



TRANSCRIPT

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SEGMENT 1

DR. KRISTIN RIEKERT: Thank you so much for coming tonight to Portraits of Adherence. My name is Kristin Riekert. I'm co-director of the Johns Hopkins Research Center, and we will be having a very exciting evening tonight.

Our learning objectives have been scrolling through all evening, so I'm not going to take the time to read them right now. I want to disclose that I have consulted Gilead Sciences, Inc., to develop the CF R.I.S.E program. Today's session was supported by an educational grant from Gilead Sciences to Johns Hopkins, but Gilead was not involved in creating any of this content.

We're going to start with a couple of presentations to make sure everybody is oriented and fully trained up to listen to these vignettes, starting with Cindy George, Senior Director of patient engagement at the CF Foundation.

SEGMENT 2

CINDY GEORGE: Thank you, Kristin. Good evening, everyone. It's wonderful to be here with you tonight. I'm going to talk about what adherence is, setting the stage for the rest of tonight's program. In this brief section we'll define the current state of adherence to CF therapies and the impact of adherence to CF treatments on health outcomes. We want to start out by having a shared understanding of what adherence is, how we define it, because many

times different words or terms or definitions are associated with it.

We have chosen the World Health Organization's definition from its World Initiative on Adherence to Long-Term Therapies. This statement is specifically relevant to CF because it goes beyond just medication. It talks about the extent to which a person's behavior — taking medicines, following a diet, or executing lifestyle changes — corresponds with agreed recommendations. I want to point out that a key principle of the agreed definitions is having an active partnership with your patient's families and care providers. That's what you will hear about as we go along tonight.

As we have suspected, adherence is very important, and research is showing us that following a prescribed treatment regimen enables better health. Adherence is linked to better health outcomes and is associated with fewer pulmonary exacerbations, higher lung function, and lower health care costs. As science advances, adherence becomes even more critical, especially for young kids who may not have complications with CF and actually may never have it if they take their medications as prescribed.

The next few slides are taken from a study done by Dr. Quittner and colleagues that was published in *Chest* in 2014. They went through retrospective pharmacy claims data for over 3,000 patients with cystic fibrosis, which is about the largest sample size so far to date on CF

adherence research.

This illustrates the impact of the importance of adherence. This is looking at adherence as measured by medication possession ratio, or MPR, on the vertical axis; and MPR, which comes from pharmacy refill data on what medication is picked up.

What we're seeing is pretty significant. This shows adherence to the various pulmonary medications, and you can see a wide range of adherence for each medication. This is a composite of the individual drug MPRs. The white bar indicates the median, so it's about 50 percent; it's telling us that about half of prescribed medications for pulmonary meds are taken. It didn't matter whether the drugs, as you see in the middle, are inhaled antibiotics, or on the first column an oral pill; adherence still varied.

By looking at adherence by age, adherence fluctuated across all age groups, with a decline particularly with adolescents and young adults.

Now let's look at adherence and its impact on health outcomes. This is data from Dr. Eakin, who also shows composite MPR and an association with pulmonary exacerbations. You can see that the more courses of IV antibiotics an individual had, the lower their rate of adherence. This study also showed correlation between lung function and adherence — higher adherence correlated with higher lung function in this study, telling us that obviously adherence matters.

Back to Dr. Quittner's study, low adherence is now also associated with higher health care costs. This is measured by the composite MPR with the health care dollars. The blue bar is the low MPR, 50 percent and under; the red is moderate, 50 percent to 80 percent; and the green is high adherence, 80 percent to 100 percent. You can see that the lower prescription refill rate is associated with much higher health care costs.

Because this is such a critical issue for people with cystic fibrosis, the CF Foundation in its 2013 strategic plan included adherence as one of the objectives. This slide depicts the multipronged approach we are taking. Here is the community effort, bringing the community together to address this situation; patient and provider communication, to find suitable and accurate assessments through objective measures; and finally finding and testing an array of interventions to help to equip people with CF and their families to achieve optimal health and ultimately their life goals.

Perhaps some of you have heard that we originally called this program Patient Engagement, but more recently with the community's help, we've changed our name to Partnerships for Sustained Daily Care Program because it gets at the essence, that we're all working together over this topic.

As we know, CF is a very complicated disease with a complex therapy to manage day to day. We have been talking a lot with individuals with CF and their parents,

and these are just a handful of comments they've been making. You can see many reasons why people may have a difficult time sustaining the care day in and day out: stress, time, balancing other life activities; I think you'll see that tonight as we go along with the case studies. Thank you very much.

SEGMENT 3

DR. RIEKERT: Thank you, Cindy. Now I want to conceptualize nonadherence so as you're looking at the upcoming vignettes we're going to show soon, as you're listening to what's being said, you can think about, how can I conceptualize this, how do I know where to start. Adherence is incredibly complex for many, many reasons, and this is an oversimplification, but it does help you figure out what direction to head. If you know there are three roads, not a thousand, if there were a thousand roads you wouldn't know where to go.

Adherence typologies: There's unwitting, which is the patient. The care team all think the patient is doing what's supposed to be done, but maybe there is a misunderstanding somewhere. Maybe the patient is only doing it once a day but was supposed to do it twice a day. Or was I only supposed to do it until I felt better and then stop? It's not on purpose, it's just the patient didn't get it.

There's erratic adherence, in which the patient understands and agrees

and wants to do therapy but has trouble consistently maintaining a routine. Some folks are just not organized and not planners, so it's hard to make it happen every day.

Then there is "rationalized" nonadherence — this is in quotes because it doesn't mean it's necessarily rational; it means they've come up with a reason. These patients rationalize to themselves, deliberately alter or discontinue, and there are many reasons for that. I don't think it's working for me, I don't feel any better when I take it, I'm depressed, or I deserve the weekend off; all sorts of things can contribute to this.

Once you've picked the typology, you have to figure out what to do. You could do a thousand and one things, but when you're in the clinic and need to keep things going, the RUBBER mnemonic is a pretty useful way to go. It starts with reviewing the regimen. What does the patient say they're taking, what do they think they're supposed to do to make sure we're all on the same page of what's happening.

The next one is understanding. Does the patient understand why it is important to take a certain drug, what this drug is doing for them, why they are supposed to eat higher fat when their friends aren't — why, how, what.

Then there's beliefs — and this is a key one — what they believe. Do they think the medication works, do they have worries or concerns about side effects, implications for their

social life, stigma burden, what are goals and values and how does adherence fit in there?

Then you have more concrete barriers. Are there financial, personal, social, organizational issues that could be worked on? We educate on all these things, make sure there is no misunderstandings, and repeat. Adherence is never a one time and you're done fix; it's coming back to this issue, making sure old problems stay old problems, not current problems, and there are no new current problems.

When you put these typologies together and RUBBER together, this is how it plays out. If someone is unwittingly nonadherent, it's important to review the regimen and then make sure there's an understanding. Provide and review a written treatment plan so that they don't have to count on their memory, because none of us are good at remembering everything that happens in a visit. Have you provided the education, have you reviewed skills and techniques? Ask them to tell you back or show you the information you just provided, tell you what they're going to do with this so you know you made sure you've told them everything they need to know.

For rationalized nonadherence, we focus a lot on beliefs, what are their beliefs and concerns about therapies, will they become addicted? They may want to save these antibiotics until they need them and don't want to take them now when their lung function is fine.

Develop the discrepancy between what they say their values are — for example, I want to live to have children, I want to be healthy enough to have children — but the behavior doesn't match that. How can we link values and goals back to therapy, personalizing adherence and health feedback.

These are your lungs, this is your behavior, this is what's happening for you personally, and then using shared decision making. Putting this altogether, what path does the patient and family want to go on, and how does that fit with what is medically recommended?

For erratic adherence, focus on the barriers. Can we simplify or tailor the regimen, can we help shift how patients are doing it because of their life schedules, focusing on behavioral strategies, reinforcements. Those sticker charts are fabulous. Is someone in their life who can help them with access social support, help them set up and clean equipment, or just be with them to be supportive? This includes mental health support. If there is any depression or anxiety, making sure patients are linked into any resources.

RUBBER again is educate and repeat, and this is elicit, provide, elicit; tell me what you already know. Here's some more information that might be important. Tell me what I just told you so I know you understand. Then follow up at every clinic, every time. If we all have the resources we'd be doing this between visits, a quick text, hey,

how's it going, I know you were going to try to do X, Y, Z, I was thinking of you.

So tonight, as we're listening to the vignettes, consider what typology might be relevant to this particular person, and then what intervention you want to start with. And I want to point out, it's not uncommon for every patient to have a little bit of everything, but there is usually one that rises to the top and that one is where we probably need to start to be successful.

And with that, we now transition into our first vignette.

