

March 7, 2007

Dear Family, Friends, and Colleagues:

Ever been submerged and trapped underwater? That scare hit the Greeley family like a tidal wave last summer while on vacation in northern Michigan. Out on Lake Michigan, Dave, Alex, and Jack were enjoying a simple ride on Grandpa's super-sized Jet Ski when things went bad. Clipped to Dave life jacket to life jacket, Jack was driving up front while Alex held onto her father in the back. While maneuvering a turn, the Jet Ski turned right and Alex went left. Like dominos, all three people fell and, shockingly and against its engineered design, the Jet Ski turned over and stayed there with Alex clear from the wreck and Dave and Jack stuck underneath the machine and the water. In what felt like an eternity, Dave unclipped Jack's life vest from his own and shot putted him to the surface. Within a few horrifying seconds, Dave accounted for both kids and then a pointed moment of perspective and real life comic relief hit. Alex, never mistaken for brave, shook off her fright, while Jack was none too pleased. In between his gasps and tears, Alex's giggling at her brother, and Dave's 45 minutes worth of paddling to shore while pushing a toppled-over Jet Ski, Jack repeated over and over again, "This is the worstest thing that has ever happened to me!" Boy, Dave thought; if only that were true.

As many of you know, Jack has seen and survived many greater challenges and horrors during his short life. With this March 17th on the horizon, Jack is ready to not only enjoy St. Patrick's Day, but also his 7th birthday. For six years now, Jack has valiantly battled against cystinosis (SIS-TIN-OH-SIS), a rare genetic-metabolic disease inherited from a recessive gene carried by both parents that can potentially ravage a variety of major organ systems in the body – muscular, skeletal, digestive, neurological, gastrointestinal, and cardiovascular. Fortunately, Jack continues to defy the odds and has surprisingly good health. For the 5th year now, we have shared our story with others; to not only help with our own catharsis, but also seek your support of the Cystinosis Research Network (CRN), a nationally recognized 501(c)3 organization whose mission is to improve the quality of life of those with cystinosis through more research, better treatments, increased awareness, and accessible family support, while ultimately seeking a cure. More information about cystinosis can be found at www.cystinosis.org.

As a refresher, what have the last six years looked like for Jack? Succinctly, Jack:

- Was diagnosed with cystinosis in April 2001, joining the ranks of about 1,000 others.
- Lived in the neighborhood of death for 18 months; vomiting 10-15 times per day, losing almost half his body mass, enduring two hospital stays and countless brutal tests to find his diagnosis, which incredibly happened in just six weeks thanks to an amazing team of doctors at Rush Children's Hospital in Chicago – pediatrician, gastroenterologist, geneticist, nephrologist, endocrinologist, and ophthalmologist.
- Had a g-tube implanted through his stomach wall in 2002. Imagine a faucet sticking out of his stomach; attach a hose and pump in medicine and liquid, which unquestionably aided his stabilization and enabled him to hydrate, grow, and develop. (Note: In a recent development, Jack has done so well that his tube was removed just prior to this past Christmas. Jack proudly now sports what appears to be a second belly button or when he wants to feel tough, a "bullet hole" as he likes to describe it.)

- Has taken roughly 55,000 pills, including Cystagon, a miracle drug that allows Jack to battle his number one nemesis, an amino acid called cystine. A healthy person is able to routinely rid cystine from each cell in the body, which Jack can only do by taking Cystagon on a daily and highly structured six hour medical routine; every 8 am, 2 pm, 8 pm, and 2 am in his case, for almost 2,200 days now. Unless better treatments are found, Jack will do this for every day of his life.
- Maintains surprisingly good kidney function and manages to have fair muscle development and swallowing capabilities. The first blow in a series of ripple effects for those with cystinosis is end stage renal failure. It is quite common in kids Jack's age to need dialysis and a kidney transplant. In all likelihood, Jack will one day need a new kidney. It is not known if he will face other life altering challenges common with cystinosis like muscle wasting, diabetes, blindness, deficient pulmonary function, hypothyroidism, and neurological deterioration.
- Wore a leg brace for three years to help fix his cocked feet and strengthen his leg muscles. Today, Jack gets around fine with a distinct gait, but does have some coordination and agility challenges.
- Has progressed socially and scholastically with his peer group, despite the understandable "emotional baggage" that he carries. Beyond the doctors who have aided Jack's body, a bevy of teachers, clinicians, and therapists (see speech, social worker, psychologist, etc.) have helped his inner being as well. Jack continues to work through issues of control and frustration manifested in his earlier fight to survive.

