Cold Open

Hannah: All parents can agree on one thing – they want their children to be healthy and happy. But every day, parents around the world experience the earth-shattering moment of learning that their child has a chronic illness, which can completely alter the trajectory of their life. Camille knows this feeling all too well.

Camille: It's not an easy thing to hear that your child is less than the perfect creation that you wanted. And however you are learning about it, it's going to be difficult. It is not something you will take lightly at all.

Hannah: Camille's son, Camren, has sickle cell disease. That means his red blood cells are hard and sticky, and each one looks like the c-shaped farm tool called a "sickle". This abnormal shape makes it hard for the red blood cells to easily move through blood vessels and deliver oxygen to the body, causing symptoms like debilitating pain, swelling, and frequent infections. The disease can make daily life excruciating and can even be deadly.

Sickle cell disease is an inherited condition, and many people believe it only impacts individuals of African descent. But it can be found in many kids, including those with Mediterranean, Indian and Middle Eastern ancestry. Camren's journey with it started before he was even two-years old. Today, he's 17.

Camren: I'm in grade 11. I like to play sports. I've been playing sports from a very young age. Three years old, if I can remember correctly. My dad put me in soccer, so I've been playing soccer ever since.

Hannah: Can you tell us, Camren, some of the symptoms that you experience?

Camren: Well, joint pain was one of the major ones because of all the sports and stuff. When I would play in some unfavorable temperatures, I would start feeling tightness in my joints, like a lot in my legs. And at first, I didn't really know what was going on. And I was like, oh, this is uncomfortable. I don't really like this. But after asking some questions, learning from mom, asking doctors, they told me that it was sickling in the blood and that was because of the cold temperatures. That's the main one that I can remember.

Camille: Whenever he was experiencing like influenza or flu, a cold, fever ensues. And as soon as the fever ensues, it could possibly cause those pains because he was sickling. Those were things that he did ongoing all the time. I just have to tell you this one little story. When he was four, we went to African Lion Safari, just outside of Toronto. Okay, so my son, we toured the whole park the whole day, and he's like, mommy, can I go into the water park? And I was like, no, because that water is ice. And he so badly wanted to go, and oh my gosh, Hannah, the joy. We recorded it, because my heart was just bursting. When he was done, he was shivering. And we know when he's shivering that bad. So, we went straight to the hospital. But it was worth it. We knew what we were doing, and it seems like it was torturous. But He had a moment of joy.

slight pause

Hannah: Camren is one of around 2,000 children in Canada impacted by sickle cell disease. About half of them will be seen at SickKids, where Dr. Isaac Odame is a blood disorders specialist. Isaac has dedicated his life to making things easier for families with sickle cell disease. He played an integral role in the

implementation of a newborn screening program for early detection and treatment in newborn babies. Now he has his sights set on toxic treatments, anti-Black racism, a potential cure, and limited care capacity globally.

Hannah: You're listening to SickKids VS, where we take you to the frontlines in the fight for child health. I'm Hannah Bank, and this is SickKids VS Sickle Cell Disease.

Act One

Hannah: The most common medication for patients with sickle cell disease is hydroxyurea.

Camille: I don't know if you've had the pleasure of reading about that drug, but any parent with that being presented to them just clenches their heart.

Hannah: Hydroxyurea is a chemotherapy medicine. For sickle cell disease patients, it makes their red blood cells bigger, rounder and more flexible – less likely to sickle. Administering the medication at home, however, is difficult for families. Because of the drug's toxicity, it needs to be handled very carefully. Here's Isaac to explain.

Isaac: So, hydroxyurea is a key drug. But this medication comes mainly in capsules. You've got to dissolve it in solution. And what we've done in the past is that parents then have to open the capsule, which has a powder in, have it go into a syringe with some liquid to dissolve it, to prepare the solution and give it to the child. And as you open the capsule, some goes into the air. Now, they have to wear gloves, they have to wear masks because you want the child, only the child who needs to be exposed to it. You don't want normal people, a pregnant woman, to be exposed to those powdery substances in the air because they are aerosolized once you open it.

Hannah: To make it easier for parents of children with sickle cell disease to administer the medication, a team at SickKids developed a specialized tool to shred the capsules.

