PULMONARY HYPERTENSION ROUNDTABLE

Transitions in Pulmonary Hypertension

In January 2022, Claire Parker, RN, MSN, CPNP-AC, Pediatric Nurse Practitioner at UCSF Benioff Children's Hospitals, San Francisco, California; Nancy Bair, RN, MSN, APRN, CNS-BC, Pulmonary Hypertension Advanced Practice Provider at the Respiratory Institute, Cleveland Clinic, Cleveland, Ohio; Neal Chaisson, MD, Director of the Critical Care Medicine Fellowship at the Respiratory Institute, Cleveland Clinic, Cleveland, Ohio; Liza Bonpin, San Francisco, California; and Maddie Bonpin, San Francisco, California, gathered to discuss the lifetime transitions in pulmonary hypertension care.

Claire Parker: Hello. My name is Claire Parker. I'm a nurse practitioner with UCSF Pediatric Pulmonary Hypertension. Today, we're here to discuss some transitions throughout the lifespan of a pulmonary hypertension patient. We've got some wonderful representatives from various PH teams, including a parent and patient. Liza and Maddie, do you want to go ahead and introduce yourself? Maybe, Maddie, you can tell us how old you are and what you've been up to?

Maddie Bonpin: Hi, I'm Maddie I'm 16 years old. I have PH, and I've had it for my entire life.

Liza Bonpin: Hi, I'm Maddie's mom, Liza. I'm Maddie's super fan since she was born. I can't believe she's almost going to be going to college! Nice to be with you all and talk about possible transitions in the near future.

Neal Chaisson: My name is Neal Chaisson. I'm an adult pulmonologist and PH provider. I often deal with adolescent transitions because at the Cleveland Clinic, each provider has a specific subset of PH that we specialize in. The primary patient population that I see are patients with congenital heart disease and associated pulmonary hypertension. Obviously, many of these folks transition up through the pediatric cardiology pathway and into our clinic.

Nancy Bair: I'm Nancy Bair. I'm the pulmonary hypertension Clinical Nurse Specialist at the Cleveland Clinic with Dr Chaisson and 8 other physicians. I've been with the group in PH for about 18 years now.

Claire: Wonderful. We'll start with a question for Liza and Maddie. We would like to know, from your perspective, what is exciting about your transition to adult pulmonary hypertension care and maybe what makes you a little bit nervous or what is unknown about that transition for you?

Maddie: I don't know. I haven't really thought about it because it's not really something I've thought about with the transition to adult care. I don't really have anything that I'm excited or nervous about.

Liza: Are you excited about almost college?

Maddie: Oh, yes, that. I know I'll be in college in a year and a half.

Liza: That's part of the transition, I think.

Maddie: Yes, it is. I'm excited about college because I'm a junior in high school now. I'll be applying soon.

Claire: Maddie, what do you think will be different for you when you come to visits with us versus visits with an adult provider?

Maddie: Probably the same thing with different doctors.

Claire: Are your parents going to continue to come with you to clinic visits?

Liza: It'll probably just be you, I'm thinking. I'm more than happy to accompany you maybe.

Claire: Liza, have you thought about what it will look like with the transition to adult care and more independence for Maddie?

Liza: A little bit. We've been training with getting to know your meds and knowing what you take and not yet how to order the meds. Just those nitty-gritty details. Then just recently talking about what happens when you're in college, if you're living away from home needing to remember to eat right and take your meds. She doesn't really cook or have any interest in cooking. I know she knows how to buy food, as she said. Having a stable job, which she does have a stable job right now, while at school with income to buy your food if you're not going to cook it. Not everyone cooks. That's okay, I told her. Being financially independent as well, which is I think really important. Just knowing how to take care of herself to face herself. That's still a work in progress, but those are all hopes and concerns that she can be independent and know also when to ask for help and who to ask for help with. It's exciting. She's come a long way since 16 years ago. We're looking forward to it. Obviously, start planning now, start practicing now.

Claire: It's been a long journey with your pediatric providers. It's going to be a shift, and it's going to be different. I can tell you from a pediatric provider perspective, we're so excited for when we're able to transition some of our patients to the adult group, and it almost feels like we're bringing you towards graduation. We are both excited and nervous for you to be more independent.

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Sometimes it's a little hard even for your providers to deal with that transition knowing that we won't be involved in your medical care moving forward. Neal, what challenges do you see when young adults transition into your practice?

