

Laney's Story: The Problem of Delayed Diagnosis of Pediatric Stroke

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Childhood ischemic stroke is rare, with an incidence between 1.2 and 8 per 100 000 person-years.^{1,2} As a result, stroke is often not initially considered when children present even with classic signs of ischemia, leading to significant diagnostic delays.³⁻⁷ Given the time dependence of stroke treatment, even short delays in diagnosis may lead to poorer outcomes.

Parent advocacy groups and collaborations of pediatric stroke specialists work to improve recognition of pediatric stroke through research, social media campaigns, and education. These groups are often born of tragedy: a patient suffers a devastating stroke, and a physician turns her efforts to champion the field while a parent rallies families to advocate for improved recognition of a shared diagnosis. In this case, the death of a bright, beautiful 4-year-old girl inspires a family to dedicate her legacy to this noblest of causes: spreading her story so that someone else's child may live.

LANEY'S FATHER

Recounting the days of Laney's first transient ischemic attacks (TIAs) and strokes and the horror and confusion that accompanied that time is something I would never wish on any parent. But if this article can help even 1 child receive a faster stroke diagnosis, then it is worth every painful word that I type.

First, I want you to know Laney. She had flowing auburn hair, an infectious smile with a gap in her front teeth, and

an uncanny elegance for a 3-year-old girl. But a 3-year-old girl she was. She loved jumping in muddy puddles, "flying" around the house as Supergirl, and running around in circles until she was dizzy. She could put together a puzzle faster than some adults I know, manipulate a chocolate from her enchanted parents, and melt your heart with her big brown eyes. She was our whole life. In our minds, we were the happiest family on Earth.

It all began on May 7, 2016. We had recently returned from vacation, where Laney hiked miles every day in Sedona, Arizona. We will always remember these days as our best days. Laney, at 3.5 years old, was taking a bath when she suddenly started speaking with a slight slur. It was subtle, and we attributed it to her being tired.

Two weeks passed. We were in the kitchen when Laney started speaking with a slur and developed a facial droop. We called 9-1-1, and Laney was taken to a local emergency department (ED) where the doctor suspected she was having seizures. Laney was transported to another hospital where an overnight EEG revealed "seizure-like" activity, and the doctors recommended an MRI. Because this was a Sunday morning, with no anesthesiologists available, we were given the choice of keeping Laney in the hospital until Monday or taking her home and scheduling the MRI soon. We took Laney home to save her from the distressing hospital setting. There was no mention of stroke during either of these hospital visits.

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Three days later, Laney experienced the same symptoms (slurred speech, drooping mouth and face, and trouble walking) but more severe. We rushed her to a different ED and showed the doctors and even a neurologist a video of the event. She was again diagnosed with seizures and discharged with a prescription for oxcarbazepine. There was no mention of stroke.

A week later, at Laney's first neurology appointment, we showed the neurologist the video and described her symptoms. The neurologist diagnosed Laney with complex partial seizures and told us to continue oxcarbazepine. He said we could wait 3 months for the MRI to give Laney a break.

LANEY'S NEUROLOGIST

Months before I met her, Laney had a series of TIAs consisting of dysarthria, facial droop, and inability to walk. In any adult, these would have been immediately recognized as a warning sign of impending stroke. But because Laney was 3 years old, they were misdiagnosed as complex partial seizures.

In 1 study, 4% of children admitted with a diagnosis of TIA had a secondary diagnosis of stroke within the same hospitalization.⁸ This is concordant with data that 1% to 10% of adults who experience a TIA will have a stroke within the next 48 hours.⁹ Despite the importance of recognizing TIA, mimics of TIA or stroke are substantially more common than ischemia in the pediatric population, and they can be easily misdiagnosed as complex partial seizures, postictal Todd's paralysis, complicated migraine, or functional disorders. In any child with a new-onset focal neurologic deficit, even if preceded by a first-time focal seizure, high suspicion for a vascular cause is imperative.

LANEY'S FATHER

Tragically, Laney had a severe reaction to oxcarbazepine. We took her back to the hospital, where doctors, in consultation with her neurologist, gave her lorazepam instead and discharged her.

