



FA Connect: Question & Answer Transcript

Session Title: Living and Coping with Fanconi Anemia

Invited Speaker: Dr. Naomi Joffe, PhD; Assistant Professor, UC Department of Pediatrics; Cincinnati Children's

Session Air Date: February 18, 2021

Access a recording of Dr. Joffe's presentation by visiting www.fanconi.org. The recording has been uploaded with Spanish subtitles.

Have additional questions for the speaker? Email Dr. Joffe directly at naomi.joffe@cchmc.org.

Q: My son is 12 years old and we've known he's had FA for 10 years now. He has some physical differences and has also experienced some academic challenges, including his speech and a variety of other things. He does work with a pediatric psychologist, which has been wonderful for him and his progress; however, he's 12 and will be going to middle school next year. Do you have any kind of recommendations on how to bridge those close grade school friends as he transitions into middle school? It's likely that he'll be a part of a specialized classroom or learning environment, so would you have any feedback on how to make this transition easier?

A: The idea of development and the challenges that accompany these times of change can be difficult. I think as a parent, you are looking for opportunities to help him maintain those relationships while also supporting the idea that there's a whole new group of friends to meet in this new school setting. It's important to continue supporting him in keeping up with old friendships, while also reinforcing that this new school setting will present opportunities to add to this friend collection. It's also a benefit that you're already connected to another psychologist in the community, since there are bound to be things that you're worried or anxious about due to this new kind of stage. It's difficult to anticipate the specifics of these feelings or situations, but that may be someone who's helpful in assisting your son to navigate those waters as different things arise, such as addressing those difficult interactions that he may be feeling worried or nervous about.

Q: I'm a 30-year-old and have been aware that I have FA since I was four. I had a bone marrow transplant (BMT) when I was 11 years old and have been doing pretty well since that point. Recently, I was diagnosed with squamous cell carcinoma (SCC) and had to receive a CAT scan. The process of receiving scan was very traumatic for me in bringing up bad surgical memories from my childhood. I found that I'm still dealing with a great amount of stress from that recent experience and recognized that it's still really challenging to navigate FA into adulthood. I think I have post-traumatic stress (PTS) from everything which I never really had to deal with as a child but am now being flooded with those memories and emotions now that I'm an adult. I'm wondering if you had any suggestions for how to mentally cope with these triggering events?

A: Thank you so much for sharing and I'm sorry that happened fairly recently but am glad that you're doing well. First, I want to normalize—not in that it's normal or okay—but normalize that your experience is not an uncommon experience in terms of some of those kinds of post-traumatic stress (PTS) symptoms. Unfortunately, these experiences are something that many other patients are likely to face because FA as a chronic illness isn't just a brief diagnosis, but

rather a lifelong journey. Therefore, there are lots of opportunities for things to happen that are traumatic and stressful. The positive outlook is that there are really good treatments in terms of PTSD and the first step would be talking with your hospital or physician and asking for recommendations for providers in the community who specialize in PTSD treatment. The cognitive behavioral therapy in combination with exposure and response prevention are two evidence-based treatments that are kind of “top tier” for treatment of PTSD, whether it’s medical trauma or other trauma, and I’d encourage you to think about whether you are interested in looking into that as an opportunity.

Q: Do you often see ways that PTSD presents in a more common way or is it just different for everyone? In thinking of individuals who have gone through or are preparing to go through transplant, perhaps there are some ways they can prepare themselves mentally or physically for that process.

A: Great question. For traumatic stress, if we’re noticing that we’re having more intrusive thoughts throughout the day about those past or upcoming traumas; if we’re noticing physical reactions when faced with things that remind us of those traumas; if our mood is being impacted or we’re feeling more down or depressed or irritable; and if those things are impacting us throughout the day and impacting our ability to do those things that we want or need to do—those are all good things to look out for as a sign of traumatic stress. They’re a good reminder for us to acknowledge what is happening in the moment and to remember that it’s always okay to reach out to a professional and seek support. Even if after seeing a professional, you’re told you might not qualify for a diagnosis of post-traumatic stress disorder, there are still a lot of things you’ll be able to address together to make things better and more manageable.

Q: Can you talk about the coping process and ways to establish some expectations when siblings of the patient(s) become involved in particular trauma-induced situations (i.e., transplant)?

