Dear Senator,

My name is Robyn Deleon. I am here today to address the importance of SB 452.

I represent the Urea Cycle Community of which I am Founder and Executive Director of Connecting Families Urea Cycle Disorders Foundation, [501(c)3], which helps families with this specific disorder. UCD's are rare and affect 1 in ~35,000 people. Our mission is to empower and engage all those affected by a Urea Cycle Disorder.

Urea Cycle Disorders are an inborn genetic error resulting in one of six enzymes in our liver that do not function either in part, and in many cases, not at all. It only takes improper function of one of the six in this cycle to result in ammonia (yes, like the industrial cleaner) to run through our blood stream which eventually results in brain damage, coma, and often ends with patients being forced to live with extensive brain damage, and even death. All of this can happen in a matter of hours. The warning signs are so subtle that untrained eyes likely won't know when it's time to go to an ER to get an ammonia blood gas drawn to find out your number. This number determines whether you will be admitted to the hospital, and due to the urgent need for treatment, many ER professionals are often skeptical or even suspicious of our illness because of the rarity. I can't even begin to emphasize the need for ER's and emergency professionals (EMT's, nurses, practitioners) to hear us when we tell them something is wrong. Time is critical for us and all too often we end up not only having to remain in our state of elevated ammonias, but we then also have to engage in a back and forth with emergency professionals who are too quick to suspect instead of listen and treat.

I am one of so many who have first-hand experience with this rare disease, and not because I asked for it, or even wanted it, but because I have lost two sons to this horrible and pernicious disease that could the ER's have been directed to act quickly, they may still be alive today. Too many newborn babies are still going undiagnosed, and too many emergency professionals do not understand the severity and need for immediate administration of treatment.

I am a carrier of this disease, and I currently have a 16 year-old daughter affected by a Urea Cycle Disorder as well. We both take a life-saving meds 8 times a day. The possibility of a hyperammonemic crisis is always a possibility with those suffering from UCD's. We need our meds, and we need to be heard. More than that, we need ER professionals to listen and be empowered to act quickly with treatment. The more swiftly treatment is administered, the higher the chances of our survival, because with each instance of elevated ammonia levels, there is no guarantee they will come back down. It's only by the grace of God that my daughter and I have survived this long and since the passing of my boys, we have made it our mission to help other families with the same illness around the country.

Have you ever been in a room full of people where almost all of them have had to bury a child or loved one because of a disease that they didn't ask for? I have, and it is heart breaking, and in some cases completely pointless because treatments were not administered quickly. Please help us get a head start on treatments by giving our ER professionals the authority to treat upon receiving our instructions we carry with us at all times. Almost every single UCD family I know receives treatment by a metabolic geneticist and a dietician specializing in these disorders. There is plenty of documentation and proof of our illness and hence the reason for the emergency letter with detailed instructions on how to treat us. But we seem to inevitably undergo major time loss during each crisis by spending hours of repeating history and saying the same thing over and over and we can spend hours upon hours in a hospital, but not receive treatment because **they aren't listening to us.**

I will continue to advocate for this community on behalf of the children that currently live with, and have been lost to their battle with this disease. It's because of them that I refuse to be silent about unmet needs in this community and I humbly, humbly ask for you to please hear us. We need you to support us in passing legislation giving ER professionals the ability to treat those with these types of disorders immediately upon learning of our diagnosis by the patient. We are seeking your help in establishing passing this bill for our first responders to be able to administer all medications and protocol stated in our emergency letters, which by the way, we carry with us at all times, and we still undergo major time delays, even when we are carrying signed documentation by our metabolic geneticists. This is why we need people like yourself to hear us, we are talking and no one is listening. People in the health care industry are supposed to act quickly in times in life or death situations. We are not being heard and we are losing too many children because of the delays in receiving treatment. Please hear us, please help us, and please do the right thing and help SAVE LIVES.

I thank you for your time today. We will hope and pray that you are able to help us make a difference to those who need this legislation passed, and on behalf of all of children and adults who have died from a Urea Cycle Disorder, we thank you for listening today.

Thank you for listening,

Robin Deleon
Executive Director
Connecting Families Urea Cycle Disorders Foundation