



Up Close and Personal

“This is a message of love. Love that moves from the inside out, love that never grows tired. I come to you with strange fire.” INDIGO GIRLS

BROOKE'S CORNER

In early June, we excitedly headed to Como Zoo with our South Dakota guests. Once inside Como Town, the girls hit Hodge Podge Park while I found the nearest bench, pulled out my phone, and started to catch up on a few things.

When I heard Brooke's shout to “watch me do this!”, I glanced up and saw 4 inelegant limbs struggle to make their way across the monkey bars. The site inspired me to snap a picture and then put my phone, with its half-completed grocery list, away.

Just as my eyes returned to Brooke, I caught the furtive glance she sent my way. As our eyes met, the face that had once held a look of deep concentration exploded with shear delight! It was clear that nothing mattered more to Brooke in that one brief moment than sharing her accomplishment with me.

How many of these glances have gone unnoticed in the past with my head buried in work, school, and event planning? I'll never know. But as I try to turn from my self-seeking ways, and focus on making my family top priority, I hope they'll forgive me for each and every one I missed.

It was twelve years ago now that a *Lutheran* pastor told me, “No.” Paul and I were picking out music for our wedding service, and I had finally convinced my alternative rock fiancée that “Me and You” by Kenny Chesney was the *perfect* song! I excitedly announced “our” selection to Pastor Steve Sampson, and he shook his head and said, “I'm sorry, but I can't approve that song.” My furrowed brows and dropped jaw must have hinted at my indignation, for Pastor Steve gently explained, “That song only talks about you and Paul. In order for your marriage to last, you'll need more than you and Paul—you'll need God, too.”

I look back now and wonder how many ugly, painful moments could have been avoided in our marriage had I acknowledged the wisdom Pastor Steve tried to impart all those years ago...

In the beginning it was so easy! Paul was the yang to my yin. I couldn't stop thinking about him. Then I got him, and let's just say, I had extra time on my hands once the getting was done. Naturally, I spent the extra time thinking about...me. *My* job, *my* problems, *my* needs, and *my* wants.

After a few years of marital bliss, I *wanted* kids. Once Paul

had helped me fill the bedrooms upstairs with pooping, screaming machines, my attention quickly shifted back to...him: *his* irresponsible behavior, *his* lack of understanding, and *his* selfishness.

Then, one bright, sunny summer day, Paige was diagnosed with cystic fibrosis (CF), and my attention shifted yet again. What little time we had left after working all day and caring for two little girls all night, was now spent doing medical treatments. I had nothing left to give Paul at the end of each day. On “good” days, I knew I loved him, but was too tired to do anything about it. On “bad” days, I felt bitter, resentful, and certain my life would be better if Paul wasn't in it.

Then, while shopping for airplane material one day, I stumbled upon “The Love Dare”; a book that teaches unconditional love. In an effort to bring that loving feeling back into our relationship, I followed each of the daily “dares” the book gave me. Dare 21 challenged me in a new way: “You may have realized that nothing can repair the damage sin leaves. Every day you place expectations on your spouse. Sometimes they meet them. Sometimes they don't. But never will they be able to totally

satisfy all the demands you ask of them—partly because some of your demands are unreasonable, partly because your mate is human. God, however, is not. And those who approach Him in utter dependence each day for the real needs in their life are the ones who find out just how dependable He is. Can your spouse give you inner peace? No. But God can. Today's Dare: Make time to read your Bible and pray.”

Later that week, when bitterness and resentment came knocking at my door, Dare 21 came to mind. Through clenched teeth I reluctantly prayed and asked God to show me what *He* loves about Paul...and you know what? He did. Thoughts of Paul's supportiveness, loyalty, sense of humor, and relationship with the girls quietly crept into my icy heart. As my heart warmed with love and forgiveness, perspective flooded in. It was not my job to judge or change Paul, and it was not his job to save me when this world left me feeling lost, empty or overwhelmed. Both jobs belonged to God.

To Pastor Steve: thank you for planting the seed. To Paul: thank you for being my best friend, my teacher, my fan, and the love of my life (quack, quack).

Issue
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With One Breath

TOGETHER WE CAN BEAT CF.

“YOUR KIND AND GENEROUS SUPPORT IS LIGHTING THE WAY IN OUR SEARCH FOR A CURE!”



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You Are Our Sunshine

“You make [us] happy when skies are gray.” JIMMIE DAVIS

Six years ago, a handful of our friends and family sat around our kitchen table to start a fundraising event that would help us find a cure for our daughter, Paige. We didn't care how much money we raised or how many people attended our event; it was simply a labor of love. We wanted Paige to know she would never have to face cystic fibrosis (CF) alone.

