girls are involved with anaerobic training, actually when boys are involved in anaerobic training, as well, believe it or not, there is an increased adherence to those training programs.

In a study that was done on anaerobic training in children with CF, the adherence rate during the study was 98 percent, that is almost unheard of. So children love being involved in those types of activities.

The other benefit of doing anaerobic type activities is that during the recovery period, let's say that you sprint for the ball in soccer, during the walking period afterwards, the aerobic system is what is responsible for recovery. So the entire oxygen transport system — blood, lungs, heart, muscle — are all being stressed during that time. It's a very comprehensive approach to training and because of the high adherence, I really like recommending these types of activities for adolescent females because their participation in

physical activity might be lower than boys' overall. It's a very useful tip for people to consider in this specific population.

MR. BUSKER: Let me side-bar a note to our listeners, that the study Dr. Wells referred to was described in the newsletter issue.

All right — Dr. Wells, summarize for us your key points about adolescent females and exercise.

DR. WELLS: For adolescent females with cystic fibrosis, we have to consider three critical things. First, aerobic training is very powerful and effective for this particular group. It improves and slows the rate of lung function decline in this population.

Second, we're looking to maintain increased amounts of attention to nutrition to support the increased physical activity in a healthy manner, and finally, incorporating some sort of anaerobic recommendations because that helps with adherence, which is critical for adolescent females with CF.

MR. BUSKER: We'll return in a moment with Dr. Greg Wells from the Hospital for Sick Children at the University of Toronto.

MR. BUSKER: Welcome back to our March 2012 eCysticFibrosis Review podcast. I'm Bob Busker, managing editor of the program. Our guest is Dr. Greg Wells, from the Faculty of Kinesiology and Physical Education at the University of Toronto's Hospital for Sick Children. Our topic is the Role of Exercise and Physical Activity in Optimizing CF Outcomes.

We've been discussing how the information in our February newsletter issue can be applied in the exam room. So far we've talked about prepubescent children and adolescent girls. Dr. Wells, you described some distinctions between adolescent girls and boys regarding CF and exercise. So let me ask you to focus now on male adolescents.

DR. WELLS: The next patient that I'd like to bring up is an adolescent boy. He has moderate FEV₁ of around 70 percent and a lower lean body mass than we would normally expect in a boy of 17 years of age.

MR. BUSKER: I think we're all going to assume that exercise is also important for these adolescent males with cystic fibrosis, so talk to us a little bit about that.

DR. WELLS: There's two pieces of the puzzle for adolescent males. The first one is maintenance of aerobic capacity.

Aerobic capacity, as we would measure with VO₂ peak or VO_{2max} during a cardiopulmonary exercise test will naturally continue to increase from 12 to 18 in boys just because of growth. But that doesn't mean that we can't train it as well, and we know that VO_{2max} is related to FEV₁ and to survival in patients with CF. So the higher we can get that VO_{2max}, according to some research that's been presented in the newsletter, the better that patient's outcome is going to be.

The second piece of the puzzle we can talk about, aerobic exercise in adolescent boys with CF, is the strength training piece, because boys naturally will gravitate towards that type of work because they feel like they like to increase their muscle mass, but this also has positive effects on FEV₁ for patients with CF.

MR. BUSKER: Let's take those puzzle pieces one at a time. The importance of aerobic capacity training...

DR. WELLS: Aerobic capacity training, or endurance training, may have positive effects on exercise tolerance, increasing aerobic capacity, increasing respiratory muscle endurance, and perhaps even more important, is an improvement in perceived health.

Many of these positive benefits persist long after exercise interventions have ended. So it's critical that we give children, adolescents, ideas about what they can do, help them implement it at least once, and then we can expect these benefits to last for an extended period of time.

And around exercise and aerobic exercise in particular, this is critical, because the higher we can get that aerobic capacity, the better we can expect outcome to be over the long-term, because VO_{2max} has been shown to be related to survival in patients with CF.

MR. BUSKER: Very good. Now the other piece: you noted that adolescent boys like to increase their muscle mass. Talk to us about how that can directly benefit their CF condition.

