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INSIGHTS

Family reflections—navigating the unknown: our family's Kawasaki disease journey

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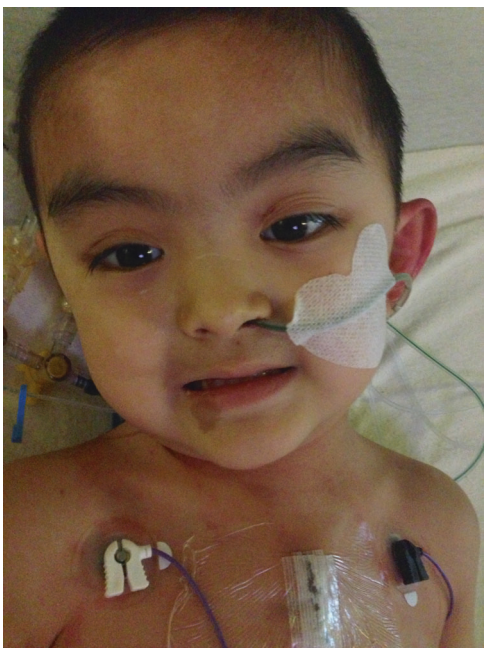
My son, Isaiah, was born with a healthy heart and lived the normal, care-free life of a typical child for 3 years. In 2011, just before his third birthday, an unexpected illness would change all of that.



A week into the start of Isaiah's preschool career he came down with a fever—not surprising, given the circumstances. Isaiah's illness began with a fever, but slowly evolved over the course of the next few days. A lump in his neck, a rash in the groin area, which later spread to his back, unprecedented lethargy, refusal to eat, refusal to walk, extreme irritability and all the while battling an unrelenting fever. We had multiple visits to walk-in clinics, being told it was "just a virus," and then two visits to our pediatrician, who eventually recommended we go to the hospital. Fortunately, Isaiah finally received a diagnosis at The Hospital for Sick Children: Kawasaki disease (KD). We had never heard of this disease before, but were relieved when we were told there was a treatment available. Unfortunately, Isaiah's course of treatment was not a straight path. Isaiah required two treatments of intravenous immunoglobulin (IVIG) and two doses of IV pulse steroids. At one point the rheumatologists started to consider alternate diagnoses, such as juvenile arthritis. After the second IV steroid infusion, Isaiah was started on oral steroids and his fever finally stayed away. We were grateful that Isaiah's first IVIG was administered within the 10-day window that is often referred to with standard KD treatment and were told that the dilatation seen on his echocardiogram, prior to treatment, would likely resolve. We were relieved, happy to finally go home and believed the worst was behind us. Little did we know, the worst was yet to be revealed.

At Isaiah's follow-up echocardiogram 6 weeks later, we received the news that would change the trajectory of Isaiah's life forever. His echocardiogram showed that he had developed multiple giant coronary aneurysms. We were shocked, devastated, and unbelievably scared.

We had no idea what this would mean for Isaiah's future and our world was turned upside down. Isaiah was started on enoxaparin right away. Twice daily injections, multiple oral