For those that know Jack, they see a great kid who goes to 1st grade, plays baseball, soccer, basketball, and video games with this buddies, participates in Cub Scouts, and loves and annoys his sister, Alex like no one else. Yes, he is every bit normal, just not as healthy as most, but much healthier than others. We count our blessings everyday at how relatively well Jack's plight has gone, yet we know our mission will never be complete until there is a cure for cystinosis, which makes the cause more than just being about Jack, although he is our driving force and centerpiece. While we have never been comfortable in making Jack a "poster boy," we know that with our small community, if we do not do our part, progress will not be made. We have shared words of hope, love, fear, perspective, the unknown, help, support, and more. To date, through this annual letter writing campaign, we have raised over \$100,000 to battle cystinosis. In the spirit of St. Patrick's Day, that deserves a toast and a hearty thanks to so many.

Of course, as Jack's parents, no progress is unacceptable. Each day through our interface with the cystinosis community, we know that others – both individuals with cystinosis and their families - are not as fortunate as Jack or our family. The battle must be waged for Jack and others and it must encompass fundraising, research, family support, pharmaceutical company outreach, and political lobbying. Part of that assault is an Internet support group, a key function of CRN. The stories from the e-group give us pause, perspective, inspiration, and thanks each and everyday...hopefully you find similar food for thought. These are some of the true stories in real words of real people throughout America:

The financial hardship and physical strain on everyone -

"We are doing dialysis at home now overnight. I have been working allot to make up for lost wages. When I work I connect XXXXX at 9pm and disconnect at 5am. I leave for work at 5:30a and return home @ 8:30pm.....She actually has dimples that I have never seen. She has not got anymore energy yet but hopefully we will start seeing an improvement. She has not returned to school but her Homebound teacher came out today.....She is still inactive on the transplant list because her EBV is elevated."

“...how many of you have had to experience a kidney transplant...is there organizations that will help with the financial part...? My husband is the only one that brings in the income, and let me tell you it isn't that much. We barely make ends meet. We get farther behind everytime we have to take my daughter to the doctors or a stay in the hospital...I am really concerned about what will happen to us financially when my daughter goes through this.”

Cystinosis does not live in a vacuum and there are other life issues to manage –

“My husband has been unable to work steadily since his stroke three years ago and more recently, he was diagnosed with Chron's disease. Ultimately, I'd like to get him on SSI as well.” (*mother of a child with cystinosis*)
“...my oldest healthy (thankfully) and in college, my middle one with spina bifida, hydrocephalus and wheelchair dependent in high school and my youngest with cystinosis and in middle school. It's a busy life but...the joy far outweighs the difficulty...I've learned to cherish each day...not everyone is cut out to handle the medical hardships. My girl's father and I divorced six years ago and he has chosen to not be active in their lives.”

