A Roundtable Discussion Addressing Patient-Centered Care and Quality of Life in Patients With Pulmonary Arterial Hypertension

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This session highlighted creative, multidisciplinary approaches to improving quality of life in patients with pulmonary arterial hypertension. Tiffani Brown shared her journey, from initial diagnosis to the present day, with the following panel of pulmonary hypertension experts: Cheryl Carmin, PhD; Emily Fedewa, NP, MSN; Jennica Johns, MD; Debbie Kittel, BSN, RN, CCRN; Laura Mudd, PharmD, BCCCP; Kristen-Allyson Ramones, MD, MBS; Allyson Rupp, MSW, LCSW, ACM-SW; and Scott Visovatti, MD.

The Initial Diagnosis: *"Everything Has Changed."*

Dr Visovatti (cardiologist): Tiffani, first of all, thank you so much for sharing your story. Could you give us an idea of what life was like before your diagnosis and how things changed leading up to it?

Ms Brown: I was living a normal life. I had started having symptoms about 3 to 4 years beforehand and was treated for asthma. The inhalers never helped, but they kept treating me for years. My symptoms started progressively getting worse in 2019, and it was in 2020 that I finally got my diagnosis. I went in for what I thought was just going to be another appointment with another doctor ordering more tests. So I thought this day was going to be just like any other. The doctor looked at me and told me that I had pulmonary arterial hypertension and that I was lucky to be alive. He told me he would be admitting me to start treatment. I was in denial. I had never heard of this disease, and so I didn't believe the diagnosis. I spent the weekend googling, which is the worst thing you can do. When it was time to be admitted to start treatment, I refused to go into the hospital. It took multiple phone calls with Debbie, my PH nurse, to get me to agree to come in for treatment because Google had me convinced that I was just going to die, so I didn't understand the point of treatment. So googling is definitely the worst thing I could've done because here I am, 4 years into treatment, and I'm still alive.

Ms Rupp (social worker): So many things to adjust to. Those of us who treat patients can't imagine what all of those things are. Dr Carmin devotes her time to this, so tell us a little bit about the kind of things people are adjusting to at this phase.

Dr Carmin (psychologist): What Tiffani described is typical: the deer in the headlights look: "Oh my God, what's going on? I'm dying." We really do try to dispel that. Everybody's reaction is different. Some people are anxious; some people are depressed. Most people are overwhelmed by worry. We acknowledge that feeling overwhelmed is absolutely appropriate for this situation, especially early on in this process. We also try to find out how resilient each person is by learning how they have coped with life events in the past. We help people to not focus on the "I can't." We focus on "I'm going to need to do this differently." The other piece is pointing out that this is not a hopeless situation and that you've got a team. That medical team is part of your journey; you're not doing this alone.

Initial Treatment Considerations: *"I am so overwhelmed."*

Dr Visovatti: Tiffani, you were in the clinic one day and admitted to the hospital soon after that. You're getting bits and pieces of information from your inpatient team, your outpatient team, and from the Internet. Must have felt like you were hit by a tidal wave in so many different ways. Were folks listening to you, or were you just being talked at.

Ms Brown: In my opinion, I was being talked at. I had shut down, so there was really no talking to me. I didn't feel like anyone could get through to me. I had to now grieve the life I thought I was going to live. It was the death of a life that I thought I would live.

Dr Visovatti: Debbie, you are often the point person for our patients once they are admitted. Well-meaning people are providing a lot of information. How do you ease people into having to make decisions?

