Every Three Hours: 
The Walk to Independence

Janet Butcher Shaw

Faculty Advisor: Dr. Susan Thorne
History Department

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Abstract

“Every Three Hours” is a memoir about raising my son Patrick who has two rare medical conditions--Glycogen Storage Disease 1a (GSD1a) and Mast Cell Activation Syndrome (MCAS). As I analyze the events in Patrick’s life when my spouse and I educated and nurtured him, it raises the central question for my research: how can parents of chronically ill children foster safety and independence for their children?

This master’s project is multi-disciplinary in that it incorporates primary and secondary data as well as creative writing. Using first-person narrative, I explore how my husband and I navigated the uncharted medical and parenting challenges of GSD1a and MCAS. Secondly, woven into this primary research is data exploring the medical and psychological aspects of GSD and MCAS. Additional insights come from memoirs of parents who have walked down a similar road.

As I chronicled Patrick’s life, I realized that this project has become not only a story of raising an ill child to become independent. This memoir has become an awareness of life choices I had to make once I had a compromised child. This memoir is also about my false sense of control as a young adult and the loss of that control. Additionally, this account acknowledges that almost all parents love their chronically ill children and only want the best for them. The reality of raising an ill child is that it takes health insurance and money—not moral superiority.
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To
Mark Shaw: a wonderful father and support
Patrick Shaw: a man of incredible inner strength
Steve Shaw: the older brother and mentor with unbelievable patience
Our family’s “angels”: Retha, Amy, Mrs. Mason, Dr. Stanley, Dr. Ripton, Dr. Weinstein

To all parents of children with chronic illness and disabilities: It’s never going to be easy. You will walk with your child’s condition the rest of your life; jump when the phone rings; celebrate his small victories. Can you help your child take those small steps forward to independence? It’s worth everything you can give to it.
And, along the way, you will find yourself changed.
Introduction

There are scores of books written about children with chronic medical conditions. Parents—mothers especially—chronicle their struggles with asthma, diabetes, cerebral palsy, sickle cell anemia, Down syndrome, cystic fibrosis, cancer, AIDS, and epilepsy.

However, this memoir is about two little-known medical conditions called Glycogen Storage Disease 1a and Mast Cell Activation Syndrome.

Why would I write about relatively obscure conditions? Well, you have to write about what you know. And I know GSD1a and mast cell. I’ve lived with these conditions for years in raising my son Patrick. And even though GSD and MCAS are relatively unknown, the principles in raising a child with these conditions are very similar to raising a child with diabetes or cystic fibrosis.

What is the so what in this memoir? Why am I re-living much of the painful past? My reasons are twofold. First, I want to share with readers my methodology for raising a chronically ill child to become an independent adult. It’s amazing what a challenged child can do—step by step by step or often, inch by inch by inch. I don’t profess to be an expert in children’s chronic illnesses. I’m simply a parent who bumbled, prayed, researched, begged, and worked to help my child become an independent adult.

The second so what of this memoir is my personal journey and growth in raising a child with two chronic illnesses. As I stumbled through an unknown future with my son, I learned that I couldn’t fix all problems with hard work or sheer grit. I couldn’t maintain control of all
situations. I had to make choices, too. Did I try to maintain my career, or did I stay at home with my children? What did that mean to me, as a woman?

My hope is that you will walk with me in this journey. You may find tears, and you may find smiles. Most importantly, you may find ways to help your child walk on, step by step by step.
PART ONE:

DISCOVERY
1 In the Beginning

It was about three a.m. when the wailing began. This time, however, it was different.

Plaintive and desperate.

*I’m an experienced mom,* I thought. Just six years ago, four-month-old Stephen would fuss each night, and I would fly into his room for the three o’clock bottle. I would rock Stephen, singing my favorite hymn: “Amazing grace--how sweet the sound--that saved a wretch like me.”

It was a peaceful time, nearly holy. No sound except the rhythmic creak of my century-old oak rocking chair and soft slurps coming from Stephen. *This is the essence of life,* I would think, rocking back and forth. I remember the moon outside Stephen’s window, caught in the branches of our massive oak tree. After a nestle-down into the crib and a hefty burp, baby Stephen would move his thumb to his mouth and drift asleep.

*This* time and *this* baby were different.

I had been feeding Patrick each night when he fussed, patting Mark’s inert body with an “I’ve got this.” I was on maternity leave, while Mark put in at least sixty-five hours a week at a pharmaceutical company in Kansas City. It was only fair.

Patrick was about four months old at the time, and I remembered how my baby books recommended that it was appropriate to let him “cry it out.” That’s what I had done with Stephen, so it was about time that Patrick should start following the plan. With my older son, I had perched on the stairs just inches from his room, hearing the wailing slowly drift to a snuffle and then into silence in just a few minutes. All the while, I would bury my head in my hands and think, *this just seems wrong. Stephen may stop crying, but I am surely going to start.* But the baby books were right. Stephen began sleeping through the night after three nights of crying it
out. While these episodes only lasted about five minutes, they seemed an endless torment to me.

It was time for Patrick to follow the schedule, too. I sat cross-legged in the hallway after his wails had reached me, but on this night, the cries sounded different. I rubbed my tired eyes in frustration as I thought of the words in my rumpled baby book: “The cry-it-out method says the babies will never learn to self-soothe if mom or dad always rescue them in the night.” Ok, then. We were working on self-soothing here.

Patrick’s crying continued, building force. Five minutes. Ten minutes. What was this? Why was he not calming down? His well check with Dr. Wigginton just two weeks ago had been uneventful. What was going on, little man?

I finally grabbed the baby bottle, thinking to heck with the baby books. I’m feeding this child. And so I did, night after night. Rocking in the old oak rocker, singing Amazing Grace, and touching my face to Patrick’s fuzzy headed softness. The soft smells of Johnson’s baby shampoo and Similac would drift up to me.

I was right to ignore the baby books. I would have killed my son.
2 Doctors

About a week after I decided to throw the baby books out, I brought Patrick in to see Dr. Wigginton, our family pediatrician. I was worried. I was feeding Patrick in the middle of each night, as well as six small feedings during the day. Patrick had been born with rounded baby cheeks, but he was getting a tummy, too.

Dr. Wigginton was a neighbor and had been Stephen’s pediatrician. The doctor had weathered vaccinations, chicken pox, and midnight earaches with us. He had guided us through teething, fevers, splinters, and vomiting. Dr. Wigginton, in short, was a beautiful combination of a wise Moses mixed with a cheerful Santa Claus. In thirty-five years of practice, he had seen it all.

I positioned Patrick on Dr. Wigginton’s examination table that day. The doctor always talked to Patrick, taking the baby’s little hands in his own spotted ones. “And how are you today, Mr. Patrick? Is your mother reading you some good stories? Have you picked out your university yet?” Then Dr. Wigginton would turn to me and ask, “Do you have any questions or concerns at this point?”

“Uh, yes. It seems like the last couple of days that Patrick is not urinating much. Can you take a look?”

Dr. Wigginton carefully laid Patrick down on the exam table and peeled open the front of the Pampers. He carefully examined Patrick’s little “man stuff.” Being from Kansas, we never called things exactly what they were or said what we really thought. For example, buttocks were labeled “bunootskies,” while a colonoscopy was called a “fooootamagoooot.” Don’t ask me why.
Dr. Wigginton thoughtfully examined Patrick, looking for lumps or malformations. Patrick responded by shooting forth a vigorous arch of urine right down the doctor’s wool sweater, hitting three suede diamond patches on the front. Patrick’s aim was spot on. Dr. Wigginton, always unflappable, looked at me with a grave face and crinkly blue eyes, joking, “Well, *that* part works!”

“Uh. I have another question.” I wrestled with momentary mortification as the pediatrician carefully blotted yellow droplets from his pricey sweater. I described to him how Patrick had to be fed every night and many times during the day. I was even putting a bit of baby cereal in the formula. Nothing seemed to curb Patrick’s appetite.

Dr. Wigginton carefully checked Patrick’s temperature and gently probed the baby’s glands, ears, eyes, and abdomen. He gently sat Mr. Cherub-cheeks up, carefully running his finger along Pat’s tummy and spine.

“Patrick is tilting his head to one side a bit. It seems his neck muscles aren’t strong enough to support his head. I think we should run some tests to rule out any possibility of cerebral palsy or PKU. We’ll also run a complete CBC.”

“Wait. What? What’s a PKU?” I had heard of cerebral palsy, and the mention of it smacked my heart.

Dr. Wigginton explained calmly, “PKU is short for Phenylketonuria. We can do a blood test to rule out this rare disorder. Children with this condition can’t break down phenylalanine,
so it builds up in their body, causing brain damage. PKU can also build up to dangerous levels if the baby or child has a lot of milk, cheese, or even grains."\(^1\)

Patrick had been having formula with a bit of baby cereal in it seven times each day. Great. Purified cow’s milk whey protein. Baby cereal with rice flour, soy lecithin, and heaven knows what else.

I didn’t understand. Mark and I were healthy: we ate broccoli by the bin-full and worked out when we could. Our six-year old Stephen was healthy too, despite having run-ins with ear infections. Our parents were in great shape. Well, my dad had Type II diabetes, but that was just because he was overweight and sedentary, right?

“Let’s just run these tests and see where we are.” Dr. Wigginton was grave, but calm. “Sometimes a baby’s muscles just need more time to develop.”

A little finger of fear wiggled in my gut. Cerebral palsy and PKU—these were serious things. They were not in my plan for life. But, Dr. Wigginton had seemed pretty calm. Patrick and I left the pediatrician’s, as I attempted to quiet the fear inside me. In the car, Patrick became busy shaking a stuffed toy monkey as he sat in his car seat. Mr. Monkey’s eyes stared at me as I glanced in the rear-view mirror of my old blue Volvo station wagon. The eyes seemed to mock me: so, you think you’ve got life figured out?

\(^1\) PKU. Accessed at www.google.com/search?q=what+is+a+PKU&rlz=1C5CHFA_enUS778US778&oq=what+is+a+PKU&aqs=chrome.69i57j0l7.2471j0j7&sourceid=chrome&ie=UTF-8, on 1/11/20.
Two days later, I got Stephen fed and ready for school. It was the usual battle over clothes. Stephen, or Tee-Tee as we called him, wanted to wear a Mutant Nina Turtles tee shirt with electric-colored shorts. He selected bright blue socks most likely, pulling them over his existing socks. Apparently, socks were always itchy and annoying: “spicy,” he called them. As Mark herded Tee into the car for school, the child looked like a circus clown ready to enter the ring at Barnum and Bailey. Tee didn’t care. He was off for a big day in kindergarten. They were making ooblik today, and lunch was going to be pizza. And Maryann had hit him yesterday, and ick, she’s a girl.

Patrick and I spent four hours at Shawnee Mission Medical center after Tee headed to school. We did blood work. Patrick howled, and we waited on turquoise vinyl couches with blank walls staring at us. We did an ECG; an electroencephalogram. Patrick howled some more, and we waited again on different vinyl couches and stared at new blank walls. We did a mouth scrape for genetic testing. More howling and waiting and more vinyl couches. With Patrick still wet-eyed from the morning’s events, I carried him into the genetic counselor’s office.

As Patrick slowly filled his diaper, I answered the counselor’s questions. The room began to smell rather close.

**Nope. I have no siblings. Well, I don’t know. I guess my parents never got around to it.**

Mom told me once that kids were too expensive—but that I was loved, wanted, and needed.

**My Mom’s still alive and eighty. I don’t think she’s ever been sick, except for a few colds.**

Even the cold viruses knew you didn’t mess with my mom. Oh, mom has one sister. Aunt Jan.

She’s five years younger. I don’t think she’s been sick much either, but she sure does have an attitude.
Dad’s seventy-eight. Well, he’s pretty chunky and has Type Two diabetes and he likes vanilla ice cream a lot. No, he’s never been around toxic chemicals—unless you consider numbers. He’s a finance professor.

No, just one other son. Stephen is nearly six. He plays soccer and has a thing about scratchy socks. He’s also stubborn, but he gets that from Mark. Yes, Stephen developed right on schedule. God knew He was dealing with parental rookies.

Mark? Oh. He’s forty-one and I’m thirty-eight. Oh, my pregnancies? Both of them were easy, with normal deliveries. With Patrick, I worked all day—even went to the gym on my lunch hour. I was at home—about 5:30 in the afternoon—when Patrick decided to arrive. I was busy making spaghetti, my favorite dish. The ob-gyn wouldn’t even let me hang around and eat my simmering masterpiece. Something about I was going to have the baby on the front porch.

My grandpas? I never got to see either of them. I think my dad’s dad died of pneumonia in 1928. The other grandpa had some allergies, but I don’t know any more. His wife, my grandma, had to rotate different foods every three days for him. She would mix together certain herbs and light them in a little pot. Grandpa would sit over the vapors and inhale the smoke. I never did know how he died.

My grandmothers? Well, Mimi, my dad’s mom, was a sweetie. A minister’s wife who baked cinnamon rolls and played the organ in a funeral home in Hutchinson, Kansas. I think maybe she had diabetes, yet she would bake up a storm. My dad’s favorite when he was growing up was raisin pie. He talked about it all of my childhood. I mean, all of my childhood. In fact, dad wrote the cooking columnist Nan Wiley. She was published in the Kansas City Star and syndicated over the country. Dad described his mother’s decadent raisin pie and sent out an
impassioned appeal. Within weeks, little old ladies all over the country sent dad recipes for raisin pie. My mom made thirty raisin pies of the course of two years, and each one, according to my dad, “was pretty close, but not quite.”

My other grandma? Well, she was fine and then got really lonesome and starting smoking cigarettes. We’d find them all over her house when mom and I would come to clean. Sometimes, I’d go visit grandma, but she’d get really cross with me when I’d play in the birdbath. Then, she got really thin. I guess she was forgetting to eat. We moved her to a rest home in Emporia, Kansas where I lived as a girl. Somehow, grandma fell and broke a hip. Mom and dad took her to the hospital, and I never saw her alive again.

Mark’s parents? Well, Mavis, his mom, is a vivacious, social redhead that is a whiz at bunco and pokeno. Grandpa Bob is a wiry little guy with a flat top and a huge smile. Bob has had fried eggs, bacon, and Tang every day of his adult life. He seems pretty healthy, despite the cigarettes.

The genetic counselor took copious notes as I detailed the family history.

“Well, I’ll be in touch with you in about two weeks. I’ll need you to complete some blood work here at the hospital before you leave. When can Mr. Shaw come in? Once all the data is in, I’ll send you a report and call you.” The counselor stood up, signaling that the session was over. It was okay. Patrick’s diaper was making our eyes water.
3 The Red-Headed Angel

The weeks flew by, with soccer practices for Stephen, mounds of dirty clothes, teacher conferences, and first grade homework. “See, Tee? Here are two tennis balls, and I give you two more. Let’s count them. How many?”

“Five!” Tee would always answer.

I had only taught for five years, but never could figure out the contrarian in Tee. If I said day, he said night. If I said yes, he said no. If I said that the sky was blue, he’d say, “Well, actually, it’s azure.”

Azure? How the heck had Tee learned that in first grade?

While Tee was in school, Patrick and I had a busy schedule. We’d stack blocks or read Pat the Bunny and Goodnight Moon. We’d look at black and white stylized images of faces. “Happy!” I’d say, pointing at one image. “Sad,” gesturing to another. This was supposed to make Patrick brilliant. Patrick, at six months of age, would sit quietly for ten minutes at a time, intently listening to my stories. I’d never been around babies except for Tee. Unlike Pat, Tee would listen to a story for two minutes and then crawl like a beetle to any shiny toy nearby. Magazines would swoosh onto the floor in piles. Tee would pound pegs into his toy workbench, pushing it aside in ten seconds to zoom over to Tinker Toys. Not so with Patrick, the contemplative Buddha.

I had another month before I had to head back to work. Mark and I decided that—at least for a while—we’d use a full-time caregiver in the home until Patrick could go to school. Mark worked long hours at a pharmaceutical company, and I was about to return to a new position at a telecommunications company. Life would turn into chaos soon.
I visited with one of my friends after church one Sunday. She was a pediatrician who was heading back to work part-time now that her son was in a full-day kindergarten. She told me about her sitter who was now available. Retha.

Angels come in a lot of sizes and shapes, some of which are red headed.

Retha was in her late thirties, too, with a husband and two nearly-grown children. A smallish woman with curly red hair and freckles, Retha hailed from the Missouri Ozarks. She’d tell me how she and Bobby, her husband, would go to “Walmarts” over the weekend, sometimes hitting a big sale on fishing equipment. Retha also loved “Ventures,” another discount store where she’d purchase Coke by the twelve-pack. She had a two-can-a-day habit. Retha also loved “Pac N’ Saves” as well as “Hen Houses.” Somehow, she’d always manage to add an “S” on the end of every store. It was just her way.

Retha and I hit it off immediately. I loved her fifteen years of experience around babies. She was so calm and just always knew what to do. I had my baby books and my two degrees, but I couldn’t match Retha’s firm but gentle ways. We worked together for four weeks, talking about the boys’ schedules. Retha and I were even on the same page with discipline. We both agreed that with Mr. Contraria, consistency was vital. An occasional time-out for Tee in his little red chair could do wonders. Daily chores were a must, combined with positive feedback. Even Patrick, once he was able, would help put away the books and Tinker Toys typically strewn all over the family room.

Over the next few months, I got to know Retha’s family. Bobby worked as a welder. Crystal and Scott were in high school—both with decent grades and part-time jobs. No one was a slacker in Retha’s family. When Crystal got married to Robbie at age eighteen, Tee was in the
wedding as the ring-bearer. That red-headed child was decked out in a white tux and a small black bow tie. He looked like Mr. GQ, and boy, did he know it. Scratchy socks and all—Tee marched down the aisle of Crystal’s wedding with dignity. At the front of the church, he announced to the minister and congregation, “I got em’! The rings! Here ya go!” The congregation burst into laughter, as Tee dropped the rings and they rolled down the steps of the church altar.

Retha and I were totally on the same page. I was ready to launch into motherhood and career with child number two. Piece of cake, right?
I loved my lazy days at home with Patrick. Reading stories. Wandering over the hills in the bright blue baby jogger. Plinking the keys on Patrick’s toy piano, singing Barney the dinosaur’s favorite song: “I love you, you love me, we’re a great big family . . ..” Lying in a patch of sun on the family room floor, with Patrick resting on my chest.

However, work was waiting.

I had been an English teacher for about five years before the boys were born. I taught seventh grade English and later ran a reading laboratory for all the “sevies.” It was rewarding work, especially when I saw a mental light switch on for my kids. When little Jason went from a third-grade reading level to the sixth in four months, it was the best. When John could contain his hyperactivity and write a good paragraph, it was the best. He would allow me to place a gentle hand on his shoulder—the only kind touch he would get during the week from anyone.

* * *

Now comes the part about me becoming a captain of industry.

It was a joke between Mark and me, actually. While I was dodging spitballs at the middle school and breaking up fights in the hallway, Mark was jetting off to New York City or having business dinners at the Hyatt in Kansas City. He was writing up white papers on FASB’s and meeting with vice presidents. I felt my teaching simply paled by comparison. Plus, I was slowly going crazy teaching the same subject matter several times a day. Additionally, the introvert in me wanted to crawl under a rock. Thirty sets of eyes looking at me at 8:00 every morning? No way.
So, I wanted to become a captain of industry. Why should Mark have a cool job, and not me?

I’ve never let details like education or work experience stand in my way. I had the goal in mind. So, I took the Graduate Management Aptitude Test. The results: my verbal and communication skills were high. The math scores were embarrassingly low. “That ‘new math’ in fourth grade did me in,” I would rant to Mark. “Those Venn diagrams—that’s what did it. Who cares about union and intersection?”


Nothing. Every letter came back to me, stating, “While we appreciate your strong educational background and teaching experience, your skillsets are not a match for the position we are trying to fill.” In other words, I discovered that two English degrees and a quarter would buy me a cup of coffee. It was depressing. Someone—anyone—needed to give me a chance.

In the midst of the job application bloodbath, I applied for the MBA program at Rockhurst University in Kansas City, Missouri. It was known in the area to have a strong accounting and finance program; one in which I could work full time and go to class two nights a week. We were told to expect eight to ten hours of homework on top of the coursework per week. Thirty-six graduate classes plus fourteen hours of pre-requisites for those of us who were liberal arts people. Easy peasy, right? At any rate, the gods were with me. The school must have looked at my university transcripts and skimmed over the puny GMAT math score. I got into the MBA program.
Not only were the graduate gods with me that week, but also the job gods. I talked to a friend one day in the checkout line at Pak 'N Save. Her aunt was leaving her position as a legal secretary at United Telecommunications, the parent company for fourteen telephone companies around the country.

_Hmmmm. Legal secretary. Get my foot in the door. Why not? I could type; I could quickly learn the Gregg shorthand system at the local community college. Sure, why not?

- Somehow, I got the job. I arrived on day one, donned in my beige suit. I began hammering out legal contracts on my IBM Selectric with Wite Out tape perched nearby, praying that I could avoid typos as I peered at my boss’ scrawling longhand that filled page after page of legal tablets. What were access charges? Would I know an analog signal if it flowed by me? I simply didn’t know the telecommunications terminology. I hammered out legal contracts. Page after page. Rewrite after rewrite. As the months clicked by, I was knee-deep in legal lingo and had completed my first two graduate pre-requisite classes.

Within eighteen months, I had completed six courses and received a promotion as an accountant in general accounting. Even though I was Miss Puny Math Score, I loved accounting and understood it. It was logical and balanced. It didn’t talk back to me. If I stuck ten dollars on the debit side of a ledger, I had to do the same on the credit side. The tick-and-tie control freak inside of me loved the symmetry—it was nearly as satisfying as organizing my Little Golden books as a child. So, it was little wonder that I decided to go for an undergrad accounting degree while I was in school at Rockhurst. What was another twenty hours? I plowed through beginning accounting I and II and into intermediate accounting I and II, the “bad boys” of an

The months continued to fly by, as I completed course after course. At work, I moved into Consolidation accounting, Information Technology, and finally, Employee Benefits. It was all challenging and so fun. I would take a bank statement and wrestle with the fact that the company records had a totally different starting balance than the bank had. How could that happen? I had to get this to agree—no ULD (Unlocated Difference) for this lady. In Information Technology, my boss Ken and I would crack open new computers and load software. We would travel to various companies to investigate new types of accounting consolidation software. I would fend off the Arthur Young year-end auditors with my impressive data schedules. I was like my lab Georgie, who loved to wriggle on her back in the back yard, kicking her feet up in sheer joy. In short, I loved each area of United Telecommunications and became accustomed to jumping into a new position every year or so. And, I finally got out of graduate school, nine months pregnant with Tee. I had been at Rockhurst so long, I halfway expected the university to name a building after me.

Guess what? They didn’t.
Home on the Range

The days and months flowed in a pattern—as much of a pattern as you can have with children.

I’d get home each day promptly at six o’clock, because Retha had a family to get home to, also. Thump. Heels flung by the front door. Thud. The briefcase dumped on the stairs down to the family room. Hug: Tee Tee. Pick up and kiss: Patrick. Debrief: Retha. Five more minutes of visiting with the kids and then I’d head straight to the kitchen, still in my suit, ready to cook. Maybe spaghetti and meatballs. A savory roast steaming in the crockpot. Beef stew re-heating after the weekend marathon of cooking. Mark usually got home at 6:30, and we all ate as a family soon after. Death, taxes, and children’s hunger would never wait.

* * *

Patrick’s tests for PKU and cystic fibrosis had come back normal, as did his blood work. Although he wasn’t extremely active like Tee, I inwardly relaxed; the fear in my gut transferred to work instead.

I had learned just before my maternity leave that I was to be promoted as Senior Manager of Employee Benefit Administration and Accounting. It was a mouthful of a title, but little did I know what I was walking in to.

Our small group at work had deferred compensation plans to manage, as well as a below-market stock purchase plan, a TRASOP plan (Tax Reduction Act Stock Ownership Plan), and an old ESOP plan (Employee Stock Ownership Plan), to boot. I learned quickly that no documentation existed at all on these plans—just reams of employee distribution printouts.
did know that any missed distributions to employees and executives would trigger massive tax liabilities for the company. It would also trigger massive anger from any employee who didn’t get his distribution or didn’t get to enroll in a stock plan. Additionally, the gentleman who was relieved of running these plans when I took over was none too pleased. All I could get out of him was that each program ran on COBAL. “But,” he’d tell me, “for these seven individuals, the program has a glitch. You need to update these seven Excel spreadsheets every quarter for these individuals.” Great, I thought. *What a big opportunity for error here.* We might as well have been using hamsters inside a spinning cage to power the benefit plans. What made matters worse: my new boss was the Pension Trust Officer. His full-time accountability was making sure the company’s pension was fully funded. For the running other benefit plans—he relied on me. Adding to this poisonous mess was the fact that one of the department members was angry at me. She had wanted my job and was *sure* she had deserved it.
To this day, I am haunted by the fact that I may not have been on top of Patrick’s growing medical issues. I cooked for my boys, read stories, played Legos. It wasn’t enough.

After one particularly grueling workday, I did my usual toss of shoes and briefcase on my scramble down the stairs into the family room. I always had to hug on my boys and visit with Retha first thing. In her calm manner, Retha was directing the boys to clean up their toys. Tee made pot shots of Legos into his red toy bin, while Patrick merely scootched his favorite book around.

“I think Patrick’s tummy is too large,” stated Retha. “It doesn’t look quite normal.”

Really? Patrick was only the second baby I’d been around in my life. True, he had chubby legs and round cheeks. He used to create quite a stir in Pak ‘N Save as the personification of the Gerber baby, with his huge blue eyes and blonde curls. Later, as I changed Patrick’s diaper, I looked at his tummy. Well, it did look a bit big, compared to Tee’s at that age. However, Tee had always been skinny and active. I made a mental note to book an appointment with Dr. Wigginton.

* * *

At Patrick’s eighteen-month mark, we saw Dr. Wigginton.

“Look at his tummy,” I gestured to the pediatrician. “Does this look too big?” Retha’s recent words echoed in my head.

Dr. Wigginton carefully touched and manipulated Patrick’s stomach, holding two fingers against the right side of the tummy and thumping on his fingers, moving the fingers, and thumping again. His head was bent, listening.
“And, um, well. Should Patrick be getting ready to walk now? He crawls but doesn’t even try to pull himself up.” I had been working with Patrick for several weeks now, standing him up and gently holding onto his arms. Pat would crumple back into a sitting position.

“Mr. Patrick, what’s going on with you?” Dr. Wigginton asked gently, as he examined feet, arms, tummy, ears, eyes, mouth, and head posture.

The pediatrician stood quietly for a moment, flipping through Patrick’s medical chart. “Let’s schedule an ultrasound of Patrick’s abdomen in the next two or three weeks. I also want you to begin taking Patrick to Childwise Physical Therapy. We need to see if we can strengthen those legs. Let’s also get an appointment with Dr. Grace Somers. She’s a cognitive assessment specialist.”

Whoa. Something scary and not on-plan was happening here.

The next few weeks were a flurry of activity. At work, I dug into benefit plans, examining printout after printout, analyzing how the numbers were calculated. I met with our legal department to find out plan parameters. I met with programming so we could move from hamster-and-wheel processing to a more effective programming language at the time: C++. I met with department members to ascertain their goals and training desires. I wrote up quarterly personnel reviews. I felt as though I were slowly crawling into a place of light and of understanding my position. It was exhilarating; I was using my brain to the fullest.

Three times a week, I’d meet Retha nearby when she was picking up Tee from school. I’d transfer Patrick from her car seat to mine, and we’d head out to Childwise PT.

Amanda was a pediatric physical therapist at Childwise. I loved her calmness—the way her bright blue eyes truly examined Patrick.
I told Amanda that Patrick was not bearing much weight on his legs at eighteen months. Nor had he attempted to pull himself up or take those few halting baby steps that I longed to see. In essence, Patrick seemed perfectly content to sit like a baby Buddha.

At each PT session, I would sit by Amanda and observe her techniques, because I would have homework from these sessions. Amanda would carefully lift Patrick onto a small stool, placing his bare feet on the floor. “Look!” she’d exclaim, pointing to Barney the dinosaur on the floor close by. Patrick was supposed to get up and grab Barney and then sit back on the stool. Patrick looked at Barney—his favorite television character—but didn’t move.

Amanda, Patrick and I also practiced “cruising.” We’d head to the nearby couch with a teddy bear placed at one end. Amanda would place Patrick at one end of the couch, his hands positioned on the cushions. “Let’s go get that teddy bear!” she’d say. Amanda and I would cruise down the length of the couch, our hands patting the cushions with happy slaps, our knees pressing into the carpeting. I would look at Patrick expectantly. “Come on, buddy!” Patrick would look at me and simply slide into a sitting position.

Amanda and I would hold Patrick gently by his upper arms and move one shoulder forward slightly, then the other. This motion was supposed to encourage Patrick’s lower body to follow suit and move the legs. One leg would come forward. Then—plop—down Patrick would go, his blue eyes blinking as if to say, “I’m done here.”

Patrick’s favorite PT activity was bicycling. Amanda would lay him on his back and “pedal” his legs, carefully extending and contracting the muscles. Pat would beam and smile. “Mo!”
Patrick and I would practice these PT moves each evening after dinner. “Come on, Tee, help us,” I’d encourage. Soon, Mark, Tee, and I would be cruising down the family room couch. Slap slap slap. Pat would stand holding the end of the couch and look at us as if to say, “You’re all kind of nuts.” Our favorite family activity, however, was the bicycling. We’d all be on our backs, pedaling in the air, belting out Barney the dinosaur’s song. We looked like a family of struggling June bugs.

One night, Mark was in a work meeting, so the remaining Team Shaw did PT. I ran upstairs for a minute to start the daily laundry load. “Tee—would you bicycle with Patrick while I throw in the laundry?” Tee was the typical, responsible older brother. “Sure, mom,” he replied. I came downstairs within a minute or two. Patrick was wrapped in his baby blanket—his dee dee—a tattered piece of flannelly love. Tee had secured Patrick won-ton style in the dee dee, slowly dragging him all over the family room carpet, crying “Booster jets! Booster jets!” I rushed into the family room, alarmed by the sound of booster jets in my family room. However, Patrick was laughing with delight on his dee dee ride. Tee pulled him slowly, around and around the family room, into the kitchen, and back out, with our lab Georgie trotting behind. Slowly, Patrick’s won-ton wrap came loose, and the child rolled once, twice, six times, coming to a stop two inches from the family room wall. “Mo mo!” he smiled. “Mo mo dee dee ride.”

You can’t make this stuff up.

Two weeks after the PT marathons began, I took Patrick to Dr. Grace Somer, a physician specializing in cognitive assessment. As each week passed by, I was becoming more concerned by Patrick’s lack of physical movement. Sometimes, he seemed like a floppy rag doll. He could sit on the carpet, heels of the feet touching, knees spread open wide to the floor. However, I
was pretty sure his brain was working just fine. Words were beginning to flow out faster and faster. “Want a deee ride,” Patrick would say, or “Have some cheese crackers.” One day, as I thumped down the stairs after work, Retha met me on the landing, slightly breathless.

“Patrick just crawled up to me and said, ‘Retha, I’m uncomfortable. I think I need a Tylenol.’”

What had been simmering in this child’s brain? “Uncomfortable? Tylenol?” Really?

At the two-hour session with Dr. Somer, the specialist performed a variety of exercises to determine if Patrick’s brain was developmentally sound. Her room was quiet, peaceful, filled with bright, enticing puzzles. I was a bit envious of Pat. This looked like fun.

“What will you be looking at?” I asked Dr. Somer.

“I’m going to observe Patrick’s verbal skills, verbal reasoning, novel problem solving, new task learning, and decision-making speed,” the specialist replied, flipping through papers on a clipboard. I told Dr. Somer about Patrick’s unusual ability to sit and listen to stories—now, for up to fifteen minutes at a time. I told her about the incident Retha reported when Patrick had gushed forth two complete sentences.

“Interesting,” she commented, scribbling away and then pausing to watch Patrick. Dr. Somer then placed herself behind a low Plexiglas wall with a red ball. Patrick was on the other side.

“Patrick. Look at this ball! Can you get it?” Dr. Somer waived the red ball enticingly.

The child immediately reached around the wall and took the ball. He bounced it away, as if to say, “Now whatcha got?”

“What does this show you?” I asked Dr. Somer.
“In slower cognitive development, the child won’t realize that he can reach around the wall. He’ll try to reach directly to the ball. He wouldn’t understand that the clear Plexiglas is blocking his grasp.”

“Well, what will all these other puzzles and games tell you?” I peppered Dr. Somer with more questions. I was thinking, was Patrick smart?

“I’m observing to see if Patrick might have any intellectual disabilities or processing problems that might impact the brain-to-body movement. Has he had any brain injuries?”

Outside of the near escape during the wild dee dee ride a few days ago, Patrick had had no incidences. I did tell Dr. Somer about his need for frequent feedings and muscle weakness and floppiness.

A week later, I received a report in the mail from Dr. Somer. She was pleased to report Patrick had “advanced cognitive ability.” So why couldn’t the child walk? Something was wrong with my baby.
7 Something Was Wrong with Patrick

When Patrick was about nineteen months old, Mark and I brought him in for the ultrasound of his abdomen at Shawnee Mission Medical Center. Patrick had been continuing to spout words at a breath-taking rate. However, Patrick was still not walking, choosing instead to crawl after Georgie. The wiggle of fear in my gut had been growing stronger.

I still remember him. The technician in the ultrasound room looked like a Kansas City Chiefs defensive end. Big, burly arms coming out of his technician’s white coat. Crinkly brown eyes that radiated kindness and a cap of curly black hair.

The technician and Mark placed Patrick on his back on the white bed and the child immediately tensed. The technician tickled Patrick’s tummy and said, “Now, Mr. Patrick, we’re going to put this gel on your tummy and then I’m going to roll this wand around and around on your belly to take a look. It’s magic.” He let Patrick touch the special wand. James placed a dollop of the warmed ultrasound gel on Patrick’s belly and began to slowly roll the “fairy wand,” first on one portion of the stomach and then the next. He would pause. “I’m looking at the gallbladder now.” James would measure the organ on the screen, capture an image, and type in numbers. “I’m also looking at the liver, spleen, and pancreas. I’ll be also viewing the inferior vena cava and aorta.” Red and white surges of light appeared on the ultrasound screen, pulse-like, almost psychedelic.

Mid-ultrasound, Pat protested, “No sauce! No sauce!” against the gooey ultrasound gel and gentle wand circling on his tummy. He wound himself up into a soppy crying fit. The technician, unperturbed, captured more images and measured each organ. “It’s okay, Mr. Patrick. We’re almost done.” I held Patrick’s hand and stroked his baby curls with one finger.
“When we get home, we’ll read Goodnight Moon,” I encouraged. “We’ll go to the park with Tee tomorrow.”

The ultrasound session was over. Mark hugged Patrick as I gathered my purse and the diaper bag. I’m not sure who was the more dewy-eyed from the ordeal. Two sets of teary blue eyes looked at me.

Two days later, the radiologist had the ultrasound report faxed to Dr. Wigginton. The pediatrician called me that afternoon at work.

“Janet, I’ve just looked at this report. Patrick has an enlarged liver.”
8 Now What?

I don’t remember the rest of the conversation with Dr. Wigginton. Something about “Please call me any time” and “Bring Patrick in tomorrow at Shawnee Mission Medical Center for additional blood work and a cat scan.”

Something was wrong. Really wrong with my baby.

I went into my boss’ office and told Bill what had just happened. I had to go home. I got home somehow, not remembering the streets I took. I tried to think through my fear. The only science courses I had taken in college were Geology, affectionately dubbed Rocks for Jocks; and Trees and Shrubs. I was clueless about medical issues, but I sure could identify igneous rocks and bud scars. I knew that the liver was in the abdomen—on the side, perhaps? All I could think of was hepatitis C, cirrhosis of the liver, liver cancer. Did they enlarge the liver? All of the conditions sounded terrible and deadly.

When I got home, I sneaked Retha over into a corner of the kitchen and told her the frightening report on Patrick’s enlarged liver.

“I wonder if that’s what’s making his tummy so large?” Retha began to tear up, her freckled hand up to her mouth to suppress a cry. She and I hugged and hurried up the stairs so Patrick and Tee wouldn’t see. I came back down and mushed on my boys. We sat in the slanted late afternoon sunshine in our family room, racing Lego fire trucks and putting together large wooden puzzles. In the sickness of my heart, I thought, how many times have I sat here with my boys, just taking this peacefulness, this grace of children, all for granted?

