Quote of the Day:

"I'm not sure how we would have endured it all without the help, advice and friendship of those involved with Barth Syndrome Foundation. They have been there to support us through good times, bad times and even the very darkest days. Barth Syndrome Foundation is our lifeline. …" ~ *Brie, Mother of BTHS Diagnosed Son, Kentucky*

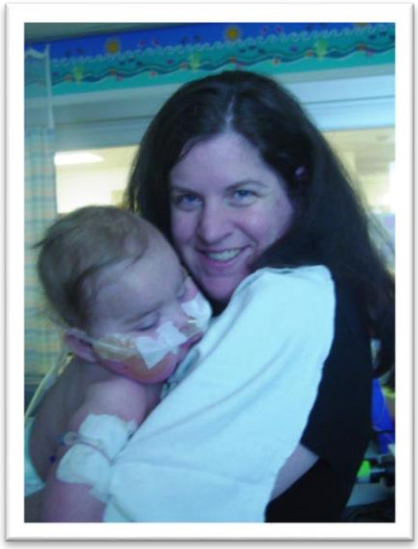
Featured Story: Our Lion Prince

Milosh

Very few families are able to say an organ donor saved their child's life and yet my family is in such a position. Before I talk about my son's heart transplant I will start at the beginning of our journey. Milosh was born with a rare, genetic disorder called Barth syndrome. Barth syndrome is a metabolic disorder and can cause cardiomyopathy (an enlarged heart), neutropenia (a reduction in a type of WBCs which results in immunity problems), muscle weakness and delayed growth.

Milosh was born in heart failure; because of this all his other organs went into failure within 24 hours of his birth. He spent the first 4 months of his life at a children's hospital in cardiac ICU. During those long months the incredibly gifted doctors, nurses and medical staff (along with God's grace) helped Milosh and his organs to recover. His heart, however, was still bad enough to require him to be listed at 1A status (the highest medical need) on the heart transplant list. We waited over two months for a new heart. Milosh even came home to wait. Then one day at a cardiology appointment an echo revealed Milosh's heart function had improved so much that the doctors wanted to wean him from some of his heart medicines. If this wean was successful they would remove him from an active status on the transplant list. We were overjoyed and at the same time very worried. With Barth syndrome, the heart condition is undulating meaning it can get better and worse and there is really no way to predict how things are going to go.

Milosh was weaned from those medicines and went inactive on the transplant list. We enjoyed 6 months of good check-ups. We counted our blessings every day and thanked the Lord for his continued presence in our lives. Then one day early in December an echo revealed Milosh's heart was not functioning so well anymore. We would have three more months of poor check-ups before Milosh's



Milosh & Brie

condition became bad enough to be re-admitted to the hospital. Although we were devastated he had to go back to the hospital, we were grateful he stayed well long enough to celebrate Thanksgiving, Christmas and his first birthday at home with his family.

Once admitted back into C.I.C.U. (Cardiac Intensive Care Unit) Milosh deteriorated very quickly. The first major setback came when his lungs were no longer able to work on their own so he had to be intubated (have a breathing tube inserted and be assisted in breathing with a ventilator). Milosh was reactivated on the transplant list and again we waited. His heart continued to worsen. His doctors talked to us about a Berlin heart (a mechanical heart). While this device has been used throughout Europe for over 20 years it is pending FDA approval in the United States. Milosh's doctors applied for a compassionate use waiver in order to receive permission to use a Berlin on him. The surgery was going fine until he was taken off the bypass machine; Milosh's lungs filled with blood. The doctor came out and told us they would have to remove the Berlin heart and put him on ECMO (a heart and lung bypass machine). We were crushed – while both devices come with many complications ECMO was the last thing we wanted because most babies will survive on this device only 2-3 weeks. When you are waiting on an organ you have no idea when that organ might arrive.

Our family gathered together and cried, prayed and then cried some more. The next time the surgeon came out he told us before they removed the Berlin Heart they called their representative. He told them not to remove the Berlin heart. Instead they should add the "mechanical lung" part of the ECMO into the "circuit." The quick thinking surgical team figured out a way to do it – and it worked! We have been told this setup has been done one other time on an adult in Germany. Milosh is the first baby in the world to have this device we are now affectionately referring to as BECMO. We knew there were still rocky times ahead of us, but we had bought Milosh more time, time to wait for his new angel heart.

The wait was agonizing. Milosh's condition was about as critical as you can get. During this time I received several phone calls letting us know we were needed at the hospital immediately as things were not looking good. I will never forget one middle-of-the-night call in particular. When we arrived in Milosh's room it was filled with every doctor and nurse in the unit. All we had been told was that Milosh was bleeding. When I rushed to his side I saw blood pouring out of all the wound sites of the tubing for the Berlin. The tricky thing about a mechanical heart is that in order to avoid blood clots a patient is given anti-coagulation medicine. It is tough to find the perfect balance, especially in Milosh's case. That morning Milosh's blood volume was replaced

(L-R) Ned, Bella & Milosh

7.5 times. My husband and I held each other and told the doctors not to stop everything they were doing. After several hours Milosh quit bleeding and became a little more stable.

Every night I would go to bed wondering if Milosh would be alive in the morning and also if this would be the night we would receive the call. I wondered that for 29 nights. Until the afternoon of April 4th – the transplant coordinator called us and said they had a heart for our precious son. I immediately thanked God – my son would now be given a second chance, he would live. We were filled with joy as well as the knowledge that somewhere a family (one we had been praying for since Milosh was listed for a transplant) was facing the worst possible thing anyone could ever imagine – the loss of their dear, precious child. We were amazed at this family's strength for being able to see past their grief in order to help other sick children, in essence saying "Yes" to life which would keep other families from having to go through the same heartache they were experiencing. We owe our child's life to them. Their incredible bravery not only saved Milosh, it also saved our family. His recovery has been slow but steady. We continue to pray for his donor family who will always hold a very special place in our hearts. Words cannot express the gratitude we feel for them. Ten days ago, Milosh returned home to his adoring big sister, his loyal dog, his playful cat and his grateful parents. This is all thanks to organ donation. Our family is one of the lucky ones. Milosh's doctor told us only 1/3rd of the children waiting for a new heart will live long enough to receive that gift of life.

That is the first chapter of our sweet Milosh's life. It has been a crazy year that I often compare to a ride on a roller coaster – keeping us on the edge of our seats, terrifying us and filling us with exhilaration all within a matter of moments. It has been a year of witnessing things no parent should ever have to see, learning complicated medical information and still taking time to savor every positive test result, reached milestone, and heart-melting smile. I'm not sure how we would have endured it all without the help, advice and friendship of those involved with Barth Syndrome Foundation. They have been there to support us through good times, bad times and even the very darkest days. Barth Syndrome Foundation is our lifeline. The information exchange via website, e-mail listserv and archives, medical communication between doctors and parents, are all vital for the treatment and care of children affected by Barth syndrome. Our amazing Foundation does all of this and so much more for each individual. The personal support and encouragement we receive daily from our Barth family is truly relentless and unwavering. That is the Barth Syndrome Foundation. That is all of us affected by Barth syndrome.

(L-R) Bella, Ned, Milosh & Brie