A: I think that similar to what we do with patients, it’s important to establish a routine for the sibling(s). Are there ways I can support? How do I support the sibling and maintain as much of their normal routine as possible while going through the transplant process? Ultimately, it’s really helpful when families have any form of extended family or friends that are already in that child’s life that continue to be in that child’s life during this time. Providing as much kind of normal consistency, whether it’s through their activities, friend group meetups, through the person they’re staying with, or weekly schedules, will ultimately be of more help to them. It’s also important to remember and discuss that things aren’t going to be normal at this time and you’ll likely have to make changes. But talking about expectations and letting them know what to expect and when, will have a great positive impact. One thing I really try to normalize for families is the use of and presence of a psychologist on their transplant journey. I appreciate my role as a pediatric psychologist, as I’m able to meet and work with a number of families who wouldn’t have normally turned for help from someone in my role. Dealing with something like a transplant is challenging and I always try to really normalize that it can be very helpful to be connected to a professional during this process, whether it’s for the caregiver, for the patient, or for the siblings. Receiving help from a psychologist isn’t something that families have to do, but I encourage them to remember that it’s an outlet that’s readily available to them. This process is hard, and families can do better when they have some additional support, particularly for going through transplant when they’re away from their other support systems. All around, it’s important to ask the families, what are some other things or systems we can put in place, whether they’re within the family circle, or utilizing a professional in the community or hospital, to help make this process as good as it can be, knowing you don’t want to be doing what you’re doing.

Q: Can you expand on tips to resolving differences and coping techniques between an adult patient and their parent? I am a 26-year-old who is about to move home for transplant and am struggling with that loss of independence.

A: Yes, that is a great question. This is something that comes up for a lot of our young adults who are in similar situations. Often times when I meet a patient who is someone working towards independence—a young adult—I will have a good conversation with them and with their parent, and will discuss and recognize that this situation is not normal. What is even normal as a 26-year-old? What are your peers doing right now? Maybe you have a job or are finishing school, or are even starting a family. At this stage, you’re likely not wanting to live at home with mom or dad and spend a lot of time with them. During these conversations, I try to normalize all sides of the equation for the family. We discuss what the son or daughter “should” be doing right now and then compare it to what they “are” doing right now. We discuss that from the young adult’s perspective, if we’re not doing something we should be doing

developmentally, we don't usually feel great about it. We talk about ways to not drive each other crazy while being in the same room or house on a regular basis, and we talk about ways that we can continue to have some of that independence as a 26-year-old.

For the young adult going through transplant, maybe they'd prefer for their caregiver to not participate in rounds. Perhaps they want to re-evaluate how much information is shared with their caregiver, or when that information is shared at all. For young adult patients, I'll often try to support working with the family and the nursing staff to have about 30-60 minutes per day that offers a quiet hour where no one comes into their patient room. Some days will be too hectic during transplant to make this possible, but most days will allow for this break. An example of what we'd do would be to post a sign on the outside of the patient's door from two to three, with written instruction to 'please not enter unless medically necessary'. If there are caregivers present during that hour, they can go take a break, and hospital staff would know not to knock on the door during this time. Ultimately, we know that when you're in the hospital, that's essentially your bedroom. It is not normal to have people like me walking into your bedroom all day long. My patients have told me that it feels different to know for an hour that no one's coming in, then to not know if someone's coming in. It simply gives them that time to be independent and in charge of their space. All of these things are good discussion points to start having with your parent(s) if you find yourself in a similar situation.

Q: Do you have any specific recommendations for once families are discharged home and are still experiencing behavioral issues? How would you suggest we manage those issues once we're home and no longer have an immersive care team around to help and support?

A: The transition home is surprisingly difficult for lots of our families. One thing we recommend for families to recognize is that preparing for, going through and then completing transplant is a huge adjustment for them, especially over such a large period of time. It is no small feat that they have succeeded in getting through, and it has required lots of changes and people to be out of their normal routine. It's required flexibility. It's important for families to recognize that it's going to take a minute to settle back to a routine—especially to children re-learning what their parents' expectations are at home. Coordinating medications in the home setting without the hospital staff is also a new adjustment. Altogether, this transition is one of those things that is easier for us to recommend suggestions while in the hospital, but then is much harder for the parents to execute once back at home. As expectations and circumstances are changed, there will likely be some push back from that child. It's normal to see behavioral challenges during this time; however, I would say that if parents are continuing to find that their child(ren) are not settling into the new home routine or are concerned about their extensive mood, worry or anxiety, that would be a good time to bring these issues to the medical team. In doing so, you could get connected to resources and get additional kinds of outside help for these extended behavioral concerns.