As soon as I heard the click-click of Mark’s dress shoes in the upstairs hallway, I bolted up to him. I needed to share my fear with someone special—someone wise and calm like Mark.
“The radiologist’s report came back today. Dr. Wigginton called me and said that Patrick’s liver is enlarged.”

I’m not sure how Mark and I got through the evening. We fixed dinner, supervised bath time, and read stories to the kids, just like always, because we had to. Mark took Pat while Tee and I headed to his room. I was in no mood to be a disciplinarian that night, so I let Tee ride the swinging wooden wolf that Grandpa Bob had made for him. It hung on sturdy ropes from the rafters in his room. Back and forth Tee raced, nearly hitting each wall—crying out “Booster jets! Booster jets!” Then, tucked carefully in his Nina Turtle sheets, we wrapped up our bedtime ritual. I raised my arms and hands apart, as if I were measuring the field goals at the Chief’s football stadium.

“I love you this much!” I’d exclaim, with my arms spread out.

“How much?” Tee would reply.

“This much!” My arms would widen.

“Howwwwwww much?”

“Thiiiiiiiiiiiis much!” My arms would spread as far as they could go.

It was beautiful. For just a moment, I forgot the sickness in my heart as I kissed Tee’s freckled nose.

* * *

Later that night, Mark and I sat with glasses of wine and a pad of paper, trying to think of where we could go for help. Although Mark worked for a large pharmaceutical company, he was in purchasing. Everyone in our extended family was either a teacher or in business. No doctors. No science backgrounds.
“What about Carol Morrison at church?” he finally suggested. “She’s a pediatrician . . .
only been out of school about seven years. She’s working part-time now, because she has two
young boys. Let’s call her tomorrow.” The two of us headed to bed, sleeping poorly. A black
mist of fear hovered over us.

The next morning, Mark headed out the door to take Tee to school while I took a
personal day to regroup. As soon as the clock hit 8:30, I called Carol. She and I had talked often
after church over coffee and donuts in the narthex. Ours was a small church of eighty
parishioners—everyone knew everyone. We all pitched in to clean the church and haul away
the trash each Saturday. When the minister’s son picked his nose during the Children’s Sermon,
everyone grinned. There were no secrets.

I described to Carol what the radiologist’s report had said. “Come on over,” Carol invited
me. Sitting at her kitchen table with coffee minutes later, I held the squirming Patrick in my
arms. Carol gently felt his tummy.

“One of my professors in school was Dr. Marris, head of genetics at Children’s Mercy
Hospital downtown. I’d go see him. He’s very knowledgeable about genetic issues and I think he
could help you get a handle on this.”

Genetic issues? My child? No one in our family had had any weird medical issues that I
knew of. People died from broken hips or diabetes or just old age. Plus, I had undergone an
amniocentesis with both pregnancies. Apparently, with my first pregnancy at age 32, I was
“mature” and therefore a risk. I was 38 and “very mature” when Patrick was born. I had
decided to receive the slightly risky treatment because my obstetrician had informed me that
an amniocentesis would detect chromosome abnormalities, neural tube defects, and genetic
disorders, such as Down syndrome or cystic fibrosis. Well, I thought. *That would not be part of my plan to have a child with a “problem.”* However, I knew I was set. Results from each amniocentesis came back labeled “within normal range.” Each Project Child was on, according to my tidy little schedule.
9 Children’s Mercy Hospital

On Monday morning I was on the telephone to Children’s Mercy. Incredibly, I got an appointment with Dr. Marris for the following week. A small beam of hope pushed at the sick feeling that had been in my stomach since I had learned of Patrick’s enlarged liver.

Our thirty-minute trek into Children’s Mercy was uneventful, as we wound up and up the layers of parking of the hospital. Patrick and I walked down hallways and more hallways, to elevators and more elevators, and into the genetics department.

The genetics waiting room was purposefully cheerful. Patrick and I checked in and headed over to a large play wall with moving gears and spinning wheels. The couches and chairs in the large room were happy colors. Framed pictures of children’s art decorated each wall. The room seemed to say: We know you’re here with a sick child. We’re going to pretend everything is pleasant. I looked around. Patrick had made a new friend, a freckled girl who appeared to be about three or four.

“See? Spin this dial,” Pat explained to the new friend.

“Da,” the child stated. She tried to push the dial but couldn’t get her hand to do it. “Da.”

By this time, Patrick had crawled over to a colorful tic-tac-toe mat, pushing each square for a satisfying “ding.” His friend was still at the dial, staring at it blankly. I looked around. Parents sat here and there in the waiting room, piled up with babies with NG tubes and loaded diaper bags. One small boy was wheeled out of the appointment room in his miniature wheelchair, an IV in his arm and a mask over his face. Another child walked by with one healthy arm but the other one was just a stub. I was taken aback. I had never seen so many children with physical challenges in one place.
“Mrs. Shaw and Patrick?” It was time for our appointment.

I looked at Dr. Marris expectantly as we entered his office, because he was my hope and my only hope for this liver nightmare. I remember his crisp shirt and pin-stripe tie. Grey eyes looked at me out of tortoise shell glasses.

“Patrick’s twenty-one months now and still not walking,” I told Dr. Marris. He’ll try to pull up but just falls down. I feed him about seven times a day. And here’s the radiology report.”

Dr. Marris took copious notes, flipped through the radiology report, and then carried Patrick over to his examination table. Gentle hands palpated the child’s tummy here and there. Capable hands sat Patrick up and looked at the cute little tilting head. Knowledgeable hands probed Patrick’s range of motion with his legs. They were pliable, floppy; knees falling open and heels together.

I wriggled a bit on the office chair. My hopes and my fears fought, making my heart thump. Patrick looked so cute and vulnerable on Dr. Marris’ exam table—huge blue eyes taking in everything around him.

After minutes ticked by, Dr. Marris handed Patrick to me and sat in his office chair.

“Well,” he said, clearing his throat. Well, what? I wondered, my gut continuing to churn.

“Based on my exam and the test results here, I believe Patrick has a form of Glycogen Storage Disease.”

I interrupted Dr. Marris. “What? I’ve never heard of this. Why didn’t Dr. Wigginton diagnose this? What is this glycogen thing?”

Dr. Marris didn’t say anything but leaned forward in his chair, staring me into quietness.
“Based on what I’m seeing, I think Patrick has a less severe form of Glycogen Storage Disease. There are about sixteen kinds that have been discovered so far. If I had to take a guess, I’d say he’s has GSD Type IIIa, which you’ll sometimes hear referred to as Forbes’ disease. We won’t know for sure what kind of GSD Patrick has until we perform a needle biopsy of his liver and take some tissue samples. If Patrick has GSD III, it means his enzyme deficiency is a glycogen debranching enzyme. All forms of GSD are rare—some more than others. Type III has a one occurrence per 100,000 babies born. Many doctors have only read a page or two about the condition—usually in medical school. Can you feel his large liver, here? It’s storing excessive glycogen. And his round cheeks are storing adipose tissue.”

“Um,” I said, as though my intelligence had all been sucked away in the fluorescent-lit room. “My degrees are in English and finance. I’m really lost here. Can you explain this to me some more? What can Mark and I do to help Patrick?” The words needle biopsy circled ominously in my brain.

Dr. Marris looked a bit irritated but continued. “Patients with GSD will have a certain enzyme in the liver that’s either missing or mutated. The condition derives from an autosomal recessive genetic disorder that both you and your husband have. When you conceived, Patrick had a one in four chance of getting GSD.”

“So . . . so . . . so . . .” I was babbling like an idiot parent, trying to take in all this foreign information and formulate questions. I had to talk fast before Dr. Marris would whisk out of the room and on to other “interesting” genetic mysteries.

“So, what this means,” Dr. Marris continued, “is that Patrick cannot convert much, if any, glycogen into glucose. When you eat a meal, for example, your body converts excess
glucose into glycogen. It is stored in the liver until more glucose is needed. As your blood sugar drops, your liver converts the glycogen back into glucose, keeping your blood sugar at what we determine is a normal level—say, between 80 and 120. A healthy liver is like a gatekeeper for the body."

I grabbed a notepad from my purse, crossing out “hamburger” and “apples” and hurriedly scribbling “glucose,” “glycogen,” “GSD Type III,” and “blood sugar 80 – 120--why?” “Healthy liver versus ???” “Adipose?” I would call Carol Morrison in the morning.

“Dr. Marris, what can we do?” I was not going to cry. I had to stay calm, think, get information. I felt as though I were pulling information painstakingly from Dr. Marris. Didn’t he realize he was my lifeline to hope? To information? To . . . everything?

“I will schedule Patrick’s needle biopsy to be performed in the next week or so. In the meantime, get some Argo cornstarch. I want you to stir two teaspoons into each baby food feeding and do the same with Patrick’s formula each time, shaking well. Don’t cook the cornstarch and mix it up fresh. Do this with every single feeding. I want you to feed Patrick every three hours."

Argo cornstarch? For a condition that was as rare as GSD? I remember Dr. Marris had said something about one in 100,000 births. Good grief. I had cornstarch in my pantry, along with Pillsbury’s flour.

Dr. Marris continued: “I want to see you and Patrick one week after the needle biopsy is performed. We’ll go through the biopsy and see how Patrick is doing on the corn starch therapy.”
The doctor stood up, signaling that we were done. I scribbled “Argo cornstarch—why?” on my pad, collected Patrick, and thanked Dr. Marris for helping us figure out what was wrong with my baby.

“Oh. One last thing. Where can I go for more information or help?” I had no idea where to turn.

“You can contact NORD, the National Organization for Rare Disorders. The front desk can give you the number.”

Patrick and I hit Pak ’N Save on the way home, buying boxes of Argo cornstarch and several jars of Gerber’s bean and bacon toddler food. I felt like Captain Jean-Luc Picard on the Star Trek Enterprise, bolstered with fierce determination as I thought: engage. The only problem was, I really didn’t know where I was going. I had no map.
10 So, What’s “Normal”?

That night, I created a cornstarch drawer, cleared a cornstarch shelf, and taped a cornstarch dosing schedule on the fridge.

I found a huge Tupperware bowl and dumped two boxes of Argo cornstarch in it. That night, because it was Tuesday, we most likely had meatloaf, mashed potatoes, and broccoli. I sneakily stirred one teaspoon of cornstarch into Patrick’s potatoes to test the waters.

At the dinner table, we hashed over the day’s events. Mark had gotten budget approval to build out more lab space at his work, because Cardizem was selling like crazy. Tee was excited because they got to handle worms in his science class, and they were gooshy and they pooped on you. I felt a bit hopeful for the first time in months. I briefed the guys on the findings with Dr. Marris, watching Patrick sideways to see if he’d eat the potatoes.

No problem. The potatoes went down and the meatloaf did too.

“So, we’ve got to feed Patrick every three hours and mix raw cornstarch into his food and formula. No, I don’t understand why exactly, except that Dr. Marris said it would help keep Pat’s blood sugar up between feedings. He hasn’t been walking because he’s just so tired. Now Patrick, the doctor told us today that your liver doesn’t work quite like Mom or Dad’s or Tee’s.”

“I know. It’s right here,” replied Patrick, patting his tummy where his liver lay. “It’s kinda big.” He had been paying attention to every word Dr. Marris had said.

“Well, if you start to feel tired, you tell dad or mom or Tee right away, okay? Dr. Marris says that cornstarch will keep you from feeling so worn out and it will help you grow strong. We’ll help you take it every three hours. I know it doesn’t taste like mac and cheese, but it’s not
too bad.” I went to the kitchen and brought out a little bowlful to show Patrick. He stuck two fingers in it and dabbed, making a fluff of snow on the table. I stuck my fingers in the bowl, too, licking off some of the powder. “See? This is what we use to make ooblek. It’s pretty cool that this will make you feel better.”

“Okay,” replied Patrick, very matter-of-factly about this new culinary turn of events. “I liked the play wall at Dr. Marris’ with the spinny wheel. I met a new friend but she couldn’t talk.”

This was Day One of Patrick’s treatment plan. He would quickly learn to come to me and say, “I’m cornstarch tired.”

* * *

Looking back at that night at the dinner table, I think about how I introduced Patrick’s chronic illness to him. “Your liver doesn’t work quite like Mom or Dad’s. Cornstarch will help you to grow and not feel tired.” Did I tell Patrick too much or too little that night? I had had twenty-four hours of psych classes in college, but I felt unprepared for the GSD diagnosis. Mark and I had no one to tell us what to do, and we were both in shock over the diagnosis. In 1993, Archie (the first Internet search engine) emerged, but those of us in the general public relied on phone calls, books, faxes, and personal contacts for information.

Today, management for children with chronic illnesses abounds in books, articles, and the Internet. Lubkin’s *Chronic Illness*, a textbook for nursing students, affirms that I was right to tell Patrick of his condition soon after diagnosis. The text states, “Education and information, given at the right time, at the right place, and at the right educational level, are instrumental in
promoting certainty.”

Lubkin lays out the ten thousand-dollar question: did I provide Patrick enough information initially? Was it at a level the child could understand? Was it at the right time, the right place? Lubkin adds: “Readiness to learn manifests in a variety of areas, such as physical readiness, emotional readiness, experiential readiness, and knowledge readiness.” In thinking about this statement, I realize that Patrick was physically lagging behind other children of his age, but he did have the emotional advantage of a loving family and numerous outside stimuli, such as play dates, church, story time, Border’s Books with its enticing cappuccino—even big brother Tee and Georgie. Patrick appeared to be fairly bright, as Dr. Somer had noted in her cognitive evaluation and was fortunate to live in a stimulating world.

It is interesting that before Jean Paget’s theories on childhood cognition, parents thought that their offspring had no cognition abilities until they started to speak. Piaget hypothesized that nothing could be further from the truth. As detailed in the “Learning RX” webpage, childhood cognitive development follows four general stages, according to Piaget:

--Sensorimotor Stage: Birth through about 2 years. During this stage, children learn about the world through their senses and the manipulation of objects.
--Preoperational Stage: Ages 2 through 7. During this stage, children develop memory and imagination. They are also able to understand things symbolically and to understand the ideas of the past and future.
--Concrete Operational Stage: Ages 7 through 11: During this stage, children become more aware of external events, as well as feelings other than their own. They become less egocentric and begin to understand that not everyone shares their thoughts, beliefs, or feelings.
--Formal Operational Stage: Ages 11 and older: During this stage, children are able to use logic to solve problems, view the world around them, and plan for the future.  


3 Lubkin, 347.

When I first told Patrick about his condition, he was nearly two years old; in other words, in the Sensorimotor Stage, but possibly morphing into the Preoperational Stage. Considering this information, it was probably useful for Patrick to touch and taste the cornstarch that he would consume every three hours, because that learning technique was using the child’s senses. However, the concept of the liver was probably beyond him. Patrick knew it was “inside his tummy,” but that was the limits of his understanding.

*Learning RX* also details the Information Processing Model that followed the two decades after Piaget’s initial theories. This model details what cognitive skills entail, such as “attention, short term memory, long term memory, logic, reasoning, auditory processing, visual processing, and processing speed. These are the skills the brain uses to think, learn, read, remember, pay attention, and solve problems.” In applying this model, I learned that at age two, Patrick’s short term and long-term memory were developing. Auditory processing, used for reading skills, would develop by about age five, and logic/reasoning would occur at this time too, as Patrick could begin to make connections between ideas. Thus, Patrick at age two could make an association between a lack of cornstarch and tiredness. However, he couldn’t conceptualize how the cornstarch worked, or how the liver processed foods. For that matter, neither could I. Additionally, Patrick couldn’t conceptualize the idea of “illness,” especially not “chronic illness.” Most likely, the child’s first perceptions of his GSD centered on Dr. Marris’


actions and our calm, matter-of-fact dinner conversation. Our tears, our uncertainties—they crawled out after our two kids were safely in bed.
"Help, Carol!" I had called my pediatrician friend after I had put Pat down for a nap. I had taken another personal day to regroup my thoughts. I was a snarled mess of emotions.

We had gone cornstarch crazy in our house after the visit with Dr. Marris, stirring in two teaspoons for a 3:00 a.m. feeding, as well as 6:00 a.m., 9:00 a.m., and so on. It was literally every three hours. Patrick seemed nonplussed over the new routine. "How do you feel today?" I asked him several times.

"Okay, mama. Can we go to Jumpin’ Johnnie’s Play Pit? And will you get me a new book at Borders for “Valemtime’s” Day? They have cappuccino there. I like cappuccino.”

Pat wasn’t kiddin’. He did like cappuccino. Mark and I had often taken the kids to Borders on a sleety winter’s day to buy a couple of books. Pat would end up with us at the coffee bar, one chubby leg crossed over the other, sipping “his” cappuccino in a debonair manner.

"Well, Tee will be home from school in three hours. Retha will get him so you and I can play after your nap. We’ll go to Jumpin’ Johnnie’s on Saturday with Tee, because you’re being such a good boy today!"

As Patrick’s breathing became regular during his nap, I dialed Carol quickly. “Carol. This is Janet. Well, we got in to see Dr. Marris. He thinks Pat has a recessive genetic condition called Glycogen Storage Disease. He doesn’t know which kind yet, but I got a call earlier about the needle biopsy at Children’s Mercy. Yes, it’s a week from Tuesday. Say, I’ve never heard of this GSD. Have you? Oh—you have? In medical school? A page somewhere in one of your textbooks? Well, Dr. Marris used a bunch of words I didn’t know—like ‘glucose’ and ‘glycogen.’
Heck, I don’t even know about healthy blood sugar. I just remember that with both pregnancies, I had to watch my sugar intake.”

Carol replied in her most soothing, doctorly voice. “My kids are still at school, so I’ll scoot over and bring you some of my medical school textbooks that I have at home. I’ll mark some passages on GSD and on blood sugar control. You can start learning about Patrick’s condition.”

Maybe knowledge would be power, as they say. Maybe knowledge would calm the feeling in my gut. Maybe I would feel just a bit in control of the situation.

Biopsy day came. I had been diligent with Patrick’s cornstarch routine, and it seemed to be helping—helping a lot. It was as if the child’s little inner computer was booting up and coming alive. Patrick was alert and happy, cruising down the family room couch with his stuffed Barney the dinosaur in hand, much like we had attempted months ago with physical therapy. It was a darn cornstarch miracle.

On surgery day, Mark, Patrick and I headed to Children’s Mercy, while Retha drove Tee to school. He clutched a “body box” that Mark had helped him with the night before. The box was anatomically correct, with objects from Hobby Lobby wired and glued into the recesses to sort of look like the liver, kidneys, the rib cage, spleen, and heart. I had raced through Hobby Lobby two days prior like Peyton Manning, tossing into my cart stuffed fabric fruits and any other object that might look like a body part. We knew Jimmy was going to have the best body box in the class, and sure enough, he did. There were flowing veins of “blood” circulating all through his body box. Not fair. Jimmy’s dad was a cardiologist. Lunch that day was going to be
“a body part buffet”—carrot sticks for fingers, Jell-O for intestines, and peeled grapes for eyeballs. “It’s going to be gross!” Tee told Mark enthusiastically.

The scheduling nurse at Children’s Mercy had told us “no food” before the procedure, because Pat would go under a general anesthesia. I noticed that on the way to the hospital, Pat’s head lolled to one side of the car seat. We carried him into the surgery waiting room, a floppy doll. Were we supposed to have skipped food and cornstarch? Well, the surgery nurse had said so, and we were rule followers.

After about an hour and a half, we headed back to recovery. Pat was wailing loudly, hooked up to a heart monitor, an IV bag dripping glucose into him. The beeping equipment was terrifying; Patrick looked like a miniature Tasmanian devil, writhing in the hospital bed, a nurse stroking his arm. “The type of anesthesia used in this procedure usually makes kids very anxious and angry,” the recovery room nurse informed me. Yes, Patrick was angry—absolutely screaming out of control. I leaned over and kissed his beet red, sweaty forehead. I felt so guilty—we had just put our son through a tough procedure. The child had already been scanned, poked, and stuck multiple times in his young life. He had been to five different specialists thus far, averaging about three appointments a week. However, Dr. Marris had said we needed to know what type of GSD Patrick had. The symptoms and treatment were different for each type, and we had to manage the condition properly.

The surgeon came into the room in the midst of the screaming chaos.
“We’ve completed a percutaneous liver biopsy on Patrick. I inserted a thin tube through his abdomen and took a small tissue sample.\textsuperscript{1} Pathology will analyze this sample to determine what type of GSD Patrick has. We should have some information for you both in about three to five days.” Duty done, the surgeon smiled encouragingly at us and headed out the door, tapping the doorframe with his palm authoritatively, as if to say, “That’s that.”

That \textit{wasn’t} that. The tissue sample taken was too small. It came back as “inconclusive for GSD type.” Two months later, at Dr. Marris’ strong encouragement, Patrick went through another needle biopsy, screaming just as vigorously. This time, we found out that Patrick had GSD1a, often called Von Gierke’s.

\* \* \*

“GSD1a is technically a glucose—phosphatase deficiency. In individuals with this condition, the glucose-6-enzyme is either missing or malfunctioning.”\textsuperscript{2}

It was 10:30 on a Monday night. Mark and I had supervised showers and stories. The dishwasher whirred and the coffeepot blinked at us, saying, “I’ll have the black magic ready for you tomorrow at 5:15.” I had thrown in a load of laundry and settled at my desk, pouring over more of Carol’s marked pages. There it was: GSD is “a malfunction of the catalytic subunit (G6Pse) which characterizes GSD1a . . . G6Pse is a nine-helical endoplasmic reticulum transmembrane protein required for maintenance of glucose homeostatis. To date, 75 G6Pse

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mutations have been identified, including 48 mutations resulting in single amino acid substitution.\textsuperscript{3}

I sighed, flipping to the next page of Carol’s book, rubbing my tired eyes. None of it made any sense to me.

“Humans express three glucose 6-phosphatase catalytic subunit genes identified as G6Pc, G6Pc2, and G6Pc3. Mutations in the G6pc gene are the cause of Type 1a GSD.”\textsuperscript{4}

I turned to textbook two, discouraged:

Glycogen storage diseases are a group of disorders in which stored glycogen cannot be metabolized into glucose to supply energy and to maintain steady blood glucose levels for the body. Type I glycogen storage disease is inherited as an autosomal recessive genetic disorder. Glycogen storage disease type I (GSDI) is characterized by accumulation of excessive glycogen and fat in the liver and kidneys that can result in an enlarged liver and kidneys and growth retardation leading to short stature.\textsuperscript{5}

I heaved open my huge Merriam Webster dictionary. All right. “Stored glycogen.” Mr. Webster told me that meant stored carbohydrates in body tissues. Well, \textit{that} I could understand; but what would that \textit{do} to the body? “Glucose,” I read from Carol’s book, determined to understand Patrick’s condition. Again, Mr. Webster informed: “\ldots an important energy source \ldots part of many carbohydrates.” Next: “autosomal recessive genetic disorder.” I remember that Dr. Marris told me that Mark and I each carried the recessive gene that caused mutations


\textsuperscript{5} \textit{Ibid.}
in Patrick’s . . . what was it? . . . I looked back at the book . . . the G6Pse. Autosomal recessive—
that was a medical term that meant when Patrick was conceived, he had to get two copies of
the abnormal gene for his GSD to manifest. Patrick had GSD in every cell of his body. Tee, the
older brother, had beaten the fateful roll of the dice.

My eyes traveled down to the end of the medical book entry. “Enlarged liver, growth
retardation, and short stature,” it said. The hope that had wavered after my visit with Dr.
Marris evaporated. Would Patrick always have an enlarged liver? Would he spend his life as
short, as different? Would he ever meet a girl? How could he go on a date, lugging his
cornstarch? My kid has to have a “normal” life, I thought, angry at what fate had thrown at us.
This is not how life is supposed to work. How can I control this? Control. I must have control.

Discouraged, I turned out the desk light, checked on Tee and Patrick, and rolled into
bed. The 3:00 a.m. alarm for Patrick’s cornstarch would come very soon.
PART TWO

BABY STEPS
12 Control Freak

As the weeks passed, our family settled into a “somewhat normal” routine: after all, we were now a GSD family. We all, in a sense, had to manage a chronic condition. Mark and I kept on the rigorous cornstarch schedule, determined to give Patrick every chance. Pat responded like a textbook case, I think, had we been able to find a textbook case. I had read in one of Carol’s medical books that before cornstarch was discovered as a way to maintain blood sugar, kids died from GSD. Apparently, the cornstarch discovery had only been since the mid 1970’s. There were very few adult humans with GSD to actually study. It was a new—and scary—frontier with no roadmap.

I recently read a beautiful memoir that pinged my heart as I thought back to Patrick’s early days and the uncertain, murky GSD roadmap. The memoir, Raising Henry, examines a mother’s role in raising a child with Down syndrome, otherwise known as Trisomy 21. Unlike childhood narcolepsy or Glycogen Storage Disease, Down syndrome is fairly common, impacting about one out of seven hundred births across all races. The extra twenty-first chromosome consists of over three hundred genes, creating a massive genetic puzzle. Children with Down syndrome must cope with reduced mental capacity, low muscle tone, sensory imbalances, eating and swallowing difficulties, and heart defects.

My take-away from Raising Henry is that raising a child with special needs is a continuous battle of investigation, of learning. Fortunately, with some chronic illnesses and disabilities such as Down and Type One diabetes, support systems in the U.S. are numerous. I must admit I was a bit jealous when I read that when Henry left the hospital as a newborn, the
mother Rachel was put in contact with an agency that would coordinate intervention services. The well-educated family would receive a team of therapists to help Henry work on physical and cognitive development. At present, early intervention for Down syndrome is mandated by federal law, but the resources are distributed on the state level. As Henry’s mother Rachel Adams writes, Down syndrome children from Colorado may be put on a waiting list; families from New Jersey pay for services on a sliding scale. Rachel and little Henry were lucky to live in New York State. In her family’s case, the state charged the family’s insurance and subsequently made up any costs not covered by insurance. In New York state, uninsured children are totally covered by the early intervention program. In poorer states with less tax revenue, the intervention programs are not as robust, entailing waiting lists. Meanwhile, the Down syndrome child and family manage as best they can, operating in the dark.

I think back to when our family finally got Patrick diagnosed at twenty-one months. Our intervention “program” was a slip of paper with the phone number for NORD. That was it. Mark and I knew little about how to nurture Patrick or what to expect of him developmentally. Part of the issue with Patrick was that Mother Fate dealt him a rare disorder, affecting about one in one hundred thousand births. Research didn’t exist for early intervention when Patrick was born in 1992, because children with GSD had only just begun to survive. Most physicians could vaguely remember studying GSD in medical school.

I remember that Dr. Marris at Children’s Mercy hospital finally gave us the name of a child nearby us in Shawnee, Kansas who had GSD1a. I think I had finally badgered the man into submission. Patrick was about six at the time, and I called to arrange a meeting with the family. One Sunday afternoon, the Shaw family eagerly met the Sawyers.
Billy Sawyer came racing into his home, wiping his sweaty face on his red-striped tee shirt. He had been out playing soccer. I was so relieved: here was a twelve-year-old with GSD who was simply out playing with his friends. Perhaps Patrick could ride a bike, swim, go to high school, go on a date some day. Possibilities opened up to me like two prayerful hands cupped upward. As Billy gulped down a glass of water, his mom Grace filled two large syringes with cornstarch and water, fitting a nasogastric tube on the end of one of the syringes. I’m sure I stood dumbstruck as Billy maneuvered the NG tube up his nose, down through his esophagus, and into his stomach, compressing the attached syringe and releasing the starch into his stomach. He grabbed the second syringe, fitted it onto the NG tubing, and squeezed again. Billy handed the syringes and NG tube back to Grace with a “Bye, Mom,” and zoomed out the door to his buddies. The cornstarch routine was just that—routine to Billy.

Hope lifted us up as we drove back that afternoon to our home at Lake Quivira.

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One day when we were still fairly new at GSD care, I headed down the stairs after work to the family room. It had been a doozie of a day. One department member came to me upset, thinking she should get to go to more training seminars each year. At her job level, company policy allowed her two per year; she insisted on four, harrumphing out of my office with a glare that nearly seared my eyebrows. The 1099-INT’s didn’t print off in the computer room, either. A programming problem. We had to get it figured out, because federal law mandated that the INT’s had to go out before January 31st. The federal government wasn’t very touchy-feely about this sort of thing. On top of all this, I heard rumblings of a new merger as I was making copies, which meant that my department would have to either assimilate or add benefit plans. That
would entail endless meetings with Legal, Ernst and Young consulting, and the programmers. Voluminous communications. Hand-holding the shareholders. It never ended, but it was also exhilarating. Come feast and famine; war and peace; snow and blazing heat—you didn’t mess with employees’ benefits.

Turning around the corner at home, I met Retha, breathless. It was only the second time I’d ever seen her breathless.

“Just ten minutes ago, Patrick took four steps! He was cruising down the couch and saw Barney over by the coffee table and went for him. Four steps!”

I was thrilled but also envious. Retha had witnessed something Mark and I had been waiting on for the last five months. And I had missed it. What was I doing, going to work every day? How could benefit plans ever compare to Patrick’s first steps? Did I have to choose my work over my child; why couldn’t I have both?

That night: joy. Patrick took more steps for Mark and me. Tee, my freckled bundle of energy, had said to Patrick, “If you walk over to me, I’ll give you a booster-jet ride in your dee dee.” And so, Pat simply walked to big brother and gave him a hug.

* * *

The next day I stood in the copy room, my feet aching from the navy heels. I absently ran off copies of executive deferred compensation plans, looking up at the bulletin board in angst. I was bone tired; or as my Grandma Julia used to say, I was all out of spizerincktom. I’d gotten up with Patrick for his 3:00 a.m. feeding; back up again at 5:15 to shower and dress for work. Mark was flying back and forth to Paris for his latest project, and so Retha and I had to power through the weeks. Would I ever understand Patrick’s condition? Would I ever feel like I
could get my arms around all the chaos at work? And then—I looked up and there it was on the bulletin board. It was a message, I think, for me.

_Footsteps in the Sand_

One night I dreamed a dream.
As I was walking along the beach with my Lord.
Across the dark sky flashed scenes from my life.
For each scene, I noticed two sets of footprints in the sand,
One belonging to me and one to my Lord.

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“When you saw only one set of footprints,
It was then that I carried you.”---Mary Fishback Powers

“It was then that I carried you.” The words felt like a warm, comforting hand on my shoulder, and unprofessional tears fell onto my stack of copies. It’s funny how twenty-six years later, I remember my sense of desperation contrasted with an extreme moment of peace. I knew that God wouldn’t sweep in and “fix” Patrick. There were no guarantees that Patrick would get better. There were no guarantees that Patrick would live. There were no guarantees that my work would settle down. Life was, well, life. Life was Georgie chasing after Tee as he somersaulted across the back yard. Life was feeding the goats at Farmstead with the boys. Life was that stolen moment of wine with Mark after a crazy day. Life was spit-up on my shoes and baby crap on my sleeve.

Well, control freak, I think, let go. You don’t “have” life; it “has” you.

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Patrick seemed to be taking his new medical condition with childlike calmness. He trotted Barney the dinosaur for short walks all over the house: on the couch, the coffee table,

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the stairs. “Barney, we’ll go to the park with the twisty-turney slide tomorrow. But, you have to put your toys away first. Here. In this red bucket. Don’t let me tell you twice. And it will be time for your cornstarch soon.” The words just exploded out of that child.

I, however, wasn’t taking Patrick’s medical condition as well as he.

Control and discipline have always governed my life. It is my white picket fence that keeps everything mentally corralled. Even when I was four, I had my Little Golden book collection sorted neatly. My Barbie dolls sat in a row on my bedroom shelf, followed by my troll dolls, tallest to shortest. As an adult, my clothes closet was arranged by color, with sub-categories of summer versus winter, and sub-sub-categories of short sleeve versus long sleeve. My spice rack was alphabetized from allspice to wasabi. I had my MBA and I flossed nightly. Check. Check. Check.

Thinking back to my childhood, I realize why I felt like I had life by the tail in my 20’s. I was taught in childhood that with education and hard work, you could get what you wanted in life. If that didn’t work, you’d grab onto your situation like a pit bull with a dead squirrel, shake it, and work some more until you got what you wanted. You stayed on plan.

I came by control and discipline naturally. Mom and dad were both teachers. They had met in 1950 at Hutchinson High School in Kansas. Dad was teaching typing and shorthand there, while Mom taught English and ran the school newspaper. Story goes, Dad met Mom at a faculty school picnic, as Mom, dark-haired and skinny, was in the food line heaping raw carrots and celery onto her paper plate. Dad wowed her with a real pickup line: “You don’t like carrots much, do you?” The magic flowed, and Mom and Dad got married in 1952. I arrived, quite conveniently, 363 days later. Right on plan, of course.
I don’t remember much of Hutchinson but more of Emporia, Kansas. We had moved there when I was six, because Dad had gotten a finance teaching position at the state university. Mom easily got an English teaching position at a small Presbyterian college just a mile from our home.

Life was tranquil and predictable in Emporia. On Sunday mornings, Mom would pop a roast into her blue speckled pan before church, dumping in peeled potatoes and carrots and two cans of beer for tenderness. Weekdays were a rush, with a glass of Tang for breakfast and Rice Krispy cereal with whole milk. Monday was meatloaf, green beans, and baked potato night. But, on Thursdays, we got wild. Mom would buy Swanson chicken pot pies at Kroger—four for a dollar. They’d bubble for an hour in the oven, the crust turning a magical tan. You see, Thursday night was choir practice at the Congregational church down the road. Mom reckoned that the pot pie shortcut was okay since we had to rush out of the house at 6:45. I’d sit in the front pew at the church as the choir hashed and rehashed “Seek Ye the Lord.” Dad had a solo part: anxiety sweat would run down his face as I would read Nancy Drew’s The Secret of the Old Clock.

When I was growing up, Dad always retreated to his office in the basement after supper to prepare for his 7:30 a.m. finance class. Mom always had a clipboard full of themes to grade, her red stick pen placed neatly across the top page. Every Saturday, Dad would cut the grass or tackle small repair work. Mom would get her hair “done” into a helmet of dark brown curls and then head to Kroger. I would clean our two-bedroom ranch to a spit shine with Pledge furniture polish, singing at the top of my lungs to “Eight Days a Week.” The Beatles. To a ten-year-old, they were hunky.
Several times a year, Mom would decide to treat Dad to a homemade pie. The raisin pie efforts had fizzled after Mom’s two-year desperate bake-a-thon, thanks to Nan Wiley’s cooking column and scores of little old ladies. Dad would be out on a stepladder in the Kansas fall glory, wearing paint-splattered khakis. He’d hang his old transistor radio on the top rung of the ladder, painting the house sideboard vigorously and listening to Kansas University football, his alma mater. His favorite game was KU against Oklahoma, the big rival. “Dammit! Come on, KU,” Dad would holler at the radio, slapping the paintbrush against the siding. “Dammit, KU! RUN!”

Mom would come into the back yard and say, “Steve, I think I’ll make a pie. What kind would you like?”

“Uh, I think I’d like cherry.” Dad always asked for cherry. Every time.

I don’t think my parents meant to be so precise, so disciplined. They couldn’t escape their childhoods, any more than I can escape mine to this day.

My dad was born in 1922, the union of a Congregationalist minister and Lillian Hayles, the organist at a church in Leavenworth, Kansas. Grandpa Stephen had lost his first wife, and at age fifty, the lovely Miss Lillian caught his eye, as she filled the church with “Faith of our Fathers.” Lillian made the pipe organ at the church absolutely rumble, and most certainly, Reverend Butcher’s heart. The woman was both pretty and smart. She had graduated from Beloit College in Wisconsin in 1918 and had gone on to the Kansas City Conservatory of Music, where she majored in the pipe organ. At age thirty-two, Lillian was considered a spinster, but the Reverend changed all that. The two married in 1921, calmed the wagging tongues in Leavenworth, and moved to Hutchinson, Kansas to a larger church. In 1922, my dad—the
blonde, curly-headed Stevie—was born. He was a gift, for Lillian had been thirty-three when she became pregnant in 1921.

Life does twist. We know this intellectually. When it happens, however, it stabs our gut because it’s not really supposed to happen to us. And, true to life’s twists, in 1929 as the Great Depression was erupting, Reverend Butcher caught pneumonia and died. Lillian was left with the parsonage, a tiny pension, and seven-year-old Stevie.

