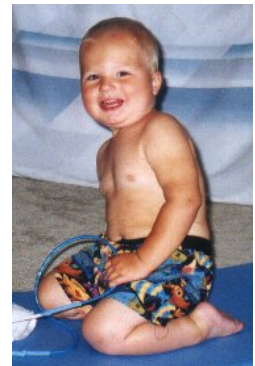


# Justice

THIS IS A PERSONAL STORY SHARED BY A FAMILY WHOSE CHILD HAS  
ISOVALERIC ACIDEMIA (IVA)

When my nephew, Justice, was born he was such a precious little thing. Did I say little? Justice was anything but little. He had very long legs for a newborn, and the rest of him was all feet and hands. Hands that looked like they could swat planes from the sky. It was obvious that he was going to be a big boy like his dad and everything seemed fine. Justice had his mandatory newborn screening and all checked out fine. Unfortunately Washington only screens for four disorders.



At about a week old Justice wouldn't eat and appeared to be losing weight. The doctors said that was normal, it was just his mom being a "panicky new parent". They even gave his mom classes on how to properly breast-feed her new baby. What a joke!

Within the next few days Justice had no appetite at all and rarely woke up on his own, so he was taken to the emergency room. They finally ran some blood tests. The results were so erratic that Justice was immediately sent to Children's Hospital in Seattle. My family was an emotional wreck and it seemed as if the world was crashing down on us. We didn't know how precious every minute was.

Once in Seattle, the doctors were still at a loss, until one of his nurses noticed a "sweaty socks" smell. She was sure it was a metabolic disorder, so Dr Scott, from the University of Washington Medical Center, was called in. Dr Scott quickly diagnosed Justice as having Isovaleric Acidemia (IVA). A rare inherited metabolic disorder that makes it impossible for Justice to properly breakdown proteins, which causes a build-up of acid in his system.

We were told Justice would need special care, diets and doctors for life, and if we didn't follow his diet precisely, he could become retarded or even die. They said he would always be small for his age, and he would be hospitalized for even the most common childhood ailments. Because of this first acute attack, Justice could be developmentally delayed, but it was too soon to tell. It seemed like a hopeless situation.

We shed a lot of tears and said a lot of prayers. Our prayers were answered; Justice responded well to treatment and had no lasting effects from the acute attack of IVA.

Justice's acid levels are being well managed now and he hasn't had any further acute attacks. He constantly amazes his doctors, not to mention his family. At his last check up he was actually big for his age! Justice is developing on schedule and his acid levels are so low the doctors can hardly tell that he has IVA.

If Justice would have had the expanded screening at birth, the first acute attack could have potentially been avoided and he wouldn't have been put at risk for retardation and sudden death. Knowing that other babies are

put at risk for these types of acute attacks, and the devastating results of those attacks, breaks my heart. Especially when it's as simple as comprehensively screening EVERY baby. No family should have to endure that kind of heartache when it can be prevented.

Sincerely,

Terilyn DePaolo

Written April 2000 by Terilyn DePaolo

Aunt of Justice, born 1998