Neal: I think back to 2 recent experiences, and I think Nancy has worked with me on both of these patients. One patient came of age to transition and was encouraged by her mom to become more independent, but was very scared to leave the *nest*, if you will. This wasn't a bad thing, but she emphasized to me the need to be patient and help her make the transition from dependence on her parents to independence on her terms. Many pediatric pulmonary hypertension patients have travelled this journey shoulder to shoulder with their parents, especially when they're on IV therapies. So navigating things can be a real challenge. We had to really sit down with my patient and establish goals together. Sometimes they worked, and sometimes we had to regroup and provide reassurance and another shot at boosting her confidence.

On the flip side, I had another patient with idiopathic pulmonary arterial hypertension since age 4 who transitioned easily. This individual went to college and insisted on being so independent that they just decided the medications weren't necessary. This obviously created some issues for him. Regrouping and helping his parents to rebuild trust in their son was a challenge. I think that the smoothest transitions occur when we help families, parents and patients alike, to anticipate and discuss what a transition looks like. What are the pitfalls of being too dependent or too independent? Questions like, "How do I get a new provider if I move? How do I choose the right insurance once I leave home? How do I interact with the specialty pharmacy?" These are all important topics and frankly, most adolescents have very limited knowledge of this side of patient life.

Claire: Neal, do you have a social worker or someone that helps with that transition in insurance?

Neal: We have a full-time pulmonary hypertension coordinator, Kasi Timmerman. Her job is to help us with navigating many of these issues, including insurance prior authorizations, obtaining patient copay assistance or grant funding, working with the specialty pharmacies to make sure that patients have as many options as possible etc. She is fabulous, especially given the current challenges we are facing with a lack of funding for patient copay assistance.

Claire: Yes. It's challenging on our end as well. We have a wonderful social worker and nurse coordinator that helps prepare for all of these transitions. Sometimes as prepared as we can be, we still face obstacles. Nancy, do you have any thoughts on challenges and excitement around transition in general?

Nancy: I think the social work issue is definitely an issue for the adult patients, and we do have access to a social worker group that we can find through Epic. They're very good about contacting the patients and following up more for financial kinds of support, not for the medication piece. We have that pretty well nailed with our program coordinator. Compliance is an issue when you have the young adults come into the adult practice and maybe not having mom or dad with them to help them get to their labs, remember to keep their appointments, remember to call us with issues or call the pharmacy back when they call them 50 times to refill their medicine. I think those are the biggest issues.

Claire: Liza asked about the age that we actually transition pediatric patients to adult care. I told her that in our practice it's a bit of a range. Sometimes it depends upon the developmental level of the patient or family dynamics. It sometimes depends upon insurance. In California, that may not be until the age of 21 because we have California's Children's Services, which covers pulmonary hypertension patients until they reach 21. Sometimes our young adults decide that when they transition to college, it would be better to have an adult PH provider which is close to their new residence rather than traveling back to wherever their home pediatric provider is. There is no absolute age cutoff or one age for transition. Do you find that as well?

Nancy: Yes. We have never had a big pediatric presence in the pulmonary hypertension world. That's an interesting point about insurance coverage through the state programs. I am not familiar with Ohio's programs to allow for transition to adulthood. For many years, we have worked together with our Pediatric Cardiology group to comanage the PH patient. The pediatric social workers have helped me with many issues surrounding medication support through our Medicaid programs.

Neal: Our pediatric cardiology group has followed several patients with congenital heart disease throughout their pediatric period and then transitioned them to our group as adults. We recently welcomed a new pediatric cardiologist to focus further on pediatric PH at the Cleveland Clinic. Prior to her arrival and even now, we work together with the pediatric cardiology program on a case-by-case basis to discuss cases that are particularly difficult. As those kids start to transition toward adulthood, they often transition to our group for more specific management of their PH. In some cases, our pediatric cardiologists continue to follow them for their congenital heart disease care. They assume the cardiology portion, we assume the PH portion.

Claire: I think that's great, comanagement over several years. Maddie, whether you've known it or not, we've been thinking about transition for even a few years now, and when we go to appointments and talk to you about what your meds are and having you tell us more, what doses you're taking, and asking if you're starting to call your specialty pharmacy. This is all part of this lead up to more independence and having part of your visit without having your mom in the room, too.

Liza: I do agree with that. That is, I think, maybe the most challenging is the insurance component. When you become an adult, you don't really realize how important insurance is or how much you need it. A lot of millennials or younger people nowadays think, "Oh, I'm just going to go see the urgent care in the clinic. I don't need to deal with insurance." For Maddie, it's a different story. She needs insurance no matter what. Transitioning from CCS, which has been really amazing regarding prior authorizations and getting the specialty pharmacies to help with the meds and making sure they come in time. Getting all this stuff in line and delivered.

