



TRANSITIONING TO ADULT HEALTHCARE IN THALASSEMIA: Q&A with Dr. Farzana Sayani

ABOUT DR. SAYANI

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Dr. Sayani attended the University of Calgary, Canada, for medical school, completed residency and fellowship training at the Universities of Calgary and Alberta, and completed a Hemoglobinopathy fellowship at the Hospital for Sick Children in Toronto. She has a Master's Degree in Haemoglobinopathies from University College London.

Dr. Sayani joined the Division of Hematology/Oncology at the University of Pennsylvania in Philadelphia in 2012. During her time there, she has established the Penn Comprehensive Adult Thalassemia Program, and developed a successful transition program for thalassemia patients from Children's Hospital of Philadelphia (CHOP). Dr. Sayani serves as the Director of the Penn Comprehensive Adult Thalassemia Program and the Penn Comprehensive Sickle Cell Program.



Why must thalassemia patients undergo a transition to adult healthcare? How does adult healthcare differ from pediatric care? What are the advantages to adult patients who transition their care to an adult setting?

The "transition to adult care" refers to the process of getting pediatric patients ready to eventually assume full responsibility for their own healthcare as independent adult patients. This may or may not require the transfer to a new physician. The American Academy of Pediatrics recommends that doctors and families begin to plan for transition as early as 12 years of age.

In an adult setting, adult patients are the ones who make the decisions about their own healthcare. (Of course, these decisions may still be made in consultation with parents, loved ones, and healthcare providers.) It is therefore important to prepare young adult patients to take on the responsibility of decision-making, while still ensuring that they feel supported and have the tools to seek out, navigate and process information related to their care. Unlike pediatric care, adult care often involves the need for the patient to be actively involved, to ask questions, to be a self-advocate, and to take responsibility for their own well-being. Adult providers supply adult patients with important health information and engage them in discussions of treatment options and goals, and



expect the patients to actively participate in decision-making. Parents and other members of a patient's support network may be involved in these discussions - with the adult patient's consent – but ultimately it is the adult patient's decisions that will be considered.

It is important for adult thalassemia patients to establish care within an adult setting for several reasons: Adult doctors are experts in providing care for the adult complications of thalassemia such as endocrine, cardiac, and liver complications. Even simple things such as transfusions may have different medical considerations in the adult patient. As adults age, they are also at risk of developing other illnesses/diseases that are best managed by adult teams. An adult hematologist is trained in how to manage adult hematological disease in the context of other adult illnesses, and may be best suited to coordinate multidisciplinary adult care. In addition, fertility and reproduction are best managed in an adult care setting. Adult patients will also need to navigate educational and occupational opportunities, relationships, and life goal-setting, which are best supported by a team of adult providers and behavioral health specialists. The adult team will help the patient with thalassemia to achieve their goals and aspirations as an adult, while managing their thalassemia.

What are transition to adult healthcare programs? How do these programs prepare pediatric patients to make the transition to adult care?

The purpose of Transition to Adult Healthcare programs is to teach skills to teenagers and young adults to help them effectively manage their own healthcare and to successfully utilize available health services. Teens and young adults are guided in this process by their healthcare providers and by their parents/caregivers. Each teenager/young adult will move through this process at their own pace. In their younger years, patients needed parents and their healthcare providers to make all of their health-related decisions for them. As they reach their teenage years, they may be able to start making their own health-related decisions with support from their parents and their healthcare providers. Eventually, they will be able to make their own independent healthcare decisions.

Even after transitioning to adult healthcare, patients are still encouraged to seek support from their parents and loved ones. Living with thalassemia is difficult, and having a support system benefits all patients, including adults.

What responsibilities do thalassemia patients take on as they transition to adult healthcare?

As young adults with thalassemia transition to adult healthcare settings, it is important for them to become comfortable with several new responsibilities, including;

- Scheduling their own medical appointments with the thalassemia provider and any subspecialists
- Scheduling blood transfusions and necessary medical tests
- Understanding the reasons why they may need blood transfusions and why they must take particular medications
- Being familiar with the complete list of medications that they take, knowing how to obtain these medications, and keeping track of when medications must be re-ordered
- Knowing how to contact their medical providers if they have any questions
- Knowing which emergency room they would go to if needed, and how to contact their regular providers in emergency situations
- Learning about health insurance and what is involved in obtaining coverage
- Learning to advocate for their own healthcare needs to help ensure that they receive the best possible care

What skills and knowledge are most helpful to patients in making a smooth transition to adult care?

