I Wasn't There

Caring for a Patient With Trisomy 21, Alzheimer Disease, and Epilepsy Until His Final Days With COVID-19

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I wasn't there.

Neither was his sister, when he had his first episode concerning for seizure. At that time, James was a 56-year-old sweetheart and diehard Elvis fan with trisomy 21 and Alzheimer disease: the latter I had diagnosed 4 years prior.

Teresa's favorite adjective for him was "amazing." He had been working and living semi-independently, a favorite at the fast-food chain where he was employed and the Habitat for Humanity where he volunteered. He loved to look sharp for any occasion and was always the life of the party. One year, the day after Thanksgiving, he came down dressed up, including a favorite polo shirt, which he preferred completely tucked in, and wearing cologne. His sister asked him why he was dressed like that and he replied that it was for Thanksgiving. He was disappointed to learn that they had celebrated the holiday the day prior. This was the first time Teresa started to be worried he was showing some cognitive changes.

With his memory loss, he transitioned to a group home with full-time live-in staff. Teresa brought him in to see me not long afterwards. I always enjoy watching a fellow lefty draw. When trying to copy a Necker cube, he managed a square. He registered 1 out of 3 recall items, which he subsequently could not remember. He was one of the first patients I had seen with trisomy 21 and Alzheimer disease.

While I'm not sure he ever fully realized why he came to me, he knew I was a doctor, as he was quick to show me a bruised finger or a gouty toe. He had a gentle, genuine smile, which he often shared with his sister, Teresa—to him, Yee Yee. (He had significant dysarthria at baseline, which I learned to understand better over the years, though his sister often "translated" his responses.) Sometimes that smile turned mischievous, which would earn a firm, "James Allen!" from Teresa. While his sister preferred him clean-shaven, at one point he grew out a strawberry blonde goatee and sideburns, of which Elvis would have been proud. I got to know him and his sister well as we were in frequent contact managing behavioral changes and medications. At every visit, he was accompanied by Teresa and caregivers from his group home.

The staff at the home had been the ones who first noted a seconds-long episode of syncope with incontinence, though no convulsions. A local doctor had started him on levetiracetam. Because he was more sedated and without more data as to the "seizure," I weaned him off of the antiepileptic drug (AED), as he had no history of epilepsy.

A year later, he had another episode: again, witnessed by his home's staff, only this time with generalized tonic-clonic activity. The emergency department identified he was likely dehydrated, so perhaps an electrolyte disturbance triggered a seizure, and started him back on levetiracetam. Since he lived a couple of hours away, I hadn't seen any of these events myself and pieced together what I could from outside records and long telephone calls with his sister. Unfortunately, no EEG had been performed. Resuming levetiracetam this time led to anger and irritability, a stark contrast from his usual sunny and mellow disposition. Weighing the risk-benefit ratio, we decided to wean him slowly off the levetiracetam one more time. If he had recurrent spells, we would start him on lamotrigine, to avoid sedation and provide mood stabilization.

I received the report that he had further spells, so we enacted the plan. We transitioned him from dual therapy with levetiracetam and lamotrigine to monotherapy on the latter. He was not having the same sedation or behavioral and mood changes he had with AEDs previously, so we chalked that up as a win.

As a cognitive behavioral neurologist who primarily practices outpatient medicine, I thankfully do not get too many emergency pages. During clinic late one morning, I got a page that he was in our academic medical center's emergency department because of seizures.

You better believe I was there.

As I bolted to the emergency department after my last clinic patient, my gut told me this had been serious enough to warrant transfer from his local hospital. My neurologist's bag was in tow to examine him for myself, despite knowing the inpatient neurology consult team would be coming soon. Every couple of minutes, his whole body would shake with a myoclonic jerk, which we eventually confirmed as myoclonic seizures with EEG. He still remained pleasant and interactive

Figure 1

throughout, a smiling marionette being pulled by invisible strings on all his limbs.[1] The duration and frequency indicated he was in status epilepticus, which thankfully broke quickly with IV levetiracetam.[2]

Because I had supervised his transition off of levetiracetam 3 times over the past 2 years, I felt guilty that being weaned from this AED had triggered this status. We had made these decisions with the full support of his sister and under the careful monitoring of his home staff, but I felt the lion's share of the burden as his neurologist. Teresa and I had bonded over the years in our mutual affection for James, trying to minimize medications and side effects to optimize his care, and I didn't want to let either of them down.