After the hospital, we took Laney to the nearby Chuck E. Cheese's. We ate pizza, took pictures, and played games. We won more tickets that day than we had in all our other visits combined. We were deliriously happy, if just for a moment. That was the last time we ever spent with our healthy little girl. I think about those joyful hours every day.

The following morning, almost a month since Laney's initial symptoms, Laney woke up unable to stand, speak, eat, or drink. She wanted so badly to communicate, but the words would not come out as tears rushed down her cheeks. We called 9-1-1. The medics arrived as Laney was writhing on the floor. They did not consider stroke but instead administered epinephrine. In hysteria, Laney was transported back to the hospital. She was unable to stand or speak. Her heart rate climbed to ~190 beats per minute, and I ran throughout the ED floor looking for a doctor to help Laney, who appeared to be dying in front of our eyes.

Our little girl was not talking and was barely moving. Laney was transferred to a larger hospital where doctors concluded that she was tired from her seizures. She had high blood pressure (which we now know was to help perfuse blood to her brain to prevent stroke), and they gave her medicine to lower it. Again, because no anesthesiology staff was available over the weekend, they could not do an MRI. There were no wheelchairs available, so I carried Laney's limp body across the hospital to radiology for a head computed tomography scan. I will never forget that long walk.

Finally, on Monday, June 6, 2016, Laney underwent an MRI. The doctors informed us that Laney has suffered multiple acute strokes and had severe brain damage. This was the first time we had heard the word "stroke" since Laney's symptoms began. We were devastated and frantically wanted to help her. We did not yet realize that this damage was permanent and that the Laney we knew and loved was gone.

Over the next months, Laney continued to have strokes as doctors struggled to stabilize her. Those days were filled with innumerable horrors. We never left Laney's side and did not leave the hospital grounds for the entire summer. After months in the hospital and brain surgery, Laney was transferred to inpatient rehabilitation. She worked valiantly in speech, occupational, and physical therapy. Laney's therapists were wonderful, and she began to laugh, smile, and regain strength. Laney was discharged that fall and continued in intensive outpatient rehabilitation. She worked so hard and even learned how to communicate with buttons and sit up on her own. Laney would even hold her little sister, whom she loved fiercely.

LANEY'S NEUROLOGIST

I only met Laney after her prolonged hospitalization and inpatient rehabilitation, but she captivated me immediately despite being devastated by moyamoya disease, a cerebral vasculopathy that, in Laney's case, was relentless and caused multiple strokes that stole her ability to move and speak. Reviewing Laney's course, it was easy to identify in retrospect the delays in stroke recognition and cognitive biases, including anchoring, that contributed to the delayed diagnosis. But hindsight is 20/20, and given how common seizures are in children, it is not hard to imagine a thoughtful, capable pediatrician or

neurologist making the misdiagnosis. Laney's case is unfortunately not an exception. Multiple studies have revealed that the average time to diagnosis of childhood ischemic stroke is >22 hours.³⁻⁷ In 1 study, arterial ischemic stroke was only considered as a possible diagnosis in a quarter of cases of childhood stroke despite focal deficits being a part of the presentation in 86% of cases.⁷

Even if the need for neuroimaging is recognized, in most centers, MRI is not available around the clock. It is even less common for centers to have emergency pediatric anesthesiology available, a necessity when the duration of an MRI far exceeds the time a child, particularly one in distress, can remain still. As a result, it is not surprising that presentation outside of normal working hours leads to delayed neuroimaging and diagnosis.⁶

Early diagnosis of stroke is critical for early implementation of neuroprotective measures, assessment for candidacy for hyperacute therapies (such as mechanical thrombectomy and intravenous thrombolysis), avoidance of harmful medications (such as antihypertensives), treatment of risk factors for stroke recurrence, and early implementation of rehabilitative therapies. Fortunately, primary pediatric stroke centers are emerging as centers with around-the-clock ability to rapidly diagnose and treat pediatric strokes.¹⁰ Emergent transfer of any child with suspected acute ischemic stroke to a primary pediatric stroke center should be considered when safe and feasible.

LANEY'S FATHER

Tragically, in December 2016, Laney suffered another stroke. After multiple failed medical interventions and visits to several medical centers, Laney was placed on hospice care. Our little girl finally succumbed and died on March 27, 2017. We kissed

her little face and body until the second the funeral-home transport took her away.