Years later, as I read about my dad’s life, I could see why my mom always took care of him; just like Lillian had taken care of dad as a child. I could see why my dad was an uncertain, anxious adult. His father and comfortable life were ripped from him in the middle of the Great Depression. Lillian raised Stevie while baking cinnamon rolls to sell out of her home. She played the organ at McPherson’s Mortuary in Hutchinson. They got by. They barely got by.

With one foot planted firmly on my dad’s backside, Lillian hustled him into a local community college after high school. After serving in World War II, dad worked summers at Sears and Roebuck, selling lawn equipment and paying his way through a BS in Finance at the University of Kansas. Later, when he was teaching at Hutchinson High School, dad would take summer school business classes for his master’s degree at Pittsburg State. Lillian’s foot on the backside then switched over to my mom’s foot. In 1960, as Dad moved to an assistant professorship at Emporia State University, he began coursework in finance at Indiana University in Bloomington, Indiana. Doggedly, Dad would teach Introduction to Corporate Finance and Management during the school year in Emporia. Mom would teach Shakespeare, steadily plowing her way through a stack of student themes each week. Every summer, for six years, my family would drive five hundred miles to Bloomington, where we sweltered in Everman married
student housing. I’d play kickball and jacks with all the other kids whose daddies were in school. After dinner, I’d work through my weekly pile of library books. We had no television, no telephone—just a main room, a bathroom, and a tiny kitchen. I’d fall asleep at night on a cot in our apartment, just inches away from Dad’s desk. I’d hear the soft flip-flip of pages as Dad poured over macroeconomics. The window box fan whirred and sucked in the humid Bloomington air. Mom typed up Dad’s papers on the thirty-pound Smith Corona typewriter. Peck-peck-peck-zing! She was fast.

When I think about my mom, she was no slouch, either. Mom’s father, George Malcolm, was an attorney in Pittsburg, Kansas. Julia was a legal secretary at his law firm, and at age thirty-two, she made George’s torts tingle. Marriage came soon after. My mom Jeanne was born the next year in 1920, followed in five years by sister Janet.

The Great Depression years were not as tough on the Malcolm family as most. Even in the midst of a crisis, the townspeople of Pittsburg still needed an attorney. Grandpa George would settle a drunk driving charge in court, and two cases of fine local wine from Chanute would arrive on the front porch of his home. When Grandpa George helped Garner’s furniture store with mortgage issues, Lonnie Garner gave the family a beautiful, forest green area rug for the living room.

Mom went to Pittsburg State College undergrad. I still don’t know how, but she ended up at Northwestern, obtaining a Master’s in English with a 4.0. By age twenty-two, she was teaching English in Ellsworth, Kansas, moving a few years later to the big town of Hutchinson. “Ellsworth is at the damn ends of the world,” Mom would often say to Dad.
Mom broke rules. When she and dad got married in Hutchinson, she kept teaching at the high school and local tongues flapped. *Did your husband lose his job? Are you in money trouble?* Blue-haired ladies would pepper Mom with questions in the checkout line of Dillon’s grocery. When Mom got pregnant with me precisely as planned, Hutchinson really talked. The high school suspended Mom’s teaching contract, because she was beginning to “show.” “The kids can’t see a pregnant lady teaching them verbs,” the principal explained to Mom, as though her growing mid-section would lead the high school kids to rampant sex in the halls. When I was one year old, Mom went back to teaching—this time—geometry, as the school math teacher had inopportunistly died in the middle of the school year. I stayed with Mrs. Prop next door, a widow woman that radiated love and chocolate chip cookies. Again, the town yapped: *Jeanne, you’re taking a man’s job. Why are you doing this? Why aren’t you home with your child? Are you both in financial trouble?* Mom just had to work; she was so damn smart. Dirty diapers were no match for congruent triangles. I get that.

Virginia Nicholson in *Perfect Wives in Ideal Homes* writes about the era I grew up in. The 1940’s represented a time of war and privation, while the 1960’s began to blow apart traditions. The 1950’s? Well, they were a forgotten decade until recently. Only 1.2% of women actually went to a university at this time, Nicholson estimates. In the midst of the fifties, my mom just didn’t fit: she was educated and career oriented.

I am a true product of my past. As the poem “Footsteps” settled into my heart that day in the copy room so long ago at work, I realized that instead of control and discipline, life was

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full of uncertain turns and a series of choices, if I were really lucky. Should I work outside the home, or stay home with the children? Could I do a little of each? Grabbing up my stack of deferred compensation data that day, I headed to my desk to scribble out a quick pro-con list for myself. Fifteen minutes later, I rolled my chair back from my desk. There it was: chicken-scratches of alternatives.

The pros of continuing work were several. First off, I listed financial security and insurance. That was so important with a child—especially an ill one. Then, there was the exhilaration of using my brain: solving those accounting problems or delivering an impactful benefits presentation. And then, the Big One: my ego. I wanted and I needed to feel intelligent and live out my dream as a career woman. I had grown up surrounded by an aura of education and careers. It’s just what you did as a woman. Anything less was sub-par.

My pro list for becoming a stay-at-home mom was equally strong. Unfortunately, internet systems in 1998 didn’t provide work-at-home capabilities. No Amazon Prime or Whole Foods existed to deliver the endless demand of diapers, cereal, and hamburger to my home. Plus, my boss was a traditionalist. My department needed one manager in the office—not job sharing. And—that manager had to be there physically, five to six days a week.

So. Working from home in 1998 or job-sharing was just not an option for me. I dealt with an either-or situation. What would staying at home mean for me?

My heart told me that staying at home meant I could see those precious boys more. As a working mom, I heard a lot off buzz about “quality time with my children.” Shouldn’t all my time be quality time, I wondered. Also, if I stayed at home, I could pull the stress load off Mark. We could have great home-cooked meals. Mark and I could have time to talk at the end of the
day, instead of falling into an exhausted stupor at 10:30 each night. I could be at all the boys’
activities. I thought back to all the mother-daughter tea events at my middle school, so many
years ago. My mom could never come because of her teaching schedule. Each year, I felt like an
abandoned orphan at the tea. It just hurt. Did I ever want my boys to feel that sense of loss?
13 Dear Mom

Mom and I wrote to each other once a week. We only lived one hundred miles apart, but long-distance calls were for emergencies only. You called when someone had died, was dying, or was going to die. One time when Mom actually called me, her poodle Nicki had gone to the great kennel in the sky. Another time, I picked up the phone, a bit anxious when I saw Mom’s area code.

“Hey, Mom. What’s up?”

“Could I stay with you and Mark for a week or two?” Mom never bothered with chit-chat. Her voice was measured and even.

“Of course! We’d love it. Is Dad coming too?” Mom and Dad were always together, like Mutt and Jeff.

“Well, he’s coming from Emporia to the Kansas University Medical Center by ambulance. He had a heart attack and is in Newman Hospital in Emporia. He’s being transferred to KU Med and the doctors think he will need a quadruple bypass.”

“Oh, good grief! When did this happen?”

“Yesterday morning. Steve was eating breakfast and then started having chest pain. I called an ambulance and got him over to Newman. Thank God it’s only a mile away. He’s being transferred to KU tomorrow.”

It had been hours and hours and Mom had finally called. Oh, good grief, Mom.

* * *

Shaw  64
Mom,

I’m so glad Dad is back home and that his heart is doing well. It was a long three weeks at KU Med, wasn’t it?

I know we talked about Patrick’s condition while you were in Kansas City and I showed you all the cornstarch supplies. Pat and I had another appointment with Dr. Marris yesterday. I peppered him with my usual list of questions about GSD. At the end of our appointment, he looked at me over his glasses and had the nerve to say to me, “You ask a lot of questions, don’t you?”

Well, Mom, I didn’t want to anger our only local GSD expert. I just gave Dr. Marris my best withering teacher stare and said, “I haven’t even gotten started with the questions yet. Patrick’s my child and I have to understand what’s going on with him.” I couldn’t muster a glare exactly like you used to give me, but it was pretty darn good.

I think Dr. Marris was taken aback. He rifled through his file cabinet and found me a listing of what Patrick should eat and what he should avoid. I know we’re coming down to Emporia to see you and dad two weekends from now, so here’s a list that may help you with meal planning. Well, the bad news is that your cherry Jell-O made with Coca Cola is out. And your cherry pie. But your beef stew is in . . . it’s the best in five states.

Mom, here are the main things Patrick shouldn’t eat. Dr. Marris explained to me that because the child’s liver is compromised, certain substances cause the liver to become inflamed or to store more glycogen. His new diet is kind of like managing a child with Type 2 diabetes. We can’t let Pat’s blood sugar go above 120 or below 80. Dr. Marris said something about if the blood sugar goes too low, lactic acid will build up. I need to find out more about that.
Dr. Marris says that Pat should have complex carbohydrates like whole grain pasta or wheat bread, but only about fifteen grams per meal and five per mid-day snacks. Those carbs, like cornstarch, take longer for Patrick to digest and so his blood sugar will stay normal longer. That’s why he has to drink cornstarch and water every three hours. Apparently, the cornstarch provides Pat a steady amount of glucose because it is more slowly digested than other complex carbohydrates, even slower than wheat bread. Anyway, we started putting the Argo in water, because I found out that milk contains sugar. Patrick and I mix up cornstarch and water in two plastic cups and make a contest of it. Who can drink the cornstarch first? Well, Mom, the mixture tastes like Milk of Magnesia gone bad, but I tell Pat the mixture will give us super-powers. He’s walking all over the house and his fingernails have even started to grow!

The handout also says that Pat shouldn’t have lactose. I looked that up—it’s in milk and cheese. Fructose, which is in fruit, is definitely out, as is Sucrose. Here’s what the handout says, “galactose and fructose are converted to unwanted glycogen stores, lactic acid, fatty acids, and uric acid which can be harmful to the body in large quantities.”¹ It looks like Patrick can have small amounts of Equal, Sweet N’ Low,, and Tapioca syrup. Maybe Crystal Lite? It sounds like it would be easiest just to avoid deserts altogether. I don’t know what we’re going to do about birthday parties.

Patrick can also have almost any type of vegetable, but not more than one packet of ketchup with a meal. Since when is ketchup a vegetable? Well, most plain meats are okay but

not too much fat. Here’s some good news. Pat can have plain popcorn, pretzels, baked corn chips, and saltines. I have a feeling that more than a bite or two of Chucky Cheese’s pizza is going to be out. Thank goodness Patrick likes crunchy and salty food. He’s just like me. Give me corn chips over brownies any time. Raw carrots dipped in a bit of Ranch dressing. We can do this. We will do this.

I must go throw in a load of laundry. Tell Dad hi and to stay off the ice cream.

Love, Janet
14 Disaster

Cornstarch. Dr. Marris. Jumpin’ Johnnie’s. Tee’s concert at school. Haircuts. Groceries. Laundry. Dr. Wigginton. Bloodwork. Ultrasounds. Life rolled on, in an ultra-busy-but-smooth way, I thought, considering we were a GSD family now. Mark ping-ponged back and forth to Paris. He hated travel, but we needed his job, his insurance. The needle biopsies, blood work, and ultrasounds had come to thousands of dollars, even though we were lucky enough to have insurance.

Tee came home one day from school, his pool-blue eyes watery; his freckled nose red and gooey. “My throat feels spicy, like my socks,” Tee whined to me.

I spent two days at home with Tee, catching up on work at night. Tee and I watched hours of Looney Tunes, sneaked in mac and cheese when Pat napped, and built gi-nor-mous Lincoln Log towers. Mostly, we rocked in my old oak rocker, reading Frog and Toad and singing “Amazing Grace.” Tee would hunch down on my shoulders, swiping his gooey nose periodically. When I took him in to Dr. Wigginton, he didn’t find a fever or strep. It was just “one of those nasty viruses” and we had to wait it out.

On Wednesday, Tee was ready to head back to school. I know he was feeling better, because we had the usual morning battle over clothes. I was tired and Tee won. He went to school wearing Ninja Turtle clothing in every color, I remember. Oh well, he was on the mend. We drove to school in my old powder blue station wagon, a small pool of oatmeal jiggling in the cup holder. It had been there for five days.

That night, when I got home from work, Retha was holding Patrick. He looked limp and his eyes were glassy.
“This little guy’s not feeling well.”

I could tell. Pat felt a little warm to the touch, and sure enough, the thermometer told me 101. I had been in meetings all afternoon and Retha hadn’t been able to reach me. Our department assistant had been out with the flu.


That night I slept poorly. Dr. Marris and I had not talked about what to do if Pat got a cold or the flu. That man was pure scientist. Dr. Marris’ idea of patient instruction was to throw the phone number for NORD at me. At any rate, I got up with my alarm at 3:00 a.m. I needed, I wanted Mark to be there. He was always the wise sage; he would know what to do. I felt so inadequate.

Pat was still hot; 100 now. I rocked Pat and sang to him, trying to get some warm milk and cornstarch down him. Patrick would not open his mouth.

At 6:45 a.m., I checked on Patrick.

Patrick lay on his side between two rolled baby blankets—the way the pediatrician taught us, to prevent suffocation. I could barely see golden curls and the curve of Patrick’s cheek—one hand curled near his mouth. Patrick’s chest rose softly up and down.

I put my hand gently on Pat’s cheek. What was it?
I hesitated for a second, then two, then three. Didn’t it seem that he was breathing faster than normal? I racked my brain. Had I ever seen this with Tee? A gentle up down, up down, like a tiny bellows. One, two, three, four. That fast.

It was nearly seven a.m. I ran to my desk and called the number on my memo pad. Dr. Marris’ emergency line. I got an irritated-sounding attendant.

“Uh, hello? This is Janet Shaw. My son Patrick is a patient of Dr. Marris. He has Glycogen Storage Disease. Glycogen Storage Disease. Glycogen—ahhh!—Listen—Patrick’s really sick. He’s breathing funny, I think; too fast. We need to come in. Yes, I’ll hold. Please hurry.”

Ten minutes later, Retha had arrived to handle Tee’s morning routine. Cornstarch bag packed, I bundled Patrick into the station wagon. Children’s Mercy Hospital was a good thirty-five minutes away, through downtown traffic. As I drove, the blocks crawled by. Tick. I looked back at Pat. A breath in, a breath out. One two three four. Tick.

I carried the limp Patrick into Dr. Marris’ office. He looked at Patrick for a few seconds on the exam table and barked, “Follow me!”

I scrambled down the hall after Dr. Marris with Patrick in my arms. I hadn’t known the doctor could move so fast. We burst through one set of scratched grey doors. A turn. Another set of doors. Another endless hallway. Into the emergency room. Onto a gurney.

“I need an IV bolus of glucose. 5 percent glucose with .45 sodium chloride and with potassium 20 mml. Non-lactated ringers. NOW!” ER personnel scrambled as Dr. Marris barked orders. I backed away from the gurney. The doctors knew we had a child clinging to life.

Within seconds, one doctor had a needle in Patrick’s arm, changing the needle for a small plastic tube and taping the area securely. Pat was so sick that his wails were weak,
pathetic. The doctor drew vials of blood from Patrick’s arm and handed them to a nurse, who scurried away to the lab. Another nurse swiftly placed a catheter from the plastic tube in Pat’s arm into the nearby IV bag and Kangaroo pump. Within just a few minutes, the pump clicked, dripping life-saving glucose into the sick child.

I sat shaking two hours later in Patrick’s hospital room, one arm draped over the rail of Pat’s bed, touching—just touching—those curls. The reality smacked me: I had nearly lost my child. As Patrick drifted off to sleep with the steady click of the Kangaroo pump, I quickly ran to the nearby nurse’s station and called Mark collect. It was about 7:00 in the evening in Paris. I caught Mark at the hotel, and he made arrangements to catch the first flight out to Kansas City. I called Retha and that saint—that red-headed saint—called her family and told them she would be at our home that night.

“What happened to Patrick?” I peppered Dr. Marris with questions as soon as he came into our hospital room at about 10:15 that morning. Guilt and shame had been creeping into me. This couldn’t be my baby: sick, lifeless. I had thought I had been doing GSD “right”—maybe I was just an incompetent mom?

“Patrick has metabolic lactic acidosis from the flu. When he wouldn’t eat or do his cornstarch, his blood sugar dropped below normal. His body’s level of ketones built up and acidified the blood, making lactic acid rise. Lactic acid should not be above 7.4. In other words, Patrick’s blood profile had begun to crumble. Also, his liver was unable to remove the excess lactic acid. That’s why he was tired and breathing so rapidly. We’re running several tests every three hours on Patrick. I’ve requested a basic metabolic panel to check his kidney functioning.
We’re also looking at Patrick’s calcium, protein, blood sugar, and electrolyte levels. We’ll also catch a urine sample.”¹

“So, I should always bring Patrick into the hospital any time he gets the flu?” I was beginning to get the picture.

“Any time Patrick won’t eat or do his cornstarch, you need to bring him in immediately. Immediately.” Dr. Marris paused to catch a quick breath and to look at me over his readers. “This goes for strep, chicken pox, high fevers, or any vomiting. Patrick’s body cannot fight off these conditions without having blood sugar issues.”

I wished I had known this from the get-go.

As if on cue, a sweet-faced nurse came in the room to complete the three-hour blood sample. She paused, a kind hand on Patrick’s forearm. “Well, Mr. Patrick! I’m Janice and I’ll be with you until 10:00 tonight. Then Miss Ira comes in. We’re going to get a little blood from George here. See? This little port here is called George. He takes all the sticks and the blood draws—not you! No more ouchies, ok?”

Janice tickled Patrick under the chin for a second and was rewarded by a weak smile and big blue eyes that studied her.

In the next eight days, George got stuck every three hours, day and night. IV bag solutions got changed; the Kangaroo pump clicked away. Click. Get better Patrick. Click. Mom’s here. Click. I’m so sorry, Patrick. Click. The days became a blur of faces: the doctors; the young residents with their clipboards and nervous faces; Dr. Marris’ assuring presence; the dietician;

the cleaners; our minister; Mark’s anxious and drawn face; friends with stuffed dinosaurs and Mylar balloons. The support was overwhelming.

And the Lord whispered, "My precious child, I love you and will never leave you
Never, ever, during your trials and testings.
When you saw only one set of footprints,
It was then that I carried you."²

Patrick and I rocked in the hospital room each day, watching Barney and Mr. Rogers for hours. Holding his dee dee, Patrick sang along weakly: “I love you; you love me; we’re one happy family . . .” One day, he turned his head to me mid-rock and said, “When can Tee-Tee come give me a dee dee ride with booster jets?” Amen. Patrick was on the mend.

Meanwhile, my little world of “tackle problems like a pit bull” had crumbled. My Plan A of a successful life in control was in shambles; Plan B had taken off, with me chasing the illusive ghost, breathlessly.

Twenty-six years ago, Patrick nearly died from the flu. In thinking back on his life, I wanted to read about other children with chronic illnesses and disabilities. What had I done wrong? Did other parents live in a world of doubt, of fear, like I still do?

*Lubkin’s Chronic Illness* textbook tells me that illness “. . . is the human experience of a disease and refers to how the disease is perceived, lived with, and responded to by individuals, their families, and healthcare professionals.”¹ A disability, on the other hand, is a “physical or mental condition that limits a person’s movements, senses, or activities.”² A chronic illness or disability is even more complex in character. Lubkin indicates that the chronic element includes “permanency, residual disability, non-pathologic alteration, and required rehabilitation or a long period of supervision.”³ The text indicates that the degree of an altered lifestyle can relate to the patient’s perceptions and beliefs about the disease—more than the disease itself.⁴ In Patrick’s case, his mutated gene was in every cell of his body. No magical medicine existed to “fix” Patrick—all we could do was manage the condition—every three hours, for a total of 2,920 times per year. A slip-up of six hours could mean a seizure or coma; a slip-up of twelve hours, death.

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¹ Lubkin, p. 4.


³ Lubkin, p. 5.

⁴ Lubkin, p. 4.
As I read twelve memoirs of children and adults with chronic conditions recently, I intentionally picked a variety of issues: spina bifida, narcolepsy, multiple sclerosis, cerebral palsy, autism, Trisomy 8 Mosaicism, severe food allergies, and Type 1 diabetes. What could I learn from other caregivers? How did their lives compare to my experience?

My first ah-ha was that as a parent, I had tended to assume that my Patrick could or couldn’t do certain things, simply because he was chronically ill. This assumption is so ironic, because a central theme of this memoir is how a parent can raise a child to be independent, given the parameters of the chronic condition.

In Cora Means Heart, Cora’s mother struggles with the same issue of assumptions: most certainly a chronically ill child can only do so much. She writes, “Children with spina bifida see a lot of doctors.”5 Yes, they certainly do. Children or adults with spina bifida have a congenital defect of the spine. Part of the spinal cord and its meninges (membranes) are exposed through a gap in the backbone. This can cause paralysis in the lower limbs and can also cause mental handicaps. Cora, like many others with this chronic condition, has a shunt or a tube in the brain. The tube tunnels under the skin with the other end of the tube ending in the abdomen, which then drains off excess cerebrospinal fluid. Unfortunately, Cora needs the shunt replaced periodically, entailing surgery. On top of that, she continues to have ultrasounds of her kidneys and bladder as well as extensive blood work to check for kidney and bladder functions. Her bladder must be drained via a catheter. Cora’s condition means that she can’t walk without braces and her growth hormones are low. Does this sound like a lot of medical issues? You bet.

J.A. Busick, Cora’s mother, is a force of courage. She writes about her older daughter, Ashley, who began playing organized basketball at age three through the YMCA in Owensboro, Kentucky. Cora, at age three, wanted to play the sport too, even though she had just started walking with braces and had a tube running through her. Busick admits that Cora’s basketball interest was “nowhere on my mental map of life.”6 Neither was having a disabled child.

Busick found a program in her hometown called Upward, where “every kid plays, and every kid scores.”7 Now, when I read this, I wondered, how can a child in braces ever play basketball? As the old expression says, “never say never” to a determined mother and a hopeful child. Busik relates that at the end of the first season of basketball, the coach put down Cora’s crutches and lifted her up so she could drop the ball into the 8-foot high basket. Busik marvels, “The crowd went wild. It was Cora’s first basket.”8 Busik reveals that by the end of Cora’s third year of basketball, she had taught herself to shoot one-handed into an 8-foot goal. Busik was amazed that the disabled or chronically ill kids could do things she herself couldn’t do.

I’m just as guilty as Busik with my assumptions. I think back on the time when Patrick was on a summer swim team with Tee. Somehow, I had volunteered as swim team coordinator, even though I barely knew a flip turn from a dolphin kick. At that time in the 1990’s, we lived in a sleepy lake community west of Kansas City called Lake Quivira. Tee was twelve and reigning

6 Ibid., p. 30.
7 Ibid., p. 31.
8 Ibid., p. 31.
king of the butterfly stroke and the breaststroke. Pat was six, with a moderate Buddha belly. Pat would typically muster through the backstroke for one lane until his energy gave out.

The city kids were daunted by swimming at Lake Quivira. Our dock formed a three-sided angular “C” into the lake, jutting from a sandy, man-made beach. We adults would get the ribbons ready, rope off the lanes, dig out the bullhorn, and ready the score pads. Then—most importantly—I’d get fish feed out of my backpack. Each swim meet, one swimmer got to do the honors. Plink! Plink! Plink! The kid would run around the dock, flinging the feed into the warm, murky lake. Within seconds, catfish and bluegill would swarm the waters—right before the city kids walked down the beach to the dock.

Our secret weapon for each meet was Catfish Charley. Now, Charley had been hanging around the Lake Quivira waters for three or four years at least. Charley loved two things: fish feed and nibbling on city kids’ toes. At a hefty length of three feet, Charley simply unnerved our competition.

Patrick would swim the backstroke faithfully each competition. I’d walk along close to his lane, and wait in angst, ready to jump in to rescue him. Stroke, gasp, sink, float up. Stroke, gasp, sink, float up. Each time, Patrick would rally to the water’s surface while I readied to plunge in. Somehow, he always finished the lane and got the yellow fourth-place ribbon. Patrick was determined. If Tee could swim, so could he. Why not?

At the end of summer championships, I drove Tee to Bonner Springs, Kansas to compete. No Catfish Charley to help us there, but earlier that day Tee had scooped up a vial of precious Lake Quivira water. Plink! Plink! Plink! The brownish lake water splattered into the fancy Bonner Springs pool. Thus, Charley swam with us in spirit. I could just see him waving
back and forth slowly, mottled, whiskers twitching, wise eyes taunting: “We’re gonna get you, city kids.”

Patrick was at Farmstead with Amy during the final tournament, because his total season points were low—a succession of fourth place ribbons. I knew there was no way he could compete against the jocks from Bonner, Turner, and Shawnee. No way.

“Patrick Shaw.”

I had just been over to the judges’ booth to check on Lake Quivira’s total points.

“Patrick Shaw.”

_Crap. Oh, crap._ The announcer was calling Patrick to compete. And Patrick was thirty miles away, oblivious, throwing feed to goats. The child could have competed—should have _competed_, and GSD wasn’t the culprit here. Amy wasn’t the culprit. Patrick wasn’t the culprit. I was. I had assumed Patrick wouldn’t compete at the championships because of his GSD. If I couldn’t truly believe in him, then who would? I resolved to never—to never—_ASSume_ what Pat _could_ or _couldn’t_ do. How could he learn independence if I didn’t truly believe in him?

Patrick’s situation at the swim trials is so much like Cora’s in _Cora Means Heart_. Never tell a child with GSD that he can’t swim and compete. Never tell Cora, a child with spina bifida, that she can’t play basketball. In all likelihood, you’ll get surprised.
After Patrick’s eight day stay at Children’s Mercy Hospital, life resumed to normal, sort of. By this time, Patrick was nearly age four. It was time again for the bi-monthly appointment with Dr. Marris.

“Hmmmm.” Dr. Marris palpated Patrick’s stomach, measured his legs, checked his fingernails. He gently touched Patrick’s pink cheeks that had become less Gerber baby-like over the past few months.

I spun into mother alert mode. What was “Hmmmm”? Dr. Marris adjusted his glasses and looked at a recent ultrasound report. “Patrick’s liver has shrunk quite a bit, which is a good thing. It means glycogen is not being stored in the liver as much and the cornstarch is doing its work. And he’s walking well. Great vocabulary—do you read to him a lot? However, Patrick’s growth rate—see on this chart? It’s not where it should be. Here’s the average for a four-year-old male. Down here is Patrick. I recommend we have a gastrostomy tube placed into his stomach for an intake of Tolerex at night. It should be the additional nutrition Patrick needs to catch up on his growth. Children with GSD, even the males, will only grow to about 5’4”, unless we take extra steps.”

I had no idea what a gastrostomy tube was, or Tolerex, for that matter. And, I had forgotten my notebook and pencil. Dr. Marris had a way of blasting me with words.

“Can you tell me how this works? How does the Tolerex get into Patrick’s stomach at night? What is this tube you are talking about?”

Dr. Marris gave me one of his looks. “Tolerex is a formula that delivers 100% free amino acids, but with very few calories from fat. It’s normally used with people who have a severely
impaired GI function. We can use Tolerex for tube feeding or oral use. After Patrick has a
gastrostomy tube placed through the abdomen and into the stomach . . .”

“Wait, wait!” I interrupted. “Surgery? A hole clear into his stomach?”

“Yes,” continued Dr. Marris, slightly irked. “Our pediatric GI surgeon will cut here.” Dr. Marris touched the left side of Patrick’s abdomen, away from the liver. “A temporary 12” Foley will be placed in the incision. The tubing on the outside of the stomach will need to be securely taped. After the healing period is over—say three weeks—the GI surgeon will place a much simpler Foley in the abdomen called a Mic-key button. It lies flat to the skin’s surface. It has a balloon inside the stomach keeping it in place, as well as a locking mechanism on the top, so you can twist and lock tubing from an IV bag containing the Tolerex. You’ll have a home health care nurse show you how to operate the Kangaroo pump that meters out the Tolerex through the night. While Patrick is on nightly Tolerex, he won’t need cornstarch until morning. The Tolerex will have to be special ordered at your local pharmacy.”

Thankfully, Mark was in town for a couple of days. That night, he read stories to Tee. I could hear him: “Yes, I love you this much. Thiiiiis much! Thiiiiiiiiiiiiis much!” In the other bedroom, I had selected a children’s book of Bible stories for Patrick. We had just finished learning about Jacob and his coat of many colors. We had looked at pictures of Moses and the burning bush. You could say we were on a theological roll that night.

“Does God ever have to take a bath?” Patrick interrupted me.

I was flummoxed. My spiritual background was definitely iffy. I’d been to church every Sunday my whole life, because my father was a “PK.” You went to church unless you were coughing or burning with fever.
I’d always thought of God like Charlton Heston in *The Ten Commandments*. I was sure he wasn’t as good-looking as Heston, of course, but thought that he would have a long gray beard and flowing robes. Perhaps stern, flashing eyes. After all, he had to do a lot of smiting in the Old Testament, and I spent a good deal of my childhood trying not to be smote. I believed in God, of course, but He was kind of like someone on the other end of a 911 call. I could get Him if I needed Him. Mostly, I could handle life. At least I thought so until Patrick was born.

“Uh. Well. Uh. I don’t know if God even has feet.” *What a lame reply, Janet.* “Maybe he’s like a gentle spirit. Like a soft fog that wraps around us and loves us very much. Do you remember that day we saw fog in the woods by our house? Actually, Patrick, I don’t really know how to tell you about God.”

Patrick reflected on this, silent. I can still picture him clutching Barney and flipping his dee dee against one cheek. Our theology was good, for now. Little did I know there would be much tougher questions later on.

With the kids in bed, Mark and I sat in our family room, debating about the gastrostomy tube. It would be Patrick’s third surgery in three years. We didn’t know what to do. All surgeries carried risks. Were we right in subjecting our child to yet another terrifying experience?

At the end of the pow-wow, we decided we had to give Patrick every chance to grow—to be his best, whatever that might look like. A child with a “forever” cornstarch habit, but a child with a chance. The surgery was on.

* * *

The gastrostomy at Children’s Mercy went smoothly. We had learned from the previous two surgeries to coordinate with the attending anesthesiologist beforehand. Patrick was
allowed some light food and cornstarch the morning of the surgery. Once he was under the general anesthesia, the specialist would pump Patrick’s stomach, switching him immediately to a glucose drip during the procedure. The drip had to stay in place for two hours after Patrick woke up until he could hold down cornstarch, water, and a light snack.

The day before the procedure, we had told Patrick he was getting a special button in his tummy for a few years—kind of like he’d seen on the Coke cans at Grandma B’s house. I showed Patrick a packet of Tolerex, which I special ordered from Rite Aid. It would become Patrick’s fifty-six dollar a week habit for some time to come. I opened a packet and let Patrick look.

“See? It’s just a powder and we’ll mix it up every night in water before story time. It will go into your tummy very slowly, all night long. You’ll grow and soon get as tall as Tee!”

“It stinks!”

Patrick was right. Once he was on the Tolerex regimen, I would dump a packet and sixteen ounces of water in my blender at night, watching the milky goo whirl and slop. The smell would knock you over; it was like rotting library paste. I typically had about thirty seconds to pour the mixture in the IV bag and scrub the blender. The Tolerex mixture would set up and I could have patched my tires with the stuff.

Patrick never knew the struggles we had with our insurance company over the Tolerex. Our first insurance claim got fired back to us like a pop fly off of Derek Jeter’s bat. I re-filed, including two letters of medical necessity—one from our pediatrician, and one from Dr. Marris. Certainly, a letter from the head of medical genetics from Children’s Mercy . . .
Nope. The claim was fired back. “Your insurance does not cover nutritional supplements. See paragraph 17, clause two.”

I fired back with equal venom. “It’s not a nutritional supplement. It is to help our child with GSD grow appropriately. This is GSD—a rare endocrine condition. Here is another copy of the letter of medical necessity.”

Nope. A three-week period of silence from the insurance company. Another rejection. The doo-loop continued four more times.

In desperation, Mark walked in one day to the office of the vice president of human resources at his work. Fortunately, the two had been in meetings together before.

“See these applications and letters of medical necessity? We’ve tried everything. Can you help us?” Mark fanned the forms out like a loaded deck of cards.

Sharon—the VP—had no children of her own. She was, as we say, a tough cookie in the business world; most of her coworkers were afraid she would gut them and string them up. In fact, her secret nickname was “Attila the Hun-y.” “Just a minute,” she said. Sharon flipped through her Rolodex and dialed a number.

“Yes, this is a claim from Mark Shaw on behalf of child Patrick Shaw. Here’s his ID number. Actually, there are six claims for Tolerex. You see them? Okay. Pay them. From now on.”

That quickly, our Tolerex costs were reimbursed the next eight years. As it turns out, Mark’s company actually paid for employee benefits while the health insurance company was merely the administrator. However, it struck me: what would it be like to raise a child with no
insurance? No job? With no Mark? With no VP to turn to? The reality was that insurance and medical care worked well for those of us with connections.

Fortunately, Tolerex is covered by our government’s Food Stamp program. However, Medicaid places many restrictions on Tolerex and, more importantly, gastrostomies. According to the Finnegan medical supply website, “Insurance will only cover feeding tubes if the patient is 100 percent tube fed for all nutrition and liquids. This means that you can never feed the patient by mouth. If a DME company is told a patient can take even a spoonful of liquid orally, they legally cannot provide the feeding tube, and all the materials will have to be paid for out of pocket.”

1 Tolerex, kangaroo pumps, and home health-care nurses are expensive. What about a chronically ill child whose parents don’t have insurance? He doesn’t get the proper care.

* * *

The harsh truth is that not all parents with special needs children are lucky enough to have jobs or insurance. *Waking Mathilda*, by Claire Crisp, illustrates just this point. The author was a physical therapist in London, with three children and a husband who was a professor. When the H1N1 pandemic of 2009 spread throughout the world, Crisp had her daughter Mathilda vaccinated with Squalene, according to the British government’s recommendations. The seemingly innocuous, five-second vaccination went awry. Crisp found out later that this vaccine had been tested only on a small portion of adults and on no children. Within weeks, Mathilda began having nighttime hallucinations. She would sometimes collapse during the day, folding in half at the waist and falling to the floor. She began to turn quiet, introverted. She

slept during the day, waking up for minutes, only to fall asleep again. She developed ataxia, or muscle weakness of the legs and speech organs. After two years of doctors and tests, Mathilda was diagnosed at age three as the youngest patient in history with narcolepsy.

*Waking Mathilda* focuses on the sheer monumental task of a mom and dad raising a child who sleeps, collapses, and hallucinates twenty-four hours a day. The parents must care for their two other healthy children at the same time—and money is tight. Oliver, the father, is a college professor struggling to stay employed. Crisp writes about the year 2005, when Oliver was between teaching jobs: “Despite the string of qualifications, degrees, publications and professional experience, we were both officially unemployed, disillusioned, and broke. Were it not for my parents owning a tiny and spectacularly cute Victorian cottage in the heart of the New Forest, we would also have found ourselves homeless.” The family literally scoured the nearby forest for firewood to heat their home and for fruit for the kids. The milkman dropped off milk for the kids but always managed to throw in a loaf of bread. To compound the injury in this dilemma, the couple had previously been at Notre Dame for two years, and thus were rejected for unemployment benefits in the UK because they had been absent from the country for a year. Imagine. Two healthy kids and a third who is tremendously ill. No fallback. No money.

The narrative is every parent’s nightmare as Claire and Oliver work for months to get a diagnosis for Mathilda. Finally, obtaining a diagnosis, the parents press and push physicians in order to get approval for an experimental drug known as Xyrem, compounded only in one pharmacy in Missouri, and costing a resounding $3,000 per bottle. Xyrem is also known as

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Sodium Oxybate—a highly controversial drug—notoriously abused as a date-rape drug with a huge street value. At Bristol Children’s hospital—one of many sources Claire uncovered, she pushed Dr. Jensen for Xyrem. His response was ambivalent: “I would have to look into it. It’s a very expensive drug and difficult to get a hold of . . . we would need to make an application to the board and justify why the National Health Service should spend thousands of pounds when Xyrem isn’t proven to help children as young as Mathilda. Then they will argue that for everyone like Mathilda, the same amount of money could treat, say, five other children who also need specialized medication.”\(^3\) Two months later, Claire pressed Dr. Jensen again at an office visit. In despair, she remembers that the doctor reminded her, “ . . . a year’s supply would cost our local health authority twelve thousand pounds.”\(^4\)

In the midst of the NHS red tape, Mathilda was fading before her parents’ eyes.