Ms Kittel (nurse program manager): As overwhelming as it is for her, it's just as overwhelming for me. I know how much information you need to get to the patient so that they understand their disease, understand their options, and it's difficult when they're thrown into this. The whole world has changed, and you have to break it down in little pieces and parts. It's really a team approach, and the leader of the team is the patient; she has to be the one to say what she is willing to do. Are you willing to do IV therapy, are you willing to do SQ therapy, or is that just not an option for you? And then we have to do a social assessment as well. Are they good patients to put on parenteral therapies? Socially, do they have a clean place

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to mix their medicine? Do they have a mixing buddy? Do they have help? What is their support system? We involve palliative medicine early on to help us with side effect management. We involve our psychologist to help with some of the emotional support. There's so much information to get to patients, and you only have so much time, so it helps to ease them into this process. One of the things that we do with some patients who need parenteral therapy is to have the specialty pharmacy nurse go to their home to teach them the pumps and get some of the teaching done at home, before the admission. This is not always possible, but that would be the ideal scenario.

Starting Medications: *"The medications are as bad as the disease."*

Ms Rupp: So what treatment plan did you and your team choose, Tiffani?

Ms Brown: Starting out, I was on subcutaneous Remodulin once I left the hospital. I was not ready for that. Now, looking back, I wish we had the ability to have another patient come into the room and talk to me to prepare me for what's about to come because, even though we have our doctors and our nurses telling us what to expect, we need that other patient telling us what's to come. They could have prepared me for the site pain that I was about to endure, and I was on subcutaneous for about 2.5 years before I just couldn't do the site pain anymore. So I am a year and a half into intravenous, and I say all the time I wish I would've done IV from the start because my quality of life on IV is so much better.

Ms Rupp: Laura, how do you think about treatment plans to help with side effects?

Dr Mudd (pharmacist): We really try to take a proactive approach to side effect management. In that initial period, we typically up-titrate much more quickly than we do in the outpatient setting. We like to schedule side effect management, and we find that this can help avoid those unmanageable, severe side effects during the initial up-titration period. At our facility, we typically utilize scheduled acetaminophen and scheduled prochlorperazine. We find the combination is really helpful for headaches as well as nausea and vomiting. To avoid delays, we also enter orders for as-needed medications to help with refractory nausea, vomiting, or diarrhea. There's a lot of education on medication side effect management that is needed. We definitely don't want patients to be surprised by them, and we want them to have open communication with us about how they're feeling. Even if there's a slight change in a side effect, we can sometimes get ahead of it and make a change that might help avoid a really severe side effect. We have a standard up-titration protocol as well as a goal dose that we're trying to get to before discharge, but I think it's so important to make sure patients never feel that they have failed if they can't get to this dose right away. We know that patients respond to these medications differently, and so we really want the patient to be the leader and go at their own pace. So we can definitely make modifications to that titration plan. Also, sometimes our patients just need a titration vacation, as we call it. They just need a day to kind of reset and see where they are with side effects. We can then go back and see if we're able to continue with up-titration. Occasionally, a patient has severe, uncontrolled side effects, or comorbid conditions that limit our usual strategies. In these situations, we're very fortunate to have our palliative medicine colleagues, so I'll turn it over to Dr Johns.

Dr Johns (pulmonologist, palliative medicine specialist): When I meet a patient for the first time, I introduce myself and ask about their understanding of palliative medicine. It's like what Tiffani was saying: They're googling things, and then they see palliative, and they say, "Oh my goodness, I am dying." I spend a lot of time explaining what palliative really is. Sometimes I'll show a big circle and say this is palliative medicine; this is all the things that we can do. Then I'll show a tiny circle inside the big circle and explain that this is end-of-life care and hospice. Modern palliative medicine is really meeting patients early in their disease process to manage their symptoms. In PH, we're really trying to manage symptoms aggressively so they can optimize maximal therapeutic interventions for that patient. I also follow them in the clinic to help optimize outpatient symptoms, like diarrhea. Ideally, I prescribe medications that address more than one symptom at the same time. The other part of what we do is help them navigate all the things that they're going through.

Settling In: *"How do I get my life back?"*

Dr Visovatti: Tiffani, I hope that the benefits of you medications have outweighed the side effects.