Isaac: It doesn't allow any aerosols to be released, so that way it is safer. It is neat. It's not injurious to the environment. Parents can feel safe and can use it confidently without all the barriers that they were facing before. And when we sent out a survey to all the children's hospitals in Canada, they all said, absolutely, yes, we need it for our families. So, there were 700 children we know who take this medicine and have to dissolve it through that process. So, what we are developing will serve at least 700 children around Canada, 300 of which we see here in SickKids.

Hannah: After several prototypes and lots of testing with SickKids families, the Capsule Shredder is now being manufactured locally and will soon be distributed to patients and families across Canada. The Capsule Shredder will make it easier for children to receive the medication and will help their parents administer it more safely. Next up? Make the Capsule Shredder available to countries around the world.

Act Two

Hannah: Pain caused by sickle cell disease can be excruciating, so it's important to seek treatment as soon as possible. Navigating the healthcare system can be tricky. Especially when you have an unfair disadvantage.

Isaac: The story of sickle cell disease cannot be told if we're going to address it without addressing anti-Black racism.

slight pause

When patients seek treatment, they face their everyday anti-Black racism. For example, you're going to the emergency department in pain, needing help. Now, you and I will agree that pain is not something you can easily measure. It is not a pulse. It's not blood pressure. It's not a temperature. Pain is what the patient says I feel. And that is where they face the most frustration because they have pain, and they are not believed.

I will tell you an example. A woman with a sickle cell pain crisis and recurrent crisis has realized that if she goes to the emergency department with better makeup, well-dressed, she gets attention more promptly than if she doesn't. Now, which one of us, when we are in intense pain, will have to spend time to get ready, put on makeup, put on clothing as though you are going out to a party, going out to a very important place because you're you are fearful that if you don't do that, you won't get the respect and recognition that you need that will address your pain issue. These are barriers that should not happen in modern day Canada. And anti-Black racism definitely is a cause for concern for many patients which make them reluctant to seek care. And sometimes that has been a mistake because it has been that cause of early mortality.

Hannah: While SickKids is a leader in child health, we are also part of a system that is still addressing existing systemic inequities. We acknowledge and take responsibility for these inherent barriers that exist for the Black community. An initiative called the Black Experience @ SickKids is working hand-in-hand with the Black community to increase awareness about inequitable treatment, raise much-needed funds, build systems of support and spread knowledge globally. A major focus of this initiative is sickle cell disease.

Isaac: You can have the best care, but if the individual cannot access it because, for example, transportation to get into the place is a challenge, then you may have the service, but it will not be accessed. And these are inequities that exist. So, SickKids through the Black Experience at SickKids is beginning to take a closer look at all these and saying for all the expertise and the services we have, are we making sure that nobody is disadvantaged in accessing that? And that all families, regardless of their race and the socioeconomic status, have equal access to these facilities and expertise.

Act Three

Hannah: Universal newborn screening—introduced in Ontario in 2006— has dramatically reduced complications from sickle cell disease by enabling better monitoring and more proactive treatment. Because of these advances, doctors like Isaac are focusing more and more on expanding options for long-term solutions, even curative treatments.

Isaac: There are curative treatments, which we are undertaking. The established one is bone marrow transplant, in which case you need another child within the family that doesn't have sickle cell disease. And in addition, it's a perfect match for the child with the disease. And as you can see, not all families are fortunate to have that. Family sizes are smaller these days. It reduces the chances that another child who doesn't have the disease is a perfect match and can be a donor.
Isaac: Less than 20 percent of our families have that option. And that is why research is now moving into what we call gene therapy, where you don't need a donor. The patient's own stem cells, which has the gene abnormality are collected. They are sent to a manufacturing site where the gene is modified within those stem cells and returned to the patient. That patient's own stem cells are wiped out and replaced by the modified cells. So, this approach really means that you don't need a donor, and everyone can have access to it. It is going through clinical trials. And then there's the CRISPR technology. The discoveries of this process won the Nobel Prize in 2020.

Hannah: CRISPR is a technology that edits genes. A genetic mutation can be replaced with a functional version of the gene by cutting out the DNA and then letting the natural DNA repair itself.
Isaac, did you ever think that you would be sitting here talking to me about something like this? It sounds almost too good to be true and a little bit sci fi.

Isaac No, it's amazing. It's a breathtaking era that we have at this point, with understanding of the ability we have in modifying genes and correcting what has gone wrong and being able to give hope to many, many, many children.

Hannah: The future is looking brighter for children and youth with sickle cell disease, like Camren. In developed health care systems, a lot of progress is being made. But what about elsewhere? Unfortunately, many of the parts of the world where the disease is most prevalent are the least equipped to manage it. Isaac is working to tackle this challenge, too.