As a final attempt to get help, the Crisp family moved to the United States near Stanford so that Mathilda could get help. Oliver was fortunate to get a teaching assignment at Stanford, and thus, incredible medical insurance became available. Had he not been employed, Mathilda’s medical options would have been slim to none. Through Dr. Emmanuel Mignot, Professor of Sleep Medicine at Stanford, the family finally got a prescription for Xyrem.

*Waking Mathilda* has a proverbial happy ending. Mathilda finally receives management for narcolepsy, and Oliver, the father, still teaches at a prestigious university with a healthy insurance program. However, everyone is not so lucky. In fact, most of the world’s population isn’t.

\(^3\) Crisp, p. 172.

As I think back on my son’s numerous appointments, tests, scans, surgeries, and ER visits, I remember the horrible stress of what is it? What if? The Shaw family paid plenty of money out of pocket to help Patrick. However, we had the benefit of jobs—good jobs—and insurance. What must it have been like for Claire and Oliver Crisp to have medical uncertainty heaped on top of financial issues and government red tape? Here is my take-away. Most parents work really hard to parent their chronically ill children. Most parents love their chronically ill children. However, what makes the difference between a thriving Patrick Shaw and a child dying in inner Chicago is money, insurance, and great health care. That’s it. There’s no moral superiority involved when it comes to healthcare—just money.

I think back to the nights in ER at Children’s Mercy with Patrick, as we waited in the 3:00 a.m. haze of despair. However, numerous children in that same waiting room—mostly black—needed to see a doctor, too. Any doctor. These were kids with fevers and vomiting episodes. They were waiting, because no other hospital would take them. They didn’t have pediatricians. They didn’t get well-checks. They didn’t get vaccinations. The kids and parents waited, because they were part of a faceless group in the US. That faceless group is the uninsured.

What happens to chronically ill children when mom and dad don’t have jobs or insurance?

In 2010, the Affordable Care Act provisions went into place in the US. Along with this was Medicaid expansion on a state-by-state basis and the establishment of Health Insurance Marketplaces. However, in 2018, “… 27.9 million nonelderly individuals were uninsured, an
increase of nearly 500,000 from 2017.” According to KFF.org, most of our uninsured are low-income families with at least one worker in the family. In fact, in 2018, 70% of nonelderly uninsured individuals worked for an employer that did not offer them health benefits.

Additionally, many of the uninsured are black—half of the nonelderly uninsured population. The website also reports that individuals with income below 200% of the Federal Poverty Level are at the highest risk of being uninsured. In 2018, eight in ten of the uninsured were families with incomes below 400% of poverty.

Why on earth would people not get insurance? Pretty straightforward, really. According to KFF.org, “. . . in 2018, 45% of uninsured adults said that they remained uninsured because the cost of coverage was too high.” Many jobs don’t provide insurance coverage and sadly, some poor adults live in states where Medicaid coverage has not been expanded. They get the double whammy: poverty, but not enough poverty to qualify. Additionally, it’s quite common for the poor to not even know where to turn for help. And if you’re an undocumented immigrant—forget it. You can’t apply for Medicaid. You try to pay for your bills with cash as you get it. Mostly, you try to stay under the radar and away from Immigrations and Customs Enforcement. Lawfully-present immigrants under 400% of poverty are eligible for Marketplace

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6 Ibid

7 Ibid.

8 Ibid.
tax credits, but only those who have passed a five-year waiting period after receiving qualified immigration status can qualify for Medicaid.\textsuperscript{9}

Like the children sitting in Children’s Mercy ER in Kansas City, one in five uninsured adults go without needed medical care, according to a 2018 study.\textsuperscript{10} Those uninsured, if they can even get to health care at a hospital, normally pay higher rates than do the insured patients. KFF.org documents that, as a result, “. . . uninsured people are less likely than those with insurance to receive preventive care and services for major health conditions and chronic diseases.”\textsuperscript{11} Lack of outpatient care then results in hospitalizations for normally avoidable health conditions. Quite often, being poor and/or uninsured simply means imminent death. A mother doesn’t bleed less for the death of a child simply because she’s poor or black.

* * *

I had never been good at working with equipment. Even when I would use a screwdriver on a pesky cabinet door, I always used the mantra, “Lefty loosey; righty tighty.” So, it was with relief the day the home healthcare nurse arrived with a shiny new Kangaroo pump, IV bags, tubing, an extra Mic-key button kit, medical tape, and assorted syringes. Not to mention a travel-sized Kangaroo pump backpack. I eyed the array of equipment with anxiety.

Retha and I trained with the home healthcare nurse; I worked with Mark and Tee later that evening. The Mic-key button and Tolerex became a nighttime routine. Quite often, Patrick

\textsuperscript{9} Ibid.

\textsuperscript{10} Ibid.

\textsuperscript{11} Ibid.
would flip and thrash around in the night. The IV line would crimp, causing a piercing alarm that made Mark and me sprint to Pat’s room. Sometimes, the securely taped Mic-key button would unlock from the IV tubing. Oblivious, the Kangaroo pump would continue its click, click, click. We’d awaken to Patrick’s howling, as the sticky Tolerex ran over his pj’s, the sheets, and onto the floor. On other occasions, about every two months, the inner balloon of the Mic-key would deflate. Out it would slip, exposing the hole going into Patrick’s stomach. Mark and I would fly into Patrick’s room to give him an emergency dose of cornstarch, because the Tolerex could not get into the stomach with a defective Mic-key. We’d tape the Mic-key back into Pat’s stomach, pack extra cornstarch, bundle up the sleepy Pat and Tee, and drive to Children’s Mercy ER. It was Murphy’s law that the Mic-key always broke at 3:00 a.m. on a twenty-degree Kansas City night, as the stars crackled in the sky. The four of us would get back home at 5:30 a.m., just in time for Mark and me to shower up and head off to work. Retha would come and let Tee sleep in for the morning, his face pale and freckled from the 3:00 a.m. scramble.

About the fourth time our family had its middle of the night sojourn to ER, I decided to consult with Dr. David Roberts, a pediatric gastroenterologist at Children’s Mercy. We had to do something about the frequent races to the hospital.

Dr. Roberts looked like Robert Redford—just in his early forties. With his boyish haircut and blue eyes, he seemed like the guy next door.

“See, Patrick? Here’s a new Mic-key button for you. Since you’ve been growing so well, we’re going to go up a size. The new button has a stem that is a little longer. And see here? This is the balloon at the end of the stem that will hold it in your stomach.”

“It doesn’t look like a balloon,” Patrick countered.
“Well, I’m going to show you how to put the new Mic-key in and then blow up the balloon. It’s kind of like magic. Okay . . . first, we always wash our hands with soap and water. We sing ‘Happy Birthday to You’ so we know we’ve washed long enough.”

Patrick, Dr. Roberts, and I sang enthusiastically. I looked over at the young doctor and thought, *Huh. Tone deaf.* A couple of nurses peeked in, smiling. “Now, let’s stand here by the exam table and gently pull out the old button. You *gently* pull. See? It comes right out, because the balloon on the end has collapsed. Now, we’re going to put a bit of lubricant on the tip of the new button. It will go into your stomach slick as a whistle. Then, we’ll push the Mic-key in until the pop-top is right up against your skin. Do it gently. Don’t worry; your little stomach hole is all healed up.”

Patrick was nearly five at this time. The world was pretty straightforward. If Dr. Roberts said to gently push the Mic-key in the hole, then that’s what you did.

“Now, we have a bowl of distilled water here on the exam table. Take the little syringe that came in your Mic-key kit. Pull it apart and dip this end in the water. Good! You’ve filled it up most of the way. Now place that black rubber tip into the other part of the syringe. You’re going to push some of the water out. You only want eight millimeters of water in the syringe. See the marking? This is when Mom can help you. Perfect! Okay--see the little valve on the side of the Mic-key? Over there--it’s on the side. There, you found it. Place the tip of the syringe into the valve and push aaaaaaaaaaaaaaaa the water out of your syringe. Yes, that’s it. You did it!”

Patrick surveyed his new Mic-key with interest. It lay flat and perfect against his skin.

“So how come you had to go to school *for so long*?” Patrick questioned Dr. Roberts.

The doctor’s blue eyes crinkled with amusement. I longed to hide.
“Well, a few things I do are harder, Mr. Patrick. You did a great job! Maybe medicine will be in your future.”

From then on, Patrick was the master of all things Mic-key.

Mic-key button profile as shown by Kimberly Clark.

From www.handtohold.org, accessed on 8/1/20. Mic-key button with tubing to a Kangaroo pump.
“I think Patrick is bored.” Retha and I were having one of our end-of-workday briefings. “I know I’m talking myself out of a job, but I think Patrick needs to go to school. We go to story time at the library, see kiddie movies, and enjoy the petting zoo in Lenexa, but it’s not enough. When Tee does his homework after school, Patrick gets his pencil and does “math,” too. He’s trying to teach Barney how to read. He’s Mr. Social. He needs other kids around.”

Retha was always right.

Tee’s school was half an hour away from home but very close to my work. He had been in school since age four, too, and loved it—despite nightmare encounters with science fair projects. I still cringed when I thought of our latest project last month—a platform made with popsicle sticks that had to hold ten pounds. Well, ours didn’t. Tee had also bitten his kindergarten teacher, Mrs. Parker, five years ago. But only once and it wasn’t very deep. At any rate, Mark and I made an appointment the next week to meet with the Early Years and Pre-K teachers after the school day.

“I recommend that Patrick start in the Early Years program, even though he could handle the cognitive activities in Pre-K,” stated Mrs. Miller, a dark-haired sunshine beam of a woman. I still remember Patrick wandering over to the Book Nook corner and then over to miniature easels. And the back of the classroom—it was magnificent! Sliding red numbers in slots. A huge clock with movable hands.

I was sold; I wanted to go to Early Years myself.

When Mark and I got the kids to bed that night, my fretting started. School meant Patrick leaving his safe cocoon of family and Retha. I had to make sure that Mrs. Miller and the
school nurse understood Patrick’s condition perfectly. Thank goodness—I worked only five minutes from school, should something happen. I made a mental note: *keep my cell phone with me at all times*. Our department assistant loved to take lengthy smoke breaks.

That Saturday, Patrick went down for his nap, because even a normal day tired him. I went to work, planning. I scribbled: *To Do’s for School:*

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--Description of GSD1a--simple but accurate.
--How to mix up cornstarch and administer. Use large syringe into Mic-key for cornstarch—like at home?
--Need schedule for dosing. Must find out snack times and lunch at school. When is PE?
--Emergency contacts
--Supplies: fresh boxes of Argo monthly, plastic containers, large syringes, tubing for the Mic-key, sugar-free Citrucel
--Have supplies in the classroom and the nurse’s office?
--Keep extra supplies in my car?
--List of foods for Patrick to avoid. How to handle birthday parties?
--Low blood sugar warning signs
--Buy string pack to hold all of Patrick’s supplies.

I set my Pentel down and rubbed my head. I think I had my to do list. I set about creating forms on my computer. Thank goodness I had gotten some helpful pages from NORD. I left voicemails with Mrs. Miller and the school nurse. I hoped I could document and communicate enough so Patrick’s school day would go smoothly. It was step one for a healthier childhood. How independent could Patrick ever be? I didn’t have a clue, but school was a big step forward.

* * *

Dear Mom,

I know you are worried about little Patrick starting school so young. But as a professor, I know that you love education and I think Patrick will, too. Remember—you were finished with your masters and out teaching in western Kansas by age twenty-two. Well, I think Patrick has
your brains. Just the other day, I was asking the boys to pick up their toys before bedtime.

Well, Tee was popping his Legos into the toy tub—he’s such an obedient kid! But Patrick, good grief. Here’s how the conversation went.

“I don’t need to put my blocks up. I’m going to use them tomorrow. It’s unnecessary."

*Unnecessary?*

“Mama, I’m pretty sure you got all these toys out, so you should pick them up.”

“Dad can do it. What has he done all day?” I looked over at Mark, sprawled out on the couch, *Time* magazine open, eyes closed and mouth open.

Mom, I just stood there during this conversation, speechless. Finally, I fell back on every parent’s lame mantra: “Because I told you so.”

Well, Mom, sad news. We’re going to lose Retha, because she needs to work full-time. She’s like a sister and a freckled wise sage, all rolled into one. I did talk to Carol Morrison at church. She knows a young girl finishing up her Elementary Education degree. Anyway, her name is Amy and she has one year left of college. She wants to work part-time, so I’ll have her come over and chat with me. When both boys are in school, we’ll need someone to pick them up and do snacks and homework.

Back to the school issue. I called NORD last week and one of the staff members faxed more articles to Mark’s office. So, here are some snippets I found. Can you read them to Dad, too? I know he’s worried about Patrick.

“In many ways school is a microcosm of society in general, and, in interacting with teachers and pupils, the child with a chronic illness will learn how to present their illness
outside school and experience how their condition is perceived by others. Common to the majority of children with chronic illness is the desire to be treated the same as their friends. “1

Mom, we don’t know how far Patrick can progress, but I just want him to do what other kids do. Well, do whatever he can. Oh here: I just found two more entries:

“Every child needs to feel part of their school community. They need to take part in sports, school fundraising, drama productions and school trips.” 2 I guess we’ll figure how Patrick can do these when we get to that point.

“Written information, detailing how the child is affected by their illness, the use of medication or treatment and symptoms to be watchful for, should be made available to the principal and then disseminated around the key staff members in contact with the child.” 3

You know Mom, as I just wrote this down, I realized that I need to get all the medical information to Patrick’s PE teacher, too. He’ll need to understand that Patrick can’t handle long stretches of physical play. He’ll need to understand what Patrick means when he says, “I’m cornstarch tired.” And speaking of cornstarch tired, I’m tired, too! Bye for now and with hugs, Janet


2Ibid., p. 130.

3Ibid.
18 The Big Day

I was anxious that morning. Very anxious.

The blue station wagon was packed to the gills. Tee had his backpack and a huge black trash bag. Each kid had to bring one hundred objects to school and teach the other kids division problems using the objects. Tee had chosen one hundred red cups. Not something small, of course, like pennies. One hundred large Solo cups. We had practiced division the prior night, lining up twenty cups here and ten over there. “See? Ten cups go into twenty” . . . (slide slide) . . “like this. How many times did we do that?”

Tee was a whiz at reading and PE, but math was mysterious.

“Two! Three! Two and a half!” Tee shot out answers, hoping to hit the target, any target.

Patrick was strapped in behind me in the car. He had a new backpack: Barney, of course. It was chock full of extra clothing, underwear, and approved GSD snacks. The string pack was inside, also, with pre-measured containers of dry cornstarch, Level Life glucose gel, a large catheter tip syringe and tubing. We had been giving Patrick’s cornstarch to him through the Mic-key button, because the starch was like drinking chalk. And Patrick was slow.

I had been up since 5:15 a.m. that morning, fortifying myself with two hits of coffee, showering, applying makeup, and selecting a suit for the day. Yes, this one. A black and red plaid pleated midi skirt with a black boiled wool top. A turtleneck. One gold chain. Earrings. My black Timex. Dark pantyhose. Demure black flats. The corporate look was complete. Watch out, world.
Mark would usually get breakfast ready for the kids at seven. I’d flip two English muffins out of the toaster, slap on some butter, and throw my gourmet breakfast into foil. At 7:15, we’d load the car: two kids, two backpacks, math project, my breakfast, and my gym bag. Mission Control: ready for departure.

I phoned Mrs. Miller at four o’clock of Day One of Patrick’s Adventures in Academia.

“How did Patrick do?” I enquired, trying not to sound like a helicopter mom, while truly being a helicopter mom.

“He was adjusted by noon,” the teacher replied. I could almost hear her beaming.

I’m sure I let forth a relieved whoooooo. “Uh. I forgot to tell you when we had our “cornstarch” meeting. If Patrick ever starts to throw up, please call me immediately. I mean, right away. If this happens, he can’t keep his blood sugar up. You can give him some glucose gel that’s in his string pack, but I’ll still need to take him to the hospital. His lactic acid will probably elevate.”

Mrs. Miller may have been quaking on the other end of the phone line, but she gave no clue. “We’ve got it under control here.”


The school schedule was going swimmingly, I felt. Until one day, a week later.

Patrick’s IV line had crimped twice during the night, sending me out of a deep sleep into a sprint down the hall to his room. Mark was in Paris that week. The middle-of-the-night chaos created bathroom trips, two rounds of “Amazing Grace” and a loooooooong back rub. The next morning, I was bleary-eyed. Despite my two cups of morning courage, I forgot to put an extra outfit and underwear in Pat’s backpack.
We pulled up to the front of campus that morning. Tee hopped out with his backpack and a huge rolled up diagram that we had helped him with last night. It was sort of, kind of, a little bit like, wanted to be, could have been a depiction of a double helix. Good grief. Why couldn’t the kid ever have a simple book report on *Gone with the Wild*? As Tee burst in the front door of his building, we circled to the back of the campus. I parked in front of the Early Years building and walked around to the back seat to help Pat.

There it was, simply everywhere. Apparently, something had disagreed with Patrick, and that disagreement, brown and sticky, had headed onto Patrick’s clothes, squishing onto the car seat in all directions. This was not a simple Wet Wipes cleanup. This was a four-star emergency.

I made a quick phone call to work, wheeling the wagon around towards home. Thirty minutes later, I carried a towel-wrapped Patrick into the huge laundry sink in our home. I stood him up, peeling layers of brown sticky clothing off of him, dropping them into a bucket filled with Spic-N-Span suds and warm water. Sprayer and soap in hand, I tackled Patrick’s tummy, legs, feet, arms, hands. Wait. How did the brown disaster get behind one ear?

Back in the car. A clean Patrick. A car seat that smelled like Lysol. Thirty minutes later, we retraced our steps to the Early Years building. Patrick joined in with art time, immediately forgetting the morning mess in the excitement of Tempera paint. I got to my office fifteen minutes later, opening up my Franklin Planner at my desk to assess the day’s agenda, taking a shaky breath. Okay. Meeting with programmers in fifteen minutes. Employee review at one. Oh no. It was the lady that was always in a huff with me. The chip on her shoulder was the size of Texas. A stock plan update with my boss at three.
I slammed my planner shut and took a swig of coffee. And there---*there it was*---on the cuff of my suit coat. A lovely, large, brown reminder of the morning. Ground deep into the fibers and there to stay.

Did I still think I had life under control?
Weekends were the best, as life slowed down by ten ticks.

On Saturday mornings, I’d grab a huge mug of coffee and milk and both boys would sit with me on the carpet, nestled inside my legs. “Birdie in the nest,” the boys would call it. We’d watch our favorite cartoons: *Scooby Do*, *The Jetsons*, and, of course, *Teenage Mutant Ninja Turtles*. Mid-turtle adventures, the boys would both hop up and yell, “Turtle Power!” That was my cue to grab cup number two and resume the nest position.

Saturday afternoons were playtime for the kids. I remember one day in particular. It was autumn in all its Kansas perfection. The tang of smoke circling from my neighbor’s leaf pile. Warm sunshine pulling dry muskiness from the earth. It was a day where nothing bad could happen.

I puttered around the cool house in my jeans and fuzzy sweatshirt. I folded dingy dish towels and tossed Legos into a cracked red toy bin. Peace. Quiet. Contentment in my heart. Tee, his neighbor Scott, and Georgie galloped across our front yard and into the woods next door. A crumbling house foundation—a cool fort enticed the three into the dense woods. Patrick, reveling in his five year-old glory, trotted slowly after the big boys. His blonde curls bounced with every step.

I circled to the front door and yelled at Tee.

“Hey! Make sure you take Pat with you, okay? Watch him.”

“Sure, Mom.” Tee’s freckled, lanky arms flashed as he zoomed through patches of sun and into the treasures of the woods.

I wandered into the kitchen and began frying up hamburger, onions, and garlic.
Spaghetti. My mouth watered as I tasted my batch. Humming a few bars of “A Bushel and a Peck,” I wiped my oniony hands across my apron. The apron was my favorite--white, with red piping and rickrack. “Kiss the Cook!” it advertised across the front.

Footsteps thumped across the back deck. Tee, Scott, and Georgie burst into the house. Tee dumped his treasures onto the Formica countertop: an old transistor radio, four bolts, and a rumpled paperback with a buxom blonde on the cover.

“Mom--we’re starving! Can Scott stay for dinner?” Scott, one of six kids next door, sniffed the simmering spaghetti sauce and looked at me expectantly. In his household, anything beyond mac and cheese was gourmet.

“Sure.” I flicked on a light switch to inspect my hamburger concoction. I poured grains of salt into the crook of my hand, letting them flit into the spaghetti sauce. “Tee, where’s your brother?”

“Don’t know.” Tee picked through his treasures, eyeing the buxom blonde on the book cover. “We were exploring the woods and got hungry. I thought he was following us.”

A tickle of fear feathered in my gut. Patrick needed his cornstarch in about an hour.

I trotted out of the house and into the network of trees, now fading gently to blues and greys. Hidden dark pockets. Leafy dampness.

“PAT!” I called.

No answer.

Tick.

In fifty minutes, Pat’s blood sugar would begin to drop. And drop. And drop--to thirty.

Running now, I sped back to the house. “Stephen! Scott! Find Dad and start looking for
Pat, NOW!”


Forty minutes left. No Patrick. The silence screamed at me.

My husband sped down our street in the dusty blue Volvo station wagon, as Pat’s empty car seat straps flapped in the wind. His new favorite toy, Penny the Panda, tipped sadly in the back seat. Meanwhile, I ran back up to the house and called the Lake Quivira fire department. Sirens soon began to wail. Police cars cruised the lake perimeters. Patrol lights revolved, piercing and slicing the blackening woods.

Tick.

I raced across the street, yelling to my neighbor Debbie for help. She hopped on her old golf cart and roared down the street and into the green velvet of a nearby golf course.

Tick.

Fear circled my stomach, pushing its black tendrils into my heart. I prayed again. *Not this, God. Not this. Help me. I’ll do anything for You.*

I ran back into the woods--circling, circling, asking God for the crunch of footsteps or a flash of blonde curls. I grabbed a tree trunk, gasping for air, listening. Faint yelling echoed across the woods.

“Janet! Janet!”

I sprinted into my front yard. What if? Sweat broke out; I smelled my fear.

I heard Debbie’s cart come to a painful, dieseling halt. There she sat. And there . . . there
was Pat with chocolate on his Gerber-baby face and an empty wrapper in his fist.

I ran up, grabbing, sobbing, thanking God.

“Pat! Don’t ever do that again! We didn’t know where you were.”

“It’s okay, mama,” Pat replied matter-of-factly. “I didn’t know where I was, either.”

Mother meltdown. Relief, tears, joy, anger, guilt.

*Stamped into my primal gut now is the preciousness of life.* The dice rolled one time and Pat wandered off. Roll two—he could have collapsed in the woods, sliding into shock. Roll three—he could have wandered into the lake. Roll four—my neighbor just happened to find him in the middle of a golf course, eating a Hershey bar.

Life can flip in a minute. Had I allowed Pat too much independence at age five? Was it fair for me to burden Tee with the task of watching his little brother—again?
Tee was a shadow child. What a perfect name for a child who lives as a second-class citizen because of a sibling’s chronic illness or disability.

I recently read about Randall Beach, who is a practicing attorney now. Growing up, he was the big brother to Jon, who had been diagnosed with Type 1 diabetes. Beach’s book *Shadow Child* hits frighteningly close to home in describing the life of a sibling who grows up second fiddle to the ill brother.

Randall writes of the time Jon was first diagnosed with Type 1. Jon was in the hospital for a few days getting the condition under control. Randall states, “I needed to know that Jon still existed and was going to come back home. It had only been a few days, but to my seven-year-old mind those days had been weeks... being without a mom was worse. Grandma and Auntie always made sure I was taken care of and fed well... they were not adequate substitutes for Mom. I needed my Mom. I wanted her back.”

I think about Beach’s reaction to Jon’s hospital visit. Ironically, my two children were about the same age when Patrick spent days at Children’s Mercy. My husband managed to juggle work and hospital visits, pulling in his mother for help and stringing together hot dog meals for Tee. Tee’s homework and sporting events went on the back burner, as did the nighttime stories.

The sibling of a chronically ill child not only misses his parents, but he also sees the ill child getting “special” treatment. Beach writes that he was a bit jealous of Jon’s novelty dinners

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in bed in the hospital. He remembers, “It seemed that my brother was having a pretty good
time in the hospital.” In like manner, this is what our son Tee saw. Patrick’s time in Children’s
Mercy meant multiple staff visitors, Mylar balloons, and meals in bed. It meant Mom rocking
him for hours. It meant all the cartoons he could handle. Meanwhile, Tee got hot dogs, school,
a missing mom, and homework.

Beach also describes his feelings when he was brought into Jon’s care routine—much
against his will. On occasions, Jon’s blood sugar would drop, or he’d have a spike after an insulin
injection. One time, the family was heading back from the Boston Science Museum when Jon
began screaming and shaking in the back of the car. Speeding down the Mass pike at sixty miles
an hour, Beach’s mother handed big brother a wad of honey packets from the glove box. Beach
hopped in the back seat. He recalled, “I pressed my body against my brother’s jerking
movements and tore open the first honey packet. When Jon’s mouth opened with a scream, I
thrust the packet into the side of his mouth and squeezed . . . his jaw clamped tight and a small
gurgle came from somewhere; then it opened again with another scream. All at once, through
slits in my brother’s eyes, I saw his eyeballs roll back into his head. He was having a seizure.”
Fortunately, mom was close to Camp Joslin, a special diabetes summer camp that Jon was
attending. She wheeled the car into the camp, where trained doctors got Jon into the infirmary
and stabilized him. Within an hour, Jon was back to normal and trott ed off, laughing with a
camp counselor, to enjoy archery. Meanwhile, Beach sat on the porch of the main camp

\[2 \text{ Ibid., 43.} \]

\[3 \text{ Ibid., 59} \]
building, sobbing from the rawness of the afternoon. He had been traumatized by being so close to a life-and-death situation. He recalls, “My most important job was to take care of my brother . . . No matter what happened to me, I was to be thankful that I was spared the disease borne by Jon.” 

In similar manner, our son Tee was often too involved in Patrick’s emergencies. Tee was dragged to ER many nights when Patrick was vomiting or needed a new foley. Tee was privy to all the midnight kangaroo pump alarms and Patrick’s wails if his blood sugar dipped.

Beach makes another important point about being a shadow child. He states, “. . . from my perspective, my baby brother fast became my babied brother in many ways. Jon’s well-being became my mother’s number one priority, and his happiness became her second . . . I am confident that my mother wished the same for me then and does to this day. The difference was that she had to be actively engaged in the management of my brother’s health and in doing so, became actively engaged in the management of his happiness.”

As I read Beach’s words, I thought, convicted! Mark and I tried our best to divide our time between Tee and Patrick. However, GSD is a jealous and demanding mistress. When Patrick would have a blood sugar episode, all attention flew his way. End of story.

Beach indicates that, for all the reasons described above, jealousy entered into his relationship with Jon. He recalls one particular incident involving a log cabin.

Jon and Randall had a crude lean-to out in the woods near their home—much like the fort Tee and Patrick loved to play in as children. Over time, Jon began to take an interest in log cabins, so his parents decided he should have a real one. This project involved clearing the

\[4\text{Ibid.}, 65.\]

\[5\text{Ibid.}, 75.\]
woods, pouring a concrete slab, and building a two-story structure, complete with four working

glass windows, an asphalt roof, and a door. It was essentially Jon’s cabin.

*Interesting,* I thought as I read about the cabin. *This sounds like the time we bought a shed for Patrick.*

When Patrick was about nine, he, too, longed for a “real” fort or playhouse. Oh yes—he also wanted a trampoline.

“You would always know where we were, Mom,” Pat would argue. “We wouldn’t track mud into the house. We could play in the shed. We could nurture our creativity.” *Nurture our creativity? Oh brother.*

The arguments would continue non-stop. “The trampoline would be a cool place for all my friends to hang.” “Think of the exercise!” “You don’t want me playing video games, do you?” “We could put our bicycles in the shed. It would give you more room in the garage.”

Patrick would inhale deeply and continue for another fifteen minutes non-stop.

It is true what Sharon Dempsey writes in *Extreme Parenting.* She describes, “It is all too common to try to over-compensate for the child’s health problems. Many parents find that they want to give their child everything to somehow make up for the deficit that fate appears to have bestowed.”

Had Dempsey been watching the Shaw family through a figurative window? Much like the journalist described in her book, Mark and I finally caved into Patrick and bought a $1,000 assembled shed. With cute white siding and black shutters, it was every kid’s dream. Additionally, a trampoline arrived, becoming the social center of the

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6Dempsey, p. 1.
neighborhood. Both boys were living the dream, except this was really all to make Patrick happy. And Tee knew it.

Did Tee act out because he was the shadow child? Of course.

When Patrick was in grade school and Tee was in high school, Tee’s favorite trick was to wait until Patrick was in the shower. Tee would head into our back yard on a hot summer afternoon, where cicadas perched chirping on our trees. Tee would scoop up his buggy friends, sneak into the bathroom, and dump twenty flailing cicadas on Patrick’s head. You could hear the screams for miles. Occasionally, Tee would keep his pranks simpler. Once, he put a bit of Ex-lax in Patrick’s almond milk. Four bathroom visits later, Patrick determined the culprit. Another time, Tee sneaked into the bathroom, again, during Patrick’s shower. This time, he sprayed copious Lysol disinfectant into the air, running out and holding the bathroom door shut. Patrick’s screams brought Mark and me running to the rescue. Minutes later, the two boys sat in separate time-outs, Patrick sulky and angry; Tee rather smug at the apparent leveling of justice.

* * *

Dear Tee,

Here you are a six-foot grown man, but you’re still Tee to me. You always will be.

I just have to write that I’m sorry. I’m sorry for all the times you got dragged to ER because Patrick’s Mic-key button came out or he started vomiting. I’m sorry for the eight days I was in the hospital with Patrick and the nurses wouldn’t let you come visit. You needed your mom. I’m sorry that I burdened you with Patrick’s care. It wasn’t fair for me to teach you about giving Patrick cornstarch or how to operate the kangaroo pump. You never really got to be a
kid; you were like the third parent. I also think about the time Patrick got lost and we found him in the middle of the golf course. I think we said some hurtful things to you when we got back home, just smearing guilt on you.

I’m also sorry about the time you were all excited about getting a dog. Remember? We were going to name him Oscar. We had stopped first at McDonald’s and had fun in the Playscape outside. It was so strange that Patrick’s sweaty leg twisted when he went down the slide, cracking his fibula. I think the ER called it a greenstick fracture, and one doctor gave me the third degree about child abuse. Patrick ended up with a bright blue cast from his groin down to his toes, and everyone fussed over him incredibly for weeks. You just took it. You wanted a dog, but you got a trip to the hospital instead.

Dad and I did our best. We tried to get to all your track events, your plays, your concerts. But, looking back on all this, I know you sometimes got the shaft. It wasn’t fair that your needs sometimes got shoved aside because I had to run to the hospital with Patrick or pick up more Tolerex—or, or, or.

I look at you now—what a man you are! Business degree, MBA—all A’s. A consultant in purchasing and supply chain functions. Flying all over. Dad and I are so proud of what you’ve done, but more importantly, who you are. You are hard-working with a strong moral code. I just can’t believe that you call us every week. You come to visit and bring us our favorite wine or help re-stain the picnic table. You are the one who remembers about Valentine’s Day flowers. Dad and I often say, “We done good!”

Tee, we love you thiiiiiiiiiiiiiiiiiiiiiiiis much!

Shaw 110
What do healthcare experts write about sibling response to a chronically ill child in the family? Did the Shaw family experience what psychologists find in their research?

* * *

Helping Children and Adolescents with Chronic and Serious Medical Conditions indicates that a chronically ill child can both unintentionally harm and help fellow siblings. The textbook relates that in one study involving a child with diabetes, “it was found that siblings both benefitted (with high levels of behavioral competence) and were penalized (by anxiety and low self-concept.)”

According to Webb, healthy siblings of a chronically ill family member often learn adaptability. They also realize that they are an integral, functioning part of the family process. According to a 2006 study by Roberta Woodgate of children with cancer, healthy siblings often understood that the parent needs extra time with the sick child. However, the healthy sibling will at the same time feel left out and isolated. Additionally, the healthy child can often feel sadness about his brother or sister’s chronic illness. That sadness is often unrecognized by parents, which causes further stress in the healthy child. However, Webb indicates that “It would seem that children act to preserve families amidst a situation of being overwhelmed . . . The healthy sibling does his or her part by laying low.”

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8 Ibid., 32.

9 Ibid.
When I think of Webb’s words and my adult son Tee, I am again struck by how “casebook” Tee’s growth into adulthood has been.

Once Tee became about sixteen, he had displayed a growing sense of responsibility in life. His grades rose to A’s and B’s. He became organized: his backpack was an orderly cataloguing of books; his bed was made; his assignments completed in time. Tee began to garner attention that had inadvertently been pulled away from him. He became a superb swimmer, winning medal after medal. He excelled in the high hurdles in track—a sport that requires both speed and strength.

Tee’s pattern in college continued. His grades moved to all A’s; he became president of his fraternity and vice president of the honors fraternity for business majors. He got runner up for homecoming king, and all the while, a bevy of cute women flocked around him. I also remember Tee’s college graduation vividly. He proudly wore the honors gold cord down the sleeve of his graduation gown. Four award medals hung around his neck. Tee was a star.

What would experts say about Tee’s experiences being the big brother of a chronically ill child? Did he benefit, or was he penalized, as the textbooks indicate? Yes, and yes.

To this day, Tee pushes himself within the paradigms of success as defined by our family. Also, he is the epitome of the thoughtful, giving son. He never misses a birthday or a Christmas celebration. His gifts are considerate and unique. For instance, how many sons understand that a mother would love hand-blown wine glasses? On family vacations, Tee will go to market for fresh fish, making a fabulous dinner for the whole family. He’ll call his grandma regularly. The list goes on and on.
Yes, growing up as a shadow child has given Tee some benefits. His accomplishments are what Western society appreciates today. Quite often, individuals who are hard-working and people-savvy are those that do well financially in the U.S. Additionally, as Tee worked to get out of Patrick’s shadow, he discovered numerous personal skills—scholastic and athletic—that may have lain dormant otherwise. Tee also has a real appreciation for getting up each day in a life that is simply “normal.” As a thirty-something, he has been illness-free. He understands that many people—40% of the U.S. population—are not so lucky.\(^\text{10}\)

The negative aspects of Tee’s compensation are several. The young man acts according to what others want—not always what he wants. Additionally, he is still “in competition” with the world. Tee continually goes for a better job, better education, better cars, better women. Can you blame him? The squeaky wheel gets the grease.

Life was busy—really busy.

Mark ping-ponged back and forth from Paris in a company position that was continually growing. My days at United Telecom—which had morphed into Sprint—were full and exciting.

Sprint periodically would gobble up a smaller company in the telecommunications industry. When that would happen, my department would inherit the new benefit plans to run. Sometimes, depending on how a plan was constructed, we could assimilate it into one of our own. More often than not, however, the new plan could not be molded into an existing one. ERISA (the Employee Retirement Income Security Act) was exacting in its requirements; the Department of Labor was not to be trifled with. Federal law was federal law, and that was it. So, our department added benefit plans like pearls on a necklace.

Fortunately, the Drama Queen in my department transferred to Human Resources to a position that paid more. We were lucky. I hired Shu to come in and tackle the SEC reporting for our benefit plans. Shu had a work ethic like no other. She cranked out perfect numbers and schedules, fending off our auditors from Arthur Young singlehandedly. I’m not sure she ate or slept. Meanwhile, the rest of us polished and perfected a below-market stock purchase plan that became entirely automated. Programming on our ESOP and TRASOP was updated; we no longer had to worry about potential errors or off-program fixes. Our group purred like a happy kitten. Occasionally, I could sit back for a minute in the hubbub and enjoy our efforts that took care of 50,000 employees. I loved talking with our shareholders—benefits plans were not just a corporate function, but real help to real people. On a daily basis, I would talk with thirty or forty Sprint retirees, helping them with the purchase prices of their various stock plans and outlining
the tax consequences of that sale of stock. More often than not, the Sprint stock plus Social
Security benefits became that retiree’s plan to support himself.