I did not blame myself, though I did indulge in some self-chiding for not taking more seriously the initial accounts of his seizures. I made the best the clinical judgements I could have with the information available to me. I finally believed he had seizures when I was personally there to witness them.

Over the next several months, he stayed seizure-free on the levetiracetam. Our most recent visit, which marked our 7.5-years-long clinical relationship, had been via telemedicine because of the COVID-19 pandemic. His facility had a number of COVID-19-positive cases, so his ability to travel was limited. We had initially planned to make this encounter in person, but another COVID-19 outbreak meant his facility was on lockdown.

I was able to set up screen time with Teresa at her home and him with staff at his facility. A caretaker provided tech support on the laptop while I conversed virtually with the facility physician, who was at his bedside. While his sister and I were disappointed we weren't seeing him in person, at least he could see the full, rather than masked, faces of us on the computer. His caretaker and physician were in full personal protective equipment, including masks and gowns. He slept most of the day now, but woke up enough to beam his megawatt smile over the computer screen. At this point, he was reliant upon caregivers for all his daily activities, requiring assistance to move from his bed to his chair. His cognition had been fluctuating with the visitation restrictions from the pandemic, as he wasn't getting the social interaction his outgoing personality craved. He was pouty with Teresa when she did come, because he didn't understand why he wasn't seeing her as often as he usually did. I made no changes to his medications. We briefly revisited goals of care, which they had discussed at length with the team when he was last hospitalized at Vanderbilt.

A week after his last visit, I was working from home, churning my way through my electronic medical record (EMR) messages, when I saw a note from my nurse. She said he had passed away.

My classic, cliched defense mechanism of denial chimed, *This has to be a mistake. I just saw him last week via teleneurology. He can't possibly have died,* I demurred. I hoped this was some sort of name mix-up from his local hospital. I opened his chart and saw he was hospitalized at our medical center, so I knew the news was accurate. I scanned the progress notes. He had been in the hospital for several days, admitted for COVID-19 complications. While he fought valiantly, his body, with baseline Eisenmenger and advanced dementia, could only take so much.

Reflexively, I called his sister. I'm not an emotional person, but I steeled myself not to cry. I failed: it only took a few minutes before I started choking on my words. I had not planned what to say, but between stammering out condolences and both of us shedding tears, we both grieved his passing. Yee Yee at least had been able to stay by his bedside throughout his hospitalization and was with him when he passed. She was there.

I wasn't.

His admission hadn't been flagged when I logged in our EMR, otherwise I would have visited, I told his sister. *Or at least I would have tried to.* We were roughly 9 months into the gestation of the COVID-19 pandemic in our state. Hospital protocols and visitation policies remained strict. I wasn't sure if I would have been allowed to come if I were not a part of his admission team or officially consulting, since I knew the COVID-19 ward was under tight security.

She told me that she purposely did not contact me to inform me of his admission because she knew I would try to come. She thought of my 2 small toddlers at home—she always kindly inquired after them and I would show her the latest photo saved on my cell phone. Since she didn't want me to risk exposure, illness, and possibly passing COVID-19 on to my family, she waited to tell me.

Guilt struck afresh. If I had known, would I have visited? Would I have justified the potential risk to myself, my family, and other patients and voluntarily swathed myself in double-gloved, double-masked, and gowned layers to hold his hand once more? Would I have been afraid to try? I ruminated over not being there for him when he died, particularly since he was at my hospital. If I had been able to visit, what would I have really achieved? In his delirium, he probably wouldn't have even recognized me. No neurologic assessment I could have offered would have changed the outcome.

I shared my conflicted feelings at a local physician emotional support group I am a part of, Finding Meaning in Medicine. The grief I felt at his passing was still raw, as it had only been a few weeks since he had died. We had all shared our battle stories and scars for how our practice of medicine during COVID-19 had changed, including our transition from in-person meetings for this group. While not physically there, we were virtually.

A wise colleague pointed out to me that over years of caring for him, my presence during all those encounters was what mattered the most. Every clinic visit, when I would hold his hand and evoke his contagious laugh and winsome smile. Every telephone call with his sister, sorting through symptoms and side effects. His death was but one moment in his rich and full life. It was okay that I had missed it.

For everything else, I was there.

Acknowledgment

The author thanks the patient and his sister.

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