DR. WELLS: We have just completed a study at the Hospital for Sick Children that shows that upper body

strength and anaerobic capacity are significantly related to FEV₁, perhaps even more highly correlated than traditional cardiopulmonary exercise testing. This is critical for boys because boys in adolescents have natural increases in a hormone called testosterone, and testosterone helps us to layer on an increased muscle mass, which has also been shown to be related to positive health outcomes in patients with CF. If we can keep lean body mass up in this population, we can expect improvements and maintenance of health around lung function for an extended period of time.

MR. BUSKER: Dr. Wells, discuss the benefits of strength training for adolescent males a little bit more for us.

DR. WELLS: Strength training increases lean body mass, especially around muscle mass because when we get into the gym and we lift some weights that result in micro tears in the muscle tissue. This stimulates the body to build new proteins in the muscle tissue, new contractile proteins, and that causes hypertrophy or growth of the muscle tissue. This has positive benefits upon physiology, but importantly, especially for boys in this age group, it has benefits for their psychology, as well, in a number of different areas.

There's lots of very interesting research on the relationship between psychology and physiology, and we don't have time to get into that now, but it's an exploding area to be aware of.

The other real benefit we can see with strength training is that even though we get into the gym and let's say lift 12 repetitions of a weight, and that only takes us about a minute, the whole recovery minute is also being done aerobically inside the body. So even though we're doing strength training, we're targeting the aerobic system as well. So it's almost like fooling your patients into doing aerobic training by getting in and doing strength training, both of which have positive effects upon health and lung function in this population.

MR. BUSKER: So your specific exercise recommendations for young males with cystic fibrosis?

DR. WELLS: The exercise recommendations we might look at for an adolescent boy with cystic fibrosis would be to incorporate both aerobic endurance and

strength training into a weekly plan. For example, on Monday, Wednesday, Friday we might recommend cardiovascular exercise, like soccer, basketball, alternated with Tuesday, Thursday, and Saturday - going to the gym and lifting some weights to try to instill that hypertrophy response in the body. Those are movable and as long as we're alternating between aerobic and strength I think we can achieve everything we're looking for in this population.

Now when we're doing that it requires slightly different nutritional support. When we're doing aerobic endurance activities, the body uses a lot of muscle glycogen, the priority is on getting carbohydrates back into the system to refuel the muscles.

When we're doing strength training, the priority shifts towards protein, because we're looking to rebuild muscle tissue and to do that we need increased amounts of protein, fish, meat, dairy products, even things like tofu that you might be able to get an adolescent boy to eat. But it's a very different approach to support each of those two things. If we do that effectively, their bodies will respond really, really well to the training stimulus.

MR. BUSKER: Under what circumstances would you not recommend exercise for these patients?

DR. WELLS: For adolescent males with cystic fibrosis, again, there are two periods where we have to be careful in terms of recommending exercise. The first one is, as I mentioned before, acute pulmonary exacerbations. When patients are sick we default back to just low levels of habitual physical activities to keep them moving. Again, we need to be very careful with adolescent males because they may get into a sports situation, and when you're in a sports situation or playing with your friends, the activity may go on for many hours. We have to make sure that these young men are rehydrating effectively, probably using a sport drink to make sure that their electrolyte levels stay at a healthy zone.

Because patients with CF sweat out more electrolytes than healthy people do, they may be at risk of dehydration and hyponatremia. So having those sports drinks available when doing an extended period of exercise would be something to consider. Dehydration may be something young adolescent males may get into just because they love playing so much.

MR. BUSKER: Those same caveats would apply to adolescent females as well, correct?

DR. WELLS: They would. If we're aware for all patients with cystic fibrosis of the three items, acute pulmonary exacerbations, exercise in the heat, exercise for extended periods of time, if keep those three items in mind, we can have people with cystic fibrosis, children, adolescents, and adults, exercise safely.

