



Complex Child E-Magazine

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Living Your Best While Preparing for the Worst by Jamie Barnes

Hayden, one of my four quadruplets, has an unknown progressive syndrome that is slowly devastating his body. This is the story of how we have chosen for our family to live life: preparing for the worst while enjoying our life together.

Hayden's Story

After an intra-uterine insemination procedure, we were surprised to find out that all four ovums developed into embryos. But even early on we knew something was different about one: it was far smaller than the other three. By 20 weeks, we had named each baby and knew where each one was positioned. We named that little fighter Hayden.

The quadruplets were born after a relatively uncomplicated pregnancy at 32.4 weeks gestation. Hayden was the tiniest at 2 lbs 15 oz, but aside from jaundice, he was a healthy preemie breathing on his own just like his siblings: Peyton, Adrien, and Makenna.



Hayden's fight through typical preemie health issues was more significant as compared to his siblings. He took longer to put on weight, maintain his temperature, and nipple a bottle, but after 31 days he graduated from the NICU and was reunited with his sibling who had come home some 10-12 days before. Fast forward through 48 daily diaper changes and 24 bottles of pumped breast milk, and we began to realize the gap in Hayden's development and his siblings was becoming greater. We were referred to a neurologist, got lots of testing including an MRI, and then got a devastating blow. It appeared that the gray matter in Hayden's brain was shrinking. His neurologist didn't know if it would stop shrinking or what the cause was, since he had never seen anything like it.

Life with four babies was busy, and adding therapies and specialists to the mix made it even more so. Over time, Hayden's development remained static and severely delayed, but his health declined. He started having seizures, silent reflux, constipation and eventually respiratory issues. Hospital stays became more frequent with pneumonias, testing, and surgeries.

Making Tough Decisions

With each pneumonia infection, most often from aspiration, we had to brace ourselves for a possible end to Hayden's life. At some point we learned that it really wouldn't be the brain atrophy that killed Hayden, but most likely an infection such as pneumonia. In many hospitalizations he was intubated, and we were asked to consider a tracheotomy. This idea was presented many times with increasing pneumonias, severe reflux, and floppy airways. At some point during a routine follow-up with the pulmonologist, he presented the idea that both my husband and I needed to have a serious discussion to consider not only a tracheotomy, but also any life-sustaining ideas. He suggested doing this while Hayden was acutely healthy, as the implications and decisions have a different feel and may not be as well thought out when in crisis.

We had to be honest and consider how a tracheotomy would impact the lives of both Hayden and our other children. It was our opinion that Hayden's quality of life would be greatly diminished with the tracheotomy. To add more artificial means of support that could lead to even more pain and infections seemed to equate in our minds as more suffering. In addition, we didn't have any skilled nursing support, and to add on tracheotomy care to the already round-the-clock complexities of his care would be overwhelming for our family. It is complete mayhem trying to meet the individual needs of each child as it is, and his siblings were already unintentionally ignored at times.

After much discussion and fact gathering, we decided not to proceed with a tracheotomy. Physicians reassured us that no matter what we decided, there really was no right answer, and they would support us in whatever choice we made.

Just after the new year of 2009, Hayden had surgeries for a hiatal hernia, as well as a far-reaching alternative to a tracheotomy, which was to remove about 50% of his epiglottis tissue, since his airway was so floppy and causing apnea spells. Those surgeries were the start of a rough journey. Hayden developed life-threatening infections and bowel obstructions. After a total of 86 non-consecutive days in the PICU with sepsis, pseudomonas infection in his lungs, delays and extubation failures, and multiple GI surgeries, it was determined that he would benefit from continued nutrition given intravenously. It was adding another artificial means to sustaining his life, but specialists and parents alike assumed it would be temporary, and so we agreed to it.



Therapeutic Support Plan and DNR

During one of the long PICU stays, we were approached about how far we wanted to proceed in maintaining Hayden's life. We met with his team of specialists, and had individual discussions with each physician in great detail. Ultimately, we put in place a conservative Therapeutic Support Plan, including a Do Not Resuscitate (DNR) order for both the hospital setting and home.

