Kasey Shakespear
Ways and Means Committee Hearing
July 12, 2024

My name is Kasey Shakespear, I am an Assistant Director for the Utah Center for Rural Health at Southern Utah University. I am here today to discuss my family's journey with medical innovation and the limitations of living in a rural community. The views that I will share are personal and not representative of SUU.

In July 2021, my wife and I were expecting our 3rd child. We found ourselves facing the unthinkable during an OB appointment at 19 weeks gestation. Our unborn son was diagnosed with Bilateral Renal Agenesis formerly known as Potters Syndrome. BRA is a congenital condition in which the fetus does not develop kidneys. BRA occurs in approximately 1 in every 4,500 live births. At the beginning of the second trimester of pregnancy, the fetus' renal system becomes active and takes over the production of amniotic fluid. In the absence of kidneys to generate amniotic fluid, anhydramnios occurs. This is a complete lack of fluid in the womb.

BRA is considered by medical text as a universally fatal condition with no treatment options. 1 in 3 BRA babies die before birth usually due to pinching off the umbilical and blood supply from the mother. The rest will suffocate and die in the minutes and hours after birth due to critically underdeveloped lungs. When we received the news, my mind immediately went to what cemetery we would bury him in, and how we would explain the situation to our two older children. My wife on the other hand had already begun researching the condition before the visit.

She had discovered an innovative clinical trial happening at a handful of centers around the country called the Renal Anhydramnios Fetal Therapy (RAFT). Originating from Johns Hopkins, the RAFT Trial was an innovative treatment program involving serial amnioinfusion of fluid consisting of saline and antibiotics into the womb done with ultrasound-guided needles. The closest centers to us that offered RAFT were in Denver, Colorado 630 miles away, and Pasadena California 380 miles away. We chose to go to Pasadena and meet with Dr. Ramen Schmidt and Dr. Martha Monson at LA Fetal Surgery.

My wife and unborn son were screened and deemed to be eligible to be enrolled as a fetal-maternal pair for the trial. We didn't know it at the time, but they were 1 of 20 fetal maternal pairs selected, out of over 600 BRA cases screened. However, Dr. Schmidt warned us that pursuing this treatment would be a very long road. If we proceeded, he warned it could threaten our marriage, my career, our financial stability, our home, and the wellbeing of our other children. And that the treatment was still a Hail Mary with a small chance of success. We considered those factors and decided to pursue treatment.

Doing so required us to temporarily relocate from rural Utah to Pasadena, California. Over the next 11 weeks, my wife received 11 serial amnioinfusions. At 31 weeks gestation, my wife's sac began to leak, and we had to halt treatment. The original medical plan was for my son to be

cared for at Children's Hospital Los Angeles, but he was measuring too small to have a peritoneal catheter placed for PD dialysis. We found ourselves suddenly forced 360 miles north to Palo Alto California, just one week before my son's birth at Lucile Packard Children's Hospital at Stanford.

In late 2021, my son was born with Apgar scores of 9 and 8. The amnioinfusions had been successful, but over the next five and a half months, my son battled for his life daily in the NICU. He was placed on hemodialysis two days after birth with an Aquadex machine developed for adults with congestive heart failure and adapted for dialysis on neonates. This innovation allows babies that are too small for PD catheters to instead have a smaller tunneled central line placed, which cycles the blood directly from the bloodstream, into the machine to be cleaned, and then returned to circulation. Our son remained on Hemodialysis daily until he was three months old, at which point, a PD catheter was placed, and he eventually transitioned to Peritoneal Dialysis at 4 and a half months old. The significance of this transition is that PD dialysis can be performed at home, whereas Hemodialysis cannot.

Our son went through blood pressure regulation issues stemming from the absence of kidneys. These issues might have required constant use of multiple daily medications to maintain appropriate blood pressure. However, our son was treated with Angiotensin-II at four months old, becoming the first neonatal kidney patient at Stanford ever to be treated with this innovative medication. Within 10 days, his blood pressure stabilized, and he was able to be removed from the medication altogether and has maintained healthy blood pressure without medications ever since. Through his stay in the NICU, he contracted a serious infection, underwent multiple high-risk surgeries, experienced seizures, and overcame it all.