MR. BUSKER: Thank you, Doctor Wells. Now we've talked about pediatric patients, we've talked about both female and male adolescents — what about your recommendations for adults with cystic fibrosis? Describe a typical adult patient for us.

DR. WELLS: The patient I'd like to bring up next is a male, 35 years of age, who still has relatively good lung function, 60 percent FEV₁s, so he's still relatively healthy.

Obviously, this is an age that we need to be very considerate of, because as patients with CF are managed well with nutrition and physical activity and with improved drug and pharmaceutical therapies, we can expect people with CF to live longer. So recommendations for this age group are really critical.

MR. BUSKER: Well, let's get directly into that then. What physical activity recommendations would you have for adults with cystic fibrosis?

DR. WELLS: The recommendations for adults revolve primarily around habitual physical activity. Habitual physical activity is positively associated with lung function, and patients with the lowest quartiles of habitual physical activity have the steepest rate of lung function decline. And habitual physical activity, because it's just part of your life and incorporated into every aspect of what you do, may be more effective than an exercise program.

As we become adults, we have to work, we have to manage multiple responsibilities, so incorporating exercise into life becomes challenging. Habitual physical activity becomes something that we have found is very effective for keeping movement integrated with people's lives, and that can be critical for adults with CF.

MR. BUSKER: And your specific exercise recommendations for these patients?

DR. WELLS: The nice thing about recommendations for adults is that it's important to be involved in as many different things as possible. We can look at the benefits of being involved in aerobic endurance type activities, like walking, running, cycling, swimming; all of those have well documented positive effects upon health.

We can also incorporate anaerobic type activities, interval training and games like beach volleyball or soccer, or any sort of sport that adults may choose to be involved with. Getting into the gym and doing strength training is also very important for this population. It's enjoyable, it can be fun, and we encourage people to do that as well.

The final piece of the puzzle which we'd like to add for this group, adults, which is different from children and adolescents, is the importance of flexibility for overall health and maintenance of injury resistance and avoidance and those sorts of things.

So the comprehensive approach is what we're looking for here: aerobic, anaerobic, strength, and flexibility. I call that the aggregate of 1 percent gains. If we can incorporate just a little bit of physical activity across a wide spectrum of exercises on a daily basis, we can just keep ourselves physically healthy over an extended period of time, and that's the magic piece is just variety and consistency.

MR. BUSKER: The concept of CONCURRENT exercise, talk with us about that, please.

DR. WELLS: Concurrent exercise is a new concept that comes out of high performance sport, where athletes in specific sports who used to do just one type of activity are now adopting a wide range of activities to improve their performance in their specific sport of interest.

We found that that has positive benefits as well for people across the spectrum of health levels from athletes down to healthy people, to people with a chronic disease like cystic fibrosis. Concurrent training simply means we want to try to incorporate as many different things as possible in a week plan.

For example, we might do some light cardiovascular activity on Monday morning, we might do yoga Monday evening, and then do a strength training session on Tuesday. Something like that could be cycled through the rest of the week, where activities are incorporated in a week plan. They're completely different, they target many different aspects of our physiology, but they are all done within a limited time frame. It keeps things interesting, it keeps people improving across a spectrum of their physiology and we think that it may be very powerful for people with chronic disease because it has benefits across the entire pathway in the body, from heart to lung to blood, right through the muscle tissue and maybe even down into the bone. So it's a powerful new concept that we're recommending people consider.

MR. BUSKER: What type of nutritional support is particularly important for adult CF patients?

DR. WELLS: When you are looking to incorporate physical activity and exercise into your life, nutritional support is critical. And in patients with CF it's even more important because we have to maintain body composition around lean body mass and also percentage of body fat.

So we're looking to maintain levels of carbohydrate intake to make sure that exercise is fueled properly, increasing levels of protein intake to make sure that when we do train our body to rebuild itself effectively, and also to increase the amount of fat intake in the form of good-quality fats. That will help improve the nervous system and also maintain base levels of energy to fuel all these activities and health. So nutrition is absolutely critical for patients with CF, especially those who are incorporating exercise into their lives.