Neal: Maddie, what are the things that you worry about the most or that you hope that your adult physician would be able to provide or continue for you?

Maddie: I haven't really thought about that.

Claire: Maddie's very in the now.

Neal: Yes.

[laughter]

Claire: Are there any tools that you like to see young adults have when they transition to your practice, Neal?

Neal: Yes. I have learned to have the frank discussion with adolescents about the fact that even if mom or dad is still at the appointment, that's not who the appointment is about anymore. I think it's really important to focus my attention on the adolescent patient as the patient and to start moving the focus of communication away from mom and dad. A progression must occur where responsibility for the PH care moves from parent to child. As I said earlier, depending on where you are, we have to meet you there and work with you to make that transition work.

That can be really hard for some people, and it can take some time. For others, they're on the opposite end of the spectrum and they don't want mom and dad to know anything, which is totally fine. We get to work with the consequences of both approaches.

Liza: That's why I encourage practicing now at least with Maddie to encourage her independence. She does get a lot of the appointments and reminders on her phone, right, Maddie?

Maddie: Yes.

Liza: I don't receive the reminders. She's the one that reminds me. That's a start, I think.

Claire: That's a great start. I think Liza, too, we've seen over the last at least year plus that I think you do a really nice job of when we ask a question in clinic, you look to Maddie. She's the one answering her health questions. That's a great move toward independence, and you can help fill in when it's needed. Maddie is getting to be the age where she's really in charge of her body and her medications and knows what's going on better than the rest of us.

Liza: Exactly.

Claire: In thinking about other types of transitions, sometimes we get to the point where we need to transition to aggressive IV or sub-q prostacyclin therapy. Sometimes the medication management support isn't there, whether it's from the patient's own capacity or lack of supportive caregivers. If you run into this, what options do you consider?

Neal: Fortunately, this has not happened in my clinical practice very often. However, in some cases, we see patients who travel from overseas to see us. Sometimes, those folks don't have access to the same medications in their country as we have here. That can be challenging and our options in these cases can be really limited, which is frustrating. We live in a world where access to health care is very unequally distributed.

In other cases, because of religious or social customs, some patients do have access to care but opt against taking certain medications. This issue most commonly arises with patients who I recommend to start parenteral prostacyclin therapy. Fortunately, in the last few years, we've been blessed to have oral prostacyclin or prostacyclin-like pathway medications as an alternative to IV or subcutaneous therapy for these folks. I don't know if this is the ideal alternative, but at least the alternative exists. When I first started, the oral prostacyclin pathway was not available.

Claire: I agree. We actually run into issues like this in pediatrics as well. Sometimes it's not an ideal social situation. There's not a secondary caregiver that can help monitor an IV or sub-q prostacyclin. We hope to give them the best option that aligns with their lifestyle and social situation knowing that it may not be your first choice.

Nancy: I think of one of Neal's patients, actually, that we met for the first time in the hospital and required a parenteral prostacyclin. We had no idea how terrible her living situation was. A specialty pharmacy nurse went to see her and told us that she had no running water. Even though you asked these social questions in the hospital, those critical details were not asked. Often times the patients are too embarrassed to talk about their living situations, whether it is the physical surrounding issues or family support. I think that's the problem when we get the emergency admissions to the hospital. We really don't know the patient.

Claire: I think that's such a good point. Our therapies are not without risk in day-to-day management. I think we need to be very vigilant of all situations. On the flip side, have you had issues where a patient tells you that they would like to transition away from a more aggressive therapy like IV or sub-q prostacyclin now that they're older or maybe their living situation has changed or any other circumstances? When you make this decision, do you tend to get another cardiac cath? Do you follow them more closely? What's your general approach?

Neal: The answer is definitely yes. There's not many people who love IV or subcutaneous therapy. At least none that I know of. I think about all the things that I did as an adolescent that are restricted for these patients simply because of the mechanics of the pump and the inability to get it wet et cetera. There's a big push in a lot of adolescents, especially as they go off to college, to get off of IV prostacyclins or sub-q prostacyclins. There are obvious limitations to that.

Sometimes we have tried to get people on oral therapy and it's worked well. Sometimes it hasn't. I don't have extensive experience transitioning these patients, but with the last few approaches we've done, what I've said to the patient is, "Let's get a baseline right-heart cath. Let's transition you. Then about 3 months after that transition is complete, we'll repeat your right-heart cath." That's where we've been and it seems to work well for everyone.

