



Bright Light Foundation

Nicole Kienzle

2013 BLF Recipient



Nicole is a sweet 11-year old girl, who has a great sense of humor and even through she has a small stature, she is a big, fun personality. She is an active member of Girl Scouts, enjoys camping and fishing, and loves animals, especially small dogs. She has two older brothers and likes to “mother” them (or boss them around as they say). She is a tough cookie considering all the things she’s been through in her life so far.

Nicole was born with DiGeorge Syndrome, which is a deletion in the 22q chromosome. This syndrome caused a heart condition called Truncus Arteriosus Type I. When she was born, she had a trunk entering her heart that served as the Aortic artery and the pulmonary artery. When she was three months old she had her first heart surgery to put in place a pulmonary artery and valve, called a homograft. The surgeon also placed a gortex patch to close a hole between the two lower ventricles in her heart. At six months she had another surgery to remove the pulmonary homograft and replace her damaged aortic arterial valve with a mechanical titanium valve.

When Nicole was nine months old she still weighted less than 10 pounds, which made our pediatrician investigate why she was gaining weight so slowly. The doctors that we consulted at Presbyterian St. Lukes Hospital in Denver Colorado realized that she has pulmonary hypertension, which was easily fixed with a Nissen, G-Tube surgery. In this surgery the muscle at the top of the stomach is tightened to prevent reflux and a G-Tube was inserted through the wall of the abdomen into the stomach. The G-Tuube was then used to feed Nicole with a time released machine that pumped formula into her stomach.

Because of the is surgery Nicole was finally able to gain weight and become more active. It was only after the that she began doing the things babies should do, like rolling over, giggling and growing. Nicole



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flourished after these surgeries. Just before her second birthday she finally began eating regular food, which was good because a month later she pulled out her G-Tube!

In 2009 at eight years old, Nicole has her third heart surgery due to the Homograph wearing out. During this surgery they replaced the Homography pulmonary artery/valve with a Contegra (bovine) replacement. Before this surgery she was only able to walk around 100 yards on our family walks, but days after the surgery she was walking and running almost as far as her two older brothers.

She will continue to undergo heart surgeries throughout her life to replace the artificial pieces in her heart as they wear out or she outgrows them

When Nicole was six years old, we began to notice a slight curve in her spine. She wore a large brace for a year or so to keep it from getting worse, but the curve continued to grow. In 2009, we consulted Dr. Mark Erickson at Denver Children's Hospital, where she was diagnosed with scoliosis. In October 2011, Nicole went through a back surgery to place two VEPTR rods on either side of her spine. The rods are special because with small outpatient surgeries every six to nine months, her surgeon can extend the rods so her back will grow as she does. Her last expansion was February 25, 2013. Her next expansion is scheduled for August 2013.

After Nicole's first back surgery, her parents noticed her breathing was becoming loud and wheezy. After a few months of working with a Pulmonologist, in January 2013, they performed a bronchoscope – a small camera was inserted through a tube into the lungs so they could be observed. It was discovered that due to the scoliosis and her aortic artery, which arches to the right instead of the left as in a normal person, the main bronchial tube entering her right lung was being constricted. Basically the spine was pushing against her Aorta, which was compressing her bronchial tube. Currently Nicole is operating on one lung, which makes it difficult to recover from colds and flu. And we are careful, but also encouraging, about physical exercise.