

“It Occupied Her Entire Childhood”: Looking Back on Limb-Lengthening

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Approximately 1 in 1000 children are born with congenital lower limb deficiencies related to underlying congenital vascular insufficiency.¹ Their parents face a difficult choice: early amputation and prosthesis-fitting or a series of painful, invasive lengthening procedures, which, despite some advances, still carry a risk of a poor functional outcome for a significant minority of patients.¹⁻³ In this Family Partnerships essay, a young woman born with congenital short limb, her mother, her current orthopedic surgeon, and a pediatric ethicist reflect on their family's choice and its consequences for her health and wellbeing over the past 30 years.

RENE MAUCHIN, PARENT

Lindsay is the younger of our two daughters. I had been advised to stop taking antiepileptic medications during my pregnancy, but this led to my having more frequent and severe seizures. Sometimes I fell but remained conscious. Although 3 doctors have told us that Lindsay's disability was unrelated, I still feel guilty that I might have caused it. Regardless of what got us into this position with our little girl, we were always going to do everything on heaven and earth for her.

The day after Lindsay was born in 1987, the pediatrician came to my room with the longest face I have seen a doctor have before. Until that moment, I had been excited to see him bring the baby in. I knew immediately something was wrong. He started talking, and I heard nothing. It was like buzzing in my ears. I was 23 years

old. The room was spinning and I did not understand any words he was telling me. “Breathe,” he said. “Do you know what a prosthesis is?” “No,” I replied.

LINDSAY ELLINGSWORTH, PATIENT

When I was born, a nurse noticed my left leg was significantly shorter than my right, by ~8 cm. The doctors figured it was from a break that happened in womb during development. Then they found that I also had never developed a knee joint and had very little hip mobility.

Despite this, I was raised like any other child and never felt that I was different or was treated differently. This is how I was born; I did not know anything else. As a kid, I did not think, “Why do I have a lift on my shoe?” When my parents bought me a bike for my fifth birthday, I hopped right on it and rode it with one leg without ever being taught how. It was natural adaptation. I think little kids do not ask, “How am I going to do this?” It felt natural because that was all I knew. Looking back, I think it is amazing how kids can adapt when they are not treated as if something is wrong with them.

RENE MAUCHIN, PARENT

Our first pediatrician referred us to a local orthopedic surgeon, who saw Lindsay when she was 4 months old. He told us that the best option was amputation with a prosthesis at age 2. As a young mother of a new baby, this was horrifying. The doctor showed us the radiographs and explained that

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Dr Hootnick coconceptualized the manuscript, led the review of the literature, drafted his individual sections, and revised the introduction and conclusion to reflect his perspective as an orthopedic surgeon; Ms Ellingsworth and Ms Mauchin drafted their individual sections and revised the introduction and conclusion to reflect their perspectives as patient and parent; Dr Caruso Brown coconceptualized the manuscript, drafted the introduction and her individual section, and organized, edited, and revised the entire manuscript; and all authors approved the final manuscript as submitted and agree to be accountable for all aspects of the work.

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there would likely be an 8- to 10-cm discrepancy. He explained that there was a new surgery that would stretch the bone but that it had limited success and that Lindsay's case was too "radical" for this surgery.

We agonized for the first few months and decided to seek a second opinion at a large children's hospital when Lindsay was 9 months old. Although we were not fully aware at the time, they recommended lengthening surgery. Soon after, Lindsay started to walk, and not long after that, another surgeon (Lindsay's third; he would become the first to operate on her) joined the local practice. This new surgeon had performed many lengthening procedures and written articles about them. Our first surgeon said we were lucky to have him here. He explained that Lindsay would need two surgeries and likely still need a lift shoe, her discrepancy was substantial. There was also discussion about slowing down the growth in the opposite leg.

DAVID HOOTNICK, MD, ORTHOPEDIC SURGEON

My personal experience with leg-lengthening procedures began during a pediatric orthopedic fellowship at the Hospital for Sick Children in London, researching leg-lengthening inequality in the 1970s. There, I examined firsthand the survivors of the Wagner technique of leg lengthening, which, by that time, had already been abandoned in the United Kingdom because of the surfeit of complications encountered. I began publishing on the subject in 1977. Thereafter, my patients were treated with prosthetics only or by amputation of the foot through the ankle with prosthetic-fitting around the time of ambulation, because I saw better outcomes with this approach.

