

Piecing together confusing symptoms

Perhaps the day my sister Sonja's physician met her, he saw a 55-year-old woman with the typical symptoms of a depressive disorder: emotional blunting, loss of insight, obsessive-compulsive behaviors, irritability, distractibility, and a tendency to overeat. He prescribed a selective serotonin reuptake inhibitor and discussed its side effects. When my sister returned to his office a year later, she complained that she couldn't do her job anymore and didn't feel like herself. The doctor ordered laboratory tests and an MRI of her brain. The best diagnosis he could offer was Alzheimer's disease, even though it didn't



exactly fit my sister's presentation or her young age. He prescribed memantine (Namenda). My brother-in-law drove my sister back to their small country home in disbelief.

My mother first realized something was wrong when Sonja, an accomplished quilter, asked her for help with a pattern. My mom had sat with my sister, helping her measure and lay out the colorful pieces for a

pattern called *Delectable Mountain*. But quickly frustrated, my sister threw down the fabric pieces and never sewed another quilt. In the following 2 years, my family and I became increasingly puzzled by Sonja's behaviors. Declining cognitive abilities and disinhibition appeared first.

One year after her frustration with the quilt, I received a frantic call from my other sister who had just spent a day with Sonja. She described Sonja's behaviors to me, and I poured through all of the books I could find that discussed dementias or any other pathology that could possibly explain my sister's symptoms—incontinence, loss of the ability to communicate, disinhibition, and a shuffling gait. Frontotemporal dementia (FTD) caught my eye early on in my reading, but I dismissed it thinking it to be a rare potential diagnosis that my sister's physician had probably already ruled out.¹ Almost 2 years later, a second MRI confirmed the atrophy of Sonja's frontal and temporal lobes. Although rare, FTD was in fact the disease that was claiming my sister's life. With an accurate diagnosis finally in hand, we knew that my sister would succumb to her illness in 1 to 2 years. As it turned out, it was only a matter of months.

On a bitterly cold and windy day in January 2008, I drove behind the hearse that carried my sister along a winding country road to the tiny cemetery where she would be buried. I kept replaying in my head the events of the previous 4 years and wondered what could have been done differently. I was

overcome not just with the profound sadness of her death but more the sheer senselessness of it. My sister was only 60 years old and, by most measures, healthy until just 4 years ago. My father's death, a few years earlier, had been difficult but expected. He lingered for 29 months with chronic obstructive pulmonary disease. Conversely, my sister's diagnosis of FTD was unclear until just months before her death. By that time, she was a mere shadow of herself.

At my niece's home after the funeral, I gazed through tearful eyes at a crowded wall of photos. My sister peered out at me from among the snapshots—her dark hair framed her rosy cheeks as she smiled with squinted eyes into the camera. This photo revealed nothing of the deterioration that was taking place in her frontal and temporal lobes. To picture that, you would have had to see her sitting in front of a plate of food, not knowing how to pick up a fork. You would have had to be on the other end of a phone conversation during which she would say hello and then hang up in the middle of your sentence. You would have had to see the stacks of fabric squares and unfinished quilts abandoned in her sewing room.

An earlier diagnosis would not have changed my sister's outcome, but it might have helped her providers and family with planning her treatment and care. An important part of palliative care for a patient with dementia is educating the family about disease progression and how the patient will change. Unlike cancer, the prognostic markers in dementia, be it Alzheimer's disease or FTD, do not accurately predict the patient's life expectancy.² However, some complications and comorbidities can signal the end of life.

Equipped with this information, families can make the best of the time that they do have left. They can plan picnics in the park while their family member can still walk and enjoy the outdoors. Grandchildren can sit at their grandmother's side and hear stories about fishing, cooking catastrophes, mud fights, and first dates. Mothers can jot down favorite recipes on index cards and favorite poems can be dog-eared. A sister can hold her sister's hand and tell her she's sorry about all the births and weddings and graduations and anniversaries that will be missed. And she can sit on the porch with her and flip through sticky pages of old photo albums and remind her of all the amazing moments of her amazing life. **JAAPA**

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