LANEY'S NEUROLOGIST

Brian and Laura were and still are Laney's fiercest advocates. In the darkest of moments, they mourned her loss while simultaneously celebrating her beautiful life. I learn from their courage, revel in my fortune in having been a part of Laney's life, and take solace in the bright eyes of her little sister gazing at me from a Christmas card pinned at my desk. To this day, they tirelessly fight for the victims of childhood stroke, and it has been a privilege to join them in this journey.

PARENT-PHYSICIAN PARTNERSHIP

During Laney's life, our partnership was focused on managing the complications of Laney's brain injury, optimizing her rehabilitation, preventing further strokes, and, eventually, keeping Laney comfortable at home. Our partnership, forged by our joint goals and mutual adoration for Laney, continued after her death, this time fueled by the "what ifs" and "should haves" of her life.

Together, we testified as a united family and physician team to the Maryland House of Delegates and Senate, along with others affected by pediatric stroke, to advocate for legislation targeted at improving recognition of childhood stroke. With a beautiful poster of Laney between us, her father powerfully depicted the toll of pediatric stroke on their family, bolstered by medical facts and statistics from her doctor. If it is passed, the proposed bill will mandate education about pediatric stroke for licensed child care providers, school nurses, and directors of student services in local education agencies.

Laney's parents established the Laney Jaymes Foundation in 2017 with the missions of expanding pediatric stroke awareness, supporting research, and providing adaptive equipment to children who have suffered from stroke. Together, we identify clinical and research priorities that merge the goals of physicians and scientists with those of patients and families. For example, with guidance from Laney's doctors, the foundation is actively funding research to better understand childhood cerebral vasculopathy, the most common cause of childhood stroke, and specifically moyamoya disease, the type of vasculopathy from which Laney suffered. The foundation both funds the research and helps guide its trajectory (with consistent collaboration between parents and physician scientists), incorporating both viewpoints and sets of priorities as we work toward the common goal of helping children like Laney.

The joint family-physician voice has the potential to be incredibly impactful. With Laney's inspiration, we continue to work together to make meaningful strides in research, education, and legislation to improve the lives of children with stroke.

ABBREVIATIONS

ED: emergency department
TIA: transient ischemic attack

REFERENCES

1. Felling RJ, Sun LR, Maxwell EC, Goldenberg N, Bernard T. Pediatric arterial ischemic stroke: epidemiology, risk factors, and management. *Blood Cells Mol Dis*. 2017;67:23-33
2. Lynch JK, Nelson KB. Epidemiology of perinatal stroke. *Curr Opin Pediatr*. 2001;13(6):499-505
3. Rafay MF, Pontigon AM, Chiang J, et al. Delay to diagnosis in acute pediatric arterial ischemic stroke. *Stroke*. 2009; 40(1):58-64

4. Gabis LV, Yangala R, Lenn NJ. Time lag to diagnosis of stroke in children. *Pediatrics*. 2002;110(5):924–928
5. McGlennan C, Ganesan V. Delays in investigation and management of acute arterial ischaemic stroke in children. *Dev Med Child Neurol*. 2008;50(7):537–540
6. Mallick AA, Ganesan V, Kirkham FJ, et al. Diagnostic delays in paediatric stroke. *J Neurol Neurosurg Psychiatry*. 2015; 86(8):917–921
7. Srinivasan J, Miller SP, Phan TG, Mackay MT. Delayed recognition of initial stroke in children: need for increased awareness. *Pediatrics*. 2009;124(2). Available at: www.pediatrics.org/cgi/content/full/124/2/e227
8. Adil MM, Qureshi AI, Beslow LA, Jordan LC. Transient ischemic attack requiring hospitalization of children in the United States: kids' inpatient database 2003 to 2009. *Stroke*. 2014;45(3):887–888
9. Wu CM, McLaughlin K, Lorenzetti DL, Hill MD, Manns BJ, Ghali WA. Early risk of stroke after transient ischemic attack: a systematic review and meta-analysis. *Arch Intern Med*. 2007;167(22):2417–2422
10. Bernard TJ, Rivkin MJ, Scholz K, et al; Thrombolysis in Pediatric Stroke Study. Emergence of the primary pediatric stroke center: impact of the thrombolysis in pediatric stroke trial. *Stroke*. 2014;45(7):2018–2023