

February 23, 2006

Dear Family, Friends, and Colleagues:

This past holiday season, the temptation proved too great and the Greeley family purchased a DVD of *It's a Wonderful Life*, the all-time classic film by Frank Capra that stars Jimmy Stewart as George Bailey. The story tells of how Bailey, through the help of his aspiring guardian angel, Clarence, assesses his life and confronts the scenario of what would have happened if he were never born. Ultimately, Clarence earns his wings as a guardian angel when Bailey realizes his success in life is based upon the blessings of family and friends. The film culminates when Bailey's young daughter utters, "Every time a bell rings an angel gets his wings." While our family was watching the movie, an amazing thing happened; our door bell rang. The UPS man had just delivered our monthly supply of Cystagon, the medicine that for all intents and purposes keeps our five-year-old son, Jack alive. The symmetry of that moment – Bailey's self-reflection and the plight of our own family – was profound through the metaphor of something as simple as a bell.

As many of you know, Jack has cystinosis (SIS-TIN-OH-SIS), a rare metabolic, genetic disease that can potentially destroy every major organ system in the body – muscular, skeletal, neurological, gastrointestinal, and cardiovascular, among others. Through many guardian angels of our own, Jack is set to celebrate his 6th Birthday this St. Patrick's Day and we are pleased to say that Jack's relatively good health continues. So, as March 17th rolls around again, we are reaching out to our personal network through this, our fourth annual letter writing campaign, to ask you to celebrate this holiday in honor of Jack and his birthday, embrace your own good fortune, and consider supporting our cause by making a donation to the Cystinosis Research Network. CRN has a mission of improving 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, including our past three outreach letters, can be found at www.cystinosis.org.

Some 25 million Americans battle a rare disease, so the daily routine our family faces is not drastically different than many. Jack maintains a 24 hours a day, seven days a week, 365 days a year strict medical regimen – up to six medicines, four times around the clock at 8:00 am, 2:00 pm, 8:00 pm, and 2:00 am. With Jack now in kindergarten, we have overcome a new twist in that he has done well with the school nurse administering his afternoon meds. Short of a cure and improved formulations of his drugs, this will not change for Jack's entire life. This "existence by alarm clock," whether literal or figurative, matters critically for Jack's well-being. When our 2:00 am bell has sounded every night since Jack was diagnosed in the spring of 2001, we have long since realized that this routine drudgery is disguised as a blessing...our own guardian angel, if you will, summoning us to help Jack with another round of Cystagon. Just like Clarence and George Bailey, the circumstances and benefits of Cystagon alone would make you think that divine intervention has helped Jack and the roughly 1,000 other Americans who battle this obscure disease:

- Someone discovered this drug. It is a medication that enables the amino acid, cystine, to leave Jack's cells, which his body cannot do as others routinely can. Without it, Jack would have cystine crystals accumulate throughout his body, particularly in the kidneys, eyes, liver, muscles, pancreas, brain and white blood cells that would minimally lead to end stage renal failure and require a kidney transplant for survival.
- Jack has been able to tolerate Cystagon treatments, which many patients cannot. This offers us hope that Jack will avoid or minimize other life threatening complications like muscle wasting, difficulty swallowing, diabetes, blindness, decreased pulmonary function, hypothyroidism, and neurological deterioration.
- One pharmacy in this country (CVS ProCare of Ohio) distributes the drug manufactured by one pharmaceutical company (Mylan Labs). In an era of a healthcare crisis and publicly traded drug companies driven by the bottom line, an "orphan disease" like cystinosis does not normally rank high on the pecking order. Yet, this little used, unprofitable drug makes a life-altering difference for cystinosis patients.
- The drug is shipped to our home via overnight delivery. In a time of great conveniences in life, the overnight delivery trade does more than serve customary business needs. Taking it a step further, Jack's blood is express shipped each quarter from his hospital (Rush University Medical Center) to the main lab in the U.S. that determines cystine levels (UCSD Medical School); such efficiency is critical.

Despite Jack's body chemistry being pretty sound currently due mainly to his early diagnosis (within six weeks of testing at the age of one) and his acceptance of harsh Cystagon treatment, we still truly cannot predict his long-term prognosis. With continued in-roads in research and treatments through activism in the Cystinosis Research Network, we are hopeful that a daunting disease like cystinosis can be managed like other more common diseases, such as asthma and diabetes. This uncertainty drives us and is largely why we serve on CRN's Board of Directors and reach out to our network via means like this letter.