The home front was a hubbub of a different nature. Patrick had graduated to
kindergarten after two years in preschool. “I was “helded” back,” he’d explain to everyone. And
indeed, he had been. Patrick’s brain wasn’t the culprit—it was his body. Despite cornstarch,
Citrucel, Tolerex, allopurinol for gout prevention, and quarterly blood work and ultrasounds,
Patrick lagged slightly in physical development. Somehow, he remained unfazed by all this. Play
dates and birthday parties sprang into his life like popcorn in a skillet. Patrick’s trips to the
school nurse for cornstarch were just part of the class routine. Also, his Mic-key button seemed
no big deal to classmates. “It’s kinda cool,” said one of Patrick’s classmates to me during a field
trip. “You could drink a bunch of red Koolade through your Mic-key and you’d never have to
brush your teeth. Hey, I wonder if you could shoot stomach goo out of your button?”

Tee seemed to like sixth grade all right. He’d bring a wad of homework home each night.
Amy would help tackle the math; I was vice president of book reports and social studies after
dinner. We’d work on pop-top rockets and balloon hovercrafts. We’d practice on spelling lists.
The words seemed to whiz through Tee’s brain with no impact.

Mrs. Mann, Tee’s teacher, was on her game. One day at work, I got a call.

“Stephen is struggling with his reading. Unfortunately, he is testing below the fifth grade
level. I can see that in class he’s having trouble keeping up.”

Mrs. Mann gave me the name of a nearby reading specialty clinic. I scheduled a day off
work to take Tee in for a battery of seventeen reading tests. Seventeen.
The resulting report indicated that Tee had dysgraphia as well as other reading issues I simply couldn’t pronounce. I arranged for Amy to pick Tee up from school each day at 12:45, drive him to Reading Solutions, and return him to class by 2:15 or so. Five days a week—back and forth. Seven months of re-training. Re-testing. Then--success. Tee finished 6th grade in June reading at the 8th grade level. I was thrilled; this was my child who seldom made waves. He had been suffering academically, and I hadn’t noticed it. My attention was focused, as always, on Patrick’s well-being.

All should have been good, but it wasn’t.

I dragged myself out of bed each workday morning at 5:15, slurping down two mugs of liquid ambition to shake off the morning coma. Often, when I’d get up at 3:00 to do Pat’s cornstarch, I’d toss for an hour, ruminating about a new tax issue or a disgruntled shareholder. Five fifteen would come screaming at me just an hour later. Then, there were the evenings:

- 6:00 p.m.: race in the door and debrief with Amy
- 6:05: hug-and-catch-up time with Pat and Tee
- 6:30: kick off shoes and cook dinner
- 7:00: dinner with the family
- 7:20: short TV time for the boys. Do up dishes and pull out frozen meat for the next night. Throw in one load of laundry. Check backpacks for forms to fill out and sign. Dump out gamey gym clothes. Wash and fill up cornstarch containers.
- 7:45: homework time with the boys.
- 8:00: shower time for Patrick; mix up Tolerex; story time; start the Kangaroo pump.
- 8:20: Patrick in bed. Begin Tee’s shower.
- 8:30: Tee’s story time and hugs.
- 9:00: Laundry in dryer.
- 9:15 Prepare boys’ backpacks and snacks for the next day.
- 9:30: Answer work-related email.
- 10:30: Bedtime.
- 3:00 a.m.: Middle-of-the-night Kangaroo pump check.
- 5:15 a.m.: Back at it.

Over and over and over.
One night, I stared at the whirring goo of Tolerex in the blender, shaking out a new IV bag in readiness for the smelly mess.

I was so tired. So tired. I just wanted to find the nearest bed, pull up the covers, and not get up for two days. What was worse, I realized that I was getting to see Patrick only two hours on a workday, and Tee only three. It didn’t feel right. Not right at all.

Had my mother gone through this angst with me when she went back to teaching? She was lucky; she had gotten home at 4:00 each day. And there had been only one of me. And I had been healthy. And my dad didn’t travel—he was home by 5:00 each day. But mom had had a good teaching career; first high school and then as department head of a small Presbyterian college in Emporia. Wasn’t that what a woman did? A woman became well-educated and then cruised through a succession of good careers. A woman could have a great husband, children, and a demanding job, right? Plus, make cherry pies? I thought of my favorite song when I was a kid. Blasting at full volume on our stereo in Emporia, Peggy Lee would belt out:

I can wash out forty-four pairs of socks and have 'em hangin' out on the line
I can starch and iron two dozen shirts 'fore you can count from one to nine
I can scoop up a great big dipper full of lard from the drippin's can
Throw it in the skillet, go out and do my shopping, be back before it melts in the pan... ¹

If my mother and Peggy Lee could do all this, I was evidently lacking as a woman. Would I have to make a choice between career and motherhood?

I didn’t want to make a choice. I loved both.

* * *

Pow-wow number five. Mark had gotten home from Paris Friday night. It was a peaceful Saturday afternoon, the leaves curling and flipping across our back deck. Tee and Pat were next door at Scotty’s home, playing. The atmosphere in our home had simmered with an undercurrent of constant tension for months. Mark’s work, my work, the kids. And the GSD. The GSD. That constant, unwelcome party.

“I can’t do this much longer, Mark.” I made a feeble attempt to scoop up a pile of newspapers. The house was a wreck of toys, tippy cups, and soccer balls.

For the first time in our marriage, Mark and I began to consider a single-income future. We looked at Mark’s job level—considerably higher than mine. I was fortunate to be a manager at Sprint, but the harsh reality of the 1990’s was that only two women directors existed at corporate headquarters and only one female vice president. It didn’t look good for me. I felt like a kid in a candy store at work, with my nose pressed against the glass ceiling that still kept women in place. It made sense to have Mark continue to work, because his earnings potential was simply better. I still love Mark for the fact that he never assumed that I should be the one to stay at home. However, we both knew finance, and the numbers didn’t lie. I didn’t want to quit work—I loved the exhilaration of problem-solving and helping shareholders. But I was tired—so tired. Also, was I being a mediocre mother?

That Saturday afternoon, Mark and I grabbed notepads and scratched out ideas. Costs for me to work outside the home: Amy, gasoline, dry cleaning, suits, costly made-to-order food, bi-weekly house-cleaning help. Wear-and-tear on the car; wear-and-tear on the family. What could we ditch in our lives? Keep the old station wagon forever. Scrap vacations. Eat more
hamburger. Embrace Pizza Hut and ditch Crowder’s Steak House. I could be the house-cleaner and the errand-runner. We could do this. We had to do this. However, I felt pangs as Mark and I discussed the options. Was this the role I was meant to play? Would I turn into a bathrobe-wearing, Oprah-watching slob? Would my mind melt into mush if I devoured Nora Roberts’ heaving bosom novels? This was a new paradigm for me, and not a comfortable one. I was also afraid of what other career women would say to me. More importantly, I was truly afraid of what I would say to me. Hmm, Janet. Not living up to your mother’s image? Living life in a mediocre fashion? Letting your brain rot?

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I’ve been gone from my work now for twenty years, and I still miss it. I loved working with company programmers and lawyers to improve the benefit plans. I loved digging into sheets of data and figuring out patterns and flows. I loved helping employees in our department grow, get training, and get promoted. I loved helping the little old lady shareholders with their stock questions as they were trying to figure out their retirement. I loved getting up in front of shareholder meetings to discuss the next stock offering. I loved chatting with Bill Esrey, the CEO, as I huffed and puffed on the stair-stepper at the company fitness center. Work simply gave me definition and challenge.

Oddly enough, within two weeks, I found that being a stay at home mom also gave me definition and challenge. Had that 1970’s “I am woman: hear me roar” just been bunk? Had I been sold a bill of goods by popular culture?

I think so. When a woman has a demanding career, a marriage, one “normal” kid, and one with chronic issues, she can’t do it all. Something will give. Typically, what gives is time.
spent with the spouse or the children. What also gives is the woman’s sanity. A career is important, but so are spouses and children and mental health. When I take my last breath, I won’t dream about Sprint’s benefit plans. I’ll think of Mark, Tee, and Patrick—my three loves. I’ll think of our bike rides in Shawnee Park or swim meets with Catfish Charley.

I had to choose between an “official” career and family, and I feel to this day that I chose wisely given our circumstances. What’s more important—I was so fortunate to even be able to make the choice in the first place. That’s a blessing that I do not take for granted. Ever.
PART III

WALKING FORWARD
I did not turn into a bathrobe-wearing, Oprah-watching slob. I threw myself into number-crunching for the PTA. I coordinated the summer swim team at Lake Quivira. I worked with the kids at church to create Christmas pageants, herding thirty angels and shepherds onstage. I scrubbed toilets at home and simmered Bolognese sauce. I made one hundred scones for Tee’s history unit on Victorian England. Every day was different, and every day was tied to my family. It was amazing. My brain still got challenged and I got to flex my organizational skills. Who started the myth that the stay-at-home mom wears a cute apron and sips martinis? She puts in sixteen-hour days, but they can be very good days.

Most importantly, I took a deep breath for the first time in five years and watched Tee at soccer practice. I cheered Patrick as he played T-ball. He seldom made it to first base, but no matter. I sipped a good Chardonnay with Mark at the end of a busy day. It was great. Absolutely great. For the first time in years, our family felt the vibration of anxiety quietly slow down. And the old blue station wagon? It just got older. Didn’t matter.

* * *

In the 60’s, Bob Dylan used to wail, “The times, they are a changin’.” They certainly were for the Shaw family.

I’d been a stay-at-home mom for about six months. I still choked on the term, bristling if I got an eyebrow raise from a snooty female attorney at a neighborhood party. Maybe I was a family coordinator/love provider/dog walker/toilet scrubber? No label seemed quite adequate.
Mark came home one night looking drawn in the face. During dinner, we witnessed a rousing discussion between the boys: “Boston is too in Texas. Boston baked beans—like what the cowboys eat over a campfire? That’s got to be in Texas.” Then, we supervised showers and story time. I loved this part of the day, because I could read stories to the boys without the nagging sense that I needed to plow through twenty emails afterward. I could simply listen to their ideas. We had some doozie concepts come out, too, as I learned to take time with my children. One night, I walked by Tee’s room to hear, “Why do you have a beard under your arms, dad? And how does Mrs. Parker know every time I’ve passed a note in class? I did not draw her with a gigandous wart!” However, Patrick was truly the question man. One night, Pat blurted out during story time, “Wait! Do you think we could connect the dots to Tee’s freckles and draw a picture? If God doesn’t need to take a bath, do you think he takes showers instead? Do snakes have ears?”

These questions always left Mark and me slightly breathless.

That night, Mark and I sat down for our usual chat with some Three Buck Chuck, I imagine. Taking a sip of the wine, Mark’s eyes twitched slightly at the slap of tannin.

“Well, stuff is going on at work,” Mark finally got out.

“Stuff?” I was instantly alarmed. “Stuff going on” was never good.

“The company is moving its headquarters to New Jersey, and the president wants me to find new headquarters and run day-to-day administration. I’d be in charge of the phone operators, flight operations, security, the cafeteria—all the things that employees get angry over. It would be a promotion for me. I’ve prayed over this opportunity but I
want to see how you feel. If you’re not on board, we won’t do it.” Mark handed me a glossy brochure on the wonderful state of New Jersey.

New Jersey! Oh, dear God. I walked to our bookcase and pulled out a big atlas. New Jersey . . . New Jersey . . . it’s somewhere on the east coast. Here it is. What??! That tiny state? Does it even have roads? Looks like it could drop off into the Atlantic. Hmmmm . . . it’s really close to New York City. All that crime. Hypodermic needles on the streets. Oh, but the Metropolitan museum of art. Broadway. Central Park. Ellis Island . . .

.. My mind flipped between excitement and dread.

Folks from Kansas are fairly certain that nothing exists other than Kansas. Why would you want to move and leave miles of rolling wheat and the smell of cattle dung? Besides, we are pretty sure that once you get to the Kansas border, the world is simply flat and you just drop into an abyss. Heaven knows what is in New Jersey.

“Let’s think on this and I’ll do some praying for guidance, too,” I replied to Mark. My world paradigm was cracking.

Mark and I have always been firm believers in divine nudges. Our spirituality has led us in certain directions and those directions have been good. So, after about a week of listening—just listening to a God that no longer smote me but had walked with me through the rough GSD journey—I knew. I felt it. New Jersey would be okay.

“How are we are going to tell the kids about this?” As usual, Mark was on point. By this time, Pat had moved to first grade and Tee was in seventh. It would be a tough time to move the kids from Kansas to New Jersey.
Mark came up with the idea of reading our kids a story about change. The prior week in a staff meeting, one of the managers had mentioned a book by Dr. Spencer Johnson called *Who Moved my Cheese?* Apparently, since quite a few families would be impacted by the upcoming corporate move, this book was supposed to help break the news to kids. The book was a parable of sorts, about two mice—Sniff and Scurry—and two little people—Hem and Haw—who lived in a “maze” and looked for “cheese” to nourish themselves. The “cheese” represented what these characters wanted in life, and the “maze” was where they would hunt for what they wanted. However, only one of the mice handled the process successfully, with minimal stress. The victor wrote his “a-ha’s” on the maze wall.

A week later, Mark brought home *Who Moved my Cheese?* After dinner, he gathered us in the family room.

“No homework tonight!” he announced. “I’ve got a special story to read to you all.” Mark proceeded with the drama of the cheese. He was good reader: I could almost see Hem and Haw blundering through the maze. The boys were riveted; even Georgie sat nearby, watching the family.

Mark finished the book, preparing to tell Tee and Patrick about our move. “Well, guys, here’s what’s going to—.”

Patrick blurted out in the middle of Mark’s grand performance. “So, are we moving???”

How did Patrick know?
New Jersey turned out to be the coolest thing since sliced bread.

We found a house on the western side of the state, although I hesitate to use “west” and “east” when referring to New Jersey. With one good sneeze, you can head out of the Newark airport onto I-78 and be halfway to Pennsylvania. At any rate, the western side of the state had lower property taxes. Coming from Kansas, we wallowed in sticker shock.

Our neighborhood consisted of twenty-eight homes surrounded by sod farms and cattle. Technically, we lived in Pittstown, named after William Pitt the Elder, an English Whig statesman of the 1700’s. I snickered the first five years in New Jersey when I said the word “Pittstown.” The town made my hometown of Emporia, Kansas look like a burgeoning metropolis. Pittstown had a bank, a deli, a B&B, and an old barn with a faded sign that said, “You R Here. So This Is Pittstown.” I could head north five minutes to the big town of Clinton, housing about ten thousand residents.

Clinton absolutely radiated cuteness. I would come into town for errands, driving slowly over an old metal bridge straight from nostalgic Christmas cards. I’d cruise slowly past a red gristmill from the 1700’s. A slow-moving river meandered on either side of the downtown, complete with lazy kayakers and circling geese. It was like Norman Rockwell had gone absolutely crazy, painting broad brushstrokes of charm. The two blocks of downtown displayed shuttered brick storefronts and planters bursting full of ivy and pansies. While the typical small Kansas town sported a fly-specked Dairy Queen
and a hardware store reeking of fertilizer, downtown Clinton had gorgeous shops filled with leather goods, Brighton shoes, and gourmet pistachio ice cream.

We landed in Pittstown just a few days before Valentine’s Day 2000. Mark and I had planned on relocating in the middle of the school year so Tee and Patrick would make friends by summer. The kids’ school was called Union Township—incorporating kids from kindergarten to eighth grade. I loved the place. The school was a throwback to the 1960’s; a red brick single story affair with playgrounds and soccer nets. The inside of the school smelled vaguely of chalk, beanie weenies, and dirty sneakers. Everyone knew everyone in the school, and the lucky teachers only had about twenty children per classroom.

On day one at the new school, Tee and Patrick walked two doors down from our home and caught the 8:15 bus along with the rest of the neighborhood gaggle. They hopped with excitement, because they had never ridden a school bus except for occasional field trips. The boys’ backpacks were filled with notebooks, approved snacks, and Valentine cards for each classmate. Not bad. The first day of school for the kids, and boom. A Valentine’s party.

I called Mrs. Mintel at the close of day one, trying to contain my helicopter-ness. Not surprisingly, Patrick appeared at ease at the new school. He would walk down the hall to Mrs. Mason’s office twice a day for his cornstarch.

I had met with Mrs. Mason, the school nurse, just a few days prior to discuss Patrick’s cornstarch schedule and to bring in boxes of Argo starch and spare syringes. I provided her an ER instruction list, contact list, and snack/cornstarch schedule list. Mrs.
Mintel had the instructions, too, as well as the PE teacher. Awareness and education were crucial for Patrick’s safety at school.

Mrs. Mason hailed originally from “Longah Islant,” where she loved to go “down sho-ahh” to Cape May and drink “caughwwwfee.” She was an island of motherliness in the middle of the school day. Kids came to her for headaches, scraped knees, test anxiety, and, well, cornstarch.

Tee, our stoic son, was not well adjusted at Union Township, I found out months later. He was a new seventh grader in a sea of kids who had known each other for six years. He was the newbie—the outsider from Kansas, wherever that state was. And Tee, not wanting to trouble mom and dad, just dealt with the loneliness. The Shadow Child was, as usual, quiet. And I didn’t notice. Again.

* * *

The best part of Clinton and Pittstown was that Dr. Charles Stanley was only an hour and a half away at the Children’s Hospital of Philadelphia. Quite accidentally, back in Kansas City I had run across an article about Dr. Stanley. He was considered a GSD guru, and boy, did we need a guru. Dr. Marris in Kansas City was technically expert, but he never explained anything to me. It was a “getting blood from a turnip” scenario. Within ten minutes of reading about Dr. Stanley and the Philly Children’s’ Hospital, I called long distance and had made an appointment for late February.

We had only just moved to Pittstown when Patrick and I made the big drive to Philadelphia. Neighbors had instructed me to take state routes 513 and 527, cutting diagonally across rolling Jersey sod farms to I-95 south. “This is fine!” I said to Patrick.
“We can do this!” Actually, I had never driven in a city larger than Kansas City. Patrick busily turned the dials of the radio, trying to find the Red Hot Chili Peppers. As we got fifteen miles outside of Philly, traffic on I-95 began to crawl, snakelike. The highway was a series of ruts, blacktop patches, and orange barrels.

Yes! There’s exit 22 to downtown Philly. With my blinker on, I slowly edged to the right. A line of cars blocked the lane I wanted, like a crawling wall. “Patrick! Give em’ your cutest smile!” I waved to the drivers to my right, batting my eyelashes. We needed to get over a lane.

Nothing. I was invisible. My exit was 200 feet away. Then 150. Then 100. Okay, here it goes, I thought grimly. Determined, I put my blinker on again, grabbed the steering wheel, and did a “Jersey Slide.” Somehow, traffic parted like the Red Sea, as the angry Kansas mom plowed across two lanes to exit 22.

“Hey lady! Learn to drive!” one cabby yelled at me, waving excitedly from his car window. I may have said, “Patrick, my goodness: look over there at the vintage red Corvette.” I then flipped the cabby off with enthusiasm. Oh, my mother would not have been proud.

The Philly Children’s Hospital is both a mecca of hope and despair.

The facility is massive, with parking garages that wind up fourteen floors to a nosebleed level. Shiny bright hallways take you to this elevator, and then down this corridor, and across this walkway, and down this escalator, then over to this waiting room, and into this door. Large silver mobiles slowly twist from the ceiling. Rows of red-framed pictures painted by children line the halls.
Amidst this exterior cheerfulness Pat and I saw children with issues. All kinds. Some kids were in wheelchairs with feeding tubes. Some had no legs. Some sat by the bells and slides of the activity walls, not knowing what to do with the toys. Some just lay, inert, in their parent’s arms.

Pat didn’t say anything as we trekked to Endocrinology that day, but he took it all in, his blue eyes wide and a little watery. Outside of a string pack filled with cornstarch and syringes, you couldn’t tell there was anything wrong with him. I think, for the first time, Patrick felt really blessed. Yes, GSD was bad; it could kill you. But, he could walk. He could think. He could go to school.

Dr. Stanley and his PA Linda Steinhaus were an aura of light in the hospital. We met for an hour, where Patrick was gently prodded and prodded; weighed and charted. But most importantly, Dr. Stanley and Linda talked to Patrick like the bright young man that he was. He wasn’t GSD Specimen Number 24, but the official class clown of the Union Township first grade. He was a drum player and a skateboarder. He was a kid of a thousand questions.

Patrick and I went from Dr. Stanley to the blood work lab to ultrasound. “We want Patrick to have quarterly lab work as well as the quarterly ultrasound. We’ll see you back here in three hours to evaluate our results.” The crinkly blue-eyed endocrinologist smiled at us and shook Patrick’s hand. “Young man, we’ll see you in a bit, ok?”
The phlebotomist captured five vials of Patrick’s blood. I was amazed; Patrick had had blood drawn so often he could just nonchalantly observe the draw, while I would hold his other hand and hum *Yankee Doodle*, looking across the room to keep calm.

After an extensive ultrasound of the liver, kidneys, gallbladder, and bladder, we headed for a forty-minute play session with a child psychologist. Pat would be followed quarterly with Mrs. Markham.

It’s no lie that kids with chronic illnesses or disabilities are at risk for psychological stress, as are their siblings and parents. Sharon Dempsey in *Extreme Parenting* indicates that “Play therapy is recognized as being beneficial in helping children to explore issues affecting them . . . The fear of hospital procedures and the uncertainty of the future create anxieties that can be explored through play therapy . . . through play, the child can communicate unconscious and conscious experiences and emotions.”¹ *The Journal of Pediatric Psychology* adds further: “The presence of GSD types 1a and 1b are associated with reduced quality of life and independent functioning, and elevated levels of internalizing distress and parental stress relative to healthy peers.”² Fortunately, the Children’ Hospital of Philadelphia was no stranger to children with unique and devastating conditions. While the Shaw family played together, ate

¹ Dempsey, p. 111.

together, and hugged together, we didn’t know if we were managing the collective
stress of illness appropriately. Perhaps Mrs. Markham could help.

So, I sat outside the therapy room, twisting my shoelaces. Patrick seemed to be
doing okay, stress-wise, as far as I could tell. Each day, he’d burst in the door from
school with Tee, talking non-stop as I fixed the boys PB&J or popcorn.

“It’s not fair! I was five feet away from Kyle when he got in trouble for making
fart noises. The teacher gave me a time out, too. I was there, but not involved. And, I did
not call Mrs. Decker a lard ass. Oh, here’s my multiplication test. I got an A. Here’s my
spelling test. I missed one.”

“Patrick, what are these five red tickets here in your backpack? What do they
mean?”

“I dunno. They’re from my teacher. Oh, Mom, Dan invited us over to roller blade
after school. Can we go?”

“Sure . . . for just a bit. Both of you—grab your kneepads and helmets. Pat, what
do you need to do when you get home from school, first?”

“Cornstarch?”

“You bet. And at what time?”

“Uhhhh. 4:00?”

“You’ve got it, Pat. And here . . . help me rinse out today’s starch containers and
re-fill two of them. That’s it. Wipe them really dry and place them rim down on this
clean towel. I know you’ll only be three doors away, but what do you need to do,
always?”
“Fill my string pack with my cornstarch containers and bottled water? And wear it?”

“You betcha. And your next starch time is 7:00. Hugs and kisses, guys. Tee, you and Patrick need to be back at 4:45 to start homework, okay? Both of you: set your watch timers.”

I was a believer in hands-on training with Patrick as well as daily repetition of his cornstarch schedule. I wanted Patrick to incorporate his healthcare into his very being. It would have to be second nature, because no GSD cure was in sight. Careful management was our only hope. I remembered only too well my own father. He had neglected his Type 2 diabetes, suffering from poor vision, a heart attack, and two failed kidneys.

Both boys tromped out the door, ready to roller blade up and down Dan’s driveway, taking miniature leaps off a cool one-foot ramp. I made a quick call to Patrick’s teacher to learn about the proliferation of red cards in the backpack. Apparently, my young man was quite social, and the teacher’s presence at the front of the classroom was merely a suggestion that Pat should listen. Mrs. Mintel mentioned something else about paper airplanes, too.

Thinking back to the Philly Children’s hospital, I felt that Patrick’s play therapy, class clown role, plentiful social invites, and good grades must have meant that all was good in the ole’ noggin. To be sure, I had seen Patrick look worried at the hospital when Dr. Stanley discussed starting him on allopurinol. Apparently, kids with GSD were at risk for gout. We were also going to get an appointment with a nephrologist at Children’s Hospital to monitor Patrick’s kidneys. Apparently, kids with GSD were also at risk for
kidney complications. In fact, the blood lab gave us a huge neon orange collection container for a specimen collection. Dr. Stanly patiently told Patrick that the urine collection would measure how much creatinine and protein were processed through his kidneys. Then, there was also the discussion with Dr. Stanley about the ultrasound today that had revealed four adenomas on Patrick’s liver.

“They’re benign little growths on the liver, Patrick. Now, yours are about half an inch long, so they’re tiny. They are called inflammatory adenomas, but they usually don’t hurt anything. We will monitor them every three months here at the hospital with an ultrasound. If you measure your cornstarch very carefully, the little growths should just stay like they are. You see, keeping your blood sugar normal with cornstarch also keeps your lactic acid normal, which then keeps your triglycerides normal. Oh—triglycerides are a mixture of fatty acids. Sometimes, high triglycerides over time can make the adenomas grow. If you keep everything controlled, your adenomas can stay the same, shrink, or disappear. Don’t worry, Patrick—we will take special care of you.”

Patrick and I sat there, a neon urine collection bottle and pages of data in hand. We both blinked, a bit stunned. The blood sugar to lactic acid to triglyceride connection was beyond me. I wished desperately that I had taken more than just Rocks for Jocks in college. Igneous rocks were of little value, now.

That night, it happened in the middle of *Harry Potter and the Sorcerer’s Stone.*

“Why did God give me GSD?” Patrick looked clear into my soul and waited.

The question seemingly came out of nowhere. I was dumbstruck for seconds, but then remembered the flood of scary information that had hit us earlier that day at the
hospital. God love Dr. Stanley. He was knowledgeable, kind, and honest. But, tough facts were tough facts. GSD was GSD.

I had read in one of my parenting books that I had to be honest with my kids, but not overly dramatic. Patrick was smart, really smart, but he depended on the family for his sense of safety and his reality.

“Well, um.” My mind raced. How do you communicate tough medical information and theology? Know your child, my gut told me. Feed your child, my gut had told me years ago, before Patrick had been diagnosed with GSD. I resolved to rely on my inner sense.

“Well, um” I repeated, stalling a bit. “Remember how we talked about how GSD happens? Dad has a defective gene and so do I. When you were created, there was a one in four chance that you could get GSD. But, Dad and I had never heard of the condition. We didn’t know we were going to give you anything bad. We love you and Tee more than anything, you know.”

I took a deep breath while my mind zigzagged. “So, I don’t think God “gave” you GSD. It just happened. I think maybe because of GSD you will always be sensitive to other people and their health issues. You’ve been to the Philly Children’s hospital and seen all the kids with no arms or who can’t walk, right? But here’s the good news, Patrick. You were given a great brain. Maybe you have been created to do something wonderful. Something that requires compassion for others and a good mind. Do you think that might be true?”

Patrick reflected a moment. “I guess so. But it doesn’t seem fair.”

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“No, it’s really not fair, for sure. But, Dad and Tee and I are here for you, always. Always and forever. That’s one thing you can count on.” I gave Patrick a mama bear hug.

Later that night, I was awakened by a stage whisper. “Mom!”

It was Patrick, standing by my bed with his comforter and dee dee. “Can I sleep down here next to you?”

“Sure.” I made a comfy pallet on the floor right next to my bed, where I could reach out and touch Pat’s cheek. Patrick hunkered down, sighing and rolling over into a child’s oblivion. Within minutes, I heard “Mom!” again. Tee wandered over to our bed, his covers in hand. Tee curled up next to Patrick. I touched the reddish curls. Georgie padded in moments later, curling up by the boys. The Shaw family slept, in peace.
We had been in Pittstown for about five years when the boys’ social schedules exploded. Football practice for Tee. Pizza parties. The sixth-grade spring dance with real live girls. Patrick’s drum lessons and roller blading escapades.

“Karthik invited me over for a sleep-over!” Pat exclaimed one April day as he and Steve burst in the back door from school. By this time, Patrick was a cocky eight-year-old.

I dried my hands on old checkered towel: I had been elbow-deep in making meatloaf. Leaning on the kitchen island, I placed snacks out for the boys as my stomach did a one-eighty. I had known this question would come up soon with Mr. Social.

“Well, let me talk to dad tonight,” I replied, stalling.

Karthik Dwarki lived cattycorner from our house, just a hop and a skip out our back door, with his father Lakshan, mother Amrita, and little sister Anupama. Dr. Dwarki was a veterinarian with an MBA. Karthik, at age twelve, was bright—the kind of son everyone wanted to have. And there was Anupama, a round-faced five-year-old who would wander into my kitchen when Karthik was over. She loved to stand by the pantry door and widen her dark eyes. “I like Oreos!” she’d inform me, tilting her head and waiting.

“I promise we’ll be in bed by ten.” I came out of my reverie when Patrick started in on his arguments for the sleep over. “And the Dwarky family is really healthy! They’re vegetarian.”

“So, no hot dogs over there?” I countered.
Patrick took another breath and went at it. “And you don’t want me to be socially awkward. And, everyone is doing sleepovers. Tee gets to. And Tee gets all the pizza he wants.”

The guilt began rolling in. Tee was healthy and Pat was compromised. Tee did get all the pizza he wanted at parties. With Patrick, I’d have him take two bites, then switch to carrots or cheddar goldfish. Walk away from the pizza, I’d coach. Just say no. I sounded like Nancy Reagan in the midst of a drug campaign.

The next day, I called Amrita and told her Patrick had been invited to a sleep over that weekend at her house. Incredibly, Amrita knew about the proposed social event. Within ten minutes, I wandered down the back yard and into the Dwarki home. Amrita and I reviewed Patrick’s cornstarch schedule and his GSD symptoms. I had her phone number and she had mine. Amrita had been a nurse before her children, and then there was Lakshan’s veterinary background. Patrick’s first sleep over would be in a medically informed household. Patrick didn’t have fleas, but still . . . a veterinarian’s home . . .

The night of the big sleepover, I stood in the backyard and watched Patrick trot to Karthik’s house, the string pack bouncing on his shoulders. Patrick had his cornstarch schedule, cornstarch, glucose tablets, bottled water, snacks, timer watch, and walkie talkie. I had the other walkie talkie: Shaw communication central was operative. Was I being obsessive? Yes, definitely.

“Remember, now.” I had kissed Patrick on his curly head as we packed up. “Cornstarch next at 8:00 p.m.; 11:00 and 3:00 are your bigger doses so you can sleep
longer. 7:00 a.m. is a regular one. See the containers marked ‘nighttime’? What do you do after the nighttime doses?”

“Call you on the walkie talkie.” Patrick impatiently glanced out our sliding glass doors, itching to see Karthik.

“Yes, please. If I don’t hear from you, I’ll traipse across the back yard in my PJ’s and curlers and knock on the basement door. You’ll be embarrassed to death.”

“Okay, mom.” Patrick trotted to Karthik’s as I heaved an uncertain breath. Would Patrick get busy with video games and forget his cornstarch? Would he go berserk over cheese pizza? My baby had just walked out of my little control zone. Yet, it was time for the first sleep over to happen; it had to happen. This night would represent Patrick’s first baby step to middle school, high school—maybe even college. I dared to dream a bit.

“The sleep over was the time of my life!” Pat burst in the next morning fairly early. I had been pacing the kitchen with a cup of liquid motivation but nonchalantly peeled a banana when Pat flew in. “We played Grand Theft Auto III and watched Shrek. It was awesome.” What was awesome was that Patrick had done his cornstarch on time and had called me after each dose. A fun, uneventful evening was perfect for any kid. It was critical for a GSD kid.

I got half a hug in before Patrick started begging.

“Next week can I go to Michael’s house?” Michael was across the street and down two doors.

“I think we can arrange that, after I meet with his mom Michelle.”
“And Josh wants me to come over, too.”

“Well, let’s do a few more sleep overs in this neighborhood, first,” I countered. Josh lived five miles away. “You have to be careful on the sleep overs, right? I need to feel comfortable with this. And you—you must become an expert in GSD.”

“I’ll be so responsible, Mom.” And so, the sleepovers continued, at our house and all around the neighborhood. Within months, they happened all around the school district.

* * *

Patrick was about thirteen when he got the flu. He vomited once and felt warm to my touch. I began giving him glucose gel immediately, but it flew back up within minutes. After the second vomit, I grabbed Patrick’s string pack and began loading up supplies: our ER instruction letter from Dr. Stanley, cornstarch, water, syringes, glucose gel, and healthy snacks. We roared fifteen minutes over to the Hunterdon Medical Center, the nearest hospital.

I had always liked the hospital. It was new and friendly, with polished floors and pink-smocked volunteers. However, it was not a place where you’d go for heart surgery or GSD.

Patrick and I screeched into ER about 7:00 p.m. that evening. Mark stayed home with Tee, heating up beanie weenies and supervising homework. I imagine that our new pup Sonnie curled up on the kitchen floor nearby, aware that life was not right.

The triage nurse checked me in. Slowly. So very slowly.
I gave her insurance information. I showed her the ER letter. I filled out symptom information. I dug my nails into my palms. The nurse painstakingly took Patrick’s temperature, blood pressure, weight, blood oxygen level, and glucose. The minutes crawled, as Patrick’s eyes began to glass over and his head drooped.

“His glucose is at 72,” the nurse announced to me with a bit of contempt, as if to say, *Lady, this place is for emergencies.*

“Yes—I know. I’ve been giving Patrick glucose packets for the last hour. But when he has the flu, his lactic acid will elevate and the rest of the blood profile will start to collapse. See here? On this letter from Dr. Stanley? Patrick has glycogen storage disease. We need to act fast: he could go into shock.” I tapped the ER letter for emphasis as a tear dripped onto the form.

The nurse seemed unimpressed as she called for a gurney for Patrick. Clearly, I was not a doctor and therefore uninformed. Clearly, Patrick was only thirteen and therefore uninformed. Clearly, Patrick was not bleeding from stab wounds and was therefore not an emergency. However, I could see my son sinking into shock, his eyes dull and hollow. **I knew my son and I knew GSD.**

Two minutes. Four minutes. How long could it take to get a gurney? Was I in an emergency room or in a darn parking lot? I looked around the ER waiting room. Two other patients were restlessly flipping through *Golf Digest* and *House Beautiful.*

It took a full hour and a half before Patrick and I were admitted into the bowels of ER and into a treatment room. Just in time, a nurse started an IV bolus of glucose and Anzemet into the crook of Patrick’s right arm. Patrick’s face had become pasty white—
his eyes closed and skin clammy. Before we finally got an IV delivered to the room, I had been trotting up and down the hall, waving my emergency forms in general panic. I flagged down every nurse, doctor, and custodian I could find. “Someone’s coming in to help soon,” they all soothed me. After the bolus of dextrose began to provide life-giving fluid to Pat, a nurse then set the kangaroo pump clicking and whirring. Within thirty seconds of the bolus, Pat perked up, like a computer re-booting. Two blue eyes fluttered at me and opened in confusion. I rubbed my teen’s forearm gently and gave him tiny sips of water and cheddar goldfish. “Let’s try a little cornstarch in thirty minutes and see if you can hold it down.” My baby.

“Patrick, we are so lucky that you got the IV in time, because it saved your life. If you’re off in college and this happens again in an ER room, grab your ER form and pretend to have a seizure if you get ignored. I might not always be with you, and even medical staff don’t always realize how quickly things get dangerous for someone with GSD.”

“I got it, Mom. I got it.” Of course, Patrick did.
25 More Steps

I always say, if you want an interesting life—raise a couple of boys.

Patrick was best buds with Josh at Union Township. Josh was a bit burly, athletic, and smart to boot.

Patrick burst in one day from school with his usual fan of red tickets from the teacher, notes from girls, complaints, stories, and requests. “Hey, mom,” he managed to get out between bites of his snack. “Josh asked me to go to a thing with him Sunday afternoon at his church.”

Evidently “a thing” was something good because Patrick seemed so excited.

Sunday came and I busily packed up Patrick’s pile of supplies into his string pack.

For that special Youth Evening, the church youth group leaders had rented a bouncy house and set up an obstacle course. A cotton candy machine spit out pink fluffy magic in the church side yard and popcorn scented the air. Over in another corner, church dads grilled hotdogs for the kids.

