STEVENS
JOHNSON
SYNDROME
FOUNDATION

Stevens Johnson Syndrome Foundation

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April 2003

Our prayers go out for the safe return of our service men and woman.
God Bless America!

SJS Foundation Website

New and Improved!

We would like to thank Amit Khatri for the new SJS Foundation Website. The new site is very easy to navigate and has many new links.

Amit Khatri suffered from SJS when he was 12 years old from a sulfa drug. He is severely visually impaired and taught himself to design website's. We are very grateful for his hard work and dedication to the SJS Foundation.

Some new links include:
SJS what do I do now?
PDF SJS article downloads
Please add your story to our Share your SJS Story section on the website
Remember you are not alone!
www.sjsupport.org

SJS Kid's support group

Message board just for Kids! Support for Children and teenagers that have suffered from Stevens Johnson Syndrome or Toxic Epidermal Necrolysis or had a family member with SJS/TEN. This group will be monitored by the SJS Foundation and no emails will be accepted until they have been reviewed for safety purposes. To join go

http://groups.yahoo.com/group/sjskidssupport

Congratulations

To the Law Offices of James C. Barber
For their recent settlement on behalf an SJS Victim.
Involving the drug DAYPRO.
In Jane & John Doe
vs.
Pharmacia Corporation and G. D. Searle & CO
for more information visit their website at:
www.stevensjohonsonsyndrome.com

SJS and the Media

The SJS Foundation has recently appeared in 2 health magazines. In December 2002 in, For The Record, and in the January issue of Today's Dietician. We also interviewed in Denver with a local ABC affiliate. We continue to spread the word about SJS. Thank you to Valerie Chereskin for her public relations work!

We would like to thank all of you that have held fundraiser's for the SJS Foundation. We are very grateful. The Foundation has supplied brochures for fundraisers and you have helped educate the public into these terrible reactions.

Disclaimer:

The SJS Foundation newsletter is for support and information purposes only. No part of this newsletter should be used for medical advise or treatment. Please contact your physician for medical advise. The views and opinions of SJS Support Group members are strictly their own and not necessarily those of the SJS Foundation

Our trip to Boston!

by Jean Farrell McCawley

On March 24, 2003 we took Julie back to the Boston Foundation for Sight. While we were there we met Warren Levy. He was there for a check up on his scleral lenses. Warren told me how terrible his eyes felt until he got the lenses. He looked Great and it was wonderful to meet him.

Julie was only fitted for a lens for her left eye this time. Unfortunately she has too much damage to her right eye and Dr. Cotter could not get a lens to fit her properly. We were so thrilled that she was able to fit her sighted eye, and even more thrilled when her vision was 20/30 with the lens in. It was a bit of a struggle teaching her how to put the lens in herself. Dr. Cotter amazes me, she has tremendous patience. I myself had to leave the room several times to regain mine.

The SJS Foundation has purchased toys for the BFS waiting room. Now children can play between fittings and parents can relax for a few minutes. It was hard to occupy our 3 year old Kerry and 9 year old Julie during the waiting times.

Dr. Rosenthal and Dr. Cotter were great as usual. We are so thankful to them for all they have done for SJS Patients around the world. They have given so many people the chance to have their life back.

In Loving Memory of Joseph Berner

By his daughter Kathleen.

This story is dedicated to the memory of my beloved father, Joseph Berner, who courageously fought SJS and TEN but lost his battle on Aug. 11,2000. As I write this two years later, visions of his horrendous rash, blackened lips, blistering and shedding/sloughing skin most of all, his terrible pain and suffering come to mind all too well.

Dad had a nasty rash for about 7 weeks, which became progressively worse. It began as an itchy, red rash and his skin ended up looking like red leather, or scalded skin. He was being given a new treatment, called Fludara, to help him into remission from Non-Hodgkins Lymphoma, a treatable, but incurable cancer he'd had for nearly 3 years. His oncologists and dermatologist felt he was just dealing with this nuisance side-effect of the Fludara, yet his rash worsened with each subsequent treatment; he had 4 treatments by the time he was hospitalized. In addition, Dad was on other medications simultaneously with the Fludara. Only Cortisone and severe itching and inflammation from the rash. Next, Dad got very sick with fever, sore throat, and severe coughing.

