

ILLUSTRATION BY PETE GARCEAU

THE INHERITANCE

A FAMILY ON THE FRONT LINES OF THE BATTLE AGAINST ALZHEIMER'S DISEASE

In 2009, writer Niki Kapsambelis took on a story assignment that changed her life.

It was an article for the University of Pittsburgh Alzheimer Disease Research Center's newsletter; Kapsambelis was asked to write about a family from North Dakota, the DeMoes. The family was partnering with Pitt's William Klunk, MD/PhD professor of psychiatry and neurology, and Chester Mathis, PhD professor of radiology. The scientists had developed Pittsburgh Compound B, an agent used for imaging brains in living patients to identify buildup of a sticky protein (called beta-amyloid) associated with Alzheimer's disease.

Five of the six DeMoe siblings carried the gene for early onset Alzheimer's. Their father died from it, as did their paternal grandmother.

Kapsambelis met the family at Pittsburgh's Omni William Penn Hotel. At that time, two of the siblings, Brian and Doug DeMoe, who'd worked in the oil fields like their dad, were starting to experience symptoms. The men were then 48 and 44, respectively. Brian was unable to make the Pittsburgh trip.

After meeting the DeMoes, Kapsambelis got into her car to drive home and found herself shaking. "I just felt they had a much more important story than a newsletter article," she told the Pittsburgh Post-Gazette.

Eight years and many, many interviews later, Simon and Schuster published Kapsambelis's book, The Inheritance, *which weaves the DeMoes' story with a narrative of the history of the disease and related research, notably the work of Klunk and Mathis. Kapsambelis came to feel she was part of the DeMoe family; the DeMoes called her their "author-in-law."*

We share edited excerpts of the book here. —Erica Lloyd

In her introduction, Kapsambelis describes the importance of what the DeMoes decided to do with their remaining years. She notes that researchers are searching for a way to predict who will get Alzheimer's so they can treat those people before they slip away.

. . . But to find such a treatment, doctors need a patient who is guaranteed, with 100 percent certainty, to get the disease—only then will they know whether an experimental treatment is successful, by testing it out on that person and then measuring its effect.

Those perfect patients do exist, as one tiny sliver of the population who stand distinctly apart from the rest. They are the people living with one of three known mutations that guarantee they will be stricken. Only about 1 percent of all Alzheimer's patients fall into this category. They are hit young: their average age of onset is between 30 and 50 years old.

These are the people we will thank when Alzheimer's itself becomes a distant memory.

By testing preventive drugs in this population, researchers hope that they will be able to translate a successful treatment before another generation is lost.

FAVORITE SON

Brian was living in a trailer on the outskirts of Tioga when his sister Karla and mother, Gail, began to think that maybe it was time to bring him closer to the center of town. They wanted to keep an eye on him as his memory and thought process deteriorated: Alzheimer's patients often wander and go missing, or people take advantage of them.

They found him a little one-bedroom house just down the street from Doug's and a few blocks from Gail's. He moved in, bringing a stray cat that he'd adopted and called Missy, until he found out it was a male. Then he named it Mr. Missy. Brian's daughter, Kassie, found it ironic that her father, who'd hated cats throughout her childhood, was mellowing under the shadow of Alzheimer's to the point where he doted on his kitty. The first night he spent in his new house, he climbed onto the roof to rescue the cat, only to fall and break his legs in the process.

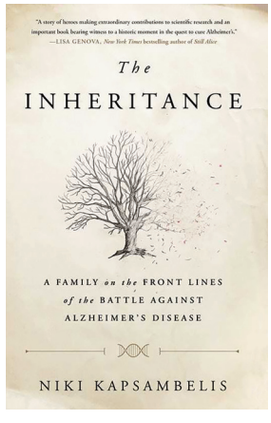
For a few years, Brian got by. In the aftermath of Trey Sunderland's firing from the National Institutes of Health, Karla was determined to steer her family to another research venue. She hoped Bill Klunk's work at the University of Pittsburgh would fill that void.

It would prove to be a fortuitous partnership. In Klunk, the DeMoe family finally found their champion: a man who cared about them as human beings as much as he cared about the scientific knowledge he could gain from their unusual genes. Gail saw Bill Klunk as a man who would watch over her family in ways that she couldn't; his entire staff treated them like old friends. One by one, the DeMoe siblings trekked to Pittsburgh each year, undergoing a battery of tests and brain scans so scientists could see how the disease was progressing both biologically, inside their brains, and clinically, in their behavior.

The study paid for travel costs and provided a meal stipend and small honorarium; in return, each study subject had to schedule at least three days off from work and convince a partner to accompany him or her—a spouse or a close friend or family member who could objectively answer questions about the subject's day-to-day level of functioning. The study partners also had to schedule time off, and in some cases, they had to handle the rigors of traveling with an Alzheimer's patient.

Gail frequently traveled with her offspring, and Klunk and his staff came to know each of them intimately. When Gail's grandchildren grew into adulthood, most of them joined the study.