Never did we imagine that by the year 2011 our little grass roots effort would develop into an event attended by more than 500 people and raise more than \$50,000 for CF research in a given year! None of us could have predicted that more than 240 businesses and individuals would come to our aid during an economic crisis by sponsoring the event, providing an item for our silent auction, or making a cash donation. And never would we have dreamed that The Billy's, Black Marauder, Roger Clyne, PH, and fans would travel from all over the state and nation to help take our humble little soiree to the next level.

Whether you joined us this year, or have been alongside us since the beginning, we thank

you for choosing to pour your compassion on the more than 30,000 individuals and families in this nation battling CF. Your kind and generous support is lighting the way in our search for a cure!

When we started our journey as CF parents, “the unknown” haunted us. Our life's pursuit was to do anything and everything we could to stop CF's final and devastating blow from leveling our Paige. But thanks to your steadfast love and support over the years, our journey has now shifted. Instead of spending our spare time and energy fighting death, our family now has the courage and confidence to focus on living life. In order for us to do this, we find that we must let go of our WITH ONE BREATH event.

Have no fear, we will *always* be committed to raising money for the CF Foundation's unbelievable research program! We'll see what we can cook up for 2012, but in the meantime, we thank you for everything you've given us over the years. The sunshine you bring into our lives truly lights our darkest hours with hope...so please don't take our sunshine away!



Roger Clyne
and fans

Photos by Justin Yunke of
PragmaticPhotography.com

"Today I lie here thinking of what to write...what profound statements can I make to my daughter the day I learn she has cystic fibrosis? I can't say the diagnosis is a surprise; after all, I'm the one that made the doctor run the test. I can say that hearing confirmation of the diagnosis has me doubled over gasping for air. I feel numb inside. I have heard people tell me that I may have to watch my daughter die today.

I am so sorry you have been chosen to bear a weight this great, Paige. My thoughts are everywhere. Part of me wants to hold you and shield you, while another part of me feels a resolve to be firm and cut you no slack—as if it might prepare you for what is to come.

When I read about all of the additional therapies, drugs, and dietary guidelines we will now need to follow each day, I feel tired. I honestly don't know if I have what it takes. As a working mom I already feel average at best, and now you need me to be better than average.

I hope and pray I don't let you down, daughter. I am so scared of losing you, Paige. I don't know if I'd be able to live without you. I will pray every night that God smiles upon you. I love you."

The CF Foundation is extending lives in their search to cure and control cystic fibrosis!

With the new treatments being developed in the Foundation's drug discovery pipeline, hope for the future is at hand.



"THIS IS A HISTORIC TIME IN THE FIGHT AGAINST CYSTIC FIBROSIS." BOB BEALL

Since Paige's diagnosis, you have helped the Cystic Fibrosis Foundation (CFF) develop 6 new weapons in their fight against CF. Better yet, an additional 7 new treatments that seek to cure or control CF are in the CFF's drug discovery pipeline right now; anxiously waiting their opportunity to hit the battlefield. The CFF has made the following unbelievable, unprecedented progress in the past year thanks to devoted volunteers and donors like you:

TARGETING THE BASIC DEFECT

The Phase 3 clinical trial of Vertex Pharmaceutical's VX-770 has shown profound results yet again. In the study, those receiving the drug demonstrated marked improvement in lung function (the best seen in any clinical CF drug trial to date!), significant improvements in sweat chloride levels, and weight gain compared to those on placebo. These results indicate that VX-770 is able to restore the proper flow of salt and fluids into the airways; targeting the basic defect in CF. These results are so incredible that Vertex is currently working with the FDA to allow CF patients

with the G551D mutation, who are in critical medical need, access to the drug prior to FDA review and approval!

The CF community also awaits the Phase 3 results of another exciting CF modulator, known as ataluren (formerly known as PTC124). An earlier, smaller study showed that CF patients who took ataluren had a correlating improvement in lung function. Both of these exciting drugs have the potential to stop the progression of CF in up to 15% of the CF population since they target the defective gene.

TREATING THE SYMPTOMS OF CF

Respiratory problems, resulting from frequent lung infections, remain the most serious complication for CF patients. In February, Gilead Sciences, Inc. released a new FDA-approved inhaled antibiotic called Cayston®. Cayston® offers a much-needed and highly effective alternative for those battling persistent, life-shortening lung infections caused by an antibiotic-resistant bacteria known as *Pseudomonas aeruginosa*. In addition, Bronchitol (developed by Pharmaxis) received "fast-track status" by the FDA due to the compelling results



WAY TO GO, PAIGE!