SEGMENT 4

(video playing)

JAMIE: I'm Jamie. I'm a senior in high school. I love playing softball. I've played it all my life. I pitch, I play first and I play second, and I have cystic fibrosis. Not a lot of people know it. I mean, my sister does, but she's my sister, she lives with me, otherwise I really don't like to talk about it. I don't want people to look at me differently saying that I'm that sick kid because I'm not. I'm just like everyone else.

I used to do my treatments all the time back when I was sick because I had to, but now I feel fine. I don't have to do my treatment two times a day with my nebulizer and clean it every time, and do my vest. That takes up so much time. I'm not even coughing up any gunk anymore, what's the point of doing it? I don't

have time for all of that now. I mean I have to go to school, I have to go to softball practice right after which is six days a week, and then I have to come home, do hours of homework, and all I want to do is go to sleep and I just don't have time to do treatments.

I don't think it's a big deal when I miss a treatment sometimes. I feel great, so I cough a little bit. I still keep up with everyone else. I take my pills, I mean I kind of have to. My mom sets them up every day on the table so she'll know if I don't take them, but I mean they're just pills, they're not that hard to take, even when I'm at school, but doing everything else, I mean, sure, if I get sicker I'll go back and do everything, but I'm feeling great right now, there is no reason that I have to take time out of my day to do it. And you know what else, it really bothers me that every time I do those treatments, it reminds me that I've got cystic fibrosis and that I am that sick kid. I hate that, I just don't want to be known as that.

So what's the point of driving myself crazy to fit in all those treatments when I'm feeling so good?

(end video)

DR. GREGORY SAWICKI: We'll get back to Jamie's case in a little bit, but how many people in the room know someone like Jamie or care for someone like Jamie? It looks like a vast majority.

I'm Greg Sawicki, a pediatric pulmonologist and the center director

at Boston Children's Hospital. The first part of my talk will be to talk about people like Jamie, who is an adolescent female living with cystic fibrosis and is thinking about why she expresses these challenges around adherence.

We're going to talk about the various types of barriers that an adolescent with CF may experience. We'll think about adolescent nonadherence as an individualized problem, taking what Dr. Riekert said is thinking about the individual barriers and problems and challenges that an individual may have related to taking their therapies, and then we'll describe ways to identify adherence barriers.

It's very well established that adolescence is a high-risk period in cystic fibrosis. This is data from the US National CF Registry, which shows the average lung function by age based on birth cohort, starting with the red line at the bottom, people who are born in the late '80s, moving up to people born in the last decade. Although lung function has improved at every age with every birth cohort, in the box you can see that the rate of decline of lung function has not changed during the adolescent years, ages 13 to 18, despite any advances in improvement and care. We can talk a lot about why that may be, but clearly adolescence is a high-risk period from a health outcome perspective, and it's also a high-risk period from a developmental perspective.

Adolescents with cystic fibrosis, and

other chronic diseases for that matter, have very common attitudes which are listed here on this slide. For them, cystic fibrosis might be a problem that their parents are taking care of. This is something they were born with, diagnosed when they were very young, and their parents have always done all their therapies and helped them with them since they were that young.

CF can be on the back burner for them. Symptoms could be a nuisance, but they minimize symptoms. Adolescents tend to minimize a lot of things including symptoms, and in fact, they may say that taking medications or completing their treatments does not result in their feeling any better or feeling any different. In fact, they may feel that some of their therapies are making things worse. Jamie in the video talked about how she felt worse sometimes when doing her nebulizers or using her vest. They didn't make her feel well.

The common thing we often hear is something to the effect of, if I skip my treatments I don't feel sick. Not doing something for a day or two or three, maybe nothing happens, and gosh, maybe that's a sign that I don't need to take these therapies. Similarly, I think we can't forget that there are people who will think if they're perfect with my meds they shouldn't have any problems, yet every time they come to clinic we decided to hospitalize them or do some more therapy despite their taking all these therapies. So we can't just always assume that it's a reaction around negative or not

taking therapies.

And as a result, adolescents can develop anger. That anger can be at the disease, it can be at their caregivers, it can be at their care team. I think when we think about adherence behaviors, we have to take all of these attitudes in mind.

For all people with CF, particularly adolescents, what factors influence adherence? Clearly there are individual factors — age, gender, literacy, what they understand about their disease and treatments, mental health concerns, behavioral issues, coping styles, and health beliefs, as was previously discussed.

But the family is equally important, the structure of the family, the socioeconomic position of the family, health insurance issues. The knowledge that the family has around disease, mental health concerns in family members and caregivers, the way a family copes with chronic disease, what their health beliefs are and their perceptions, the quality of relationships and their involvement in care.

We can't absolve the health care system from leading to problems. Access to care, continuity of care, communication between patients, providers, and care teams. Biases providers may have, how frequently people are seen in clinic, all can affect adherence. And then at large, the society and community in which an individual lives has a big impact. The school, the neighborhood, their job, their peer support, their social

support and the stigma they may feel, as Jamie introduced in the video, of not wanting to feel different from other kids that she is going to school with.

This is why adherence is a challenging problem. It's not just one thing that we can do or think about to address an issue, it can be a multifactorial issue.

I will highlight a few challenges that I think are most prominent in adolescence. The first is that of treatment burden and treatment complexity. I deliberately separated these two because they're not necessarily the same thing. By treatment *complexity*, I talk about how complicated it is to do some type of therapy, how long it takes, how many times you have to do it, how complicated it is to set up. Treatment *burden* is what an individual perceives about that therapy.

Many people may have to do only one treatment a day but perceive that as very burdensome; many others do seven treatments a day and perceive none of that as burdensome because they've managed to fit it into their lives. And I think we see a big spectrum in our patients and families around this interface between complexity and burden, and I think it's important for us to disentangle and think about these two concepts separately.

Treatment complexity in cystic fibrosis has been increasing. This was data from a national database about 10 years ago looking at three

years, 2003 to 2005, when there weren't many new CF medications available to patients. You can see that over the three years, regardless of age group — these three different bars represent three different age groups — the number of medications as defined by a treatment complexity score, increased during those three years. We at CF care centers are practicing more aggressive early intervention therapies and recommending many, many more therapies. That increase in complexity occurred not just for adults and not just for teenagers and not just for sick people and not just for those with better lung function, it was across the board.

We've looked at the time it takes for patients to complete a day's regimen. This is a study of adults with cystic fibrosis who were simply asked how long it takes for them to complete their therapies on a regular basis during one day. They estimated on average about two hours, or almost 108 minutes per day for therapies, most of it on nebulized care, airway clearance, and exercise. So thinking about it in the context of someone like Jamie who has to go to school and softball practice, and wants to play with friends and have homework and everything else that she needs to do with her life, two hours a day is a lot to ask her to carve out.

But it's not just how many therapies we ask our patients to do. Adolescents have a desire for greater independence. They have less parental supervision, a more erratic lifestyle in terms of sleep and

schedules. They are concerned about social acceptance, and that was clearly seen in the video. There's experimentation and risk-taking, which is a normal developmental process during adolescence, and a sense of invulnerability, which is also a developmentally normal milestone in adolescence. Often adolescents don't have long-term goals or long-term views, so challenges around daily care and daily therapies can be heightened because of that.

When you talk to adolescents about adherence, do identify barriers. They identify barriers similar to what Jamie has stated in the video: time pressures, lack of time, uncertainty in schedules. They display forgetfulness, whether accidental or purposeful. An awareness of their disease trajectory can sometimes cause some nonadherent behavior and be a barrier.

They may recognize the futility in therapy, in adhering to a therapeutic regimen saying, I'll get sick anyway, why should I bother, or they may avoid therapies in favor of other activities because they have a sense that they want to do other things, they know that their life is limited, they have cystic fibrosis, they have heard all these messages, and they're getting that kind of a picture in their heads.

Adolescents can have competing priorities. They need to balance time tradeoffs. They want to be normal, they have privacy concerns, they don't want to seem different, and they may not perceive the

consequences. This is something that has to be thought about and elucidated in conversation.

But when you ask them, adolescents will also tell you what may facilitate adherence. These include understanding and learning about the importance of their therapies, getting that education and accepting responsibility. Sometimes an adolescent may say they've never actually heard that message from their care team, and in fact, their care team may have been spending all their time educating parents about this but not talking to the adolescent directly.

Adolescents identify relationships with their CF care team as an important facilitator of adherence behaviors, and they understand that when the CF team is creative in problem solving it actually leads to better outcomes. That goes along with the concept of empowerment. Adolescents want to feel empowered, they want to have some control over what they're doing. They don't want to lose total control, they don't want to have a lack of a safety net. They'll say that they want their parents or caregivers available for help and be around, but they also want to make sure they are able to exert some independence. They identify self care skills through repeated practice as an important factor in improving adherence behaviors — I think establishing a structure or routine is something all of us can benefit from when it comes to routine activities — and adolescents have also identified structure as a facilitator of

adherence.