The transition to adult care makes many patients uneasy, as it means leaving the comfort of the pediatric setting which is familiar and which offers a lot of support. Some of the skills that are most helpful in



making a successful transition to adult care include:

- Being able to ask for help when needed, especially when facing new life experiences
- Making healthcare a priority, and learning more about thalassemia
- Bringing up any concerns about health or about the transition process itself with healthcare providers
- Giving new healthcare providers the opportunity to get to know the patient and their life goals so that they can be partners in care
- Learning how to become an effective self-advocate. This requires patients to believe in themselves, to express their needs clearly, to assert themselves, and to obtain support from others. Learning how to advocate for themselves can give patients an important sense of control over their lives and helps them to achieve their goals, healthcare-related and otherwise.

When should a child begin the process of transitioning to adult care?

The earlier the transition process begins for a patient, the more prepared the patient will be in the long run. Many patients start preparing for transition during their early teenage years to allow sufficient time to practice the necessary skills in making their own decisions and managing their own healthcare. Early teenagers should start to become knowledgeable about their condition and their medications. They can be responsible for taking their own medications. They can also start to identify when a medication refill is needed and know how the medication is obtained. Late teenagers may also practice making their own healthcare appointments and ordering prescription refills. In addition, they should start to see their doctors alone – for at least part of the doctor’s visit – to gain independence in managing their own healthcare. Teenagers may have conversations with their doctors about what their privacy rights will consist of once they turn 18 years of age. They may also talk to their parents and healthcare providers about the age at which they would like to make the transition to a new doctor for adult care – or if they will need to find a new doctor if they end up going away for college.

What can parents do to encourage their child to take a more active role in their own healthcare management?

Parents have a key role to play in helping their teenagers learn how to manage their healthcare on their own. Parents may ask their children’s doctors the following questions to encourage their child’s move towards independence:

- What can I do as a parent to help my child with the transition process?
- What does my child need to learn to prepare for the transition to adult healthcare? Do you have a checklist of skills that my child should begin to work on?
- When should my child start to meet alone with you, for at least part of the visit, to start playing a more active role in their own healthcare?
- Can I work with you to prepare an Emergency Care Plan for my child so that he/she is prepared in the event of a medical emergency?
- How will privacy and consent change when my child becomes 18 years of age?
- What is the process of transition and what will care look like after he/she turns 18 years of age?

How can patients find an experienced adult provider in their local area? How can they determine whether the new doctor might be a good fit for them? What questions should they ask about the adult provider’s practice?

The pediatric provider should be able to recommend one or more experienced adult thalassemia providers in a patient’s local area as they begin to transition to adult care. If they are unable to do so, the Cooley’s Anemia Foundation (CAF), which advocates for U.S. patients with thalassemia, can help patients to find an adult provider with thalassemia experience in their local area. To assess whether the potential new adult provider may be a good fit, patients should contact their office and arrange for a new patient visit. This will give transitioning patients an opportunity to meet the program physician and nurse in order to get a sense



of their thalassemia knowledge and experience. It may also allow patients to take a tour of the transfusion and clinic facilities. It is very important for patients to ask questions during this initial visit regarding their care, so that they know what to expect if they were to become a patient within that practice.

Some important questions to ask of the new adult provider include:

- **Transfusions:** Where are the transfusions administered? What are the hours of operation for the transfusion clinic? What is their protocol for typing and screening, as well as for performing transfusions? Is transfused blood matched for C, E, and Kell antigens? How are transfusion reactions managed? Will it be necessary to see the physician before each transfusion? What are the pre-transfusion hemoglobin target ranges? How would they adjust the transfusion regimen if a patient's pre-transfusion hemoglobin level was lower or higher than the target range?
- **Iron overload monitoring and management:** How often is serum ferritin monitored? Do they have an MRI machine that specifically quantifies liver iron concentration (LIC) and how often is this testing performed? Is cardiac MRI T2* available, and how often is this testing performed? When putting patients on a new iron chelator, how often do they monitor to see if the medication is effective or if it causes any side effects?
- **Monitoring for complications:** Does the practice have specific subspecialists they can refer patients to for any thalassemia-related complications? These subspecialists might include an endocrinologist, a hepatologist (liver specialist), a cardiologist, an ophthalmologist, an audiologist, and a reproductive medicine physician.
- **Emergency care:** What is the protocol if a patient should develop a fever (for example, stop chelation immediately and contact the hematologist)? Who should patients contact if they feel unwell or develop new symptoms, or experience a transfusion reaction? In the event of an emergency, which Emergency Room are patients referred to, and what protocols are followed regarding their care?