Act Four

Chioma: My name is Chioma Okechukwu. I'm a trained paediatrician from Nigeria. I'm currently in my second year of fellowship training at SickKids. I'm doing a subspecialty fellowship in sickle cell disease. Dr. Odame is my boss, and he's my trainer. I had a particular friend of mine in elementary school who was always sick, always out of school, and funny enough, any time she returns to school, she will still do better than most of us who had been in school all the while. I was particularly fond of her. But I really felt sympathy for her because she appeared sick and was always unwell. She was very thin, and so while other children avoided her, I stuck close to her and was her closest friend. When I got into medical school, I got to understand that what she had was sickle cell disease. I found myself naturally wanting to be around sickle cell patients and help them to live their best lives.

Hannah: Chioma, what has it been like working alongside Isaac at SickKids and seeing the kind of diagnosis and treatment that's happening here?

Chioma: It has really been a life changing experience for me. Because like Isaac said, in Nigeria, we see a lot of sickle cell patients and most of them are not diagnosed at birth. They are diagnosed when they show up in crisis somewhere around five to 10 years of age. And most of the preventive care has already been missed. I believe that newborn screening is the beginning of tackling the problem of sickle cell disease in Nigeria.

Also, there's a problem of poverty where patients are unable to pay for access to care. In Nigeria, there is majorly no health insurance, so patients have to pay out of pocket for their clinic appointments and even their medications like hydroxyurea. So, I've really learned a lot about what should be done and what the standard of practice should be. And I believe that there is much that can be implemented back home, even in spite of our limitations.
Hannah: While practices such as newborn screening are now the standard of care in Canada, countries like Nigeria are not there yet.

Chioma: Diagnosis is often not at birth. It's usually at the point of a crisis or hospital admission. And so that point is difficult breaking such news to the patients in the first place. Typically, the child with sickle cell disease in Nigeria is more likely to have the characteristics of sickle cell habitus—that's sickle cell children—can have, if they are if they are not well managed from birth. By habitus, I mean the jaundice, the thin limbs, the prominent forehead or prominent jaws and even poor growth. You see these children looking sick, thin and generally unwell. And also there's this overlap of the burden of malaria on sickle cell disease patients in Nigeria, where they have to cope with frequent malaria infections, unlike children in Canada who don't have to deal with malaria. There's a clear difference in the physical appearance of a sickle cell patient in Nigeria against the ones I see here in Canada. Those who have been well managed from birth in Canada do not look differently from their peers. They grow well. They look healthy. They have easy access to care, and overall, they do better.

Hannah: Although children with sickle cell disease in Nigeria and Canada fare differently in many ways, in other ways they face very similar challenges. One of those key challenges in stigma.

Chioma: So back home in Nigeria, any family who has a child or children with sickle cell disease is seen as being cursed by the gods. And it's even made worse if they should actually lose a child to sickle cell disease. It really is quite bad, especially in rural areas in Nigeria.

Hannah: When we spoke to Camille, Camren's mom, she described a similar challenge existing right here at home.

Camille: One of the social workers—she said there's a lot of support groups that we have for sickle cell, but a lot of parents don't come. And I said, well, why is that? I would think that they would want a lot of information. She said, well, people don't want to show up because they're embarrassed, and they don't want other people in their community to see them with a child that is marked, shall we say. My heart was broken. I mean, that's your baby. Who cares? And she made me look deep and say, well, I'm not walking around telling anybody, but I'm not keeping it a secret. A light bulb went on for me, and I said, oh, my gosh, by not telling anybody, you are keeping it a secret. So I became loud. I became vocal. I told everyone.

Hannah: So much progress has been made, but there is still so much to do. That's why the efforts of doctors like Isaac, parents like Camille and initiatives like the Black Experience @ SickKids are so important. Tackling the greatest challenges in child health cannot be done alone.

Closing

Hannah: From SickKids Foundation, this is SickKids VS. Thanks for listening. If you want to support work like this, visit SickKidsFoundation.com/podcast to donate. And if you like this podcast, please subscribe and rate us on Apple or Google Podcasts, Spotify or wherever you listen to SickKids VS. SickKids VS is produced by me, Hannah Bank, Jasmine Budak, Kate Daley, Emily Holland, Neil Parmar, and Gillian

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