Ripple effect of years of damage and the potential death sentence –

“XXXXXX's year has not been good and she doesn't have long left. After Christmas she will be stopping dialysis and the Dr's say that she will probably have somewhere between a week and a month to live after that. The last 10 months she has spent in so much pain that constant narcotics have been the norm...She has fought a good long fight and just doesn't have any fight left...we don't know if she will be home for Christmas or not.” (*an email from a husband regarding his 34-year-old wife...she died a few days later*)

By-product health issues and the impact of physical development –

“XXXXXX has had the knee stapling to correct her bowed legs...gets the staples taken out of her left leg in the next month or so...they staple the growth plates that lets the outside of the leg catch up with the inside.”
“XXXXXX has problems with tetany. The first time about killed her because they didn't know what it was and she was only 11 months old so she couldn't tell us what hurt. By the time they figured out what it was, it headed down her neck/airway. Her whole entire body was stiff/rigid and they figured it was just a matter of time before it would have closed off her airway.”

Ripple effect of treatments causing new problems –

“XXXXXX was transplanted 4 years ago but then got cancer (Post Transplant Lymphoproliferative Disorder) caused by her anti-rejection medicine. She underwent chemo all summer and had to stop taking her anti-rejection meds. Her transplanted kidney, now unprotected from her immune system, began to reject...She is not doing great on dialysis...vomits...stomach hurts...does not eat anymore, maybe 4-6 bites all day...she gets feeds at night through her g-tube...she is eligible for a new transplant one year cancer free.”

Kidney problems and the bed wetting/diaper repercussions –

“XXXXXX is 4...our insurance pays for diaper inserts. They soak up about 13 oz or urine and make the nighttime much easier. We went from changing her 4-5 times in the night to twice.”

Diagnosis/healthcare challenges and the mental/emotional anguish –

“...daughter that is five that was just diagnosed with cystinosis. We have thought for the past 4 years that she had Barters syndrome. We are now facing a kidney transplant in the next 3 to 4 months.”
“My son wasn't diagnosed until he was almost 4 years old either. I was blamed for all of medical problems and his small stature. They wrote failure to thrive on all of his medical reports and blamed me for poor care.”
“...our three month old twin boys were tested and it came back they do not have cystinosis like their sister.”
“We have the disease in both kids under control. I think for now we are dealing with the emotional and psychological effects of this disease. It's a roller coaster for sure! Our marriage has taken a back seat but now we have been in counseling for almost a year.”

This reality continues to compel us to act and is why we ask for the consideration of people like you to help. In recent developments with CRN, an all-volunteer organization, Christy has expanded her role from VP of Research to include Executive Director, which also precipitated the move of CRN headquarters from Boston to the Greeley home in Chicago. A key current task is the planning of the 2007 CRN Family and Medical Conference that will be held this July in San Antonio, TX. Meanwhile, Dave continues to serve on the CRN Board. His wife's dedication amazes him and he often jokes about how she just doubled her pay, really capitalizing

on her pre-med and Master of Public Health degrees. What was the first rule of multiplication? Ah, yes, anything multiplied by 0 is still 0! ☺

It is not easy being objective with a “sick” child at home, but cystinosis has provided us a tremendous gift in a world of growing complexity and materialism. In his best-selling book, *tuesdays with Morrie*, Mitch Albom writes about his dying college professor, Morrie Schwartz, whose body but not mind is being destroyed by a horrific disease, ALS. Using Morrie’s words of wisdom, Albom writes – “As you grow, you learn more. Aging is not just decay...it’s growth. If you’ve found meaning in your life, you don’t want to go back. What really gives you satisfaction...is offering what you have to give. Devote yourself to creating something that gives you purpose and meaning.” For us, cystinosis has provided us purpose and something to share. From our hearts, we thank you for hearing our story and wish you and your family the best.

Sincerely,

Dave “Jack’s Dad” Greeley

Christy “Jack’s Mom” Greeley

Please consider supporting the Cystinosis Research Network, including a tax-deductible contribution. Inquiries can be made and checks sent to:

Jack’s 7th Birthday Celebration
c/o Cystinosis Research Network
302 Whytegate Court
Lake Forest, IL 60045
crn@cystinosis.org
www.cystinosis.org



Jack enjoys a happier moment on the beach at Lake Michigan.



Jack takes a break from picking pumpkins to hug his sister, Alex.