After a frenzy of bouncy house jumping and hot dogs, thirty sweaty kids tromped into the church. A young man stood at the altar, his curly brown hair just grazing his collared white shirt. His eyes were heavy-lashed, earnest. He looked like a twenty-first century Jesus, if Jesus had insisted on shopping at Polo.

“Boys and girls,” he began, spreading his palms out to the gathering. “I know you think church is just for old guys and that Jesus is for losers. But, do you want to be a winner? DO YOU WANT TO BE A WINNER FOR JESUS?” The young man brushed back his wavy brown hair and narrated his story. He had been with a biking crowd, spending his
days drinking and drugging. Until one day—he had rested on a rock near a wheat field by Wellington, Kansas. There—he had seen Jesus and had changed his life.

Electricity and spirituality sizzled through the air. “Yeah, yeah!” the kids echoed back. Patrick settled his sweaty back into the pew, filled a cornstarch container with water, gulped the chalkiness down, and began thinking about his next project: building a hovercraft.

The hovercraft had been an obsession for months. Patrick and I would wander through Home Depot for two hours at a time, while the young inventor selected this bolt and that cabling and those sheets of tin. Using our leaf blower as a propulsion device, Patrick was positive he could build a hovercraft and sail over to Karthik’s house, magic carpet-like. He could take our new pup’s droppings and zoom over to the science teacher’s yard, scattering them like confetti. The possibilities were endless and exciting.

“You all stand up!” the young Jesus-man beckoned. “I say, stand and come on down!”

Patrick jolted back into reality, his hovercraft plans disappearing into the mist of his brain. All he had heard was “Stand up,” so Patrick rose uncertainly. He felt something was amiss when he heard people begin to clap all around him. Looking around, he realized that no one else was standing. No one.

“Let’s hear it for this young man, here. Come on down! Son, can you tell all these fine folks here that you’ve given your heart to Jesus? Everyone, let’s hear a big AMEN!”

Patrick blinked, ending up stunned and alone at the front of the church, as the congregation applauded. The next thing he knew, he was signed up with a church
sponsor. Fifteen church members lay their hands on him and prayed for his sins, especially for all his red tickets from the teacher. In the midst of hovercraft planning, Patrick had been saved.

* * *

It was about a week later that our family got an exciting pamphlet in the mail. Emblazoned across the front, it said, “Enroll NOW for the Institute for the Gifted summer camp!” Ooooh. Summer classes at Bryn Mawr, Pennsylvania. Ivy-covered stone walls. Classrooms reeking of academia. Hands-on science. I wanted to go to camp, too.

That evening, I slid the glossy pamphlet over to Patrick at the dinner table. Tee was going to work that summer at ShopRite in the produce department, to help pay for Eleanor, his four-wheeled pride and joy. Patrick was too young to work, of course, but I had a household rule: no more than two hours per day of video games. Each boy had to work, play outside, or take classes.

“See, Patrick? You’ll share a dorm room with a friend. You’ll have classes and recreational time each day, plus art projects, improvisation, and campfires . . . .”

Patrick interrupted my spiel, meatballs skewered on his fork. I absent-mindedly took my own fork and gently poked his elbows off the table. Fancy-pants camp or no, manners were manners in our house.

“I wonder if I can build my hovercraft there,” mused Patrick. “Hey, I wonder if girls will be there?”

The next day, it hit me. What had I just committed to? Sure, Patrick had done about twenty successful sleepovers by this time, but Bryn Mawr was another deal. The campus was almost two hours away. A lot could happen in two hours. Also, it was three weeks long: five hundred and four hours during which Patrick would have to remember cornstarch, alarms, class schedules, and diet restrictions. It was a lot for a twelve year-old.

I took a deep breath and called the toll-free camp number.

“Ah. Will there be a nurse on campus? She’s one building over from the dorms? Can I get her name and office number? What about getting a few special food items during the meals? You have gluten free? Oh, that’s great. What about sugar-free alternatives? Wonderful. Okay, here’s my charge card number . . ..”

My next call was to Mrs. Meeks, the summer camp nurse at Bryn Mawr. Her friendly voice reminded me of Mrs. Mason, the loving school nurse at Union Township.

“Yes, Mrs. Shaw, we do have some children at summer camp with medical needs. Let’s see. I think in the June session we have twenty-five children total. Two use an epi-pen. Bee and food allergies. I’ve got one with exercise-induced asthma, and one with Type 1 diabetes. No, I’ve never worked with a child with GSD, but tell me about it.”

Mrs. Meeks listened intently on the other end of the line. I went through the basics of Patrick’s condition and care routine. Did she think she could work with us to help Patrick go to camp? Could Mark and I drive to Bryn Mawr before camp to go over information with her and form a game plan for nighttime cornstarch?
While we went to Bryn Mawr, Tee was to spend two days with his friend Brett. Brett’s dad had dirt bikes, and the three would head out to zoom trails each day. On the Big Planning Day, Mark, Patrick and I hopped in the car to meet with Mrs. Meeks. I was all a-flutter with excitement for Patrick, yet filled with fear, too.

Bryn Mawr and Mrs. Meeks did not disappoint. Mark, Patrick, and I walked across a grassy campus and through a dignified granite archway of rocks into the nurse’s office. After an hour of reviewing Patrick’s forms, showing off his Mic-key button, and hearing a vigorous discourse on how to build a hovercraft, it was set. The Plan. Mrs. Meeks took her place alongside the other angels I had met thus far in life: Retha, Dr. Stanley, Amy, and Mrs. Mason.

As we discussed with Mrs. Meeks, Patrick would set his alarms at nighttime for larger cornstarch doses that would buy him four hours of sleep at a time. After each nighttime dose, Patrick would call the nurse on her dorm room phone, using his new cell phone. If she did not get a call, she would physically head to Patrick’s room to make sure the dosage had happened. This nighttime routine meant that Mrs. Meeks would set her alarm, too, every night, for three weeks.

I pulled on a cuticle nervously. “Maybe we should call Patrick, too, each night? Just to make sure?”

“Mom, we’ve got it. Patrick is responsible and I’m responsible. I’ll call you the first morning of camp to let you know how it’s going.” Mrs. Meeks looked at me with sympathetic eyes; she was a mom, too.
We lined up the supplies on Mrs. Meek’s office shelving. Twenty-five boxes of cornstarch, sugar-free Citrucel, plastic spoons, syringes, tubing, bottles of water, extra plastic containers, Level Life glucose gel, goldfish crackers, allopurinol, oral Zofran for nausea, and an extra Mic-key button. Phone contact list, ER instruction list, cornstarch schedule. Extra timers. That was it. Mrs. Meek’s shelving was ready for end-times.

A week later, Mark, Patrick and I made the trip back to Bryn Mawr. As we got “the baby” settled into his room, I compulsively laid out spiral notebooks, pencils, and cornstarch supplies. I placed two weeks’ worth of shorts, tops, and underwear in his drawers. I hung up towels in the bathroom and generally obsessed. Toothbrush: check. Comb: check. Antibacterial hand wipes: check. Had we forgotten anything?

“Janet, calm down.” Mark looked at me, shaking his head. He was always patient and philosophical. “We’re leaving Patrick at Bryn Mawr with a nurse three hundred feet away. He’s not in outer Mongolia.” Mark was right, of course. In two weekends, we would head back to Bryn Mawr for Parents’ Day. I could swap out fresh clothing then, sample the camp food, and watch the kids do their improvisations. This was not outer Mongolia. But still--my baby would be almost two hours away from me. It was a scary first.

I gave Patrick one last hug before he and his new roommate dashed out the door to the first organized event of the camp: Frisbee tag, followed by a picnic dinner, a campfire, and a storyteller from nearby Haverford.

“Remember to—.” I started to remind Patrick of his next cornstarch time.
Mark touched my arm. “Stop, Janet. Patrick set his watch alarm with me while you were hanging up towels in the bathroom. He’s playing Frisbee right out there. See? Two hundred feet from his cornstarch supplies. His room key is on a lanyard around his neck. There are three teachers out there. Let it go.”

The control freak in me softened a bit. I took one last desperate look around the dorm room, smoothed Patrick’s bedspread compulsively, and headed off campus, grabbing Mark’s arm for support. The drive home was long and endless, as mile after mile separated us from our child. Our child with a chronic illness.

That night back in Pittstown was also long and endless. I finally slept at about midnight, waking up automatically at 3:00 a.m. I had gotten up at that time for years, and sixteen years later, I still wake up at 3:00 in alert mode. That night, I eased out of bed, wandering down the hall to Patrick’s room. His roller blades leaned in one corner. I opened the top drawer of Patrick’s dresser. Pogs and Beanie Babies. The faded blue deede, long forgotten. I looked over to Patrick’s bathroom, his toothbrush holder empty and crumpled red boxers on the floor. Was Patrick up right now, doing his cornstarch? The sixty miles to Bryn Mawr seemed so far, so far.

* * *

Dear Mom,

You never told me being a mom was so hard!

I wrote you last week about Patrick’s first time to go to camp. It’s a lovely place, and thank goodness, there’s a fantastic nurse there! She called me the first morning of camp. Somehow, I think she knew I was pacing the night before at 3:00 a.m. Well, I was
up, looking at Patrick’s dee dee and sniffing a bit. Mom, he’s growing up to be such a bright, funny, ornery kid. Can you believe he’s as tall as I am?

The cornstarch routine at Bryn Mawr is going fine, according to Mrs. Meeks. Oh—she’s the nurse. Can you believe she sets her alarm each night to make sure that Patrick has done his cornstarch? I’m going to write the director of the summer program and tell her what a dedicated person Mrs. Meeks is.

Patrick has fallen in love with improvisation. His little group does all kinds of practice drills, like “Same Circle” and “Word at a Time.” For Parent’s Day, we got to come up for a picnic and look at the art projects. I’ll never tell Patrick this, but when it comes to art, he’d better not make it his day job. I looked at his acrylic painting, hanging neatly with the rest of the kids’ artwork. I know you can’t say, “What is this supposed to be?” But really, Mom, what was it supposed to be? Patrick had painted a big red circle on a black background. That’s it. A red circle. The title written on the painting was “My Cornstarch.” Hmmmmm. Makes me think. Makes me worry. I’ll talk to the therapist at CHOP about this. I have to say that, on a brighter note, Patrick’s improvisational skills were amazing. He’s so quick. Patrick’s skit group did a scenario where two of them were in a taxi and he was the driver from “Joisey.”

At the end of the three-week camp, Mark, Tee, and I hauled Patrick home. The back of the station wagon was filled with stinky laundry, art projects, and several notes from girls. The front of the station wagon was filled with victory. Patrick had successfully managed his health care for three weeks. In the meantime, Tee had reigned king at home. We had cooked his favorite foods and had rented his favorite movies. He was taking a week off work and about to go to soccer camp. The Shaw family felt almost “normal.”

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26 Our Saint with the Knee-High Cast

Dr. Stanley looked at Patrick with his kindly blue eyes. “Would you consider doing this?”

I looked over at Patrick, who was now a strapping thirteen-year-old. He was so tall that I looked eye-to-eye at him. Patrick gazed at Dr. Stanley with an expression of interest. The three of us were sitting in an exam room at the Children’s Hospital of Philadelphia after an ultrasound, bone scan, blood work, and McDonald’s.

“We really need smart young people like you to help with research on GSD,” continued Dr. Stanley. “Wouldn’t it be great to help with a new type of cornstarch? One that would allow teens like you to sleep seven hours at night instead of only four hours? Plus, you would really get to know Dr. Weinstein and his staff. I consider Dr. Weinstein the top expert in the world on GSD. That’s all he does. He lives it and breathes it.”

Dr. Stanley had just told us the devastating news that he was retiring from endocrinology at the age of seventy. The new endocrinology department head was from Boston, but she had little experience in GSD. Unfortunately, the Shaw family would have no expert close by to turn to.

I looked again at Patrick who, in a single, fluid movement, poured water into a cornstarch container, shook it vigorously, and gulped it down. It was 3:00 p.m. and Patrick’s watch had beeped. Patrick nodded to me yes, wiping a cornstarch mustache off his lip. He rattled in his backpack for saltines.

“Let me talk to Patrick on the drive home.” I rose, shaking Dr. Stanley’s hand for the last time. “I am so grateful to you. Patrick is doing so well. He’s doing sleep overs and did a camp at Shaw 151
Bryn Mawr. Before long, he’ll be learning to drive. Then maybe college. We’re on a good road to independence.”

Dr. Stanley stood and shook my hand. Patrick looked down, embarrassed, hesitated a second, and dove in to give Dr. Stanley a big hug. The doctor beamed. I swear I saw just a hint of a tear in one eye. “Patrick, I’m going to read about you one day in some professional journal. Keep studying, young man.”

That night, the family gathered over dinner as we all shared our day. Mostly, we took turns sorting through the materials Dr. Stanley had given us that afternoon. It was exciting stuff. Really exciting.

We learned that Dr. Weinstein was a pediatric endocrinologist who had specialized in GSD the past twenty years. As a fellow at Harvard University, Weinstein became intrigued by GSD—“ . . . this disease that no one else was touching.” Weinstein moved to Florida in 2005 to work with the Gene Therapy Institute to develop genetic therapies for GSD. I scanned through the information further. “At present, he and his small staff operate on part of a floor at the University of Florida hospital in Gainesville, Florida. Dr. Weinstein sees approximately 500 patients each year; including those from 49 states and 45 other countries.” I read these words to the family, as a bit of hope flamed in my heart. “Oh, guys, listen to this.” I read further. “Iris Ferrecchia is a nurse for Dr. Weinstein. She has four grown children, three of which have GSD.”

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2 Ibid.

3 Ibid.
Oh, my. Here’s what Iris says . . . “When you have a doctor who cares so much, it’s easier to have kids with it [GSD] because you don’t feel like you are alone . . .”^4 Apparently, one of her sons needed a liver resection because of tumors. “Listen to what Ferrecchia says!” I waved another page of information at the family. “I felt like I needed a doctor at 2 a.m., and [Weinstein] answered the phone.”^5 The nurse adds in her comments that Weinstein is often at the office until midnight; he is known to sleep just a few hours a night . . . Dr. Weinstein is an interesting combination of science and love.”^6

“Yeah, Mom. Let’s do this.” Patrick shoveled in food with enthusiasm. “I can be a science experiment.” Testing a new type of cornstarch wasn’t exactly like building a hovercraft, but still, I could tell Pat was intrigued.

“And Pat. Are you okay with what it says here in the information? You’ll be in the GSD wing of the University of Florida’s hospital while I’ll be across the street at the Holiday Inn at night. Three days, it says here. A get-acquainted meeting with Dr. Weinstein and staff first. That night, after dinner, a nurse will insert a port into your arm. You’ll drink a dose of regular cornstarch, but at eleven o’clock, you’ll drink the experimental stuff. It says here . . . oh, where is it . . . the new starch is a high amylopectin—a high molecular weight. You can keep your port in during the day because they’ll tape your arm up really well. Then, we can take a taxi to nearby museums or something. We just have to be back at the hospital by 7:00. Yes, that

^4 Ibid.

^5 Ibid.

^6 Ibid.
means TGI Friday’s chicken fingers, young man! You’ll go through the same routine the second night. The third morning is a debriefing on your medical data.”

“Why do I have to get a port in my arm?” Pat didn’t seem afraid, just irritated.

I flipped through two more pages of information. “It says here that all through the night, monitoring equipment will track your blood sugar and lactic acid minute by minute. Whenever your blood sugar drops below 70, the experiment ends. You’ll take a dose of your regular cornstarch at that time, but the IV monitoring will continue until after breakfast that morning. You’ll have a continuous record running. So, are you okay with all of this?”

“Yeah, this will be cool.”

Cool, indeed. Patrick could help with GSD research, becoming the first “guinea pig” in history to test the new type of cornstarch. Even better—Patrick could become a patient of the top GSD guru in the world.

* * *

The University of Florida and Dr. Weinstein’s GSD research floor were amazing. How could a huge hospital and rows of research labs feel so welcoming? It was Dr. Weinstein and his staff. They were simply wondrous.

Patrick and I flew into Gainesville mid-afternoon for our three-day Beta testing. At the hospital, we wound our way through hallways and elevators until we got to the GSD stomping grounds. I glanced to the right and left as we headed to Patrick’s room. Hmmm. A GSD baby and her parents. A teenager. A twentyomething? Patients of all ages were waiting for GSD hope.
I remember that a freckle-faced nurse wheeled in a small cart with GSD-approved snacks and drinks. Patrick consumed cheddar goldfish with great relish, busily checking off selections for dinner. Check. Check. Check. Check. Being a GSD test subject was hungry business, indeed.

Dr. Weinstein breezed into Pat’s room about five, dressed in a white button-down shirt and a loosened tie picturing Tweety Bird. He clomped over to a chair near Pat’s bed. Wow. A bright blue cast went from his toes to his knee. Dr. Weinstein extended his hand.

“Mr. Patrick. I’m so glad you were able to come and help with our GSD Beta testing. Can I sit and visit for a while?”

“Uh, sure. What did ya do to your leg?” Patrick was not one to beat around the bush.

“Well, I’ve got a patient from Clifton, New Jersey, and his insurance company is refusing to pay for his GSD blood work and treatments. I lost my temper last week and kicked the wall of my office. I’ve been trying to help the family get medical coverage and nothing is working.”

Wow. Dr. Weinstein was a man of passion and caring.

I sat across from Dr. Weinstein with my yellow legal pad, busily writing down the conversation with Pat. In an hour, the two talked of Pat’s career interests. Who knew Pat wanted to be a lawyer some day? Who knew he wanted to start weight training? Who knew he liked Samantha? Furthermore, who was Samantha? Then, Dr. Weinstein drew out a diagram of how Pat’s liver processed—or mostly didn’t process—glycogen into glucose. The two talked of high school and college and the inherent risk of alcohol. Apparently, Pat faced a double whammy: alcohol would inflame his already-challenged liver. Too much alcohol could also make him miss his cornstarch alarm. And with weight training—steroids were especially lethal to folks
with GSD. Unfortunately, I learned that some GNC weight-training supplements contained small
bits of steroids and had to be avoided at all costs. I scribbled away, amazed at the vast amount
of information flowing from Dr. Weinstein. Where had this saint been all my life? I glanced at
Pat. He was engaged with the earnest doctor as information passed back and forth.

With the excitement of a twelve-year-old, Dr. Weinstein thumped into Pat’s room the
morning of day two, sporting a Tasmanian devil tie this time. He had a koala bear wrapped
around his stethoscope, to boot.

“Great news!” he announced to the two of us. “Guess how long you went last night on
the new starch?” Dr. Weinstein didn’t wait for an answer. Waving sheets of data, he exclaimed,
“Eight hours! You went eight hours! That’s ground-breaking. Now, we’ll need to see how you do
tonight. And, of course, the new starch will be tested on scores of GSD patients of all ages. We’ll
do refined dosing tests in a few months. I hope you’ll come back to help us, because we sure
can use young people like you.”

Mid-scrambled eggs, Patrick was full of smiles. He had never had eight hours of
uninterrupted sleep.

At the end of the three days at the University of Florida, Patrick also received a two-
week nutrition plan for GSD, as well as a detailed cornstarch dosing schedule based on his age,
weight, and activity level. Cornstarch had to be measured precisely for each dosage; something
that I hadn’t been doing so carefully. I hadn’t realized that too much cornstarch could elevate
Patrick’s blood sugar, but too little would allow the sugar level to drop. On day two of the GSD
study, Pat’s port arm was securely wrapped; he pulled on a long-sleeved checkered shirt.
 Apparently, it was one Samantha liked. We headed two floors down to the hospital pharmacy
to buy a glucometer and test strips for blood sugar testing. Then, according to Dr. Weinstein’s
instructions, we walked three blocks west of the hospital to a local head shop. There, amid the
marijuana pipes and bubblers, we bought a really good gram scale. A gram scale has a “tare”
function, so that the weight of the plastic container does not factor into the weight of the
cornstarch. The dosage for Patrick would be precise—36 grams per dose during the day; 65 at
night.

In thinking about those three groundbreaking days in Gainesville, I remembered the
practicality of Dr. Weinstein’s help. We received custom-tailored ER instructions on his
letterhead, a revised dosing schedule, and a travel letter (“Uh, young man, what are those little
containers of white powder in your backpack?”). We had pages of nutritional information and
easy-to-understand GSD medical data. One of the nurses gave us an application to join Medic
Alert so that Patrick would have a life-saving data card in his wallet at all times. As it turns out,
Medic Alert could even scan in Pat’s ER letter into its database, so ER physicians would know
exactly how to treat Patrick if he were brought into the hospital unconscious. Additionally,
nurse Iris came into Pat’s room one day to provide us with a pamphlet showing how to buy car
accessories for those with chronic medical conditions. I looked at the pamphlet. Amazing! I
could buy bright orange sleeves that Velcro’d around a car’s seat belt. Each sleeve stated
“MEDICAL INFORMATION” in bold, black letters. On the back of each was a zippered
compartment where I could slide in a copy of Pat’s ER instructions and cornstarch schedule. I
remember that when we got home to New Jersey, I bought four of the neon wallets—two for
Mark and me, one for Tee, and one for Pat’s car. The kid couldn’t even drive yet, and certainly
didn’t have a car, but we would be ready for that day.
One year later, we headed to Gainesville again to be part of Dr. Weinstein’s final study of nighttime cornstarch. In this study, the endocrinologist and his team fine-tuned dosing requirements for the new starch. Again, Patrick “lasted” eight hours on the new starch both nights. By this time, Patrick was five-foot eleven. Before we’d get ready to leave the hospital for our day’s fun and adventure, he’d go visit a patient’s room, with Dr. Weinstein’s permission. He’d normally visit a GSD baby or toddler, the parents often sitting by the bed in numb despair.

Patrick was a chatty guy. Within thirty minutes, the parents would learn that Patrick was a ninth grader. He was going to take Driver’s Ed. He had been on the swim team. He had a girl named Samantha that liked him. He’d been on sleepovers. He’d been to camp. He had once eaten a roly-poly bug on a dare. His big brother threw cicadas on him two years ago.

Each time, we’d see hope creep into the parents’ faces. Patrick symbolized a “normal” kid—if “normal” really exists. I would talk the parents through steps that would help their child become more independent. Patrick and I tried to be Team GSD hope.

When Patrick was a sophomore in college, the new cornstarch product launched—Glycosade, by Vitaflo USA. To this day, hundreds of individuals with GSD use this product to achieve something you and I take for granted. A good night’s sleep. However, here’s the irony of Patrick’s beta testing adventures: he can’t use Glycosade because it makes his stomach hurt. But, you know what? Other GSD kids can.

* * *

A few months after our second testing session at the University of Florida hospital, I got an email from Dr. Weinstein. In addition to GSD1a, his research team was studying GSD1b. Would Patrick and I be available to go to the NIH in Bethesda, Maryland to provide a blood
sample during summer break? An anonymous GSD1b patient would simultaneously provide a sample, too. The blood work had to be drawn at the same time for effective testing and data storage. Dr. Weinstein and other scientists were attempting to determine differences in blood composition that might account for the additional difficulties incurred by Type 1b patients.

Dr. Weinstein knew that Patrick was somewhat a science whiz-kid. So, the endocrinologist provided us basic information on the differences between GSD1a and b. Essentially, the enzyme deficiency in 1a is the Glucose-6-phosphatase-α catalytic subunit, impacting the G6PC gene. The chromosome location is 17q21.31. On the other hand, the enzyme deficiency in GSD1b is the Glucose-6-phosphate transporter, impacting the gene SLC37A4. Its chromosome location is 11q23.3. Both diseases are considered autosomal recessive and present themselves as hepatic, meaning in the liver.\(^7\) 1a occurs in 1/100,000th of the general population, impacting specifically the Ashkenazi Jewish, Mormon, Mexican and Chinese heritages. On the other hand, 1b accounts for only 10 – 15% of all Type 1 GSD cases. High-risk populations for 1b include Native Americans, Iranian Jewish, and those of Italian heritage.\(^8\)

According to Dr. Weinstein, GSD1b may appear clinically similar to 1a for many years. At some point, however, patients with 1b will develop cyclical or permanent neutropenia, which means that neutrophils are lower in number than in a healthy patient. Neutrophils are cells in

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\(^8\) Ibid.
the patient’s immune system that attack bacteria and other organisms when they invade the body. So, a patient with neutropenia is subject to numerous infections and fevers, much like a cancer patient who has undergone chemo. Another major complication of GSD1b which impacts nearly every patient between the ages 5 and 12 is inflammatory bowel disease, resembling Crohn’s. This condition is often called “enterocolitis;” it impacts the small intestine and creates diarrhea and abdominal pain. A GSD1a patient under poor metabolic control can face growth retardation, hepatic adenomas, osteoporosis, sclerosis of the kidney, small fiber neuropathy (peripheral neuropathy and tingling), and renal complications. The 1b patient must also manage these conditions plus neutropenia and enterocolitis.

After I got the email from Dr. Weinstein, I snagged Patrick after school, luring him to the kitchen island with unbuttered popcorn. I showed him Dr. Weinstein’s email and the information about GSD1b patients.

“Jeez. They have it a lot worse than I do. Oh, my God.” Patrick slid his hand, blue and grubby from art class, up through his curls.

“So, what do you think?”

“Can I do this before I start Driver’s Ed? That’s in late June.”

“Sure. Let me call the NIH tomorrow. Dr. Weinstein has provided us a testing case number to refer to. He said something about paperwork we will need to fill out before we go.”

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When Dr. Weinstein said “paperwork,” it was an understatement. Within a week of calling the NIH, I received a hefty packet in the mail. I dove into the pile of forms. I had to list my current address plus the three prior ones. Jobs—date of birth—social security number—parents’ last names. The make and model of my car, as well as the license plate number. Countries outside the U.S. the family had traveled to, and when. An official passport photo of myself and of Patrick. The NIH did not mess around.

In mid-June, Patrick and I drove three and a half hours to Bethesda to the massive NIH campus. We pulled up to the Gateway Center, stopping at the guardhouse.

“Hi!” I smiled at the guard, hoping my Kansas dimples would let us breeze through quickly. We had to be in the lab in one hour, first dumping off our suitcases at the Children’s Inn adjacent to the lab building.

The guard was not impressed by my Kansas dimples or fluttering of eyelashes. “Out of the car, please. Both of you.” From seemingly nowhere, two other guards appeared, along with a bomb-sniffing dog. Patrick and I stood, as directed, in a painted square about twenty feet away. A German Shepard circled the vehicle, sniffing intently for explosives, cocaine, and Uzis. He found nothing exciting, other than packages of snacks. One guard popped our trunk, took out the suitcases, and zipped them open, lifting our shorts and my pj’s with a stick. The other guard walked around the car with a mirror attached to a pole, examining the car undercarriage for more explosive devices. Guard number one circled to the interior of my station wagon, zipped open Patrick’s backpack, and took out boxes of Argo cornstarch and plastic containers of mysterious white powder. He took them inside the guardhouse, scanning the containers and
boxes with a gizmo much like we had seen at security at airports. Sure enough, we had no explosives, heroine, or cocaine. Just cornstarch. Lots of cornstarch.

Just a few minutes later, Patrick and I headed back to the lab building, up several floors, down more halls, and into a sterile waiting area. I felt like a rat in a maze.

After ten minutes in the phlebotomy lab, Patrick sauntered out, his blood drawn and mouth swabbed. Somewhere in the maze of laboratories nearby, a child with GSD1b did the same thing. Both sets of parents signed paperwork that the genetic data of each child should go into databases at the NIH, the Children’s Hospital of Philadelphia, the University of Florida, and Duke University.

Patrick and I will never know exactly how his genetic information benefitted patients with GSD1b. These patients can often manage the hypoglycemic issues of their condition, but the neutropenia and inflammatory bowel complications continue to plague them. However, as with many medical mysteries, scientists have made progress. In 2017, researchers at the NIH successfully used AAV mediated gene therapy to restore GSD1b mice that experienced fasting hypoglycemia.\(^{11}\) In 2019, researchers led by Veiga da Cuntha published that GSD1b neutropenia “could be treated by administration of kidney glucose transporter inhibitor (SGLT2 inhibitor).\(^{12}\) The hope and research continue.

Somewhere, amidst the cornstarch, the Mic-key button, the blood work, the syringes, the restricted diet —- lies purpose. Patrick knows he has stepped forward to help others with GSD. I think back to story time with Patrick when he bored into my soul with his large, blue


eyes, asking, “Why did God give me GSD?” Perhaps God knew that Patrick would help another child.
“I want to get rid of it.”

I was sitting next to Patrick at the kitchen bar, his backpack spewing out its usual contents of rumpled homework and red tickets. Tee would be home after track practice, so I always seized this time to eat popcorn with Patrick, do a little English homework (“Mom, The Yearling stinks”), and catch up on his life.

“Dr. Stanley said my growth is right on track. I’m fifteen and five-foot eleven. But, the kids in gym class look at me funny with this Mic-key button.”

My heart felt a slight pang. I remembered my middle school days of braces, glasses, and zits. I remembered how, in the locker room, some girls had cool turquoise bras and an awesome body to go into them. I remembered how, at the time, I did not have a cool turquoise bra or an awesome body. I just wanted to be just like everyone else, because to be different in middle or high school was humiliating. Patrick had been fairly indifferent about his GSD condition thus far, but the Mic-key button. This was different. It was an outward sign that he was not the same as others.

“Okay, okay. I’ll call Dr. Stanley tomorrow. Now, Patrick, do you realize what this means when the Mic-key button is gone?”

“Yeah. I know. I’ll have to get up in the night and mix up my cornstarch and gulp it down.”

“Right. The rest of your life. No more Mom doing the dose through the Mic-key while you sleep. You’ll have to do it. Every night. Even when you’re tired.”

“Mom, I can do it.” Pat was dead serious.
“Okay, then.”

The next day, I tracked down Dr. Stanley.

“Actually,” he said, “You can use the small syringe that comes with the Mic-key button. Remove the liquid from the balloon in the stomach and gently pull the button out. Clean the area three times a day, apply Vaseline, and keep a large square of gauze securely taped over it. The hole should begin to heal up in two to four weeks.”

It didn’t heal up. Patrick was a complicated guy.

About six weeks after Patrick had removed his Mic-key button, he was in GI surgery at the Children’s Hospital of Philadelphia. The hole into his stomach was stitched shut. Within months, all that remained was a thumbnail smile of a scar. Mic-key was gone.

* * *

Time seemed to accelerate during the rest of Patrick’s high school years. Meanwhile, Tee had gone off to Radford University. The nine-hour drive to see him for parent’s weekend had left Mark and me sobbing all the way back. One day, the following week, I even found myself sniveling into my shopping cart at ShopRite grocery. The separation was hard for us, but Tee was in absolute heaven.

More saints entered into Patrick’s life when he was in high school.

Dr. Ripton taught American History, British History, and Comparative World Cultures at Pat’s school. His wife, Jane, taught A/P English. She caught on in a New York minute that Patrick was not actually reading the assigned novels. Pat was smart, of course. He’d read the first five chapters of each book, and Cliff-Note-it through the rest. His footnotes in his papers only ranged from chapters one through five.
Patrick, a good writer, got a “C” on his first paper.

“Do you find nothing worthy to footnote in the rest of *The Rule of the Bone*?” Mrs. Rushton pinned Patrick to his chair one day with a deadly stare, while the rest of the class smirked.

“Uh. . . uh. . . uh.” The vocal Patrick was convicted. Amazingly, he began to read each of the remaining five novels of the course. And, the goof-off student found he loved reading.

Dr. Ripton opened Pat’s eyes in his class on Comparative World Cultures. Pat, along with his other classmates, had known little injustice in fifteen years of middle-class whiteness. Their worst battles involved “unfair” coaches or “mean” teachers who objected to paper airplanes sailing through the air. Dr. Ripton, however, taught the kids about atrocities conducted by the “good guys” during the Bosnian war. He dealt with the Tongzhou mutiny, the Bodo League massacre, and the ethnic cleansing of Circassia. Patrick, long the conspiracy theory-lover, became enraged as he learned about atrocities in Latin America, mostly sponsored by the US government at the behest of the boards of directors of various fruit companies. He, fellow students, and Dr. Ripton gathered with others to protest the killings in Darfur. Patrick picketed against the policies at Wal-Mart that hedged health benefits. I was thrilled. In the sea of apathetic teens that cared only about the next party, Patrick became engaged and morphed from a class clown to a passionate student. I owe all to Dr. and Mrs. Ripton and others that nudged and encouraged Pat. As a Congolese priest friend of ours says, “You can’t pick up a nut with one finger.” Raising a child is indeed a team effort. It’s a long, massive effort.

* * *

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At the end of each school year, Patrick’s school would conduct two full days of final exams in every subject. At the end of final exams, the students were given an opportunity to take two weeks of optional, outside-of-class learning. Ninth graders had to stay locally, so, of course, Pat took improvisation. The “Joisey” cab driver in him longed for more expression. At the end of his tenth-grade year, Patrick was given the opportunity to travel with teachers and classmates to Israel.

“I really want to go! Dr. Ripton and his wife will be going with us. Twelve of us will be selected to go for the nine-day journey.” What Patrick neglected to tell me was that the trip would include areas near the bombings at the Gaza strip.

Mark and I have always believed that at least half of a person’s education occurs outside the classroom. We wanted both of our sons to know more than just the immediate bubble surrounding their lives. But nine days in Israel. Tel Aviv was 5,700 miles away and a six-hour time difference. Israel was a high-tech country, but also a country with political and spiritual tensions. We didn’t know the country’s hospitals or the doctors. This was no little jaunt to Bryn Mawr, but I was determined that Patrick would have opportunities similar to his classmates. Independence for Patrick was a journey—not five steps, but twenty, or forty.

Mark and I met with Dr. Ripton and his wife, much like we had done with Mrs. Meeks at Bryn Mawr. We reviewed Pat’s ER schedule and cornstarch schedule. We discussed nearby hospitals where the group would be staying in Israel, should Patrick get the stomach flu. We detailed food restrictions. The Riptons were “on;” Mark and I began gathering supplies, updating medical forms, drafting a power of attorney for medical care, and checking Patrick’s vaccinations. A lawyer friend of us had advised us on a power of attorney, which had to be
signed and stamped by a notary public. Apparently, some hospitals and medical practices would not admit a minor child if not accompanied by a parent or guardian. Mark and I would take no chances regarding this trip to Israel.

The differences between Israel and Bryn Mawr are huge, as you might imagine. For the trip to Israel, we had figured out with Dr. Ripton’s assistance, how close the various hospitals were at each point of the itinerary, obtaining their addresses and telephone numbers. Fortunately, the sponsors had a mobile phone with international calling abilities. Equally important, we had to make sure Patrick had a box of Argo cornstarch for each day he would be gone, plus several more in case of accidental spillage.

The average traveler doesn’t know that Argo cornstarch can’t be purchased in a typical grocery store in Israel, or in Britain, Ireland, or Martinique, for that matter. Sometimes you can find “corn flour,” but structurally, it’s not the same; it fails to keep blood sugar up adequately. Sometimes, you’re lucky and can find tapioca starch, which is a good substitute for Argo. Also, the average traveler doesn’t know how nervous the TSA becomes when a teen shows up in security with small bottles of white powder, a bottle of Citrucel, and multiple boxes of Argo. It’s a showstopper, GSD travel letter or no travel letter. To ease the potential panic at the airport, we always insisted that Patrick get a decent haircut before travel and wear a collared polo shirt and dockers. It wasn’t the time to look scruffy. It was sad but true; stereotyping was alive and well.

Patrick contacted several of his buddies on the same flight to Israel. Sure, they could also carry on several boxes of Argo apiece. So, on the evening of the nearly eleven-hour flight from Newark to Tel-Aviv, five friends carried boxes of Argo cornstarch in their backpacks, along
with Pat. All told, fourteen boxes would travel to Israel. I knew the TSA was going to have a hissie over the massive amounts of white powder making its way to Tel Aviv.

The big day in early June came, with Pat and his buddies scattered throughout the crowded El Al flight. Most of the passengers on Patrick’s flight were Hasidic Jews. During the long flight, the passengers repeated the Tefillot (prayers) twice. Patrick, as he told me later, remembers being antsy. He had read a bit of Fear and Loathing in Las Vegas and had dug around his backpack for snacks. He drifted off to sleep periodically, only to be awakened by the soft murmuring periodically resonating through the plane. He arrived in Tel Aviv early in the morning, bleary-eyed from little sleep and multiple time zones.

