February 24, 2016

Senator Peter Courtney
Oregon State Capitol
sen.petercourtney@state.or.us

Dear Senator Courtney,

I am writing to you today as the Executive Director of Cystic Fibrosis Research, Inc. (CFRI), a 40-year-old cystic fibrosis (CF) nonprofit patient advocacy agency based in Palo Alto, CA, but with constituents across the world, including Oregon. CFRI applauds and supports your efforts to designate May as "Cystic Fibrosis Awareness Month." It is vitally important to this physically and emotionally fragile population that their voices are heard. By designating May as "Cystic Fibrosis Awareness Month," the State Legislature will increase public awareness of the nature of the most common fatal genetic disease in North America and the many debilitating challenges faced by those diagnosed with CF and their family members.

As I say, many times a day, "I hate cystic fibrosis." I hate that it begins to take the breath away from our children the day they are born, and ravages their bodies until they can fight no more. I hate that they suffer excruciating pain from endless lung exacerbations and gastrointestinal problems; that there is no cure, and that the fate of so many is to die from drowning in their sputum or blood from hemorrhaging lungs… or from lungs that simply cannot take another breath.

SB 1522 can be a significant step in educating the public about the realities of life with cystic fibrosis. CF is an orphan disease, a rare disease impacting less than 200,000 people in the U.S. With approximately 30,000 people in the U.S. diagnosed with CF, the need to heighten understanding of this complex disease is critical for the health and well-being of this population. Just because the numbers are small, doesn't mean the disease is insignificant. It is a painful, progressive, expensive, and deadly disease that drains the life from those with CF day by day. It is heartbreaking that the median age of death for an individual diagnosed with CF is in the mid-20s. And, these are not 20 years of a gentle and healthy life – these are 20 painful years of increasing hours spent each day waging a battle to slow the progression of the disease. This requires multiple sessions of daily respiratory therapy, dozens of daily pills, frequent rounds of IV antibiotics, at times - supplemental oxygen, and many hospitalizations. For those with advanced lung disease, transplantation is the only option to prolong life, but due to a variety of factors, including a shortage of organs, many do not survive the wait. And the survival rate is not impressive. New drugs and a cure are desperately needed to end this vicious cycle. Yes, the past few years have brought remarkable new therapeutic drugs that are changing the course of the disease for so many. But for many, the drugs have not come in time.

How does one acquire CF? Both parents must be carriers of the mutated gene to pass it on to their children. For each pregnancy, there is a 25% chance that the child will have CF, a 50% chance that the child will be a silent carrier of one copy of the gene, and a 25% chance that the child will not be a carrier, and not have CF. One in 29 Americans is believed to be a silent carrier of the CF gene. I am a carrier, but the odds are that there is at least one other carrier in the room that will be hearing about SB1522 today.

The basic issue with CF is a defective exchange of fluids across cells, caused by a mutation in the gene for the CFTR protein. Secretions that are usually thin become thick and sticky, clogging ducts and passageways, especially in the lungs and pancreas. In the airways, this thick mucus creates a chronic state of inflammation, and a perfect breeding ground for opportunistic infections which destroy the airways, often leading to respiratory failure and death.

While most people associate CF with lung disease, it is systemic, impacting the sinuses, pancreas, and reproductive systems.

My role as ED of CFRI is extremely rewarding, but it is my role as the mother of a 31-year-old daughter with CF that lights up my world. She is why I have dedicated my adult life to fighting the disease that is constantly trying to kill her. To better understand life with CF, I would like to share excerpts of an article I wrote that was published by the Huffington Post in October 2014 –

Cystic fibrosis came to stay on Good Friday 1986 at 5 p.m. when my toddler was 22 months old. "Cystic fibrosis?" I said to the voice on the phone. "What is that?" The voice that I'll never forget responded coldly, "She'll be dead before she's five. Call a specialist." And the line went dead. And our world crumbled.

Cystic fibrosis. The most common fatal genetic disease in North America. Those born with it live through endless bouts of painful lung infections, liver damage, CF-related diabetes, and severe and painful gastrointestinal complications. It is debilitating to say the least.

