



Megan Gray

LYMPHANGIOMATOSIS

By Patti Perkins Gray, Megan's Mother
"After all we have been through, I look at Megan today and feel extremely blessed to still have her here with us."

Our journey with Lymphangiomas did not begin with any difficulty breathing. There was no chronic cough or cold that would not go away. In fact, Megan hardly ever had a cold. In the beginning it didn't look like a lung problem at all.

Megan was 15 months old in October 2009 when her collar bone broke for no apparent reason. Much testing followed. X-rays, MRI, and CT scans revealed cystic lesions in almost all of Megan's bones, her thyroid, abdomen, and in the soft tissue behind both of her collar bones reaching down into her chest. Finally, a biopsy of her thyroid provided the diagnosis of diffuse lymphangiomas.

Things were fairly uneventful until January 2012 when Megan got very ill with group A streptococcus (strep) bacterial infection in the blood. What began as a fever one evening quickly spiraled out of control. Within 24 hours our 3-year-old little girl was admitted to the pediatric ICU with sepsis and toxic shock syndrome, and soon after, all of her organs began to fail. It was a very long traumatic experience, and it is a miracle Megan survived at all.

For those first seven months in 2012, we were in the hospital more than at home. It was during this time that the pleural effusions began. Megan's disease had now progressed to her lungs. She required repeated chest tube placement to drain the fluid from around both lungs. With no FDA approved treatments for her disease, she was started on a combination of drugs (sirolimus, vincristine, and prednisone) that have been used off-label for lymphangiomas, alone, and in various combinations. After only a couple of weeks on this regimen the effusions stopped. Megan remained on this cocktail for more than a year until the side effects of the drugs got to be too much and we decided to take a break.

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Lymphatic vessels reach every part of the body except the central nervous system (the brain and spinal cord). The lymphatic system has three main functions:

- Maintain fluid balance
- Defend the body against disease by producing lymphocytes
- Absorb fats and fat soluble vitamins (A, D, E, and K) from the small intestine and transport them to the blood, bypassing the liver

This condition is marked by the presence of cysts that result from an increase both in the size and number of thin-walled lymphatic channels that are abnormally interconnected and dilated. The deranged lymphatics tend to invade surrounding tissues and cause problems due to invasion and/or compression of adjacent structures.

*Source: Lymphangiomas and Gorham's Disease Alliance. "What Is Lymphangiomas?"
lgdalliance.org.*

In June 2014, Megan's pleural effusions came back with a vengeance. Three times in the span of five weeks, she had to be hospitalized and have chest tubes placed to drain the fluid from around her lungs. The sirolimus, vincristine, and prednisone regimen was restarted and seemed to be working, but when the effusion returned a third time, her doctors and I decided it was time to go ahead with pleurodesis, a surgical procedure to scar the lung to Megan's chest wall in hopes to stop effusions from returning. Five days after the procedure Megan was home and the surgery seems to have been a success. She has some shortness of breath but nothing compared to how it was with the effusions.

Lymphangiomatosis is so rare that most doctors have never heard of it and have no idea how to deal with the complications that it can cause. With that brings many fears and frustrations. It has been a blessing that I found the Lymphangiomatosis & Gorham's Disease Alliance (LGDA) and got in touch with others dealing with this disease. Megan has even met a couple of patients, which has helped her to know she isn't the only one.

After all we have been through, I look at Megan today and feel extremely blessed to still have her here with us. She is an inspiration to us all. I pray that one day this disease will be cured or at least better managed.