- **Patient support:** Does the practice have a nurse with experience in thalassemia care? Does the practice have a social worker available to thalassemia patients?
- **Patient services:** How many other patients with thalassemia are currently managed in the practice? Do they offer thalassemia support groups?
- **Optimizing patient care (for patients in smaller centers):** Does the provider consult with a thalassemia specialist in a Thalassemia Treatment Center when needed? Would they consider jointly caring for a patient together with a thalassemia specialist at a Thalassemia Treatment Center? How does the provider keep up-to-date with advances in thalassemia care?
- **Clinical trials:** Can the provider directly enroll patients in clinical trials, or can he/she refer patients to another provider who can enroll patients in a trial?

Do patients' pediatric providers and their new adult providers generally work together communicating behind-the-scenes during transition to ensure a smooth handover of care? If not, what can a patient do to help facilitate the handover of care?

The extent of collaboration between a patient's pediatric center and adult center at the time of transition varies a great deal between medical practices. Some larger pediatric centers that care for a large number of patients have established transition protocols in place and work closely with adult centers at transition time to make sure that all appropriate documents have been shared, that the Blood Banks have been in communication with each other, and that the pediatric and adult providers have been in contact. Some adult centers will schedule a visit with the transitioning patient to give them a tour of the new facilities and to allow them to meet the new adult team and to ask questions about their care.

However, if there is not much collaboration between the pediatric and adult centers then it will be necessary for the transitioning patient to play a more active role in the transition process. In such cases,



the transitioning patient should make sure that the relevant pediatric medical records are sent to the adult provider prior to the first adult appointment. They should also request that the pediatric and adult providers communicate with each other about the patient's case prior to transition, so that the adult provider has been brought up to speed on key areas of care management. Such patients may also find it helpful to ask the adult center if they can have a tour of the new adult facilities and an opportunity to meet the new, adult team and to ask them questions about their medical practice.

What are some of the biggest challenges associated with the transition to adult care?

Young adults with chronic health conditions are a vulnerable patient population. Sometimes their health conditions worsen as they become fully responsible for managing their own healthcare, especially if they have not been adequately prepared to take on these new responsibilities. This worsening of their health status may be associated with reduced use of medical services, and reduced adherence to their prescribed medications – especially iron chelation medicine.

Many patients who undergo the transition to adult care report on the difficulty of leaving their pediatric providers, with whom they have had long-standing relationships. Sometimes these patients feel abandoned by their pediatric team, even though they are aware that hospital rules sometimes prohibit pediatric providers from treating and/or admitting adult patients into a pediatric hospital if necessary. Transitioning patients may also feel distrustful of the new adult care team, who may spend less time assisting them than their pediatric providers did and may therefore appear to be “less invested” in the patient's care. Processes and procedures in the adult clinic/hospital may also be different from what the patient experienced in the pediatric center, which can take time to adjust to. Finally, the transition to adult care comes at a time of life when young adult patients are undergoing many other life transitions, all of which may leave the patient feeling overwhelmed.

What advice do you have for patients who are struggling during/after the transition to adult care?

The transition to adult care is a major life change, and it may not go smoothly at first. Patients may find that they are often forgetting to take their chelation medicine without their parents to remind them, or they may find that scheduling their transfusions may be trickier while balancing other school/work commitments. Patients may also miss seeing familiar faces and friends from the pediatric setting at transfusion time. Patients who find that they are struggling with any aspect of their healthcare following transition should understand that this is normal – but they should also reach out to their new adult thalassemia providers as soon as possible to let them know so that the issues can be addressed. The new healthcare providers may not have had a long history with a recently transitioned patient, and they may not have been aware that the patient was struggling. But once they realize that an adult patient needs assistance, the doctor/nurse/other members of the team will work with the patient to help resolve the issue. There may also be a patient support group that patients can participate in, or there may be a social worker available to talk to. It may sometimes be helpful for recently transitioned patients to reach out to the providers in their former pediatric center – with whom they have a long history – if they are in need of advice and assistance.