After notifying his doctors of this worsening condition, our family initiated his hospitalization, where he was diagnosed with pneumonia. His lips were already a scabby, black color and he was very dehydrated. He was immediately hydrated with IV fluids and was given steroids and antibiotics for several days. We were so hopeful that while his pneumonia was being treated, his terrible rash could be better evaluated as well. Dad even seemed to improve a bit from all of the fluids, but he still had no voice and an extremely productive cough; he coughed so hard that he often choked on the profuse mucus. On Dad's fifth day in the hospital, pustules and blisters began appearing on various parts of his body and we all became very concerned, but were told that the rash was "running its course." A few days later the blisters were weeping and his skin began cracking and sloughing off at those areas. The friction of moving his blistered, sore bottom, back and arms on his bed was extremely painful and he needed help to sit up and change positions. Even his tongue began sloughing off layers, making swallowing yet more difficult. Never had I seen anything so bizarre. All of this time Dad was being thickly smeared with a white cream called Nystatin, combining a topical steroid, antibiotic and some other property I can't recall. No bathing took place at all. Also, Dad's eyes felt gritty and dry, but the ophthalmologist didn't find any vision abnormalities, he just prescribed moisturizing eye drops like natural tears for the discomfort. Dad's eyes looked red-rimmed and sore; the drops did very little and Dad found reading or looking at TV undesirable.

Most of the time I found him praying when I came o see him daily now I think I know what he was praying for. We all questioned the staff caring for Dad constantly, but nobody told us Dad had SJS until Dad's 11th day in the hospital, when it was mentioned to my husband. It wasn't explained in depth, only that it was caused by an adverse drug reaction. The doctor said it could be far worse than it was. We later found out that each doctor who saw him from the 1st day he was admitted to the hospital wrote SJS as a diagnosis on his medical chart records (which we obtained after his death). These doctors included his oncologists, infectious disease doctors, dermatologist, and ophthalmologist. After being sponge bathed on the 9th night in the hospital. Dad took an extreme turn for the worse, although we had just been old he wasn't ready for bathing after my sister inquired about it. He was given morphine but he still shook from the severe pain. They discontinued his IV fluids for some strange reason this time; he couldn't eat and barely drank. Yet he was moved from an oncology to unit a transitional care unit on the 12th day, usually a place where improving patients are sent. After all of this time they suddenly felt he needed a more sterile environment; the nurses and our family, the only visitors allowed, wore masks, gloves, and gowns.

The nurses here openly admitted that they didn't know how to care for someone in Dad's sloughing skin condition; theirs was the first recommendation of a burn unit for proper care of his skin loss and what now many became raw wounds, like that of a burn victim. These nurses were the first to show any honesty there.

Not without a verbal hassle with an infectious disease doctor and one oncologist, my brother-in-law, a pediatrician, who had just returned from traveling, helped facilitate having Dad moved by ambulance to a hospital with a burn unit on his 12th day. When the elevator doors to the burn unit opened and Dad was wheeled out, they knew immediately that they had an SJS patient by the smell of his rotting flesh. It was determined that Dad had lost more than 30 of his skin and his wounds were deeper than 3rd degree burns.

He would need skin graft surgery on his back, where the wounds were the deepest, and much rehabilitation; he was stripped of the good muscle tone he once had. After all that had transpired, the risk of infection was also very great. Debriding bathing sessions began immediately to remove the dead skin along with the thick white cream slathered on him at the previous hospital no easy task. IV fluids were quickly started and medicated/sterile gauze was carefully placed on the afflicted areas. The burn unit staff treated him with dignity and compassion as they cared for him. In addition, they communicated and were honest — something new and much welcomed. Even Dad weakly whispered to one of my sisters that he wished he'd come to the burn unit sooner. Despite looking better on the outside from the debriding sessions. Dad still didn't feel good and still had severe pain, even after all they were doing to help him. As everything unfolded, what was saw going on outside of his body was also happening internally. The SJS had already progressed to TEN, Toxic Epidermal Necrolysis. By the 3rd night in the burn unit, Dad began to crash so fast that the staff there was shocked. He was intubated and his kidneys were failing. Dad went into profound septic shock, an extreme bacterial infection carried through the bloodstream, which causes the vital body systems to shut down. None of their valiant all-night efforts or machines could keep him alive. On the 4th day in the burn unit, we all said our good-byes as Dad lay in a coma, for we had made the decision to end life support; only his heart was beating, everything else was in failure.

There was no hope for survival. Dad's battle ended—SJS/TEN won. Dad's team of doctors at the first hospital terribly mismanaged his care, which made his death from this horror show condition, even more difficult. In hindsight, we all wish we knew at the 1st hospital what we now know about SJS/TEN all we learned at the burn unit and more information obtained about it since his death. There was never a doubt about his care at the burn unit.

I hope and pray that this horrendous condition can become more widely recognized by the medical profession. More warnings need to be given to the general public about the havoc a severe adverse drug reaction can create if the culprit drug is continued and SJS is treated improperly/allowed to progress. Perhaps these warnings and better information could eventually lead to improved diagnosis, proper treatment and delegation of care to a burn unit, if needed. As we already know, far too many have suffered permanent health damages or death from SJS/TEN—God bless the survivors and the souls of those who lost the battle...forever in our hearts.

