

February, 2003

Dear Friends & Family,

Your help is needed. On March 17, 2003, the Greeley family will be celebrating a milestone, the third birthday of Jack David. With Jack's birth on St. Patrick's Day 2000, we received a gift of "cheers." When Jack's sister, Alex was born on Thanksgiving Day in 1996, we were blessed with a gift of "thanks." Two angels from Heaven combined to give us so much to be thankful and cheerful of; no one could ask for more, then, the mysteries of life kicked in.....

Jack's first year was great – a big, happy beautiful boy, who ate, cried, slept, and played much like his sister. The Year 2000 was relatively uneventful and Jack was a perfect addition to our extended family in Portland, OR during that three-plus year phase of our lives.

In November 2000, we moved to Chicago. As Jack grew, he developed a signature move of scootching across the floor on his bottom to grab hold of things, but never crawling. Plus, he drank a lot like any big, scrappy lad should, so it was logical that he would wet diapers like crazy. It all made sense to us, because Jack was a BIG kid, so he adjusted his mobility to fit him and he pee-peed a lot, no big deal.

Then, things started to change in March – April 2001. Jack stopped eating; it wasn't picky or fussy, he *stopped* eating. All he wanted to do was drink. Then, another dynamic developed; he started to vomit regularly and we added a twist to Newton's theory of relativity...what goes in, is going to come out. *Everything* we gave Jack came back up. It became a mathematical exercise; put more in then came out so that he could retain something. If the "vomit catch" became an Olympic sport, Christy and Dave would have brought home the Gold!

Jack's odyssey in life truly began during those early spring days. Quickly, our pediatrician led to a pediatric gastroenterologist. Then, add in a geneticist, a pediatric nephrologist, and a pediatric ophthalmologist, all of whom put Jack through so many tests – the picking, the prodding, and the probing of every surface area and body part. You name it, Jack saw it and suffered through it; being strapped to backboards for x-rays and cat scans, tubes in every orifice, biopsies where bodily tissue was taken from him for lab work, and connections to many machines. We sat in doctors offices and went down the check list of what Jack's illness could be; cancer – nope, cystic fibrosis – nope, diabetes – nope, plus so many other obscure diseases.

As the events unfolded, we remember vividly a moment of comic relief. Prior to some radiology work, Jack had to drink a radioactive, barium milk shake...sounds yummy, huh? The radiologist warned us that most kids his age refused to drink it, so they would likely have to insert a tube through Jack's nose into his tummy, forcing the barium inside him. Remember, here was a kid who lived to drink, so without batting an eye, he guzzled about 12 oz. of this foul solution in-between screams and it gave him a brief moment of pleasure before being strapped to a backboard! Dave cried hard that day, but today he laughs about it.

Within six weeks, which is still a miracle to us, Jack was diagnosed with Cystinosis (pronounced SIS – TIN- OH – SIS), a rare metabolic, genetic disease. Dr. Sandler, Jack's gastroenterologist, nailed it with a team of other amazing doctors; to this date, we think he is Jack's guardian angel because identifying this disease is like finding the proverbial needle in the haystack and the chances of getting it are less than that of winning the Lotto. Some kind of luck, for sure!

Anyway, cystinosis is a terribly complex disease passed along to a child when both parents carry a recessive gene. The fundamental problem with Jack and all others with cystinosis is that they cannot process an amino acid called cystine. When cells in the body cannot rid themselves of cystine, it forms destructive crystals and from there a large ripple effect is cast. The major effects of cystinosis include: huge thirst/dehydration, profuse urination, swallowing difficulties, failure to

thrive (growth problems), muscle weakness, Rickets (bone disease), gastrointestinal problems (including frequent vomiting and reflux), photophobia (corneal sensitivity to light that affects the ability to see), and kidney malfunction, which almost always ends in end-stage renal failure where dialysis or transplantation is a must.

So, how has life been the last two years? Certainly 2001 was rough, really rough, but life is good and it gets better everyday for us, especially Jack. (Note: Life gets better too later this year when the Bears finally open the new Soldier Field!). Clearly though, the word "normal" has taken on a new meaning. Jack presently has a gastric feeding tube inserted into his stomach just above his belly button through which we can give him liquids, nutrition, and medicine. It should be there for a few more years at least, but it has enabled us to efficiently give him feedings and medicine every night at 2:00 AM. When Mom and Dad say "Good Night" now it is invariably mentioned with a simple question being asked – "Whose turn is it to get up?" Fortunately, Jack's everyday six-hour medical routine has enabled him to grow surprisingly well and maintain pretty decent body chemistry. Much of this good fortune has come through Christy's near full-time role as Jack's primary medical care-giver and administrator. Weekly trips to physical therapy and speech therapy with almost certain insurance company battles and routine doctor visits are now coupled by only occasional puking episodes. "Normal" for us is now getting kicks out of seeing some wild reactions from any random adult in public when they see Jack guzzle a 20 oz. bottled water or at diaper changing time when a little kid in a play group perhaps stares intently at the little spout coming out of Jack's tummy. "Normal" is a very relative term and our boy is certainly normal, but just not too healthy.

