

## Praise for Article on Prions

Dear Editor:

I wanted to formally acknowledge the fact I have had the chance to carefully read the recent paper by Janyce Hamilton in relation to TSEs. I felt this was a very well-balanced and informative paper that is extremely up to date in content.

One of the most significant developments over the last few years without question is secondary transmission of the infectious agent(s) by iatrogenic methods. A sustained human-to-human epidemic is entirely plausible if nothing is done. The precautionary principle is completely relevant due to many current uncertainties. Variant Creutzfeldt-Jakob disease has only ever been studied in symptomatic patients (except for the one blood recipient who tested positive at postmortem biopsy and died of other causes). Therefore, we

still do not know what happens early in the disease in humans who will become symptomatic later.

The CJD International Support Alliance fully recognizes, understands, and is engaged in the challenges that are before us. We have to accept the fact that due to the rarity of these diseases, we will never get the level of external support required to fully eradicate them. The level of understanding and passion of ordinary families affected by TSEs should not be underestimated.

Combined with the vigor and passion of key individuals and various research groups, the remaining uncertainties about these diseases I am certain will be understood and then acted upon.

Work on the diagnostic and treatment fronts are equally encouraging; they very

much go hand in hand. The multidisciplinary approach is extremely important.

As with most situations, taking preventative measures is the first “firewall.” I hope this paper goes some way to better educate many with regards to several key aspects of TSEs.

Kind regards,

**GRAHAM STEEL**

*Information Resource Manager  
CJD International Support Alliance*

*Editor’s note: As mentioned in the letter, variant Creutzfeldt-Jakob disease, rare as it is, has an overwhelming human cost to the few people and families affected directly by the incurable disease. The follow-up article, by Janyce Hamilton, author of the prions paper in the January issue, tells the story of one such family.*

### One American Family’s Experience With Variant Creutzfeldt-Jakob Disease

Growing up with her dad, brother and sister in Miramar, Fla., where the family settled after moving from the United Kingdom when she was 12, Charlene Singh, being the oldest girl, took on the role of “mom” in the house and used “her loudest voice” to boss around her siblings. She taught her sister how to tie her laces and how to ride a bike. When she baked brownies, she gave strict orders of when they were and were not permitted to touch them.

Charlene was helpful and caring around the house and always set a good example for her sister and brother, especially when it came to studies. She graduated from high school fourth in her class, got scholarships to college, and organized the March of Dimes walk at work. She had the gorgeous skin tone of her Indian ancestry, and was proud of it — participating in a local Indian fashion show. Yet, she strove to exceed the expectations of a business world whose

leaders often had lighter skin. And she did. She got straight-As in her courses.

Then, three months after proudly becoming the first in the family to graduate from university, there would be another first: an ominous invader that would make history.

Something silent started making its presence known inside her body. Charlene found herself becoming anxious and a bit forgetful, even teary at times, and this new moodiness dimmed the glow of her recent achievement. After uncharacteristically losing her temper, she told her little sister, “You know, Lisa, I think something is wrong with me.”

She usually had her head in a book, but she barely read anymore. Her e-mails to her mother began not making much sense and then she totaled her car in a crash. Soon, she lost her job due to her forgetfulness. When she saw a doctor, he prescribed an antidepressant for anxiety.

But nothing changed, and her family noticed her hands began to shake. She even started to trip and stumble when



**CHARLENE SINGH** was the first U.S. victim of “mad cow” disease.

walking. Her eyes now had a permanent look of fright. Not trusting the doctors here, her family flew her to London, where she got worse. Meanwhile Christmas was upon them, and Charlene’s dad bought her a black dress in just her favorite style as a gift. When her aunt asked her if she was happy with her dad’s gift, Charlene said, “What gift? I didn’t get a gift.”

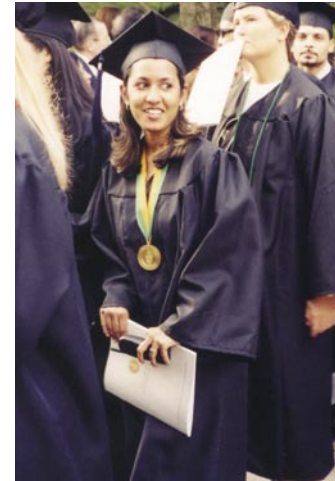
It wasn’t long before London hospital doctors familiar with the symptoms took

a tonsil biopsy and delivered the worst news possible: Variant Creutzfeldt-Jakob disease — Charlene was the first such human “mad cow” victim in America.

While the family struggled with the shock, her mother flew with her back to the United States to die at home with her father, who raised her. Charlene’s highly infectious and incurable disease would make the media, doctors, the Centers for Disease Control and Prevention, and even her dentist, appear to go a little mad themselves as they rushed to react to the news. The news had “implications.”

Charlene’s parents did not want any media coverage at that time. The CDC and

the Florida Health Department advised the family to keep her identity out of the public when they first issued the public health statement. They did not want any unnecessary public alarm. Charlene’s aunt, Sharon Singh-Passley of Pembroke Pines, Fla., said her niece’s identity would have remained private, and did for a while, until she approached her parents about going public with Charlene’s story. “I personally wanted her story to resonate at the USDA and hoped that it would encourage them to step up their screening and testing for bovine spongiform encephalopathy so that such a disaster would not happen here, and also to show the devastation that this disease does



**CHARLENE SINGH** at her college graduation ceremony. She died three years later.

to a human life.” Her parents’ condition was that her last name not be used to protect her family, but one media outlet slipped and gave her full name, so that was that.

Charlene never underwent blood transfusions or tissue transplantations, nor had major surgeries or dental work. “As a child she liked hamburgers, sausages, and steak-and-kidney pie, growing up in the United Kingdom — all the usual foods children enjoy,” Charlene’s aunt explained, “but back then people didn’t know beef cattle were widely infected with BSE, and the public was certainly not warned about the potential threat to human health. We don’t even know if we ate the same food as her; if we will still come down with it. There’s no test.”

At Sunday dinner prayers, Charlene would mouth the words to her dad, “Am I going to die?” Her dad would say, “You are very sick, but we’re doing everything we can to help you.”

Bouts of madness knocked family members off guard as Charlene would kick, punch, and bite. A priest came with a blessing to tame the possessing illness. In the months ahead, she went down to about 70 pounds, stopped walking, and could no longer communicate. A feeding tube was fitted into her stomach because she also lost her gag reflex and ability to swallow. She was unrecognizable as the teenage girl who would get together with her girlfriends, dress up, and go out partying.

Charlene died on June 20, 2004, almost on the exact day she graduated from college, three years earlier. ■■■■