Kathleen Lutterschmidt

SJS RESEARCH

My name is Jean McCawley and my daughter Julie, had Stevens Johnson Syndrome when she was just 11 months old. Julie was prescribed Phenobarbital for pediatric epilepsy. Fourteen days later the nightmare began. Julie's eyes swelled shut and she broke out in blisters. No one knew what was wrong. Finally 4 days into her hospital stay she was correctly diagnosed with SJS. The medication was withdrawn and supportive care was started.

Julie's young life was changed forever that day. She suffers from eye, esophageal and lung damage. She is now 9 years old. I was desperate for information and answers to why this happened to my baby. No one could tell me. Every doctor I asked gave me a different answer. There was no information available to parents so we decided to start the Stevens Johnson Syndrome Foundation. That was in 1995. Since then we have heard from people from around the world. Unfortunately, research into SJS has been slim to none. The National Health Institute just has so much money and as rare diseases go, SJS is not getting funding for research.

I for one would like to know why my child was stricken with SJS. Could this happen to my younger child? What about my own siblings, my future grandchildren? I want answers and research. Could there be genetic markers that make our families more susceptible to adverse drug reactions? Isn't research what we all want?

I am happy to say that First Genetic Trust wants help give us those answers. They are a privately funded research company. They have the funding for this project. In our economy more and more research is going to the private sector. Grants are few and far between and we need the help now. First Genetic has the way and the means to provide the answers to our questions. Anyone that has had a loved one suffer from SJS/TEN knows the horror and helplessness we all feel. We have a common bond.

Through this research we may be able to save a future victim and spare a family from this devastating illness. We cannot wait for the government to help us. My heart breaks everyday as I hear from grieving parents that have lost their child to SJS. It will continue as long as we delay research.

The Boston Foundtion for Sight needs your help!

Most Health Insurers will not pay for the scleral lens.

There are many SJS/TEN patients who can benefit from the sight-restoring Boston Scleral lens. The vast majority of SJS patients are young, in the prime of life. Some are children. Most have exhausted all other options for regaining functional vision and many face a lifetime of relentless eye pain and disabling sensitivity to light. The Boston Scleral Lens is their only hope for regaining functional vision and finding relief from the pain that dominates their every waking moment. For others it represents an alternative to corneal transplant surgery with its potentially serious complications, long recovery time and uncertain visual outcome. What is the monetary value of giving them the opportunity to reach their full human potential, eliminating their need for welfare and disability support and enabling them to become productive tax-paying members of society? The answer of most medical insurance companies in the U.S. is: Fitting **the Boston Scleral Lens is not a covered service."** This is not an experimental procedure. Approved by the FDA in 1994, the Boston Scleral Lens has proved its effectiveness in hundreds of patients and our results have been reported in the top peer-reviewed medical journals. This is the ultimate irony: a safe procedure that provides the immediate and priceless gift of sight for less than $8,000 is denied coverage while corneal transplant surgery is covered.

Because many of their patients have been profoundly disabled for years some for decades, most have exhausted their savings and that of their families. **Can we deny them the right to see because they can't afford to pay?** Our answer is a resounding **"No"**. As a result, more than half of our patients depend on the compassion, generosity and charity of strangers in order to rejoin the mainstream of society. This should not happen in the wealthiest nation in history whose citizens are protected by the Americans With Disability Act. What is wrong? The answer is: they are the victims of powerless bureaucracy and a medical delivery system that is unresponsive to one of the most basic medical needs of society.

What is the process for securing insurance reimbursement for a new medical procedure?

A new CPT Code: the first step (a catch-22 in our case)

This code defines each medical procedure and establishes its relative reimbursement value. Since none of the existing codes or their modifications are applicable to fitting the Boston Scleral lens, we applied for a new CPT code **It was denied as was our appeal.** The reason: Your procedure is not widely available." **The requirement that a new medical procedure be widely available** in order to be issued a new CPT code, has in this instance, insured that the Boston Scleral Lens will never be widely available since other academic eyecare centers in this country are hesitant to offer a costly non-reimbursable procedure. This provision, presumably meant to protect the interests of consumers, serves in this case to deprive tens of thousands of citizens of a basic human right: the right to see. We need to change this rule or be granted an exception.

Securing coverage: the second step

Health insurers are not legally required to provide coverage for a medical procedure even when it is defined by a CPT code. Their decisions are based on competitive market forces. Orphan devices like ours that have a very limited constituency and lack the power of numbers are not even on their radar screen. **But they do yield to public and political pressure.** How else can one explain the enlightened policy of many insurers of covering breast implants while denying coverage for a procedure that, for many, represents the only means of restoring functional vision.