Ms Brown: Yes. About a year into treatment, I was finally feeling well enough to get up and do things again, but with that came a whole new routine, and I had to figure out how to go about my daily life outside of the house. I needed to make sure I had the alarm set and that I was taking everything I was going to need with me. I had to pack that extra bag of supplies and the extra pump. You really have to plan ahead. You couldn't do anything just on a whim anymore. So you have to find your new routine, find your new normal. You also have to remember that, even though you're feeling good, it doesn't mean that your body can keep up with everything your mind wants to do. Sometimes you wake up the next day not feeling so great because you overdid it on your good day. So it's about learning how to take your good days and not overdo it so the next day can be a good day as well. Also, learning how to navigate the site pain was a big one for me because I was on subcutaneous. When I was hanging out with friends, I had to think, "Okay, where am I at with my site?" Do I need to take all the ice packs and all the creams? Can I be out of the house for an hour or two and be okay with my site? Once you've gotten your routine down, it does make life a lot easier.

Dr Visovatti: Tiffani, were you on supplemental oxygen at some point in your journey?

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Ms Brown: No, I got very lucky.

Dr Visovatti: We are so grateful that our delivery systems are getting easier and easier for patients in many ways. They are becoming less visible to the outside world. Emily, I wonder if we could ask you for your thoughts about Tiffani's situation up until now and discuss the concept of an "invisible disease."

Ms Fedewa (nurse practitioner): Yes, certainly. I think it's really important to recognize that pulmonary hypertension is often an invisible disease which, honestly, can be a double-edge sword. Patients receive this diagnosis, go home, and they look often like they did prior to the diagnosis, but they're likely not feeling their best physically or emotionally. They have this new full-time job that is managing their pulmonary hypertension. They are coordinating with specialty pharmacies; they're staying on top of refills (often from 2 to 3 or more pharmacies); they're contacting pharmacies when their medications aren't delivered; they're contacting their PH clinic when they have trouble contacting the pharmacies. I mean, the list goes on, and these are not one-and-done tasks, right? These are ongoing for life: taking medication 2 to 4 times a day, planning those medications around meals, planning to take their medications and their supplies anytime they leave their house, coordinating oxygen deliveries, making sure they have enough oxygen when they leave the house. We have patients that require 10 to 15 tanks just to attend a medical visit. Doing all of these things and processing all of these things when a patient still looks how they looked prior to their diagnosis can make it really hard to communicate this experience to their friends and families. Of course, not all patients go through this, and for those that do, there's an opportunity for education, but education takes time. So I think it's really important to recognize the challenge of having this invisible diagnosis, and on the other side, once somebody is wearing oxygen or sometimes parenteral therapy, that previously invisible disease is now visible, and that can bring into play a whole host of other psychosocial considerations. This can be especially hard for younger people in school and people in their 20s that are trying to go out. I think it's really important that we, as providers, recognize the complexities and challenges and that we acknowledge these things (out loud) to a patient. I think this acknowledgment goes a long way towards validating their experience.

Ms Kittel: I remember a time when Tiffani would never use a scooter in the grocery store. Now, in order to be able to do everything else she wants to do in a day's time, she'll use one of those little motorized carts to get through the grocery store.

Dr Carmin: I encourage patients to bring partners, support people, with them to their doctor's appointment because it is an invisible illness. It's important that people who are close to you understand what your needs are or what the course of the disease is. This is important information for them.

Dr Johns: I've had a patient ask me to tell their significant other that they can't do things because it's hard for them to tell their significant other themselves.

Ms Brown: Sometimes we can tell people, but they don't always hear it. We can say it over and over and over again, but they don't hear us.

Dr Visovatti: Kristen, you had your own personal journey with PAH. You were in the prime of life when you were diagnosed: You were accepted into medical school; you were thinking about your career, thinking about starting a family. Could you give us your perspective on how you struggled with life as you had envisioned it and then the life you were suddenly handed?