The Cooley's Anemia Foundation (CAF) is also a resource for thalassemia patients who may be struggling with transition. CAF has a social worker who patients can speak to about the process of transitioning to adult care. CAF can also provide introductions to other patients who have gone through the transition process and who can share their experiences and insights. CAF and the Centers for Disease Control and Prevention (CDC) have also developed a series of videos which feature thalassemia patient Aaron Cheng, MD, discussing his experiences living with thalassemia – including the transition to adult care – which can be found at: <https://bit.ly/2KPGXda>. The most important point is that patients do reach out to someone for help when they are struggling so that they don't feel alone in this process. There are many resources available to help with transition.



Many patients end up transitioning to adult care when they move away from home to attend college. This can be especially challenging as these patients do not have their parents around to support them during transition. Do you have any special advice for patients whose transition to adult care coincides with a move away from home?

Going off to college can be an exciting time of life, and a key step in developing independence for young adults. Because their health needs are of critical importance, many college-bound patients find it helpful to visit the campus student health center as well as a nearby medical center with thalassemia expertise when they go on tours of potential colleges. Patients who need help finding an experienced thalassemia physician/medical center at or near their college-of-interest are welcome to reach out to the Cooley's Anemia Foundation (CAF), which is in contact with thalassemia physicians throughout the U.S., for assistance.

Once a patient has identified a college or university which offers an academic program that suits them, they can begin to investigate the thalassemia treatment center options that are nearby. If the college is located near an experienced thalassemia treatment center, patients can reach out to them to see if they are able to accept new patients. Many pediatric programs will accept college students, who may remain with the program through college graduation. However, if there isn't a large, well-established thalassemia center nearby then patients may still be able to get excellent thalassemia care while in college by partnering with a local hematologist for ongoing care (transfusions, monitoring for iron overload, chelation monitoring, and referrals for local subspecialty care). This care can be supplemented by visiting a specialized thalassemia treatment center that cares for a large number of thalassemia patients for a comprehensive annual evaluation once per year. The highly experienced thalassemia center will review the patient's health records, care plan, and latest test results and will collaborate with the local hematologist to optimize the patient's care in order to keep the patient healthy for the long-term.

Another important step is for patients to figure out which type of insurance coverage they will have in college and whether this will be accepted by the thalassemia clinic that is at or near the college. It will then be helpful to connect the new thalassemia provider at college with the pediatric provider so that the medical records can be transferred and the physicians can speak about the patient's healthcare needs. Patients may want to remain in contact with their pediatric thalassemia program until they feel that they have established a good relationship and management plan with their new adult care team. In fact, patients attending college may continue to see their original pediatric providers when they are home on holiday breaks or for summer vacation.

Patients may find that their adult care providers do not have as much experience with thalassemia as their pediatric providers did. Patients should know that they may still be able to get very good care in such a situation, but this will depend on the patient knowing the recommended care guidelines for thalassemia, and advocating for themselves if the new care team is not aware of these guidelines. The Cooley's Anemia Foundation has some Thalassemia Management Checklists available on their website on topics such as managing transfusion therapy, monitoring of iron overload, and monitoring of deferasirox therapy, which may help with this. These Checklists can be found at: <https://www.thalassemia.org/learn-about-thalassemia/about-thalassemia/#thalassemiabrochures>.

Also, patients may want to prepare for the possibility of an emergency by letting trusted people – such as their roommate, college residence advisor, and/or friends – know about thalassemia and how it may affect their health. It can also be helpful for patients to stay connected to their family and friends from back home – even though they may have moved far away from them – as they can continue to be a strong source of support. Friends from the thalassemia community can be another key source of understanding and support, regardless of where patients end up living. Finally, the more patients can do to maintain a healthy lifestyle (getting a proper amount of sleep each night, eating healthy foods, and exercising) while in college, the better they will feel as they begin to manage on their own for the first time.