As we talk through some strategies over the next hour or so, we'll think about ways to address some of these challenges. We can think about addressing treatment complexity, about how to make interventions and therapies practical and fit into someone's lifestyle, identify ways to reduce treatment burden.

But more importantly we need to think about how to design interventions tailored to the appropriate developmental trajectory of an adolescent and think about their youth-derived goals. We need to incorporate clinician input, parent input, peer input, support system, and the adolescent's input themselves. We do want to foster a system in which we promote adult milestones through early initiation of these behaviors and repeated practice of these skills.

So we're going to continue with Jamie's case, with the second part of her story.

(video playing)

NURSE: Hi, Jamie, how are you doing today?

JAMIE: I'm doing good.

NURSE: I'm going to go over your after-visit summary, but I've been hearing great things.

JAMIE: I go to the clinic every third month with my mom. It takes away from going to practice and doing the

things I really want to do, but I guess it's something I still need to do.

NURSE: And I wanted to point out your BMI, because it's really good. So whatever you're doing, keep up the good work.

Let's go over your medications. You have quite a list here.

JAMIE: Now here's the part where she's going to ask me about doing my treatments. Okay, I admit it, I always get nervous, I just hate talking about this.

NURSE: Your oral medications, your tablets, your enzymes, how are you keeping track of those and keeping up with taking them on a daily basis?

JAMIE: They're usually set out for me breakfast and dinner, and I keep really good track of them. I take them all the time; I'm going really good with my pills.

JAMIE'S MOTHER: I do set them out in the morning for all day for her.

NURSE: I see that you have some nebulizer treatments. How are you doing with doing those on a daily basis with some airway clearance? That can be a challenge.

JAMIE: I do them most of the time. I try to do them as much as I can.

JAMIE'S MOTHER: Did you do them yesterday? I don't remember hearing the vest or the nebulizer?

JAMIE: I'm really healthy right now, I'm doing great, I'm just, I'm so busy

doing my sports and my homework, I just — I keep trying to tell them that I'm feeling good, I don't need to do my treatments. I don't have time to do that every day.

JAMIE'S MOTHER: And with my schedule, it's hard to keep up with making sure she's doing her treatments.

JAMIE: If I'm feeling good, do I really have to make time to keep doing them?

(end video)

SEGMENT 5

DR. SAWICKI: How many of us have had this conversation in a clinical setting? Oh, more than the five people that raised their hands, I'm sure. Sometimes I think we wish that there was a little bubble cloud over the family that we were talking to so we could find out what was in their inner workings. What we don't know here is what the clinician was thinking in her thoughts. And I think bringing that out into the open is an important step.

One of the challenges that comes up with adherence is exactly illustrated by this second vignette: how are we actually able to measure this, how can we quantify what is actually being done in the home? We're going to describe some challenges in measuring adherence to chronic therapies, identify some strategies to measure adherence, and talk a little bit about the advantages and disadvantages of these

methodologies.

Broadly speaking, we can think about measurement in a couple of different ways. One is self report — just asking the individual to tell us what they've been taking, that's what was done in the vignette. Another is clinician report, asking the clinician what they think has been taken and what the adherence level has been. We can look at pharmacy records, at things like the medication possession ratio, or the number of refills given, or the proportion of days covered; there are different metrics for that. Now we have technology that has electronic monitoring capability. For oral medication there is something called the MEMSCap. For nebulized devices or airway clearance devices like the vest, there are chips that download and record information, and other types of monitors on MDIs or other types of equipment.

Let's first talk about self and clinician report. That's the easiest and cheapest thing we can do. A very important study we all should know about from the UK in 2011 compared both patient report, clinician report, and objective adherence as measured through what's called an I-neb, which is an electronically monitored nebulizer that's available in the UK. On your left is the patient report vs objective adherence. The X axis is the objective adherence rates and the Y axis is the patient reported adherence rates. A red line would be perfect parity and agreement, and you can see that the vast majority of individuals have high self reported adherence but low objective

adherence. Over and over they're reporting a lot more adherence than what is actually being completed. Almost nobody has high objective adherence and low self report, so if they're saying they're doing everything, they are likely over-reporting. Similarly, if someone reports that they're not completing all their therapies, they're probably more accurate; it's less likely that you underreport such behavior.

But I think even more important, when we think about provider report vs objective adherence, this circle is someone who has an objective adherence of about 70 percent, but the clinician has rated their adherence as 10 percent. So this is person doing a lot of therapies but the clinician doesn't believe them, and that causes and immediate disconnect in their conversation.

Similarly, some people have almost no objective adherence, 10 percent, but the clinician believes their adherence is 70 percent. This person would have a conversation and might report doing all their therapies, when they're not, but the clinician believes they're doing all their therapies. Then the person can leave a clinic visit and say, I'm doing just fine, I fooled them, they don't know that I'm not doing anything. This is not a source of good communication.

We have to move away from self report and clinician report. This isn't a good metric when we're trying to identify truly nonadherent behavior or challenges. We can think about electronic monitoring. Its advantages

are continuous monitoring, which provides are long-term, real time data; it's more objective than self report. It can identify a large spectrum of issues, including under-dosing, delayed dosing, holiday periods, white coat adherence. What I mean there is, if someone is scheduled to come to clinic and does great the three days before clinic, you can see that and pick that up.

The disadvantages are, the devices can malfunction and recording may not happen; technology failure or lack of technology in the home such as wi-fi, for instance; cost; and privacy concerns. Many people just don't want every minute and everything in their day monitored, and I think that is a real concern that we have to think about.

Some studies have looked at electronic monitoring. The first was the I-neb in the UK, looking at adolescent adherence weekdays vs weekends, and school time vs vacation time. It's hard to tell in these graphs, but the average line basically shows that during weekdays adolescents were more adherent than during weekends, and during school time they were more adherent than during holiday time. Think that's important because when we think about strategies to improve adherent behavior, it's not an everyday approach either; it can depend on the time of day, the day of the week, summer vacation or winter vacation, or during school.

More recently there's been some interesting data on ivacaftor. We all think that ivacaftor is a medication

that everyone will be adherent to. It's an oral medication, it's easy, and it's one of these novel CFTR modulators which has great clinical benefit. This is data out of Cincinnati Children's Hospital, which looked at only 12 patients, comparing self report, MPR, and electronic monitors. Each bar represents one patient; the gray bar is self report. You can see that pretty much everybody reported 100 percent adherence to ivacaftor.

However, when they looked at MPR or prescription refills, there was fluctuation up and down. Some people were filling or overfilling, and others were not filling their prescriptions at all. The blue bar, which was the electronic monitor on a MEMSCap, which recorded every time the cap was opened to take the medication, the mean adherence was only 61 percent.

This shows the duration between ivacaftor doses and weekly adherence rate. As the study progressed, adherence went down and the inter-dose duration went up. People were taking longer and longer times, 18 hours or 20 hours in between doses, instead of twice a day. These were people who knew they were being monitored, yet their adherence rates went down during the study. This suggests a behavioral and a human nature challenge that we have to deal with.

Pharmacy records identify what medications a person has obtained, not what was prescribed, but they allow for evaluation of adherence over a longer period of time. Pharmacy records only measure

what a person has picked up, but don't tell us what's happened in the home once that medication has left the pharmacy, and it's always not always clear what has been prescribed. We, as clinicians, might sometimes overfill prescriptions; enzymes are a good example. We might write for a thousand enzymes for the month but they only need 600, so over time they're going to build up a stock so they won't fill a prescription. They're not nonadherent, they just don't need the prescription. Or we might write for a bid med, knowing that the copay is higher so we want to give people a two month supply with a one month prescription. We may do that, especially in a situation when someone has multiple medications, so it doesn't account for changing treatments over time.

Let's think about Jamie's adherence typology. Dr. Riekert presented three typologies — erratic, unwitting, and rationalized. On your iPads, please let us know what you think is the predominant typology in Jamie's case.

The crowd seems to coalesce around rationalized nonadherent behavior, and when I was reviewing Jamie's case I certainly felt the same way. She had periods of unwitting and erratic behavior, as well.

(One of the questioners asked us why is she not using a spacer in the video, and as a pulmonologist, that was the first thing I noticed, too, but that could very well be unwitting nonadherent behavior. Maybe no one's actually ever educated the

family that the spacer had to be used. I think that is definitely a reality in a real world situation.)

But in general, Jamie's behavior, as she explained it, is this rationalized behavior. She says to us, I feel well, so why do I need to take my meds. I'm too busy, I have school, I have softball, why should I take my meds and take time to do that. She has come up with her own views around why she doesn't need to take her medications. I feel fine, despite doing that. Look at me, I'm no sicker than any of my friends.