How did we come to this decision? We had to consider the impact not only on Hayden, but also on our entire family. We had to examine his level of functioning and need for artificial life support, as well as consider the severity of his disease and the unknown nature of its progression.

We also accepted that after countless blood, urine, spinal and tissue samples being analyzed across multiple labs with no diagnosis, that we may never know the name of a

syndrome or have any causative answers. We realized that having a name for his syndrome doesn't equate a cure. The hard reality is that brain cells are not replaceable, which is needed to be able to slow down or stop progression. We still search, although not as passionately or as far-reaching, for an answer, not for Hayden, but for his siblings and their future children. It is possible that with an autopsy answers can be found.

My husband and I shared similar perspectives, which helped us to be honest with each other and ourselves. We decided that with all that Hayden has going on, it would stand to reason that no heroic measures should be started. However, we would sustain his condition at a reasonable level. In terms of quality of life, we would rather limit treatment than make the decision to remove treatments. Our plan includes the following:

- No CPR
- No inotropic or heart-stimulating agents
- No intubation
- No mechanical ventilation
- No dialysis
- Blood products can be used to treat an acute and/or easily correctable condition

Supportive Care

Since Hayden had a Therapeutic Support Plan in place as well as IV therapy for nutrition and medications, it was suggested that he would benefit from supportive care. The term supportive care encompasses both palliative and hospice support.

It was explained as additional support so that we don't have to see a physician or make an emergency room visit every time Hayden gets sick. The terms palliative and hospice were not really used with this particular company, preferring the term Supportive Care. The Supportive Care agency has a nurse who comes out every other week to draw labs and to check vitals. Soon, we started to get calls seeing if we were interested in having hospice volunteers. My husband and I were taken aback by that term--hospice--and started to wonder if the physicians failed to tell us our son was dying. I asked the nurse the next time she was out, and it was explained that Supportive Care encompasses palliative and hospice support, but that does not mean that Hayden has to have an expected death date.

We soon allowed these so called hospice volunteers into our lives. A group of teens from a high school about 20 miles away come out once or twice month for a couple of hours to play with the kids so that my husband and I can get stuff done around the house or even just get some quality time in with Hayden. The quads adore the "big kids."

Our Supportive Care also has available an Expressive Therapist who uses play therapy as her medium to help the children cope with the constant medical crises and to learn about death. The therapist has a session with the kids every month, and has introduced the concept of death through Marc Brown's book, *When Dinosaurs Die*. The kids can tell

you reasons why a living being might die and have heard of people dying due to a bad illness, but have yet to make the connection that their brother could die from a bad illness. They do know that he gets sick easily and that's why they have to be careful about hand-washing and avoiding a lot of facial contact with him when they have "sickie germs."



Some of the projects in the therapy include finding ways to honor their brother through craft making. Since the kids are really into imaginative play and mimicking adult and child roles through play with their baby dolls and other toys, the therapist builds onto this. It is not uncommon to see the baby dolls with a discarded syringe, extension tube, or even a saline eclipse taped to the doll's chest. In the next couple of play therapy sessions, the therapist plans on using doll play to explain that the doll is very sick and building upon this to explain that the doll can no longer get help from medicine, nurses, doctors and hospitals. Eventually, there will be a funeral for the doll.

The quads also get support through a therapy dog and his handler, the grandmotherly hospice volunteer. The dog makes monthly house calls. Sometimes just petting and loving this dog is all they need to calm down.

Daily Decisions

The summer Hayden came home with Supportive Care in place, we were never sure how much we should plan ahead. There were the simple things such as buying clothes for the next season. If I buy both the boys matching outfits, will only one be wearing it by the time it fits? There are also more expensive decisions, such as getting home modifications to our existing house, or building a new house with main floor accommodations for

Hayden. How far or how soon do we go into this? Will he live long enough to benefit from this?

There were other decisions about Hayden's involvement in the community. Do we keep him isolated so that he is less exposed to potential viral/bacterial threats? Do we send him to preschool? We decided if we have a limited amount of time with him that we'd make the most of it. I think it's been the best decision, and well worth the risks. I can't imagine sitting at home all the time and not allowing him to experience the world with the time he has left. We tried it in the past before surgeries, keeping him isolated from activities and events so as not to get sick beforehand. Since mom and dad are his full-time and round-the-clock nurses, it meant we often had to break up the family. One parent would stay home with Hayden while the other with the three active kids.