Everyday since that diagnosis, perspective became the battle cry of our daily lives with cystinosis. Part of our perspective comes from knowing more about cystinosis. We have been blessed by working with the Cystinosis Research Network (CRN), an all-volunteer group of parents from coast-to-coast who have kids with the disease. Not only has this group educated us and supported us in countless ways, but they have also given us great hope for Jack and his future. Please take a look at their web site, located at www.cystinosis.org.

In the past six months, we have taken positions on the CRN's Board of Directors. Dave is on the CRN's Executive Committee as the group's Vice President of Research & Development, while Christy chairs the Research Committee. The set-up is kind of like home life – you know, Dave is the "head of the household" and presumably the boss, but Christy is really in charge and does all of the work!

After a wonderful and dedicated family from Burlington, MA launched the Cystinosis Research Network in 1996, the CRN has taken on an amazing transformation in the past several months with an even brighter future. In its quest for improved treatments (and eventually a cure) for cystinosis and to help families deal with the effects of the disease, the CRN works to:

- Build relationships with the medical community (doctors, researchers, pharmaceutical companies, medical schools, etc.)
- Support families with medical, developmental and other emotional matters
- Lobby the government for support of rare disease research
- Form strategic alliances to position the CRN for future growth
- Raise funds and awareness to enhance research on cystinosis

So, what does the future hold? For Jack personally, so much could happen. Potty training difficulties, childhood taunting, life-long medicine regimens, special education and/or other accommodations at school, kidney transplantation, doctors, therapists, hospital stays, the challenges of preparing for an independent adulthood – all of this is a partial list of what could be on the horizon. This story, however, is not one of despair, rather one of hope for Jack and us. With the CRN and others committed to battling cystinosis and other rare diseases, the future is full of promise.

Presently, there are some brilliant doctors and scientists who are committed to fighting cystinosis, highlighted by hospitals and labs at the National Institute of Health (NIH) in Bethesda, Maryland, Tulane University in New Orleans, and the University of California-San Diego. Other medical professionals in Europe are also studying cystinosis. These scientific “miracle workers” have been able to synthesize a drug called Cystagon; even though it is harsh on the body and smells horribly, it is Jack’s No. 1 medication as it helps his body remove cystine from its cells. Moreover, they have been able to find the gene that causes cystinosis and have discovered how to inflict mice with this disease so that new treatments and the effects of the disease can be better studied. Some researchers have also examined the wide-range of developmental effects on kids with cystinosis (i.e.– cognitive, digestive, neurological, gastrointestinal, muscular, etc.). Others are on the brink of developing new formulations of Cystagon which could cause fewer side effects and mean the medication would last longer in the body (taking it two or three times a day instead of four, so no more 2:00 AM wake-up calls and fewer compliance issues). All of these efforts will aid rare disease research in general and ultimately help 25 million Americans who suffer from a variety of these rare inflections, like cystinosis.

With such promise and hope, Dave’s goal of kicking some “cystinos-ass” is plausible, but so much more needs to be accomplished. The limitations of these researchers and the CRN are only due to a lack of financial resources. This is where your help is needed. In celebration of Jack’s birthday next month, we ask that you consider sending him a gift in the form of a donation to the Cystinosis Research Network. Whatever amount that works for you and your family in these uncertain times works for Jack and the CRN. Since the CRN is a nationally recognized 501(c)(3) organization, your donation would be fully tax deductible.

Starting this year then, your support of Jack and the CRN can begin by sending a check payable to the “Cystinosis Research Network” to us in Chicago. We will make a group donation to the CRN in Jack’s name and the CRN will send you a statement of your tax deductible contribution. We also respectfully ask that you circle March 17 on your calendar and remember each St. Patrick’s Day as not only “**Jack’s Birthday,**” but also a date to send “cheers” to those around you who might be less fortunate. As for us, this is the beginning phase of our efforts with the CRN and we hope that when you consider your annual giving and donations to organizations of interest to you, you can please give special consideration to the CRN. We look forward to your assistance with our cause as things develop over time.

Again, this is not meant to be a sad story. So many horrible things exist in today’s world and this is merely part of our family’s journey. To fully understand the total who, what, when, where, why and how of this journey is beyond our capabilities and rests in the hands of God almighty. To grab strength and perspective on a much more comprehensible level, reflecting on a line from a wonderful movie called “The Shawshank Redemption” helps. The film stars Tim Robbins as Andy DeFran, a man with an unbeatable soul and spirit who is wrongly imprisoned for the murder of his wife and her lover. The line simply is “You can get living or you can get dying”...we choose to get living and we would appreciate your support of Jack and others who suffer from cystinosis.

Sincerely,

Dave “Jack’s Dad” Greeley

Christy “Jack’s Mom” Greeley

P.S. Please hug and kiss your family everyday and tell them that you love them.