Though the tests were grueling, Klunk and his group worked hard to make the trip palatable. The subjects were encouraged to sightsee, attend ball games, explore the city. When the



COURTESY KARLA HORNSTEIN



The DeMoe siblings with their mother in 1986. BACK ROW, FROM LEFT: Jamie, Doug, Dean, and Brian. FRONT ROW: Karla, Gail (their mom), and Lori. Karla was the only sibling who tested negative for the gene for early onset Alzheimer's.

workday was over, the DeMoes and the doctor often ate dinner together. Klunk did this with many of the patients in his study, except for certain cases: If study participants chose not to learn their genetic status, he politely declined dinner, fearing he might accidentally tip someone off to what he knew.

Klunk admired the DeMoes for their courage and their selflessness. Their contributions gave science a window to a baffling disease. He would come to know three generations of DeMoes; he would witness their losses and triumphs, learn their deepest secrets and most paralyzing fears. He was humbled by their sacrifices and their trust in him. He would become their touchstone, a lifeline as they were battered by the relentlessness of the disease. Unlike anyone else they had met in the research world, he always made himself available, always returned their calls.

"There's only one family like the DeMoes," Klunk said. "They're the salt of the earth."

Brian made it to Pittsburgh twice to participate in Klunk's study before the disease claimed too much of his brain to make travel possible. His small world grew smaller; he walked across the street from his little house to the Skol Bar, which was populated and run by his friends. They made sure nobody stole from him; it would have been easy to take his money.

At the local drive-in, family friend Kim Johnston worked behind the counter and waited on Brian when he showed up for lunch. Knowing that too many questions flustered him, she placed his standing order for a Philly steak wrap without asking and took the correct amount of money from Brian's billfold when he silently handed it over to her. Then he'd settle into a table across from her and wait for his food, saying, "Ayup," like he was relaxing after a particularly hard day at work.

Brian had always been generous; now he bought small trinkets to give his mother. They were all he could afford, but he made the gesture to show her his affection. He walked to her house, often woefully underdressed for the harsh weather, loping along on his bum leg that had never quite healed right after the fall from the roof.

Kim Johnston remembers teaching him to ride a three-wheeled bike, thinking it would help him get to Gail's more easily.

At 5-foot-2, Kim struggled to hold the small bike while Brian moved the pedals, forgetting to coordinate each push.

"Okay, now, now, now!" she yelled, as Brian started to get the hang of it.

"I can't!" he shouted.

"Can't never could," said Kim, borrowing one of Gail's favorite expressions, and Brian pedaled faster.

They wobbled down the street, past an oil company office; workers stuck their heads out of the window, fascinated at the spectacle of a burly man in a trucker cap shakily pedaling a bike several sizes too small for him while a short woman cheered him on like an exultant mother. By the time they reached Gail's house, Gail was outside, laughing so hard she had to cross her legs to avoid wetting her pants.

"That was probably one of the last times that I thought part of him was still in there," Kim said.

Once he was home all the time, he would spend hours playing the practical joke that had been a DeMoe family tradition for decades: canning cars. The trick never got old. As his disease progressed, it got more hilarious. He canned cars so often that the people of Tioga came to expect it when driving past Gail's house; a few times, he forgot to let go of the fishing line and nearly lost a finger.

And sometimes, though he loved her, Brian was uncharacteristically cruel to his mother. When Brian's mood swung low, he unleashed his temper on Gail, much the way his father once had. Though she knew by now that the viciousness was Alzheimer's, it was impossible for Gail not to take the abuse personally.

Occasionally, she brought him with her when she went across town to volunteer at the nursing home. Sometimes he slipped outside to smoke a cigarette while she was doing a patient's nails. That worried her because she knew he was a wanderer, and the nursing home wasn't a locked facility. What would happen to him when he was no longer capable of living on his own?

The answer came in February 2008. From the other side of the state, Karla had been monitoring her brother's living situation. She called the woman who tended the Skol Bar and learned that Brian was sometimes emerging from the bar's restroom without pulling up his underwear. And Karla knew that the ugly incidents with Gail were escalating.

Because Brian was no longer married, his children were scattered, and Dean had been diagnosed with the mutation, Karla alone held the power of attorney over his affairs. It was up to her to decide what to do next. After consulting with Brian's children and some of his friends, she made the first of what would be many difficult choices. She decided her big brother would enter a memory care facility—one that was locked to safeguard against wandering. It was in Minot, 80 miles away.

Karla told Lori and a few other family members and close friends about her plan, but made sure to keep the news from Gail. No matter how hard it was for her to relive the terror and pain of her final years living with her husband, Moe, Gail could never have agreed to send her favorite son away.

"That was probably one of the last times that I thought part of him was still in there."

The morning they decided to take him, Brian's sisters Lori and Karla—who were staying overnight at Gail's, ostensibly for an ordinary visit—met in the kitchen with Doug's daughter, Jennifer, as well as Kim Johnston. Gail woke to the sound of the gathering and wandered down the hall into the kitchen. When they told her what they were doing, she began to cry.

"Mom, I just know you would never want to let him go," Karla said gently, reassuring her that it was the only choice they could make. Karla accepted full responsibility for that decision, hoping to spare her mother some of the pain. Let Brian hate her; she could recover. Gail never would.

"It was awful," Karla said. "I didn't want her to feel that way."

The decision to move Brian to a locked facility—however pleasant the accommodations, however necessary the move—would be the first of many difficult turning points for Karla. With each decision, a limb of her family tree fell to her feet, her peace of mind eroding just a little more. —*Niki Kapsambelis,*

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