



BOSTON FOUNDATION *for* SIGHT

a patient STORY



kelli

Last Sunday Kelli developed a fever.

David's Journal

March 22

She toughed it out for a couple of days, but Monday night she called me (I was at a conference in Atlanta — my first trip away from her and Zoe in 2 years) and said I might need to come back if she didn't feel better in the morning.

March 23

Tuesday night she developed a rash on her back. It extended to her arms and face later in the night. By early Wednesday morning it was looking pretty bad and she was feeling even worse, so we went to the hospital.



A fever of 106.5°

Antibiotics

Big doses of steroids

Morphine for the pain

Kelli and David, successful Internet entrepreneurs and the parents of a two-year-old daughter, Zoe, were living a happy life. Out of nowhere, Kelli, 36, contracted toxic epidermal necrolysis syndrome (TENS), a rare disease thought to result from an adverse reaction to medication or a virus — or some combination of the two. In Kelli's case, the cause would remain unclear, and the next six years would prove to be a monstrous ordeal.

A sky-high fever and painful rash that would soon cover 70% of Kelli's body landed her in intensive care. An alert resident recognized the condition, which has a frightening mortality rate of 30-40%. The rash had begun to blister, and the top layer of her skin was sloughing off; infection was a deadly threat. TENS, a severe form of Stevens Johnson syndrome (SJS), also attacks the mucous membranes, including the ocular surface of the eye, causing extreme pain, ulcerations, light sensitivity, and potentially blindness.

David, a self-professed geek, went into overdrive, surfing the web for leads on effective treatments for this mysterious condition. He found an Internet user group whose members urged him to make sure that Kelli's eyes were looked after. They also told him about a specialist in Miami who could advise on amniotic membrane transplantation to save and possibly rehabilitate her corneas. Kelli's medical team was hopeful and, in consultation with the specialist, performed the surgery twice in six weeks.

March 25

The doctors had pretty much worked out that this was classic TENS (toxic epidermal necrolysis syndrome) and suggested we either transfer to intensive care or move to the burn center of a local hospital.



Cardiac arrest: Kelli is revived
 Intravenous Immunoglobulin (IVIg)
 Pig skin xenograft to replace and regenerate her skin
 Atrial fibrillation
 Pneumonia

Doctors put Kelli into a drug-induced coma to manage her unrelenting pain. They also administered a sedative that would erase her short-term memory to reduce the trauma she'd experienced.

April 29

Another amniotic membrane transplant onto the cornea. Surgery went just fine. Did as promised under light general anesthetic. One area in right eye off to the side was also affected, so patched that. Plugged all 4 tear ducts. Did patch both eyes to help healing.

June 4

Recovery will mean regaining a lot of the ground. But not necessarily all of it... One small symblepharon (adhesion) in lower corner of right eye. Injured tissue remodels. Scarring can develop in this phase.



“ I WENT TO BED EVERY NIGHT at seven o'clock. I couldn't tolerate the pain. I was depressed, I have to admit. I was sleeping half my life because I couldn't function, but I had to because of my daughter.”

— Kelli

July 20

Eyelid margins are not good. Tear film compromised. As part of disease course.

December 1

20/20 in good eye

Her medical team's herculean efforts saved Kelli's life. David's research and advocacy played no small role. Considering what might have happened, her visual acuity was barely compromised. In fact, the vision in her “bad” eye was 20/30. However, the disease had damaged her ocular surface system, resulting in trichiasis, a condition in which the eyelashes grow into the cornea, causing excruciating pain with every blink and leading to corneal ulcerations. TENS also caused severe dry eye and its related chronic pain and extreme sensitivity to light.

David and Kelli learned about scleral lenses, which rest on the less sensitive sclera, or white of the eye. Although she couldn't fathom putting anything into her fragile eyes, in the fall of 2005 she made an attempt to be fitted. It was not successful, and she entered into a long period of coping and despair. It was a foreign frame of mind for a young woman accustomed to being not only fully engaged in life, but absolutely driven.



In the summer of 2009, she spent ten days at the Boston Foundation for Sight. And everything changed.

Knowing the emotional toll that her corneal disease (and TENS) had taken and the anxiety she felt about placing anything in her eyes, Kelli's medical team at BFS tailored her treatment to support her needs. This included more one-to-one sessions with her trainer — ten, in fact — to ensure that she could insert, remove, and care for the organization's innovative prosthetic devices with total confidence.

She was fitted with five trial devices from the 2,000 found in the BFS library. The refinement process continued with the custom fabrication of several devices, the last pair of which she wears comfortably every day.

Kelli is now doing everything she did before she fell ill; in fact, she's venturing into new territory, having joined the gym and started swimming. Seeing her today, you'd never know what she's been through; you'd have no idea that seeing pain-free was the breakthrough she'd been waiting for.



“**HAVING MY CORNEA BATHED** in liquid all day has affected my whole body.... With my prosthetic devices, I forget that I had this terrible thing happen to me. It's really turned things around.”

— Kelli





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Boston Foundation for Sight
464 Hillside Avenue | Suite 205
Needham, MA 02494
781-726-7337
www.bostonsight.org