Paige raised **\$1047.89** for CF research this summer by hosting her first garage sale! A huge thanks to everyone that contributed!

of two long-term Phase 3 clinical trials. Bronchitol, an inhaled dry powder, is designed to restore normal airway clearance and reduce the buildup of mucus. Results reported that CF patients receiving Bronchitol showed significant improvement in lung function and had fewer respiratory problems.

NUTRITION

In CF, thick secretions build up in the pancreas and prevent the release of important enzymes that help with digestion. About 90% of people with CF take pancreatic enzyme replacements to help their body absorb the essential vitamins and nutrients necessary for growth. While enzyme replacement therapies have been in use for decades, FDA recently required that all pancreatic enzymes on the market be reformulated and undergo clinical testing to ensure their safety and efficacy. In April, the pancreatic enzyme replacement therapy Pancreaze™, by Ortho-McNeil-Janssen Pharmaceuticals, Inc, was approved under the new, more rigorous FDA testing and review procedures, and is now available for CF patients that do not respond well to the traditional enzyme replacement therapies.



Cepacia Scare

Burkholderia cepacia (*B. cepacia*) is a group of bacteria found in wet soil and decaying plants. The mere mention of this organism in casual conversation can fill those in the cystic fibrosis (CF) community with a sense of impending doom, or remind them of tragic loss.

B. cepacia is a rare but significant threat to those with CF. Once *B. cepacia* enters the mucus-filled, CF lungs, one of three things happens: 1) nothing, 2) the lungs become infected and deteriorate at an increased rate, or 3) the organism ignites "cepacia syndrome". In "cepacia syndrome", the bacteria spread violently through the

bloodstream and cause severe lung problems, fever, and a rapid decline in health.

The marked difference in prognosis among individuals infected with *B. cepacia* appears to be attributed to the 8 known strains of the bacteria. With the milder strains, like *B. multivorans*, early and aggressive treatment with antibiotics may eradicate the organism, or at the very least, slow the deterioration of lung function from approximately 13% per year to 7% (1-2% per year is expected with CF).

With the most dangerous and devastating strain of *B. cepacia*, known as *B. cenocepacia*, infected individuals are

not expected to live more than 100 days after contracting the organism. This is due to the fact that "cepacia syndrome" is resistant to treatment with antibiotics.

Since *B. cepacia* is usually transmitted from one person with CF to another, infected individuals must stay away from others with CF. Thus, those with *B. cepacia* are not permitted to attend CF fundraising events, and are seen in separate clinic and hospital areas. In addition, the presence of *B. cepacia* typically precludes an individual from receiving a potentially life-saving lung transplant. Thankfully, it is estimated that fewer than 2.7% of the CF population will ever have to encounter a *cepacia* scare.

GREAT STRIDES in St Paul

Over the course of a lifetime, people come and go. But every once in a while, a select few weave their way into your life and become perma-



nent fixtures. These individuals stand beside you through thick and thin (and tornado warnings!); asking nothing in return.

Thanks to several of our true-blue friends, team WALK FOR PAIGE raised \$2200 at the Cystic Fibrosis Foundation's GREAT STRIDES event in downtown St Paul this year. Thirty-five of our permanent fixtures even braved thunderstorms and severe weather advisories to be at Paige's side on her annual trek for a cure. Of the 60 teams registered, ours was one of only 25 teams that

actually showed up to participate!

Our group's adventurous spirit paid off; not only did the weather hold out, we also ended up leaving with backpacks full of free goodies and hearts filled with hope. In two short hours, we helped the CF Foundation raise more than \$200,000 for CF research!

Our family knows that your unwavering support is part of what keeps Paige so healthy. Thank you for loving her like we do, and for being there to help her weather the storm.

CF MISSIONS

Helping Paige Beat CF

When cystic fibrosis (CF) strikes, Paige's eyes fill with pain and anguish. When those eyes turn to us and plead for help, we can usually do nothing but hold her. We try so hard to "get better" at watching her fight and endure, but it never gets easier. When chronic disease makes its move, it renders us furious, defeated, and terrified. But then we remember the \$240 the Rule's & Friedner's raised for CF research last Fall, and realize Paige will smile again.

We remember the \$1300 Jim and Angie Fritz raised for CF research this past Winter at their St. Patty's Day party, and know the sparkle in Paige's eyes will return.

We remember the 4 kids in our neighborhood that trained for a 1-mile race with us because "it helps Paige's lungs", and understand that CF will never stop Paige from living the life God intends her to lead.