Oh the joy when the gene that causes the thick mucus which clogs up the airways and passages in other organs was discovered! Now -- all of our friends said -- she'll live! Wait, said those of us with our ethereal children. There is no cure yet.

We turned to CFRI for support and information and to CF specialists for ongoing care of this progressive disease. To gain weight, our daughter needed to take pancreatic enzymes with meals because the thick mucus blocked the ducts in her pancreas, thus preventing the release of her own enzymes to digest food. We never missed a day of multiple inhalation treatments and chest percussion therapy, so that her lungs would remain open and give her the precious breath that she needed to survive. Oral and IV antibiotics, and fungicides, became her best friends.

But at age 12, my daughter went into unexpected liver failure. She hemorrhaged and lost nearly half her blood supply. After weeks of endless painful procedures to save her, we neared the end. Mother's Day 1997 -- she had three days to live. And, miraculously, a perfect liver became available and she received another chance at life through a liver transplant.

Eighteen years later -- after graduation from UC Berkeley with highest honors, 11 years of marriage, countless life highs and health lows, and the discovery of nearly 2,000 CF genetic mutations -- she is still lighting up the world with her gentle nature, her passionate opinions about matters most important, and her continued fight to live with CF.

Now much can change with CF on a daily, weekly or monthly basis. Nine months after I wrote that article, our worlds were turned upside down once again. In 2015, my family spent the longest summer of our lives in the course of only four weeks. We endured all the challenges, agony and heartbreak that can come with loving someone with cystic fibrosis. These are the times when we cry out, "I hate cystic fibrosis," and feel helpless as we are dragged into the dark abyss of this destructive disease.

Yes, the past few years have brought remarkable new therapeutic drugs that are changing the course of the disease for so many. But for others, the drugs are not effective or their disease has progressed too far, like it had with my daughter.

My daughter with end-stage CF lung disease turned 31 in June. Two days after her birthday, she was admitted to her CF care center with a cold. Four days later she went into acute respiratory failure and was dying. Her husband had to make the decision to let her go, or to try the only option that might save her at that very moment: life support. **She could not breathe. She was in agony. Her lungs had failed.** We were told to be prepared to say good-bye. The thought that we might never see her beautiful green eyes again or hear the sweetness of her voice or feel the warmth of her embrace was excruciatingly painful.

We kissed and hugged her, and let her doctors do what they could. She surprised everyone. She lived!

After four painful and breathless weeks in the hospital's intensive care unit, she suffered another life-threatening setback, and at that very moment her transplant team walked in and said that they had accepted the perfect set of lungs. My daughter received her transplant the very next day. Afterward, I asked her surgeon, "How much time did she have left?" He placed a gentle hand on my shoulder, looked me in the eyes with compassion and said, "She didn't." The dark summer suddenly became brilliant with the joy of my daughter's new life. An incredible new season was about to begin: One of saying good-bye to her former life of living with ravaged CF lungs and of welcoming the endless and wondrous opportunities of living with non-CF lungs.

We had become accustomed to life with CF and the progressive nature of the disease. The emotional and physical burdens endured by our precious family members and friends living with this debilitating disease are enormous. Our family is now on a different CF journey. We are extremely grateful for this extraordinary gift of life and are counting our blessings!

Living with cystic fibrosis is a challenge. Our hope is that those living with CF will live life to the fullest, in spite of stormy weather. As the ED of CFRI and as the mother of my daughter, I thank you for sponsoring SB1522, for I know that by designating May as "Cystic Fibrosis Awareness Month," you will put a face on CF and your efforts will have a positive outcome on the approximately 400 individuals living with CF in Oregon.

You and your colleagues can make a difference in the lives of these extraordinary people, and I thank you for your time and efforts in recognizing the overwhelming challenges of this life-shortening disease. The Oregon Legislature will be better informed as a result of adopting SB1522, and can help with future issues such as providing immediate access to life-saving FDA approved drugs and therapy, affordable and reliable insurance, and even funds marked for CF research. Your decisions can mean the difference between life and death for this fragile population.

I hope I have been able to put a face on CF for you, and again, I applaud your efforts to designate May as "Cystic Fibrosis Awareness Month." If CFRI can ever be of assistance in the future, please do not hesitate to contact me.

Warmly,

Sue Landgraf
Executive Director
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