Since we last wrote a year ago, we have enjoyed much success, which many of you have warmly conveyed you have heard about through the CRN newsletter. Most significantly, we held our bi-annual Family and Medical Conference in Salt Lake City last July that was attended by nearly 50 families, over 160 people, and 19 doctors and scientists. We have continued to make national and international in-roads in our community by building awareness of the disease via the CRN's internet support group; such information exchange between families and practitioners really benefits many, particularly those less fortunate. With every "ding" of an incoming email from this e-group, it is another bell sounding that could help make a difference. Much public awareness was also generated, including a feature on Jack in Chicago's largest suburban newspaper, *The Daily Herald* (see enclosed article), and a segment on the Discovery Health Channel's program, *Mystery Diagnosis* that featured the Jordan family of California and their three-year, bewildering trek in trying to diagnosis their son, Joey. Additionally, many fundraising events were held. Through our network alone, this has included the 2nd Annual Jack Greeley Family Fun Day at Tamarak School and Day Camp that yielded \$15,000; a stranger, having read the *Daily Herald* article, ear-marked \$1,000 raised from his annual golf tournament to CRN; and numerous people who have received our letter extended their personal donations into corporate involvement, such as company donations and corporate matching. All told, this type of generosity has rung a critically important bell, the cash register for research. This has helped fund new cystinosis research this past year totaling nearly \$200,000 in several areas. One study will aim to develop a better tolerated and more efficient form of Cystagon through viral-mediated gene transfer. Another seeks to design and synthesize new forms of Cystagon chemically that will be odorless and tasteless, some of the most bothersome side effects of the medication. The third study will establish a tissue repository using kidney biopsies from cystinosis patients; these samples can be utilized for a wide range of research into the disease.

Even though Jack continues to do well, he must still contend with some issues. His right ankle and foot cock out, causing challenges with his walking and running. He will always fight a battle with hydration and presently still has more protein spilling into his urine than is ideal, which can hinder muscle and growth development. For example, while Jack's height and weight are both surprisingly good, his weight has really leveled off this past year, which has prevented us from considering removing his G-tube implanted through his skin and stomach wall. Additionally, crystals are clearly building up in his eyes as is evidenced by his growing sensitivity to sunlight. Unfortunately, Cystagon does not enable cystine depletion in the corneas (blood does not flow there). Fortunately, eye drops exist that, when topically applied, remove the cystine crystals there. Unfortunately, the drops must be taken every hour while awake and they sting considerably. Ah, the ups and downs of rare disease health maintenance! Through all of his trials and tribulations, it bears mentioning that Jack made a major contribution to the healthcare of his family this past year when he gave his big sister, Alex - one who would never be described as brave - a pep talk that shots of her own would not hurt when she needed to receive an IV (needle) for a medical procedure. Jack went so far as to show her how to roll up her sleeve and point out where the needle would go. If she ever needs advice for swallowing a pill, using a syringe for a G-tube, chugging a bottle of water, or giving a urine sample, she has the right brother for those, too.

How far have we come in the first five years of our journey with cystinosis? After being diagnosed in spring 2001 at a year old, the first 18 months saw Jack vomiting 10-15 times per day, two hospital stays, dramatic lack of growth, and countless tests and treatments from doctors and specialists in pediatrics, gastroenterology, nephrology, genetics, nutrition, radiology, endocrinology, and ophthalmology. While Jack still has a bevy of doctors that he visits quarterly, he has made tremendous progress also through work with speech, physical, and occupational therapists, plus marked scholastic improvement through his IEP (individual education program) plan with our county government and local school district. He continues to progress well in school and stands to not have the profound learning challenges of many kids with cystinosis, which buffs our enthusiasm for Jack's development when combined with his two above average intelligence tests to date. Jack

also has numerous friends and participates in a slew of activities with them, ranging from play dates, soccer, summer day camp, and T-ball, where he played on the Cubs last spring. (Note: In addition to being happy that his dad was his manager, Jack proudly proclaimed that Sammy “Salsa” was his favorite player. Trust us; the fairly stubborn Jack cannot be convinced to pronounce it any other way. Yes, perhaps he received more than the recessive gene for cystinosis from both of his parents. ☺)

For our family, like George Bailey, we know we have been blessed in many ways – family, friends, doctors, modern conveniences, and contemporary science, to name a few. Some simple bells metaphorically remind us of these blessing on a daily basis; whether the UPS delivery man ringing the door bell after a Cystagon delivery, an alarm clock going off for medicine, an incoming email from the CRN internet support group, or fundraising ringing the register of research. This last example calls for so many of our family, friends, and colleagues to take bows as we have raised almost \$80,000 for cystinosis research through our first three letters alone. This kind of support for Jack and the Cystinosis Research Network is humbling and we thank everyone for their compassion, indulgence, and consideration with this current letter.

In another seemingly simple situation, we were reminded that Jack and cystinosis is part of our life. Recently, when Jack attended a buddy’s Tae Kwan Do birthday party, Dave was hoping to discreetly take Jack aside from the festivities to an adjacent empty room to give him his 2:00 pm meds. Quite perturbed about being removed from the action, Jack looked at Dave with his chin out, hands turned, and palms faced up and said, “Dad, it’s okay. Right here.” Dave quickly read Jack’s mind – “Dad, I take medicine all the time and have done it for five years. All my friends know I need medicine. They know I have a “tube” in my stomach. I do not know any other way. Let’s go and get this over with...there are some balsa wood boards that need to be chopped!” Immediately, the bell went off in Dave’s head...this kid is going to be just fine, especially given the help, support, and love so many have demonstrated. Thank you all for helping us to make a difference. God Bless.