One day my husband Andrew and I were talking about childhood memories, and with a basic understanding of child development, we came to decide that if long-term memories are forming within the third year of life, than we will make a conscious decision to create memories for the quads that have Hayden as part of their life. We set forth to have a summer full of adventures. As Hayden healed from abdominal incisions from the GI surgeries and responded really well to intravenous nutrition, he took off. Even though Hayden does not have the motor skills, speech, or vision to experience the adventures in the same way that his siblings do, we actively helped him to engage in the activities by hand-over-hand assistance, or through sensory exploration. He responded really well with lots of smiles, contentment, and laughs.



As he responded, his siblings shared in the excitement. They began to help him engage in his surroundings and experiences. Some of the proudest moments were when one of the quads would shout out, "Mom he smiled at me! He laughs when we are laughing! He looked right at me! He turned his head when the cat walked by!" His siblings adore him and often come running when he is distressed. They know just the right tricks to

provide comfort. At four years old, the children have an amazing sense of compassion and love for their brother. We take tons of pictures. We never know when Hayden's brain will fail to send the signals to breathe, cough out secretions, start seizing uncontrollably or whatever. I take at least one picture of him everyday, since we never know if it will be the last.

He did indeed go to preschool and even rode the bus twice a week. He responded really well, and it has been an incredible experience. Hayden has really started to interact in his own way and made developmental "inchstones." We couldn't be more proud of his progress. He shows excitement around his siblings with lots of kicking and turning toward them as they play from afar and directly with him. This is a boy who lost his smiles and they have returned with great frequency and intention. His laughter is an indescribable experience too. The school has gone above and beyond in his special needs classroom. School specialists have taught us so much more about our son, and truly enjoy and have great affection for him. I am confident that with these experiences that Hayden gets the best quality of life we can offer.

A Unique Approach For Every Family and Situation

Today we face even more questions about his fragile life. It appears on a chest X-ray that he has another hiatal hernia. Based on Hayden's history and what we learned, he may not survive anesthesia again if he needs surgery. He may not be successful with extubation post-surgery. If we decide not to proceed with surgery, does this mean that his GI system will push further up into his diaphragm compromising his lung capacity and creating a slow suffocation? For now, we wait until we can get some answers before getting consumed with worry.

Like other families in this situation, there are often numerous compromises to be made when balancing between treatment and quality of life. The burden of chronic illness and the care it demands can exhaust a family physically, emotionally, and financially. Thankfully, at this point we do not have the latter worry, but that can certainly change if we need to pay for nurses to care for Hayden as his illness progresses. To be perfectly honest, the demands of life with three very rambunctious healthy children and one with a chronic illness send my husband and me into an exhausted and often times interrupted sleep. If you ask either one of us, though, we both will tell you how proud and happy we are with this life we've created with our perfect family.

My perspective on how we've approached and are handling the gravity of having a child with a life-threatening and progressive illness may not be agreeable to others and that's OK. We've drawn upon the wonderful experiences to find the strength to make the hard decisions proactively, such as developing a DNR. My husband and I have to get real in our thinking patterns to avoid superstitious or "wishful thinking" that can often times lead to false hopes. Moreover, rather than putting our heads in the sand, we accepted the gravity of the situation.

Every situation is unique, but what I hope to pass on is how we developed a pragmatic approach based on the love we have for him and his siblings. In this family, we have actively engaged in memory creation and family togetherness through experiences and activities that allow us to take advantage of the time we have left.

Jamie and her husband Andrew have been married for 10 years and were blessed with quadruplets 4 1/2 years ago. Before the quads were born, she had a brief stint in social work before becoming a special education teacher, while earning her Masters Degree in Education. That background has been a huge asset in finding resources and understanding Hayden's needs. Hayden is truly the hero in his family's eyes.

Jamie regularly updates Hayden's Caringbridge page with the good, the bad, and sometimes the downright funny antics of life with four 4-year-olds. Follow along at www.caringbridge.org/visit/haydenbarnes