Lindsay was diagnosed with 30% congenital shortening of her left femur at birth, which persisted as she grew. The severity of the discrepancy

made her a nonideal candidate for lengthening: the likelihood of complications increased with each centimeter of length gained. Sadly enough, however, the more severe the shortening of the affected femora, the more normal are the feet in these patients.³ Lindsay had a normal-appearing foot, making it all the more understandable that her parents balked at removing an apparently healthy part of their beloved child. When a 2000 study of fibular hemimelia, for example, reported that children who had undergone amputation were more active, with less pain and greater satisfaction, and had undergone fewer procedures with fewer complications and lower medical costs than those who underwent lengthening, proponents of the lengthening procedures responded that amputation was an "irreversible mutilating procedure."⁴ In my experience, many surgeons develop preferences of their own, weighing innovation more heavily than evidence and pursuing mechanical solutions to biological problems, and these can be obstacles to meaningful shared decision-making.

LINDSAY ELLINGSWORTH, PATIENT

Everything changed when I turned 8 and went through a living nightmare: my first lengthening surgery. To lengthen the limb, surgeons have to break the affected limb with a subtle "thud" of a hammer. My affected limb was my femur, which is rare; >50% of these cases are done on the tibia-fibula. The femur is the largest bone in our bodies. Imagine having that bone broken and then having 3 screws drilled into the top of the limb and 3 into the bottom. I still remember waking up in recovery and screaming. I was in so much agony, and I kept wondering, for the first time in my life, "What is wrong with me that I would need to endure this much pain?"

I understand now why my parents were not able to prepare me for this experience. They did not understand what this procedure was going to be like any better than I did. They were young, with very little medical knowledge. All they knew was that the lengthening procedure had been "talked up" as the very best option. Limb lengthening dramatically interrupted my childhood. I had to relearn how to adapt to my condition. I was restricted and held back by an external fixator containing 6 screws, for what would feel like most of my childhood. Every day for 8 months after the first procedure, I took an Allen wrench and rotated the 6 screws in my external fixator a quarter of a turn. Eventually, I did this 3 times a day, separating the bone on each side of the break to allow new bone to grow in-between. I endured excruciating pain that woke me screaming during the night, side effects from pain medications that never seemed to work anyway, muscle spasms, frequent dressing changes, having to wear dresses even if it was below 0, endless questions from other kids, stares from adults... the list is almost endless. I stopped wanting to go to school. On Halloween, people asked me, "Is this part of your costume?" and said, "Wow! You did great on your costume makeup." I wondered how I could really look that out of place. On the worst nights, I cried from muscle spasms unless I fell asleep sitting upright.

Eventually, my doctor realized that the limited range of motion in my hip caused my muscles to tense, which led to excruciating pain, and recommended lengthening my hamstrings, which took place 6 weeks after my first surgery. I gained 6 cm in femur length and the external fixator was finally removed. My second lengthening, 3 years later, went more smoothly, until the day the external fixator was removed. I still had internal screws, and there was

a chance my femur would fracture while waiting for the screws to be removed. That night, I turned just right in my sleep. As I screamed, my dad struggled to reapply the external fixator. The pain was indescribable. By then, I had learned to fear and dread surgery, especially the inevitable risk of complications leading to more pain and more surgery.

DAVID HOOTNICK, MD, ORTHOPEDIC SURGEON

I first met Lindsay ~10 years ago, when, as a young nursing student, she was referred to me for a second opinion regarding her ongoing left leg problems. Her knee was painful and unstable, and her left leg was, and still is, scarred and atrophic, ~2 in shorter than her right. When I heard that she refused to walk with a shoe lift, I wrongly assumed it was because vanity. I quickly realized my mistake: Lindsay's knee is so stiff that her leg needs to be shorter to allow her to swing it in the absence of being able to bend it. However, her gait is still lurching, and each step twists her spine, resulting in lumbar and cervical imbalances.

At our first meeting, I unsuccessfully urged Lindsay to reconsider her intended career in nursing for something more sedentary. Unfortunately, over the next several years, Lindsay returned to me several more times to perform additional procedures to address her ongoing complications and chronic pain, as she describes in the next section, and she was eventually forced to leave nursing and apply for Social Security disability benefits. For Lindsay, the results of the lengthening procedures on her left leg are nothing less than catastrophic.