MR. BUSKER: And the potential risks in recommending exercise for patients with CF?

DR. WELLS: We need to be aware of several risks. The first is to remember that the sweat glands of patients with CF are different from those of healthy people and that they excrete more sodium and chloride. So when patients with CF are exercising, we do want to recommend sport drinks that are high in electrolytes. This becomes even more important when patients begin to exercise in the heat; for example, going for a run outdoors in the summer, or doing prolonged activities. I would define anything

prolonged as being longer than an hour at a time. That may lead to hyponatremia and dehydration and we want to be very careful of that in patients with CF.

Now it is encouraging because patients with CF have pushed the limits of what humans are capable of. You have an example of Lisa Bentley who's a professional Iron Man triathlete, and she is doing exercise in the heat for prolonged periods of time in a healthy fashion, but her nutrition is absolutely perfect to be able to support that. So it is possible, we just have to do it very, very carefully.

The other group that I would like to bring to everyone's attention is patients with severe lung disease, and that would be defined as anyone an FEV₁ of less than 50 percent. Here we have to be extremely careful because patients run the risk of exercise-induced arterial hypoxemia or not being able to deliver enough oxygen to the blood.

So in patients with severe lung disease, we do want to be very careful. It is possible to participate in an activity, but we have to be careful when we're recommending it.

One paper suggests that there is potentially a risk of pneumothorax in this population, so again it is important to incorporate exercise on a case-by-case basis which the clinician can do individually with the patient would be something to consider here.

MR. BUSKER: Thank you for those cases, Doctor. I'd like to shift gears now and ask you to look to the future for us. What is the latest new research telling us about exercise for patients with cystic fibrosis?

DR. WELLS: I believe the future is exciting because we have the potential to incorporate exercise into the lives of people with cystic fibrosis, and to do so from a mechanically justified perspective. New research that's just come out demonstrates how the cystic fibrosis transmembrane regulator, the protein that's dysfunctional in CF, affects muscle tissue, and we also have really new interesting research that shows that in a seven year follow-up, FEV₁, lung function, and habitual physical activity are closely related, and the people with the highest levels of physical activity have a 50 percent slower rate of decline of lung function. This is exciting new research that we'll be publishing shortly.

The final piece I'd like to introduce is that we've also found that there is a very close relationship between anaerobic function as well as strength and FEV₁ in children with CF. This is in addition to the traditional perspective that cardiopulmonary exercise testing and VO_{2max} are closely related. So we may be able to provide new opportunities and new options for people with CF to incorporate exercise into their lives and to do so with the confidence that they're having an impact on their health over the long-term.

MR. BUSKER: Thank you, Doctor. Now to wrap things up, I'm going to ask you to very quickly review what we've discussed today. The exercise interventions for prepubescent children?

DR. WELLS: The exercise recommendations for prepubescent children revolve primarily around the nature of children's play. If we can get children to participate in activities that mimic their type of play, bursts of short activity and recovery, I think we've really helped them to incorporate an important aspect of exercise into their lives.

MR. BUSKER: Differentiating exercise recommendations between adolescent females and males?

DR. WELLS: For adolescents with CF, we would like the girls to participate in aerobic activities as a foundation and for boys to participate in strength activities as a foundation.

The reason for that is that aerobic function plateaus in adolescent females, so incorporating aerobic function will continue to target the oxygen transport system, and incorporating strength is critical for boys. It's something that boys like to do and it also has aerobic benefit. So it targets both positive changes in the muscle as well as in the oxygen transport system.

MR. BUSKER: And the specific recommendations for adult patients with CF?

DR. WELLS: The recommendations for adult patients with CF are to incorporate as much habitual physical activity into their life as possible. Habitual physical activity has been related to lung function, so it's critical that adult patients with CF do as many different things as they can as often as possible.

MR. BUSKER: Dr. Greg Wells from the Hospital for Sick Children at the University of Toronto, thank you for being part of this eCystic Fibrosis Review Podcast.

DR. WELLS: Thanks very much for having me on the show. I really enjoyed it.

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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