Dr Ramones (pediatric pulmonary fellow, patient): I was in the hospital, and my doctor was so good. She explained what pulmonary hypertension is, what the management would be, what the prognosis was. The sentence that really stuck with me and probably will sit with me for the rest of my life were the words, "You can never get pregnant," and that really just broke me. I just felt like my life was taken away at that moment. I got discharged, and I went to my bridal shower the next day. It was hard to adjust to that new life. Even now, I'm dealing with it. Although I have a lung transplant, I'm on immunosuppressants that are teratogenic. So PH has a lasting effect, even though I don't have PH anymore.

Dr Visovatti: You are devoting your life to pediatric pulmonary hypertension. We talked a lot about how adults have felt and what Tiffani has gone through. Could you give us an idea about how things are different for kids?

Dr Ramones: In pediatric PH, we focus on the challenges for patients in school and also caregivers' support. One of the most challenging things is how patients miss school because of so many appointments and hospitalizations. They're unable to participate in gym class, or they have to carry an oxygen tank when they're on the playground. We even start seeing them dissociate from other kids because other kids just don't understand what they're going through. As pediatricians first and foremost, we really want to protect the overall development of the child. We now have multidisciplinary clinics where patients see pediatric pulmonology, pediatric cardiology, social work, and nutrition. This really cuts down on the number of appointments that patients have to make. I think connecting families is really helpful, too, because you just don't see kids with PH that often. So they see someone like them, which helps a lot. Caregiver support is important in pediatrics because most of our patients can't take care of themselves. So relying on parents, guardians, caregivers and really focusing on their support is important. I went to a conference a couple of months ago, when they mentioned the concept of the forgotten patient. They were basically saying that the caregiver goes along the journey as the forgotten patient. When the patient has good days, the caregiver has good days. When the patient has a bad day, the caregiver has a bad day.

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Giving Back: *"How can I help others?"*

Ms Rupp: We all talk so readily, whole-heartedly, and passionately about support; it looks different for everyone. Everybody's support network is certainly important. Tiffani and Debbie are a prime example of how to make a good support network. Would you share with us about how support groups, peer support, and the support of your team has been really integral to your path?

Ms Brown: There was a pivotal point for me. I was just in a really bad place. I was in the hospital. I just had a heart catheterization, and Debbie walks into the room and says, "I want to talk to you about starting a support group, and I want you to co-lead it with me." I said, "Let's do it." It was like music to my ears because I had been thinking that I wished I had a support group to go to. I wished I could talk to other patients who understood PH. So we started. Our first meeting came around, and we were both so nervous and excited. It was a wonderful first meeting. The patients were just as excited as we were. We had a great turnout, and to this day, we still have an amazing group.

Ms Rupp: Can I ask: Reliving your PAH experience can be negative, right? It can be very traumatic to have to go through it again. How does it feel to share that on a regular basis in the support group?

Ms Brown: I feel like it helped heal a part of me that I didn't know needed healing. Your initial diagnosis is so raw and full of so many emotions, but you just get through it, and you keep going. Then you meet another patient who's going through it, and you're like, "Let me hold your hand. Let's get through this." As you're helping them or guiding them, it's healing a part of you that you didn't know was damaged. So the support group has just helped me immensely in a lot of different ways.

Ms Kittel: I learned from patients just how much misinformation you find on social media. I wanted a more supportive place for patients. During the first couple years, we did a lot of lectures or talks about pulmonary hypertension. Now I feel like our support group has shifted to not just learning about PH but also about supporting each other. For example, Tiffani was going through a really bad time. We had a support group meeting that week, and I secretly pulled everybody aside and asked if they would please write a little note of encouragement for Tiffani.

We put them into a "jar full of support" and gave it to her. She sat in bed and just cried and cried as she read through every note that everybody sent. We've given out multiple jars of support to different patients who are struggling. Our biggest event of the year is a low sodium Thanksgiving dinner. Tiffani and I cook Thanksgiving dinner for all the families in our support group.

Ms Rupp: I love what you've done with your group. We want to make sure that people are supported and informed, so we've put together a list of resources that people can explore further. We list things you can do as a community to rally people around pulmonary hypertension awareness, diagnosis, management, and support.

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