Normally, I’d expect the sponsors to let the kids re-group the first morning. After all, they’d been on a long overnight flight from Newark, New Jersey. But no. That morning the kids and sponsors headed off on a rigorous bicycle ride through Masada National Park. Somehow, Pat made it through the ride, feeling sick and exhausted. Some of the crazier kids had even decided to hike to the top of the Masada fortress at 5:00 a.m. Pat had decided to sleep in a bit, reckoning that a sunset would look just as good as a sunrise.

However, the time in Israel was magical. In addition to Masada, Dr. Ripton’s group visited the Baha’i Shrine, the Sea of Galilee, the Mount of Olives, and the Western Wall. The bell towers rang daily with the call to worship for the Jews, the Muslims, the Christians, and the Druze. Patrick’s travel group feasted on hummus, tahini, and sliced tomatoes. For a kid from Kansas, Israel was a place of magic, of incredible technology, of unique sights and sounds.
“I hate this expression,” said the bedraggled Patrick as Mark and I picked him up at the Newark airport, “but the whole time on my trip, I knew I wasn’t in Kansas anymore. But I’ll tell you—if I see one more plate of hummus . . . .”

“Did you click your ruby red slippers together to get back?” I teased.

“Oh, mom.” Eyeroll. Eyeroll. I had missed my teen with the attitude. Most importantly, my son had managed his GSD well on the trip, despite different time schedules, food, climate, and activities. Patrick came back exhausted, but alive and determined that GSD, that ever-present companion, would not hold him back.
It seemed like I had just quit sniveling over Tee’s absence, yet we were about to launch child number two into the halls of academia. More likely, the halls of little sleep. Of fraternities. Of drinking. Of temptations galore. Of frightening independence.

Dr. Ripton’s influence was ever-present as Patrick applied for various colleges. Would it be Dickenson College, Franklin and Marshall, or the University of North Carolina? Duke? Nope. He had good grades but not good enough. Perhaps American College or George Washington University? Patrick was intent on studying international relations, with an eye towards grad school.

Patrick and I looked at colleges, while Mark worked away, preparing for the tuition blast. Patrick and I would go on college tours, absorb the admissions hype, then hang in any student common area we could find.

“Yeah. The kids at Franklin and Marshall look pretty serious.” Pat crossed his legs and flipped through the glossy college brochure again. We sat at a picnic table in the commons area, sipping sugar-free lemonade and eating Cheetos.

“Well, maybe they’re here, uh, to study?” I said in my most motherly tone.

I got my third eyeroll of the day. It was a banner day for being a mom.

After all the tours, forms, essays, interviewing, testing, and gnashing of teeth, it was down to two universities: George Washington University in D.C. or the University of Delaware in Newark. George Washington had the feel of a city school and a powerhouse international relations department with significant government connections. On the other hand, U Del had wide, green lawns, red brick buildings with ivy, and, most importantly, cute girls.
“I don’t know what to do!” Patrick would wail to me five times daily. “GW has all kinds of government ties which might be helpful to me in my future career or maybe law school. But U Del looks like a college is supposed to look. The kids seem to be happy and connecting with each other. There are fraternities, and I saw some blondes . . .”.


“Well, I’m going upstairs to take a shower and think on it.” I had never heard of thinking in the shower, but it was close to singing, right? Within two minutes, I heard the soft whoosh of water above me, followed by a sharp clunk of the soap hitting the shower floor, followed by “arghhhhhh.” More thunks of soap hitting the floor and then the frustrated slap of flip-flops. Another prolonged “arghhhhhh” and frustrated pounding on the shower wall.

Patrick came into the kitchen slightly damp, sporting only his plaid boxers. I pretended to be busy rolling up Italian meatballs. “I’ve decided,” he announced. *Good thing,* I thought. The deadline to pay by credit card was *tonight,* at midnight. Nothing like taking it down to the wire.

“It’s going to be U Del. I gotta learn but have fun at the same time.” Pat pulled raw carrots and Ranch dip out of the fridge and began munching. Contemplating in the shower apparently made a person ravenous.

“You know, Pat, I lived at home my first year of college and was absolutely miserable. It was for financial reasons, you see. But I worked hard the next summer and made enough money to live on campus my sophomore year. Strangely enough, my fun level went up one hundred percent,
and my grades went up to all A’s. Play hard, work hard, as they say. But in your case, don’t play too hard--.”

I got yet another eyeroll for my efforts and dug out my credit card to phone in the tuition deposit. The card would soon be burning and continue to flame brightly each semester.

* * *

I wish I could report that Pat’s college years were uneventful, but of course, they weren’t.

Pat’s dorm roommate and soon-to-be-best-bud at U Del was dark-haired Cody, a kid from a speck of a town called Magnolia, Delaware. Cody had black-fringed eyes and a slow drawl that belied his incredible intelligence.

“Uh. Cody. I wanted to let you know about these containers of white powder that you’ll be seeing here on Pat’s bookcase. Well, it’s not cocaine.” I was busy unpacking school supplies in the dorm room while Mark vacuumed. I grinned inwardly at my feeble attempt at a joke, continuing to explain Patrick’s condition to Cody.

“It’s no problem. I’m cool with it. Say, looks like you could use some help with that futon.” Cody leaned back in his desk chair and eyed a jumble of parts on the dorm room floor.

Cody was right. Mark and I, sporting six degrees combined, had thirty parts of a futon scattered around the doom floor. It was a hopeless mess, but Cody, the budding engineering major, dragged out his blue toolbox and began to assemble parts. I didn’t know what kind of kid brought a full-fledged toolbox to college, but I did know this. Cody was smart and patient—an all-around good guy with four—count ‘em—four types of wrenches.
Within weeks, Cody and Patrick pledged a fraternity and jumped into coursework. They also jumped into parties and girls. Most importantly, Patrick found his passion at U Del: improvisation. He became a member of the campus group called The Riot Act, practicing four nights a week. Somehow, amidst all the frantic activity, Patrick did study, double majoring in international relations and economics. His minor was philosophy, which meant that, over the next three years, he and I would have interesting conversations on the Copenhagen concept or determinism and freedom in society. I loved these exchanges with Patrick and would scurry to Wikipedia afterward to look up what the heck we had been discussing.
U Del had a trimester system, which meant that the students had the entire month of January to work or do a short travel abroad class. Now, I had never traveled abroad in college—that was just for rich kids. Instead, I worked between terms at the campus instructional media department, cleaning sixteen-millimeter films with toxic chemicals and wheeling around carts loaded with projectors and clunky monitors.

Patrick arrived home for fall break his freshman year at U Del. Nearly breathless, he burst in the door and handed me a mesh clothes hamper. In a moment of madness, I had volunteered to do laundry, since Pat had been busy studying for mid-terms and performing improv.

“Mom! You’ll never guess what I can do this January for a class!”

I gave Pat a massive mom hug as he lugged the mesh container back to our laundry room. I pulled out two white tee shirts. Within seconds, plaid boxers and ragged jeans sprang out of the container, sending up a sweaty, college-boy smell with them. I turned my head, pulling out an item at a time with my thumb and forefinger. One load of laundry, washed twice. A second load, washed twice. A third load. A fourth. A fifth. As it turns out, Patrick had done no laundry for two months. Zip. Nada. I guess he was too busy performing with The Riot Act and thinking about metaphysics. Apparently, clean boxers were a plebian detail of life.

Pat followed me into the laundry room, impervious to the wafting stink. “U Del offers a January travel abroad class. It’s offered by Dr. Gita Barua, a professor of Indian literature. We would travel for twenty-five days, going to Assam, Agra, Goa . . . uh, I forget the other states. It
would be so cool. We would get to see the Taj Mahal, the Amber Palace, and the Agra Fort. We would study Indian literature which has been totally ignored in our culture.”

Patrick rolled this information off his tongue as casually as if he were driving down the street to Walmart. For a kid from Kansas, how did he know about India?

Deep breath and a forward verbal launch. “This would be so good for my international relations major at U Del. I’ve got to start building my resume now.”

I had to hand it to Pat. He was a smooth talker. I stood in the laundry room, a bit stunned. India? I sat down at the tiny table in the corner of the room and Googled rapidly. India. About 8,200 miles away. Over one billion people. Oh . . . over nine hours’ time difference. Holy smokes.

The Kansan/mom/perfectionist/worrier started thinking about germs. Real germs. I had read somewhere that India had more than its share of germs. I Googled some more, and there it was: “India has one of the world’s highest number of bacterial infections; tuberculosis, cholera, typhoid, pneumonia and a host of them. In fact, the country has the highest number of tuberculosis patients in the world, accounting for one-fourth of all TB patients in the world. Multi-drug resistant (MDR) and extensively drug-resistant (XDR) tuberculosis cases are increasingly reported.”¹ I pushed back from my table, feeling a little sick in my heart. Multi-drug resistant cases?

“I don’t know, Pat,” I said, rubbing my forehead with anxiety. “Look at this data. If you get any stomach issues and can’t get to a hospital within about two hours . . ..”

“Mom! We’re staying all over India, but with families that own schools. They’re ‘comfortable,’ as you and dad like to say. Their houses will be much nicer than ours. They have servants and doctors and hospitals there. Dr. Barua grew up in India and went undergrad there. Are you seriously going to keep me at home the next forty years and tuck me under your arm?”

“Maybe.” I smiled weakly at Pat, knowing that every fiber in my body wanted to protect him. Our family had been through so much, so much.

* * *

It wasn’t until we got pages and pages of data about the month in India that I began to relax. Just a tiny bit.

Mark and I faced this dilemma: just how much do you “let go” of a child with a chronic condition? Sure, Patrick had navigated his time in Israel well. He had gone the next year to Japan for nine days with a class group. Both trips went by without incident. But the stakes were higher for the trip to India. Further away. Longer. Scarier. Would I have let Tee go on such a trip? Were we being proactive in letting Pat take such a huge step to independence? Or, were we just being crazy parents?

In looking at research today about chronically ill children and nurturing independence, it’s not always helpful. Nearly all of the advice is framed in terms of parenting a five-year-old. For example, have him participate in household chores. Walk the dog. Be responsible for homework. What if the “child” is 19 years old and six feet tall—and the independent act is one of traveling to India? It’s a different scenario entirely. There’s no roadmap for this.

Despite all this, some helpful suggestions exist. The University of Michigan Health System website reminds me that “Parents who have been very involved in their teen’s care for
many years may find it difficult to let go of their role as primary caregiver.” To that I respond, *Good grief! Has someone been inside my head all these years?* The website also provides additional information to guide me through Patrick’s development. For example, our family must stay involved with Patrick; give information; plan for procedures; give him choices, support his friendships; be hopeful; listen; teach coping skills; stay organized; be aware of the risks unique to Pat’s illness. It’s all great advice. The problem is, when your chronically ill child is a flesh-and-blood Patrick, all you want to do it hug him, throw out the scholarly articles, and never, never let go. Your heart can hardly take it.

I do remember our family’s decision process in allowing Pat to participate in the India seminar. Our questions looked something like this:

--How is Pat’s general health and GSD control?
--How well is he trained to manage his day-to-day health?
--Is Pat a responsible individual, laundry aside?
--How does he conduct himself in the emergency room?
--How has Pat handled his previous travel?
--Has he been responsible and health-conscious at college the last two months?
--What are his grades?
--How trustworthy are the faculty sponsors on the trip to India?
--Were hospitals available near every point in the itinerary?
--Could Pat get a working cell phone once he landed in India?
--What were the ramifications of the multiple vaccinations Pat would require?
--Could Pat learn and follow explicitly the many food restrictions in India?
--Could we get students to help Pat lug 35 boxes of cornstarch to and around India?
--Would we have let Tee take such a trip? How important was brotherly equality in the family?

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As it always does, the dust settled after many discussions. Patrick would go to study in India with Dr. Barua. After all, we had let Tee go on a school trip to Peru when he was twelve. Tee had to dodge snakes and spiders on his trip, as he and his classmates stayed in a lodge near Machu Picchu. Tee came back home with an evil-looking, two-foot machete as well as a carved blowgun. Ironically, airport security had allowed him to put his machete in the overhead compartment of the airplane, as long as the weapon was securely wrapped in newspaper. Who knew that newspaper was an effective anti-terrorist device twenty-five years ago?

“Pat, if you want to do this, you must obtain the name of a hospital for each location on the itinerary in India. Dr. Barua has this information, I’m sure. You will also need to meet with her in November to go through all your medical forms. This week, please call Dr. Rushton to get a list of vaccinations needed for your trip and I will schedule them. We probably need to start on some shots this week. This is all your responsibility.”

Even when Pat was in high school, we had done some of the preparatory work for his camps or trips, but now, he was a college student. He had to step up. It wouldn’t be that long before he was out working, we hoped.

Flash forward. Patrick got listings of hospitals and phone numbers for his travels, as well as names of host families and phone numbers. He endured vaccinations for hepatitis A, hepatitis B, typhoid, cholera, yellow fever, Japanese encephalitis, rabies, meningitis, polio, MMR, Tdap, pneumonia, and influenza. The kid became a human pin cushion. While Pat was enduring multiple shots, we were navigating the painful Visa application process. It seemed, like the NIH, that India wanted every vestige of information about us.
It was mid-morning at the Newark airport. Ten kids gathered with Dr. Barua, ready to go through airport security to board a United Airlines flight. The kids would be airborne for about fifteen hours, barreling through several time zones. During this time, Pat would have to manage numerous small snacks and his cornstarch, measured out to last four hours apiece. Once the kids landed, the group would buy cell phones in the airport.

I met all the students in the group, along with Dr. Barua. True to his word, Pat had talked to each one about his cornstarch and medical condition. No big deal. Each kid put three boxes of Argo in a backpack while Pat toted five. I gave Pat what I hoped was a non-embarrassing hug as the group prepared to go through security.

“Don’t forget to buy water once you go through! Use your antibacterial wipes! Call me! Protect your passport. Keep your money safe. Have—”

“Okay, Janet.” My husband put a restraining hand on my shoulder, but I think I detected a quiver in his voice. “Pat’s got this.”

I felt excitement for Pat. What would he see and experience? And I also felt real dread—that sick-in-the-heart feeling. My youngest was going to be far, far away—out of my efforts to help him. What had I just done here?

In the days ahead, my heart simply raced when Patrick called us.

“Mom, India is full of the most beautiful sights in the world and also the most wrenching.” Pat went on to tell us of shanties built in the road medians and the exotic, crowded spice markets. “I never forget that I was in India,” he would tell us excitedly on the phone. “And guess what! We go to the Taj Mahal tomorrow.”
I found out two weeks after Pat’s return what had happened at the Taj Mahal. He and two others had sneaked around the reflection pond to one side of the monument. They had all stuck their tongues out, licking the ivory white marble of the sacred mausoleum.

“Why!???” I exploded when I heard this distressing tidbit of news. “Why would you have done something silly like this? Talk about picking up germs. And it’s a sacred shrine!”

“Well, we figured that no one in history would have licked the outside wall of the Taj Mahal. It opened in 1648, and we were the first to do this!”

Perhaps there had been no one this foolish visiting the Taj Mahal since 1648.

* * *

Sometimes, four or five days crept by before I heard from Pat, because certain states in India did not permit cell phone contact with the U.S. During those days, I prayed often. I knew from the itinerary which state Pat was in on which day, but it was just a name for the Kansas girl. I was being tested on both faith and on letting go. Both concepts were uncomfortable for me, so you could say as Pat was growing and experiencing, so was I.

Reuniting with the group at the end of twenty-five days was exhilarating. I’d never seen a group of kids so excited yet so bedraggled.

Over the next two weeks, Mark and I heard more and more stories about India. There was the time when Pat and another student were heading back from a market to their group, only to be chased by a pack of wild dogs. There were occasions when Pat would be staying with a “comfortable” family, only to be confused and upset by the extreme deference of the servants. In the Shaw family, we did our own cleaning, our own yard work, our own home repairs. Pat had never seen a formal caste system in operation before, and it disturbed him
greatly. That understanding in itself was worth the entire price of a college education. Patrick was beginning to whiff the scent of social inequality.

The last bombshell Pat dropped on us was that he had been sick in India. Sick! I hadn’t known about it.

Apparently, Pat had been staying with one of the many families on the itinerary that owned a school. In fact, one of the sons became friends with Pat, giving him a designer scarf. At any rate, on day three with the family, Pat began to feel feverish and achy. The family called their doctor, who came out, examined Pat, and gave him a round of antibiotics. Within two days, fortunately, Pat rallied and was able to move on with the group. When Pat got home, he gave me the physician’s receipt for services. The house call plus antibiotics equated to sixteen dollars, U.S.

We will never know what “bug” Pat got. The blessing was that he got home safely, with a lifetime of adventures, his eyes wide open.
30 Tesoreria General de la Republica

Pat’s sophomore year of school rolled along smoothly. Schoolwork, improv, and fraternity obligations filled the days. In mid-October, Pat starting conspiring for his upcoming January course. By this time, he was knee-deep in international relations and economics courses.

Pat burst into the house at fall break, this time without lugging a loaded laundry bag. Hmmmmm. Could this be maturity in the works? Or did a mountainous pile of laundry exist back in Newark, Delaware?

At the table that night, Pat was full of stories about his roommate Cody, who pulled all-night study sessions before tests and still got good grades. And why could Cody eat fries and drink Cokes and not get fat? Then there was the incident of a college kid caught by the police at the intersection of Main and Chapel streets. The problem was that he had only his boxers on and may have been inebriated. Mark and I listened intently. The crazy stories made us want to go back to college, except we’d both have to be in bed by ten to make it through the next day.

“Anyway, guys, here’s what I found out about this winter break. There’s an organization called World Wide Chile that helps students find internships in Chile and also shows them around the country. I could work for a month in Santiago, Chile for their Treasury department. I would be the first American intern to do that. Ever--ever! Wouldn’t this be a great work experience to help me get my first job?” Patrick, always the marketing man. His blue eyes, now behind navy frames, blazed with intensity.
It did sound intriguing as a brief experience in international work. I got to Googling again. Santiago. 5,100 miles from the Newark airport, give or take. A decently high standard of living. A volatile history. And then I probed Pat further: he would be with a group of other students interning various places in Santiago. The group had a sponsor who would organize cultural gatherings on the weekend. Pat would stay with a host family and get food there. He could walk from his lodgings to his place of work during the month of January.

Planning for Chile was like planning for India, only there were fewer shots involved.

We had our medical checklists down cold. The dilemma of this trip was the cornstarch. Pat was traveling independently from Newark airport, meeting up at his host family’s home in Santiago via cab. That meant 35 boxes of cornstarch traveling with him. After some pondering, Pat came up with the solution. He’d have one large suitcase full of work shirts, khakis, and shorts, and an equally large suitcase of Argo. Add a computer bag. Add his backpack full of water, cornstarch containers, and snacks for fifteen hours. It would work, as long as both suitcases arrived with Pat to Santiago. What would happen if the suitcase with the Argo became lost? Pat could only carry four boxes in his backpack. He was fairly certain that packages from the U.S. to Chile got waylaid by CorreosChile, the postal service. Then what? Then what?

One day, I sat down at my computer, arranging airfare for Patrick’s big internship. It was all pricey, but finally, I found a decent fare on Continental Airlines to Santiago. Pat would fly from Newark to Haiti to Panama City to Santiago. A lot of transfers, but hey—it was affordable, and it was Continental Airlines. A big, stable company.

About twenty hours later, we got a call from Pat: he had landed in Santiago and had bought a temporary cell phone. He sounded shaken and rolled into a rant.
“Mom, don’t ever have me fly again to Haiti!” I could hear a quiver in Pat’s voice. “I’m not coming home that way. You’ve gotta check the flights back. I’m not doing it. You won’t believe this. When we landed in Haiti, the airline officials made us get off the plane, totally leave the terminal, and walk around outside—outside, Mom!-- to another terminal with all of our luggage. We had to go through security again. I walked out of the terminal and onto the tarmac, and then, presto! A man grabbed my suitcases and wouldn’t let go without me giving him forty dollars. I mean, I had TWO huge suitcases, my computer bag, AND my backpack. Then, as I was approaching the front of the airport, it happened again with a different man. It was a bad situation.” I could hear Pat inhale with agitation.

“Oh my God!” I could picture Pat, bogged down with baggage and trying to ward off people intent on “helping” him with his luggage. “Well, somehow you got out of this. What did you do?”

I had never dreamed that Pat would be forced to exit one terminal in Haiti, only to battle his way into the second one. I felt terrible. How had I assumed flying would be the same everywhere? Thank goodness Pat was a daunting six foot two by his sophomore year. That didn’t hurt.

“Then, mom.” Another massive inhalation. “When I got to security in the second terminal in Haiti, the guards gave me crap about having a whole suitcase full of Argo cornstarch.”

“Maybe they thought you would be doing a lot of baking.” My joke was wasted on Pat.

“I couldn’t even talk to the guards, because I don’t know Haitian Creole. I showed them my travel letter and they just shoved it back at me. These were big, burly dudes with Berettas.”
“How’d you get through? Did you know anyone else in line?”

“Well, on the flight over I sat next to a young doctor who flies to Haiti each year for a volunteer medical clinic. Thank goodness—she saw the ruckus and came forward to explain to the guards. I got through. But I’m just sayin’, I’m never goin’ . . . .”

“I’m so sorry, Pat. I had no clue that you would have to go through with this experience. But, a good reaction and thinking on your part. I’ll check the return flights when I get off the phone.” Needless to say, Patrick flew back home a different way.

* * *

Patrick made it to his host family in Santiago, a woman with a teen-aged son at home. Pat soon found out that the woman never really cooked; all he could find in the refrigerator was an occasional pastel de choclo—a corn casserole with meat. Sometimes, he’d spot an empanada. The problem was, most of the food in the home contained wheat, cheese, and rather greasy meat—absolutely nothing that Patrick could eat. Pat would take to wandering the food stands after work, picking up items he could eat for breakfast or pack for lunch. Sometimes it was raw carrots or roasted chicken. Pat made do.
“Regional Metropolitana de Santiago de Chile.” The gold lettering across the front of the building was formal, impressive. I found out one phone call later that the treasury building sat near the presidential palace called La Moneda. The palace had been stormed when Augusto Pinochet took over Chile in a 1973 coup. Former president Salvador Allende was overthrown and civilian rule ended.

“I’d walk to the treasury building and see bullet holes in the side of the palace,” Pat told me during one phone call. “Legend has it those bullets were from Pinochet’s troops. It’s pretty cool to walk to work and see history.”

“Tell me what your internship is like.” It had only been three days, but I was so excited for this opportunity for Pat. I had never been to South America. We saved our dollars for Tee and Pat to travel and learn.

“Well, the Metropolitan Treasury is kind of like the U.S. IRS. I’m working in the tax collection section. I create legal files regarding tax collection and update records. The job is really kind of like glorified data entry. My boss is really cool, though. He’s an attorney and can speak some English.”

“Are the others in the office pretty friendly?”

“They don’t speak English. I remember some Spanish from high school, but not much. After work, I usually walk to a nearby bodega and get food. I’ve found shops where I can buy children’s books in Spanish. My favorite is My Name Is Gabriela. Well, it’s written for an eight-year old, but I can teach myself more Spanish that way at night.”

So, Pat was essentially reading Run, Tip, Run and working for the Chilean IRS. Not a bad gig. By the end of January, Pat could talk to fellow workers in the office. By the following
January, he had been invited again for a return internship. *Patrick era un tipo con suerte.*

*Patrick was a lucky guy.*
31 Five Steps Backward

During Pat’s senior year in college, I started getting phone calls of a different nature from him. He was noticing tingling in his extremities and all over his skin. Headaches. Brain fog. GI issues. Tiredness. Rashes. You name it. Yes, he’d been to his doctor in Newark. No, she couldn’t find anything wrong. Blood work normal. Blood pressure normal. No arrhythmia. Lungs were clear. Good oxygen intake. Pat got kicked down the line as a “whiner” to a GI specialist nearby. Meanwhile, Pat’s symptoms swelled and diminished, like a black tide.

Pat graduated from U Del cum laude, only .001 of a point away from Magma Cum Laude. As a mother, it was all I could do not to put a banner across the front of the house. In May, he came to his new home in Chapel Hill, North Carolina. Mark and I had moved there for retirement in 2013 when Pat became a senior. We loved New Jersey, but the property taxes made us choke. At any rate, Mr. U Del graduate took a class that summer and worked near the University of North Carolina campus at ‘Sup Dog, bussing tables and wiping up pools of beer. Patrick had also applied at Border’s Books, deeming that would be more “fun” for himself since he was an avid reader. However, he committed what I call a Career Limiting Move, or CLM. In other words, on the way to the interview, Pat was running late. So, what was his plan of action? Cut off that white Buick to his left and screech into a parking spot right in front of Borders. An issue arose during Pat’s interview when the interviewer recognized Pat as the cocky young man that had just cut her off as she, too, was trying to park. Needless to say, the interview was over quickly. Mr. Pat was relegated to ‘Sup Dog.

* * *

Shaw 189
One beautiful Carolina evening in June, Pat, Mark and I sat out on our tiny screened in porch, taking in the sixty-foot pine trees. We each had a goblet of Chardonnay that cast off a golden light—I remember holding the glass up and admiring the glint. Pat sipped a Diet Coke, his clothing still grubby from running the ‘Sup Dog dishwasher.

“I’m gonna have hot dog under my nails forever!” he moaned. “I’m losing my soul at ‘Sup Dog.”

Mark and I reminded Pat that we had both worked at small-time drug store cafes. We, too, had washed dishes, scraped off the blackened café grill, and made peanut hiccups. We had both walked two miles in the snow to get to work. Our tips were dimes and quarters. I had to wear a hairnet, looking like Ruth Buzzi’s character Gladys Ormphby. We had lost our souls working, too, but it was character-building.

Pat listened to Tough Times Parental Lecture 101 with an eyeroll or two. Parental lecture aside, the three of us hammered out a plan for fall. Pat jotted down the list:

--Study for the LSAT’s and attend Princeton Review classes
--Continue losing his soul at ‘Sup Dog
--Take another improv class or two
--Handle his laundry and wash dishes at Chez Shaw
--Help mom tutor at a nearby Latino community

Pat’s spring semester would be at Malaca Instituto in Malaga, Spain. Grandma B had sent Patrick a nice check for graduation, specifying that it had to be spent on education. Then there was Aunt Carol’s check, and Grandma Mae’s. We could do this. Spanish fluency and international relations? It did make sense as a next-step career plan.

So, the plan for life was on. Except it wasn’t.

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Shaw 190
As summer progressed into fall, Pat’s condition worsened, with headaches, nausea, and GI symptoms smacking him without reason. We wedged medical appointments between ‘Sup Dog and LSAT classes. We had Patrick’s internist mystified. In fact, we had specialists in neurology, infectious diseases, rheumatology, dermatology, and gastroenterology mystified. Pat was tested for Lyme disease, Epstein Barr virus, Mono, low vitamin D levels, Chronic Fatigue syndrome, Fibromyalgia, Sjogren’s Syndrome, Celiac disease, intestinal parasites, Helicobacter pylori infection of the stomach, mold exposure, and lupus. At the end of the massive testing, Patrick was labeled as an “anxious patient.” After four months, we had no answers. Just strange, apparently unrelated symptoms.

“Let’s go back to the University of Florida to see Dr. Weinstein,” I suggested one day. I was a tangle of anxiety. Some days, I’d come home from the grocery store and Pat would be stretched out on the couch, pale and sick. He’d struggle to go to work. Some evenings, he’d feel better, locking himself in his room with on-line LSAT practice tests. The symptoms would flare up and down, up and down, with no discernable pattern.

Pat agreed to the Florida trip.

Dr. Weinstein did a “tune-up” on Pat, measuring blood sugar and lactic acid for two days. I stepped out for many of the conversations with Patrick and Dr. Weinstein; after all, my son was twenty-two. I did learn later from my son that the two had talked a lot about law school. In between nurses and Dr. Weinstein, we’d go visit the young GSD patients on the floor. The parents’ eyes would light up when they’d see the 6’2” curly-headed hulk walk in the room. Pat represented hope to them. In a similar way, Pat’s face brightened when Dr. Nadia made her rounds, clipboard in hand and an ever-present smile on her face. She was the first GSD patient
to finish medical school and take her boards. Very simply, Dr. Nadia epitomized the future to Pat. He could dare to think about law school and beyond. Pat’s happiness grew even more after one particular conversation with Dr. Weinstein. “You will live to play with your grandchildren,” our saint assured Pat. “However, you have to manage your GSD health rigorously.”

As the next few weeks progressed, Pat adhered to the adjusted cornstarch schedule, measuring out Argo precisely to the gram. We watched our food for non-approved GSD items and Pat drank only water. Yet, the tiredness, foggy headedness, and GI symptoms continued. Something was wrong with Patrick.

* * *

It became very clear that Patrick wasn’t going to Spain for his language immersion. In between LSAT practice tests, he began researching online. It turns out that a malfunctioning GI system can create tiredness and foggy headedness, in addition to bloating, cramps, and diarrhea. One afternoon, Pat wandered out of his room, tapping his computer screen at me.

“The FODMAP diet, Mom.” Pat gestured to the screen. “It stands for “fermentable, oligosaccharides, disaccharides, monosaccharides, and polyols—all hard-to-digest sugar and fibers that pass through the small intestine without being properly broken down.”

“But why does that cause a problem?”

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“It says here, Mom, that when these FODMAP’s pass into the colon, they ferment and create gas. They also pull water into the gut, and thereby stretch the intestines. This causes bloating, pain, cramping, diarrhea, or constipation. Also, maybe bacterial overgrowth.”

Pat and I sat together, reading on. “The low FODMAP diet is a three-phase nutritional intervention. It includes an elimination phase, reintroduction phase, and personalization phase.”

“I’m going to print out the list of potential “danger” foods,” Pat continued. “Corn, garlic, onions, spinach . . . can you work with this? On top of the GSD ‘danger’ foods?”

We soon ate very differently as a family.

* * *

Over the next six weeks, junk food and fast food flew out the door of our home, as did pork, spinach, ramen noodles, butter and bacon—the list was vast. We ate “safe” foods: grilled steak with olive oil, salt, and pepper. Plain baked potatoes. Sautéd fresh kale. Steamed eggplant. Rice, sometimes, even though it was technically a FODMAP safe food. It appeared that limiting FODMAP foods was merely a floor, not a ceiling to the dietary changes Patrick needed to make to feel better. Patrick later learned that all meats had to be cooked fresh and eaten or frozen immediately, as histamines would build up quickly and disrupt the GI system. During this period of clean eating, Patrick also actively searched for help. He reasoned that

5 Ibid.

6 Ibid
there had to be a medical system that functioned similarly to the methodology of lawyers; in other words, payment by the hour. Pat was tired of physicians that pushed him out the door after his allotted fifteen minutes. During the search for help, Patrick ran across an article in The Wall Street Journal about a service called “Patient Navigator.” Essentially, it was a company that would employ a researcher for a person’s unique medical conditions. The researcher, an individual with a PhD in the biological sciences, would take that patient’s symptoms and investigate extensively, coming up with a short list of possible medical conditions within a few weeks. It was a phenomenal concept. Our family was desperate to put a name and a definition on Pat’s ailment. We knew him. Pat was anxious, of course, as the doctor’s charts indicated. However, he was anxious because he battled cramps, nausea, and dizziness for no apparent reason. He was anxious because he wanted to get on with life. He was anxious because his symptoms were indicative of neurological problems, autoimmune problems, or possibly infectious disease problems. Was he just sick, or really, permanently sick? Worse than GSD sick, at a mere age twenty-two?

“Mom, the deposit for the Patient Navigator research is $500.00. That money goes toward the hourly research fee of $50.00. After that, you have to pay an additional hourly rate. Can we try this? I don’t know what else to do, and I want to feel good again. I want to take the LSAT’s and go on to law school. The way I feel now, I can’t do much of anything. I’m scared I’ll spend the rest of my life lying on your couch.” Pat sat next to me, the pages of the Wall Street Journal scattered around him. He rubbed his eyes, hollowed-out and dull. My vital college grad was incapacitated, sitting on my couch, thirty pounds heavier than when he graduated. His life
was going south fast. Where had my perky, confident son gone? I felt as lost as that night many years ago, when Patrick cried and cried—and I couldn’t help him.

Mark got home from a church meeting that day and I nabbed him fast with the information on Patient Navigator. “What do you think?” I patted Mark’s arm impatiently. All my years of living with GSD ambiguity rolled right over me.

“We’ll make it work,” Mark reassured me. “We’re parents, aren’t we? I’m making a bit these days.”

Within a day, we had Patient Navigator on board. I didn’t want to get my hopes up too much. Patrick and I had seen so many experts over the past few months. Each one had her own paradigm, but no one had answers. “I can’t find anything wrong.” “Anxious.” “Let’s wait a couple of months and re-test.” “I’m going to refer you to ____ (fill in the blank.)” Pat began filling in detailed pages of his medical information and emailed it off to Patient Navigator.

Miracles do happen. Within three weeks, Pat received an email backed with pages of data from Dr. Willingran, one of the researchers. I was elbow-deep in meatloaf when Pat bounded in. I hadn’t seen any “bounding” from Pat in months, only an overweight, sluggish guy.

“This makes sense! I’ve been reading about this online!”

“What does the report say?” I smeared Sweet Baby Rae’s barbeque sauce on my apron anxiously.

“Mast cell activation syndrome. MCAS.”

I had never heard of mast cell activation syndrome, but I had never heard of glycogen storage disease a few years ago, either. Pat certainly had a knack for getting rare diseases.

“Mom: listen here.” Pat read quickly, his hands shaking.
MCAS occurs when the mast cells in your body release too much of the substances inside them at the wrong times. These mast cells are part of your immune system. They’re found in your bone marrow and around the blood vessels in your body. When you’re exposed to stress or danger, your mast cells respond by releasing substances called mediators. Mediators cause inflammation, which helps your body heal from an injury or infection. This same response happens during an allergic reaction. Your mast cells release mediators to remove the thing you’re allergic to. For example, if you’re allergic to pollen, your mast cells release a mediator called histamine, which makes you sneeze to get rid of the pollen. If you have MCAS, your mast cells release mediators too frequently and too often.  

I looked over Pat’s shoulder at the printout with a strange mix of anticipation and dread. What if? What if this new “thing” were like cancer? What if it were worse than GSD? Was this even fair for Pat to have TWO chronic health conditions? Strike two, I thought, mentally shaking my finger at God.

“Mom, right here it says that ‘too many mediators can cause symptoms in almost every system in your body. However, the most commonly affected areas include your skin, nervous system, heart, and GI tract. Depending on how many mediators are released, your symptoms can range from mild to life-threatening.’” Pat’s finger scanned down the document rapidly. He wanted, so wanted, to get a diagnosis. The medical unknown was growing seeds of fear in him.

“Here. Look right here. Itching skin. Runny nose. Swelling of the tongue. Trouble breathing. Low blood pressure. Mom, remember that time when I almost passed out in the bathroom? GI stuff here. Cramping, diarrhea, nausea. Nervous system also, mom. Headaches, dizziness, confusion, extreme tiredness. Oh, my God. These are a lot of the symptoms that have been hitting me.

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8 Ibid.
They’re all over my body. No wonder each specialist couldn’t figure out the whole picture. They just thought I was a nut case.”

Pat and I sat at the kitchen table, reading page after page from Dr. Wellingran. “Let’s look at some of the triggers,” I suggested. “Maybe that plus the FODMAP diet will help you to feel better over time.” We scanned down the literature. There it was—our list of triggers to consider:

--allergic-type triggers, such as insect bites or certain foods
--drug-induced triggers, such as antibiotics, ibuprofen, and opiate pain relievers
--stress-related triggers, such as anxiety, pain, rapid temperature changes, exercise, being overly tired, or an infection
--smells, such as perfume
--hormonal changes, such as those related to a woman’s menstrual cycle

Pat began the lengthy process of identifying lifestyle triggers as well as dietary.

“You know, mom, when I take a hot shower, I feel kind of sick afterwards. Also, being out in the sun. And after exercise.” Pat’s computer was open, and he pecked away on a new spreadsheet. I glanced at the columns. “Patrick Shaw. Symptom. Possible Cause. Time of day. Date. Comments.” Pat was on it.

“So, we can’t really identify troubling foods, yet?” I continued to hover, hopeful with the new information.

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9 Ibid.
“Nope. I have to eat super clean for the next six weeks. Then, I can add one potential trigger food back in at a time. See here. I’ll add those foods in on the spreadsheet when it’s time.”

“Look down here, Pat. Ways to determine if you truly have MCAS.”