Claire: Yes. We've had similar strategies. We've had a handful of adolescents especially that have not done well with sub-q or IV therapy. I think it makes us very nervous to transition away from it, knowing that the other options are not as effective. We also really value the patient's quality of life and want to improve med adherence and prevent bad site pain. I agree, I think we've had some good experiences transitioning from parenteral to oral or inhaled therapy. We've had other kids that did not do well. We've had to have those hard discussions that maybe it's best, even though it's not their favorite, to go back to IV or sub-q therapy.

Nancy: When I was thinking about that question, I was thinking about our elderly population that all of a sudden, whether there may be a change in their mental status and they can't remember to change their cassettes or they no longer have a family member that lives with them, or perhaps they just voluntarily want to move to an assisted living situation. We try to accommodate that because it's not a safe situation that they're going into.

Claire: I think that speaks to the fact that quality of life is a spectrum which balances with medical management.

Nancy: I think it's important to have serious discussions with the patient and family. If you have an elderly patient that you meet first time that has a bad heart cath in the hospital, the first thing everyone wants to do is be really aggressive. We have all likely had to deal with a situation when a patient needs care beyond the hospital. I think it's important to have that discussion if the family thinks that at some point their loved one is going to need an assisted living or skilled nursing facility. Parenteral prostacyclins are generally not accepted in those facilities, which puts everyone in a stressful situation. Some of our oral medications can be quite complex because the patient has to put together different doses to make the final dose. In someone with even early dementia, an error can be life threatening.

Claire: That's a good point. The last question we have is in regard to transitioning to hospice. How have you dealt with urgent hospice situations for patients who are on parenteral medications? Or when the patient is leaning toward hospice, but the family is reluctant to stop the pulmonary hypertension medications?

Neal: Oh, boy. This is a hard one. I think that this is a very personal decision. I think first and foremost, we have to remember to put away whatever is on our agenda and recognize that we're there to support a family who's in a state of crisis. Sometimes that takes a little bit of time and listening and working on a timetable that isn't our own. I heard a quote one time that before patients can accept your care, they need to know you care. I think that that's part of the key in hospice discussions.

If a patient doesn't trust that you have their best intentions in mind, then the ability to navigate conversations that are delicate, such as those related to hospice, tend to be fraught. Stopping to just listen to what the concerns of a patient and family are can be really helpful toward assuring them that you're working on their timeline. Sometimes that requires months, or years, or sometimes it requires very little time. At the end of the day, I have never pushed somebody against their will to come off of medication. What I have done when the situation presents itself is help to try to explain what the purpose of hospice is and how that's different from what the purpose of the medications are.

Giving patients and families the space to recognize that hospice is not trying to treat the disease, but helping to treat the patient provides an alternative to the narrative we're just withdrawing support. In fact, we're actually providing a different level of support that is wholly dedicated to empowering patients to pass away on their terms, with dignity and without suffering. That is powerful. That can't happen until families trust that in fact, the support you're proposing to provide is actually in their love one's best interest.

Claire: I absolutely agree. Aligning with your patient and really listening to what they're asking for in life, and how they want to live out whatever time they have left, can really change your strategy on how to best treat your patient. That may mean aggressive medical management or more comfort care and time outside of the hospital. It is a broad spectrum.

Nancy: I think this is always such a difficult situation. I know there've been times where we've had patients who wanted to transition to hospice but did not want to abruptly stop their PH medications. Before finalizing the hospice contract, the patients ordered their medications which allowed them to transition more comfortably. It is important to discuss with hospice and patient to develop the plan of care.

Claire: Does anybody have any last thoughts? I think we're just coming to the end of our time.

Neal: There isn't a magic formula for the transition between pediatric and adult care. It's not like, once you hit 18, there's some other human being inside that body that magically changes or flips a switch which necessitates care by one provider or another. It's a continuum. In my opinion, the best programs provide a continual off-ramp and an on-ramp. At the end of the day, recognizing that the patient is the customer, and it's our job to work with them and not the opposite, is important. This was engrained in me when I first showed up at the Cleveland Clinic. But in the hustle of our jobs, it is easy to lose sight of why we're here. As Nancy said, our pediatric PH program is pretty small. The patients who we have had transition and the congenital heart disease patients we have had transition have been an opportunity for us to build fantastic relationships with patients and their pediatric providers. Over the years as those relationships have grown, we've built mutual trust. This, in turn, helps build rapport among patients who have been with that practitioner for years and years and are now transitioning.

Claire: Yes. I agree. Absolutely. I think that is a wonderful way to wrap up our roundtable discussion. Thank you all so much for taking the time out of your day to join us.