What can we do for someone like Jamie? There's a lot of things we could do and I'm not going to give you the answers in 30 seconds, but I think we need to go back to that RUBBER mnemonic and think about what we can do. We need to think about her beliefs and develop a mutual understanding of what her therapeutic goals for school and her relationships.

We need to discuss her concerns about therapy. She's expressed some concerns, and we need to understand what they are. We need to think about problem solving around those barriers and concerns. We also need to think about education and why these therapies work and make sure she understands why they are important and what the long-term ramifications are, and we need to engage in shared decision making. Having a conversation with her, her support team, her parents, her friends, and us as a care team, and come to some plan and then review, educate,

review, repeat, and consider all these things iteratively. Clearly we could do a lot more in addition to this, but this is the first place to start.

With that I'm going to turn our attention to a younger child, and we'll go on to our next video. Thank you.

SEGMENT 6

(video playing)

COLLIN'S MOTHER: This is Collin. He's almost 5 years old. He has cystic fibrosis and also eosinophilic esophagitis, which means eating has always been a struggle for him. Despite it all, his lung function is still very good.

COLLIN'S MOTHER: Collin, it's time for you to come eat your lunch, baby.

COLLIN: Five more minutes, okay?

COLLIN'S MOTHER: Collin, you've already had your five minutes, you need to come have your lunch now, let's go. Come on.

COLLIN'S MOTHER: Mealtimes have always been a big problem. His CF clinic has told us he's lacking in good nutrition and we constantly worry about his slow weight gain and how small he looks compared to other kids his age.

COLLIN: Can I eat for you, mommy?

COLLIN'S MOTHER: Collin's been good at self selecting out of foods that cause him pain or digestive

issues, so we feed him whatever he'll eat. Right now it's peanuts. He likes them in the shell and he likes to crack them. He'll sit here forever cracking the shells but eats very little of them.

Collin, I need you to take some bites of your food, okay?

(Collin shakes head "no.")

COLLIN'S MOTHER: Now come on, I need you to take one more bite.

We really tried everything we can think of to get him to eat, sticker charts, TV time, we've begged, we've pleaded, we've even started explaining that if he doesn't eat, he won't grow up to be a big boy, or worse, that he'll get sick. Collin's got a whole bag of tricks to avoid eating. He'll play with his food, he'll push it around on his plate, he'll try to get up from the table.

COLLIN: I'm done.

COLLIN'S MOTHER: He's too tired, he's too cold, he'll try anything to avoid having to eat. Collin's latest trick is taking a bite or two of food and then telling us he's full, or that if he eats any more he'll be sick.

COLLIN: I feel like I have to throw up.

COLLIN'S MOTHER: The only thing that seems to work now is bribing him.

Collin, I need you to take a sip of milk, then you can have your surprise, okay?

COLLIN: You mean that dinosaur?

COLLIN'S MOTHER: Yes, this dinosaur, so take a sip of your milk, please. Stop stalling, please take a sip of your milk. Thank you. Good job, buddy.

COLLIN: Can I go play?

COLLIN'S MOTHER: Now you can go play.

But we can't keep giving Collin new toys at every meal, and he starts school next year, we have to find another way.

(end video)

MARY MARCUS: Good evening, my name is Mary Marcus. I'm a registered dietitian and clinical nutritionist at the University of Wisconsin Pediatric Pulmonary and Cystic Fibrosis Center in Madison, Wisconsin, and I'm excited to be here tonight to talk to you about something that is near and dear to my heart, and that is feeding kids.

Feeding, or food and fluid, is fundamental to life, and psychosocial issues surrounding eating and drinking are critical to parenting and family life. In the case of children like Collin, eating and drinking are even more critical. My learning objectives for tonight are to identify the types of barriers children with CF may experience, recognize that each child has individualized reasons for nonadherence, and describe ways to identify children's adherence barriers.

Collin had multiple things going on. He has cystic fibrosis, but he also has eosinophilic esophagitis. That means he's going to be seeing multiple care providers, and maybe multiple care providers on multiple days and different times and visits. As care providers, we also may not have the time to provide continuity of interaction, and we may be providing unclear information or recommendations that may be perceived, at least, as conflicting. Families may also struggle with understanding what we've asked them to do, and they may or may not feel comfortable sharing with us the complications of trying to put all these recommendations together into a plan.

Also, concerning health literacy, we can't fall into the trap of assuming that a family that is more highly educated and perhaps living at a higher socioeconomic status has a higher health literacy than a family at a lower socioeconomic status who maybe doesn't have the same education level and advantages as other families. We've had situations where the parents weren't the ones providing most of the care; perhaps a nanny was providing a lot of the care, but the parent was the one coming to clinic. We also need to find out what are the child and family characteristics structure and function and the care-giving environments the child may be spending time in where they're being fed, maybe school, maybe daycare, maybe different parents' homes if families are divorced. We need to find out all the different environments in which a

child may be eating and growing and receiving treatments and therapies.

Also, cost and food security play a big role in this younger child population. In the United States, up to 25 percent of all children live in poverty and hunger, and up to 50 percent live in low income homes. This means many of the families we're caring for may not always have the money to purchase the food or the recommended vitamin supplements out of pocket and still have money for medicine. I've had families tell me they've had to choose between food and medicine, and that is a real concern with this population and with all of our families.

Parental stress and depression are also huge factors. Any time you have a child with special needs, particularly when there's a nutrition issue involved, parental anxiety and guilt can lead to less structured meals and snacks and increased grazing, more intrusive feeding practices, and greater acceptance of mealtime disruptions and negative behaviors, as we saw in the video with Collin.

A family's culture and beliefs about food and diet are also important to ascertain, how necessary they feel the nutritional interventions may be. A family may construct their own view of what adherence is when they're trying to factor in all the treatments and the nutritional therapies and so forth, and because we all have to eat to live, sometimes nutrition isn't viewed as a "real treatment."

The child's age can also play a role. Young children don't understand the long-term health implications of nonadherence, and they also may not be at the age of reason, like Collin. Food and eating are one thing, though, that young children know they can control. They can't control very much else in their life, but they can control what they eat, and that gives them power.

Nutrition nonadherence in young children is twice as likely in kids with CF than without CF. We often see nutrition nonadherence in young children with CF exactly as we saw in the Collin video: food refusal, stalling. He's eating peanuts, but why did he pick peanuts? It takes a long time to crack a peanut, it takes a long time to eat a bunch of peanuts, and then just play with your food, basically. He wanted to get down and leave the table, so he was trying to distract his mom from needing to eat. It's also about finding out what's distracting.

Negotiating — could he play for five more minutes before he came to the table. Young children will often have a fear of new food, so introducing new foods may take 10 to 20 times before a child will eat them. A child may be perceiving mixed messages, depending on how many care giving environments and how many people are feeding him and what all those family members and care providers may themselves feel about food and nutrition.

And then, of course, the autonomy and power of no. Rewarding Collin

for his behavior just further fueled his negative behaviors.

How do we find out what the adherence barriers might be for a young child with CF? We don't have the opportunity to call up the grocery store to find out when they last refilled their groceries. We don't have video cameras in people's homes seeing what's happening at mealtime as we did with Collin. So we have to do an interview. That's a lot of patient self report, which we know is not always very accurate, so we need to ask the question in lots of different ways and lots of different times over to try to get that picture in our minds of what's going on.

Ask who's responsible for meals? What happens at mealtime? What happens when the meal or snack is not eaten or the food is refused? What distractions might be present for both the child and the parent or caregiver — phone, tablet, TV, video games, other toys, siblings, other people around the table that may be distracting. I've had families tell me that they basically stopped talking at dinnertime because it was so important for the child with CF to eat that it totally changed the mealtime landscape. When you guys are all eating, you'd hear lots of discussion and chatter and eating and communication. Eating is so much more than just fueling yourself; it has such a social component. So to have to take that away to get a child to eat is pretty extreme.

How long meals and snacks take is also important to know. Problem behaviors are more likely to show up

in the second half of the meal, and the number of bites per minute is inversely correlated with the length of the meal. In other words, the longer you sit there doesn't necessarily mean the more food will be consumed.

Find out how enzymes, vitamins, and supplements are given and how often they're missed and what happens when they're missed, and also ask who is responsible for administering them. You'd be surprised at how young a child sometimes is responsible for taking their own enzymes and supplements at mealtimes.

To go along with this, of course, find out whether the family has any concerns about meals or snacks. Do they feel the meal and snack time are going all right or not? What does a typical meal and snack time look like, much is eaten at one time, and who decides what to eat and who decides how much is enough.

