C.A.F.E. TEXAS Compassionate Access for Epilepsy

Jaxon

Jaxon Lane Huffman was born on December 17, 2010 at Ennis Regional Hospital. He is our second son and was perfectly healthy baby boy at birth.

When Jaxon was two and a half months old, we woke up to weird movements while he was sleeping. It lasted just under 1 minute. At first we did not realize what this was. After several occurrences of these movements while sleeping, we took him to Children's hospital in Dallas 4 times and they would not do anything. They kept telling us it was probably acid reflux. We finally started videoing these movements while he was sleeping. His pediatrician confirmed these were some form of seizures. He made a phone call and sent us to Medical City Dallas ER. They immediately admitted us to the epilepsy monitoring unit and set Jaxon up for EEG monitoring. Later that day Jaxon was diagnosed with partial simple seizures with an unknown cause at that time. Jaxon was prescribed Phenobarbital for his seizures at that time. He responded well to the medication, and continued to develop like any other baby boy would. He was seizure free on the medication, and we were hopeful that this was the end of the seizures for our son. What little did we know, was that this was only the beginning for Jaxon. Things would soon get worse.

When Jaxon turned 6 months old we took him in for his normal checkup and immunizations. A few hours later Jaxon started having seizures again. His neurologist at Medical City Dallas said that he had gained weight and that he needed to increase the Phenobarbital for it to be therapeutic. After 2 increases the seizures continued. We took a video in to the next appointment to let the neurologist review it. When Jaxon's neurologist reviewed the video he knew immediately was happening. He told us that these were no longer partial simple seizures, that Jaxon was having infantile spasms. I thought to myself well spasms have to be better than full blown seizures. I was completely wrong. He immediately sent us upstairs for an EEG to confirm the diagnosis. While waiting I did some research about infantile spasms. My husband and I were completely devastated when reading about the prognosis of kids with infantile spasms. The EEG confirmed that Jaxon was having infantile spasms.

An infantile spasm (IS) is a specific type of seizure seen in an epilepsy syndrome of infancy and childhood known as West Syndrome. West Syndrome is characterized by infantile spasms, developmental regression, and a specific pattern on electroencephalography (EEG) testing called hypsarrhythmia (chaotic brain waves). The onset of infantile spasms is usually in the first year of life, typically between 4-8 months. The seizures primarily consist of a sudden bending forward of the body with stiffening of the arms and legs; some children arch their backs as they extend their arms and legs. Spasms tend to occur upon awakening or after feeding, and often occur in clusters of up to 100 spasms at a time. Infants may have dozens of clusters and several hundred spasms per day. Infantile spasms usually stop by age five, but may be replaced by other seizure types. Many underlying disorders, such as birth injury, metabolic disorders, and genetic disorders can give rise to spasms, making it important to identify the underlying cause. In some children, no cause can be found.

Jaxon was immediately put on a medication called ACTH. This is a high dose steroid that was supposed to eliminate the infantile spasms. We had to give Jaxon IM injections 2 times per day for several months. At this time Jaxon had some genetic testing, metabolic testing, 2 MRI's and many other test and still no cause for his seizures were found. All tests came back normal. Jaxon's neurologist told us that since Jaxon's test were normal that this medication worked for 90% of all patients. We were sure this was going to be the answer to our prayer. Not only did the medication not stop the infantile spasms, it did not even decrease them. We were on the ACTH for months and Jaxon developed all of the horrible side effects of this medication. He had high blood pressure, a swollen face, was fussy, and gained a lot of

weight being on the steroid. Our neurologist tried several more medications including ones that could cause peripheral blindness, liver failure, rashes, developmental delays and many more before telling us that he did not have much experience with Infantile spasms and suggested that we find a neurologist that did.

We decided to go to Cook Children's in Ft. Worth. I had heard wonderful things from several of my patients about the neurologist there. At our first appointment we went through all of his records and what treatment options we have tried. Jaxon's new neurologist ordered another MRI since he was now almost a year old, and to see if anything had changed. Sure enough, the MRI showed that Jaxon had atrophy of his brain, and abnormality in the white matter in several areas. I cannot describe the feeling when we heard the news. As a parent you have these hopes and dreams for your child to live a wonderful life. Not only was our child not developing, but now we find out that his brain is not developing properly. All we knew to do from that point on was to pray. Pray that God would give us the strength to not be angry, and the strength to be strong for our son. We prayed that eventually we would find a treatment that would give our son relief from these seizures.

Jaxon has tried numerous treatments including ACTH, Prednisone, Phenobarbital, Sabril, Vitamin B5, Fycompa, Topamax, Keppra, Onfi, Banzel, Diastat, Klonopin, Vimpat, Clorazepate, Felbatol, Lamicatal, Depakote, the ketogenic diet, and many more treatments including chiropractic care. Jaxon has been on up to 4 seizure medications or more at a time and not one of them have even decreased the amount of seizures that he has per day. Jaxon participates in physical therapy, occupational therapy, and speech/feeding therapy 2 times each per week.

At age 4, Jaxon has very little head control, no trunk control, no use of his arms or legs, and is non verbal. He has no muscle tone, and is not able to sit, crawl, stand or walk. He has 20-40 cluster seizures every day which consist of up to 100 spasms per cluster at times. He now also has 3-5 grand-mal seizures daily and in some cases stops breathing. As Jaxon continues to get older, his seizures are continuing to get worse with no relief. Recently Jaxon had a swallow study done which confirmed he is now aspirating all liquids and foods. After much discussion with Jaxon's doctor, he was referred to palliative care. We have exhausted all treatment options for Jaxon at this time. We are asking for the opportunity for Jaxon to be able to try CBD. This is the only option left for Jaxon.