Sincerely,

Dave “Jack’s Dad” Greeley

Christy “Jack’s Mom” Greeley

As a nationally recognized 501(c) (3) advocacy group, your donation made payable to the Cystinosis Research Network is fully tax deductible. To be part of Jack’s 6th Birthday Celebration, please send your check made payable to CRN directly to Jack as part of a group donation or forward to CRN:

Jack Greeley
302 Whytegate Ct.
Lake Forest, IL 60045
(847) 482-0857
greeleycd@aol.com

OR

Jack’s 6th Birthday Celebration
c/o Cystinosis Research Network
10 Pine Avenue
Burlington, MA 01803
(866) 276-3669
crn@cystinosis.org
www.cystinosis.org



Neighbor

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SECTION 5

FRANK & LINES
BY BOB THALES

'Our greatest blessing'

5-year-old Jack Greeley battles cystinosis with parents in his corner

BY JANET DERALEAU
Daily Herald Correspondent

Cystinosis is a big word for a little boy like Jack Greeley. Although the 5-year-old from Lincolnshire doesn't have to pronounce it, he does have to live with it.

His understanding of the rare genetic disorder comes down to the plastic feeding tube protruding from his stomach and the awful-tasting medicine he must take every six hours. He doesn't remember when as a baby he vomited food — sometimes 10 times a day.

At nine months, prior to the diagnosis, he was thriving at 24 pounds, said his mother, Christy Greeley. A few months later, his weight had plummeted to the 10th percentile on the growth charts.

But in the world of cystinosis, Jack is lucky. He was diagnosed correctly and early by a doctor who understands the illness. His body is able to tolerate a miracle medicine called Cystagon that flushes most of the buildup of the amino acid, cystine, out of his cells. Without the drug, people develop kidney failure and require transplants to survive.

The cystine buildup also causes a host of complications, including diabetes, blindness and muscle wasting. Approved by the Food and Drug Administration in 1994, the drug is a lifeline for cystinosis sufferers, despite the ordeal of having to take the medicine for



Here is some of the medicine that Jack Greeley has to take on a daily basis.

the rest of their lives. "The reality is that Jack's medical care and his acceptance of Cystagon treatments have essentially commuted his death sentence to a life with a chronic disease, which is our greatest blessing," wrote Jack's father, Dave Greeley, in a letter to relatives and friends announcing Jack's fifth birthday and asking for donations to the Cystinosis Research Network. "Beyond the medical end of things, we see our good fortune in so

many ways through Jack's development. He is a normal 5-year-old, just less healthy than most."

In true 5-year-old fashion, Jack likes Pokemon cards and talks about his friends in his class at Tamarack Preschool in Lincolnshire.

He has a fondness for his cat, Murphy, and his 8-year-old sister, Alex. His sky-blue eyes twinkle beneath a head full of silver-blond hair. Surprisingly, his eye and hair color are characteristic of those with cystinosis.

Jack's health requires constant monitoring. He sees a team of 14 doctors, therapists and others who help him manage his illness. At the helm is his mom, who keeps her family's life and Jack's health routine on schedule. In addition, she serves on the Cystinosis Research Network board of directors.

"Christy is my hero," said Colleen Hammond, founder of the Cystinosis Network, forerunner of the Cystinosis Research Network, headquartered in Massachusetts. Hammond's 15-year-old son also has the disorder. "She cares for Jack 24 hours a day, but she finds time for the Network."

With a master's degree in public health and experience in pharmaceutical research, Christy Greeley can talk the talk with cystinosis researchers. She was instrumental in the creation of a National Institute of Health symposium that brought scientists together to share their findings about the disorder.



Jack Greeley, 5, Lincolnshire, smiles with his mother, Christy. Jack has a rare disease called cystinosis. Jack's parents are active in trying to raise research money to fight the disease, which primarily strikes children.

Dave Greeley, who works in sales and marketing for the Chicago Bears football team, also serves on the board of directors using his talents to help the network better sell themselves.

"His marketing skills have been a godsend," Hammond said. "We have gone from a mom-and-pop group to a corporation."

Like all of the network's parent volunteers, the Lincolnshire couple are driven by trying to save their child's life, Hammond said.

"It's almost painful for me to ask people for money," Christy Greeley said. "But if we don't do it, it's not going to get done."

The Greeleys started their letter-writing campaign in honor of Jack's third birthday and raised \$18,000 through donations. Last year, that figure rose to

\$37,000. In addition, the community took the cause to heart with Jack's preschool raising \$5,000 at a family fun day and another group making the cystinosis organization the beneficiary of their golf outing.

Fifteen years ago, children with cystinosis routinely died by the age of 10. Hammond calls Jack and his generation pioneers.

"Because of improved treatments, we can be as optimistic as possible," she said. "We hope and pray they will have the same future as healthy kids. The reality is: We don't know."

For information on the Cystinosis Research Network call (866) 276-3669 toll free or visit the Web site at www.cystinosis.org.

Call Dave and Christy Greeley at (847) 482-0857.