--Symptoms affect at least two body systems and are recurrent, and there’s no other condition causing them.
--Blood or urine tests performed during an episode show you have higher levels of markers for mediators than you do when you aren’t having an episode.
--Medications that block the effects of the mast cell mediators or their release make your symptoms go away.\(^{10}\)

Pat and I kept skimming the data for how to control MCAS. “Here it is,” Pat pointed.

“Thank God. Not just food control. If I can find a knowledgeable doc, here’s some stuff I could try.”

--H1 or H2 antihistamines. These block the effects of histamines, which are one of the main mediators released by mast cells.

--Mast cell stabilizers. These prevent the release of mediators from mast cells.

--Antileukotrienes. These block the effects of leukotrienes, another common type of mediator.\(^{11}\)

“Wow. This is easier said than done, right?” I tapped my finger on item one. “Figuring out if no other condition is causing your headaches or your GI issues? Isn’t that what we’ve been investigating the past six months?”

“Yeah. Let me type in down here what all systems I’ve had tested so far.”

\(^{10}\)Ibid.

\(^{11}\)Ibid.
The list was *long*. Pat and I began Googling for doctors familiar with MCAS. That list was *short*. Turns out, MCAS was just as rare as glycogen storage disease, if not more.
In the midst of symptom-tracking and eating cleanly, Patrick had finished his Princeton Review class and had taken multiple practice LSAT exams.

“It’s showtime,” Pat announced one day as I folded laundry. “Wish me luck!”

I gave Pat a motherly hug. “I’m so proud of you, no matter how the LSAT comes out. You’ve been working at it so hard, and I know you’ve felt crappy for months. Don’t know how you do it—you’re just an amazing guy.” Pat’s hands felt cold and clammy; I could feel the nervousness flowing from him.

Pat arrived home a few hours later, his eyes bloodshot. He had worked with LSAT officials ahead of time so that he could eat a snack and do his cornstarch during the testing break after section three. He just knew he couldn’t make it through the entire testing. Mental energy actually lowered his blood sugar, much like physical exercise.

“Oh, my God,” was all he said as he sat down at the kitchen bar after the test. After a brief lunch, Pat zonked out in his room, coming to three hours later.

* * *

About four months later, it came. The envelope. I was fairly tap dancing in the kitchen with anticipation.

Pat ripped the envelope open, breaking into a beautiful smile—something we hadn’t seen for a while. “Oh, man,” he breathed. “These scores will get me into quite a few schools. Well, not Harvard. And I’m a point shy of qualifying for Duke.”

Mark rounded the corner with groceries just as I smarter off to Pat. “Well, you slacker! I guess we’ll keep you.” Mark, Pat and I had the best family hug you’ve ever seen. I grabbed onto
Pat, thinking of all the times he was in the hospital, or all the times he couldn’t eat pizza at a birthday party, or . . . or. It had come. A break.

Pat applied to several law schools, much as he had done for undergrad: a “stretch” or two, moderately hard, and several “easy” ones, with the idea of “easy” rather relative. In a few weeks, he heard back about two free rides and a couple of reduced tuitions. “The one I really want . . .” Pat pecked away at his symptom spreadsheet. “Uh, corn: no. Broccoli: no. Any meat with nitrates: no. Carrots: no. Sautéed kale: yes. Potato chips cooked in coconut oil: yes. Okay. What I really want is Georgetown Law Center. I could make some great connections there—and the professors! They’re amazing. I’m scared to even dream it, but some day, I’d love to work for the Department of Justice. It’s a big stretch, though.” Dr. Ripton’s presence from Pat’s high school days was alive and well, nudging my son’s brain. Pat knew he couldn’t change the world, but maybe reverse an unfair ruling? Maybe stop an anti-trust violation?

Pat got wait-listed at Georgetown. Somewhat hopeful, he and I combed through the admission data. “Look here.” I pointed at the information. “Says you can appeal in writing and write a supplemental essay. What do you think?”

“I have to try. I just have to try.” Pat wandered off to his room. He had just cooked himself some scrambled eggs, and turns out, they were on the “potentially evil” list. Patients with MCAS can have bad reactions to a number of foods, and often there’s no rhyme or reason to what they can handle. The only way to find out is by trying a bit of the food and waiting to see how the body reacts. So, with eggs, Pat felt nauseous and exhausted within five minutes. Later, however, I heard insistent typing. Pat had logged in his egg reaction and then began hammering away on his Georgetown essay. Within three days, he had appealed his admission.
Two weeks later, Pat got another letter from Georgetown. The response lay on the kitchen counter, hopeful yet ominous. Pat circled the envelope like a stray dog eyeing a fresh bone. Finally, he tore into it, while I held my breath. “Come on, God,” I prayed, trying to make a bargain with the Almighty. “Just let Pat go for it. Give him a chance!”

Pat got to “go for it.” His wait-listed status at Georgetown became a full admission.

* * *

For the second time in his life, Pat prepared to head to school.

One day, he came out of his room smiling. “Guess who I’m going to live with at school? Karthik!” I had forgotten that Karthik was studying at George Washington Medical School. His years of Saturday morning studying had paid off. I was sure that his mom and dad were busy now, working on the daughter, Anupama. I smiled, thinking of the little Anu who would hang around my kitchen cabinets, mooching Oreo cookies. “Karthik has an apartment with another guy in the Foggy Bottom area of D.C. I can take the metro over to the law school every day.”

*Perfect*, I thought. *Living with a friend during law school would be comforting. And, a future doctor, no less.*

Late that summer, Pat enrolled for classes. He was assigned to “Section A” of the L1 year, so the first year’s classes would include Civil Procedure, Constitutional Law, Criminal Justice, Legal Practice: Writing and Analysis, Property, Torts, and International Law. We had gotten a partial student loan for tuition; Mark and I felt strongly that Pat would feel plenty of skin in the game if he, too, were paying tuition. Plus, it was a lot of money going out the door for us as a family.
The summer before law school should have been one of joy. Instead, it was a summer of horrible uncertainty.

As the end of August loomed, Pat was a sick as ever, despite his careful diet. He had been to about twenty doctors during the past year, but still had no real answers. Dr. Willingran’s research indicated MCAS, but what did you do with that kind of diagnosis? What if Pat got to Washington, D.C.—six hours from us—and became even more ill? How could he manage the rigors of law school? On and off. On and off. Pat struggled with the decision to start law school.

Pat decided to push forward into life. On a muggy day in late August, the three of us loaded up the SUV with clothes, a new comforter, toiletries, and cornstarch. Lots of cornstarch and a big bag of empty plastic containers and lids. Fortunately, the small bedroom Pat would use was furnished with a desk, a bookcase, one lamp, and a bed. The kitchen had pots, pans, and dishes. All the rooms were amply furnished with mice. Since the apartment was right next door to George Washington University hospital, it shook with blaring sirens day and night.

All that said, it was an easy move-in. As we helped Patrick unpack, I fussed with Pat’s computer and pencil cup, true to my nature as a mother/worrier. I felt anxious, like the time we moved Pat into camp at Bryn Mawr ten years ago. This time, the stakes were high.

Back home, I prayed daily for Pat to make it through the craziness of law school. The kids at Georgetown were bright and go-getters. I remember many times Mark talking of his law school experience, stating, “It was terrible. It was the armpit of my life back then.” Patrick would go through that same awful “armpit period,” I was afraid. Only he wasn’t a typical, healthy twenty-three year-old.
About a week into school, I got a call from Patrick that fluctuated between panic and a rant. “Oh, my God, Mom. On the first day of my Civil Procedures class, Professor Halton had us get out a piece of blank paper and write an ‘original writ.’ We all just sat there and looked at each other. What the heck is an ‘original writ’? After about four minutes of all one hundred of us sitting with blank looks and even blanker paper, the professor said: ‘I want you to remember how you feel right now. You don’t know the answer to the question. In fact, you’ll never know answers to the questions your clients bring you. It’s going to be this way your whole career. It is up to you, and you alone, to research and battle with the issues.’ Mom, I felt so stupid. What have I gotten myself into?”

“Sounds like everyone was stumped. And you’re in school with a bunch of bright kids.”

“And then!” [I heard a long, shaky breath]. “In Constitutional Law, well, thank God I had read the assignment and briefed the cases. Remember The Paper Chase? Our professor was like Charles Kingsford grilling James Hart. One kid in the second row hadn’t read his cases and tried to fake an answer. The professor had this guy in knots and tears in three minutes. I’m just petrified I’ll get called on.”

“But you, Pat, will have read and analyzed your cases. I know you.”

“Yeah, I’m working like sixteen hours a day. But, if I accidentally get any food in me that my body doesn’t like, I feel like crap for hours. I’m trying to analyze these cases sometimes and my head just hurts so bad. Or, I get Montezuma’s revenge.”

“Keep writing down your foods and your symptoms, Pat. You, more than any of the doctors, can figure this out, because you know you.” I motioned for Mark to come talk with Pat. “He’s panicked about analyzing all the cases,” I whispered.
In the next thirty minutes, I heard Mark talk Pat “off the ledge.” In his beautiful, calm manner, Mark recounted the times he, too, had been panicked in law school. “You’ll get faster and faster in combing through a case for pertinent details,” he reassured Pat. “It will go twice as fast in about two weeks. Stay with it. You can do this! We are just so proud of you and how hard you are working. This is a steady marathon, not a sprint.”

Off of the figurative ledge, Pat gulped a dose of cornstarch, ate a few saltines, and dove into Marbury v. Madison. I resolved to research doctors throughout the U.S. that worked extensively with MCAS patients. Merely watching trigger foods was not cutting it for Pat. He was sick yet attempting to study sixteen hours a day.

Only later did I find out how horrific year one of law school was.

Pat would take the metro to Georgetown law school each morning with his heavy pack of law books, snacks, and cornstarch. After he’d exit the metro, he’d walk an additional six blocks to McDonough Hall for his first class. It would take him about forty minutes of class time to shake off the nausea and exhaustion from the walk to class. After a full day on campus, he’d get back to the apartment in Foggy Bottom, sick again.

In the evenings, Pat would pour over his cases and briefs, making comprehensive outlines. However, unlike the other law students, Pat would study for forty minutes, then lie down for fifteen; study forty, lie down again. It was the only way he could make it through school.

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Back in North Carolina, I sighed after a particularly heart-wrenching conversation with Patrick. The next day, I researched and Googled all afternoon, trying to find a specialist familiar
with MCAS. Just as GSD had been twenty years earlier, MCAS was an unknown. For years, patients lived with this terrible disorder, yet often became horribly dysfunctional over time or died from anaphylactic shock. Some patients had to go on full disability, unable to even leave their homes. Additionally, because they complained of multi-system ailments to their physicians, many became labeled as “whiners and anxious.” Management of the condition eventually fell into two realms: H1 or H2 antihistamines that could block the effects of histamines, a type of mediator released by mast cells. Or, some medications, called mast cell stabilizers, could prevent the release of mediators from mast cells in the first place. Pat was attempting to prevent the release of mediators by eating correctly. It was just not enough.

The next day, I wailed about this situation to my internist during my annual checkup. Dr. Powell had probed a bit when she saw my blood pressure. “What’s going on in your life?” She looked concerned, listening as though she had all day. Quietly, she wrote down five important words for me on a Post-It note: “Lawrence Afrin, M.D.: MCAS specialist.”

I called Pat that night amid his pile of casework, telling him about the expert in MCAS. “Dr. Afrin practices out of Armonk, New York. Maybe after your December break we can both fly to White Plains for an initial consult.” I agreed to do some legwork to get an appointment set up.

I worked hard during October to get a January appointment with Dr. Afrin. Pat was not a minor, so that was roadblock one. Also, he was in D.C., knee-deep in study outlines and a study group when he wasn’t in class. Dr. Afrin was in New York and tied up with countless patients. I felt like I couldn’t talk to anyone. As it turned out, too, Pat had to “prove” that he had one of the indicators of MCAS. Just feeling horrible most of the time was not enough.
“Remember about a year ago when you had a colonoscopy and an endoscopy? It was when we were trying to figure out what was wrong with you. Well, Dr. Afrin will accept data from your biopsies. Apparently, the cell blocks from the biopsies can be stained, so mast cells can be detected. I’ve been doing a bit of reading. There is some controversy about the number of mast cells that are needed to make the diagnosis. Experts in the field often try to detect chemical evidence of the disease to support abnormal biopsies from the gastrointestinal tract. That’s why Dr. Afrin will do blood work and urinalysis at your first visit. Anyway, Pat, can you make some calls and get this sent to him? I can’t do this because of HIPAA, of course.”

Pat got the data sent. Would Dr. Afrin accept Pat as a case study?
The wind whipped around the White Plains airport, rattling the windows with icy blasts. I had lived in “North Cackalackie” long enough to grab a parka and earmuffs when the temperature dropped to fifty. That day in White Plains, it was a bracing fourteen degrees. As I sat in the terminal waiting area, I felt excited to meet up with Pat and perhaps get help. Pat had made it through his first semester of law school, declaring that the final exams were brutal. He had slept for two days off and on after he finished, then took the train to Chapel Hill for three days of Christmas break. He had gone back to D.C. early to buy textbooks and begin reading for the second semester classes.

Our time with Dr. Afrin was helpful, despite the chilling blasts that shook our southern souls. The specialist had reviewed the colonoscopy biopsies, ordered massive blood work as well as a twenty-four-hour urine collection. Dr. Afrin explained that laboratory tests looking for evidence of MCAS included: 1) serum chromogranin A and tryptase and plasma heparin, histamine and prostaglandin; and 2) 24-hour urine collections for N-methylhistamine, 2,3 dinor 11-beta-PGF2-apha, and leukotriene E4.¹ The specialist also reassured Pat that MCAS could reveal itself in many, random symptoms, including throat pain, prostatitis, chronic pelvic pain, non-bacterial cystitis, general sensitivity to pain, diarrhea, nausea, asthma, cognitive fogginess, or anxiety. After three days, Pat flew back to D.C., hopeful and waiting on test results. “There are a lot of alternatives we can try,” Dr. Afrin had reassured Pat. “It’s different with each

person, and you have to work in one medication at a time, just like you would a new food. Otherwise, you have no clue what is actually helping you. It’s trial and error for each person.” In other words, MCAS had no quick fix, just management. Just like GSD.

It wasn’t until Pat’s second year of law school that he began medications to truly manage MCAS. While he continued Face Time calls with Dr. Afrin, he also discovered an allergist only a block away from his apartment, very close to the George Washington University hospital. The young woman had worked with several MCAS patients. Most importantly, this specialist was one of the first physicians ever to acknowledge Pat’s personal knowledge of his conditions. Between the two specialists, Pat began to painstakingly test several medications, one by one, while monitoring his FODMAP diet.

Both of Pat’s new doctors recommended that he start on a combination of H1 and H2 blockers—to “wall off” mast cell mediators. According to the OpenAnesthesia website, H1 antihistamines are mostly used to treat allergic reactions and mast cell-mediated disorders. H2 antihistamines are indicated primarily for gastric reflux disease because they reduce the production of stomach acid by blocking the H2 histamine receptors in the parietal cells of the gastric mucosa. Because Pat experienced system-wide symptoms, he would be taking both H1 and H2 blockers.

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2 OpenAnesthesia, accessed at https://www.openanesthesia.org/h2-blockers_onset_time/#:~:text=H1%20receptor%20antagonists%20are%20typically,to%20suppress%20gastric%20acid%20secretion%20on%206/10/20.
For H1 blockers, patients often take Zyrtec (Cetirizane), Claritin (Loratadine), or Allegra (Fexofenadine). Common H2 blockers include Zantac (Ranitidine), Pepcid AC (Famotidine), Tagamet (Cimetidine), or Axid (Nizatadine). At the present time, the FDA has banned both Zantac and Ranitidine for possible carcinogenic ingredients. On the other hand, to stabilize mast cells, specialists often recommend oral Cromolyn, mixed with water about four times daily before meals. This medication targets the GI tract in particular. Cromolyn also comes in a nebulizer form. Oral Ketotifen is an alternative to Cromolyn and can be purchased at compounding pharmacies. Many times, patients take both medications. Still another medication, Quercetin (Neuroprotek) may have a mast cell stabilizing effect in some patients.

Most MSAC patients find that they feel better using compounded prescriptions, as sometimes the inert ingredients or dyes can cause allergic reactions. These compounds are expensive and not always covered by insurance.

Other recommendations exist for the condition. MCAS patients must avoid certain drugs that trigger mast cell release, including narcotics, muscle relaxants, certain antibiotics, anti-seizure drugs, local anesthetics, IV dye, and ACE inhibitors. To promote overall health, MCAS patients may often take probiotic therapy that includes Lactobacillus rhamnosus (Culturelle) and Bifidobacter species, according to Dr. Afrin.

Pat began taking 30 milligrams of levocetirizine daily. This over-the-counter medication, commonly known as Xyzal, was an antihistamine working to block Pat’s reactions to hot showers, perfume, certain lotions and shampoos, and bright sunlight. The second prescription

3 Wong, Henry. Medscape. 5/21/18.

4 Ibid.
Pat began was 2 milligrams of oral Kototifin daily. This drug is often used for the treatment of asthma, food allergies, anaphylaxis, irritable bowel syndrome, and general MCAS issues. Pat also initiated 600 milligrams of gabapentin daily, as well as oral cromolin sodium. Although gabapentin has no direct impact on managing MCAS, it can help with migraines and sensory overload—secondary symptoms of MCAS.

I knew what Pat could eat because I had cooked for him during his summer breaks. I can tell you, it wasn’t much. And, the trigger foods at first glance appeared random. For example, Pat would react to peanuts, but not Brazil nuts. Salt and pepper were okay, but not rosemary or chives. He could eat sautéed kale, but not spinach. Fresh sea bass was okay, but not salmon. All meats had to be cooked and eaten immediately or frozen to avoid histamine build-up.

* * *

After Pat’s visit with Dr. Afrin and his local allergist, he continued testing various foods for reactions and worked in one medication at a time, according to his physicians’ advice. Sundays became Pat’s day to buy groceries for the week. He’d walk everything back to his apartment and begin cooking—a two-hour production. All food would be frozen immediately in small zip-lock bags. The apartment freezer was bulging with baggies of frozen, cooked ground turkey (absent rosemary seasoning). Other baggies contained frozen, steamed kale, cooked eggplant, or new potatoes. The cabinets were full of potato chips cooked in coconut oil, Brazil nuts, dried kelp, cornstarch, plastic cornstarch containers, Citrucel, and probiotics. It looked like a science lab. During the busy week, Pat would microwave a bit of food for breakfast and then pack lunch into a thermal pouch filled with four ice blocks. The lunch, along with the day’s measured cornstarch and medication, all fit into Pat’s backpack, along with three or four law
books and spiral notebooks. During the thirty-minute commute to Georgetown Law, Pat lugged the heavy pack.

Pat’s second semester of year two went better. Sometimes he’d have several days at a time when he felt good. Occasionally, if he tried a new probiotic, Pat would experience two days of nausea and exhaustion. Unfortunately, the crush of law school study never stopped; not for Saturdays and Sundays; not for GSD; and certainly not for MCAS. I’m not sure where it was that Pat reached for strength and determination to keep studying. He had only what I can describe as “grit”; a blinding desire to finish law school and to do so *cum laude.*

After writing about fifty application letters, Pat obtained a summer unpaid internship with Judge Calabria with the North Carolina Court of Appeals in Raleigh. He planned to live with us and drive my car, since his old clunker had been sold when he went off to law school. There were few parking spots in D.C.; besides, we needed the money to pay for all the law books. I volunteered to cook for him that summer, because with the hour commute twice daily, he’d be gone ten hours a day.

It was during this internship that Pat experienced the feeling of actually practicing law. He researched cases for Judge Calabria eight hours a day and loved it. Our standard family joke ran like this:

Mom: “So, what case are you working on now? Anything interesting?”

Pat: “I’m loving the research! Well, I could tell you about the case, but then I’d have to kill you. It’s super sensitive.”

Mom (with Joisey accent and hands on hips): “You talkin’ to me???? You got a problem wid dat????”
At this point in Pat’s life, he had the management of his GSD and MCAS fairly well worked out. That summer, our kitchen looked different. We had a clean towel on the counter with a large, covered bowl of cornstarch and six clean plastic containers, resting upside down. We had a gram scale out. Pat’s string pack lay nearby. Levocetirizine, Ketotifen, cromolyn, and ranitidine were lined up on the counter. Inside the pantry were piles of new potatoes, blocks of dried kelp, boxes of Argo cornstarch, baggies of Brazil nuts, and Jackson’s Honest Chips. Our freezer was packed with pouches of cooked turkey, eggplant, and kale.

Working for Judge Calabria was definitely not ‘Sup Dog. Pat had to wear a dress shirt each day. A backpack just didn’t quite look right in this new job. But what to do? Before the internship started, Pat obsessed about appearing “normal” in the workaday world. What would the other clerks think when he had to microwave his frozen, crumbled turkey and eggplant or shake up mysterious white powder in water? What if he was having a bad MCAS day and had to run to the restroom six times? How would he transport his frozen lunch and cornstarch bottles to work each day discretely?

“What if there’s a special end-of-summer lunch for us interns? What if people want to go out after work to a bar? What am I going to do?” Somehow, Pat had gotten through most of childhood with only moderate anxiety about his conditions. However, he was stressing now. I think that he really wanted to make a go of a career. Would being ill impact his hiring chances?

We got on Amazon and searched for leather satchels. That one, there—the dignified oxblood leather one. It would look better than the beaten-up backpack from law school. Hmmm. Could Pat cram in his refrigerated lunch carrier, plastic blocks of ice, four cornstarch containers, and a water bottle? Looked like it could work.
The summer blew by, and in late August, Pat was back in D.C. for his third and final year of law school. It went well, for a while. Pat had decided to restart the defunct anti-trust club at school, recruiting new members and hosting informational seminars for fellow students. Various law professors were normally good about presenting at the seminars. The anti-trust club would have free pizza afterwards, with Pat nervously mingling about, Brazil nuts in hand.

Mid-fall, Pat began complaining to me about various foods that set off nausea and exhaustion. Chicken and rice, former staples, suddenly became off limits. Why? Then, corn really began to bother Pat. That normally would not have been a deal-breaker, but Argo cornstarch was... **corn**. Cornstarch and food were what kept Pat alive. Without them, Pat’s blood sugar would plummet to thirty within a few hours. He’d be dead in less than twenty-four hours or hooked up to an IV in a hospital. It was a high stakes problem. **Really?** I questioned God. **You let Pat get to law school, have him do well, and now this? Really? Have you gone on a coffee break?** My spirituality wavered. I was just angry.

That semester, I could feel Pat’s anxiety simply vibrating whenever he’d call. The year he’d lived with us, his weight had increased to 245 pounds—a bit heavy, even for a 6’2” fellow. With the new food allergies emerging, Pat could eat less and less. Soon, he was down to about ten foods and weighed 185 pounds. When he’d do his cornstarch dose—every three hours—he’d feel sick for two hours afterwards. Yet, he had to take his cornstarch to keep his blood sugar up. On top of this, Pat was studying like a fiend. He wanted to get all A’s his 3L year, to boost his GPA enough to make **cum laude**. The situation was misery personified as Pat dragged through each day. How would he do in law school? Unknown. Would he shrink to eating four foods? Unknown.
I began digging into research again, trying to keep Pat relatively free to begin his outlines for the December final exams. I turned desperately to Dr. Weinstein for help, setting up a conference call between the specialist and Pat.

“Honestly, I’ve never had a GSD patient who suffers from mast cell issues,” Dr. Weinstein had told Pat on the call.

“Did you get any guidance at all?” What were we going to do without Argo? I picked at a cuticle anxiously, making myself bleed. Here we are again, in totally uncharted territory. How long, God, are we going to do this?

“Well, Dr. Weinstein did say I could try Authentic Foods Tapioca Flour instead of cornstarch. Apparently, it will keep my blood sugar up as well as Argo. He said there are other flours, too, like almond, amaranth, barley, chickpea, and rice flours. They don’t work as well for sugar control. So, I guess I’ll begin working in tapioca flour gradually tomorrow. See what happens. Mom, what if it doesn’t work?”

My mind couldn’t go to the “what if.” “Sounds like a plan,” I reassured Pat. A bit of tension seeped out of me. “I’ll dig around on the MCAS website—see what I can find about food problems. Other people must be going through these same issues.”

“Yeah, and some of those people are incapacitated. They can’t even leave their apartments. They can’t work.”

“Hang on. Just hang on. Let me research. Hey, I love you thiiiiiiiis much!” I started digging into information fast, my gut churning with anxiety.

* * *

Shaw 215
Patrick actually spent two hours of poking around the MCAS website, somehow packing this search into the incredible workload. He found a name. A name of hope, perhaps.

Lucinda Williams was a registered dietician who worked solely with mast cell patients. She used tried-and-true techniques to help her patients work problem foods back in. That was what Pat needed desperately. He set up a conference call with Ms. Williams the following week.

“You won’t believe this, Mom.” I could hear beeping in the background. Apparently, Pat was microwaving dinner—turkey, new potatoes, and kale—again. “So, it’s a rotational thing. The first two weeks, I pick out a food I really want to work back into my diet. I think I might start with chicken. So, I’ll eat like one little sliver of cooked chicken on Monday. Wait a day. On Wednesday, two little slivers. You keep on like that, increasing just a bit on alternate days, until you can tolerate a full serving. Ms. Williams says that at first, I might feel a bit sick when I test. Well, then I work in the next food. I’m gonna try rice, then green beans.”

I was amazed. My very own grandfather had eaten a rotational diet in the early 1900’s. I never knew much about the venerable attorney that lived in Pittsburg, Kansas, other than he had asthma and food allergies. Huh. Maybe I had passed down the mast cell problem along with GSD.

Pat was able to work in chicken, rice, and green salads over the next few months. But not green beans. With only one bean, he felt sick in minutes. But, over time, food introduction worked. Pat went from eating only ten foods to twenty. It was a miracle. It was a testament to eating carefully and tracking all foods, week after week.

* * *

Shaw 216
The end of Pat’s final year of law school was a blur of cases, outlines, study groups, and grueling final exams. It was a blur of tapioca starch, FODMAP rules, levocetirizine, and Ketotifen.

It was mid-May when Steve, Mark and I arrived in Washington. It was hot that day. It was a Washington, D.C. sticky hot. It didn’t matter. Pat was graduating from law school.

We sat in makeshift wooden camp chairs, surrounded by four hundred other parents, grandparents, and assorted screaming children. The wooden slats of the chairs pleated our hindquarters as sweat trickled down our backs and in front of our ears.

I fanned myself with the Georgetown Law program and perched on the tip of my chair, hoping to see the students better. It was a sea of twenty-something’s out there, all dressed in heavy black robes and eight-sided tams. Each graduate sported a purple cord on the tam and a heavy purple stole lying over the black robe.

Two o’clock approached. We could all feel expectation build and crest, as grandparents and parents fanned and waited, fanned and waited. I looked for my son. Not that row—maybe the next? No. Shaw. Maybe in the back.

I scanned the young law school students as they chatted with each other in anticipation. And then—I found my son. His eyes spotted mine across the sea of black and purple. Blazing blue eyes behind navy glasses. Dark curls flipping over the tam. Standing tall above the others, he flashed me a huge grin and a thumbs up.

A single chime echoed across Healy Lawn from the bell tower on campus. The students and law professors stood in a sudden whoosh, while cadets marched the U.S. flag and Georgetown flag to the back of the stage, slowly, sedately, to Pachelbel’s Canon in D. I heard name after name. Student after student. And then—Patrick Shaw: juris doctor, cum laude.
I grabbed Mark’s hand and then Steve’s. Thank you, God. Just--thank you.

* * *

Today, Pat works for a corporate law firm based in D.C. He handles litigation upon occasion, but also works on multi-billion dollar mergers. Attorneys on the team must ensure that the buyout doesn’t create anti-trust issues. In his spare time, which is minimal, he plays the drums and watches improv. And—I hear something about a lady named Samantha.

Pat calls about every two weeks to touch base. Whenever I ask him about his current work with the huge life sciences company, I get this reply:

“Well, mom, I could tell you, but then I’d have to kill you.”

Oh, Patrick. Forgeddaboutit.
34 Un-asked for Advice

It seems easy to look back on the past twenty-eight years and pontificate. Yes; we did this and investigated that. We researched this and tried that. The reality is: the Shaw family bumbled through Pat’s medical conditions quite a bit. However, amidst the confusion and map-less journey, we did manage to learn. Patrick has grown up to be a fully functioning adult in a career he loves. He grew up this way perhaps because of us, or perhaps in spite of us. So, what’s the “so what” of my learnings?

--When you learn you have a child with a chronic disability or illness, make a comprehensive list of everyone you know that could help you—either emotionally or practically.
--Relish any and all improvements you see in your child. It may be finally getting that smile from him or seeing him eat on his own.
--Share setbacks and frustrations with trusted family members or friends.
--Research, research, research! Physicians and specialists are wonderful, but you know your child the best. You are also the person who will push the hardest for him. Specialists tend to have a particular paradigm—they are, after all, human. You will need to cross paradigms all the time.
--Teach your child about his condition by involving him in his daily care.
--As best as you can, have your child assume family responsibilities.
--Don’t assume your child can’t perform a task. He’ll often surprise you.
--Set small, achievable goals of independence for your child. Look for new opportunities for him to “stretch.” Baby steps can become adult steps, but it may take months or years.
--Document your child’s condition, ER instructions, and medical schedule. Have a power of attorney ready at all times. Make sure all medical supplies are available. Redundancy is always good.
--Ask for help and be kind to yourself. Check with trusted friends or relatives so you can have a day off. Perhaps take your other children out for a treat. You’re in a long marathon, not a sprint.
--Celebrate how hard your child is working, not the actual end event.
--Listen to your child. What is he feeling and fearing? Talk with him.
--Join a group of others who have the same condition. Share knowledge, pain, and frustration. It feels good to know you’re not out there alone. Try to . . .

Whoops. I have to go. Patrick’s on the phone . . .

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EPILOGUE

THE WALK TO HOPE
Raising a child with a chronic condition involves a lot of steps—steps forward to new goals, and steps backward as adversity rises. The journey is never stagnant, because a chronic illness is just that--chronic.

While research in MCAS is still in its infancy, Glycogen Storage Disease Type 1 has been a recognized and defined condition since 1929. Strides toward managing the condition evolved in the early 1970’s with the discovery of Argo cornstarch as a blood sugar management tool. Additionally, while Patrick had to be definitively diagnosed with GSD1a via two liver biopsies in 1993, the process is easier today. According to Rarediseases.org,

GSD type I is diagnosed by laboratory tests that indicate abnormal levels of glucose, lactate, uric acid, triglycerides and cholesterol. Molecular genetic testing for the G6PC and SLC37A4 genes is available to confirm a diagnosis. Molecular genetic testing can also be used for carrier testing and prenatal diagnosis. A liver biopsy can also be used to prove specific enzyme deficiency for GSD Ia.¹

What is on the research horizon today for GSD1a? In a word, gene therapy. The UltraGenyx company has developed a therapy called DTX401—an “investigational AAV gene therapy being developed for the potential treatment for patients with Glycogen Storage Disease Type 1a. DTX 401 is designed to address the defective gene for the

enzyme glucose-6-phosphatase-α, a defect that results in the inability to regulate blood sugar (glucose).”

The path to the UltraGenyx gene therapy for humans has been slow and tedious. As the website CureGSD describes, in 2007, the University of Florida research team began working with Maltese dogs, the only canine species in which GSD occurs naturally. Maltese dogs affected by GSD1a normally die within hours of their birth; even with medical treatment (cornstarch), the dogs typically die within four weeks. In 2007, Dulce, a Maltese with GSD1a, received the first experimental gene therapy. CureGSD reports that

Within 2 weeks, the duration of fasting markedly increased, and visually the dog improved with decreased hepatomegaly, increased energy, and improved growth. The effect of gene therapy waned, so a second gene therapy was performed in January 2008. Following this treatment on Dulce, the lactate concentration normalized, and she was able to fast for 9 hours without development of hypoglycemia. All glucose support was stopped at 6 months of age, and Dulce had no problems clinically while weaning off therapy.³

Researchers performed a liver biopsy on Dulce after the second gene therapy and determined that her liver was demonstrating 7% activity in converting glycogen to glucose. The website reports that twenty-one months after the second gene treatment, Dulce went into heat and as a result, developed increased lactic acid. Dulce then went onto low dose glucose therapy; she

³ CureGSD, accessed 10/9/18.
received her third gene therapy in 2010. Researchers report that at four years of age, Dulce has no “adenomas or evidence of complications.”  

Dulce later gave birth to Tucker and Jasmine in 2010, with each puppy receiving gene therapy “...using a modified vector.”  

At two weeks of age, both Tucker and Jasmine could fast for six hours; glucose support was discontinued at three weeks. A second gene therapy treatment was performed when each pup was eight weeks of age. At the time of the website writing, both were doing well.

* * *

I was amazed when I talked with Dr. Weinstein nearly four years ago that the gene therapy dosing would move directly from Maltese dogs to humans.

In typical pharmaceutical drug testing, animals are dosed with a new drug, with higher and higher levels of the drug administered. Then, human testing (clinical trials) incorporates a four-phase process.

In typical human FDA testing, Phase One represents a small group of young, healthy males that receive dosing of a new drug. Phase Two consists of a small group of individuals that have the targeted disease. Phase Three of the study involves hundreds/thousands of people with the disease. And finally, Phase Four occurs after FDA approval of the new drug. In this phase the drug is tested for other applications.

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4 Ibid.

After months and months of searching, Dr. Weinstein and fellow researchers at the NIH entered into an agreement with UltraGenyx, a biopharmaceutical company focused on the development of novel products for rare and ultra-rare diseases. The company announced that on July 26, 2018, the first human patient was dosed “in the Phase 1/2 study of DTX401, an adeno-associated virus vector based on gene therapy for the treatment of patients with GSD1a. This study began under the clinical trials government identifier NCT03517085.” 6 UltraGenyx states that the DTX401 “has been shown in preclinical studies to improve G6Pase-a activity and reduce hepatic glycogen levels, a well-described biomarker of disease progression.” 7 Also important is that the FDA has granted a Fast Track designation to DTX401 for the treatment of GSD1a. According to Globenewswire, “The FDA Fast Track program is designed to facilitate the development and expedite the review of drugs that are intended to treat serious conditions and fill an unmet medical need. Fast Track designation allows for more frequent interaction with the FDA review team.” 8

UltraGenyx describes the Phase 1/2 GSD gene therapy study as “open-label, dose-finding.” The company also describes several success factors of the study, including time to hypoglycemia, impact on lipids, uric acid, and measurement of glycogen in liver. The company states regarding the testing protocol: “There are three potential dosing cohorts in the study,


7 Ibid.

8 Globenewswire, accessed 11/18/18.
and three patients will be enrolled in each cohort. Patients in the first cohort receive a single
dose of DTX401 of $2.0 \times 10^{12}$ GC/kg. The decision to proceed to the next, higher dose cohort
will be made after the data monitoring committee evaluates the safety data for all patients in
the previous dosing cohort."  
As of this writing, UltraGenyx is in the first phase of testing, with the third cohort completed.

* * *

Imagine the excitement. For the first time in history, humans can potentially have their
GSD1a “fixed.” In 1971, the year of my high school graduation, the disease was almost always fatal. And now—forty-nine years later—there’s a chance.

Dr. David Weinstein attends to over 600 patients in 48 countries, so he knows well the
anxiety and hope of patients and parents. As he prepared for a historic moment with Jerrod,
the first human in the GSD gene therapy trial, Weinstein stated, “We’ve been tirelessly working
toward and anxiously awaiting this milestone for 2 years, and so have our patients.” 10 At an
interview, the first human gene therapy recipient Jerrod, remarked, “I can’t imagine a life
without cornstarch. It’s life or death.” Patrick would agree, as he’s administered cornstarch
doses on airplanes, on the beach, in the movies, and even on tree top platforms while zip-lining. He’s carted multiple boxes of cornstarch to Japan, to Israel, to India, to Chile.

As illustrated by the photograph below, Jerrod received his first gene therapy as a
simple IV infusion; he was awake and alert during the entire procedure. Six weeks after the

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infusion, doctors stopped the patient’s cornstarch therapy. Results of the study are not available at this time but will be released by UltraGenyx at some point.

Five steps forward . . . to the walk to independence.

(images from Globenewswire.com, accessed 3/18)

The “high-five” from Jerrod; Dr. Weinstein is to the right.


“Today@uconn-edu,” accessed 11/1/118.