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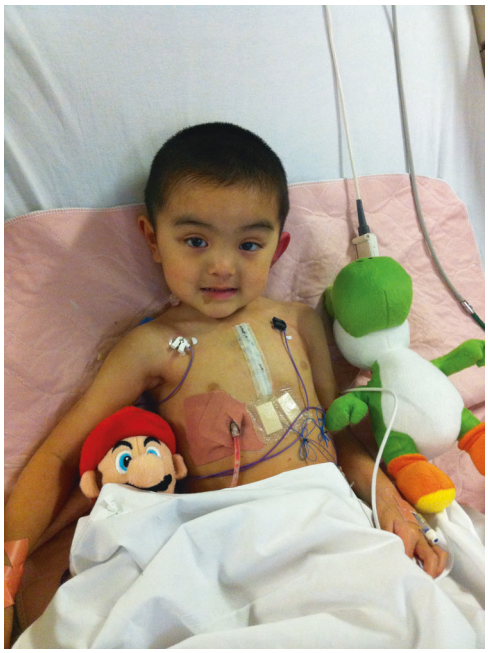
medications, clinic visits and echos every 3 months, and annual magnetic resonance imaging or cardiac cath testing are all now part of his life. Since that day, Isaiah's journey has been a roller coaster ride of ups and downs. He had a second bout of KD in January 2013, but fortunately, with no additional artery damage. That same year, Isaiah experienced chest pain and the results of a cardiac cath determined that immediate intervention was required. He underwent a double bypass surgery 4 days later. More recently, Isaiah was admitted to the hospital for multiple blood transfusions, due to extremely low hemoglobin as a result of uncontrollable nosebleeds. The low hemoglobin resulted in episodes of ischemia, which prompted further investigation.



Shortly after, Isaiah had a stent placed to relieve some narrowing in one of his arteries. Who knows what his future holds?

I have always thought of Kawasaki disease as a disease of uncertainty and unknowns. Most KD parents have never heard of Kawasaki disease until their child is diagnosed with it. Most of us were taken by surprise by this mystery illness that seemingly appeared out of nowhere and plunged us headfirst into a nightmare. A disease we never knew existed, managed to swiftly threaten the health of our children and sadly, permanently changed the lives of some of them. Many families have never fully recovered from the trauma of their child's KD diagnosis. The diagnosis catches parents off-guard. It is something unexpected and unheard of, making it difficult for parents to navigate and advocate for their child, and often makes the diagnosis more frightening compared to illnesses that are more commonly known. For a Kawasaki disease parent, the fear of the unknown is very real.

I live with this fear of the unknown every day. What will Isaiah's next echocardiogram or MRI show? New stenosis? A new clot? Reduced function? How long will his next nose bleed last? Is his antithrombotic therapy balanced enough to protect this heart, but not cause dangerous levels of blood loss? Will Isaiah experience chest pains again? Will Isaiah require another intervention soon? If further intervention is needed, what will be the best option and what are we willing to agree to? Will Isaiah have another heart attack?

For parents whose children have coronary artery complications, the path is uncharted. I have learned to expect the unexpected. Even though our children are monitored closely, we can never predict exactly what will happen next. As parents, we deal with each challenge to the best of our ability, using all the resources at hand. My family is beyond grateful for the outstanding care Isaiah has received from his dedicated and caring cardiology team. We are exceedingly fortunate to live near The Hospital for Sick Children, one of the top pediatric hospitals in the world and to be cared for by world-class medical professionals who truly care about Isaiah and not just his heart. Their support provides confidence and comfort, as we travel through this unpredictable journey.

As Isaiah grows older, his understanding of his disease has changed. We are aware that we need to start preparing for his transition to adult care and it has forced us to come to terms with the fact that we can no longer shelter him from the burden of his disease. We have never wanted him to feel restricted or defined by his heart disease. We quietly steered him away from contact sports and downhill skiing (due to his antithrombotic therapy), but encouraged other activities, like swimming, golf, and tennis. We have not been able to spare him from the countless days in the hospital and unpleasant, sometimes painful medical procedures, but we have tried our best to make the other aspects of his life as "normal" as possible and not allowed him to feel the fear that we feel. While Isaiah's life is not what we envisioned for him when he was born, we hope that he is still able to live fully, or even more so, despite or even perhaps, because of, his additional challenges.