Securing adequate payment: the final step

The battle is not over with the issuance of an appropriate CPT code and classifying the Boston Scleral Lens fitting as a covered service by health insurers. A far greater threat to the survival of this vision-restoring technology would be the decision of health insurers to assign a fee that is less than our clinics' costs. This is not a theoretical possibility. It has happened in the past. Since most provider contracts prohibit balance billing, this decision would be tantamount to imposing a loss on the clinics for every patient fitted with the Boston Scleral Lens even for those who can afford and have the desire to cover the costs of the procedure. Is there a way to bring forces to bear to correct this injustice?

We need your help!

As our vision-restoring contact lens devices become more widely known among our colleagues in the eyecare profession, the number of desperate patients referred to us continues to grow and this success is testing the limits of our financial and physical resources. The need to establish this technology in other centers is becoming more urgent. Yet, the uncertainty of insurance reimbursement represents a major disincentive for academic eyecare clinics to offer this life-changing treatment and we need your help to correct this injustice.

Cont:

Jo

The public media

Journalism is one of the bulwarks of a democratic society. We need to get the word out and recruit this important resource. Help us reach the print, radio and TV journalists who can make a difference in our quest to correct this discriminatory practice.

The political establishment

Who are better equipped to correct injustice in our society than our elected officials? Who are more responsible for protecting the rights of the underserved in our communities than our elected representatives? Help us to enlist their support for this mission.

Financial support

In the absence of insurance reimbursement, it has been necessary for us to subsidize the costs of fitting the Boston Scleral Lens for over half our patients. And as their numbers rapidly increase, our ability to serve them is jeopardized. **The right to sight should be a birthright and not a privilege.** But until it is, we need your help if we are to continue to offer our life-changing lenses to those in need and turn no one away because of their inability to pay.

If you are moved to support this effort, please contact us at or call the Boston Foundation for Sight at 617-735-9330.

Perry Rosenthal, M.D.

Founder and President

The Boston Foundation for Sight appeared on OprahWinfrey

We were all very excited to see The Boston Foundation for Sight on the Oprah Winfrey show. A great segment on the scleral lenses, entitled Medical Miracles was aired Feb 3, 2003. Our good friend Joe Zienowicz was featured ! Joe as most of you know had TEN resulting in a great deal of eye damage. Since Joe was fitted for the lenses he was able to return to work, drive a car and golf. The Scleral lens gave him his life back. We are very happy for him and his lovely wife Susan. To learn more about the Boston Foundation for Sight,

Please visit their website at:

www.Bostonsight.org

The Genetics of SJS" study is progressing well and we now have 12 sites actively recruiting across the United States. The investigators and locations of the sites are as follows:

- Boston, Mass Dr. Perry Rosenthal
- New York, New York Dr. Roger Yurt
- Livingston, New Jersey Dr. Hani Mansour
- Miami, Florida Dr. Nicholas Namias
- Cleveland, Ohio Dr. Charles Yowler
- Nashville, Tennessee Dr. Ronald Barton
- Minneapolis, Minnesota Dr. John Twomey*
- Milwaukee, Wisconsin Dr. Ramesh Sachdeva
- Denver, Colorado Dr. Paul Bauling
- Salt Lake City, Utah Dr. Jeffrey Saffle
- Los Angeles, California Dr. Warren Garner
- San Diego, California Dr. Daniel Lozano

We will be able to enroll study participants aged 6 and above in this study. This will require additional approval by the Institutional Review Boards that approve research involving human subjects; Dr. Rosenthal and Dr. Twomey have received approval to enroll pediatric research participants already.

Thus far we have 24 research subjects with SJS enrolled in the study and nearly 20 more that we are contacting to schedule for appointments. Our goal is to enroll 200 research subjects that have been admitted to the hospital with SJS or TEN as a result of taking any medicine.

If you are interested in participating in the study and have been admitted to the hospital (stayed over night) with SJS or TEN within the last 10 years, please send an email to info@firstgenetic.net or call the FGT Helpdesk at 866-259-1772 (toll free). We will forward your contact information to the closest site and they will contact you.

Penny Manasco

Executive Vice President,
Chief Medical Officer
First Genetic Trust

The Stevens Johnson Syndrome Foundation is a non profit organization. We rely solely on contributions help us continue to provide information packets, borchures and newsletters to patients and their families suffering from these severe reactions. Your donations are tax deductible and very much appreciated. .

Our special thanks to those people listed below for their generous donations:

General Donations

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In Memory of Grace Corey

Sylvia Koskey

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