Do you have any suggestions for parents on how to “let go” of managing their child’s healthcare at the time of transition?

Some parents find it difficult to “let go” of managing their child’s healthcare as their child enters early adulthood. In such cases, it may be helpful to remember that the transition to adult healthcare is one of many phases of their child’s move towards independence. For their child’s long-term well-being, it is best to teach their child the skills that will enable them to eventually manage their healthcare on their own.

Parents of young children find themselves in the role of overseeing all aspects of their child’s healthcare. As their child gets older, this role will gradually change. Instead of completely overseeing their child’s care, they will start to function more as “consultants” to their late teenage/young adult children as they begin to take a more active role in their own care management. The transition process therefore affects parents as well as children, and involves a gradual series of steps to make sure that everyone is adequately prepared:

- Children should be encouraged to ask questions of their healthcare providers as early as 12 years of age. This will engage them and help them to become more active participants in their own healthcare.
- In the early teenage years, children should be given the opportunity to speak privately with their healthcare providers. This will enable them to gain confidence in their abilities to manage their own care. It will also give them the opportunity to ask personal questions of their doctors while allowing them some measure of privacy.
- It may be helpful for parents to engage their teenagers in open discussions about their healthcare and any medical decision-making that must be made.
- Teenagers should be encouraged to manage their own medications. However, parents should check to make sure that their teens are, in fact, taking the medications as prescribed, and that medications are reordered when the supply is getting low. It is normal for teenagers to make some mistakes in taking their medications, and this can be a good

opportunity for parents to serve in an advisory capacity to help them to improve. This might involve exploring the reasons why they didn’t take the medication as prescribed, and helping them try to find solutions to overcome any obstacles so that they can better stick to their prescribed medication plan in the future.

- It may also be helpful for parents to have open discussions with their teenagers regarding their thoughts about their future (for example, heading off to college or the military, as well as hopes for eventual marriage and children) and the importance of taking care of their own health so that they can realize these dreams.

What legal changes in healthcare occur when a patient reaches 18 years of age, and how can this impact their medical decision-making and treatment?

Patients become legal adults at 18 years of age and are therefore legally responsible for their own care. At that point, the doctor will talk directly to the patient – not to his or her parents – about the patient’s health. In fact, parents/guardians cannot access the patient’s medical records, and need the patient’s approval to be in the room at the doctor’s visit. Confidentiality between a patient and his/her doctor is legally known as the Health Insurance Portability and Accessibility Act (HIPAA). HIPAA is a federal law that gives a patient legal rights over his/her health information and sets rules and limits on who can look at and receive his/her health information. Any health related information found in a patient’s medical records, conversations with providers about the patient’s care, as well as patient information in insurance providers’ computer systems and billing information are all protected under HIPAA. The patient’s consent is required for anyone to access this information. As an example, transitioning patients may be asked to provide consent prior to sharing their information with their new adult thalassemia doctor. Parents will also not be given any information directly from the adult thalassemia doctor without the adult patient’s permission.



How does the transition to adult healthcare impact the other transitions (for example, in academics, housing, social life, and employment) that a young adult may be undergoing at the same time? How might a successful transition to adult healthcare set a young patient up for success with the other transitions they are making to adulthood?

The transition from adolescence to adulthood represents a major life change that impacts many dimensions of a person's life. This stage of life is characterized by growing independence and self-determination. Many individuals with thalassemia leave home at this time to attend college or to begin a career. The preparation patients have undergone to get ready for their transition to adult healthcare may help them with other major life transitions they are making at this time as well. Some of the useful skills patients may have learned in preparation for their transition to adult healthcare, and which may help them in other areas of life, include:

- Learning to set goals and to outline specific steps to achieve them.
- Taking responsibility with regard to their own healthcare.
- Learning to ask for help when needed, which shows maturity and confidence. It also requires the ability to identify people who are likely to provide the help that is needed.
- Not giving up easily if asking for help didn't immediately produce the desired outcome. Sometimes obtaining help requires multiple requests and/or ongoing effort.
- Problem-solving when unforeseen issues arise, or when things don't go according to the original plan. In this way, the preparation for transitioning to adult healthcare teaches patients many useful life skills that can benefit them even outside the realm of healthcare.

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