I know all KD parents hope that researchers are able to find answers to some of the many unknowns. From the perspective of a family that has a child with aneurysms, our main concerns are medication, monitoring, and intervention. There are many different anticoagulant, antiplatelet, and other heart medications that are being prescribed to KD patients for long-term use. I appreciate the efficacy studies currently being done for some direct oral anticoagulants in KD patients and hope this type of research continues, in order to provide KD patients with improved medications that will be safe and effective in treating *their* specific concerns, but also improve their quality of life. I would like to see continued advancements in available imaging technology, monitoring strategies, and data collection to elucidate and predict how KD arteries change as children age and ensure that aneurysms can be accurately analyzed and monitored to allow for appropriate intervention at the ideal time. Isaiah has already been through two interventions in his short 10 years of living with his heart disease. I understand that with any medical intervention there is hardly ever a perfect solution, but due to the relatively limited number of KD patients, there seems to be so much uncertainty with regard to which choice of intervention is "best" in certain situations. I am hoping that as more data are gathered, the "best" choice becomes clearer for surgeons and interventionists. My ultimate wish would be advances that provide a way to repair Isaiah's damaged heart muscle (due to a previous infarct) and normalize his arteries. I recognize this type of discovery, may be much farther into the future, so for now, I wish for new interventions that are effective, specifically in Kawasaki disease patients, whose vessels may not be like that of a typical heart disease patient.

In my role as the Chair of the Support Committee at Kawasaki Disease Canada (kdcanada.org), I hear the concerns of many other Kawasaki disease parents, primarily whose children do not have coronary involvement. While their fears may differ from mine, they are equally as valid.

Overwhelmingly, the most common concern is what possible long-term effects Kawasaki disease may have on their children that have yet to be determined. Are these children more at risk for heart disease later on in life? If they have been fully discharged, do they still need their heart health monitored? Are there other long-term effects related to inflammation, joint pain, mental health, or cognition? Or are there health issues that a child may develop as a result of having had Kawasaki disease? It may be unreasonable to link every future ailment to a past KD diagnosis (realizing that

correlation does not imply causation), but with so many unknowns surrounding Kawasaki disease, it is difficult for parents who have been through that unexpected, and often traumatic, diagnosis to put their worries to rest. Research that uncovers some of these unknowns would surely bring these parents some peace of mind.

A definitive and accessible diagnostic test for Kawasaki disease, a reliable way to identify the most at-risk children, and effective individualized treatments for patients would save parents from heartache and many children from potential heart damage. I know these are lofty goals and in order for them to be achieved many KD unknowns will need to become "knowns." The ultimate unknown to conquer, and likely the holy grail of Kawasaki disease research, would be determining the cause of Kawasaki disease. My hope would be that if a cause was determined, a way to prevent Kawasaki disease would soon follow. So many little hearts would be spared from damage and no more children would have their lives permanently changed. No more parents would mourn the loss of their child as a result of this disease. No more families would be plagued by the fear of the unknown. No more children would have to live with the burden that I wish I could take away from Isaiah every day.

I know my list is long and that the science is complex, but I remain hopeful. I recently had the privilege of attending the 13th International Kawasaki Disease Symposium and was exceedingly impressed and extremely grateful to see all the ongoing research that is being done by investigators across the globe. Knowing that so many amazing minds are dedicating their time, and for some, almost their entire career to researching Kawasaki disease gives me great hope, not only for Isaiah, but for all Kawasaki disease patients: past, present, and future. On behalf of all Kawasaki disease families, I thank you for your perseverance and dedication to unraveling the mystery of a disease that quietly has had a profound effect on families around the world. Your research may not have the prestige or funding that is seen in other pediatric diseases, but please know that the work you do is appreciated by KD families more than we can ever express. Many had lost faith in the medical community after struggling to get their child diagnosed; we thank you for working tirelessly to restore that faith. Dr. Kawasaki was passionately committed to his patients and to finding a cause for the disease that bears his name. I am optimistic that as each piece of the puzzle is discovered and each unknown is revealed, the Kawasaki disease research community will come full circle to finish the work that he started 60 years ago.

COMPETING INTERESTS

The author declares no competing interests.

ADDITIONAL INFORMATION

Correspondence and requests for materials should be addressed to Carin Lin.

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