



Complex Child E-Magazine

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Juvenile Polymyositis: Moving Past the Diagnosis

by Marlowe Futrell

Our Matthew is a happy, laid back, exuberant, smart little six-year-old. He loves to be outside--swinging, going down the slide, chasing bubbles, throwing balls. One of his favorite things is the tunnels at McDonald's, Chuck E. Cheese, and the zoo. He loves sweets, cookies, cakes, ice cream, and popsicles. He plays his Gameboy and races Hot Wheels. He loves to go places--zoos, museums, just about any store, but especially amusement parks. He adores his two big brothers.

In other words, he's a normal kid.

Not that he doesn't have his challenges. He was diagnosed just before he turned three with Pervasive Developmental Disorder, Not Otherwise Specified (PDD-NOS), a disorder on the autism spectrum. He also has moderate to severe speech issues, making him hard to understand. He has been making good progress, thanks to many teachers, and speech, occupational and physical therapies. He has worked very hard.

New Symptoms

This past February, Matthew was sick for four days with a low-grade fever. Usually, when he has a slight fever, you would never know it--he's almost as active and usually bouncing around as normal. This time was different. He was wiped out. The pediatrician said it was probably a virus. Looking back, we realized that he hasn't been the same kid since.

Soon thereafter, Matthew started to walk more slowly. We barely noticed it, and just dragged him along. He started to complain about leg pain, first in the right and then in both legs. Later that month, the pediatric orthopedic surgeon, after reviewing X-rays, concluded that Matthew was in the early stages of Legg-Calvé-Perthes Syndrome, a degenerative disorder causing loss of bone mass in the hip. He dismissed us and said to come back in a few months.

By early March, his aquatic therapist noticed weakness in shoulders. It was hard for him to put hands "up to the sky." His physical therapist also found that he was not able to do things that used to be easy for him. It became hard for him to climb up a curb. He didn't want to get off the couch to get anything out of the fridge.

By mid-March, Matthew needed help ascending the five stairs in front of the house. He could hardly throw a ball and started to be scared to descend stairs outside. Both legs still hurt. One of most heart-wrenching things was that he only wanted to play outside for 10 minutes. It used to be hard to get him inside after an hour.

The Search for Answers

Meanwhile, doctor appointments were becoming a big part of Matthew's life. On a Friday, our pediatrician became very concerned and got us appointments the following Monday with the pediatric orthopedic surgeon and Tuesday with a pediatric neurologist. At this second appointment, the orthopedic surgeon still believed that Matthew may possibly be in the very early stages of Legg-Calvé-Perthes Syndrome, which addressed the leg pain but didn't explain the weakness that he had in his shoulders. He suggested testing.

The pediatric neurologist concluded that whatever Matthew had was probably muscular in nature. After the second appointment with her, we were referred to a second neurologist who had a greater expertise in muscular diagnoses, and was the co-head of the department. At the second appointment with her, an EMG was performed and it was confirmed that Matthew's issues were muscular.

The next test was to rule out Duchenne muscular dystrophy, which required an eight week wait in order to receive test results. The neurologist said that she did not want to subject Matthew to more tests (such as a muscle biopsy or an MRI) without ruling out muscular dystrophy. When told about the test, his physical therapist (very experienced), flat out said that Matthew did not have muscular dystrophy.

Meanwhile, over this eight-week period, Matthew could not get up off of the floor, could not go up or down stairs, found it hard to get on and off of the potty, needed help getting in and out of the car, couldn't get out of bed, and had difficulty picking up small objects off the floor. At his worst point, he couldn't get up off of a chair. Mostly he just laid around. We also noticed that he was becoming depressed and frustrated that he couldn't do the things that he used to do so easily. By this time, we had applied for a handicapped parking placard.

In three months, Matthew had gone from being a normally functioning kid to someone who couldn't get out of a chair. This was very much a low point as we knew that if Matthew's regression continued, in several more months, there would be no more Matthew.

A Diagnosis

The neurologist, realizing that muscular dystrophy does not present this quickly, referred us to a rheumatologist. Several days later, he was able to diagnose Matthew on the spot, although he wanted to do some additional testing to confirm the diagnosis. Soon thereafter, Matthew had an EKG and an MRI. The EKG was normal, which showed that the muscle weakness did not affect the heart and likely did not affect the lungs. The MRI showed many places of muscle weakness, as expected.

The next day, a muscle biopsy was done, which, with the other tests, confirmed the diagnosis of Juvenile Polymyositis (JPM). We were relieved to finally get a diagnosis and to find that there was hope.

Juvenile Polymyositis is one of two primary types of Juvenile Myositis, a rare autoimmune disorder that causes inflammation in the blood vessels under the skin or in the muscles, called vasculitis. The other and more common form of Myositis is Juvenile Dermatomyositis, although kids with this form develop a wicked rash, which Matthew does not have. It is estimated that 3000-5000 children in the United States have Juvenile Myositis.¹ Of those, only five to ten percent have JPM. The primary symptoms are muscle weakness, fatigue, and sometimes fever. “Poly” means that many muscles are involved. It is believed that Matthew’s JPM may have been triggered by the virus he had in February.

In the 1960’s, approximately one-third of children with Juvenile Myositis died and another third had serious long-term disabilities.² In the 1970’s, when steroids began to be used as treatment, less than ten percent died. Today, with the use of steroids and other medications, the death rate has dropped to one percent. Still too many.

In June we were back in the hospital to see how Matthew would react to the steroid treatments. He had an IV steroid treatment both Monday and Tuesday. Several days thereafter, we began to see an improvement in muscle strength.

An Unknown Prognosis

Four months later, Matthew continues to receive the steroid treatments, now every four weeks (down from every two weeks), and also takes oral medications, and will for the next year or two. The medications have, to this point, done their job, as we have seen great improvements. He is walking faster, can get out of the car, go down a flight of stairs (holding on tight to the rail), get off the potty, go to the fridge, get out of bed, and is spending time playing outside. His charming personality is back.

We don’t know what will happen in the future, but we have a lot of hope. There are essentially three outcomes of JPM: a monocyclic course, a polycyclic course and a chronic continuous course.³ With a monocyclic course, Matthew’s Polymyositis would be a one time occurrence and go into remission permanently. With a polycyclic course,

the illness goes into remission, but reoccurs at least once and often multiple times. A chronic course is when there is continuous active disease for more than two years after the initial diagnosis. Often it remains active to some extent for four to ten years, and for some, into adulthood.

We, of course, hope and pray that this is a one-time occurrence. We are buoyed by the quick reaction that Matthew has had to the steroids (we *love* steroids!), and other medications. But if that's not the case, we'll be able to deal with it. And Matthew will be fine.

Marlowe Futrell and her husband, Jim, live outside Pittsburgh with their three boys, 13-year-old Jimmy, nine-year-old Christopher, and Matthew who is six, as well as with Bo the basset hound. She also is the owner of MMc Marketing Research & Consulting, specializing in primary and secondary marketing research.

¹ Brian M. Feldman, *et al.* "Juvenile dermatomyositis and other idiopathic inflammatory myopathies of childhood." *The Lancet* 2008;371(9631):2201-2212.

² Feldman.

³ Lisa G. Rider, *et al.*, eds. *Myositis and You, A Guide to Juvenile Dermatomyositis for Patients, Families, and Healthcare Providers.* (Myositis Association, 2007): 60.