Using the RUBBER mnemonic again, I modified it slightly to apply it to nutrition. We'd ask what the family and child say they're doing and what's their understanding of why, how, and what they're doing for their nutrition care plan specifically. What beliefs do they have about nutrition? What worries or concerns do they have? And what are the family's goals and values? I can't tell you how many times I've heard, oh, but I was so skinny when I was little. Are there any personal, financial, social, or organization system barriers, and by that we mean the food security issue I mentioned earlier. Do any of

the family's beliefs or goals conflict with any cultural or religious concerns. And then, of course, clarify the treatment plan, correct any misunderstandings and answer questions. Repeating all this is important, because we know it takes multiple times of going over the plan.

(video playing)

NURSE: How are you doing today?
Hi, Collin.

COLLIN: Hi.

COLLIN'S MOTHER: Bribing him has become the only thing we can do to get him to eat. Maybe it's a mistake, because we can't afford to buy him new toys for every meal, but nothing else we've tried works, I really don't know what else to do.

NURSE: We're concerned that he's really at a higher risk nutritionally, so we wanted to spend a little bit of time talking about that.

COLLIN'S MOTHER: I know, this is something we worry about constantly, and we've tried everything it feels like, and I fight this battle three times a day every day trying to get him to eat, and we just don't seem to be making any progress.

NURSE: How about dad, is he involved at all at mealtime? Maybe that father/son bond would help. Is he in the picture?

COLLIN'S MOTHER: Yeah, dad's involved, and we really see this as a team effort, we're in this together to

really try to get him to eat. And we've tried everything.

This is a daily struggle, three meals a day every day, and it's very frustrating. Many times we end up letting him not eat so we can enjoy our meal. We know it's not right, but it's really hard on our family.

NURSE: But I think we are really at this point now really concerned that he's at a very high risk, and we may have to consider some other options.

COLLIN'S MOTHER: Well, what else can we do?

(end video)

MARY MARCUS: So following along the same pattern of the Jamie case review, with the Collin case review I'm going to ask you what do you think the dominant typology is? I agree all three typologies were present. When I looked at it I thought there was more rationalized than unwitting and erratic, but certainly all three were present.

Bearing that in mind and also keeping in mind the new Cystic Fibrosis Foundation Preschool Care Guidelines that will be coming out fairly soon, I would look at the family's goals and make sure they are on board and we're in agreement on that. I'd also discuss any beliefs and concerns about nutrition that the family may have.

I'd also provide some education on nutritional behavior therapy, along the lines of the work that Scott Powers has done, and why that's

necessary. I'd point out how rewarding Collin's eating behaviors with attention and toys is just making the situation worse, using the elicit, provide, elicit approach. Then I'd do some shared decision making, making sure there is consistency with parenting and consistency with the other providers who may be in the picture.

Finally, I would probably also begin to plant some seeds for potentially a future G-tube if all other methods of working at improving his nutritional status were not working. It's good to plant those seeds early and just present it as another tool we have that some families may choose, so that they can be thinking about it and there can be a shared positive decision for the family, so they don't feel they have no choice but they just have to do it.

(video playing)

SEGMENT 7

AMY: My name is Amy, I'll be 30 years old next month, and I've got cystic fibrosis, and lately it's been really hard. Maybe in some ways I was lucky. I didn't really get a lot of symptoms until I was in college, so when I was growing up I could pretty much be a normal kid. none of my friends had to know that I had CF. I still don't want anyone to know, but now I'm getting a lot of symptoms that I never had as a kid, so it's getting tough to hide.

I've had a really hard time holding down a job because of how often I

get sick and I have to go to the hospital. (on telephone) "Hey, Mr. Raval, yeah, it's Amy. No, listen, I'm really sorry but I'm not going to be able to make it in again today. No, I understand, but, yeah, I'm just really still feeling sick. All right, I'm sorry. Thank you."

I've got a few close friends, but I always feel different and like I'm an outsider left out of things. They're all in relationships, falling in love, they're all going to get married, have kids, start life with someone, (complimenting friend's ring) "it's beautiful," but I'm never going to have that, I'm always going to be alone.

FRIEND: If you would want to help me plan the engagement party —

AMY: "Yeah, sure." How is anyone going to love me or accept me when I have cystic fibrosis, I'm always sick or in the hospital. I've been to a few CF centers, they're always nagging at me, Amy, do this; Amy, you aren't doing that, like that vest. Cough up gunk for half an hour while getting pounded on, how's that even supposed to help? It's just embarrassing and gross.

I moved to Baltimore last month to take care of my aunt, but most of the time it's hard enough just taking care of myself. I honestly don't believe that there's anything that's going to help me.

(end video)

DR. RIEKERT: For those adult providers, how typical is Amy of

experiences you have where there's just not a lot of hope and not knowing where to start. So my goal for tonight is to talk about adult challenges, things that are unique to adults. Greg talked a lot about beliefs and all those things, and they still apply with adults, but some specific challenges come up once people move into adulthood. Our goals are to look at the various types of barriers, the individual reasons, and ways to identify those barriers specific for adults.

In qualitative interviews with adults, they can identify many barriers, as well as things that are helpful. This is important to get a perspective of what's challenging and how we can help. And much like teenagers, adults have the treatment burden and social demands that are high up on things that get in the way of doing therapies, and once they become adults, work demands come in as well. Forgetting absence of perceived benefit, so this medication's doing nothing for me, I'm just too darned tired to do anything, and feeling some stigma and embarrassment. Again, this is very similar to teenagers, and is still very common in adults.

But things that help: coming to CF clinics. Sometimes this is because I'm scared of what you're going to tell me, so I do better right before and maybe get my lungs cleaned out enough that that number won't be so scary, or maybe I found the visit motivating and supportive, so that's helped me get reenergized to keep doing therapies.

Supports: family members sitting with them, helping them out is key, and helping provide reminders when they feel better. Sometimes adults, because they're already so sick, there are more likely to feel better once they do therapies. They can tell when I've missed therapies for a week: I'm coughing more, I'm more congested, I don't have energy, and then if I can get the habit back, I start to feel a little better. That can be very reinforcing.

Burden: How easy is it for me to do this, how easy is it for me to fit this into my life, and can I make it a habit and a routine. That's challenging, so how do you do that?

Distraction and rewards: Is there something I can do while I'm doing my therapies to keep me entertained. We know the teenager was on her phone, and that may or may not be a good thing if it affects technique, but it at least gets them doing therapies. The same with adults; many folks will say the only time I let myself go on social media is when I'm doing therapies, that way at least I feel like I'm doing something, I'm distracted and it's a bit of a reward. And some adults will say I do my therapies because I'd feel too guilty if I didn't. So that's a challenge.

Health beliefs are important across all age groups, but particularly in adults because it's ultimately on them. A child might not feel it's important to do it, but mom does and so it happens; but once you're an adult and you're on your own and making these decisions,

understanding if the patient thinks it's important. If there is any inkling of why bother, that's probably a red flag.

All these are on one to ten scales, saying on a one to ten scale, how motivated are you to do your therapies every day?

For self efficacy it's a series of questions: when faced with certain challenges — you have work to do, you want to go out with your friends, you're feeling tired — how confident are you that despite those challenges you can get your therapies done. In each case you can see that those who rate importance, motivation, and self efficacy higher, are those who have higher adherence.

So these are very easy things that folks can integrate into clinical practice to get a good sense of where we need to start.

Employment is unique to adults. They're no longer in school, they don't get any accommodations, they're in the workforce. The good news is, most adults with CF do find jobs and are independent from their parents, and that's a fabulous thing. But CF does affect adherence. For example, Amy was having a hard time keeping down a job because she has to keep calling in sick, so CF can affect the career you choose. Sometimes you have to work part-time or stop working completely. Sometimes it's because you're too sick, but sometimes it's just because you're prioritizing doing your therapy and therefore will work part-time or

not work at all to fit this in.

Many adults do talk about either actual or fear of workplace discrimination. Because I'm out so often, I'm not going to get put on that account or have that opportunity because my boss is worried that I won't be there when it's time for the big meeting or the big project or the deadline, so I don't get opportunities. Or I can't go because they know I can't travel so I won't get a promotion to a job that has more travel. They feel that sometimes it is a hindrance.

Very few adults are comfortable doing therapies at work, and for most therapies it's not an issue; they do it before work, or after coming home from work. But enzymes, that middle dose of a three times a day treatment, might have to happen at work to be adherent, but you don't want to do it. You don't want to be the person with CF, the sick person, you want to be known as who you are and the type of work you do. It can be incredibly stressful balancing employment and CF care.

There's a lot of unique social aspects, as well, once you're in adulthood. When you're a child, your parent is there. When you're feeling sick they're there to take care of you. Once you get married, your spouse, hopefully, if you have a good one, is there to take care of you. And take care of you not just when you're sick, but just day in, day out, setting up the equipment, reminding you to do things, sitting with you, empathizing with how hard this is, doing the grocery shopping so you can get

therapies done, you know, all those sorts of things.