At 6 months old, my son was transported via air ambulance to Salt Lake City, where he was admitted for two weeks at Intermountain Health's Primary Children's Hospital. During that time, my wife and I were trained in how to perform all necessary functions to care for our son and administer dialysis on a nightly basis at home. Settings for his PD dialysis machine are transmitted electronically directly from the provider, and the data from each round of dialysis can be transmitted back, allowing us to work in tandem with our dialysis team located 310 miles away to constantly monitor and adjust the dialysis as needed. He was discharged from the hospital for the first time at six and a half months old. He is one of approximately 20 children who have survived BRA and made it home.

It has been just over two years since our son was discharged and over that time, he has been readmitted five times, for stays varying from 3 days to a little over 3 weeks. Each stay required a life flight from St. George due to the complexity of his condition. Our local hospital cannot perform his dialysis care, which means he cannot be admitted here for any reason. In all, our son has faced certain death on five occasions, including most recently in April 2023. He was admitted and transported to Salt Lake City with an unknown virus causing respiratory distress. The illness appeared minor, and precautions were taken to keep him stable. I remained in St. George the morning he was life-flighted, preparing to travel up the following day with my older children. My wife called me that afternoon in tears telling me that I had to come now, that our

son was not going to make it. I drove the 4 and a half hours to Salt Lake City, my wife on the phone, racing to be by my son's side to say goodbye. I don't know how, but I made it to the hospital safely, and my son pulled through despite going into cardiac arrest the following morning, and weeks of intensive care. These types of frantic trips to the hospital are not something most urban counterparts will ever experience.

Our son now has numerous specialty providers including nephrologists, urologists, cardiologists, pulmonologists, neurologists, gastroenterologists, immunologists, and transplant surgeons, all of whom are in Salt Lake City. The only provider he can see locally is his primary care provider and his neurologist who travels down to see patients once a quarter. Although telehealth has helped connect with some specialists, we have also traveled 9 hours round trip from St. George to Salt Lake City every month to attend mandatory in-person dialysis appointments and other specialty visits. We make these trips, rain, shine, or in traffic. We even traveled home during a blizzard that created white-out conditions making it nearly impossible to see and drive. Our little Isaac is now nearly 3 years old and active on the kidney recipient lists in multiple regions. We hope to receive a call any moment to bring him in to receive his kidney. He will become just the second male BRA survivor to receive a kidney transplant With so few who have gone before him, it's impossible to understand the long-term ramifications of our son's condition, but we know for certain that he will continue to rely on innovations in treatment throughout his life.

My wife and I have walked through hell to save our son, and we were only able to do so because the stars aligned for us. We received tremendous support from family, friends, members of our religious congregation, and strangers. We found RAFT and were able to navigate the American healthcare system because of our education, we were able to overcome the costs of treatment thanks to insurance, the ability to continue working remotely, and the generosity of so many. And we continue to receive adequate care thanks to programs such as Utah Medicaid's Medically Complex Children's Waiver Program. We didn't have farm animals, pets, crops, or other responsibilities that prevented us from being absent for long periods.

Our story stands as a testament to the innovations and technology that our healthcare system has to offer. According to the HRSA, a little over 25 million rural Americans live in Health Professional Shortage Areas. These individuals struggle to access basic care, let alone specialty care or the type of treatment BRA and other rare conditions require. Innovations in healthcare brought about lifesaving procedures for my son, but my family's opportunity is not representative of so many others. My wife receives messages on social media from frantic parents trying to find options for treatment for BRA and other similar conditions on almost a weekly basis. Many simply cannot make it work to pursue care, usually because of the logistical challenges and economic burden incurred to pursue treatment.

Put yourself in their shoes. The economy of rural America is typically labor-intensive jobs, which cannot be carried out remotely. Typically, these are hourly jobs, with no paid time off. Rural folks often have livestock and pets, even if it is not their primary occupation. They grow gardens to supplement their food supply and do whatever it takes to get by. For those lucky enough to own homes, many would not be able to purchase their own homes at today's market value. When

these individuals face serious illness, they often must travel several hours, if not more, to find care. For the rarest conditions, they may have to traverse multiple states. Bear in mind, that not all urban cities are created equal, the care we needed for Isaac wasn't available anywhere in Utah. To pursue care in these circumstances, when the outlook for recovery is bleak even with treatment, simply cannot be done.

I applaud this committee for your efforts to improve care, especially in rural communities. I want you to know that they make a difference for families like mine. But I implore you to keep working to ensure that every American has an equitable opportunity to receive the innovative care they need in their most dire circumstances. Thank you.