LINDSAY ELLINGSWORTH, PATIENT

Because of those complications, I now have severe scoliosis. I eventually underwent 13 surgeries, including

two major neck surgeries as a young adult. I receive injections bilaterally every 3 to 6 months so I can walk. I cannot have children; I lost a child at 5 months because of my anatomy. I can no longer work and left a career in nursing, something Dr Hootnick anticipated when we first met, because of my back pain. Frustration is a word I use a lot. I get mad at myself for not being able to do certain things. I think about what might have happened if my parents had chosen amputation for me instead of limb lengthening, but I will never know what that road would have been like. I think the doctors put a pretty bow on limb lengthening, but it was nothing short of a nightmare.

RENE MAUCHIN, PARENT

Although we thought we understood the possible outcomes and risks, we never realized just how many surgeries Lindsay would need to address unexpected complications (hamstring lengthening, fractures after hardware was removed, problems with growth plates, scoliosis, and neck and back pain) and how difficult it would be to treat her pain. With each new complication, our doctors always focused on how to fix it and how to get to the goal, even when the goal seemed to be moving further from our reach. We did not know that stopping was possible: we were never asked if we wanted to stop or told what that would look like. We prayed with everything we had that lengthening surgery would positively change her life. I believe everything she has been through has made her appreciate how difficult this decision was for everyone. Through her pain, I also see how she is able to empathize with other people. Although she has endured so much, Lindsay still laughs and celebrates life. The only disability Lindsay truly has is in the eyes of others. Lindsay is my heart. If I can do one last thing for her to make her life better, I will.

AMY CARUSO BROWN, MD, MSC, MSCS, PEDIATRICIAN AND BIOETHICIST

Two years ago, a colleague reached out to me to ask if I would be willing to talk with an orthopedic surgeon who was interested in writing about decision-making around limb lengthening. After meeting Dr Hootnick, it was clear that his interests were deeply driven by his relationship with Lindsay and her family, and I encouraged him to ask her if she wanted to write about her own story. She did, and a few months later, we all sat down together to begin the conversation that eventually became this essay.

Lindsay's story embodies one of the most challenging decisions in pediatrics: a decision that must be made before a child is old enough to express any preferences and for which there are two or more choices deemed to be within the zone of parental discretion but that will irrevocably change the course of that child's life, as Lindsay vividly describes. In cases like these, there is not an obvious "right" answer, and as Dr Hootnick and Ms Mauchin both note here, it is difficult to accept that a procedure that sounds as drastic and anachronistic as amputation might have fewer complications than the seemingly more sophisticated alternative.

In an ideal scenario, Lindsay's doctors would have ensured that her family fully understood the risks, benefits, and alternatives to each of the available options, but even then, a challenge remains. How do clinicians avoid unduly weighting a much-hoped-for best-case scenario in our decision-making, and how do we help families do the same? Given what we know now about opioid exposure in childhood, how does that affect how we counsel families regarding paths that may involve long-term pain? We have to ask parents, and children when they are old enough to participate, to envision

the worst-case scenario as well, asking themselves, “If neither path produces the desired outcome, which decision can we best live with? Which set of complications is more acceptable to us? Will it be harder knowing that we tried something innovative and it failed, or that we took a more conservative approach?”

These are difficult questions, to be sure, and ultimately, families rely on their doctors to help them navigate the tension between caution and innovation.⁵ As we worked on the essay, I asked Ms Mauchin what she would want other families to take from hearing this story. She wrote back, “I don’t know if we did do the right thing. To other parents facing this situation, I would say this: You

have a right to know everything the doctors know. When you make a decision, make sure you’re making it for the right reason: what is best for your child. Don’t hesitate to see several doctors and ask for evidence to back up their recommendations. Speak up. You have the final say.”

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REFERENCES

1. Hootnick DR, Vargesson N. The syndrome of proximal femur, fibula, and

midline metatarsal long bone deficiencies. *Birth Defects Res.* 2018; 110(15):1188–1193

2. Hasler CC, Krieg AH. Current concepts of leg lengthening. *J Child Orthop.* 2012; 6(2):89–104
3. Hamdy RC, Bernstein M, Fragomen AT, Rozbruch SR. What’s new in limb lengthening and deformity correction. *J Bone Joint Surg Am.* 2017;99(16): 1408–1414
4. McCarthy JJ, Glancy GL, Chnag FM, Eilert RE. Fibular hemimelia: comparison of outcome measurements after amputation and lengthening. *J Bone Joint Surg Am.* 2000;82(12): 1732–1735
5. Csoka AB. Innovation in medicine: Ignaz the reviled and Egas the regaled. *Med Health Care Philos.* 2016;19(2): 163–168

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