But when you're a young adult, you're in that transition period. You may be living alone or living with roommates. I had some fabulous roommates but I don't know how many would help me with IV therapies if I needed help, or would run to the grocery store for me, or help take care of me when I didn't feel well, or perhaps even notice if I didn't feel well on a particular day. That's a very vulnerable time where you lose some of the social supports.

And because you're sick and you have a lot of therapies to do, social functioning goes down. That's more the quality of life of social functioning, not that they don't have social skills anymore, but the quality of their social life goes down, and therefore satisfaction with life goes down. Like I just feel like all I do all day is take care of my CF. Am I taking care of my CF to live or living to just take care of my CF.

A desire to be normal is another challenge. We all want to be normal, some of us do a little better than others, but, I want to get married, I want to have kids, I want to advance my education, but I don't know if I can or will. All that it makes it very hard to plan and it can leave individuals feeling hopeless and questioning and wondering, and feeling sometimes alone and just very different. And that all can snowball and build up some resentment to CF. Then not wanting to do therapies just to avoid.

One way this plays out is in disclosure. When you're a kid, your parent sometimes has told everybody and you have no control. Everybody at school seems to know, but as you become an adult, you have more control over who knows. This was an interesting study that asked, who have you told, ranging from people who are very close to you, such as relatives, all the way over to just acquaintances. There's close friends, people you're dating, your bosses, supervisors, teachers, coworkers, neighbors, et cetera. The dark blue bars are all of them, I've told all of them, they all know; the very light blue bar is none of them. And what you can see is even with relatives, about 15 percent of people say not even all my relatives know I have CF. And as you move toward the 20s you're with roommates, and so on, so maybe close friends know, but coworkers and neighbors don't. I live by myself and I'm sick, who's going to notice, because they don't even know I have CF, they don't know what's wrong. So this is a challenge.

And that's just who you tell. We've done some research about relationships and CF that's going to be presented at a symposium tomorrow afternoon. We've shown that if you ask people how comfortable they are talking about CF with different groups and how comfortable they are doing therapies in front of them, people who are more comfortable have higher social support and higher self efficacy, both of which are linked to adherence.

So thinking about disclosure and

who have they told, how we can help them be more comfortable talking about CF, how we can help anticipate what challenges they might face in telling other people is important.

Hopefulness comes along, but there's something called fatalism, and Amy had a bit of the fatalism: am I ever going to get married, who would ever want to marry me. Around adherence that could manifest itself as what's the point; what will be, will be; my life is in God's hands, fate's hands, any higher power; it's not in my control so why bother doing my therapies. Or even if I do my therapies, I'm not going to get any better.

And this data shows that the higher fatalism scores are associated with lower adherence. So when you hear these kind of statements of what's the point, hopelessness, helplessness, you should start thinking perhaps this will affect their adherence.

Now we're going to see Amy with her care provider.

(video playing)

NURSE: So welcome to our clinic, Amy. When was the last time that you were seen in your old clinic?

AMY: My aunt made me go to this CF center here in Baltimore, and I hate these first time visits, same stupid questions all over again, same stupid lists of things for me to do. You need to be adherent, Amy, you need to be compliant. Really, like

any of that's going to make a difference?

NURSE: I'm a little bit concerned today, it looks like your PFTs have worsened since your last clinic visit at your old center. Now your old center put you on aztreonam for four weeks. Have you been able to take that therapy?

AMY: I've tried that, it does not work for me. It doesn't do anything. I told them this, too. It doesn't do anything for me.

I tried those inhaled antibiotics once, spent all that time learning how to set it all up, used it for about two minutes, then my throat started burning and I began coughing like crazy. Yeah, that's really helpful, makes me feel a lot better.

NURSE: Aztreonam is an important inhaled antibiotic and it's going to treat the bacteria that you have in your lungs. Typically we'd want you doing it for four weeks, is that a commitment that you can make?

AMY: Honestly, like I don't think so, I guess that takes up all my time, how about two weeks, can I try it for two weeks and see how I do?

NURSE: No, unfortunately, we need for you to do the prescribed therapy, which is a four week course. So we want to be on your side and get your lung function up, and that's how we know to do that.

AMY: None of it helps. She just doesn't get it. It's hard enough just getting through the day, how am I

going to do more treatments, get to work on time, or have a social life? Will doing one more medication really make a difference? Who wants to live that way?

The stuff takes up my whole life, all I do is spend time on nebulizers, and vests, and airway clearance, and then I end up in the hospital. Can I, I'd rather at least just have somewhat of a life until I die rather than spend my whole time doing stuff that doesn't make any difference.

(end video)

DR. RIEKERT: These are some sentiments we've probably heard from patients that we see. As we've done before, let's do our question of what do you perceive is the dominant typology for Amy — unwitting, erratic, or rationalized?

All right. Rationalized, that is also how I saw it. But throughout there's many aspects we want to consider. You see that all three of us picked rationalized, and I think that's interesting as we're coming up with our cases, because I think what we typically do as clinicians is try to provide a lot of education — if they just know what they need to do, they'll do it, so that unwitting. But when we think about these typologies we say maybe that's not the most predominant thing. So we need to think about what should we be doing first.

With Amy I think it's important. It's her first clinic visit with this care provider, and already she's ready to

go to battle and there's eye rolling, so it's going to be important to start that partnership. I personally would start a lot not even talking about CF, but rather who are you, what do you like to do, you came to Baltimore, have you made some friends; so you get to know her and so create a sense that there's a partnership in that she's understood. Because I think right now she feels like nobody gets me, they just want to tell me what to do, they all say the same thing, they tell the same thing to everybody, what's the point. So that's where I think she needs to be believed. We probably don't need to start with education about why an antibiotic is so important — not that she might not need to hear that — but it's probably not the place to start.

I would start with discrepancies, what are her beliefs and concerns about therapies. It sounds like she might have a lot of negative experiences with trying therapies, so there might be some concerns about starting again. Only after I understand her beliefs and her perceptions will I start with education, because then I can pinpoint the places where she's accurate and places where she might have some misperceptions.

The education has to be provided with empathy. It can't be, oh, you don't know what you're talking about, this is going to work for you. It has to be, I understand it's challenging, this doesn't work for everybody, but this is the best way we know to help you, and so how can we work together to get this done.

And I think for Amy, shared decision making is going to be key. Again, she just feels like people are piling stuff on her, and once she feels like I've made that decision, I'm choosing to do this, it becomes a lot easier to do that. So think about it, we're all oppositional 2 year olds, even if we're 30 years old. No one wants to be told what to do, but gosh, if it was my own idea, then it's brilliant.

We have to try to engage our families into thinking, not tricking them. I don't want it to sound like we're trying to trick them into wanting to do it, but we've come to this agreement and that is what they do want to do because it has been developed with them in mind and tailored for them.

For Amy I'm a little concerned about some depression. She has a lot of hopelessness statements — all my friends are getting married, what's the point, why do these therapies, I'm going to die anyway. So it would be important to screen her for depression and make some referrals for counseling and therapy. If she says she's willing to try this antibiotic, my job isn't done, because she also says how do I fit this all in.

It's not just convincing patients to do therapies, but also sitting with them and saying, okay, it's great you want to do this, now let's talk about what your life looks like and how we can make this the least intrusive possible. What could we adjust, what could we combine, what could we stop. What we're doing is to try to make it manageable and most likely

to be successful. Because we don't want Amy to feel she's failed again, because she might just move on to another care team, and it's not helpful to just keep moving care team to care team and not having any continuity.

That's all I wanted to say about Amy.

SEGMENT 8

DR. RIEKERT: This section is going to be a little more interactive, and we're going to engage the patient and family and try to create a dialog, even in this large group setting where we can't see you, through the tablet.

We're going to pose some questions to you, and you will use the Ask a Question function to give your best practices and your help. Because it's fine and dandy for me to sit here and give you what I think might work, but you all are experts as well and have come up with lots of fabulous things, and we need start sharing those ideas.

We're going to have a conversation here and highlight some of the most common themes. After that we're going to have a Q and A and do the post test. Then we have a video showing where all our vignettes ended up.

The learning objectives for this section are to talk about patient and family centered communication; recognize when we need additional support beyond the capacity of a care team in the clinic setting do;

describe characteristics of difficult conversations; and think about tips that can promote positive conversations between patients, and families, and care teams.

Our first discussion point is to talk about what makes it difficult to have conversations about adherence with our patients and families. Use the Ask a Question tab. It doesn't have to be a big paragraph, because I know that's a lot to type, but just some phrases about what makes it difficult to have that conversation.

So time is number one. There is just not enough time in clinic, and that is a problem we all have. There's not enough time, there's not enough staff. How do you panelists work on it in your clinics to build the time to establish relationships and come up with solutions?

DR. SAWICKI: I think it's a challenging thing. It looks like in our audience, time is coming up that much, and I think part of the challenge is the number of encounters you have with an individual or family, how often you see them and how you develop that relationship. I think fundamentally if you have a strong relationship with a family, time should be less of an issue, and that relationship can be built over a period of visits. I think if we can try as a care team to accomplish everything in one visit, it's never a recipe for success because we'll never have the time to do all this, despite our best intentions.

So sometimes it's bringing someone

in more frequently, establishing contact between visits. It doesn't have to be in the clinic; it can be with phone calls or some other type of communication. I think we, as a community, are not always as creative as we could be in doing that.

MARY MARCUS: I see a lot of things here about emotions and guilt, and families and team members feeling uncomfortable talking about these issues with care providers, and feeling like we're placing blame or judging patients. I want them to be as honest as possible, and there is no right or wrong answer. That's why I don't ask the patient are you taking your vitamins; I say how often are you missing your vitamins. Trying to set the stage that I know that I don't take my medicine and vitamins and stuff every day, so I don't expect anybody to. So just trying to lay that foundation and building the rapport, as Kristin was saying earlier, and finding out what's going on with them, what's new, trying to figure out what's motivating them and what else is going on that's taking their time and priorities.

DR. RIEKERT: I think defensiveness came up a lot. One of the greatest things I learned through my training and motivational interviewing is, when you're feeling that defensiveness, that resistance, it probably means you've pushed a little too far and you're asking for something they're not ready for. It's not that they don't necessarily want to have the conversation, but perhaps they're not ready to trust you to have that conversation with

them. So how can we build that trust and rapport, and I think that's time.

I think it's changing your mindset, as Mary said, of it's okay to be nonadherent, we all are; I think not even using the word adherence is sometimes — I think that's just the ugly word in the room.

So how do you get it in, how can I be helpful, what can I do now to be helpful to you, is a good way to start. Because if you're saying I want to help you, what can I do, that puts the patient in control and it puts them in the driver's seat so they can start thinking of the solutions. Because ultimately what I found is, once you open the conversation to possibilities, patients and families usually come up with a better solution than I have. So trying to engage and have that conversation, what can I do, what have you tried, what hasn't worked, so we don't go down that path.

CINDY GEORGE: I want to comment that I noticed some folks are talking about what discipline should do this. I was wondering if you're in clinics if you have a specific discipline or do you spread that evenly.

DR. SAWICKI: I think that's a great question, and it's one that is important to address as a care team, because if you do rely on one discipline I think that can sometimes paint a picture to the family that, well, when they're going to talk to the social worker, the nutritionist or the nurse, or the doctor, they're going to be getting one message, and they're going to be getting

another message from another person, and another message, and they won't know who to believe or who to talk to. Or they'll talk to the person who they feel partners with them, even if they're just listening, not listening and just going through their motions, and they don't want to interact with the people who actually challenge them or ask the complicated questions.

I think a lot of us try this already, but I think making a focus on the unified understanding and unified message. Some comments here have been, well we don't want to over-ask the same questions so the family feels like they're repeating themselves, but I think there is some power in repetition and power in knowing that there's a community of clinicians who are all on the same page. Especially for families, getting the parents and the adolescents to agree and discuss in a room, having that big discussion think is quite critical.

DR. RIEKERT: Someone brought up an important point here of open ended questions. I'm doing some audiotaped doctor/patient visits in a different chronic illness and we transcribed them, and it's fascinating how rarely health care providers ask an open ended question. And even when they do, they ruin it by then making it a multiple choice question; we jump right into the multiple choice because the patient doesn't answer right away. It's important to remember they're hearing this question and are actually thinking about their answer, so we need to pause and count to five to give them

time to answer. If we do that, I think they'll feel more heard and then more participatory and know that we care about what they're talking about.

Our next question is, knowing that this is a difficult conversation, how can we promote that positive conversation, about adherence and self care with patients and families, particularly in a collaborative way? We've come up with a few things but we'd like to hear your ideas as well.

DR. SAWICKI: Some people have already started answering this question with, it's very related, listening, acknowledging, opening the door, all those kinds of comments are coming through here. It's absolutely true what Kristin said about not just open ended questions, but listening to the responses. I think we all have a tendency sometimes to say if we don't get the answer we want or we are driving an agenda, we will start by saying an open ended question — or we think it's an open ended question. We actually don't listen to the answer, but pride ourselves in saying we've asked something open ended. And I think a challenge in the health care system right now is all the things that we need to do at every visit, especially for physicians, but also for the whole care team. Review medication list, review allergies, asking if the patient is in pain, see if they're smoking, fill out checklists, document on a charge everything in front of a computer and not actually look at the patient.

MARY MARCUS: I was just going to

say the same thing. Sometimes you've got your list in your mind of everything you've got to get through, and they give you an answer that then takes you way over there, and you try to get back over here, and needing to take that time to let that happen and let that conversation go in that other direction. Because you may actually get much better answers and come to a better care plan and better outcomes in the end by allowing that.

DR. RIEKERT: And knowing that the agenda you walked into the room with might not be the conversation that needs to be had at that point in time, so we need to be flexible and ready to go, as you're saying.

I think Greg makes a great point: we're sitting and filling out the computer, so we're completely missing the body language then. I think all three of our vignettes had some pretty clear body language in our clinic scenarios, because it's there if you see it. It's pretty easy to tell when someone is engaged and when someone is completely disengaged. And to keep that focus we need to make eye contact with them if we want them to make eye contact with us. We need to have open body language, look like we're settling in for a good conversation, not looking like we're trying to rush out the door because we're busy. Because looking like we're trying to rush out the door says we don't want to have a conversation with them. So we need to try and open that up.

CINDY GEORGE: Just to make a comment, I know we've heard from a

lot of teams and patients and families who talk about sometimes how the clinic is set up, and sometimes you have to have your back to folks. I love in those vignettes you could look across from each other, but sometimes you might have your computer situated in a clinic room where you might have your back to it or you're just not facing and making that eye contact. So sometimes you have to kind of take into account the context of the environment, as well.

DR. SAWICKI: I think the concept of relationships has come up a lot here, too, it's about developing that relationship. It's not just the one time encounter, it's the multiple encounters. We do that already as a system but I think we can probably try to do that even more, particularly when it comes to these types of issues.

DR. RIEKERT: When it comes to time, if someone had a decrease in their lung function or increase in symptoms, we would bring them back sooner. If they're having challenges with fitting in treatments, we should also bring them back sooner. We don't want to burn them by having lots of visits, but it also points to how important it is. If we can't bring them back, those follow-up contacts are key. It doesn't necessarily have to be lengthy, but we're thinking about you, wondering how it's going, we talked about you were going to try and do X, how is it going.

It doesn't necessarily have to be a lot, but you can't underestimate how

powerful that is that someone was thinking about me when I was not face to face with them, therefore, I'm important to them, and what I'm going through is important to them as well. So trying to be creative about different ways to stay in contact between clinic visits is going to be key.

With many of these cases we talked about possibly needing more support. In Collin's case, maybe he needed behavioral therapies for nutrition, and the parents needed some parent training. Clearly that is not being done in a clinic setting. Amy might need some depression counseling, that's not happening in clinic visit. What do you do when you know referrals are needed? That it's beyond what you can handle, how, what can be done?

We're getting a lot of refers, but who do you tend to refer to? We can either say like nutrition who I refer to, or mental health issues who I refer to, just so we have a sense. I find that some of the biggest challenges are finding the right folks to refer to. I know a lot, I'm trained as a psychologist, and a lot of psychologists are not necessarily knowledgeable about CF. They don't understand the burden, they don't understand the regimen, some of them can't fathom that you should eat high fat, high salt, and so they might give information counter. They look at the life expectancy and almost get giddy to talk about death and dying issues when that's not the issue.

MARY MARCUS: I like the comment

here somebody made about refer with patient buy-in, and I think that is key in order for it to be successful, looking at the patient as an equal member of their care team. Sometimes I envision it as the patient in the middle like a bull's eye. The patient's in the middle and then all the rest of us are little satellites around them. But it's the patient and the family who need to be driving the show and remembering that, and keeping that at the forefront of everything is what's going to help drive things forward.

DR. SAWICKI: But I also think the question was loaded in the where do you ask for help domain. I think even if we try to promote referrals, if we have a referral and system that works, which is sometimes a challenge as you mentioned, even in that situation, even with buy-in from the family, we can't absolve ourselves as a care team from continuing relationship building. We can't just say, oh, we can't handle it, we're going to find a referral out and hope they can handle it because it's not our issue any more. I think care teams can sometimes try to shift move their responsibility. I think these comments about making sure the family and the patients are buying into it is quite important.

DR. RIEKERT: I think buy-in is really important, and I think as soon as you bring up the word psychology, people say I'm not nuts. I would walk into the room and I couldn't even start the consult or something because it would be, I'm not crazy, and I'd have to say, yeah, I don't work with crazy people, I work with

people who have problems in life and could use somebody to talk to and strategize how to move forward. So I don't work with super crazy people; that's for psychiatrists — I work with normal people who just need some help talking some stuff through. I try to take away some of stigma around needing more help.

It's challenging to get them to any other specialist because it's just more time, more burden, and why do I have to take this on. I think mental health is particularly is challenging to get folks to, and recognizing that behavioral health is more than just depression and anxiety; it's also problems swallowing pills, problems with getting swabs or giving sputum samples. Behaviorists can help with those things.

MARY MARCUS: Even looking for referrals outside of the health care system. Somebody here is commenting about looking to faith based and other community resources, especially if your center serves a more rural community where it may not be reasonable to get those kinds of mental health services in particular on a regular basis, to find somebody local at the community level.

DR. RIEKERT: One person brought up a very important point: we're talking about referring to professionals, but there's also engaging families and neighbors, friends, other supports, who are not necessarily trained but can be very helpful.

It's beyond what I can do here in the

room, but can grandma get more involved, as an adult, can your parent be a little more helpful. I think that's key because those are more sustainable.

DR. SAWICKI: One more referral that people have mentioned here is peers. I think some peer to peer, even though we have infection control limitations, using some technologies in other ways. I know the CFF is working on that, and I know local centers have developed peer programs, and those can be quite powerful for certain individuals.

MARY MARCUS: Yes, and as to the comment about getting community and neighbors and so forth involved, there are websites like Lots of Helping Hands that some CF families use. The mother has initiated it and families can come in, neighbors can come in and help with laundry or doing dishes and whatever. That's just a way to help support that family in other ways.

SEGMENT 9

DR. RIEKERT: So now we're going to transition. Throughout the evening you have been submitting questions, and we're going to go to those now.

DR. SAWICKI: One of the questions was an important one when it comes to adolescents. Understanding adolescent cognitive development, what level of nonadherence should we as health care professionals actually accept? Where is there a balance between pushing for treatments and promoting

achievement and normal development and accepting some level of nonadherence. That's an excellent question and one need to grapple with because there may be a point at which we do accept some level of nonadherence. Do we term it nonadherence, or just a level of a treatment plan that may not be exactly what we as a clinician, wanted. I think the language we use with an individual, particularly an adolescent and their families, is critical there. Because it is part of that shared decision making and behavioral strategies and planning to say, what works for you, what are your barriers, let's make a plan. That may build up to an agreement that works on both ends.

I think we can accept some levels of nonadherence as long as we understand the context in which those are happening.

DR. RIEKERT: I think it has to be an informed nonadherence that everybody understands the pro and con of doing more or doing less. I'm a data geek, so I always want more data showing me the curve of how much is enough. For me, how much do I have to take to get the benefit. Some patients and families do this themselves, they say this is enough and I don't want to do more. As long as that's an informed decision that I'm doing the tradeoff of I'd like to trade a little bit of, I'll be a little symptomatic to not have to do this much more therapy so I can go out with my friends at night or I can get to my children's events. That is a thought-out decision that we have to agree and work with and move

forward with. I think that is key.

DR. SAWICKI: Another question was raised about how about CF patient support groups. I think that's a good question and we started alluding to that, and Cindy, I'll turn it to you. Are there efforts nationally around those kinds of things?

CINDY GEORGE: I think you're alluding to, Greg, a peer mentoring pilot we're going to offer across several centers to see how well it goes. We'll be using video technology and online, as well as other types of communication channels to connect adults with their mentors. We'll have a list of mentors who have been screened by your care centers to make sure that folks are okay for the mentoring, but we also want to stay out of the medical realm. We want to make sure that it's just about living with CF. We'll have a coordinator at the Foundation, so this is very much definitely something we have to start promoting. We hear that from folks, as well.

DR. RIEKERT: I think though there is something to think about. I think a lot of people are very interested in some peer mentoring, some peer support. There's also a not small group who have zero interest in it at all, because they've already known someone who's died of CF, and they don't want to watch somebody else get sicker who's been doing everything and they're not doing, so sometimes it can evoke some guilt. I think it has to be fairly well moderated to make sure that it stays as positive as possible. I think when

you have elderly folks who are not sick, soon everybody is dying and you see all your friends at the funeral because look who died now. You don't want it to become that; it's sad and depressing and not the support that we want it to be.

So it has to be well thought out and very positive, and I think short-term is probably good, and if they want to build longer term relationships outside of the structure it probably will then be up to individuals to create those relationships.

CINDY GEORGE: I'll just make one last comment, that was actually just built by adults with CF, parents, as well as care providers came together, psychologists, so that we definitely did put all those elements in because we know that that's very critical to the success of those programs so no harm comes.

I think there is another good question about focusing more on wellness rather than preventing deterioration, and I think that's key of how do we stay healthy. All people need to work to stay healthy, and so we normalize it, everybody has to get exercise, everybody has to eat healthy, yours has a slightly different bend to how that is, but everybody needs to do something to stay healthy.

And so when you put that swing on it, it's much more positive. I think folks who are comfortable using exercise for airway clearance versus a vest, I've heard lots of people say I'd rather run on the treadmill because healthy people run on a

treadmill, sick people do vest. That's how they've conceptualized it. So as long as the care team is willing to partner with that, you know, a half hour on a treadmill a day, I'm impressed.

I want to remind everyone that the group that helped us create this CME course also does eCysticFibrosis Review. These are experts who provide reviews of current topics and do podcasts that you may be interested in.

(video playing)

JAMIE: When I was younger I wanted nothing to do with my CF, I didn't want to know that I have CF, I didn't want other people to know, so I thought, oh, if I didn't do my treatments, didn't take my pills, I don't have it.

Because I didn't do my treatments regularly, didn't take the pills I was supposed to be taking, it made me go into the hospital for a week, and the antibiotics made me have an allergic reaction to them, and it just, it was, none of that is worth not doing treatments for me. There's just the treatments are bigger than anything else for me now. I've just, once I started doing my treatments, I've become like proud of the way I've made myself so healthy in like such a short span of time and the treatments really helped me like grow this confidence with my CF by doing my treatments. By doing, taking my pills, I've become so much stronger with my CF, and telling people about it and being like open about it.

COLLIN'S MOTHER: There is nothing harder for a parent than to not to be able to feed their child. It's a huge feeling of failure when it seems like such a basic thing to feed your child and not being able to do it. And so to make the decision to get a feeding tube almost feels like you're admitting defeat, we couldn't do this, we couldn't make it work, and when it's such a basic thing it can be a horrible feeling to not be able to get them to eat. But the feeding tube, it's a blessing because it allows you to give them that nourishment, and so for us the feeding tube was the right decision to make for Collin.

My hope for Collin is he's starting kindergarten next year so it's really, we're really working towards the goal of him not needing to be tube fed while he's in school because there is obviously a whole host of issues around having to do tube feed at school, and we want him to feel like he fits in with the rest of the kids as much as possible. He already has cystic fibrosis, so his life is already different, but we want to eliminate one other piece of what makes him different from other kids and let him eat with everybody else.

COLLIN: I have to eat my food so I get big and strong. Look how strong I am.

AMY: So when I was younger I was at a different CF center than I am now, and, you know, I wasn't compliant, they'd tell me do this, do that, and I didn't really do it. And that was kind of it, you know, every time I'd go into clinic it was, are you doing

this, no, well you should; okay, and, you know, you go home. And then when I ended up moving here I started with a new clinic and this is, started the same process and this time, no the doctors didn't just, well, you should, and let me go home and not do it, they were, well, why aren't you doing it; well, because I don't want to. But do you understand why we ask you to do this, do you understand the benefits of what we're asking of you, the benefits from the treatments; no, not really, I don't know what any of this does. Okay, well sit down, take a look at this CAT scan, take a look at, you know, this literature, let's talk about this, and it made me kind of open my eyes and say, oh, there's a purpose to this, like I really do have a little bit of control here, you know, maybe I can take back some of my life and be a little healthier, and so that's what I did. I started being healthier, I started being more active and social and I did end up meeting my husband now. We met and we dated and he was with me through when I did still have down periods, he was with me through that, and he's been a huge support keeping me motivated. He's a huge reason why I want to stay healthy is because I love our marriage, and we just bought a house, we have two little puppies, it's awesome. So I actually have all the things I never thought I would have. So it was a combination of motivation and support of someone who loved me, and knowing what to do, why to do it, and seeing the time doing it as valuable time spent. So that really changed everything, and I wouldn't be this happy if I was sick all the time, you

know. I mean, yeah, I take care of myself and it takes a lot to do that. It's a lot of work, but it's totally worth it.

(end video)